Pediatric Transesophageal Echocardiography

Slokdarm echocardiografie in de kinderleeftijd

PROEFSCHRIFT

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Chapter 1

- General Introduction -

Over the recent decade cardiac ultrasound has largely altered the practice of pediatric cardiology. Cross-sectional imaging provides a detailed and non-invasive assessment of intracardiac morphology and thereby allows the accurate diagnosis of even complex congenital heart disease in the vast majority of cases (1-5). With the more recent adjunct of spectral and color Doppler techniques the majority of the resultant haemodynamic changes can be defined (6-10). Thus, not surprisingly, the decision for surgical correction in children with congenital heart disease is more frequently based on precordial ultrasound studies alone (11-15). In addition, such studies largely influence the clinical decision making in the follow-up of both operated and unoperated patients.

However, difficulties and ambiguities are not uncommon in the clinical practice of precordial echocardiography. One of the general limitations is that one cannot exclude what one does not visualize. For example, the site of connection of all four pulmonary veins in children beyond infancy routinely cannot be defined by precordial or subcostal ultrasound investigations. Thus, in cases where there persists the suspicion of abnormal venous connections, cardiac catheterization and angiocardiography appear to be indicated, in order to effect the ultimate diagnosis. Other structures, such as the atrial appendages can be visualized from the precordium only in the neonate and young infant. Therefore the precordial diagnosis of atrial situs, which is the first diagnostic step in the diagnosis of congenital heart disease, remains indirect and relies on the definition of the position and relation of the abdominal great vessels (16). Further limitations in the precordial echocardiographic evaluation of cardiac morphology include the poor ultrasound windows which most often are encountered in patients with thoracic cage or spinal abnormalities, or in those with cardiac malposition. Sometimes, in complex congenital heart disease, the presence of several of these limiting factors preclude the definitive diagnosis, which is only obtained by subsequent surgical inspection.

In the follow-up of children with congenital heart disease, precordial ultrasound studies are subject to a variety of problems and limitations. Firstly, in those children who have undergone surgical correction via a midline sternotomy, the presence of fibrous tissue adhesions results in a marked reduction of image quality to be obtained from the precordium. Secondly, the presence of prosthetic material used in the surgical repair produces ultrasound shadowing of the areas posterior to it. In addition, in all older children, imaging from the subcostal areas becomes more difficult and sometimes impossible. Thus, precordial ultrasound studies frequently fail to provide all the information required for decision making and optimal patient management. In particular in those (symptomatic) patients who have undergone either a Fontan or Mustard procedure for correction of their congenital cardiac malformation, (repeat) cardiac catheterization often is required to effect a complete assessment of the hemodynamic results. Magnetic resonance imaging has been reported to

be a versatile tool both in the primary diagnosis and in the follow-up of congenital heart disease (17,18). However, the high costs and the low availability militate against its routine use. In addition, the technique does not provide real-time imaging and information on intracardiac blood flow abnormalities can, to date, be obtained in only few centers (19,20).

Transesophageal echocardiography, after its clinical introduction in 1982 by Hanrath and colleagues (21), has rapidly been established in the practice of adult cardiology (22-24). Over the recent years it became the investigative technique of choice for a wide spectrum of acquired cardiac disease, such as endocarditis (25), aortic pathology (26-28) or prosthetic valve dysfunction (29-31). In addition, its value in the perioperative period, and in particular as a monitoring technique during cardiac surgery or for anesthesia is widely acknowledged (32-34). However, to date, little is known about the potential value of this new imaging technique in the diagnosis and follow-up of patients with congenital heart disease (35-37). This is in part related to the fact that the proportion of adolescents and adult patients with congenital heart disease is, although steadily increasing, small compared to the number of patients with acquired cardiac disease. Although those studies might be of additional value in the younger patient population, the standard (adult) transducer designs precluded such studies in children below twenty kilograms of bodyweight (38,39). Besides, patient tolerance is poor in this age group, and thus requires studies to be carried out under heavy sedation or general anesthesia.

Only recently small dedicated pediatric transesophageal probes have become available for use in clinical practice (40, chapter 2). In theory, the esophageal ultrasound approach would allow for an unrestricted and more detailed assessment of the cardiac structures and chambers closest to the esophagus, that are the venous return, the atrial chambers and the atrioventricular junction. Thus, the technique might be expected to provide additional relevant information in selected pediatric cardiac patients.

Purpose of the study

In face of several limitations of precordial ultrasound investigations in children with congenital heart disease, it was attempted to define the additional value of pediatric transesophageal echocardiography. Its comparative value, firstly, in the preoperative diagnosis of congenital heart disease had to be determined (Chapters 4-8), secondly, its place as an intraoperative monitoring technique in the surgery of congenital heart disease had to be assessed (Chapter 9), and thirdly, its role in the follow-up of congenital heart disease, when compared to precordial ultrasound techniques and cardiac catheterization, had to be defined (Chapters 10,11).

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Chapter 2

Technology and technique of paediatric transoesophageal echocardiography

Transoesophageal studies in the assessment of congenital heart disease should be carried out according to a standardized scheme, which observes the rules of sequential chamber analysis most often used in the categorization of congenital malformations of the heart (1,2). Since this scheme was followed in all of the study protocols described later, it will be outlined here in more detail. The equipment used for these studies will be described first.

Technology Paediatric transoesophageal probes consist of an ultrasound transducer mounted at the tip of the shaft of a paediatric gastroscope or bronchoscope. The shaft itself, which has a total length of some 70 to 80 cm and a maximal circumference of some 7 mm, contains the steering mechanism and the connecting cables of the ultrasound transducer. Steering facilities of the transducer tip is, for the sake of miniaturization, restricted to anterior-posterior movements only. Anterior tip angulation is achieved by counter clockwise turning of the steering wheel; posterior angulation by clockwise turning. The neutral position of the tip of the transducer is indicated by markings engraved on the steering wheel. The position of the transducer tip can be fixed by various mechanisms, however, as a general rule, the unlocked position should be used, and contact between the transducer and the oesophagus should be achieved by gentle tip angulation. In particular, the probe should be unlocked while being manipulated within the oesophagus over great distances.

The transducer itself is in most cases a phased array system, that allows for 5 MHz cross-sectional imaging, colour flow mapping, and pulsed wave Doppler sampling. The first paediatric probe developed (Aloka Company, Japan) used a 24 element transducer, in order to minimize the external dimensions of both the shaft and the tip of the probe (6.8 mm shaft diameter and 7 x 8 mm tip dimension). The reduction of the total number of ultrasound crystals resulted in a considerable decrease in image quality. In addition, ultrasound penetration, both cross-sectional image and Doppler information, was low and the near field artefact, which extended to some 1.5 cm of distance from the oesophagus, limited the investigation of small children. The second generation of paediatric probes, which are now being tested for production was developed by the Department of Experimental Echocardiography, Thoraxcenter Rotterdam. This probe for the first time provided 48 element cross-sectional imaging, which resulted in a much improved image resolution. The near field artefact was much reduced (some 3-5 mm) and the ultrasound penetration was much increased, allowing for diagnostic studies in children with bodyweights from some 4 to 50 kilograms. Although the width of the transducer had to be increased to 10 mm, in order to incorporate twice the number of ultrasound crystals, the height could be reduced to 5 mm. Thus the tip dimensions of both probes are comparable measuring some 30 mm. The shaft diameters of both probes are identical (7 mm). Recently, dedicated transoesophageal probes

using a phased array system and allowing for additional continuous wave Doppler facilities have been introduced. However, our experience with these probes is, to date, limited.

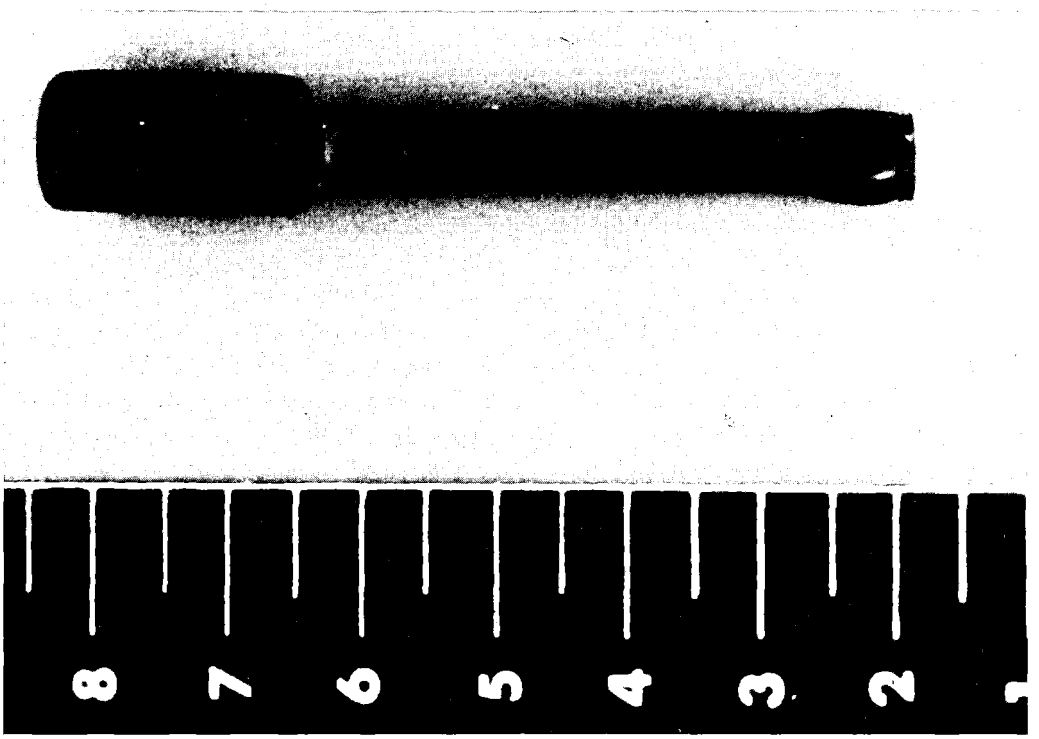

Figure 1: The tip of the paediatric transoesophageal probe developed by the Department of Experimental Echocardiography, Thoraxcenter Rotterdam. Maximal shaft diameter is 7 mm; maximal tip dimensions are 5 x 10 mm.

All transducers produce single-plane images at right angles to the shaft of the probe, that is images in the transverse axis of the heart. Thus, a series of tomographic scan planes of the heart is obtained from within the oesophagus and the stomach. Rotation of the probe within the oesophagus allows for an assessment of cardiac structures beyond the standard 90 degree angle of cross-sections.

Potential future developments in the field of paediatric transoesophageal probes include higher frequency imaging (e.g. 7.5 MHz), higher resolution imaging (e.g. 64 element transducers) and biplane or multiplane imaging. In particular the later appears to be of particular value in the assessment of complex congenital heart disease.

Technique Transoesophageal studies in children below the age of 8 - 10 years are performed routinely under general anaesthesia given for simultaneous cardiac catheterization or for cardiac surgery. However, with increasing experience with the technique a growing number of children are being studied electively under general anaesthesia (e.g. Mustard patients). In older children heavy sedation may be used. Although our experience is limited, initial experiences, using Flunitrazepam (0,02 mg/kg) or Etomidate (0,2 mg/kg), have been

promising. Routine anaesthetic monitoring is performed during the entire study (electrocardiogram, blood pressure, and pulse oximetry). Currently, we do not administer any additional antibiotic prophylaxis for transoesophageal studies. Sterilization of the probe is carried out by thorough washing of the probe and by subsequent emersion in a Cidex solution for 20 minutes. Alcohol should not be used for disinfection since it may degrade the silicon material used to cover the ultrasound transducer. All transoesophageal probes should be routinely checked for electrical safety, which is endangered when even microscopic cracks of the coating material occur.

Orally intubated and ventilated children can be studied in a supine position. Nasogastric tubes which are frequently used during anaesthesia for cardiac operations, should be removed prior to the study. Before introduction of the probe the steering mechanism is checked carefully, as are all electrical connections. The operator stands at the top end of the child with the cable and handle of the probe placed around his neck. The lower third of the probe is held with the right hand, while the mandible of the child is elevated and the neck is hyperextended with the fingers of the left hand. Then the probe is inserted into the mouth and is advanced above the endotracheal tube along the palate. The probe should be advanced gently along the midline, in order to prevent positioning in the (lateral) piriform recessi. The probe is then further advanced to the lower third of the oesophagus. In most cases the probe can be introduced without any difficulties. In case some resistance is felt during insertion, the probe should be removed and a second attempt should be undertaken. If unsuccessful, insertion under direct laryngoscopic vision is advised. In addition, this latter method should be applied in all children with nasotracheal intubation.

Children who are studied under heavy sedation without intubation should be studied in a left lateral decubitus position and should be fasted for at least three hours. A venous canula should be placed on the patients right arm. Sufficient local anaesthesia of the hypopharynx should be given 3 to 5 minutes before introduction of the probe. Introduction of the probe with the patient in a lateral position is facilitated by flexion of the neck, so as the chin approximates the sternum. The operator sits on the patients left side. A bite guard, to protect the patients teeth and the probe should be used routinely. Thereafter the probe is inserted blindly with the tip of the probe being partially flexed.

Standard imaging planes The standard cross-sectional imaging planes that can be obtained by transoesophageal imaging are described below (see Figure 2). The image orientation of transverse-axis transoesophageal imaging is standardized. The posterior aspect of the heart is displayed on the top of the screen, the left on the right as seen by the operator.

Transgastric planes With the probe positioned in the stomach short axis cuts of the ventricles are imaged. The inferior wall of the ventricles, being closest to the transducer, are displayed on the top of the monitor as is the postero medial papillary muscle of the mitral valve. By withdrawing the probe from this position the right ventricular outflow tract can sometimes be imaged, and is then depicted on the bottom of the monitor. Clockwise rotation of the probe together with further minimal withdrawal will demonstrate the liver and the inferior caval vein. Gradual withdrawal then demonstrates the hepatic veins and their connection with the inferior caval vein.

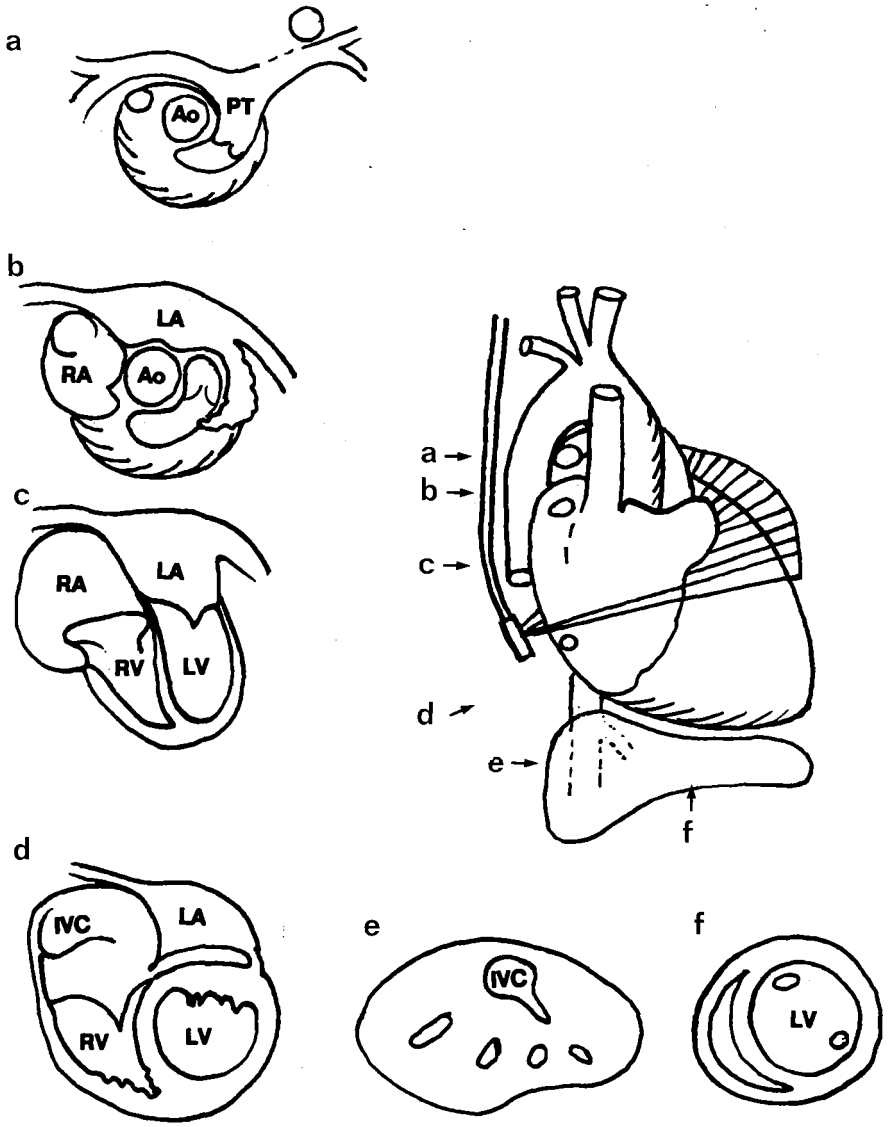

Figure 2: Diagram showing the standard transoesophageal imaging planes

Lower oesophagus Scan positions obtained from the lower oesophagus demonstrate the connection of the inferior caval vein with the right atrium. Anteriorly the floor of the right atrium is visualized together with the inferior leaflet of the tricuspid valve. In addition the inlet component of the muscular ventricular septum is visualized on this view (figure 3). Posteriorly and to the right the right lower pulmonary vein is visualized. Counterclockwise rotation from this scan position brings the coronary sinus a vue, together with an oblique section of the left ventricle and a section of the posterior leaflet of the mitral valve. The

floor of the left atrium is seen posteriorly.

Left atrial views With the probe positioned on the back of the left atrium the classical four chamber view is obtained (figure 4), demonstrating the atrial septum at the level of the oval fossa, both atrial cavities, the atrioventricular valves and the ventricular chambers. The same plane frequently demonstrates the junction of the right atrial appendage with the venous component of the right atrium. The left lower pulmonary vein is demonstrated by counterclockwise rotation of the probe from this position together with a short axis of the descending aorta near the diaphragm.

Mid oesophagus Scan planes obtained from a transducer position in the mid oesophagus produce basal short axis cuts of the heart. The first section to be obtained demonstrates the left ventricular outflow tract, including the area of fibrous continuity between the anterior mitral valve and the atrioventricular septum (figure 5). Minimal withdrawal from this scan position demonstrates the aortic root, and, anteriorly, the right ventricular outflow tract in short axis (figure 6). Clockwise rotation of the probe demonstrates the superior portion of the atrial septum and the junction of the superior caval vein with the right atrial cavity (figure 7). Minimal withdrawal from this scan position will allow the visualization of the distal superior caval vein in short axis and, posteriorly, the right upper pulmonary vein entering the left atrium. Counterclockwise rotation of the probe at this level shows the aortic root and the proximal ascending aorta in short axis, the proximal pulmonary trunk and the left atrial appendage (figure 7). Posteriorly the roof of the left atrial cavity is still visualized. Separated by only a crest of tissue from the left atrial appendage, the left upper pulmonary vein is visualized following counter-clockwise rotation of the probe. Repositioning the probe in the mid line and using anterior tip angulation the pulmonary trunk together with its bifurcation can be visualized. Whereas the right pulmonary artery normally can be followed from this scan position by clockwise rotation as far as its first peripheral branching, the visualization of the proximal left pulmonary artery, however, remains difficult in most cases due to interposition of the left main bronchus between this vessel and the oesophagus. Only a distal segment can be demonstrated anterior of the descending aorta using counter-clockwise rotation of the probe.

Upper oesophagus Further withdrawal of the probe in a midline position demonstrates the aortic arch, together with a segment of the distal pulmonary trunk. Scanning towards the right the right pulmonary artery is visualized together with a proximal segment of the superior caval vein. Scanning towards the left the descending aorta is demonstrated. The latter can be followed down to the diaphragm and cranially up to the aortic arch and the head and neck vessels. Following the aorta upwards from the diaphragm to the aortic arch minimal counter-clockwise rotation of the probe has to be employed, since the relation of the oesophagus and the descending aorta vary at different levels within the mediastinum.

A synopsis of the standard transoesophageal views used in the assessment of relevant examples of congenital cardiac malformations is listed in table 1. As a general rule cross-sectional imaging should be carried out first for the definition of the atrial arrangement and the determination of the mode of atrioventricular and ventriculo-arterial connections. Thereafter the venous return to the heart should be assessed in more detail, as should be the atrial chambers themselves, the atrial septum, the atrioventricular valves, the ventricular chambers and the ventricular septum and, finally, the great arteries. Colour flow mapping

Table 1: Summary of the sequence of imaging planes used during transoesophageal echocardiographic investigations in the assessment of congenital heart disease.

Standard examination planes	Visualization of cardiac structures	Examples of pathology visualized
Transgastric planes	Ventricular morphology	ventricular dominance
	Ventricular relations	supero/inferior ventricles,
	Ventricular function	
	Chordal apparatus MV	parachute MV
	Inferior caval vein	interruption
	Hepatic veins	individual drainage
	Atrioventricular valves	common valve orifice
Lower oesophagus	Coronary sinus	unroofed CS
		dilated CS
	Muscular inlet septum	muscular inlet VSD
	Tricuspid valve	Ebstein's malformation
		tricuspid atresia
	4-chamber view	offsetting AV valves
		atrioventricular septal defects
		perimembranous VSD
Left atrial views	Mitral valve	valvar regurgitation
		endocarditis
	Atrial septum	patent oval foramen
		deficiencies of the oval fossa
	Pulmonary veins	anomalous connection
	Atrial chambers	atrial arrangement
		Mustard baffles
		cor triatriatum
	LV outflow tract	membranes
		arterial override
Mid oesophagus	Atrial septum	sinus venosus defect
		juxtaposition
	VA junction	transposition/malposition
	Superior caval vein	anomalous drainage
	Aortic valve	valvar aortic stenosis
	RV outflow tract	infundibular stenosis
	Pulmonary trunk	patent ductus arteriosus
		supravalvar stenosis
	Pulmonary arteries	palliative shunts
		peripheral stenosis
Thoracic aorta	Aortic arch, descending aorta	coarctation
		collaterals

Legend: CS = coronary sinus; LV = left ventricle; MV = mitral valve; RV = right ventricle; RUPV = right upper pulmonary vein; VA = ventriculo-arterial; VSD =ventricular septal defect.

Note: In the sequential analysis of congenital heart disease atrial morphology has to be determined first, thereafter atrioventricular and ventriculoarterial connections should be defined and systemic and pulmonary venous drainage determined. Only thereafter the examination should focus on the relevant lesions.

should be performed at all cardiac levels for the rapid identification of flow turbulence, and pulsed wave Doppler interrogation should be performed at relevant areas of interest. The use of colour M-mode studies, in our experience, proved to be particularly valuable in young children with rapid heart rates, since it allows a precise temporal resolution of abnormal flow patterns.

In order to further elucidate the transesophageal assessment of cardiac anatomy a series of anatomic cross-sections corresponding to the standard transesophageal imaging planes are displayed on the following pages.

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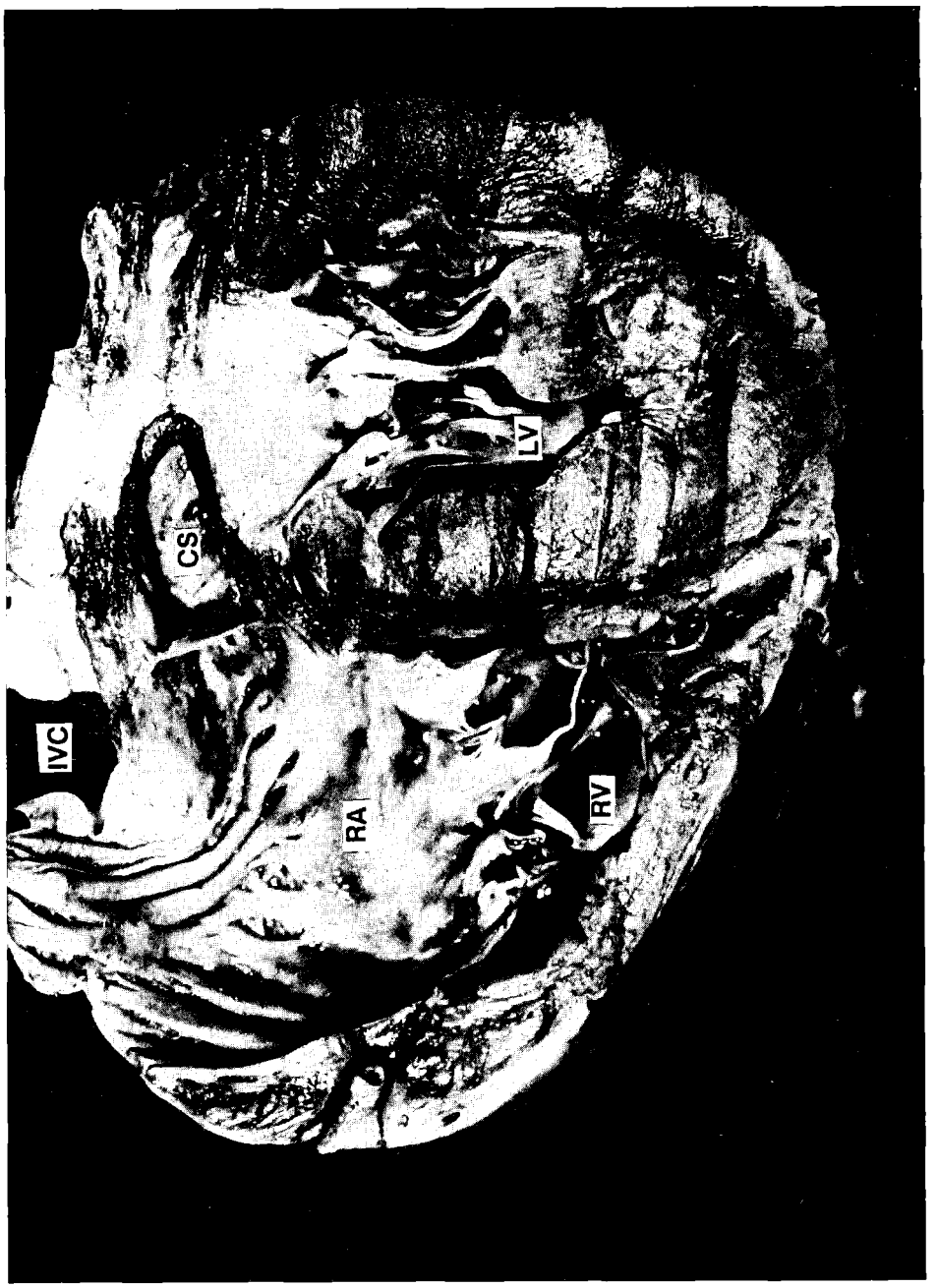

Figure 3: Transverse axis views obtained from scan positions near the oesophageal hiatus cut through the coronary sinus and demonstrate the inferior tricuspid valve leaflet and the muscular ventricular inlet septum. To the right and posterior the Eustachian valve is located at the site of drainage of the inferior vena cava.
Legend: CS = coronary sinus; IVC = inferior vena cava; LV = left ventricle; RA = right atrium; RV = right vemtricle.

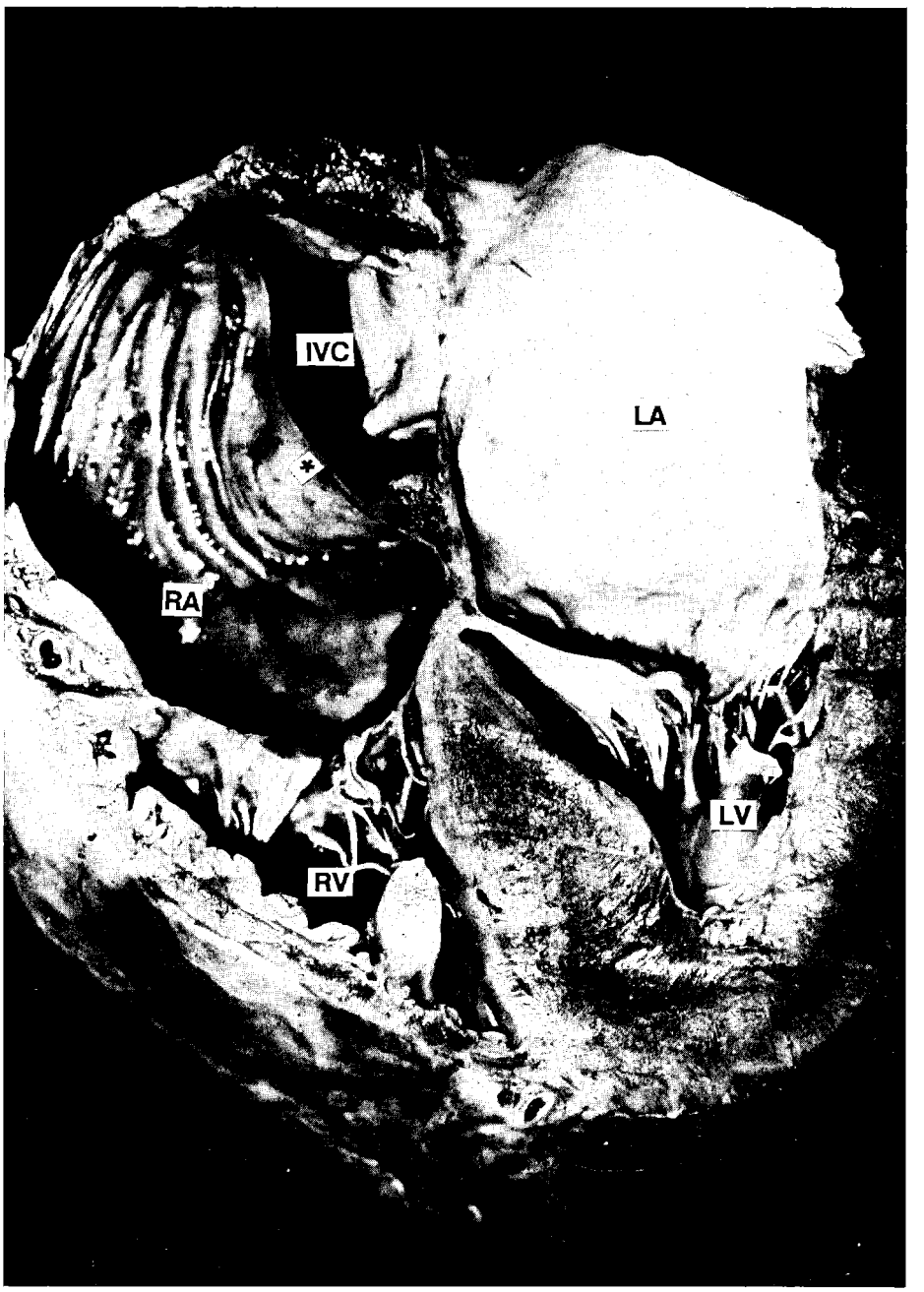

Figure 4: Anatomic correlate of a standard transoesophageal four-chamber view. The offsetting of both atrioventricular valves is nicely demonstrated as are their chordal attachments. Note the course and attachment of the Eustachian valve (asterix) to the atrial septum.
Legend: LA = left atrium; others see figure 3.

Figure 5: Anatomic cross-section corresponding to a standard left atrial view, for the assessment of the atrial septum (oval fossa, asterix) and the left ventricular outflow tract. Note the atrioventricular septum (arrow) and its relation to the septal leaflet of the tricuspid valve.
Legend: LVOT = left ventricular outflow tract; others see fig 3.

Figure 6: Slight advancement of the probe results in an oblique cross-section through the aortic root. The aortic valve leaflet seen closest to the transducer is the non-coronary cusp. Note the appearance of the (muscular) atrial septum and the trabecula septomarginalis within the right ventricular outflow tract.
Legend: RVOT = right ventricular outflow tract; others see fig 3,4.

Figure 7: Mid oesophageal view at the level of the cavo-atrial junction and the level of the arterial valves. Both atrial appendages are seen in cross-sections. Note the crest of atrial tissue at the site of the cavo-atrial junction, and the bifurcation of the left main coronary artery.
Legend: Ao = aorta; LAA = left atrial appendage; RAA = right atrial appendage; RUPV = right upper pulmonary vein; SVC = superior vena cava.

Chapter 3

Transesophageal Echocardiography in Children with Congenital Heart Disease - An initial experience

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Summary Transesophageal echocardiography, using a single plane (transverse axis) dedicated pediatric probe, was performed prospectively in 25 anesthetized children undergoing routine cardiac catheterization or intracardiac surgery, in order to assess any potential role it may have in the initial diagnosis, perioperative management and post operative follow-up of children with congenital heart disease.

The age ranged from 1 year to 14 years 10 months (mean 6.1 years); the weight from 6.5 to 52 kilograms (mean weight 22.4 kg). Studies were successful in all patients; no complications were encountered. The results of the transesophageal studies (combined imaging, color flow mapping and pulsed wave Doppler) were correlated both with those of prior precordial studies and the information obtained at cardiac catheterization. Transesophageal echocardiography allowed a more detailed evaluation of the morphology and function of systemic and pulmonary venous return, both atria, interatrial baffles, both atrioventricular valves, and the left ventricular outflow tract. Additional information was obtained in 15 patients (60 %). Problem areas for single plane transesophageal imaging were the apical interventricular septum, the right ventricular outflow tract and the left pulmonary artery. The intraoperative use of transesophageal echocardiography allowed the assessment of the surgical repair and monitoring of ventricular function and volume status while the patient was weaned from cardio pulmonary bypass.

Transesophageal echocardiography in pediatric patients is of additional value in three main areas a) in the morphologic diagnosis of congenital heart disease, b) for perioperative monitoring and c) for post surgical follow-up.

Introduction Since the introduction of transesophageal echocardiographic high resolution imaging allied to color flow mapping into cardiological practice, some three years ago, it has gained widespread acceptance both as a diagnostic technique with specific applications in the adult outpatient clinic and as a perioperative monitoring technique in cardiac surgery (1-4).

Although experience with this new technique in the adult cardiac patient group has rapidly expanded over the recent years its use in the pediatric cardiac patient group is still very limited (5,6). The reasons for this are multifold. Firstly, all standard adult transesophageal probes exceed 12 mm in maximal diameter and thus are too large for safe use in small children. Secondly, patient tolerance towards transesophageal investigations is poor in older children even under sedation and should not be attempted in children under eight years of age (7); general anaesthesia is required for studies in small children. Thirdly, the precordial ultrasound approach in unoperated children with congenital heart disease normally allows the investigator to obtain high quality diagnostic information, thus limiting the need for an alternative acoustic window to the heart. However, in unoperated children with chest wall abnormalities or cardiac malposition and in operated children precordial studies are frequently compromised.

Clinical studies with transesophageal echocardiography in both the adult outpatient clinic and operating theatre have shown that the technique provides excellent image quality and allows detailed insights into atrial pathology, the morphology and function of both atrioventricular valves and the left ventricular outflow tract (8-11). From this experience in adult patients, and our own recent experience in adolescents and adults with congenital heart disease (12), it might be expected that a small dedicated pediatric transesophageal probe could be of additional diagnostic value in specific areas of congenital heart disease.

In the prospective study reported below, we used pediatric transesophageal echocardiography in twenty five unselected children with congenital heart disease to evaluate its potential benefits and limitations in the initial diagnosis, as a perioperative monitoring technique and for postoperative follow-up. The results were compared and contrasted with both the prior complete precordial ultrasound studies and findings at cardiac catheterization and/or surgical inspection.

Patients and Methods

Study patients Twenty five unselected pediatric cardiac patients were studied with a small single plane (transverse axis) transesophageal probe. Twenty four had congenital cardiac malformations and one child had rheumatic mitral valve disease (Table I). The age at investigation ranged from 1 year to 14 years 10 months with a mean age of 6.1 years. The patient's weight varied between 6.5 and 52 kilograms (mean weight 22.4 kg, median 18 kg). Nine patients weight less than 15 kilograms, nine less than 25 kg and 7 patients more than 25 kg. Transesophageal studies were performed in three patients undergoing cardiac surgery (no. 1-3) and in 22 patients undergoing routine cardiac catheterization on clinical grounds under general anesthesia. Of these 22 patients 7 patients were studied preoperatively, 6 following palliative procedures and 9 after prior total surgical correction (Table II).

Approval by the hospital ethical committee (Academisch Ziekenhuis Rotterdam) was granted prior to commencement of the series; informed parental consent was obtained prior to individual studies. Appropriate anesthetic monitoring was conducted during the studies. Antibiotic endocarditis prophylaxis was not administered in any patient.

Transesophageal studies A specially designed pediatric transesophageal probe (Aloka Company, Japan) was used together with an Aloka SSD 870 ultrasound system. The ultrasound transducer is mounted onto a pediatric bronchoscope with a shaft diameter of 7 mm

Table I: Forty-three lesions studied in 25 patients

Lesion	Number
Secundum atrial septal defect	7
Partial anomalous pulmonary venous drainage	2
Tricuspid atresia	3
Ebstein's anomaly	1
Complete AVSD	1
Ventricular septal defect	8
Tetralogy of Fallot	2
Double outlet right ventricle	2
Left ventricular outflow obstruction	2
Aortic valve stenosis	2
Pulmonary atresia / VSD	4
Peripheral pulmonary stenosis	2
Rheumatic mitral valve disease	1
Post op coarctation / aneurysm ascending aorta	1
Post op Mustard procedure	3
Post op Fontan procedure	1
Post op tetralogy of Fallot	1
Note: most patients had more than one lesion	Total n = 43

and a maximal tip dimension of 7 x 8 mm. The ultrasound transducer (24 elements, frequency 5 MHz) allows cross-sectional imaging, color flow mapping and pulsed wave Doppler investigations. The ability of probe angulation by use of the steering wheel is restricted to anterior/posterior directions. Transesophageal imaging is performed in a series of basal short axis views, which are obtained by varying the level of probe insertion within the esophagus and by probe rotation and angulation (4).

In every case the transesophageal studies were performed with the patient under general anesthesia and with endotracheal intubation. A small amount of anesthetic gel was applied to the tip of the transducer in order to allow more gentle movement within the esophagus and to further improve contact. The probe, with the tip partially flexed, was introduced into the hypopharynx and thereafter, allowing the tip to move freely, was advanced to the lower third of the esophagus. In small children insertion of the probe was facilitated under direct laryngoscopic vision of the hypopharynx.

The studies followed a predetermined standardized scheme. Using a series of basal short axis cuts from within the esophagus cross-sectional imaging was used to assess 1. the atrial appendage morphology, 2. the structure and function of the atrial cavities and the interatrial septum, 3. the precise pattern of systemic and pulmonary venuous drainage, 4. the structure and function of both atrioventricular valves, 5. the morphology of the ventricular chambers and the interventricular septum, 6. the ventricular outflow tracts and the ventriculo-arterial junction, and 7. both great arteries. Emphasis was placed on detailed assessment of the individual specific lesions. The cross-sectional imaging studies were complemented in every case by color flow mapping of relevant areas of interest within the heart and by pulsed Doppler sampling for the assessment of intracardiac blood flow velocity profiles.

Table II: Patient data: age, weight, cardiac malformations and previous surgical procedures

Patient No.	Age yrs.	Weight kg	Cardiac malformation	Surgical Correction / palliation
1.	5 6/12	17.3	Solitus, AVconcord., VAconcord., ASD II	ASD closure
2.	4	15.6	Solitus, AVconcord., VAconcord., ASD II	ASD closure
3.	6 1/12	19.2	Solitus, AVconcord., VAconcord., ASD II	ASD closure
4.	12	35.9	Solitus, AVconcord., VAdiscor., ASD II	Mustard procedure
5.	8 3/12	21.0	Solitus, AVconcord., VAconcord., PA, VSD	Left Blalock, central shunt
6.	4 1/12	13.3	Solitus, AVconcord., VAconcord., ToF, supravalvular PS, peripheral PS	VSD closure Transannular patch
7.	5 7/12	18.2	Solitus, AVconcord., VAconcord., mesocardia, ASD II, peripheral PS	ASD closure, Patch plasty left PA
8.	13 9/12	40.3	Solitus, absent right, VAconcord., ASD II, PAPVC	Central interposition shunt
9.	13 8/12	40.6	Solitus, AVconcord., VAconcord., rheumatic mitral valve disease	none
10.	14 10/12	42.5	Solitus, AVconcord., VAconcord., PAPVC bicuspid AoV, aortic coarctation	Repair aortic coarctation Aortic root replacement
11.	7 10/12	20.0	Right atrial isomerism, complete AVSD, right hand ventricular topology, DORV	Right and left Blalock shunt
12.	5 7/12	18.2	Solitus, AVconcord., VAconcord., severe AS	none
13.	1 8/12	11.0	Solitus, absent right, VAconcord., ASD II, PA	Left Blalock shunt
14.	2 3/12	13.6	Solitus, AVconcord., VAconcord., VSD, PS	none
15.	1 1/12	11.1	Solitus, AVconcord., VAconcord., ToF	none
16.	12 7/12	39.2	Solitus, AVconcord., VAdiscord., ASD II	Mustard procedure
17.	1 9/12	9.4	Solitus, AVconcord., VAconcord., VSD	Pulmonary artery banding
18.	13 5/12	52.1	Solitus, AVconcord., VAconcord., Ebstein	ASD closure
19.	12 9/12	49.5	Solitus, DILV, VAconcord, ASD II, VSD	Glenn shunt, Fontan
20.	1 5/12	10.7	Solitus, AVconcord., VAconcord., ASD II, PA	Transannular patch
21.	1	6.5	Solitus, AVconcord., VAconcord., VSD, PS	none
22.	8 4/12	18.2	Solitus, AVconcord., DORV,	none
23.	5 4/12	18.3	Solitus, AVconcord., VAdiscord., ASD II	Mustard procedure
24.	1 4/12	8.9	Solitus, AVconcord., VAconcord., PA, VSD	none
25.	1 8/12	9.0	Solitus, absent right, VAconcord., ASD II	Right Blalock

Legend: AoV = aortic valve; AS = aortic stenosis; ASD II = secundum atrial septal defect; AV = atrioventricular; AVSD = atrioventricular septal defect; concord. = concordance; DILV= double inlet left ventricle; discord. = discordance; PA = pulmonary atresia; PAPVC = partial anomalous pulmonary venous connection; PS = pulmonary stenosis; TGA = transposition of the great arteries, ToF = tetralogy of Fallot; VA = ventriculoarterial; VSD = ventricular septal defect; yrs. = years.

The ultrasound studies were continuously recorded on video tape and were analyzed and interpreted on-line and off-line both at normal speed and by frame-by-frame analysis. Off-line analysis was carried out by two independent observers. The findings were correlated with the findings of a prior precordial echocardiographic study and with the results of simultaneous cardiac catheterization (22 patients) and/or the findings at surgical inspection (6 patients). The time allowed for each study was half an hour, and could routinely be completed within this time range. Studies were performed while the lines for cardiac catheterization were introduced or, in case of intraoperative studies, while the patient underwent opening or closure of the chest.

Results

Transesophageal studies could be performed successfully in all of the twenty five patients without clinical difficulties. No patient was found to show any signs of trauma or bleeding from the esophagus following introduction and manipulation of the probe. No episodes of arrhythmias were encountered. Cardiac catheterization was delayed in only one patient (no. 11) with complex congenital heart disease, in whom the total time of the study was about 50 minutes. In all of the other patients complete studies could be obtained without delaying the scheduled procedures.

Atrial situs The distinct morphology of both atrial appendages could be determined in all patients. The morphologically right atrial appendage, with its broad junction with the atrial cavity, consistently could be differentiated from the morphologically left atrial appendage with its narrow junction and its long, narrow and largely crenelated appearance. Thus direct diagnosis of atrial situs was possible in every patient. Situs solitus was documented in 24 patients. In one of these patients (no 17) a previous subcostal ultrasound scan had demonstrated an interrupted inferior vena cava with hemiazygos continuation, thus suggested left atrial isomerism. The bronchus morphology was not delineated on a routine chest x-ray. The transesophageal study in this patient documented situs solitus of the atria. Right atrial isomerism (bilateral morphologically right atrial appendages) was clearly documented in one patient (no 11), in whom both subcostal scans and a chest x-ray suggested this diagnosis.

Cardiac position One patient studied was known to have mesocardia (no 7). In this patient the precordial approach was severely restricted due to the overlying sternum and lung tissue; however no limitations were encountered to perform a complete transesophageal study. The remaining 24 patients had levocardia.

Pulmonary venous return The left upper pulmonary vein could be visualized with ease in all 25 patients. The right upper pulmonary vein was clearly demonstrated in 23 patients just posterior to the site of drainage of the superior vena cava. In two patients (no 8,10) in whom the right upper pulmonary vein could not be demonstrated at this site (figure 1) anomalous pulmonary venous drainage was suspected. Subsequent scanning along the superior vena cava, including color flow mapping and pulsed wave Doppler sampling, identified the site of drainage of the right upper pulmonary vein into the superior vena cava to be approximately 5 cm cranial of the right atrium in one (no 8) of the two patients. In the second patient the site of drainage could not be defined; presumably due to interposition of the right bronchus. Cardiac catheterization confirmed the anomalous drainage in both cases.

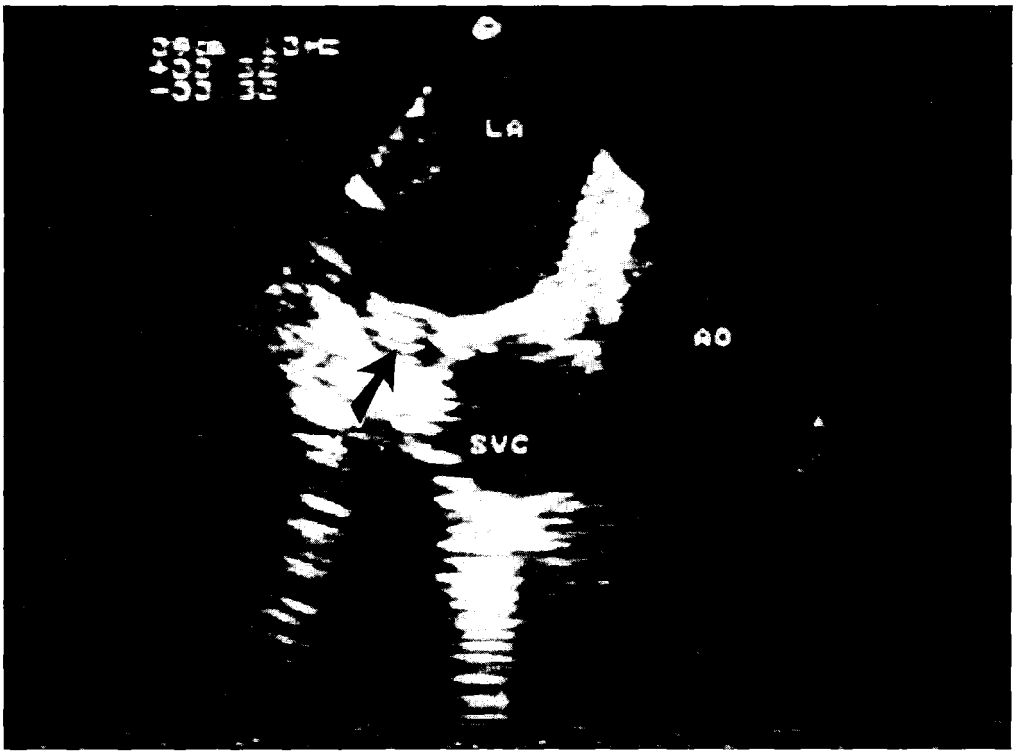

Figure 1: Transesophageal basal short axis scan in the assessment of anomalous drainage of the right upper pulmonary vein. In this patient, the vessel could not be demonstrated at its normal position just posterior to the superior vena cava (arrow). It was subsequently defined to drain into the superior caval vein some 5 cm above the cavoatrial junction. The other three pulmonary veins connected normally.
Legend: Ao = aorta; LA = left atrium; SVC = superior vena cava.

Drainage of the left lower pulmonary vein could be identified, either by cross-sectional imaging or by color flow mapping, in 21 patients and of the right lower in 18 of the 25 patients studied. Pulsed wave Doppler sampling of individual pulmonary veins could be performed with very good alignment in all patients.

Systemic venous return The site of the systemic and hepatic venous return could be defined in every patient. It was better demonstrated than by precordial studies in one patient with right atrial isomerism (no 11). Transesophageal studies demonstrated a left persistent superior vena cava in two patients. In the patient with right atrial isomerism (no 11) the vessel drained directly into the roof of the left sided atrium. In a second patient (no 10) the exact course of the vessel, which was interposed between the left upper pulmonary vein and the left atrial appendage, (figure 2) and its drainage into the coronary sinus were clearly demonstrated by cross-sectional imaging and color flow mapping. Following a Fontan-type procedure for double inlet left ventricle in one patient (no 19) combined cross-sectional imaging and color flow mapping allowed demonstration of a Glenn shunt, which was documented to be unobstructed. In this patient an arteriovenous fistula was detected that was previously unknown; the characteristic flow patterns were documented by pulsed wave

Doppler sampling. A Eustachian valve of varying size was demonstrated in the majority of patients. No case was encountered were it caused an intraatrial obstruction or turbulent flow patterns. A Thebesian valve was appreciated in only two patients.

Figure 2: Transesophageal demonstration of a left persistent superior caval vein (l SVC), which connected to the coronary sinus. The vessel is found interposed between the left atrial appendage (LAA) and the left upper pulmonary vein (LUPV).

Atrial septum The integrity of the atrial septum or the presence of atrial shunting could be assessed with ease by combined cross-sectional imaging and color flow mapping as the ultrasound beam is almost perpendicular to the septum. The atrial septum was documented to be intact in 5 unoperated children studied. In none of them there was any evidence of atrial shunting by any other correlative technique. A patent foramen ovale was readily diagnosed in 5 patients by the appearance of a two-layered structure on cross-sectional imaging (figure 3). On subsequent color flow mapping studies evidence of blood flow within these layers was found. Left atrial catheterization via these defects could be performed in 4 of the 5 patients, in none in whom the transesophageal study defined an intact atrial septum. A secundum type atrial septal defect was demonstrated in 7 patients studied. In one patient (no 13) with an imperforate tricuspid valve a massively enlarged fossa ovalis flap was demonstrated; color flow mapping across the defect revealed unobstructed right to left shunting. The atrial component of a complete atrioventricular septal defect was clearly demonstrated in one patient (no 11). Intraoperative monitoring of atrial septal defect closure in 3 patients revealed minuscule residual shunts post bypass in two.

Figure 3: Documentation of a patent foramen ovale by transesophageal cross-sectional imaging. These are readily identified by their two-layered appearance and the evidence of blood flow contained within the space on subsequent color flow mapping studies.
Legend: Ao = aorta; LA = left atrium; RA = right atrium.

Mustard baffles Three patients underwent cardiac catheterization for reevaluation after a Mustard procedure for transposition of the great arteries. Both the systemic venous and pulmonary venous pathways could be assessed completely by transesophageal echocardiography. Combined cross-sectional imaging, color flow mapping and pulsed wave Doppler tracings allowed detailed information on baffle function. In particular the inferior limb of the systemic venous atrium and the junction of the superior vena cava with the superior limb were demonstrated with much more clarity than that obtained by precordial studies. Three baffle leaks (2 tiny, 1 moderate) were demonstrated in two patients, in whom precordial scanning suggested a single leak in one. Pulsed wave Doppler tracings of pulmonary veins allowed the exclusion of individual pulmonary venous obstruction in all three patients. Suspicion of mid baffle pulmonary venous obstruction on the basis of a precordial study was confirmed by the transesophageal study in a further patient (no 23). In addition, the transesophageal study in this patient revealed mild obstruction of the inferior limb of the systemic venous atrium, which could not be demonstrated by prior precordial scanning. Cardiac catheterization under general anesthesia in the three patients confirmed the transesophageal findings except the existence of two tiny baffle leaks; no additional

abnormalities were noted.

Coronary sinus The site of drainage and the distal third of the coronary sinus could be demonstrated in 24 patients. Absence of the coronary sinus was documented in one patient with right atrial isomerism (no 11).

Tricuspid valve Three patients with tricuspid atresia were studied. Precordial studies suggested an absent right connexion in all of them. However transesophageal cross-sectional imaging defined an imperforate tricuspid valve in one patient (no 13) (figure 4). The finding was subsequently confirmed by intraoperative echocardiography and by surgical inspection.

Ebstein's malformation of the tricuspid valve was known in one patient (no 18). The transesophageal study allowed good insight into the morphology and function of the septal (apical displacement and tethering) and posterior (redundant tissue) tricuspid valve leaflet. The anterior leaflet, however, was difficult to analyze due to restricted views from within the esophagus; the large size of the patient (52 kg) precluded the definition of all chordal insertions. One patient (no 14) with a perimembranous inlet ventricular septal defect was shown to have multiple tricuspid tissue tags inserting into the crest of the interventricular septum. Chordal straddling was excluded in all patients studied.

Color flow mapping and pulsed wave Doppler investigations were used to assess tricuspid valve function. Whereas any tricuspid regurgitant was readily demonstrated, assessment of inflow patterns was generally limited due to poor alignment to the Doppler beam. Routinely the information derived from precordial studies was superior. In one patient (no 19) who underwent prosthetic patch closure of the tricuspid valve in combination with a Fontan procedure for double inlet left ventricle, the perfect closure of the valve was documented.

Mitral valve An additional commissure of the mitral valve with chordal insertions into the interventricular septum was demonstrated by cross-sectional imaging in three patients studied. Prior precordial scanning did not show this anomaly in any patient. In one patient (no 4) the chordal insertions contributed to an associated left ventricular outflow obstruction on the basis of a fibromuscular "membrane". A parachute mitral valve with a single papillary muscle, not appreciated on the prior precordial scan, was documented on transgastric short axis views (no 12; figure 4). M-mode studies allowed the registration of the opening and closing patterns of the mitral valve and pulsed wave Doppler sampling excluded any inflow obstruction. In one patient (no 11) with a complete atrioventricular septal defect transesophageal scans allowed a more detailed insight into the common valve morphology and function than did the prior precordial study.

Ventricles, interventricular septum Eight patients studied had a ventricular septal defect. Transesophageal echocardiographic studies clearly demonstrated these defects and their relationship to the atrioventricular valves. Insertion of chordae into the crest of the interventricular septum were demonstrated in two patients. A false tendon in the left ventricle was detected in one patient (no 12). Assessment of the size and function of both ventricles by transesophageal and precordial studies were in agreement.

Ventricular outflow tracts Two patients had a left ventricular outflow tract obstruction. Whereas precordial imaging suggested a discrete subaortic fibromuscular membrane in both, the transesophageal study defined involvement of the mitral valve apparatus in one (no 19). However, the almost perpendicular direction of the ultrasound beam to the outflow tract precluded pulsed Doppler evaluation. Additional information concerning the morphology of

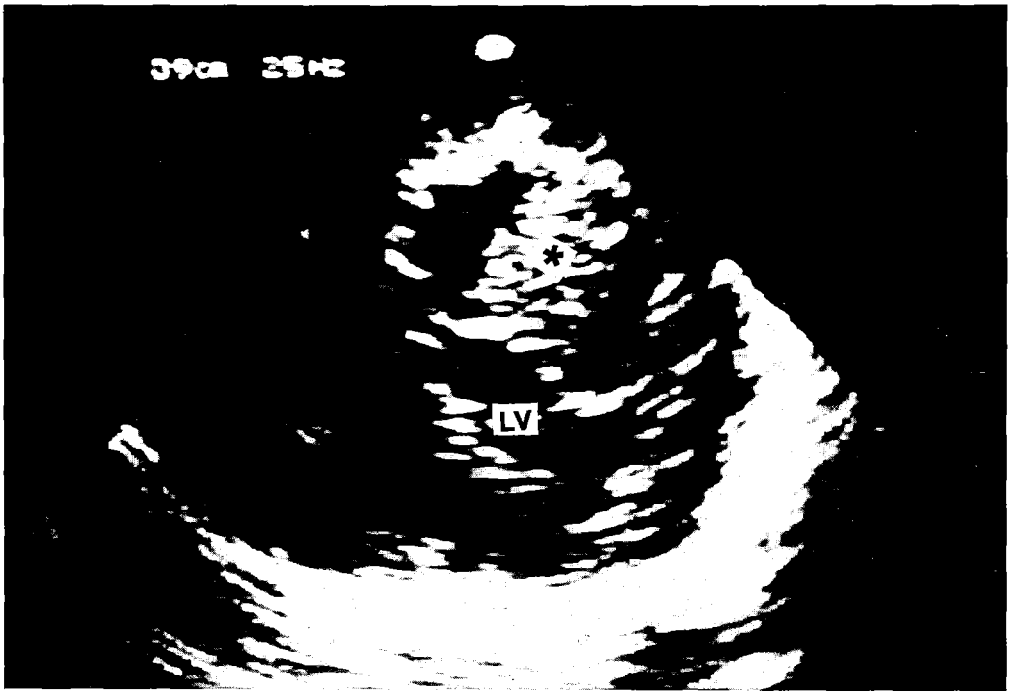

Figure 4: Diagnosis of a parachute mitral valve by a transgastric short axis scan. The morphology of the single papillary muscle (asterix), its insertion and the chordal insertions could be assessed closely by a sequence of scans. Pulsed Doppler studies excluded any inflow obstruction.
Legend: LV = left ventricle.

the right ventricular outflow tract was obtained in only one patient (no 15), in whom precordial scans suggested a doubly committed ventricular septal defect. Cross-sectional imaging from within the esophagus better defined the existence of a tiny (fibrous) remnant of the infundibular septum; both arterial valves were demonstrated in a single plane. Assessment of the degree of arterial override of the aorta in two unoperated patients with tetralogy of Fallot and two with a double outlet right ventricle was unreliable by transesophageal imaging; praecordial studies yielded better information.

Pulmonary arteries Four patients studied had pulmonary atresia. The central pulmonary artery system could be demonstrated in all of these patients; in one (no 24) with much more clarity than by precordial scans (figure 5). Transesophageal studies in the remaining 21 patients allowed demonstration of the size and course of the main pulmonary artery, the bifurcation and the proximal branch pulmonary arteries. Cross-sectional imaging allowed exclusion of a suspected supravalvular pulmonary stenosis in one patient (no 14). The pulmonary valve itself was routinely better demonstrated by precordial studies. The course of the right pulmonary artery up to the crossing with the superior vena cava could be demonstrated in 21 patients; in the remaining 4 patients interposition of the right bronchus precluded imaging further distally than 2 or 3 cm from the bifurcation. Pulsed wave Doppler

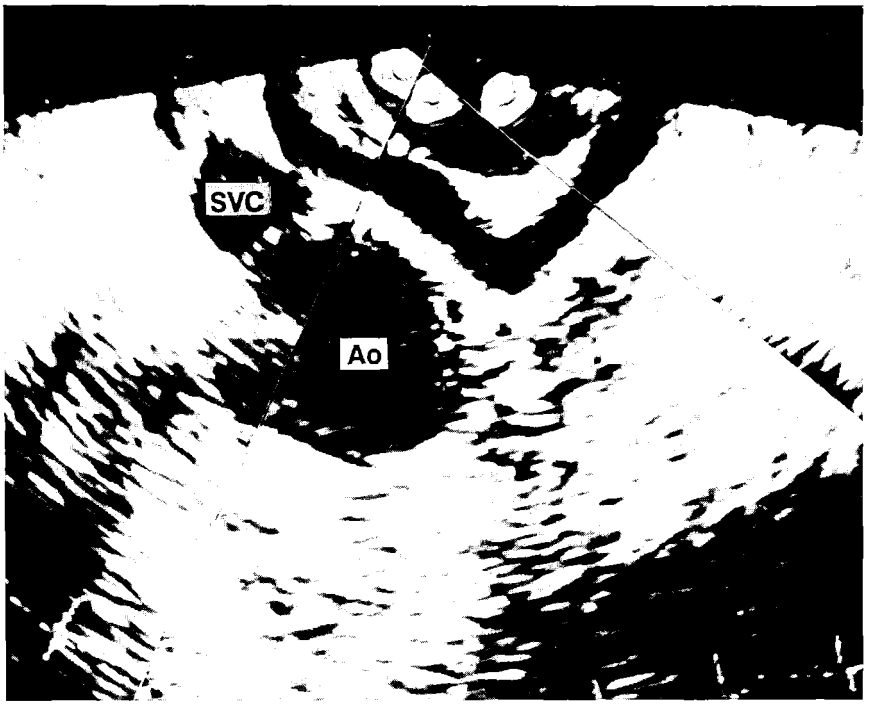

Figure 5: Composite picture of the central pulmonary arterial system in a patient with pulmonary atresia, ventricular septal defect, as seen on a series of basal short axis views at this level.
Legend: Ao = aorta; SVC = superior vena cava.

interrogation allowed rapid identification of the vessel and documented the patency of systemico-pulmonary shunts and the related flow patterns. The precise site and the morphology of the anastomosis could only be demonstrated in 1 (no 25) of the 5 five patients with palliative shunts. The origin of the left pulmonary artery could be assessed in 20 patients and was found to be obstructed in three patients. In one of these patients (no 5) the stenosis was better defined by transesophageal than by precordial imaging and color flow mapping. In 8 patients a distal segment of the left pulmonary artery could be demonstrated at its crossing with the descending aorta.

Aorta Two patients studied were found to have a functionally bicuspid aortic valve. The morphology was better assessed by transesophageal than by precordial imaging. Assessment of valvular aortic stenosis or incompetence was found to be largely limited due to poor alignment of the Doppler beam. The precordial approach consistently yielded better results. Of the ascending aorta only the proximal portion could be demonstrated from within the esophagus. No abnormalities were noted in any patient. The aortic arch and the descending aorta could be examined in every patient. One patient (no 10) had discrete narrowing of the descending aorta, after end-to-end anastomosis for aortic coarctation. No patient had a ductus arteriosus.

Coronary arteries The left main coronary artery was visualized in 17 of the 25 patients

studied, whereas the right coronary artery was appreciated in only four. Neither coronary artery could be demonstrated in all 3 postsurgical patients with transposition of the great arteries. An intramural left main coronary artery (figure 6) was identified in one patient (no 10) with normally related great vessels. Slight variation of the level of probe insertion demonstrated the origin of this coronary artery just above the commissure between the left and non coronary cusp of the aortic valve. Color flow mapping revealed laminar flow patterns, however pulsed wave Doppler sampling was unsuccessful.

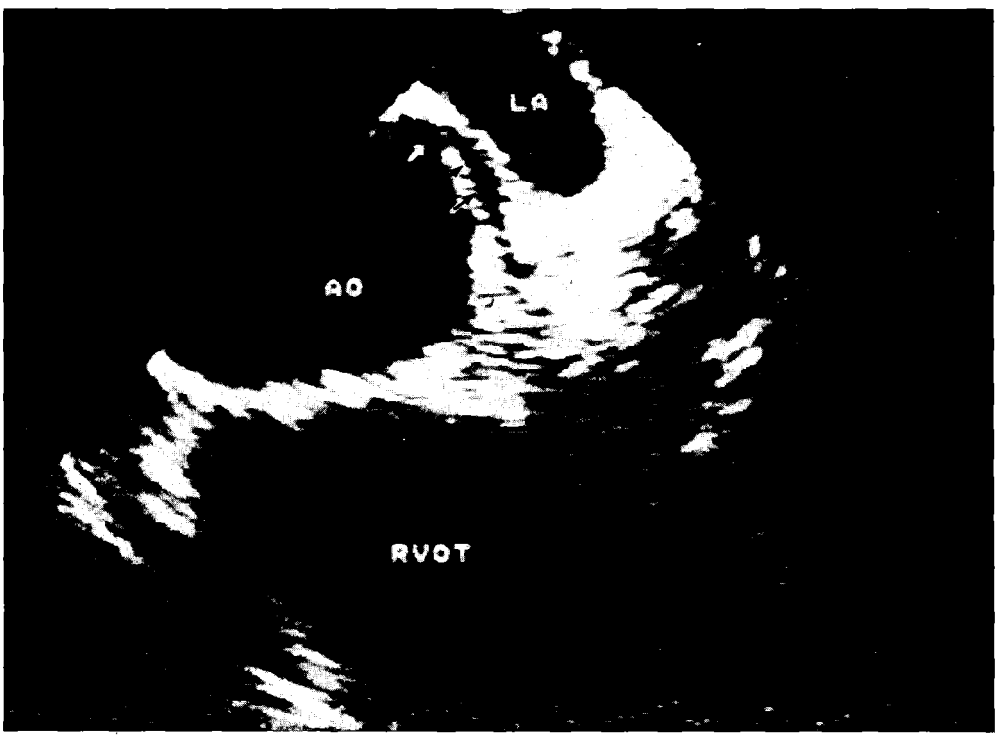

Figure 6: Transesophageal demonstration of a left intramural coronary artery (arrows). Slight variation in transducer insertion documented the origin just above the commissure of the left and non coronary cusp of the aortic valve. Color flow mapping documented laminar flow patterns, excluding any obstruction. Note the acute angulation of the vessel and its intimate relation with the aortic wall.
Legend: Ao = aorta, LA = left atrium, RVOT = right ventricular outflow tract.

Discussion

Experience with transesophageal echocardiography in children with congenital heart disease is still limited (5,6). Current transducers which are used in the adult clinics are too large to allow investigations of small children. This study, to our knowledge, is the first report on a correlative transesophageal versus precordial ultrasound study in children with congenital heart disease, in which the findings were further correlated with the results of cardiac catheterization and/or surgical inspection.

Practicability The availability of a special dedicated pediatric probe with a maximal tip dimension of 7 x 8 mm allowed safe investigations of children with a body weight of more than six kilograms; with increasing experience the investigation of even smaller children under general anesthesia should become both safe and practical. The technique of introducing and manipulating the transducer is relatively easy to learn. Studies could be performed successfully in all patients and no complications were encountered. The scheduled procedure was delayed in only one patient. With increasing experience the majority of studies can be completed within ten to fifteen minutes.

Morphologic information Transesophageal imaging in small children was felt to be easier compared with investigation of adult patients as the acoustic window is relatively large. In contrast to the adult practice the main and right pulmonary arteries can be assessed in the vast majority of children. However single plane (transverse axis) transesophageal imaging in children with congenital heart disease has inherent limitations. Due to the interposition of the left bronchus only small segments of the left pulmonary artery can be visualized. The right ventricular outflow tract, although it is visualized in most children, is cut in oblique sections which are unsatisfactory to define pathologic changes. Definition of arterial override above a ventricular septal defect by transverse axis imaging was found to be unreliable as demonstration of the crest of the interventricular septum together with the arterial valve is impossible. In older children assessment of the antero-apical portion of the interventricular septum remains incomplete due to the large distance from the esophagus.

Transesophageal studies yield the maximal morphologic information on those cardiac structures that are closest to the esophagus. In this series additional information, when compared with prior precordial ultrasound studies, was obtained in fifteen patients (60 %) (see Table III). The additional information was provided on 1. the venous return, 2. both atrioventricular valves and their subvalve apparatus, 3. the morphology of the left ventricular outflow tract and 4. the central pulmonary arteries. The clarity with which chordal insertions of the atrioventricular valves and obstructive lesions of the left ventricular outflow tract can be demonstrated implies that transesophageal imaging may become the diagnostic technique of choice for the identification or exclusion of chordal straddling and for definition of left ventricular outflow obstruction. In addition, transesophageal echocardiography is the only in-vivo technique that allows direct diagnosis of atrial situs.

Hemodynamic information The hemodynamic evaluation of intracardiac blood flow by means of combined color flow mapping and pulsed wave Doppler studies from within the esophagus is largely limited due to generally poor alignment to intracardiac blood flow. In particular the assessment of blood flow across both ventricular outflow tracts and the arterial valves is compromised by an almost ninety degree angle of the ultrasound beam to the direction of blood flow. The hemodynamic evaluation of these aspects of congenital heart disease will thus remain the domains of precordial ultrasound studies and cardiac catheterization. In contrast the right upper and both left sided pulmonary veins and mitral valve flow can be assessed by pulsed wave Doppler with an optimal angle of incidence. The detailed insight that the technique provides into pulmonary vein flow patterns is likely to open up a new field for the hemodynamic evaluation of both valve regurgitation and ventricular diastolic function in congenital heart disease. Transesophageal studies for post operative evaluation of the Mustard circulation were found to be superior to precordial

Table III: Additional information provided by transesophageal studies when compared to prior precordial studies.

Patient	Precordial study	Transesophageal study
4	LVOTO, fibromuscular membrane	Precise attachments
		Additional commissure mitral valve
5	? Stenosis left PA	Severe stenosis left PA
7	Normal mitral valve	Additional commissure mitral valve
	? Pulmonary artery stenosis	Peripheral pulmonary stenosis
8	? Pulmonary venous drainage	Right upper pulmonary vein to SVC
10	? Coronary artery anatomy	Intramural left coronary artery
	? Left persistent SVC	Left persistent SVC
11	? Coronary sinus	Absent coronary sinus
12	Small mitral valve	Single papillary muscle
		False tendon in left ventricle
13	Absent right connexion	Imperforate tricuspid valve
14	? Supravalar pulmonary stenosis	No supravalar stenosis
		Tricuspid tissue tags
15	? Doubly committed VSD	Doubly committed VSD
16	? Baffle leak inferior limb	Moderate baffle leak inferior limb
		Tiny leak superior limb
17	? Left atrial isomerism	Situs solitus
	LVOTO, fibromuscular membrane	Precise attachments of membrane
	? Mitral valve abnormality	Additional commissure mitral valve
19	? Function Glenn shunt	Unobstructed Glenn shunt
		Arteriovenous fistula
23	Pulmonary venous obstruction	Mid baffle pulmonary venous obstruction
		Mild obstruction inferior limb
24	? Central pulmonary artery system	Good size pulmonary artery system

Legend: LVOTO = left ventricular outflow obstruction; PA = pulmonary artery; SVC = superior vena cava; VSD = ventricular septal defect; ? = the precordial ultrasound study remained uncertain about the morphology and / or function of the specific structure.

studies, with their results comparing favorably with those of cardiac catheterization.

Intraoperative use Transesophageal echocardiography was found to be a versatile intraoperative monitoring technique. Its major advantage, compared with intraoperative epicardial echocardiography, is that it does not interfere with the surgical procedure and that it allows continuous monitoring of ventricular performance and volume. However, as imaging planes are limited and demonstration of the right ventricular outflow tract is poor, single plane transesophageal echocardiography can not be expected to replace intraoperative epicardial echocardiography. Future perspectives for transesophageal echocardiography in pediatric cardiac patients may be its use as a monitoring technique on the intensive care unit and during interventional catheterization procedures.

The results of this study indicate that the role of transesophageal echocardiography in children with congenital heart disease both in the primary diagnosis and as a peri- and postoperative monitoring technique is likely to be rapidly increasing in the near future. Its main advantages are that specific areas of the heart are better visualized and that the

technique allows more detailed morphologic information on these aspects of congenital heart disease than does the precordial approach. However the semi-invasive nature of this procedure, the fact that general anaesthesia is required in (small) children, and the comparatively inferior diagnostic information other aspects of congenital heart disease largely has to dictate its clinical use.

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Chapter 4

The diagnosis of atrial situs by transoesophageal echocardiography

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Summary In a prospective study direct visualization of both atrial appendages was attempted during transoesophageal echocardiographic investigations in 132 patients with congenital heart disease. High quality cross-sectional images, delineating the unique morphologic details of both atrial appendages were obtained in every patient. Abnormal cardiac position, such as dextrocardia (4 patients) or mesocardia (2 patients) did not pose any problems for transoesophageal assessment of both atrial appendages. Thus, direct the diagnosis of atrial situs was allowed in every patient. Atrial situs solitus was present in 127 patients studied. Three patients were found to have situs inversus, one patient left atrial isomerism and one patient right atrial isomerism. No patient with juxtaposed atrial appendages was encountered. All patients had prior subcostal ultrasound scans for assessment of the morphology and relationship of the suprarenal abdominal great vessels and the related patterns of hepatic venous drainage. Patients with abnormal atrial situs had correlative high kilovoltage filter beam radiographs for assessment of bronchus morphology. The results of situs determination obtained by either method were in agreement.

In this series transoesophageal echocardiography allowed the direct and accurate visualization of both atrial appendages and thus the determination of atrial situs in all patients studied. Transoesophageal echocardiography may prove to be the most reliable in-vivo technique for determination of atrial situs.

Introduction

The determination of atrial situs is the initial fundamental diagnostic step for sequential chamber localization in the diagnosis of complex congenital heart disease (1-3). In clinical practice atrial situs is normally determined non-invasively by either radiographic definition of the bronchus morphology (4-6) or by the assessment of the morphology of the abdominal great vessels, their relationship to one another and to the spine (7). These two techniques used in combination yield reliable results in the majority of patients (8,9). A complete abdominal ultrasound investigation including the definition of the splenic status should be performed in all cases where abnormal situs is suspected. However, as there are reports on

discord between the arrangement of the atria and the thoracic and abdominal organs (10-12) all indirect diagnostic approaches for the definition of atrial situs have potential limitations.

The ultimate diagnostic investigation for the clinical definition of atrial situs would be a technique that would allow identification of the unique morphologic characteristics of either atrial chamber. As both the systemic and pulmonary venous connections in complex congenital heart disease can be anomalous their determination is of little value for the diagnosis of atrial situs. The anatomical characteristics of the christa terminalis or the right atrial venous valves are too subtle to allow differentiation on either praecordial echocardiographic or angiographic grounds. Therefore only the visualization of both atrial appendages, with their constant unique morphologic characteristics (13) would allow a direct diagnosis of atrial situs. In newborns praecordial echocardiography sometimes allows demonstration of both atrial appendages by a series of parasternal short axis views (7). However, with increasing size of the patient and the natural reduction in the praecordial ultrasound window direct visualization of both atrial appendages from the praecordium becomes a virtual impossibility.

Transoesophageal echocardiography has gained widespread clinical acceptance in adult practice during the recent years (14-16). The technique allows high quality imaging in virtually every patient. In particular the cardiac chambers closest to the oesophagus can be assessed with much more detail than this is usually possible from the praecordium. Experience with this new diagnostic technique in the field of congenital heart disease is still limited (17,18). However, our own recent experience in adolescents and adults with congenital heart disease suggested that both atrial appendages could be demonstrated in every patient. Therefore transoesophageal echocardiography may be expected to be a reliable (albeit semi-invasive) diagnostic tool for the direct morphologic diagnosis of atrial situs.

Patients and Methods

Complete transoesophageal echocardiographic studies were performed in 132 patients with congenital heart disease as part of a larger study into the evaluation of congenital cardiac lesions from the oesophageal approach. The age at study ranged from 3.7 to 68 years (mean age 28.2 years). The atrial appendage morphology was assessed and atrial situs was defined in every patient as part of a complete study protocol. Transoesophageal studies were performed either on an outpatient basis in 123 patients who belonged to the adolescent and adult patient group and during routine cardiac catheterization in 9 children. Approval by the hospital ethical committee, Erasmus University Rotterdam, was obtained prior to commencement of the study protocol. Informed consent of the patients, or their parents, was obtained prior to individual studies.

Transoesophageal studies Studies were successful in all of the 132 patients. For investigations on an outpatient basis patients were fasted for at least 4 hours. No antibiotic prophylaxis was administered; mild sedation using a short acting benzodiazepine was used in seven patients. Following local anaesthesia of the pharynx the probe was introduced into the hypopharynx, and then, with the patient being asked to swallow, was advanced to the lower part of the oesophagus. A continuous electrocardiographic tracing was used for monitoring during the studies; no other monitoring technique was employed in outpatients.

Isolated supraventricular premature beats were occasionally seen, however no patient had a sustained supraventricular tachycardia, and cardioversion (either medical or electrical) was not required in any patient. The adolescent and adult patients were studied with various 5 / 5.6 MHz transoesophageal single-plane probes (maximal tip dimension 13 x 15 mm) on either a Toshiba SSH 160 A or a Vingmed CFM 700 ultrasound system.

Studies in ten children were performed during routine cardiac catheterization with the patient under general anaesthesia, as part of a prospective study into the role of transoesophageal echocardiography in the paediatric patient with complex congenital heart disease. In these patients a specially dedicated 5 MHz paediatric transoesophageal probe was used with an Aloka SSD 870 ultrasound system. The small dimensions of the probe (maximal tip dimension 7 x 8 mm) allowed investigation of children with more than 10 kilograms bodyweight without any complications (e.g. signs of bleeding, oesophageal trauma).

Imaging technique The initial step of every investigation was the assessment of atrial appendage morphology by cross-sectional imaging. The probe was advanced within the oesophagus to the level of the atria and then an optimal scan position was chosen by varying the level of insertion and by rotation of the probe. Care was taken to obtain almost transverse sections of both atrial appendages by slight up-down- and sideward movement of the tip of the transducer by use of the probe steering mechanism. Only these transverse sections allowed the demonstration of both the junction with the atrial cavity and the internal morphology of the atrial appendage at the same time. The right atrial appendage is usually visualized when scanning a basal short axis view, which is obtained by following the superior vena cava into the right atrium. The cavity of the right appendage is shown in transverse sections, to the right of the aortic root and above the tricuspid valve. Following anticlockwise rotation of the probe the left atrial appendage is seen to the left of the aorta just above the mitral valve and anterior to the orifice of the left upper pulmonary vein.

Correlative investigations All patients had prior complete transthoracic ultrasound investigations, including subcostal scans for assessment of the morphology and relationship of the suprarenal abdominal great vessels and the related patterns of hepatic venous drainage. Patients with detected abnormal atrial situs had additional correlative high kilovoltage filter beam radiographs for assessment of bronchus morphology.

Results

Appendage morphology Transoesophageal echocardiography allowed direct visualization of the morphology of both atrial appendages in all of the 132 patients studied. The morphologically right atrial appendage was demonstrated having a short blunt appearance and a broad junction with the right atrial cavity. In contrast the morphologically left atrial appendage was demonstrated having a long, narrow and crenelated appearance and a narrow junction with the atrial cavity (figure 1). The delineation of these unique features consistently allowed the differentiation of either atrial appendage. Thus the determination of atrial situs was allowed in every patient studied. Cardiac malrotation or malposition, present in 6 patients (4 dextrocardia, 2 mesocardia), did not prevent complete assessment of appendage morphology in any patient, although variations in the examination technique had to be adopted to scan the relevant imaging planes.

Atrial situs solitus was present in 127 patients. The right sided appendage was identified

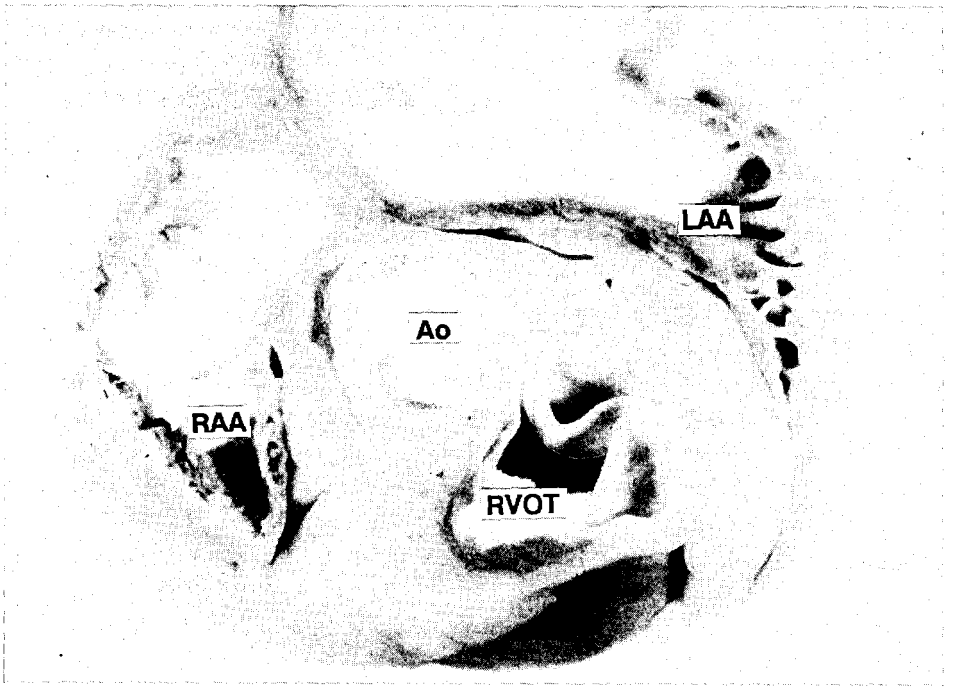

Figure 1a: Anatomic section through both atrial appendages, simulating a transoesophageal basal short axis view. The morphologically right atrial appendage (RAA) has a broad junction with the venous cavity, whereas the morphologically left atrial appendage (LAA) has a narrow junction. Note the different patterns of appendage trabeculation and crenelation. (Ao = aorta; RVOT = right ventricular outflow tract).

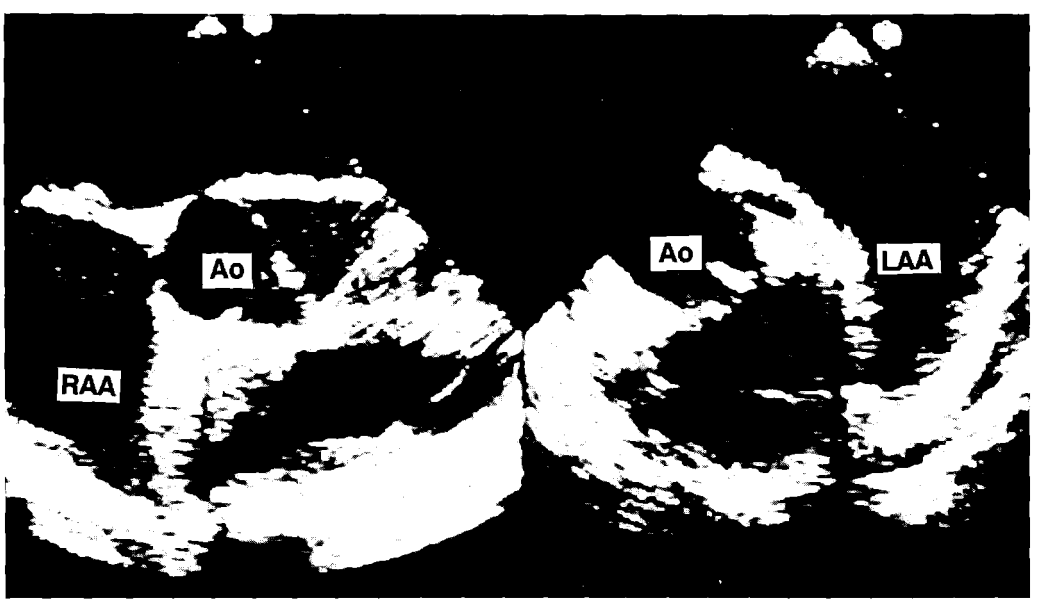

Figure 1b: Transoesophageal study in a patient with atrial situs solitus. The unique morphologic characteristics of either atrial appendage are readily demonstrated.

to be of right morphology and the left sided appendage of left morphology in all of these patients (figure 1b). There was no case of discrepancy between the transoesophageal diagnosis and the previous clinical, ultrasonographic and radiographic diagnosis or the anatomical findings at previous surgical repair.

Atrial situs inversus was demonstrated in 3 patients, all of whom had a congenitally corrected transposition (atrioventricular discordance and ventriculoarterial discordance). One patient had dextrocardia. The right sided atrial appendage in these patients was identified to be of left morphology, and the patient's left sided appendage of right morphology. All three patients had a high kilovoltage filtered beam radiograph for identification of bronchial situs and a repeat subcostal ultrasound scan for demonstration of the abdominal vessel morphology and splenic status. Either technique demonstrated mirror image arrangement of the thoracic and abdominal organs. No discrepancies between the results of determination atrial situs by these indirect investigations and the transoesophageal studies were found. Praecordial echocardiography did not allow demonstration of either atrial appendage in any of these patients.

Atrial isomerism Bilateral morphologically left atrial appendages were demonstrated in one patient with dextrocardia. Both appendages were found to have a narrow junction with the corresponding atrial cavity, clearly identifying them to be of left appendage morphology (figure 2). This result of the transoesophageal study was in agreement with the diagnosis made on the basis of a filtered beam radiograph, showing bilaterally left bronchi (long, curved bronchi with late bifurcation). A previous subcostal ultrasound investigation of the upper abdomen had shown drainage of the hepatic veins to a single sub diaphragmatic

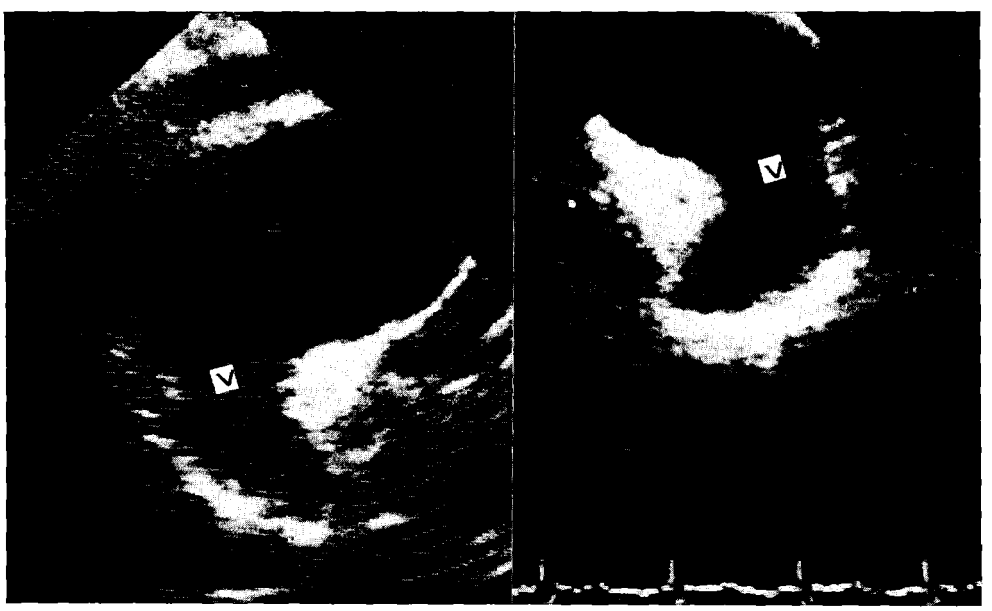

Figure 2: Left atrial isomerism in a patient with congenitally corrected transposition and dextrocardia seen from within the oesophagus. Both atrial appendages have a narrow junction with the atrial cavity (arrows); their cavity is long and narrow. These features identify both appendages to be of left morphology, thus the patient has left isomerism.

confluence into the right sided atrium. There was a hemiazygos continuation of the inferior vena cava. Thus, the patient, on the sole basis of the subcostal ultrasound investigation, could well have had situs solitus with interruption of the inferior vena cava. Dextrocardia prevented a complete praecordial echocardiographic investigation, and in particular the demonstration of either atrial appendage. The transoesophageal study clearly demonstrated left atrial isomerism, with left hand pattern ventricular topology (19) and discordant ventriculoarterial connexions.

Bilateral morphologically right atrial appendages, thus right atrial isomerism, was clearly demonstrated in a seven year old child with a normally positioned heart. The broad junction with the venous component of either atrium and a short, blunt shape of both appendages led to their identification as being of right morphology (fig 3). The child was found to have a left persistent superior vena cava, which drained into the roof of the left sided atrium, interposed between the left sided atrial appendage and the left upper pulmonary vein. Visualization of either atrial appendage from the parasternal or subcostal windows was impossible during praecordial studies. A previous filter beam radiograph had demonstrated bilateral short eparterial bronchi with early bifurcation, suggesting right atrial isomerism. Subcostal echocardiography identified a right vena cava inferior which was anterior to the abdominal aorta, but hepatic venous drainage remained unclear. Absence of the coronary sinus and complete drainage of all hepatic veins into the right sided inferior vena cava could

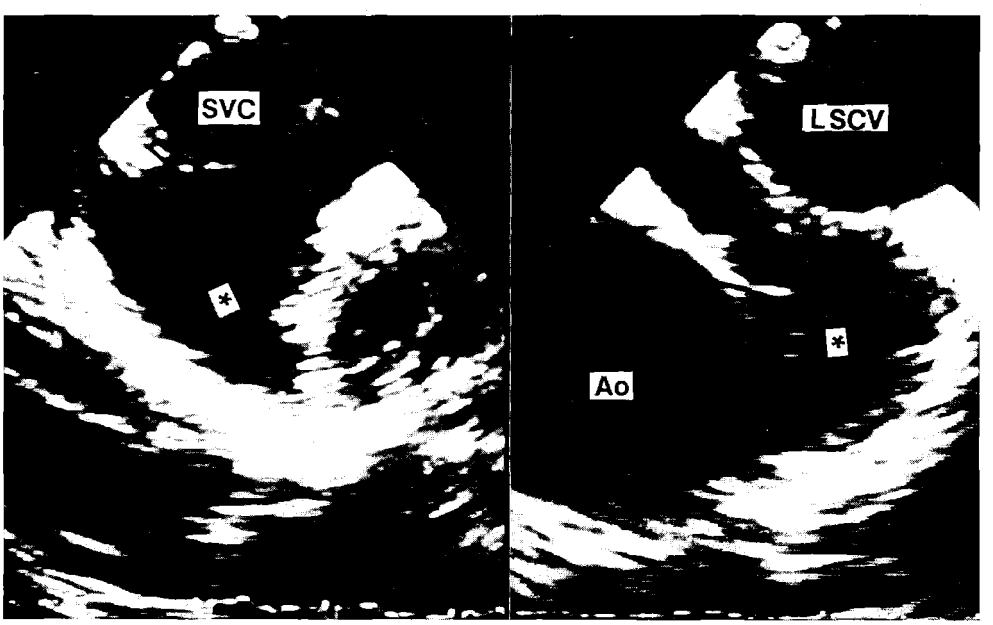

Figure 3: Right atrial isomerism. The right sided appendage has a broad junction with the atrium; the cavity (star) is wide and has a blunt ending, thus is of right appendage morphology. The left sided appendage has a wide cavity (star) and a blunt ending. The junction with the atrial cavity is narrowed due to a left persistent superior vena cava (l SVC) draining into the left sided, morphologically right atrium.
Legend: Ao = aorta; L = left; SVC = superior vena cava)

only be demonstrated by transoesophageal echocardiography. The transoesophageal study further showed a complete atrioventricular septal defect, right hand pattern ventricular topology and double outlet right ventricle with the aorta in front and to the right of an unperforated pulmonary ostium.

Discussion

Abnormal atrial situs is, by itself, of little clinical importance. The complex associated malformations that are almost always present with either right or left atrial isomerism are the determinant factors for (surgical) management and natural history (9,13,20). However, identification of atrial situs has to be the initial step for the complete diagnosis of all congenital heart malformations (3).

Conventional diagnosis High kilovoltage filter beam radiographs for identification of bronchial situs, introduced by Deanfield et al (6), yield reliable information of atrial situs in the vast majority of patients. The low radiation dose and the non-invasive character of this investigation allow routine use in patients with congenital heart disease. Subcostal echocardiographic investigations of the suprarenal great vessels, as first described by Huhta et al (7), is the most extensively used screening method for detection of abnormal atrial situs. Again reliable information is obtained in the majority of patients. Both techniques, combine the advantages of non-invasiveness and high sensitivity and specifity, as cases with discord between the arrangement of the atria and the thoracic and abdominal organs are rare (10-12). Furthermore they are not restricted to selected patient groups and yield information regardless of the patient's age. However, the unique morphologic details of either right or left atrial appendage cannot be demonstrated.

Transoesophageal diagnosis The high quality imaging that was obtained by transoesophageal cross-sectional imaging in every patient of this study allowed consistent differentiation of both atrial appendages by detailed demonstration of their unique morphologic characteristics. The morphologically right atrial appendage is visualized as a short, blunt cavity that has a broad junction with the smooth-walled atrial chamber. In contrast the left atrial appendage is demonstrated to be a long, narrow and largely crenelated cavity with a narrow junction with the venous component of the left atrium. The junction of the atrial appendage with the atrial cavity is the most constant feature for identification of appendage morphology, as it is independent from any degree of atrial distention, which may result from valvar dysfunction or intracardiac shunting. Abnormal cardiac positions, such as dextro or mesocardia, required slightly modified transducer manipulation but never represented a limitation for the transoesophageal approach. In general the probe manipulation technique in patients with dextrocardia is a mirror image of the series of probe maneuvers carried out in patients with normally positioned hearts. If present, a left persistent superior vena cava will be found interposed between the left atrial appendage and the left upper pulmonary vein. The misinterpretation of this extra "cavity" for an atrial appendage is excluded by following the course of the individual structures. Identification is furthermore aided by combined colour flow mapping and pulsed wave Doppler studies.

Clinical implications Although the findings of this study would suggest that transoesophageal echocardiography should allow the direct identification of both atrial appendages in

every patient, its role solely for the determination of atrial situs will remain limited in clinical practice, as it is an unpleasant semi-invasive technique. Only recently small dedicated paediatric transoesophageal probes have been developed, that allow the safe investigation of small children under general anaesthesia or heavy sedation. At present transoesophageal studies in patients with congenital heart disease would appear to be indicated in those symptomatic patients with poor praecordial ultrasound windows who have problems related to either systemic or pulmonary venous return, atrial lesions, abnormalities of the atrioventricular junction or pathologic changes of the left ventricular outflow tract. In all patients with congenital heart disease studied by transoesophageal echocardiography the determination of atrial situs should be carried out routinely, as it is an easy and reliable method.

Transoesophageal echocardiography would appear to be the most reliable in-vivo technique for the identification of atrial situs, as it consistently allows the delineation of the unique morphologic details of either atrial appendage. It may be considered the ultimate diagnostic technique in cases were ambiguity of situs determination persists after combined radiographic and transthoracic ultrasound studies.

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Chapter 5

The assessment of juxtaposed atrial appendages by transoesophageal echocardiography

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Summary As part of a prospective study into the diagnostic role of transoesophageal echocardiography in children with complex congenital heart disease, the atrial morphology was assessed in sixty-two unoperated children. Using the direct visualization of atrial appendage morphology, fifty-eight were shown to have usual atrial arrangement, two were documented to have right atrial isomerism and two left atrial isomerism. Of those with atrial situs solitus, four children were documented to have left juxtaposition of the atrial appendages. Only two of these patients were identified during praecordial echocardiographic re-evaluation, and three on angiocardiographic examination. Surgical confirmation was obtained in three, and juxtaposition was excluded in the remaining cases. The transoesophageal cross-sectional imaging features of left juxtaposition of the atrial appendages are unique and readily diagnostic of this entity. They include 1. a lateral deviation of the mid-portion of the atrial septum and 2. a frontal orientation of the antero-superior portion forming the floor and the posterior wall of the junction of the right-sided atrial appendage with the venous component of the atrial cavity. The knowledge of these morphologic characteristics is important, as, otherwise, this malformation may be misinterpreted as being an atrial septal defect.

The results indicate that transoesophageal echocardiography would appear to be the most sensitive preoperative diagnostic technique in detecting or excluding juxtaposed atrial appendages.

Introduction

Juxtaposition of the atrial appendages is a rare anomaly which is normally associated with complex congenital heart disease (1-3). Left juxtaposition, a condition in which both atrial appendages lie side-by-side on the left side of the arterial trunk, is the most common form and occurs about six times more often than right juxtaposition (4). As a direct consequence of the abnormal location of the atrial appendages there is a change in the geometry of the atrial

septum. Knowledge of the presence of this malformation is important to the surgeon when considering either a Mustard or Senning procedure for complete transposition or a Fontan procedure (5,6). In addition, a Rashkind septostomy may be difficult in this malformation (7).

The preoperative diagnosis of juxtaposed atrial appendages can be made from the angiographic appearances, when special emphasis is placed on identifying this anomaly (8). Praecordial cross-sectional imaging is an alternative and valuable investigative technique when used in infants and young children (9,10). However, difficulties may persist in making the diagnosis in the older child with a limited praecordial ultrasound window. Other indirect methods previously described for the diagnosis of juxtaposed atrial appendages include changes on either the electrocardiogram or the chest X-ray (11,12).

Transoesophageal echocardiography, which has recently been introduced as a diagnostic technique in the paediatric age group (13,14), provides detailed morphologic insights into the atrial and atrioventricular levels of the heart. Thus, it may provide the paediatric cardiologist with a sensitive diagnostic method for the identification or exclusion of juxtaposition of the atrial appendages. In this report, we describe the transoesophageal echocardiographic features of left juxtaposed atrial appendages.

Patients and Methods

Patients Sixty-two unoperated children were studied by transoesophageal echocardiography, as part of a prospective study into the role of transoesophageal echocardiography in complex congenital heart disease . Approval by the Hospital Ethical Committees of the Academisch Ziekenhuis Rotterdam and the National Institute of Cardiology, Mexico City, was obtained prior to commencement of the study and prior informed parental consent was obtained in each case. The age at the transesophageal investigation ranged from 2.5 months to 14.8 years with a mean age of 4.3 years. Their body weight ranged from 3.7 to 48 kilograms with a mean weight of 14.2 kilograms.

Scanning equipment Seventeen patients were studied using an Aloka twenty-four element paediatric transoesophageal probe connected to an Aloka SSD 870 ultrasound system. Forty-five patients were studied with a prototype forty-eight element paediatric transoesophageal probe (Department of Experimental Echocardiography, Thoraxcenter Rotterdam) connected to either a Hewlett Packard Sonos 1000 or 500, or a Toshiba SSH 160 A or 140 A ultrasound system. Both phased array transducers allowed 5 MHz cross-sectional imaging in the transverse-axis plane, colour flow mapping and pulsed wave Doppler interrogation. The maximal tip circumference of either probe measured 30 mm (7x8 mm and 5x10 mm respectively) and the maximal shaft diameter 7 mm. Probe tip manipulation was restricted to anterior/posterior angulation only.

Technique Of the total of 62 patients, 43 were studied under general anaesthesia either while undergoing concomitant cardiac catheterization (n = 32), or as elective transoesophageal studies on a day care basis (n = 13). The remaining 19 patients were studied in the immediate preoperative period prior to sternotomy. All transoesophageal studies were performed without knowledge of the echocardiographic or angiocardiographic results concerning atrial morphology.

After sufficient lubrification with anaesthetic gel, the transoesophageal probe was inserted

either blindly or under direct laryngoscopic vision into the hypopharynx and then was advanced into the lower third of the oesophagus. No additional antibiotic prophylaxis was administered, as is our general policy. A predetermined standardized protocol of investigation (to include cross-sectional imaging and colour flow mapping studies) was completed in every patient.

Imaging planes Both atrial cavities were scanned by recording standard transoesophageal four-chamber planes, using right or left lateral rotation of the probe. In the normal situation, the left sided atrial appendage can be visualized lying anterior to the left upper pulmonary vein, when scanning a basal short axis view and using left lateral transducer orientation. The right sided atrial appendage can be demonstrated using high four chamber views by scanning anterior to the cavo-atrial junction and supero-lateral of the tricuspid valve annulus. The appendage morphology was closely studied, so as to allow the direct diagnosis of the atrial arrangement in keeping with the methodology described previously in our work (15). The morphologically right atrial appendage was identified by the demonstration of its broad junction with the venous component of the atrium, its short, triangular cavity and its blunt ending. The morphologically left atrial appendage was identified by its narrow junction with the venous component of the atrium, its narrow and largely crenelated lumen and its pointed ending.

The orientation and the integrity of the atrial septum was then assessed by a series of four-chamber and basal short axis views, which were obtained by gradually withdrawing the probe within the oesophagus. The superior portion of the atrial septum could be visualized best, when using image planes which demonstrated the ventriculo-arterial junctions. In addition, the systemic and pulmonary venous connections were assessed in relation to the atrial septum. The integrity of the atrial septum was assessed both on cross-sectional imaging and subsequent colour flow mapping studies.

Evaluation The transoesophageal studies were continuously recorded onto video tape. The determination of atrial arrangement and the assessment of the atrial morphology was performed on-line. The results of the transoesophageal studies were then correlated with the information derived from prior praecordial ultrasound studies, which were reviewed, (n =62), with the results obtained by either simultaneous or cardiac catheterization (n = 54), and with the findings during subsequent or later surgical inspection (n = 42). Photographs were later taken from still-frame pictures displayed on a Video monitor.

Results

Atrial arrangement Fifty-eight children were defined by the transoesophageal study to have a usual atrial arrangement (atrial situs solitus), two were documented to have right atrial isomerism and two left atrial isomerism. The determination of atrial arrangement by trans-oesophageal, praecordial echocardiographic and radiographic criteria was in agreement in all but one patient. In this patient the prior subcostal scan had documented an interruption of the inferior caval vein with an azygos continuation, thus suggested that left atrial isomerism was present. The transoesophageal definition of atrial situs solitus was confirmed by subsequent surgical inspection.

Juxtaposed atrial appendages Of the fifty-eight patients with atrial situs solitus four patients were documented to have left juxtaposition of a morphologically right atrial appendage. Both atrial appendages were demonstrated to lie to the left of the arterial trunk (figure 1).

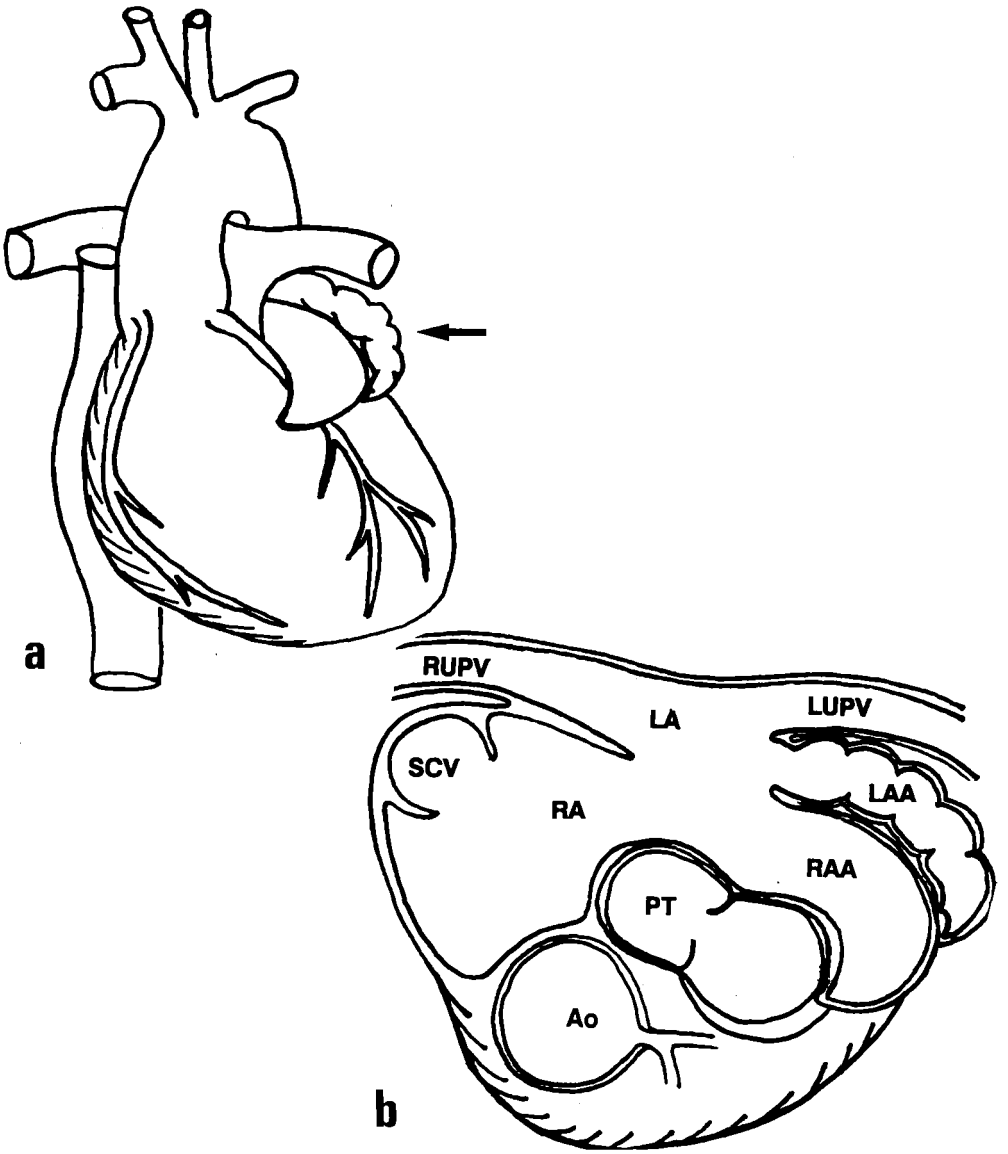
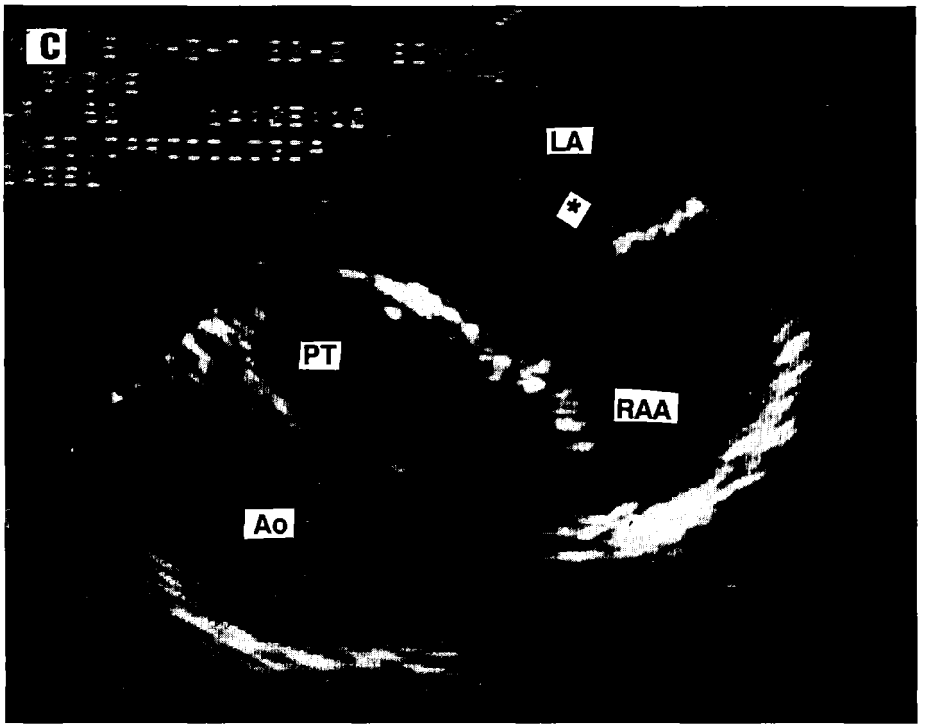


Figure 1: Left juxtaposition of the atrial appendages.
a Schematic drawing of a heart with complete transposition and left juxtaposition.
b diagram of a basal short axis cut obtained from scanning at the level of the ventriculo-arterial junction (arrow). Both atrial appendages lie posterior and to the left of the arterial trunks. The supero-anterior portion of the atrial septum is deviated to a frontal plane, forming the posterior wall and the floor of the venous component of the right atrium at its junction with the right atrial appendage.
c The corresponding transoesophageal cross-sectional image defines the atrial morphology. The right atrial appendage is clearly visualized, as is the large single deficiency of the oval fossa ("secundum" type defect, asterix) communicating with the left atrium.
Legend: Ao = aorta, LA = left atrium, LAA = left atrial appendage, LUPV = left upper pulmonary vein, LVOT = left ventricular outflow tract, PT = pulmonary trunk, RA = right atrium, RAA = right atrial appendage, RUPV = right upper pulmonary vein, SCV = superior caval vein)



A true side-by-side arrangement was documented in only one case; in two cases the left atrial appendage was found to lie to the left and superior of the right atrial appendage, and in one case to the left and inferior. The direct recognition of appendage morphology was not compromised by any of these appendage arrangements. Two children had absent right atrio-ventricular connections, two concordant. The ventriculo-arterial connections were discordant in three, and double outlet of the right ventricle in one (Table).

Table: Left juxtaposition of the atrial appendages - patient data

No	age yrs	weight kg	sex	AV junction	VA junction	sPS	Relationship of great arteries
1	2.7	11.5	f	concordant	discordant	yes	Ao right and anterior of PT
2	1.1	8.4	f	concordant	discordant	yes	Ao right and anterior of PT
3	2.9	11.5	m	absent right	discordant	no	Ao anterior and right of PT
4	1.6	8.1	m	absent right	DORV	no	Ao anterior and left of PT

Legend: Ao = Aorta, AV = atrio-ventricular, DORV = double outlet right ventricle, f = female, kg = kilogram, m = male, PT = pulmonary trunk, sPS = subpulmonary stenosis, VA = ventriculo-arterial, yrs = years.

Prior praecordial studies had failed to anticipate left juxtaposition of the atrial appendages in all four patients. On a subsequent review of these studies evidence of juxtaposed atrial appendages was found in one, and during further praecordial reinvestigation left juxtaposition

could be demonstrated in another patient using the criteria previously described by Chin and Rice (9,10). Angiocardiographic studies (performed in all four patients) documented left juxtaposition in three patients. The findings were confirmed during later surgical inspection in two patients. No other patient in this series was documented by either surgical inspection or angiocardiography to have juxtaposed atrial appendages.

Atrial septum Using standard transoesophageal four-chamber planes the atrial septum in patients with left juxtaposition was found to be deviated to a more frontal plane, giving the impression of a relatively large left atrial chamber and a small right atrial cavity (figure 2). On more cranial planes, at the level of the ventriculo-arterial junction, the antero-superior portion of the atrial septum then was found to be orientated in a true frontal plane, forming the floor and the posterior aspect of the venous component of the right atrium at its junction with the right atrial appendage.

All four patients had deficiencies of the oval fossa ("secundum" atrial septal defects). On subsequent colour flow mapping studies these defects were documented to be single, un-restrictive defects in three patients, and multiple restrictive defects in one patient.

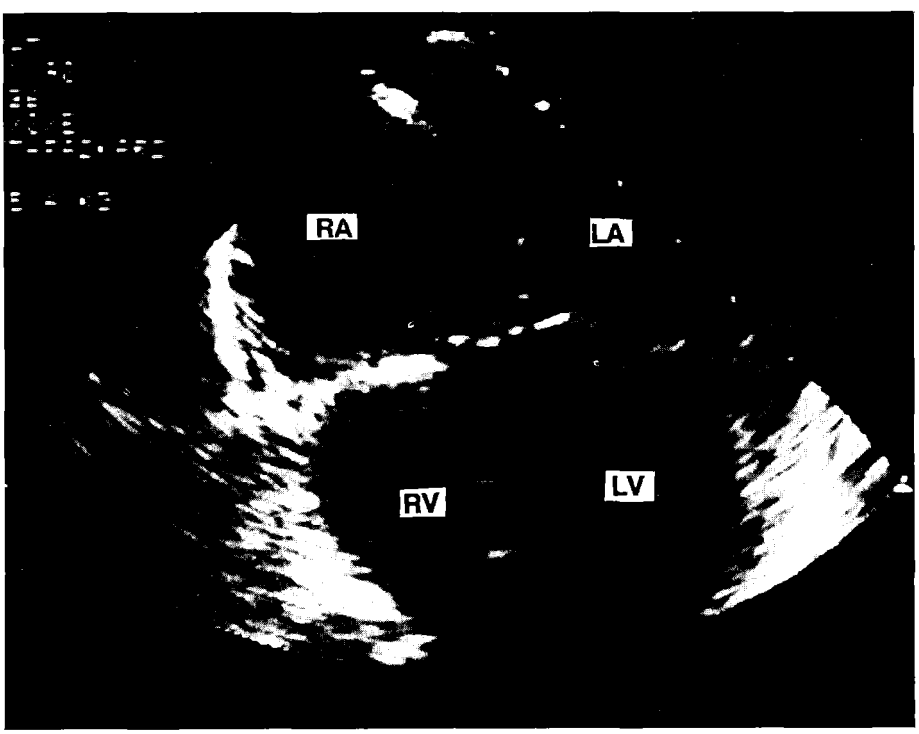


Figure 2: Transoesophageal four chamber view in a patient with atrial situs solitus, left juxtaposition of the atrial appendages, concordant atrioventricular connections and discordant ventriculo-arterial connections. The posterior portion of the atrial septum is typically deviated to a more frontal plane in this standard view. Note the hypoplastic tricuspid valve and the right ventricular cavity. The interatrial septum is intact on this view.
Legend: LV = left ventricle, other abbreviations see figure 1

Ventriculo-arterial junction Three patients were demonstrated to have discordant ventriculo-arterial connections and one patient double outlet right ventricle. Evidence of a bilateral infundibulum, preventing fibrous continuity between either atrioventricular and arterial valves, was documented in three of these patients (figure 3). The abnormal muscle bundle contributed to subpulmonary obstruction in two patients with almost side-by-side relationship of the arterial trunks (aorta to the right and slightly in front of the pulmonary trunk). There was overriding of the pulmonary valve in both cases.

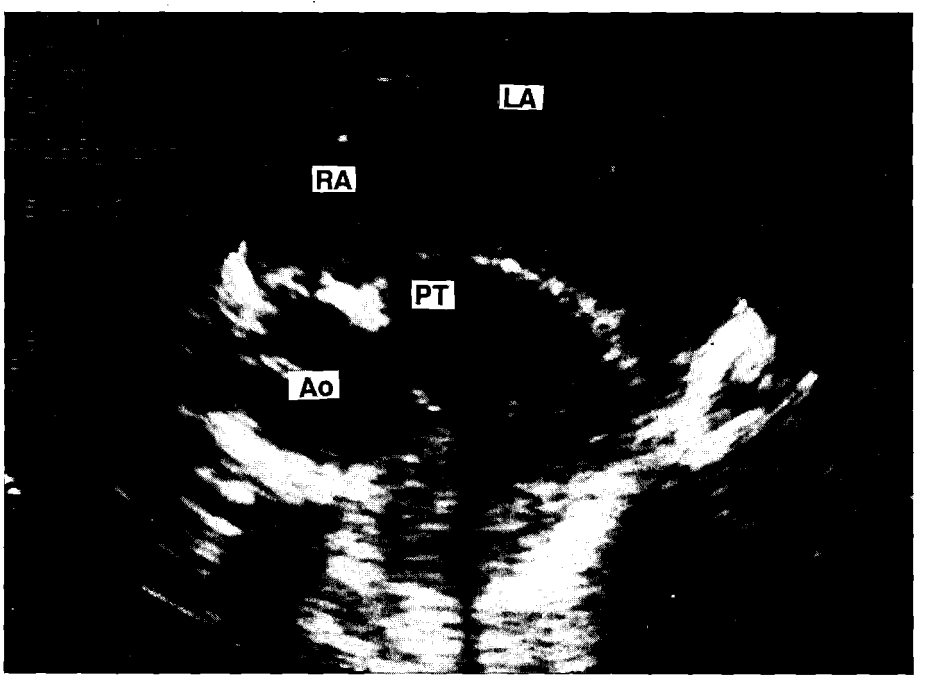


Figure 3: Bilateral infundibulum contributing to subpulmonary obstruction (same patient as in figure 2). Note the relationship of the arterial trunks with the aorta being to the right and slightly anterior of the pulmonary trunk.
Legend: Ao = aortic valve, other abbreviations as in figure 1 and 2

Discussion

Juxtaposition of the atrial appendages is a rare entity, which is associated with complex cyanotic congenital heart disease (1-3). It is most frequently encountered in cases with discordant ventriculo-arterial connections, which are either isolated (as in complete transposition of the great arteries) or are associated with an absent right atrioventricular connection (4). Left juxtaposition of the atrial appendages is far more common than right juxtaposition and can coexist with all possible atrial arrangements (16,17). Although this congenital abnormality is of no immediate haemodynamic relevance, its presence can influence the surgical approach in cases were correction is being considered.

Of the diagnostic techniques previously employed to define juxtaposition of the atrial appendages preoperatively, only praecordial cross-sectional imaging and angiocardiography

have proved to be of value. The direct determination of juxtaposed atrial appendages by angiocardiography is largely dependent on whether a sufficient contrast effect is achieved in both the atrial cavity and the juxtaposed appendage. In addition, the abnormality must be specifically looked for. Although the praecordial cross-sectional echocardiographic criteria of left juxtaposition have been described in detail by Rice et al. (10), the recognition may be difficult - not only in older children or in those with poor praecordial windows. In addition, the morphologic characteristics of either atrial appendage routinely cannot be delineated by praecordial imaging.

Transoesophageal echocardiography, using dedicated paediatric probes, was performed prospectively in a series of children with unoperated complex congenital heart disease. The results of this preliminary study suggested that the use of this new imaging modality is of additional value in the preoperative diagnosis of complex lesions. The assessment of lesions involving the venous connections to the heart, the atrial chambers and the atrioventricular junctions were largely enhanced by this technique. Thus, not surprisingly, juxtaposition of the atrial appendages is readily appreciated on transoesophageal cross-sectional imaging, regardless of the complexity of associated lesions. The transoesophageal features of left juxtaposition of the atrial appendages include 1. a right lateral deviation of the inferior and posterior portion of the atrial septum and 2. a more frontal orientation of the antero-superior portion of the atrial septum forming the floor and the posterior wall of the junction of the right-sided atrial appendage with the venous component of the atrial cavity. Since transoesophageal echocardiography is increasingly often being used in the assessment of adolescents and adults with congenital heart disease, these findings are also of relevance to the adult cardiologist, who is not entirely familiar with complex congenital heart disease. The abnormal orientation of the atrial septum may otherwise give the false impression of an atrial septal defect, located at the site of the junction of the atrial appendage with the venous cavity.

The determination of the atrial appendage morphology is not compromised by this altered appendage and septal orientation. Although not encountered in this series, right juxtaposition of the atrial appendages, or juxtaposition in the setting of ambiguous atrial arrangements (atrial isomerism), should, in view of the great morphologic detail that is obtained, also be reliably documented on transoesophageal echocardiographic studies. In addition, the assessment of the atrial septal integrity by means of colour flow mapping studies is not limited by the abnormal septal orientation - multiple septal defects can reliably be excluded. The association of left juxtaposition of the atrial appendages with subpulmonary outflow obstruction is well documented (4). In our patient population, three patients were documented to have evidence of a bilateral infundibulum, contributing to subpulmonary obstruction in two. The finding of an almost side-by-side arrangement of the arterial trunks in these two patients suggests that this situation may be a co-factor to obstruction.

The results of this study indicate that transoesophageal echocardiography may be expected to be the ultimate preoperative diagnostic technique for the definition or exclusion of juxtaposition of the atrial appendages.

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Chapter 6

The assessment of anomalies of systemic and pulmonary venous drainage by pediatric transesophageal echocardiography

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(submitted for publication)

Summary Pediatric transesophageal echocardiographic studies were prospectively performed in seventy-six consecutive unoperated children (age 0.2 - 14.8 years, mean age 4.1 years) with congenital heart disease in order to determine the value of this new diagnostic modality in the preoperative assessment of either systemic or pulmonary venous abnormalities. The results obtained were correlated with prior precordial ultrasound studies (n=76), cardiac catheterization (n=62) and surgical inspection (n=47).

Fourteen patients were documented to have anomalous venous connections, two had both systemic and pulmonary venous abnormalities. Anomalous systemic venous connections were identified in nine patients by transesophageal studies. This was confirmed by cardiac catheterization or surgical inspection in eight; one patient awaits surgery. Six of these were missed during prior precordial studies. Seven had anomalous pulmonary venous connections (1 mixed total, 6 partial), all of which were correctly diagnosed by transesophageal studies. Prior precordial studies missed anomalous pulmonary venous connections in four of these cases. No other patient was found by angiocardiography or surgical inspection to have anomalous venous connections. Thus, in this series, the sensitivity of transesophageal echocardiography to detect anomalous systemic or pulmonary venous drainage was 100 %.

Transesophageal echocardiography is a highly sensitive tool in the preoperative definition of systemic or pulmonary venous connections. It is superior to precordial ultrasound studies and can contribute additional relevant information to cardiac catheterization and angiocardiography.

Introduction

The spectrum of anomalies of either pulmonary or systemic venous connections to the heart is extremely wide, both in their clinical presentation as well as in their underlying

morphology (1,2). Whereas total anomalous pulmonary venous drainage in the critically ill neonate is usually readily diagnosed by precordial ultrasound techniques (3-5), other forms of anomalous venous connections may be difficult to diagnose and in the majority of cases cannot be definitively ruled out (6,7). Because of the important implications such anomalies may have for the surgical approach (8), the patterns of the venous connections to the heart have to be defined prior to surgical correction. Cardiac catheterization with angiocardiography is still considered by many to represent the ultimate diagnostic test. However, diagnostic difficulties may persist following this investigation.

Transesophageal echocardiography is a relatively new adjunct to the ultrasound evaluation of children with congenital heart disease (9-11). During a prospective study protocol into the value of transesophageal echocardiography in the diagnosis of congenital heart disease, seventy-six consecutive unoperated children were studied by pediatric single-plane transesophageal ultrasound. The information derived from these studies on the connections of either systemic or pulmonary venous return was correlated with that derived from prior precordial ultrasound studies, and with the findings obtained by cardiac catheterization or by surgical inspection, in order to determine whether transesophageal echocardiography may be a useful diagnostic adjunct in the preoperative diagnosis or exclusion of anomalies of the venous connections to the heart.

Patients and Methods

Patients Seventy-six unoperated children were studied. Approval by the Hospital Ethical Committees of the Academic Hospital Rotterdam and the National Institute of Cardiology, Mexico City, was obtained prior to commencement of the study and prior informed parental consent was obtained for the transesophageal study in each case. The age at investigation ranged from 2.5 months to 14.8 years with a mean age of 4.1 years. The body weight ranged from 3.7 to 48 kilograms with a mean weight of 13.9 kilograms.

Scanning equipment An initial seventeen patients were studied using an Aloka 24 element pediatric transesophageal probe connected to an Aloka SSD 870 ultrasound system. Subsequently, fifty-nine patients were studied with a prototype forty-eight element pediatric transesophageal probe (Department of Experimental Echocardiography, Thoraxcenter Rotterdam) connected to either a Hewlett Packard Sonos 1000 or 500, or a Toshiba SSH 160 A or 140 A ultrasound system. Both phased array transducers allowed 5 MHz cross-sectional imaging in the transverse-axis plane, color flow mapping and pulsed wave Doppler interrogation. The maximal tip circumference of either probe measured 30 mm (7x8 mm and 5x10 mm respectively); the maximal shaft diameter 7 mm. Probe tip manipulation was restricted to anterior/posterior angulation.

Technique Of the total of 76 children, 64 were studied under general anesthesia which was given for scheduled cardiac catheterization (n = 46), or in the immediate preoperative period prior to surgical correction (n = 18). The remaining 12 patients were studied electively by transesophageal ultrasound on a day care basis under general anesthesia. All transesophageal studies were performed without knowledge of the results obtained at cardiac catheterization and angiocardiography.

After sufficient lubrification with anesthetic gel, the transesophageal probe was inserted

either blindly or under direct laryngoscopic vision into the hypopharynx and then was advanced into the lower third of the esophagus. No additional antibiotic prophylaxis was administered for the study. A standardized scheme of investigation (to include cross-sectional imaging and color flow mapping studies) to assess the atrial arrangement, the atrio-ventricular and ventriculo-arterial junction and the intracardiac morphology, using standard transverse axis scan planes (12,13) was first completed in every patient. Determination of the atrial arrangement was performed by direct visualization of both atrial appendages, using the criteria previously described by our group (14). Thereafter the morphology of the systemic venous and the pulmonary venous connections was thoroughly examined using the techniques described below.

Assessment of systemic venous connections The morphology of the inferior caval vein and the hepatic veins was assessed by using transgastric views of the liver. A complete scan of the upper abdomen was performed by rotating the probe in a clockwise fashion, in order to determine the relationship of the infradiaphragmatic great vessels one to another and to the spine. Thereafter the probe was gradually withdrawn in order to demonstrate the presence of any venous valve within the atrium (figure 1c), the roof of the right sided atrium and the connection with the superior caval vein. The superior caval vein could be routinely demonstrated anterior to the right pulmonary artery. A more cranial segment of this vessel was demonstrated by withdrawing the probe further within the esophagus above the right main bronchus. The innominate vein was searched for anterior of the aortic arch. The coronary sinus was demonstrated by scanning low transesophageal four chamber views.

The existence of a left persistent superior caval vein was documented by using high left atrial views, and views of the left pulmonary artery and the left sided mediastinum. An azygos vein was searched for by combined cross-sectional imaging, color flow mapping and pulsed wave Doppler by sampling posterior and to the right of the right atrium and the right pulmonary artery. A hemiazygos vein was looked for posterior to the left atrium and next to the descending aorta.

Assessment of pulmonary venous connections The right upper pulmonary vein was visualized when the probe was slightly advanced from the position used to scan the superior caval vein and the right pulmonary artery (figure 1a). Variation of transducer tip angulation allowed improved imaging in cases where this was initially difficult. After demonstration of the right upper pulmonary vein, the right lower pulmonary vein was searched for by further introduction of the probe and slight clockwise rotation, as this vessel normally drains into the left atrium in a more posterior-anterior direction (figure 1b,c). The left upper pulmonary vein was readily demonstrated after visualization of the left sided atrial appendage. This vein lies posterior and to the left of the left atrial appendage, and is separated from it only by a crest of atrial tissue (figure 1a). Further slight advancement and anticlockwise rotation of the probe allowed the visualization of the left lower pulmonary vein (figure 1b). After the detection of the individual pulmonary veins the scan position was varied in order to demonstrate the exact course and the site of connection of these vessels to the atrial chambers relative to the atrial septum.

Color flow mapping was used for the rapid identification of venous connections to the atria. In addition, pulsed wave Doppler sampling was used for the unequivocal identification of venous flow, and for the exclusion of obstruction to venous return.

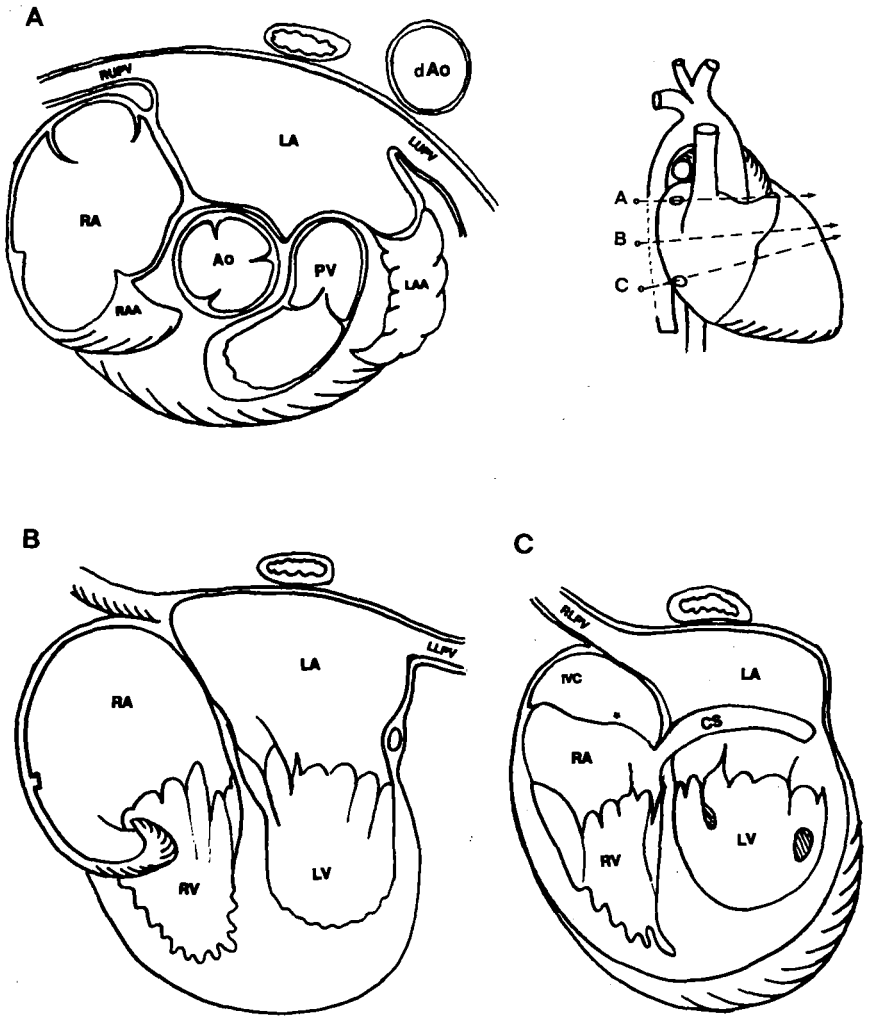

Figure 1: Diagram of standard transesophageal views used in the assessment of both systemic and pulmonary venous connections and atrial morphology. a. basal short axis cut at the level of the arterial valves, b. standard four chamber view, c. low esophageal view at the level of the hiatus.

Legend: * = Eustachian valve; Ao = aorta, CS = coronary sinus; dAO = descending aorta; IVC = inferior vena cava; LA = left atrium; LAA = left atrial appendage; LLPV = left lower pulmonary vein; LUPV = left upper pulmonary vein; LV = left ventricle; PV = pulmonary valve; RA = right atrium; RAA = right atrial appendage;RLPV = right lower pulmonary vein; RUPV = right upper pulmonary vein; RV = right ventricle.

Precordial studies All patients were studied by a complete precordial ultrasound examination using the full range of parasternal, subcostal and suprasternal views. Cross-sectional imaging, together with color flow mapping and pulsed wave Doppler studies were completed in all patients.

Cardiac catheterization Sixty-two children had correlative cardiac catheterization and angiocardiography. Blood samples for oximetry were taken at frequent sites including the high superior caval vein, the inferior caval vein, the right atrium and the right ventricle. Angiocardiography was performed using biplane equipment and included pulmonary artery injection in all children.

Evaluation The entire transesophageal studies were continuously recorded onto video tape and were analyzed both on-line and later off-line at reduced speed by two independent observers. Thereafter the results of the transesophageal studies were correlated with the findings obtained by reviewing of prior precordial ultrasound studies (n =76), with the results obtained by either simultaneous or recent cardiac catheterization (n = 62), and with the findings during subsequent surgical inspection (n = 47). Photographs were taken later as still-frames from the video monitor.

Results

Systemic venous connection The individual patterns of systemic venous connections were readily demonstrated by the transesophageal studies in all 76 patients studied using the views described above. Systemic venous return was documented by transesophageal studies to be normal in 67 children.

Seven patients were found to have a left persistent superior caval vein (table). The course of the vessel (anterior to the left pulmonary artery and interposed between the left sided pulmonary veins and the left sided atrial appendage) could be documented in all patients using combined cross-sectional imaging and colour flow mapping. The left persistent superior caval vein drained into the roof of the left sided atrium (figure 2) in three patients (No 3,8,11). None of these patients were identified on prior precordial ultrasound studies. Cardiac catheterization confirmed this finding in two, and was incomplete in the third patient, who had later surgical confirmation. In the remaining four patients (No 2,5,12,14) the left superior caval vein connected to the coronary sinus (figure 3), which was appreciated during prior precordial studies in three patients. Transesophageal studies allowed the documentation of an innominate vein (bridging vein) in 4 patients with a left persistent superior caval vein, precordial studies in six.

Anomalies of the right superior caval vein were documented by the transesophageal study in two patients. One child with atrial situs solitus (No 12) was detected to have an interrupted right superior caval vein. The persistent left superior caval vein was documented to drain to the coronary sinus. The precordial study remained uncertain about the presence of a right superior caval vein, which was definitively excluded during subsequent cardiac catheterization. In the second patient, who had right atrial isomerism (No 13), the right sided superior caval vein was demonstrated to be connected to the left side of the remnant of the atrial septum (figure 4). Neither precordial ultrasound studies nor angiocardiography delineated this abnormality. This finding awaits surgical confirmation.

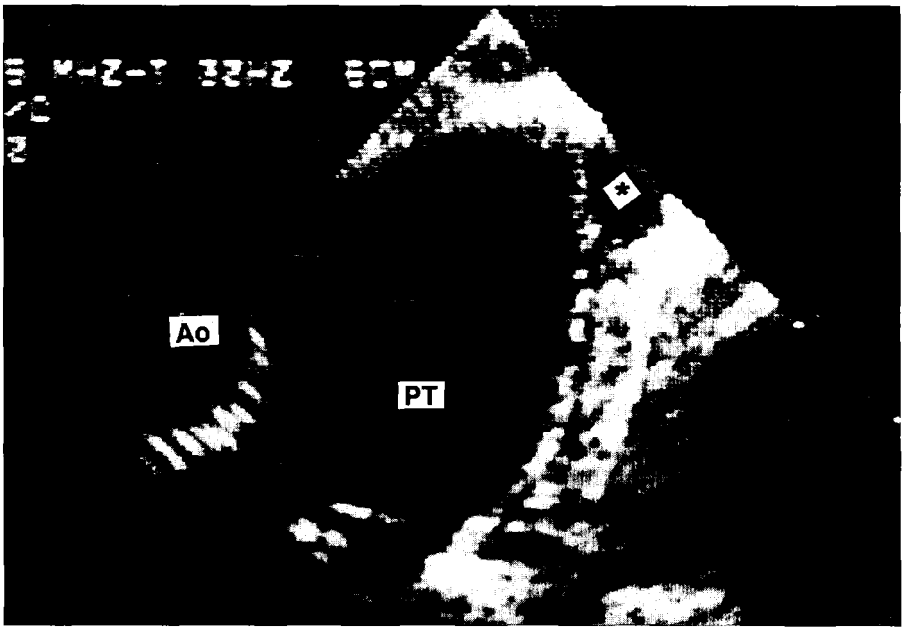

Figure 2: Left persistent superior caval vein draining into the roof of the left sided atrium in a patient with left atrial isomerism (No 11). The vessel (asterix) is seen to the left of the (dilated) pulmonary trunk near its bifurcation.
Legend: Ao = ascending aorta; PT = pulmonary trunk

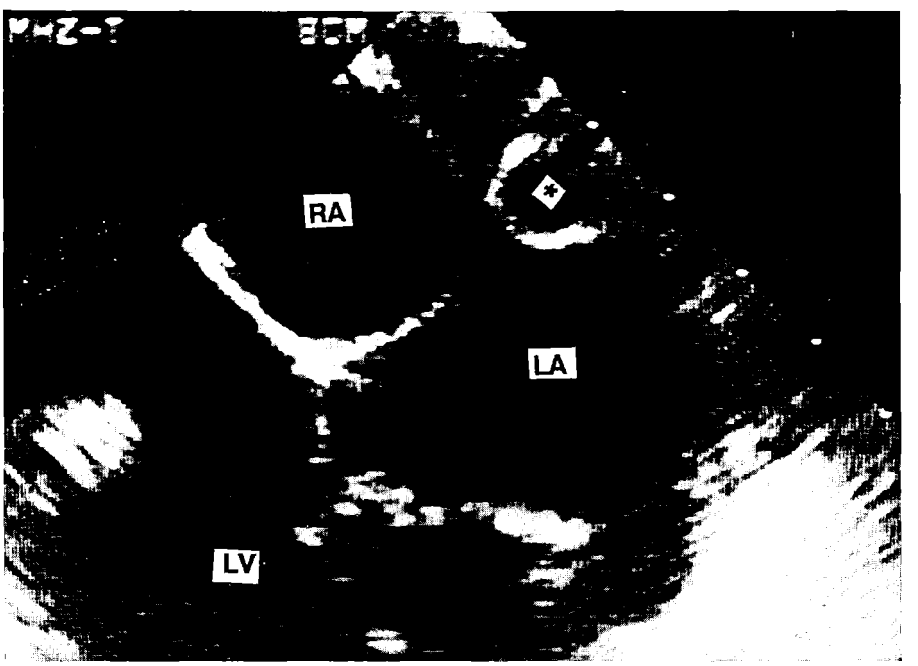

Figure 3: Demonstration of a persistent left superior caval vein which connected to the coronary sinus (asterix) in a patient with dextrocardia, atrial situs solitus and absent right atrioventricular connection (no 14).
Legend: LA = left atrium; LV = left ventricle; RA = right atrium.

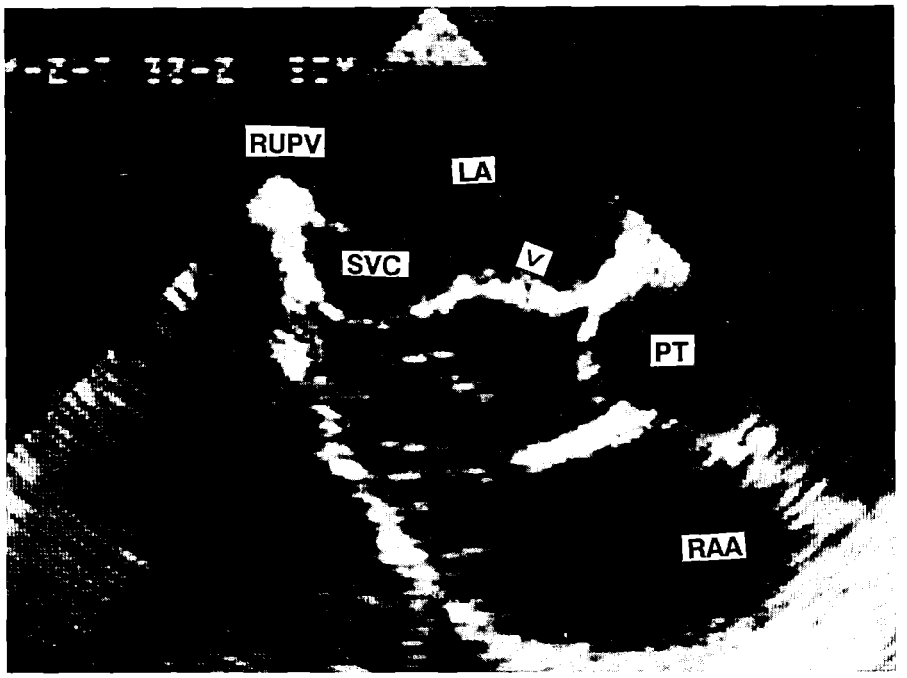

Figure 4: Documentation of a right superior caval vein which entered the left sided atrium in a patient with right atrial isomerism (no 13). The crest of atrial tissue of the cavo-atrial junction is in continuity with the remnant of the atrial septum (arrow).
Legend: LA = left sided atrium; PT = pulmonary trunk; RAA = right atrial appendage; RUPV = right upper pulmonary vein; SVC = superior caval vein.

The morphology and connection of the inferior caval vein (interruption in 3 patients) were equally well documented by precordial and transesophageal studies. An azygos vein was correctly identified in three patients with interrupted inferior caval vein. Two of these patients were documented to have left atrial isomerism (No 8,11), the third patient atrial situs solitus (No 4). The pattern of hepatic venous drainage in four patients with atrial isomerism (3 individual, 1 common) was better documented using the transoesophageal technique in one patient with individual drainage.

Pulmonary venous connection The site of drainage of all four pulmonary veins could be documented by either cross-sectional transesophageal imaging or color flow mapping in 69 of the 76 patients studied (91 %). Both the right upper and left upper pulmonary veins could be identified in all patients, the left lower pulmonary vein in 74 patients (97 %) and the right lower in 70 patients (92 %) studied.

Pulsed wave Doppler sampling in individual pulmonary veins revealed mild pulmonary venous obstruction in three patients who demonstrated continuous turbulent flow patterns with maximal velocities of more than 1.2 m/s. None of these patients were identified during the prior precordial studies. Pulmonary wedge pressures of the corresponding pulmonary artery branch were obtained in two of these patients, and reflected mild elevation in one.

Six children were documented by the transesophageal study to have partial anomalous pulmonary venous drainage (Table). Two had partial anomalous connection of the right

TABLE: Anomalous systemic and pulmonary venous connections - Patient data and diagnosis.

No	age yrs.	sex	weight kg	Diagnosis Situs	AV junction	VA junction	Systemic venous connections	Pulmonary venous connections
1	13.7	m	40.2	solitus	absent right	concordant	normal	RUPV to r SVC
2	14.8	f	42.5	solitus	concordant	concordant	l SVC to CS	RUPV to r SVC
3	7.7	f	20.0	RAI	ambiguous	DORV	l SVC to left sided atrium individual HV drainage	all four PV to left sided atrium
4	2.0	f	9.8	solitus	concordant	concordant	interrupted IVC azyos continuation	normal
5	0.7	m	4.7	solitus	concordant	concordant	l SVC to unroofed CS	normal
6	4.7	m	19.5	solitus	concordant	concordant	normal	RUPV to RA
7	0.2	m	5.1	solitus	concordant	concordant	normal	RLPV to RA
8	11.0	f	23.2	LAI	ambiguous	DORV	interrupted IVC hemiazygos continuation bilateral SVC absent coronary sinus	all PV to right sided atrium
9	5.0	f	15.4	solitus	concordant	concordant	normal	RUPV to RA
10	0.6	f	4.3	solitus	concordant	concordant	normal	three PV to CS LUPV to innominate vein
11	11.0	f	37.2	LAI	ambiguous	concordant	interrupted IVC azygos continuation bilateral SVC absent coronary sinus	all four PV to left sided atrium
12	1.5	f	6.2	solitus	concordant	concordant	absent r SVC l SVC to CS	three PV to left sided atrium LUPV to l SVC
13	5.8	m	15.8	RAI	absent right	discordant	r SVC to left sided atrium l IVC,individual HV drainage	three PV to left sided atrium RLPV not identified
14	1.6	m	8.1	solitus	absent right	DORV	l SVC to CS	normal

Legend:CS = coronary sinus; DORV = double outlet right ventricle; f = female; HV = hepatic veins; IVC = inferior vena cava, kg = kilograms; l = left; LAI = left atrial isomerism; LUPV = left upper pulmonary vein; PV = pulmonary vein; r = right; RA = right atrium; RLPV = right lower pulmonary vein; RUPV = right upper pulmonary vein; SVC = superior vena cava; TAPVC = total anomalous pulmonary venous connection.

upper pulmonary vein to the superior caval vein (No 1,2). The site of drainage was correctly identified in one patient (figure 5). In the second patient the site of drainage could not be demonstrated due to interposition of the right bronchus. Partial anomalous connection of the right upper pulmonary vein to the superior caval vein was suggested by precordial studies in only one patient (No 2).

In two patients (No 6,9) the right upper pulmonary vein (figure 5), and in one patient (No 7) the right lower pulmonary vein, were documented during the transesophageal study to be connected to the right sided atrium. All three had a sinus venosus atrial septal defect. Additional defects in the secundum septum were documented in two of these patients (No 7,9), in whom prior precordial studies did not define partial anomalous pulmonary venous drainage. Anomalous connection of the left upper pulmonary vein to a left persistent superior caval vein was documented by the transesophageal study in one patient (No 12), in whom the prior precordial study suggested normal venous drainage. Cardiac catheterization, angiocardiography or surgical inspection confirmed partial anomalous pulmonary venous drainage in all 6 patients, and excluded it in the remaining patients.

Figure 5: Transesophageal visualization of an anomalous connection of the right upper pulmonary vein to the superior caval vein (arrow), near the cavo-atrial junction. Both vessels lie in front of the right pulmonary artery.
Legend: Ao = ascending aorta; PT = pulmonary trunk; RPA = right pulmonary artery; SVC = superior caval vein

One child with total anomalous pulmonary venous connection was found to have a mixed pattern of drainage (No 10). Three pulmonary veins drained to the coronary sinus; the left upper pulmonary vein drained via a vertical vein to the innominate vein. The demonstration of turbulent flow patterns in this vessel during the prior suprasternal study led to initial misinterpretation of this finding as being total anomalous supracardiac pulmonary venous connection. On subsequent review of the precordial study mixed total anomalous drainage was suggested, however the individual site of drainage of all four pulmonary veins was not demonstrated. The transesophageal finding was confirmed by subsequent cardiac catheterization and surgical inspection.

Discussion

The documentation of both the systemic and pulmonary venous connections to the heart is an integral part in the diagnosis of congenital heart disease. It is of essential value in every patient being considered for total surgical correction.

Precordial ultrasound has been proven to be a sensitive tool in detecting anomalous venous connections (4,6). However, in older children or in those with complex cardiac malformations the precordial approach may yield incomplete information. Problem areas include the detection of a left persistent caval vein draining into the roof of the left sided atrium, the definition of partial anomalous pulmonary venous connections, or the differentiation of total from mixed total anomalous pulmonary venous connection and the subsequent determination of the exact site of connection. In addition, the site of connections of all four pulmonary veins can routinely not be visualized in children beyond infancy. Pulsed wave Doppler interrogation of individual pulmonary vein flow patterns, which potentially allows for the detection of pulmonary vein obstruction (15,16) is problematic in the older child.

Angiocardiography, which still is regarded as being the gold standard in determining the venous connections to the heart, can identify the presence of anomalous venous connections in the majority of cases. However, since the angiocardiographic demonstration of an anomalous pulmonary venous connection is most often performed by means of pulmonary artery contrast injection, the identification of any abnormality of pulmonary venous drainage can be hampered by the inherent dye dilution. This is particularly true of cases with mild obstruction of the abnormal connection. Subtle morphologic details, such as the relationship of the site of venous connections to the interatrial septum, in patients with sinus venosus defects or complex congenital heart disease, are frequently difficult to appreciate using angiocardiography.

Transesophageal echocardiography is a relatively recent imaging modality in congenital heart disease, which, with the availability of dedicated pediatric transesophageal transducers, has become a feasible and safe technique even in small children (10,11). The proximity of the transducer to the atrial chambers and the sites of venous return potentially allows a better evaluation of these structures. During a prospective study protocol, we have evaluated the value of this technique in the determination of the morphology of both systemic and pulmonary venous connections. With increasing experience patients with complex congenital heart disease involving the atrial or atrioventricular levels of the heart were selected.

Systemic venous connections A left persistent superior caval vein draining to the

coronary sinus could be easily identified on both cross-sectional imaging and color flow mapping. Such vessels were always found interposed between the left sided atrial appendage and the left sided pulmonary veins. Their course could then be followed cranially lying anterior to the left pulmonary artery by slight changes in the level of probe insertion. Drainage of a left superior caval vein to the roof of the left sided atrium was also readily diagnosed. The technique proved of great value in documenting very subtle morphologic details of the relationship of the systemic venous connections to the interatrial septum, which could not be derived from either precordial or angiocardiographic studies. Limitations in the technique include the difficulties in documentation of a bridging vein by single-plane transesophageal imaging, this is more reliably performed using suprasternal scan positions. Azygos and hemiazygos veins could be demonstrated only in cases where these vessels were enlarged. The definition of inferior caval vein or hepatic venous connections was not enhanced by transesophageal studies when compared to precordial studies.

Transesophageal studies defined anomalous systemic venous connections in nine patients and excluded these in the remaining patients. None of these remaining sixty-seven patients was documented by any other investigative technique to have anomalies of the systemic venous connections. Thus, in this series, the sensitivity of transesophageal echocardiography in detecting anomalous systemic venous connections was 100 %. Eight of nine children with anomalies of the systemic venous connections had subsequent surgical or angiocardiographic confirmation of the findings, reflecting the high specifity of the technique. One patient awaits surgical confirmation.

Pulmonary venous connections Pulsed wave Doppler sampling of individual pulmonary vein flow patterns allows the rapid exclusion of pulmonary vein stenosis. Transesophageal echocardiography proved to be more sensitive than precordial ultrasound and cardiac catheterization in detecting even minor degrees of venous obstruction. The site of connection of all four individual pulmonary veins could be demonstrated in the vast majority of cases (91 %). Only the connection of the right lower pulmonary vein remains difficult to demonstrate. This is related to the rather posterior/anterior course of this vessel far on the right lateral aspect of the heart. However, with increasing experience with this technique during the study the detection of this vessel became less problematic. Partial anomalous venous connections should be assumed in every case where the pulmonary veins cannot be demonstrated at their normal position during the transesophageal study. Subsequent scanning of the adjacent structures, using cross-sectional imaging and color flow mapping, will then delineate the exact site of drainage in the majority of patients. Problems persists only in cases where the site of connection is at the level of the bronchi.

Seven of the seventy-six children studied were documented to have anomalous pulmonary venous connections. The findings were confirmed by cardiac catheterization or surgical inspection in all of these, and were excluded in the remaining sixty-nine patients. Thus, in this series, the sensitivity of transesophageal ultrasound in detecting anomalous pulmonary venous connections was 100 %.

Limitations Interposition of the trachea and the main bronchi precludes the visualization of a distal segment of the superior caval vein, and limits the definition of anomalous pulmonary venous connection in this area. The near-field artefact of the first generation of pediatric transesophageal probes is now much reduced, and the initial problems to identify

all four pulmonary veins in even small children by color flow mapping studies no longer exist. Single-plane, transverse axis imaging of the caval veins produces short axis cuts. Thus, in order to fully appreciate the course and hence the connection of these vessels, the level of probe insertion has to be repeatedly changed. In this respect longitudinal plane, or biplane imaging may be expected to be of substantially additional value (17), however it will contribute only little in the assessment of the pulmonary venous connections.

Potential indications Since transesophageal echocardiographic studies can be performed in children only under general anesthesia or heavy sedation, indications for elective studies should be strict. They can be performed safely during simultaneous cardiac catheterization or immediately prior to surgical correction without requiring additional procedure or anesthesia time. The results of this study suggest that such studies may be indicated in the child with complex congenital heart disease, those with abnormalities of the atrial arrangement or in older children with poor precordial ultrasound windows, in whom anomalous venous connections to the heart are either suspected or their existence would alter the surgical approach.

Conclusion Transesophageal echocardiographic studies would appear to be a most valuable adjunct in the preoperative diagnosis of anomalous venous connections to the heart. The sensitivity of this new imaging modality, in this series, was 'largely superior to precordial ultrasound studies, and was comparable to angiocardiography. In addition, it can contribute relevant morphologic and hemodynamic details not available from any other diagnostic technique.

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Chapter 7

The comparative value of surface and transesophageal ultrasound in the assessment of congenital abnormalities of the atrioventricular junction

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Summary The information obtained from surface and transesophageal ultrasound (two - dimensional imaging, spectral Doppler and color flow imaging) was compared in 32 patients with major congenital abnormalities of the atrioventricular junction (10 discordant atrioventricular connections, 1 criss-cross connection, 6 double inlet ventricles, 12 absent right and 3 absent left connection). The findings obtained by either modality were correlated with findings at cardiac catheterization (27 patients) and at surgical inspection (11 patients). In 2/15 patients with an absent atrioventricular connection, as defined by surface echocardiography, transesophageal imaging demonstrated an imperforate atrioventricular valve, in 1 a common atrioventricular valve was documented. In 11/11 patients with discordant or criss-cross connections, assessment of atrioventricular valve and ventricular morphology (by defining the chordal attachments of both AV valves) was possible with transesophageal echocardiography (3/11 patients by surface echocardiography); chordal straddling was detected in two patients and excluded in three others with an associated inlet ventricular septal defect. Anomalous venous connections (4 patients), atrial septal defects (19 patients), and subpulmonary stenosis (11 patients) were better assessed by transesophageal imaging. Atrial appendage morphology could be demonstrated in all. The technique was less useful in demonstrating the anterior subaortic infundibulum, or aortopulmonary shunts.

Transesophageal echocardiography is an important new adjunct in the preoperative evaluation of major congenital abnormalities of the atrioventricular junction. It provides unique morphologic information frequently not obtainable by either surface echocardiographic studies or angiocardiography.

Introduction

Transthoracic cross sectional echocardiography has been shown to be a sensitive investigative technique in the definition of morphologic abnormalities of the atrioventricular

junction, particularly in the younger patient (1-3). High resolution cross sectional imaging however, is a relatively recent development, and a significant proportion of older children, adolescent and adult patients with such abnormalities diagnosed in early life have not had adequate surface echocardiographic studies in infancy. In addition, some patients, particularly those with discordant or criss-cross AV connections may present for the first time in later childhood or adult life. In such cases, surface echocardiography may not provide detailed morphologic information, and alternative investigative approaches may be required both for planning surgery, and for long term follow-up.

Transesophageal echocardiography is now an established investigative technique for acquired cardiac disease in the adult outpatient clinic. In older patients it has been shown to be an excellent technique for the demonstration of congenital atrial morphologic abnormalities and assessing atrioventricular valve function (4-6). The improved image quality, particularly at the level of the atria and AV junction, should make it a useful technique in the assessment and serial follow-up of congenital lesions affecting these regions of the heart. Thus, we have attempted to define the role of transesophageal ultrasound in the assessment of congenital malformations of the AV junction and of the commonly coexisting structural and functional abnormalities in a wide age range of patients.

Patients and methods

Transesophageal echocardiographic studies were performed in 32 patients, (age range 0.2 to 67 years; mean age 14.5 years) who were either referred for the first time for cardiac evaluation (9 patients) or were being followed up (23 patients) with a known congenital morphologic abnormality of the AV junction. A primary diagnosis of AV discordance in ten patients, double inlet ventricle in six patients, criss-cross AV connection in one patient, absent right connection in twelve patients, and absent left connection in three patients had been made by a combination of prior surface echocardiography and angiography. Sixteen patients had previous cardiac surgery: corrective surgery in three cases and palliative surgery in thirteen patients. Twenty-two patients (including all with previous palliative procedures) were assessed with a view to definitive surgical repair, and two others with systemic ventricular failure were evaluated prior to cardiac transplantation.

All patients underwent standard surface echocardiography during the period of review, using Vingmed 770 or Toshiba SSH 160A or Hewlett Packard Sonos 500 or 1000 ultrasound systems. All available scan planes (precordial, subcostal and suprasternal) were utilized, and standard two-dimensional imaging, pulsed and continuous wave Doppler and color flow imaging studies were performed.

Transesophageal two-dimensional imaging was performed in all, using a Toshiba SSH 160A, an Aloka SSD 870, or a Hewlett Packard Sonos 500 or 1000 ultrasound system. Adult patients were studied using standard 5 or 5.6 mHz transesophageal probes. Seventeen children were studied using either an Aloka pediatric transesophageal probe (24 element, tip dimensions 7x8 mm), or a prototype 48 element pediatric probe (tip dimensions 5x10 mm; Department of Experimental Echocardiography Thoraxcenter Rotterdam). All studies included cross-sectional imaging combined to color flow mapping and selected pulsed Doppler examinations. Prior ethical approval was obtained, and informed patient's or parental consent was given prior to individual transesophageal studies.

Studies were performed on an outpatient basis in 14 patients. For outpatient studies, no patient required prior sedation, and apart from local anesthetic spray (lidocaine 10%) to the hypopharynx no other pre-medication was given. In the remaining 18 patients, transesophageal studies were performed under general anesthesia given in the perioperative period, for cardiac catheterization, or for elective transesophageal studies in eight children. Routine antibiotic prophylaxis was not given, as is our general practice (7). In every case, a standard study protocol was used to obtain the transesophageal echocardiographic images (8).

Transesophageal studies The transesophageal examination commenced with a transgastric short axis section at the level of the ventricles to assess ventricular morphology and function. Withdrawal of the probe to the lower esophagus and clockwise rotation demonstrated the hepatic venous confluence and the inferior vena cava, which could be then followed to the right atrium in a series of short axis sections. Modified long axis and short axis views at the level of the atrioventricular junction were performed to assess AV valve morphology, the chordal attachments of both AV valves, and the area of fibrous continuity between the AV valve and the posterior great vessel. Further withdrawal and clockwise rotation enabled identification of the right sided atrial appendage. From this position, with serial anti-clockwise rotation the atrial septum, left atrium and the left atrial appendage could be scanned in the short axis.

The left upper pulmonary vein could be visualized laterally and posteriorly to the left atrial appendage, and separated from it by a muscular ridge. Pulsed Doppler sampling of pulmonary venous flow patterns was performed in all. At a slightly higher level, the proximal portions of the aorta and pulmonary artery, including the pulmonary artery bifurcation could be seen. Clockwise rotation demonstrated the junction of the superior vena cava with the (right) atrium, and the right pulmonary artery. A search was made for the drainage sites of the other individual pulmonary veins. Similar scan planes could be obtained in patients with abnormal positions of the heart within the chest. All scans were continuously recorded on videotape to enable subsequent off-line analysis.

Analysis Sequential off-line analysis of the surface and transesophageal echocardiographic studies was performed by the same three observers, all of whom were aware of the prior cardiac diagnosis. Twenty-seven patients had cardiac catheterization and angiography during this reassessment, and subsequent verification of the echocardiographic findings at surgery was possible in eleven cases.

Results

Cardiac position and atrial situs Six patients had dextrocardia, 1 had mesocardia, and the remaining 25 had levocardia. Using surface echocardiography, and from the position and relations of the infradiaphragmatic portions of the aorta and inferior vena cava (9), usual atrial arrangement (situs solitus) was inferred in 25 patients, and inverted atrial arrangement in three patients. Left atrial isomerism was suggested in two patients, and right atrial isomerism in one. In a further patient, the abdominal vessels or pulmonary veins could not be identified with sufficient clarity to enable the diagnosis of atrial situs.

In all 32 patients, transesophageal echocardiography enabled direct visualization of both atrial appendages, and thus the direct diagnosis of atrial situs (10). Situs solitus was documented in 27 patients, of whom 2 were defined to have left juxtaposition of the atrial

appendages. Situs inversus was confirmed in three patients. In one patient who was suspected of having left atrial isomerism, the transesophageal study defined atrial situs solitus. One patient each was documented to have left atrial and right atrial isomerism.

Systemic and pulmonary venous drainage. Both surface and transesophageal echocardiography were useful in detecting the drainage site of the inferior vena cava; absence of its suprarenal portion (2 patients) and direct drainage of the hepatic veins (2 patients) could be demonstrated by both techniques. In contrast, the superior cavo-atrial junction could be seen in only 19/32 patients with surface imaging, compared with 32/32 with transesophageal imaging. Bilateral superior caval veins were documented in two patients by transesophageal imaging, whereas in only one by prior surface echocardiography. In one patient with right atrial isomerism, the right sided superior vena cava was defined to be connected to the left side of the remnant of the atrial septum.

Partial anomalous pulmonary venous drainage - right upper pulmonary vein to the right sided superior vena cava - was documented in one patient only on transesophageal imaging, and was subsequently confirmed by cardiac catheterization.

Atrial septum In all patients with AV and ventriculoarterial discordance and an intact ventricular septum, the characteristic malalignment of the atrial septum with respect to the inlet ventricular septum, produced by the wedged position of the pulmonary outflow tract from the morphologic left ventricle could be seen by either technique (fig.1). A similar deviation of the mid portion of the atrial septum was noted on the transesophageal study in the two children with left juxtaposition of the atrial appendages. In three older patients atrial septal defects were detected by the transesophageal approach (two secundum defects, one coronary sinus defect) which were missed on surface echocardiography; the presence of the defect and the direction of shunting was confirmed by both color flow imaging and pulsed Doppler. In the remaining 16 patients with an atrial septal defect the transesophageal study was felt to be more reliable in the definition of their exact site and number. One patient was documented to have multiple defects, not appreciated during prior surface studies. This was confirmed during subsequent surgical inspection.

Atrioventricular junction

Discordant and criss-cross AV connections In 5/10 patients with AV discordance and intact inlet ventricular septum the reversed pattern of offsetting of the septal leaflets of the right and left AV valves was present. Thus, in four patients with situs solitus, the septal leaflet of the left AV valve (morphologic tricuspid valve) could be seen attached to the septum at a lower level than the corresponding leaflet of the right AV valve (figure 1). The reverse arrangement was observed in one patient with situs inversus. This relationship of the septal leaflets of both AV valves could be appreciated by both surface and transesophageal imaging. In three patients with an associated inlet ventricular septal defect, there was no offsetting of the septal leaflets. In all patients, the chordal attachments of both AV valves were precisely demonstrated by transesophageal imaging (figure 2), compared with adequate imaging in only two patients by surface echocardiography. Imaging of the site/s of chordal attachments enabled correct distinction of the morphologic right and left ventricles in all patients. Chordae from the morphologic tricuspid valve could be shown to have insertions into the ventricular septum, or to the crest of the septum in three patients with inlet septal defects. In contrast, the morphologic mitral valve had no septal chordal attachments.

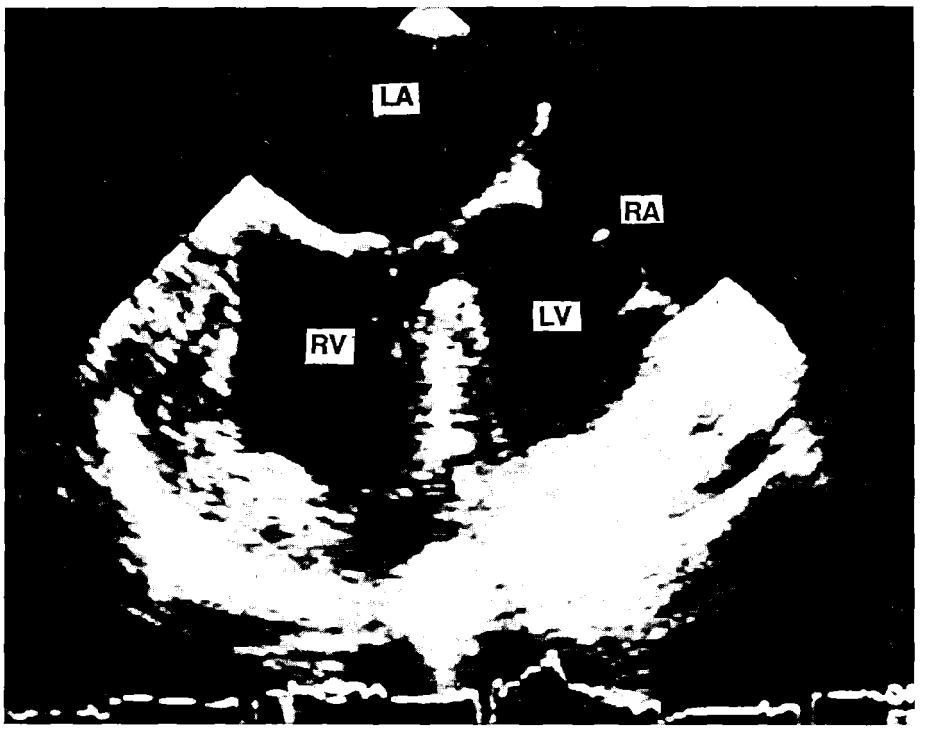

Figure 1: Transesophageal four chamber view in a patient with dextrocardia, situs inversus and congenitally corrected transposition. The atrial septum is malaligned with respect to the ventricular septum. The reversed offsetting of the two AV valves is clearly seen.
Legend: LA=left atrium; LV=left ventricle; RA=right atrium; RV=right ventricle.

In three patients with discordant AV connection and inlet ventricular septal defect, the chordal attachments of the septal leaflet of the morphologic tricuspid valve (left sided in two patients and right sided in one patient with situs inversus) were to the crest of the ventricular septum. In every case, detailed imaging of the septal leaflet and its tension apparatus was possible from the transesophageal approach using modified four chamber views, and chordal straddling could be definitely excluded. Overriding of either the tricuspid (two patients) or mitral (one patient) could however be detected by both precordial and esophageal approaches, and was <50% in every case. In a further patient with criss-cross heart (situs inversus and AV concordance) straddling of the chordae from the morphologic (inferior and right sided) tricuspid valve was demonstrated only by transesophageal imaging. Confirmation of the chordal attachments as defined by transesophageal imaging was obtained in two patients who subsequently had corrective surgical procedures.

Absent AV connection versus imperforate valve. In 1/12 patients diagnosed by surface imaging to have an absent right connection, and in 2/3 patients with an absent left AV connection, transesophageal echocardiography showed an imperforate valve with a potential communication with the underlying ventricle (figure 3). In all three cases, the imperforate

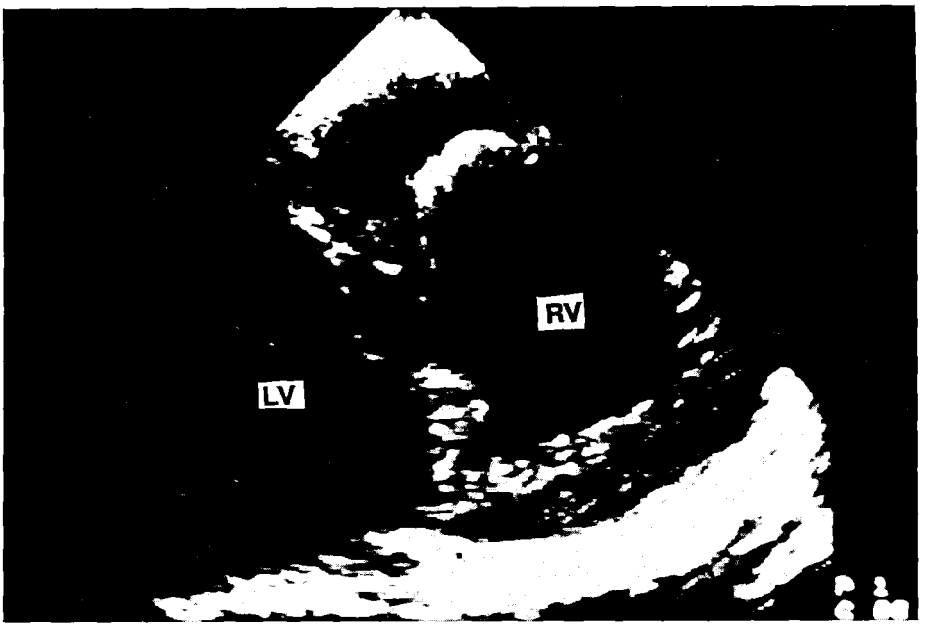

Figure 2: Transesophageal view for definition of chordal attachments in a patient with congenitally corrected transposition and associated inlet ventricular septal defect. Chordae of either AV valve are demonstrated to insert into the crest of the ventricular septum (arrows), however, there is no chordal straddling. The patient subsequently underwent biventricular repair.
Legend: LV = morphologically left ventricle; RV = morphologically right ventricle.

"membrane" domed into the atrium in systole. A rudimentary subvalvar apparatus to the underlying ventricle was documented in two patients with an imperforate left AV valve (figure 4). The diameter of the imperforate valve measured 4 to 7 mm, and the underlying ventricle was severely hypoplastic.

Double inlet ventricles Five children studied were diagnosed by surface echocardiographic studies to have double inlet of the left ventricle, one to have double inlet right ventricle. In the latter patient, the transesophageal study documented the presence of a rudimentary chamber anterior and superior to the main ventricular chamber (thus of right morphology), and thereby changed the diagnosis into double inlet of the left ventricle. The transesophageal studies in these six patients allowed for an improved insight into the individual chordal insertions of both AV valves, when compared to surface echocardiographic studies. Two children were studied in the preoperative evaluation for a septation procedure. In both, the transesophageal study contributed substantial additional information on the morphology of the subvalvar apparatus and its relations to the ventricular chambers and the ventricular outflow tracts (figure 5). However, no child so far underwent septation.

Atrioventricular valve morphology and function Determination of AV valve morphology, and distinction of the tricuspid from the mitral valve could not be satisfactorily achieved from surface or transesophageal short axis scans in any of the patients with AV discordance. In all

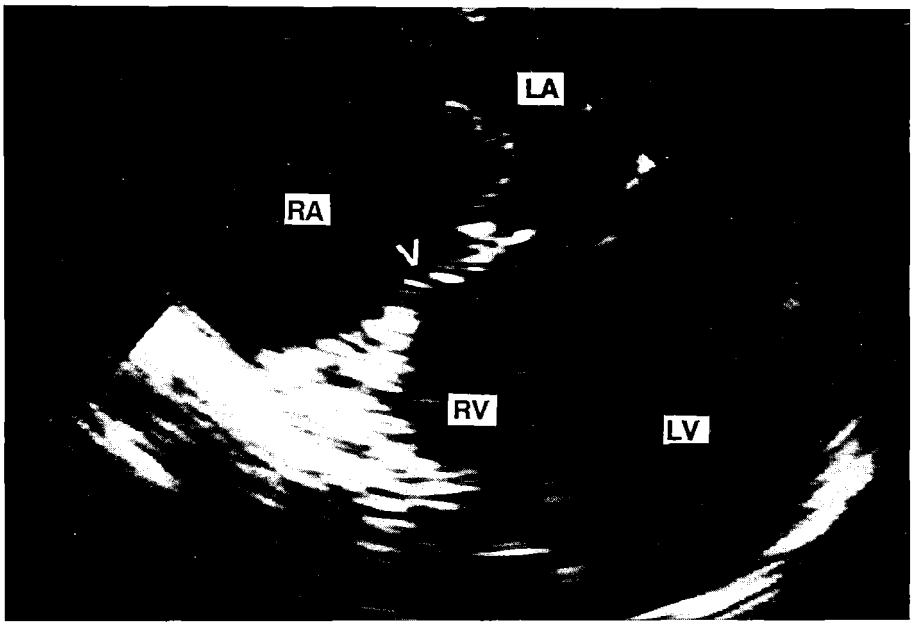

Figure 3: Detection of an imperforate tricuspid valve (arrow), in a child who was diagnosed absent right connection. (Abbreviations see fig 1)

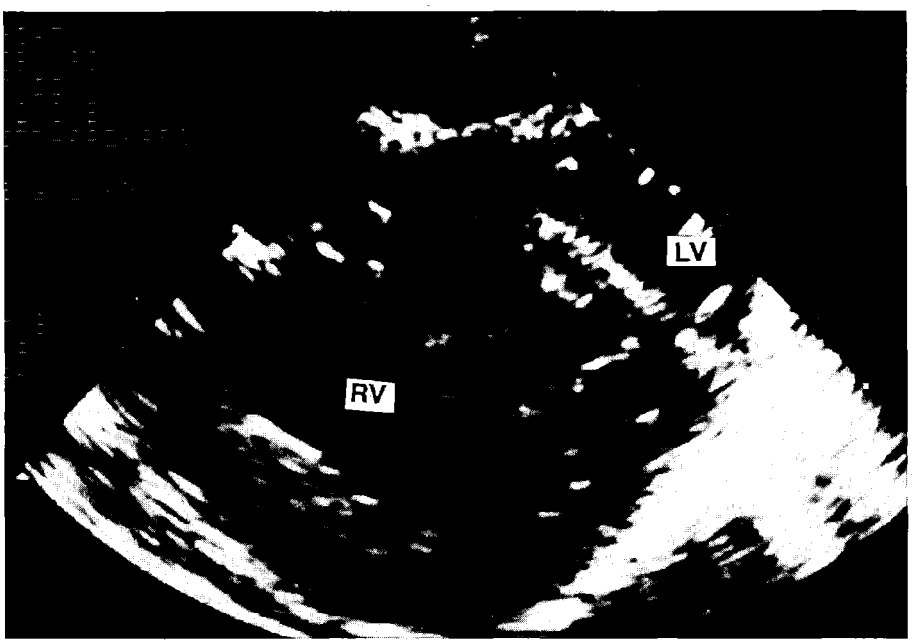

Figure 4: Imperforate left AV valve in a child. There is evidence of a subvalvar apparatus within the hypoplastic left ventricle (Abbreviations see figure 1)

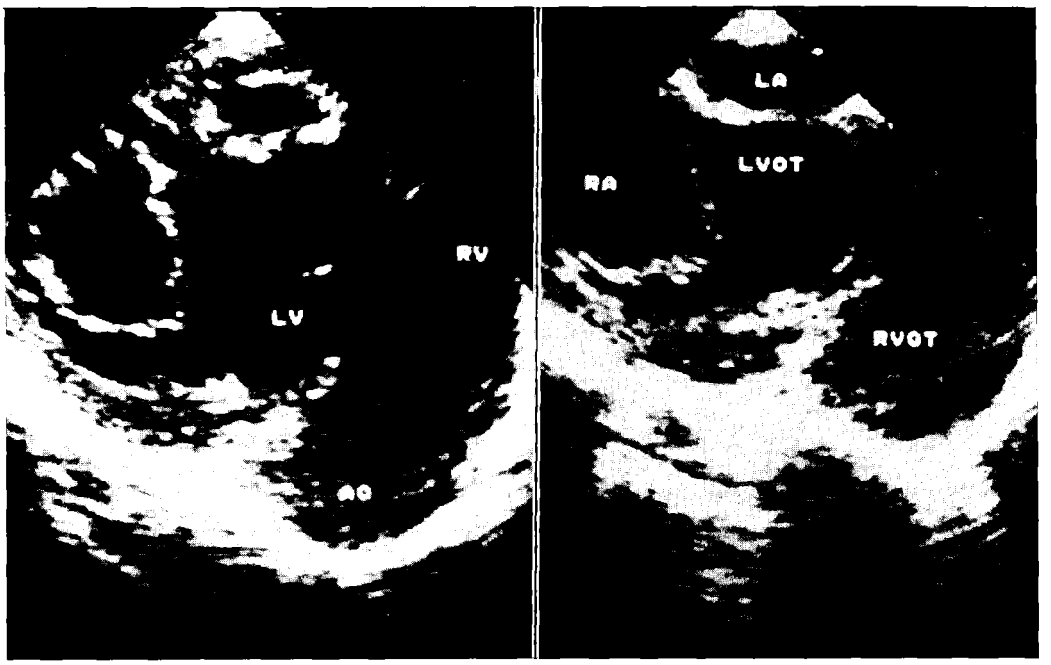

Figure 5: Sequence of transesophageal views in the preoperative evaluation of a child with double inlet of the left ventricle. The chordal attachments of either AV valve and their relation to the ventricular septum and outflow tracts were clearly demonstrated.
Legend: LVOT = left ventricular outflow tract; RVOT = right ventricular outflow tract.

patients studied the demonstration of the distinct patterns of chordal insertions of either AV valve was the most reliable predictor for definition of AV valve morphology. In one child who was diagnosed to have an absent left AV connection, the transesophageal study revealed a common atrioventricular valve, both concerning the valve morphology and the morphology of the subvalvar apparatus. There was severe right ventricular dominance in this patient.

Assessment of AV valve function was comparable in the majority of patients using either ultrasound approach. In only two patients the transesophageal study revealed minor additional regurgitant jets on color flow mapping studies. However, in a further two patients, after prosthetic left AV valve replacement, only transesophageal color flow mapping enabled the detection of valve dysfunction and paravalvular regurgitation.

Ventricular morphology and function In only two of the 10 adolescent and adult patients with discordant AV connections could the morphology of either ventricle be accurately determined based on the appearance of the trabecular septum by either approach. In view of the difficulty in visualizing AV valve morphology in the short axis, this meant that ventricular morphology had to be deduced from the offsetting of the AV valves in those with an intact septum, or from the chordal attachments of the valves when there was an inlet ventricular septal defect. Although the distinct patterns of ventricular trabeculation could be assessed in the younger patient population by transesophageal studies, the determination of

the position and the relation of both ventricular chambers were the most reliable predictors for ventricular topology in patients with double inlet of the ventricles. However, in two children an anterior rudimentary chamber, demonstrated by the prior surface echocardiographic study and on angiocardiography, could not be visualized during the transesophageal study.

Systemic ventricular function could be assessed by either method and M-mode measurements of ventricular cavity and wall dimensions could be obtained in all patients. Poor biventricular function was demonstrated in two patients, who subsequently underwent cardiac transplantation. In one other patient who was thought to have good systemic ventricular function on cross sectional and M-mode studies, transesophageal pulsed Doppler studies of pulmonary vein (marked elevation of the atrial retrograde flow velocity) and AV valve flow patterns suggested impaired ventricular diastolic function.

Arterial connections and relations The mode of ventriculo-arterial connection was more reliably assessed by surface than by transesophageal echocardiographic studies. Due to a combination of factors, including the interposition of the bronchial tree and cardiac enlargement, transesophageal studies failed to demonstrate the pulmonary artery bifurcation in 6 of the 32 patients, precordial imaging in 2 of the 32.

Surface echocardiography was superior to transoesophageal echocardiography in demonstrating the anterior aorta, and the subaortic muscular infundibulum in patients with discordant ventriculo-arterial connections (17/21 patients by surface echocardiography, compared with 11/21 by transesophageal echocardiography). Double outlet of the right ventricle (4 patients) was better defined using the precordial ultrasound approach. In particular, it was not possible to determine the degree of arterial override by transverse-axis transesophageal imaging.

Irrespective of the position of the heart or atrial situs, transesophageal echocardiographic studies were superior in assessing posterior ventricular outflow obstruction. In fifteen patients with mitral - pulmonary fibrous continuity and associated subpulmonary stenosis, the morphology of the stenosis was better shown by transesophageal imaging. In twelve, the obstruction was caused by a combination of subarterial muscular hypertrophy and abnormal tissue tags in the subpulmonary region arising from the mitral valve apparatus. In the remaining three patients isolated muscular hypertrophy was documented. However, transesophageal pulsed wave Doppler, due to the generally poor alignment, failed to determine the pressure gradient across these obstructions in all patients.

Discussion

Congenital morphologic abnormalities of the AV junction constitute a significant proportion of cardiac abnormalities diagnosed in early life. A detailed morphologic and functional evaluation is required prior to planning surgical repair of these lesions. This is particularly true in those patients who survive into adolescent and adult life.

Although precordial echocardiographic examinations have been reported to be a sensitive technique in the preoperative evaluation of AV junction abnormalities (1-3), several difficulties are encountered in clinical practice. In particular, in patients with abnormal cardiac position or cardiac malrotation the transthoracic ultrasound windows are often

limited. In addition image quality is much reduced in the older patient population and in those who had prior (palliative) surgical correction via a median sternotomy, due to the acquired fibrous tissue adhesions. Thus, in selected cases, there would appear to be a need for an additional imaging technique.

Both transesophageal echocardiography and magnetic resonance imaging (MRI) have been utilized to assess cardiac morphology in older patients with congenital heart defects. Magnetic resonance imaging has the potential advantage of allowing multi-plane imaging of both the heart and great vessels, compared with transesophageal imaging with its limited planes (11,12). MRI however is static gated for high resolution imaging, and flow abnormalities cannot be accurately assessed. Although cine MR imaging has recently been used to detect flow disturbances within the heart and great vessels, to assess ventricular function and calculate intracardiac shunt volumes, the images are compiled from the cumulative information obtained over several heartbeats (13-15). Until the problems of assessing intracardiac flow in real time are solved, two-dimensional echocardiography with pulsed Doppler and color flow mapping will continue to play an important role in the assessment of the morphology and flow patterns within the heart and great vessels in patients with complex cardiac malformations. All patients described here were investigated by single plane transesophageal imaging; biplane probes became recently available, and potentially can further increase the diagnostic yield of transesophageal echocardiography.

Patients in this study fell into three main groups: those with a discordant AV connection (10 patients), those with an absent atrioventricular connection (15 patients) as determined previously from surface imaging and angiographic investigations, and those with a double inlet ventricle. Although all patients in this study had already been diagnosed to have major morphologic abnormalities of the AV junction, some of the advantages of transesophageal imaging were readily apparent.

Atrial situs could be determined by direct imaging of the atrial appendages, irrespective of the position of the heart. Although atrial situs could be indirectly inferred by determining the sites of drainage of the systemic and pulmonary veins, morphologic classification of any chamber should be based on the recognition of its most consistently present parts, and in this respect transesophageal imaging was superior to surface imaging in all 32 patients. The pattern of systemic and pulmonary venous drainage, and also the integrity of the atrial septum could be determined more consistently by transesophageal studies. Unsuspected anomalous venous return was defined in three cases, unsuspected atrial septal defects in four patients. The impact of this information for planning surgical correction in these cases is obvious.

Congenitally corrected transposition A characteristic echocardiographic feature of congenitally corrected transposition is the pattern of offsetting of the septal leaflets of the AV valves (2,16). While this feature could be easily recognized in patients with an intact ventricular septum, problems arose when there was an associated inlet ventricular septal defect. In this situation the septal leaflets of both AV valves were inserted at the same level, and ventricular morphology had to be determined by either the chordal attachments of the AV valves, or the appearance of the trabecular portion of the ventricular septum (2,17). Septal trabeculation patterns, with either surface or transesophageal imaging, were a poor guide to ventricular morphology in these older patients with AV discordance. With

transesophageal imaging however, the attachments of the chordae from either AV valve could be demonstrated with precision in every patient. The morphologic tricuspid valve could be differentiated from the mitral valve by the presence of chordal attachments to the septum, or the crest of the septum in patients with inlet septal defects. In contrast, the chordae from the mitral valve could be shown to attach to papillary muscles on the lateral wall of the ventricle only. Detection of the presence and degree of chordal straddling has great importance in planning surgical correction, particularly in those with biventricular hearts. Angiography is a poor technique for the demonstration of chordal straddling, and relies on the detection of AV valve override, and the streaming patterns following contrast injections into the atria. As shown in this study, overriding of an AV valve was a common feature in every patient with an inlet ventricular septal defect. Although surface two-dimensional imaging has been shown to be reliable in demonstrating straddling AV valves (18), it may not be possible always to define the chordal attachments from this approach in older patients. In two patients who have subsequently had intracardiac repair of their defects, chordal straddling could be definitely ruled out preoperatively by transesophageal imaging, and both patients had biventricular repairs. In two other patients with associated ventricular septal defects who are awaiting surgery, transesophageal imaging clearly demonstrated chordal straddling in one patient which was not evident from precordial imaging.

Absent AV connections Imperforate AV valves (two left sided and one right sided) were detected in three patients diagnosed previously to have an absent AV connection. In either case, however, the information was not of value in planning the subsequent surgical procedure, in view of the small diameter of the potential communication and extreme hypoplasia of the underlying ventricular chamber. Recent reports using MRI have shown the ability of the technique to distinguish absent connection from an imperforate AV valve (19); our experience indicates that this information can be equally well obtained by transesophageal imaging.

Double inlet ventricles In this group of patients the transesophageal studies contributed additional insight into the individual chordal implantation of either AV valve and into their relations to the ventricular chambers and outflow tracts. In two patients studies were performed electively in the preoperative evaluation of a septation procedure. Whereas ventricular morphology is best determined by definition of the chordal insertions of the AV valves in patients with biventricular AV connections, this most reliably performed by documentation of the relative position of the ventricular chambers to one another in those with univentricular AV connections. In this respect it is noteworthy that transesophageal studies may not visualize rudimentary anterior chambers, however, posterior chambers and their dimensions are more readily identified.

Posterior ventricular outflow obstruction Visualization of the subpulmonary outflow tract in patients with discordant AV and ventriculoarterial connections has been a serious limitation in previous surface echocardiographic studies, even in younger patients (2). In this eries, excellent morphologic assessment of the subpulmonary area was possible by trans-esophageal echocardiography. In contrast, the technique provided less morphologic information about the anterior subaortic infundibulum when compared with surface imaging.

Ventricular function Although ventricular function could be assessed by a combination of cross sectional and short axis M-mode scans using either approach, pulsed Doppler studies

of the pulmonary venous flow patterns provided a new insight into the diastolic properties of the systemic ventricle. Little is known about pulmonary venous flow patterns in various forms of complex congenital heart disease. From the results of animal studies, and some precordial pulsed Doppler studies in patients with total anomalous pulmonary venous drainage, or following atrial repair procedures for transposition of the great arteries the pulmonary venous flow patterns appear to reflect left atrial dynamics (20-22). In one patient with apparently normal systolic function of the systemic ventricle as judged by M-mode echocardiography, pulmonary venous flow patterns by transesophageal pulsed Doppler reflected an increased end-diastolic pressure in the systemic ventricle and diminished ventricular compliance. It has been shown in previous studies of adult patients with congenitally corrected transposition that the morphologic right ventricle can function adequately as the systemic ventricle even in the presence of associated lesions that may impose additional hemodynamic loads (23). An important caveat however, must be the early detection of ventricular dysfunction, and appropriate surgical measures before overt systemic ventricular failure occurs. Long term follow-up of either the natural or postsurgical history of these patients should be possible with the use of transesophageal imaging combined with pulsed Doppler and colour flow mapping.

It is concluded that transesophageal echocardiographic studies provide an improved insight into morphology and function of complex congenital atrioventricular junction abnormalities. The chordal attachments of either AV valve can be defined with improved accuracy particularly in the older child or in the adolescents and adult population. In addition, the technique allows a better insight into subpulmonary obstruction in patients with ventriculo-arterial discordance.

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Chapter 8

Left ventricular outflow tract obstruction in childhood - improved diagnosis by paediatric transoesophageal echocardiography

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Summary

Paediatric transoesophageal echocardiography was used in 4 children with either documented or suspected left ventricular outflow obstruction to determine whether this technique may provide additional morphologic information when compared to praecordial ultrasound. Transoesophageal studies defined unsuspected mitral valve abnormalities in two patients and involvement of the subvalvar mitral apparatus in one. In one patient the study documented a suspected fibrous ridge assocciated with a malaligned ventricular septal defect.

Transoesophageal echocardiography would appear to be a new diagnostic technique with definitive advantages in the evaluation of the spectrum of lesions which cause left ventricular outflow obstruction in childhood.

Introduction

The diagnosis and differentiation between the various forms of left ventricular outflow tract obstruction in children (1) is performed routinely by praecordial cross-sectional imaging (2). Tunnel-like lesions, the variants of hypertrophic obstructive cardiomyopathy, and discrete fibromuscular obstructions can all be defined with ease in this age group.

Transoesophageal echocardiography was recently reported to be of improved diagnostic value in the diagnosis of left ventricular outflow tract obstruction in adolescents and adults (3-5). However, in children with excellent praecordial ultrasound windows in the majority of cases, the additional use of transoesophageal echocardiography would appear to be of limited value. We studied four children with evidence of left ventricular outflow tract obstruction by both transoesophageal and praecordial ultrasound in order to determine whether the technique could contribute additional information in the morphologic diagnosis.

Patients and Methods

The age at investigation ranged from 1 year 11 months to 12 years, the weight from 9.5 kg to 36 kg (mean 21 kg). Transoesophageal studies were performed with specially dedicated paediatric transoesophageal probes (single plane, transverse axis) with a maximal tip circumference of 30 mm working at an ultrasound frequency of 5 MHz. The shaft diameter of the gastroscope measured 7 mm; steering facilities were limited to anterior / posterior

angulation. One patient was studied under general anaesthesia during preoperative cardiac catheterization, and three patients intraoperatively prior to surgical resection of the obstruction; in two of these patients post bypass studies were performed. The results of the transoesophageal studies were correlated with the prior praecordial studies and with the findings at surgical inspection.

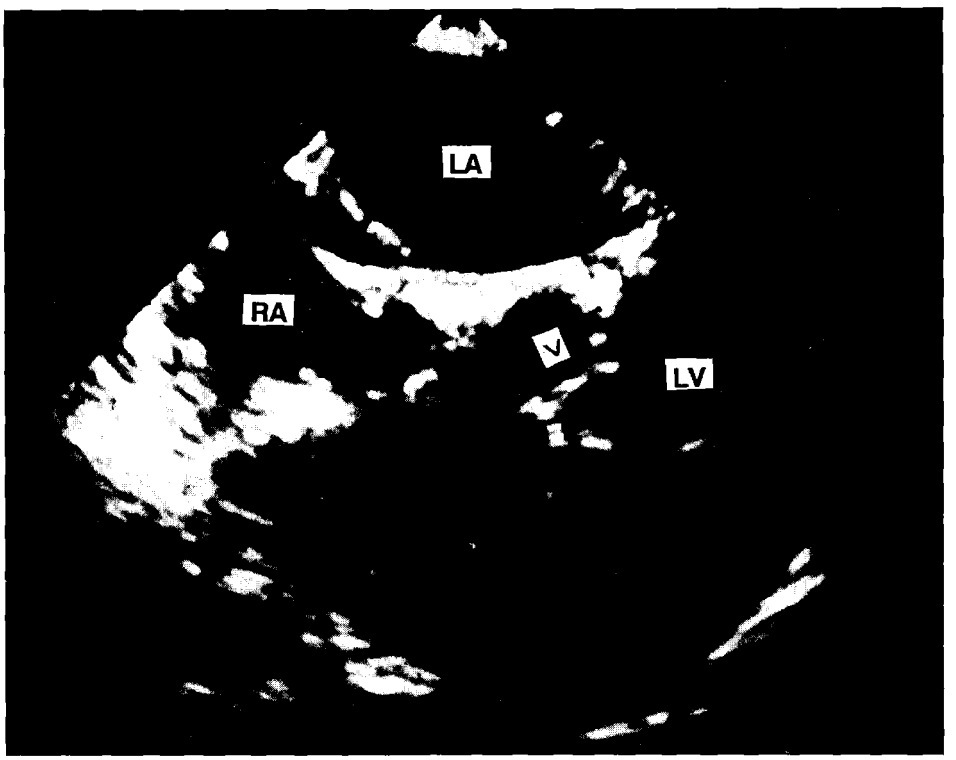

Figure 1: Transoesophageal demonstration of an additional papillary muscle (arrow) of the mitral valve in a 9.5 kg child producing left ventricular outflow obstruction. The prior praecordial study suggested a "membrane-like" lesion and documented abnormal diastolic movement of the anterior leaflet of the mitral valve; however the underlying pathology of the subvalvar apparatus was missed. Note the widely patent foramen ovale.
Legend: LA = left atrium, LV = left ventricle, RA = right atrium

Results

The praecordial studies (5 and 7.5 MHz transducers) had defined left ventricular outflow obstruction in three patients, and suggested it in one further patient. In the latter patient only mild systolic turbulence in the left ventricular outflow tract was detected on colour flow mapping; no morphologic correlate was demonstrated by cross-sectional imaging. Praecordial scans suggested membrane-like lesions in two patients and demonstrated a narrow left ventricular outflow tract in one patient 11 years after a Mustard procedure for transposition of the great arteries. No patient was found to have mitral valve abnormalities; diastolic movement of the anterior leaflet of the mitral valve was found to be abnormal in

only one patient.

Transoesophageal studies defined mitral valve abnormalities in two patients. In one an additional commissure of the mitral valve was documented on transgastric short axis cuts; insertion of mitral valve chordae into the interventricular septum were demonstrated using transoesophageal four chamber views. In one patient an additional papillary muscle of the mitral valve was documented. The existence of a membrane-like lesion, as suggested by the praecordial study, was excluded (fig 1).

In the third patient, in whom only mild systolic turbulence in the left ventricular outflow tract was demonstrated by praecordial colour flow mapping, a pre bypass transoesophageal study revealed the existence of a fibrous ridge on the crest of a malaligned ventricular septal defect. The subsequent post bypass study revealed absence of any remaining tissue and laminar flow in the left ventricular outflow tract. The existence of a fibromuscular membrane and asymmetric septal hypertrophy was diagnosed by praecordial ultrasound in one patient. Whereas only the septal attachment of the "membrane" was demonstrated on praecordial cross-sectional imaging, the transoesophageal pre bypass study documented the circular shape of the lesion, including the attachment onto the mid portion of the anterior leaflet of the mitral valve. A post bypass study in this patient showed remnants of the mitral valve attachments (fig 2). However these were of no haemodynamic significance.

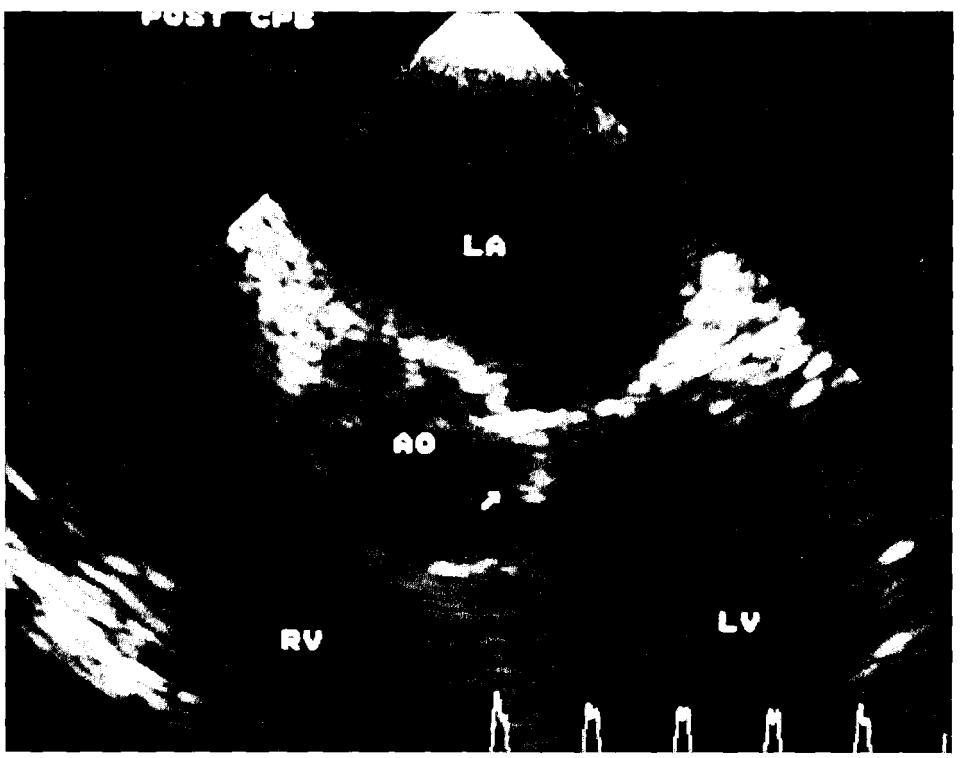

Figure 2: Immediate post bypass transoesophageal study revealing a remnant of a discrete fibromuscular membrane attached to the mid portion of the anterior leaflet of the mitral valve (arrow).
Legend: Ao = Aorta, LA = left atrium, LV = left ventricle, RV = right ventricle

Discussion

We would conclude from this initial experience that transoesophageal echocardiography in children with left ventricular outflow tract obstruction allows an improved insight into the morphology of this spectrum of lesions. In particular any involvement of the mitral valve apparatus or coexisting mitral valve abnormalities such as additional commissures or papillary muscles can be better defined by transoesophageal than by praecordial cross-sectional imaging. The additional morphologic information provided by transoesophageal echocardiography was of great value for the surgical management of these lesions, since the surgeon can achieve only limited exposure of these lesions when the repair is performed via the aortic root. In addition, the intraoperative use of the technique allowed for the immediate and accurate post bypass demonstration of the surgical results. Following atrial correction procedures for transposition of the great arteries obstructive lesions of the left ventricular outflow tract are a frequent finding. However, the praecordial assessment of this range of lesions his largely limited. In this subgroup of patients transesophageal examinations are particularly rewarding.

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Chapter 9

Intraoperative transesophageal versus epicardial ultrasound in surgery for congenital heart disease

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Summary Twenty-eight patients (age 0.7 to 65 years, median 6.1 years) undergoing correction for congenital heart disease were entered into a prospective study using both intraoperative transesophageal and epicardial ultrasound to determine the relative values of these techniques in the pre and post bypass situation.

Introduction of the transesophageal probe was successful in 26 patients (93 %). Children were studied using dedicated pediatric transducers. Epicardial studies could be performed in all 28 patients, and allowed for higher resolution imaging and a more complete pre bypass assessment of intracardiac morphology (ventricular septum, right ventricular outflow tract) than was obtained by the transesophageal approach. In the immediate post bypass period the transesophageal technique allowed a more detailed insight into atrioventricular valve function (valvar regurgitation (5 patients) and ventricular inflow patterns) and the continuous monitoring of left ventricular function and volume. Residual interventricular shunting (3 patients) or residual outflow tract obstruction (4 patients) could not be reliably documented by transesophageal studies.

It is concluded that intraoperative transesophageal and epicardial ultrasound in surgery for congenital heart disease are complementary rather than alternative techniques.

Introduction

Over the recent years intraoperative echocardiography has become an established monitoring technique in the surgery of congenital heart disease (1-3). The epicardial ultrasound approach can provide excellent quality images and thus diagnostic information in the vast majority of patients (4-7). Transesophageal echocardiography is an alternative intraoperative approach which can now be used. This technique, which has been widely adopted in adult cardiac surgery (8-10), has the major advantage that it does not interfere with the surgical procedure and that it allows continuous monitoring of ventricular function and volume (11,12). However, reports on the use of intraoperative transesophageal echocardiography in surgery for

congenital heart disease are still limited. Initial experiences have been reported by Cyran (13) and, using a pediatric transesophageal probe, by Kyo (14). With the introduction of dedicated pediatric transesophageal probes, this new intraoperative ultrasound approach is likely to be the subject of much interest. However, the relative benefits of intraoperative transesophageal echocardiography in the surgery of congenital heart disease as opposed to those of epicardial ultrasound remain yet to be defined (15).

In a prospective study, both intraoperative transesophageal and epicardial echocardiographic studies have been attempted in twenty eight patients undergoing correction of congenital heart disease, in order to determine the relative values of these techniques in the pre bypass assessment of intracardiac morphology, in the post bypass exclusion of residual hemodynamic lesions, and for monitoring of left ventricular function and filling.

Patients and methods

Study patients Twenty eight patients were studied. The age at surgical correction ranged from 8 months to 65 years (median age 6.1 years). Their weights ranged from 4.7 to 72 kilograms (median weight 19 kg). Twenty-one patients belonged to the pediatric age group, of whom fifteen weighed less than 20 kilograms. The preoperative diagnoses and surgical procedures are listed in Table I.

Before starting the study, ethical approval was granted by the Ethical Committee of the Academisch Ziekenhuis Rotterdam. Before individual studies, informed consent was obtained from the patients or their parents.

Intraoperative echocardiography In patients who were large enough to be intubated with a cuffed endotracheal tube or who underwent a short bypass time (i.e. less than one hour) both pre and post bypass transesophageal echocardiographic studies were scheduled. In those children who were intubated with an uncuffed endotracheal tube while undergoing long surgical procedures, transesophageal studies were restricted to pre bypass studies alone (n = 9). Reinsertion of the probe immediately after bypass was not feasible in our operating theatres because access was restricted by a plastic frame protecting the patient's head. Epicardial studies were performed both before and after cardiopulmonary bypass in all patients.

A continuous electrocardiogram was displayed on the ultrasound monitor, and studies were recorded continuously onto video tape. Interpretation of the studies was performed both on-line real-time and off-line at reduced speed by two independent observers who were familiar with the techniques. The results obtained were then correlated.

Transesophageal studies All studies were performed using single plane (transverse axis) transesophageal probes. In adult patients (n = 7) a 5.6 MHz 64 element probe with a shaft diameter of 10 mm and a tip circumference of 54 mm (12 x 15 mm) was used on a Toshiba SSH 160 A or SSH 140 A ultrasound system. The steering mechanism allowed both anterior/posterior and right/left lateral flexion of the probe. Children were studied with either a 5 MHz 24 element pediatric probe on an Aloka SSD 870 ultrasound system (n = 3) or with a 5 MHz 48 element prototype pediatric transesophageal transducer (Department of Experimental Echocardiography, Thoraxcenter Rotterdam) connected to a Toshiba SSH 160 A or SSH 140 A ultrasound system (n = 18). The dimensions of both pediatric probes were comparable with a maximal shaft diameter of 7 mm and a maximal tip circumference of 30 mm (7 x 8 mm and 5 x 10 mm respectively). Steering facilities were restricted to anterior/posterior only.

Table I: Patients studied

Patient No.	Age yrs.	Weight kg	Cardiac malformation	Surgical Correction
1.	17.4	68	Ebstein	Reconstruction
2.	5.5	17.3	ASD II	Direct closure
3.	65	69	Partial AVSD	Patch closure, MV reconstruction
4.	4	15.6	ASD II	Direct closure
5.	19.3	72	LVOTO	Resection
6.	62	63	Coronary artery fistula	ligation
7.	6.1	19.2	ASD II	Direct closure
8.	13.6	40	MV stenosis	Valve replacement
9.	2.3	13.2	Perimembranous VSD	VSD patch closure
10.	17	56	Ebstein	Reconstruction
11.	5.7	17.	Tetralogy of Fallot	VSD patch closure, transannular patch
12.	2.0	9.8	LVOTO, malalignment VSD	Resection, VSD patch closure
13.	10.4	28	LVOTO, perimembranous VSD	Resection, VSD patch closure
14.	4.9	18	Post op pulmonary atresia	ASD closure
15.	3.7	14.4	LVOTO	Resection
16.	1.3	6.9	Transposition, VSD	Arterial switch
17.	2.2	12.2	Muscular outlet VSD	VSD patch closure
18.	6.3	22	DILV	Fontan-type procedure
19.	13.9	52	Ebstein	Reconstruction
20.	9.3	21	Complete AVSD	patch reconstruction
21.	54	77	Tetralogy of Fallot, MV endocarditis	VSD patch closure, MV repair
22.	2.4	13.3	DILV	Fontan-type procedure
23.	12.7	51	Severe AS, ASH	Ross procedure
24.	0.7	4.7	VSD, left SVC, unroofed CS	VSD patch closure, CS patch
25.	2.0	9.8	Tricuspid atresia	Fontan-type procedure
26.	24	58	Tricuspid atresia	Glenn anastomosis
27.	1.8	9.2	DORV, subpulmonary VSD	Arterial switch
28.	5.0	18.7	Perimembranous VSD	VSD patch closure

Legend: AS = aortic stenosis, ASD II = secundum type atrial septal defect, ASH = asymmetrical septal hypertrophy, AVSD = atrioventricular septal defect, CS = coronary sinus, DILV = double inlet left ventricle, DORV = double outlet right ventricle, LVOTO = left ventricular outflow tract obstruction, MV = mitral valve, SVC = superior vena cava, VSD = ventricular septal defect.

The transesophageal probe was introduced in the anesthetic induction room. A small amount of anesthetic gel was used for lubrification. In younger children the probe was introduced under direct laryngoscopic vision. The probe was first advanced to the stomach in order to obtain short axis views of both ventricles. This was followed by gradual withdrawal of the probe in order to obtain the complete range of transesophageal short (transverse) axis views (16). In adult patients modifications of these views were obtained by using right or left lateral tilt of the transducer. Firstly cross-sectional imaging including M-mode studies was performed, secondly color flow mapping together with color M-mode studies were carried out and lastly pulsed wave Doppler sampling of relevant areas of interest was performed.

Epicardial studies Standard precordial transducers, packed in long sterile plastic sleeves, were used in every case. Cross-sectional imaging was performed using a 5 MHz 64 element short focus transducer in children, or a 5 MHz 96 element transducer in adolescent and adult patients. Color flow mapping, pulsed or continuous wave Doppler studies were performed using a 3.75 MHz 64 element transducer. Studies were performed using either a Toshiba SSH 160 A or SSH 140 A ultrasound system or an Aloka SSD 870 system. The sterile packed probes were passed to the operation field were they remained during the entire operation. Warm saline was poured into the pericardial cradle in order to improve image quality and to reduce mechanical irritation of the heart.

Complete epicardial pre and post bypass studies were performed in every patient. These included firstly cross-sectional imaging, secondly color flow mapping and thirdly pulsed or continuous wave Doppler studies of specific areas of interest. Scan positions were chosen to fully assess the intracardiac morphology and the related hemodynamic lesions. The scan planes recorded included long and short axis views comparable to those obtained from the precordium, modified four chamber views, long axis right ventricular outflow tract views, views obtained from the right atrium, and short axis views through the ascending aorta for assessment of the pulmonary arterial system.

Results

Success rate Insertion of the transesophageal probe was successful in 26 of the 28 patients entered into the study protocol (93 %). In two patients (no 8,26) attempts to introduce the (adult) transesophageal probe remained unsuccessful even after direct laryngoscopic vision of the hypopharynx. Both pre and post bypass epicardial studies were performed in all patients (100 %).

Image quality The resolution of the cross-sectional images obtained by transesophageal echocardiography was inferior to that obtained during direct epicardial studies using standard precordial equipment. The difference in image quality was most marked in children, in whom transesophageal transducers with fewer elements had to be used.

Assessment of cardiac morphology Pre-bypass transesophageal studies allowed the rapid and detailed morphologic assessment of the systemic and pulmonary venous returns, the interatrial septum, the mitral valve and the left ventricular outflow tract. No difference between the epicardial and the transesophageal approach was noted in the assessment of these structures. Only the pulmonary venous return and the interatrial septum were more rapidly assessed by the transesophageal technique.

The assessment of atrioventricular valve morphology in two patients with atrioventricular

septal defects (no 3,20) was comparable using either technique. In two patients who underwent surgical correction for double inlet left ventricle (no 18,22) the transesophageal approach allowed good insight into the morphology of the subvalvar apparatus and into valvar function (minimal mitral regurgitation in one). However, the epicardial study was superior in both patients, as the complete range of epicardial views (fig 1) demonstrated better the relationship of either atrioventricular valve to the ventricular outflow tracts, the semilunar valves and the remnant of the ventricular septum.

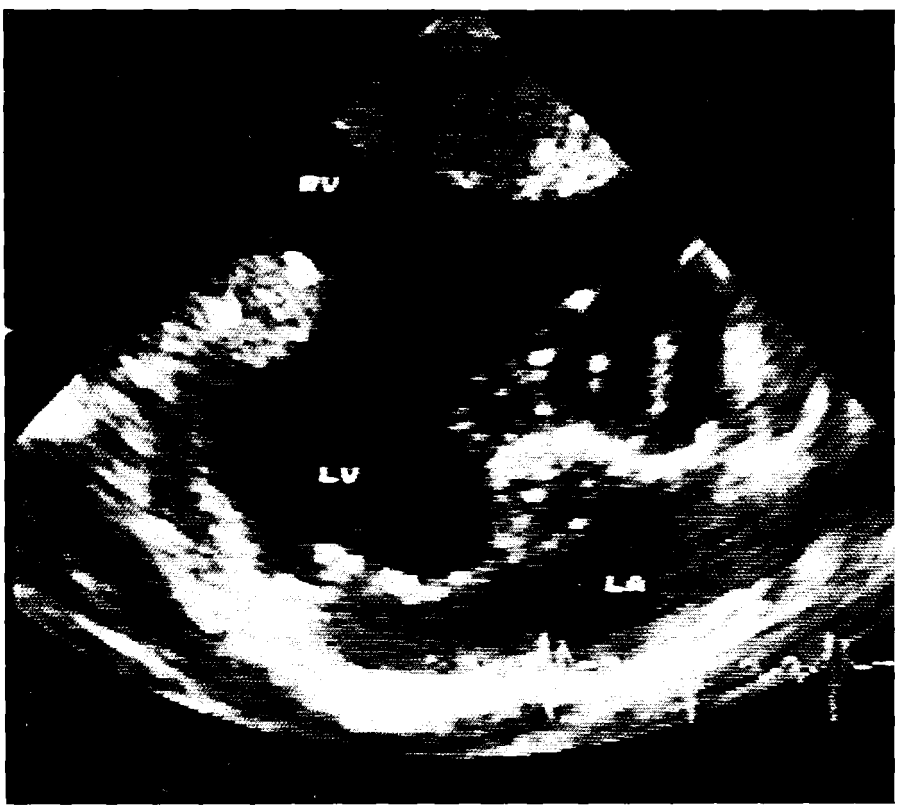

Figure 1: Cross-sectional image taken from a sequence or epicardial scan planes used in the assessment of complex atrioventricular valve morphology, and its relation to the ventricular outflow tracts.
Legend: LA = left atrium; LV = left ventricle; RV = right ventricle.

Three patients underwent reconstruction of the tricuspid valve for Ebstein's anomaly (no 1,10,19). In two of these, the fixed transverse axis transesophageal scan planes together with the large right atrial dimensions precluded a full assessment of the morphology of the tricuspid valve (figure 2). Although the epicardial approach contributed significant detail in the pre bypass assessment of the tricuspid valve morphology and clearly was superior to the transesophageal approach in all three patients, it remained incomplete in one (no 10). In this patient the septal and anterior tricuspid valve leaflets were severely displaced and almost completely tethered to the right ventricular apex.

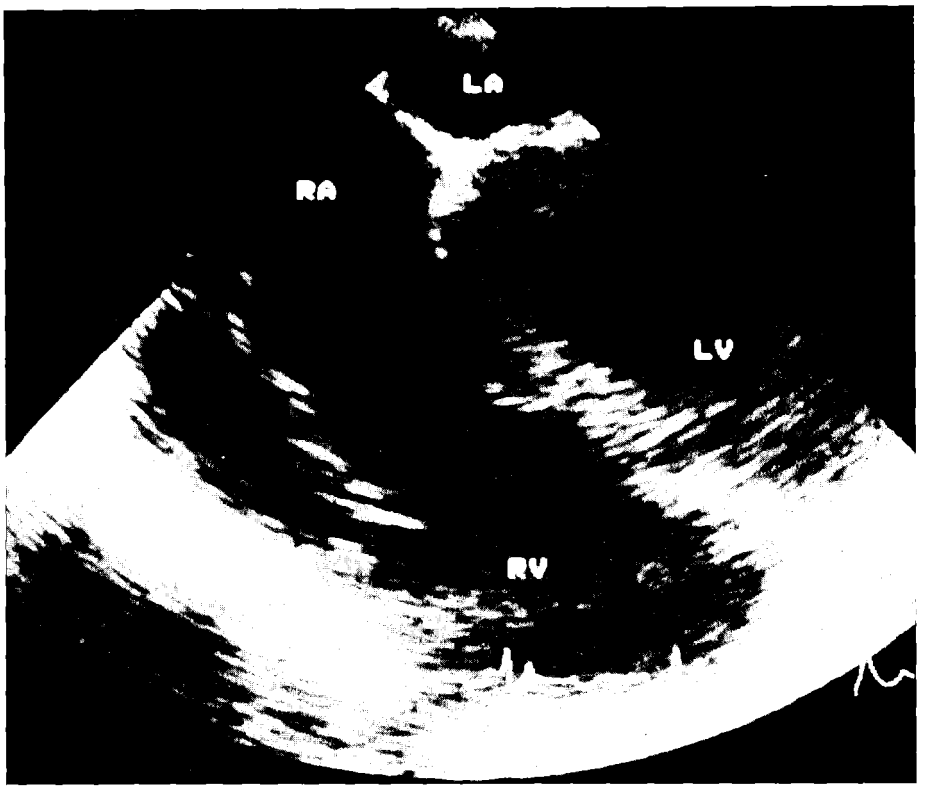

Figure 2: Transesophageal four-chamber view in a patient with Ebstein's anomaly of the tricuspid valve. Tethering and apical displacement of the septal leaflet of the tricuspid valve is readily identified. However, the assessment of the precise morphology of the subvalvar apparatus is limited due to the great distances from the transducer.
Legend: RA = right atrium; others see figure 1.

Twelve patients had ventricular septal defects. Whereas perimembranous ventricular septal defects and their relationship to the atrioventricular valves were reliably identified by either technique, muscular ventricular septal defects (2 patients) posed a problem to transesophageal echocardiography. In one patient (no 16) an additional apical muscular defect was missed during the transesophageal study. Arterial override over a ventricular septal defect in 3 patients (no 11,21,27) was not assessed reliably using transverse axis transesophageal imaging planes, nor was the precise morphology of right ventricular outflow obstruction identified in 2 patients (no 11,21). In contrast, a complete and detailed assessment of these lesions was possible by the direct epicardial approach. The exact morphology of left ventricular outflow tract obstruction in 4 patients (no 5,12,13,15) was equally well assessed using either approach.

The assessment of the morphology of the great arteries was readily performed using the epicardial approach, whereas in particular the left pulmonary artery and a large segment of the ascending aorta could not be visualized by transesophageal imaging. No differences between the two techniques were noted in the assessment of the morphology of the main and right pulmonary arteries in children. In adult patients the pulmonary valve and the central pulmonary

arterial system were not visualized satisfactorily in four patients by transesophageal studies. Assessment of the surgical repair One of the twenty-eight patients (3.6 %) underwent immediate revision during a second period of bypass for left ventricular inflow obstruction as detected by the epicardial ultrasound study (no 24). Because of the patients size (4.7 kilograms) he had not been studied by transesophageal ultrasound in the immediate post bypass period. No other patient was found by either technique to have significant residual lesions that warranted immediate revision.

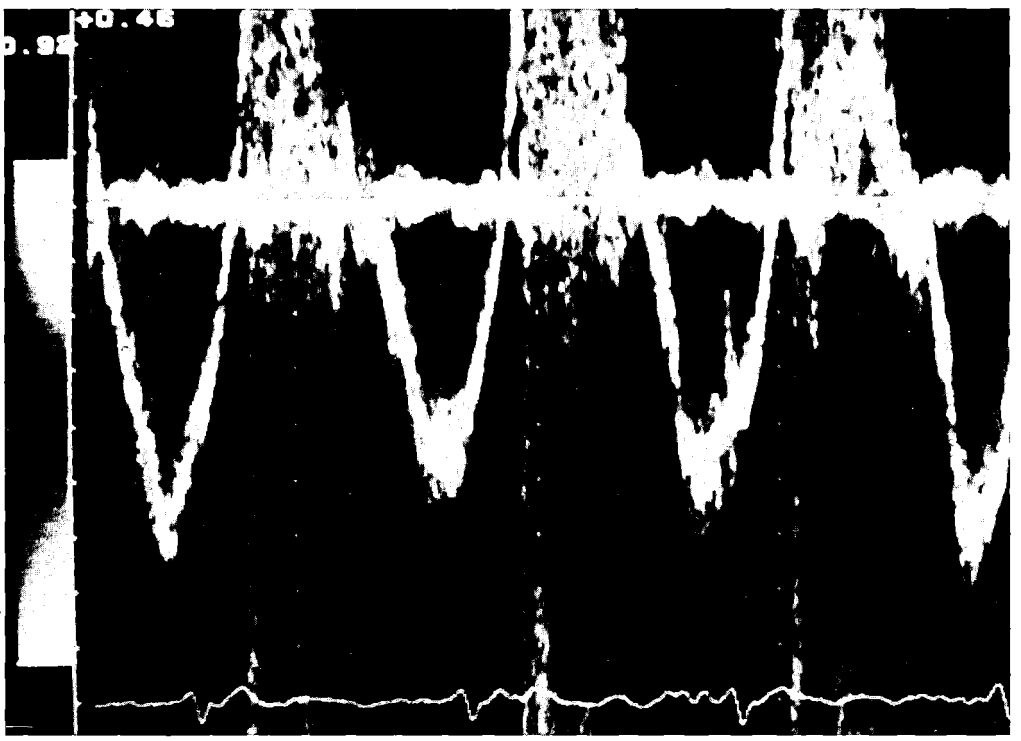

Figure 3: Transesophageal pulsed wave Doppler study for the immediate exclusion of tricuspid inflow obstruction after surgical reconstruction of Ebstein's malformation of the tricuspid valve. Maximal velocities are less than 1 m/s. There is mild tricuspid regurgitation.

Minor degrees of residual atrioventricular valvar regurgitation were noted in five patients. These were readily demonstrated in every patient by transesophageal studies. In two of these patients, both after patch closure of a ventricular septal defect, high left atrial views had to be used to detect the regurgitant jets during the epicardial studies, since flow masking precluded their demonstration by epicardial four chamber views. Atrioventricular valvar inflow patterns were much more reliably documented using the transesophageal technique (figure 3), as the views that could be obtained from the epicardium did not allow a good alignment to blood flow. This was of relevance in particular in three patients after reconstructive repair of Ebstein's malformation.

Mild residual interventricular shunting immediately after patch closure of ventricular septal defects was detected in three patients by epicardial post bypass studies. Colour flow mapping

studies not only detected the existence of residual shunting but also allowed the precise demonstration of its site. Transesophageal post bypass studies, because of the flow masking properties of the prosthetic material used in the repairs, did not show residual shunting in any patient (fig 4).

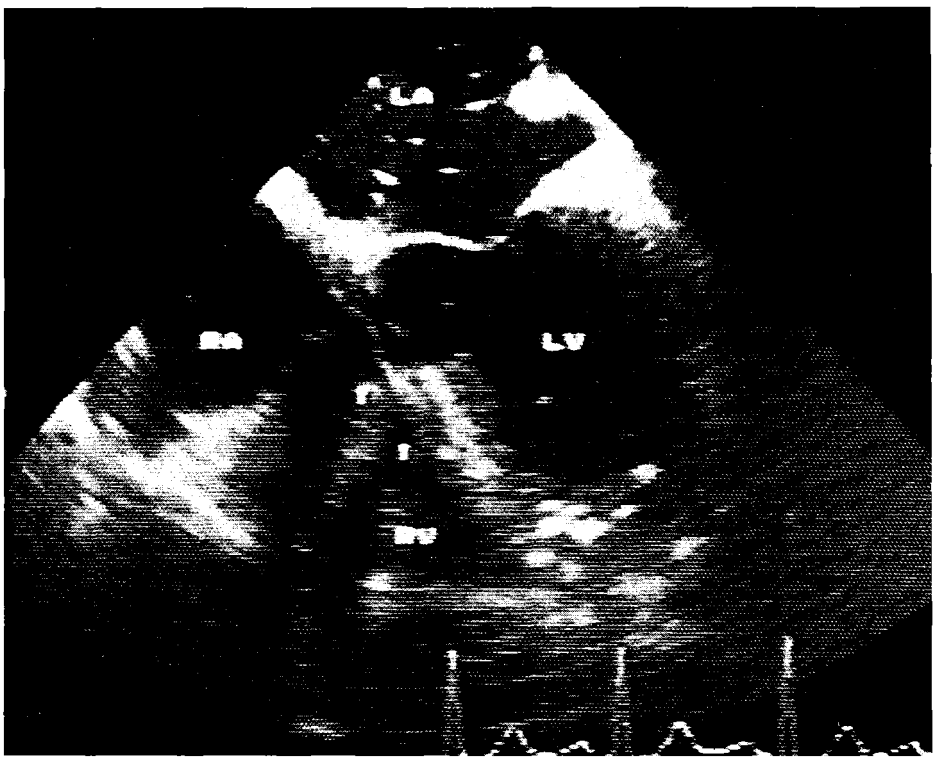

Figure 4: Transesophageal study immediate after patch closure of a ventricular septal defect in a patient with tetralogy of Fallot. Note the large areas of ultrasound masking distal to the patch, which preclude the meaningful acquisition of color Doppler information. (abbreviations see figures 1,2)

Residual ventricular outflow obstruction was suggested by epicardial colour flow mapping studies in four patients. In all four, epicardial Doppler derived pressure gradients (figure 4) were less than 20 mm Hg and no further steps were taken. Three of these patients had post bypass transesophageal studies which documented turbulence in the left ventricular outflow tract in two (no 5,15), and failed to demonstrate the obstruction in the right ventricular outflow tract in one patient after patch closure of a ventricular septal defect (no 11). Doppler evaluation of the resulting pressure gradients was unreliable using the transesophageal technique, due to the poor alignment to blood flow that could be obtained, and the availability of pulsed wave Doppler facilities only.

Assessment of ventricular function Transgastric short axis images of the left ventricle obtained by transesophageal echocardiography were used to monitor left ventricular function and volume in every patient studied. Similar views obtained from the epicardium provided an equally good assessment of ventricular function, and were able to exclude regional wall motion abnormalities. However, epicardial studies provided only a momentary picture, interfered with

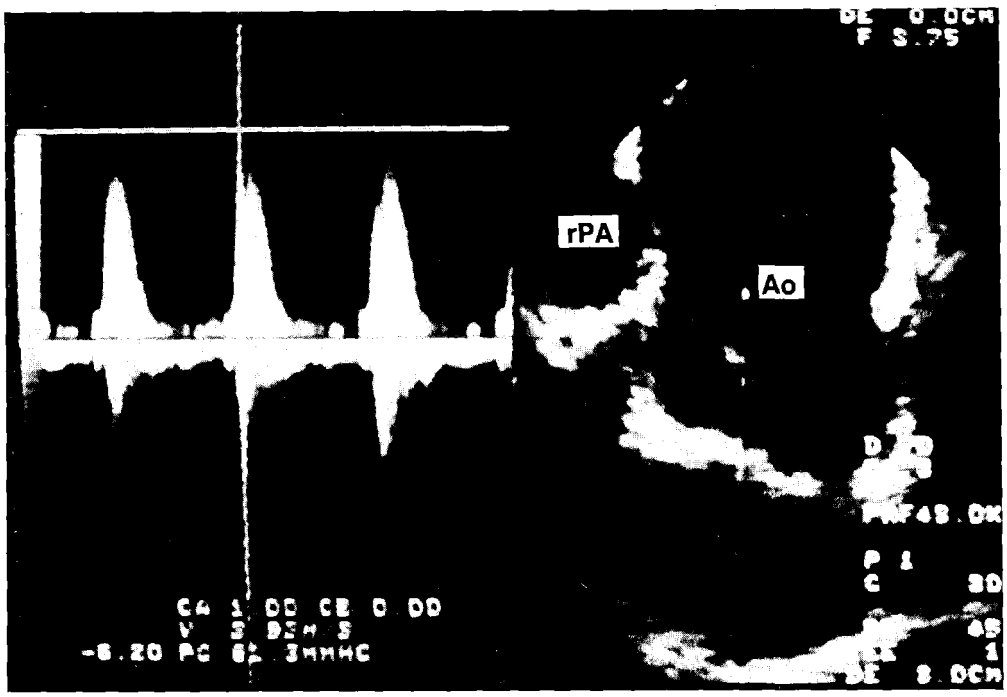

Figure 5: Epicardial assessment of the pressure gradient of a residual left ventricular outflow obstruction by placing the continuous wave Doppler probe on the ascending aorta and scanning downwards with the interrogating Doppler beam positioned through the aortic valve.
Legend: Ao = aorta; r PA = right pulmonary artery.

the surgical procedure and were not feasible during chest closure and the early post operative period. The continuous monitoring of ventricular function by transesophageal echocardiography was of particular relevance in one patient in this series who underwent a Ross procedure (no 23) who showed diffuse left ventricular wall motion abnormalities immediately after bypass and which resolved gradually during the early intensive care unit period. In this and a further patient who had a Fontan procedure (no 18) the technique was used to monitor volume replacement and inotropic support in the early post operative period.

Complications In one child with severe pulmonary hypertension the ventilation pressures increased notably following introduction of the transesophageal probe. After appropriate anesthetic intervention (oxygen, manual ventilation, anesthetic drugs) a rapid resolution of this hypertensive crisis occurred and the study was completed successfully. Complications such as esophageal trauma, esophageal bleeding and arrhythmias were never encountered.

During epicardial studies isolated ventricular premature beats were a frequent finding, and three patients had short runs of ventricular premature beats (maximum seven in succession) which ceased spontaneously. Medical intervention was never required and studies were completed in every case by choosing less arrhythmogenic epicardial sites.

Discussion

The value of intraoperative epicardial ultrasound in surgery for congenital heart disease has been well established during recent years (1-7). The principal goals of intraoperative ultrasound are to provide the cardiac surgeon with detailed morphologic information prior to the repair and to assess left ventricular filling and function and the surgical repair immediately after bypass. The majority of residual hemodynamic lesions can be detected prior to chest closure, and thus are amenable to immediate revision during the same operation on a second period of bypass. The introduction of transesophageal echocardiography means that there is now an alternative approach to the standard direct epicardial technique. Since dedicated pediatric transesophageal probes have recently become available, this new intraoperative ultrasound approach is feasible even in smaller children. Initial experience with transesophageal cross-sectional imaging in the surgery of congenital heart disease has been described by Cyran et al. who used adult transesophageal probes in children aged more than 7.5 years of age (13). Kyo et al. reported the intraoperative use of a dedicated pediatric probe which allowed cross-sectional imaging, color flow mapping and pulsed wave Doppler interrogation in 10 children (14). However no study has yet addressed the question of whether or not intraoperative transesophageal echocardiography has advantages over the direct epicardial approach in surgery for congenital heart disease.

In a prospective study we attempted to evaluate both techniques in 28 patients undergoing correction of congenital heart disease. The information obtained by both techniques was compared for 1. the pre bypass assessment of the intracardiac morphology, 2. the assessment of the surgical repair and 3. the value in monitoring ventricular function and volume. Whereas the ability of either technique to detect residual hemodynamic lesions can be quantified, the evaluation of the benefits and disadvantages of each in the definition of intracardiac morphology remains rather subjective. Table II summarizes our experience in this respect.

The adult transesophageal probe could not be inserted in two patients in this series. Potentially, the alternative use of a small pediatric probe would have allowed a study in both. Before starting this study, it was agreed that children with a body weight of less than 4.5 kilograms should not be studied. Although a successful study of a 3.9 kilogram child has been reported (14) we feel there are definite weight limits below which the transesophageal approach is inappropriate (and potentially dangerous) even when using pediatric probes. In this series transesophageal studies in children who were intubated with an uncuffed endotracheal tube while undergoing a long surgical procedure were restricted to pre bypass studies alone. This was in an attempt to avoid any potential adverse effects of leaving the transesophageal probe in place during the entire operation. Reinsertion of the probe immediately after termination of cardiopulmonary bypass was not practicable in our operating theatres.

Transesophageal studies allowed a rapid and comprehensive assessment of both the systemic venous and pulmonary venous return, the interatrial septum, and the mitral valve morphology because of the proximity of these structures to the esophagus (Table II). In contrast, the muscular ventricular septum, the right ventricular outflow tract, arterial override and the pulmonary arterial system were only poorly assessed, because of the unsatisfactory transverse axis imaging planes and the large distances from the esophagus in the adolescent and adult patients. With introduction of biplane pediatric transesophageal transducers, some of the inherent shortcomings of transverse axis transesophageal imaging may be overcome.

Table II: A comparison of intraoperative transesophageal (TEE) and epicardial (EPI) echocardiography in the assessment of intracardiac morphology and function in surgery for congenital heart disease.

	Epicardial		Transesophageal	
	morphology	function	morphology	function
Systemic venous return	+++	++	++	+
Pulmonary Venous return	++	+	+++	+++
Atrial septum	+++	++	+++	+++
Tricuspid valve	++	++	+	++
Mitral valve	+++	++	+++	+++
Ventricular septum	+++	+++	+	+[1]
Ventricular chambers	+++	++	+	+++
LVOT	+++	+++	+++	+
RVOT	+++	+++	-(+)[2]	-
Thoracic aorta	++	++	+	-
Pulmonary arteries	++	++	+[2]	+[2]

Legend: - = inadequate; + = adequate; ++ = good; +++ = excellent
LVOT = left ventricular outflow tract; RVOT = right ventricular outflow tract;
[1] = inadequate after prosthetic patch closure of ventricular septal defects,
[2] = largely dependent on the size of the acoustic window.

After cardiopulmonary bypass the transesophageal technique provided detailed insight into atrioventricular valve function and the integrity of surgical repairs performed at atrial level, but it was unreliable in excluding residual interventricular shunting or determining residual outflow tract gradients. These limitations of transesophageal echocardiography are of major relevance when estimating the utility of the technique in the surgery of congenital heart disease, as these lesions represent a large proportion of residual lesions that may warrant, and are amenable to, immediate surgical revision. Flow masking behind prosthetic material means that transesophageal color flow mapping cannot be used to exclude residual interventricular shunting. The only means of circumventing this problem would be to perform contrast echocardiographic studies while scanning the main pulmonary artery. In fact, in our experience, contrast echocardiography is more reliable than color flow mapping for grading residual shunting after ventricular septal defect closure, but it cannot demonstrate the exact site of residual shunting (17). Invasive pressure recordings could compensate for the shortcomings of intraoperative transesophageal echocardiography in the detection of residual ventricular outflow obstruction. Nonetheless, these are of limited value in determining whether a residual stenosis is anatomical rather than functional, and they cannot determine the level or the exact morphologic correlate of the obstruction. In both areas the direct epicardial approach is, in our opinion, by far superior, even when transesophageal echocardiography is combined with both contrast studies and surgical pressure recordings.

Transesophageal echocardiography proved to be of great additional value in continuously monitoring left ventricular function and volume immediately post bypass and during the early post operative period. This allows for a much improved monitoring of volume replacement and dosage of inotropic support. In addition, the technique should prove to be of considerable value

in the diagnosis and management of early postoperative complications, since precordial studies are then often largely compromised due to entrapped air and the presence of suction tubes.

It is concluded that intraoperative transesophageal echocardiography is a complementary rather than an alternative technique to direct epicardial ultrasound in surgery for congenital heart disease. The maximal value of this new technique lies in the monitoring of surgical repairs at atrial and atrioventricular level, and in the continuous monitoring of left ventricular filling and function in high risk children.

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Chapter 10

The Comparative Values of the Precordial and Transesophageal Approaches in the Ultrasound Evaluation of Atrial Baffle Function following an Atrial Correction Procedure

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Summary Methods previously used to assess atrial baffle function have included precordial ultrasound and cardiac catheterization. To evaluate if single plane transesophageal echocardiography might provide additional information, its findings were correlated with information derived from both precordial echocardiography and cardiac catheterization in 15 patients (14 Mustard-, 1 Senning procedure; mean age 16.3 years). Precordial ultrasound, using combined imaging, color flow mapping and pulsed Doppler, visualized the supra-mitral portion of the common systemic venous atrium in every case, but could only identify superior limb obstruction in 3/6 patients, mid-baffle obstruction in 0/2 and inferior limb obstruction in 0/2. Transesophageal studies, using the same range of ultrasound modalities, demonstrated superior limb obstruction in 6/6 (4 severe, 2 mild), mid baffle obstruction in 2/2 and inferior limb obstruction in 2/2 patients. The pulmonary venous atrium was equally well interrogated by either ultrasound approach, with both identifying 3 cases of mid pulmonary venous atrium obstruction (2 mild, 1 moderate). However, individual pulmonary vein velocity profiles could only be recorded using transesophageal pulsed Doppler. Precordial studies identified baffle leaks in only 3 patients (1 large, 2 small) whereas transesophageal studies identified 11 such baffle leaks (1 large, 10 small) which in 2 patients were multiple.

It is concluded that transesophageal echocardiography provides a more detailed and accurate assessment of atrial baffle morphology and function than either precordial ultrasound or cardiac catheterization. It would appear to be the investigative method which derives the most information and which is applicable on an outpatient basis in adolescent and adult patients, but requires general anesthesia in children.

Introduction

Since 1964 correction of transposition of the great arteries using an atrial baffle procedure (either a Mustard or Senning) has been the preferred surgical approach in many centers (1-3). A recently reported surgical series (3) has demonstrated a 20-year survival of 80% after such a procedure. However, following atrial correction procedures a range of late complications can occur. These include the development of right ventricular dysfunction, tricuspid insufficiency, progressive left ventricular outflow tract obstruction and life threatening arrhythmias (3-7). In addition, subacute or chronic obstruction of the systemic venous pathway in the superior baffle limb may occur and be progressive in 3-17% of the surviving patients (3,4). This is often well tolerated and clinically inapparent because of the related development of adequate collateral systemic venous drainage channels. In contrast, the late development of obstruction within the inferior limb of the systemic venous atrium is uncommon (5) although this may occur acutely in the immediate postoperative period. The late development of obstruction within the pulmonary venous pathway has been reported in 5-7% of such patients (2,3). Furthermore there is a significant incidence of persistent baffle leakage in the late post-operative period. The incidence of this has been reported to vary between 25% and 90% (4,5). Such baffle leakage was frequently unsuspected prior to cardiac catheterization.

Previous reports have suggested that a combination of two-dimensional imaging allied to contrast echocardiography, pulsed Doppler studies and color flow mapping may provide an excellent non-invasive technique to analyze baffle function (8,9). Early experiences with magnetic resonance imaging for anatomical and functional assessment of atrial baffles have been reported (10). However, these are at present limited as the definition of complex flow abnormalities by this technique is not yet widely available. Thus, where atrial baffle malfunction is suspected, cardiac catheterization remains the investigative technique of choice in the opinion of many cardiologists.

Transesophageal echocardiography is well established as a safe and practical investigative technique for acquired cardiac disease in the adult outpatient clinic (11-14). With its superb visualization of atrial morphology and the flow patterns within both atrial chambers it might now provide an alternative investigative approach to the spectrum of complex atrial baffle problems. However, it remains a technique which is inappropriate in the fully conscious child but one which can now be carried out with safety in the heavily sedated or anaesthetized child (over 5 kg) using specially designed pediatric probes. In view of the potential ability of this method to allow a better evaluation of the systemic and pulmonary venous pathways a prospective study was performed to assess any potential advantages and disadvantages which transesophageal echocardiography might have when compared to the precordial approach in patients following an atrial correction procedure.

Patients and methods

Study patients: Fifteen patients aged 4.2 to 33 years (mean age 16.3 years) were included in a prospective study to compare and contrast the relative values of precordial and transesophageal imaging in either the acute or long term follow up of patients following an atrial baffle procedure. The study protocol was approved by the Hospital Ethical Committee

(Academic Hospital Rotterdam). Fourteen patients had been followed up for 3.6 to 22.7 years (mean 11.6 years); all of whom had undergone a Mustard operation between the ages of 3 months to 11 years (Table 1). Teflon was used as the baffle material in 10 patients (cases 1-10), whereas a trouser-shaped pericardial patch was chosen in 4 patients (cases 11-14); a pericardial patch was used for enlargement of the pulmonary venous atrium in all but one patient (case 10). No patient had undergone a reoperation to refashion the Mustard baffle prior to inclusion in this series.

Seven patients were asymptomatic at the time of inclusion in this study. In four patients (cases 5,7,9 and 13) there was clinical evidence of subpulmonary stenosis, in one case (case 10) clinical evidence of pulmonary venous obstruction was present; one patient had persistent mild cyanosis at rest and signs of left ventricular hypertrophy on ECG (case 6), another had episodic paroxysmal atrial flutter (case 2). Three patients (cases 11,12 and 14) had developed symptoms of superior caval obstruction with facial and peripheral edema. The fifteenth patient - a 33 years old man with situs inversus and discordant atrioventricular and ventriculoarterial connections - had required heart transplantation after he had developed a severe dilatative cardiomyopathy. The transplantation of a levocardia donor heart necessitated the rerouting of the systemic and pulmonary venous drainage by a Senning operation; the left sided caval veins were linked to the anterior lying morphological right ventricle. One month after the heart transplantation the patient developed severe obstruction of upper and lower systemic venous inflow with associated facial edema and hepatomegaly.

Echocardiography: The precordial echocardiographic examinations were performed with either a Toshiba SSH 160 or a Vingmed CFM 700 ultrasound system using a standard cross-sectional ultrasound examination protocol. This included imaging, color flow mapping and pulsed Doppler studies which were performed with either a 3.75 or 5 MHz transducer. An attempt was made in every case to visualize the complete systemic and pulmonary venous pathways by using multiple precordial transducer positions. From both the parasternal and apical transducer positions color flow maps within the portions of the systemic venous atrium visualized were recorded and any areas of abnormal flow subsequently interrogated with pulsed wave Doppler. The subcostal view was chosen for the investigation of the inferior vena cava and interrogation of the inferior baffle limb whereas a high right parasternal or suprasternal transducer position was used for attempted visualization and pulsed wave Doppler interrogation of the superior vena cava and the area of its junction with the superior limb of the baffle. The apical four chamber view was normally used for two-dimensional imaging of the pulmonary venous pathway. Pulsed wave Doppler interrogations of flow velocity profiles at the orifice of individual pulmonary veins and at the mid-portion of the pulmonary venous atrium were recorded. Residual interatrial shunts and flow disturbances within the systemic and pulmonary venous pathways were assessed by a combination of color flow mapping and appropriate pulsed Doppler studies.

In the adult patients transesophageal echocardiography was performed on an outpatient basis using standard 5 or 5.6 MHz transesophageal probes linked to a Toshiba SSH 160, Vingmed CFM 700 or Hewlett-Packard ultrasound system. Either probe tip could be angulated in right/left and anterior/posterior directions by using the steering mechanism. No prior antibiotic prophylaxis was given as remains our routine practice. In eight children the studies were performed under general anaesthesia at cardiac catheterization using

dedicated 5 MHz pediatric probes with a maximal tip circumference of 30 mm on either an Aloka SSD 870 or Toshiba SSH 160 ultrasound system. Probe tip angulation of the pediatric probes were restricted to anterior/posterior movements.

In both, children and adults the transesophageal examination was commenced with short axis ventricular scans which were used to assess ventricular function. Then a hepatic scan was carried out in order to trace the course of the inferior vena cava, to identify the hepatic vein confluence and, by withdrawing the probe slightly and rotating it anti-clockwise, to follow the inferior limb of the systemic venous pathway towards the junction with the superior limb. Further probe withdrawal allowed the demonstration of the entire systemic venous atrium including the superior limb of the systemic venous atrium, its anastomosis with the superior vena cava and the lower portion of the superior vena cava. By advancing and rotating the probe the individual sites of drainage of all four pulmonary veins and the entire pulmonary venous pathway were scanned. A complete morphologic and hemodynamic evaluation of both venous atria was then performed using cross-sectional imaging in combination with color flow mapping allied to pulsed wave Doppler. Pulsed Doppler sampling was used to assess flow patterns in both systemic venous limbs, the pulmonary venous pathway, over both atrioventricular valves and for documentation of individual pulmonary vein flow. The entire examinations were continuously recorded on video-tape and were evaluated both on-line and later off-line at reduced speed. Twelve of the patients underwent cardiac catheterization on the same day and the remaining 3 within at most 2 to 3 months after the transesophageal echocardiographic study. The invasive data in these patients was then correlated with both the findings of the precordial and transesophageal echocardiographic examinations.

Results

It should be emphasized at the onset of this section that the shape and size of each baffle component varied according to the surgical technique employed. However, the essential ultrasound scan planes required to record a complete baffle study could be obtained by minor modifications of the transesophageal examination technique in every patient regardless of surgical technique or cardiac malposition.

The normal atrial baffle - Transesophageal imaging technique and appearance: The posteriorly located inferior vena cava and the extrahepatic confluence of the hepatic veins was best demonstrated by a transesophageal scan with the transducer positioned in a high transgastric position (Fig.1a). Then by a combination of probe withdrawal and counter clockwise rotation the region of the inferior limb of the systemic venous atrium could be visualized (Fig.1b,2). In this plane a narrow segment of the pulmonary venous atrium and the drainage of the right lower pulmonary vein could be identified lying posterior to the systemic venous atrium. Anterior to the systemic venous atrium the right atrial appendage and the anterior leaflet of the tricuspid valve were demonstrated. Further gradual rotation of the probe then allowed to scan the upper portion of the pulmonary venous atrium, the systemic venous atrium, the mitral valve and the left atrial appendage (Fig.1c). In this latter sectioning plane both left pulmonary veins were demonstrated lying posteriorly and to the left of the left atrial appendage. Further probe withdrawal and clockwise rotation opened shown in short axis views. The right upper pulmonary vein could then be visualized by

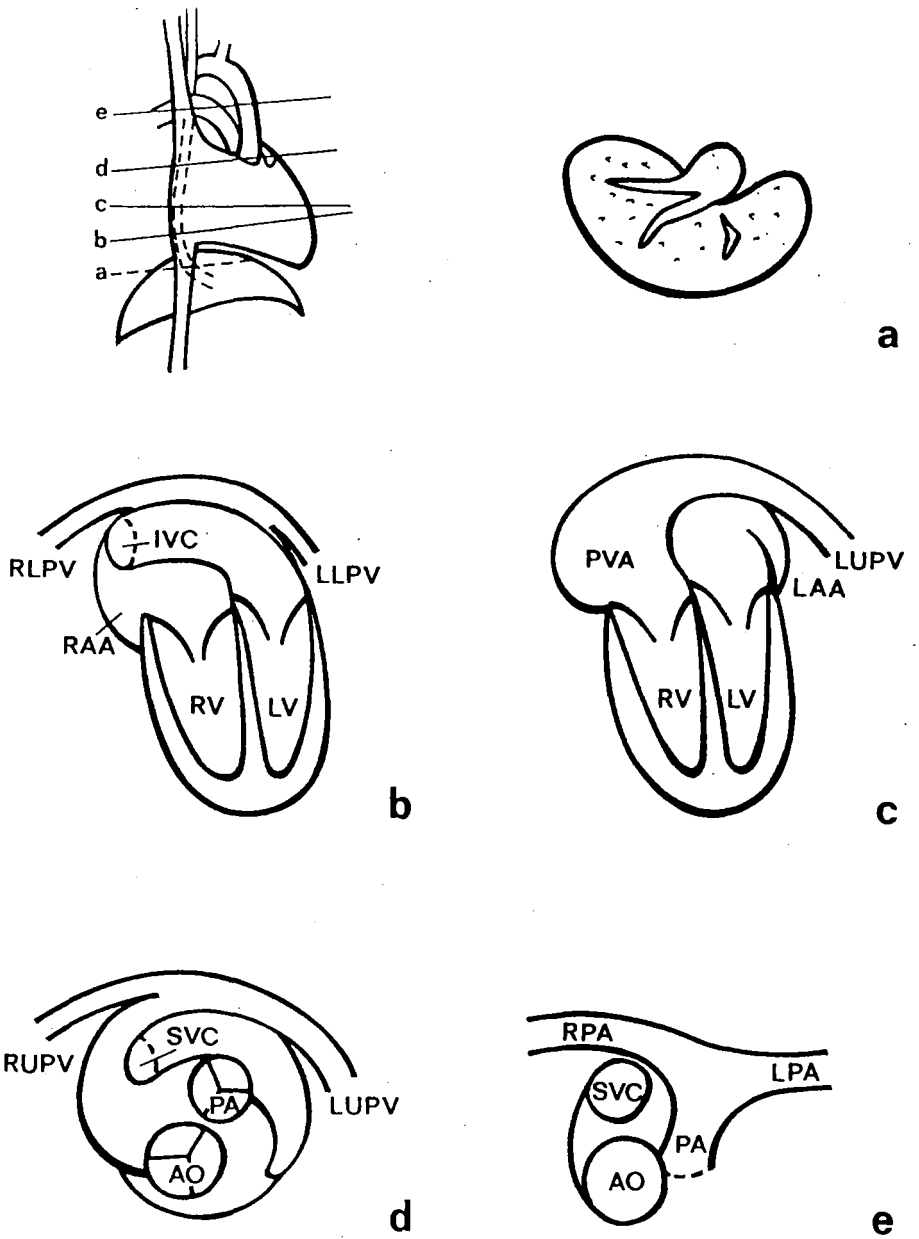

Figure 1: Transesophageal assessment of atrial baffle morphology
- diagram of the standard imaging planes (a-e).

Legend: Ao = aorta; IVC = inferior vena cava; LLPV = left lower pulmonary vein; LUPV = left upper pulmonary vein; LPA = left pulmonary artery; LV = left ventricle; PA = pulmonary artery; PVA = pulmonary venous atrium; RAA = right atrial appendage; RLPV = right lower pulmonary vein; RUPV = right upper pulmonary vein; RPA = right pulmonary artery; RV = right ventricle; SVC = left superior vena cava;

Figure 2: Anatomic cross-sections corresponding to the transesophageal imaging planes used in the assessment of the inferior limb (upper panel) and the superior limb of the systemic venous atrium (lower panel). For explanations compare with figure 1b and 1d.

up the superior limb of the systemic venous atrium demonstrating a wide segment of the pulmonary venous pathway lying posteriorly (Fig.1d,2). Anteriorly both arterial valves were shown in short axis views. The right upper pulmonary vein could then be visualized by further clockwise probe rotation. Further probe withdrawal opened up the central pulmonary arteries. In this view the superior vena cava was visualized in short axis lying anterior to the right pulmonary artery and posterior to the ascending aorta (Fig.1e).

Systemic venous atrium: The transesophageal echocardiographic assessment of the entire systemic venous atrium was complete in all 15 patients studied. The proximal portions of the superior and inferior vena cava as well as their respective junctions with the superior and inferior baffle limb of the systemic venous atrium were visualized in every case. The differing surgical techniques used in this series predisposed specific sites for obstruction within the systemic venous pathways. In 2 asymptomatic patients (cases 1 and 4) with rectangular shaped patches a mild narrowing was visualized at the transition of a medially displaced superior vena cava into the superior baffle limb. In 3 of the 4 patients with a trouser-shaped baffle and severe superior vena cava obstruction a severe stenosis was visualized at the transition of the superior baffle limb into the common portion of the systemic venous atrium (cases 11,12 and 14) (Figure 3). In these 3 patients the superior vena cava was markedly dilated and color flow mapping revealed a continuous turbulent flow pattern downstream of the obstruction detected by a continuous turbulent flow on pulsed Doppler interrogation. In patient 15 (modified Senning procedure) a marked stenosis of the superior vena cava/isthmic segment was identified which caused distension of the superior vena cava. With pulsed

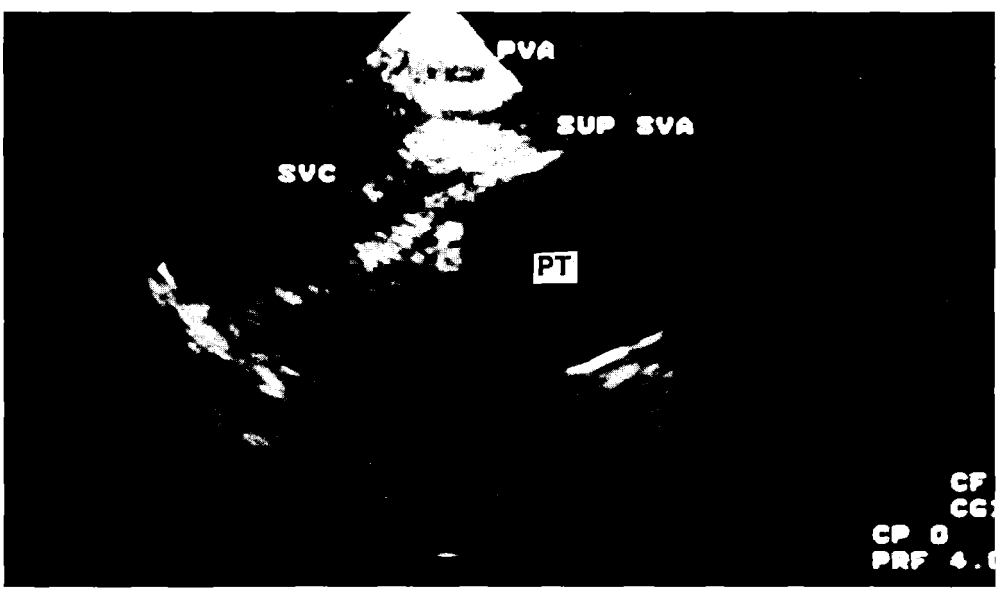

Figure 3: Transesophageal demonstration of an almost complete obstruction of the superior limb of the systemic venous atrium (SVA). The calcific stenosis and prosthetic baffle material reduces cross-sectional image quality.
Legend: PT = pulmonary trunk; PVA = pulmonary venous atrium, SVC = superior vena cava; Sup SVA = superior limb systemic venous atrium.

Doppler interrogation sampling within the systemic venous atrium just downstream to the stenotic area a continuous turbulent flow velocity waveform indicating baffle obstruction was recorded by pulsed Doppler with a peak velocity of 1 m/sec. The same patient also had mild obstruction at the transition of the inferior vena cava to the inferior portion of the systemic venous atrium. In 2 further patients (cases 1 and 2) an obstructing flap-like structure, formed by a redundant fold of baffle material, was found at the transition of the superior vena cava and superior baffle limb. Color flow mapping revealed a turbulent flow pattern that originated proximal to this structure. In the tenth patient an abnormal continuous low velocity pulsed Doppler flow pattern was recorded in the inferior baffle limb. Such continuous flow velocity profiles are normally always associated with significant intra-baffle obstruction. Eight patients (cases 3,5,6,7,8,9,10 and 13) had the appearance of a widely patent course of the systemic venous atrium on cross sectional imaging. However, despite the apparently normal imaging findings, color flow mapping demonstrated non-turbulent laminar flow pattern in the systemic venous pathway in only 3 patients (cases 3,8 and 9). In 5 patients areas of turbulent flow could be detected either in the superior or in the inferior limb of the systemic venous atrium by color flow mapping. Despite the flow disturbance, pulsed Doppler examination in every case demonstrated bi-phasic flow velocity profiles with low peak velocities. It is both the increased peak velocity of flow and the presence of continuous high velocity which indicates that baffle obstruction is present.

In contrast, using precordial ultrasound the complete systemic venous atrium could be scanned from both cavae to the mitral valve in only 1 child (case 9), in whom it appeared unobstructed on both imaging and color flow mapping. The portion of the systemic venous atrium immediately above the mitral valve could be imaged in 11 patients and had no apparent structural or flow abnormality within it in 6 patients (cases 2,3,5,6,7 and 8). In patient 11,12 and 14 the site of the clinically obvious systemic venous obstruction could be visualized to be at the transition of the superior limb into the common systemic venous atrium. In the fifteenth patient (post heart transplantation) only a short segment of both the superior and inferior limb could be demonstrated, but the findings of a turbulent flow pattern on color flow mapping within both the superior and inferior limb, taken in conjunction with the abnormal continuous flow in the pulsed Doppler velocity waveforms, confirmed the presence of two obstructions within the systemic venous pathway.

Pulmonary venous atrium: Both transesophageal and precordial echocardiographic studies permitted a complete assessment of the pulmonary venous atrium in every patient. No obstruction was noted within the pulmonary venous atrium in 12 of the patients using either ultrasound approach. Both techniques identified the same 2 patients with a mild narrowing of the isthmic segment of the pulmonary venous atrium on cross-sectional imaging (cases 4 and 5). Turbulent flow was present across both these mild narrowings on color flow mapping. With pulsed Doppler interrogation just distal to this narrowed isthmic portion in both cases biphasic flow patterns with elevated peak velocities of 1.3 and 1.5 m/sec respectively were recorded. One further patient (case 10) was found to have a marked stenosis at the mid portion of the pulmonary venous atrium on both transesophageal and precordial imaging; both methods revealed a continuous turbulent flow pattern with a peak velocity of 1.8 m/sec on pulsed Doppler interrogation (figure 4). Transesophageal color flow mapping showed all but 2 patients to have a turbulent diastolic flow pattern originating either at the isthmic

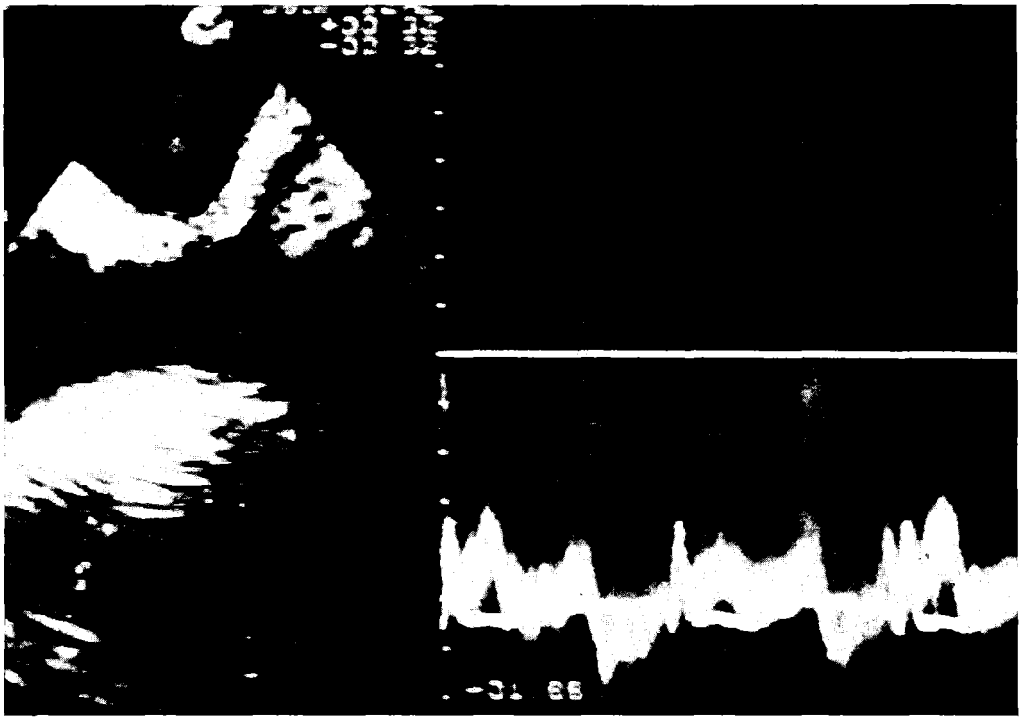

Figure 4: Moderate pulmonary venous obstruction as seen from the esophageal approach. The Doppler sample volume is placed at the site of the obstruction and reveals a continuous velocity profile (1.8 m/s), which does not reach baseline throughout the cardiac cycle.
Legend: inf SVA = Inferior limb systemic venous atrium; PVA = pulmonary venous atrium,

segment or in the posterior part of the pulmonary venous atrium. Pulsed Doppler examination of the turbulent flow in these patients revealed in every case a low velocity biphasic flow profile which was considered to constitute normal intra baffle flow.

Pulmonary vein flow: In 12 patients all 4 pulmonary veins could either be directly visualized or their return flow demonstrated on color flow mapping during the transesophageal study, whereas the precordial approach allowed interrogation of only the upper left and right sided pulmonary veins in 11 patients. It was also possible with transesophageal echocardiography to align a pulsed Doppler sample volume to flow within the pulmonary veins visualized and thus obtain flow profiles. Differing pulmonary vein flow velocity profiles were recorded in a number of patients. No patient had continuous turbulent pulmonary venous inflow indicative of pulmonary venous obstruction. A biphasic laminar flow was detected in 11 patients, in another 2 patients (cases 8 and 10) two peaks occurring during early and late ventricular systole and a third during ventricular diastole were observed. Reversal of pulmonary vein flow in early ventricular systole was found in one patient with junctional rhythm (case 2) and another with moderate tricuspid regurgitation (case 5).

Baffle leakage: Using transesophageal color flow mapping definite evidence of baffle leakage was obtained in 11 patients. Both transesophageal and precordial echocardiography identified a large interatrial communication in one patient (case 6). The wide patch dehiscence above the atrioventricular junction (figure 5) resulted in bidirectional laminar

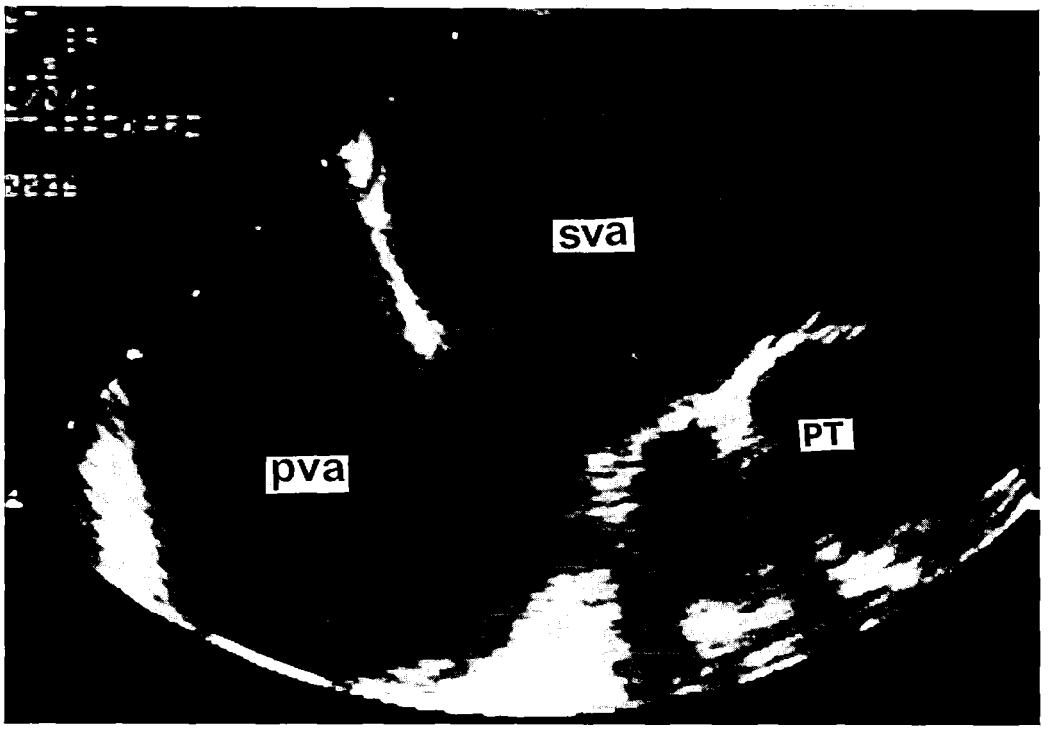

Figure 5: Large baffle leak at the site of the baffle suture line at the crux cordis, resulting in bidirectional shunting.
Legend: SVA = systemic venous atrium; others see figure 1

shunting. In 4 patients (cases 1,2,3 and 5) a bidirectional or right to left interatrial shunt was detected at the transition of the superior vena cava into the superior baffle limb. In a further 4 patients (cases 7,10,11 and 12) the shunt was found at the connection of the inferior baffle limb to the inferior vena cava and within the course of the inferior baffle suture line, respectively. In 2 further patients there were baffle leaks at both the superior and inferior limb into the pulmonary venous pathway (cases 4 and 8) In contrast, precordial ultrasound detected only 2 of these 10 patients (cases 8 and 10) to have a small residual atrial shunt on colour flow mapping.

Correlative catheterization data: Subsequent correlative catheterization data were available in all 15 patients. The transesophageal echocardiographic findings in all the patients with either systemic or pulmonary venous pathway obstruction were confirmed on both hemodynamic and angiographic investigation (Table 1). Oximetry suggested significant bidirectional shunting (systemic arterial saturation <90%) in only 2 patients. A total of 7 patients were shown to have minor degrees of residual interatrial shunting on angiography. Thus 2 patients who had definite residual baffle leaks on both transesophageal imaging and color flow mapping were missed. In addition angiography, although identifying that baffle leakage was present, did not allow to distinguish between single and multiple baffle leaks in the two patients in whom there were multiple leaks.

Table I: Correlative results in the assessment of atrial baffle dysfunction.

Lesion	PE	TEE	Cath
Systemic venous obstruction			
- inferior limb	0/2	2/2	2/2
- mid baffle	0/2	2/2	2/2
- superior limb	3/6	6/6	6/6
Pulmonary venous atrial obstruction	3/3	3/3	3/3
Baffle leakage	3/11	11/11	9/11

Legend: Cath = cardiac catheterization/angiocardiography; PE = precordial ultrasound; TEE = transesophageal echocardiography.

Discussion

Deterioration in the function of atrial baffles may develop both early and late following an atrial correction procedure (15,16). Severe baffle obstruction may occur insidiously in clinically asymptomatic patients. Therefore a sensitive and non-invasive method for long-term postoperative assessment of atrial baffle function is required. In young children precordial ultrasound studies using a combination of cross-sectional imaging and pulsed Doppler studies from the suprasternal, parasternal and subxiphoid windows have been extensively used for this purpose (8,9). However, in many older children, adolescents and adults following intracardiac repair the precordial ultrasound windows are frequently restricted, thus limiting any attempted complete evaluation of the systemic and pulmonary venous pathways. Even in small children the precordial approach cannot consistently visualize either the junction of the venae cavae with the atrial baffle or a large portion of the inferior limb of the systemic venous atrium. In addition, although the whole intra-atrial pulmonary venous atrium is clearly demonstrated from the praecordium, the evaluation of individual pulmonary vein flows by either precordial imaging or pulsed Doppler is restricted at best to interrogation of one or both upper pulmonary veins. In contrast, transesophageal echocardiography consistently allowed a complete assessment of both the entire systemic venous and pulmonary venous pathways.

Systemic venous atrium: In all of our patients - 7 children and 8 adults - the transesophageal approach provided a much more detailed analysis of the morphology of both the superior and inferior baffle limbs and their junction with the venae cavae when compared to the precordial approach. The systemic venous pathways could be visualized in their entirety, whereas from the precordial approach this was possible in only one child. Twelve patients in this study demonstrated a turbulent flow pattern in their systemic venous atria on color flow mapping. The correct site and the morphology of the obstruction as it was present in 10 patients could be determined in every case at the transesophageal study. In comparison, the prior precordial study had only demonstrated systemic venous obstruction in 3 of these cases. In cases where both ultrasound approaches defined the presence of systemic atrial obstruction the transesophageal images were of far greater quality and the sectioning planes available from this approach allowed a much more accurate evaluation of the morphology of the obstruction.

Previous reports have suggested that pulsed Doppler is a sensitive technique for use in the detection of systemic venous pathway obstruction. Such obstruction may be identified by using precordial pulsed Doppler in a number of differing ways. The presence of a non-phasic disturbed flow pattern in the jugular veins can reliably indicate the presence of systemic venous return obstruction but does not demonstrate either the site or the morphology of the obstruction(17,18). Contrast echocardiographic studies have been reported as reliable in detecting partial or complete superior vena caval or superior baffle limb obstruction when the contrast injected into a peripheral arm vein is visualized to enter the heart from the inferior vena cava (19). However, this method is again non-specific for both the site and morphology of the obstruction. In comparison, the results of this study would suggest that the definition of the site and the morphology of any obstruction within the superior or inferior portion of the systemic venous pathway can consistently be identified by transesophageal imaging.

Pulmonary venous atrium: From the precordial approach a two-dimensional echocardiographic assessment of the pulmonary venous atrium can reliably demonstrate an intra-atrial pulmonary venous pathway obstruction. This normally occurs at its isthmic portion. Pulsed Doppler interrogation of the flow velocity profile across this area proved to be accurate in predicting significant obstruction when there was a change in the pulsed Doppler flow pattern from a normal biphasic low velocity to a continuous turbulent higher velocity flow profile (20,21). However, differentiation of the level of obstruction - either at mid portion of the pulmonary venous atrium or at the orifice of an isolated pulmonary vein - cannot be reliably determined from the precordial approach. Furthermore, as pulsed Doppler is often not well aligned with the blood flow direction the true pressure gradient at the isthmic portion is likely to be underestimated. As documented in our study, the entire intra-atrial pulmonary venous atrium could be equally well visualized in every patient by transesophageal and precordial ultrasound. However, the advantage of the esophageal approach was in the close proximity of the transducer to the atria, which allowed additional information to be gathered on low velocity flows. Using transesophageal color flow mapping a diastolic flow disturbance was detected in virtually all the patients with unobstructed pulmonary venous atria within the posterior or isthmic portions of the pulmonary venous atrium. Whereas intra-atrial obstruction within the pulmonary venous pathway was equally well assessed using either ultrasound approach, the site of drainage of all four pulmonary veins, and their individual pulsed Doppler flow patterns were only reliably documented from the transesophageal approach.

Pulmonary vein flow: No patient with documented individual pulmonary vein obstruction was encountered; the majority of the patients studied demonstrated a bi- or triphasic low velocity pulmonary vein flow pattern both within the veins and at their site of drainage into the pulmonary venous atrium (22). We would consider this to represent the normal pulmonary venous return flow profile in patients in this group who have good right ventricular function, no severe tricuspid incompetence and no pulmonary venous obstruction. In the patient with moderate isthmic pulmonary venous atrium obstruction a biphasic Doppler tracing was obtained from the pulmonary veins indicating that pulmonary venous return still was influenced by pressure changes within the supra-tricuspid portion of the pulmonary venous atrium.

Baffle leakage: With the exception of 1 patient with a very large interatrial communication which was visualized on both precordial and transesophageal studies the accurate identification of both the anatomical site and shunt direction of a baffle leak could only be derived by transesophageal color flow mapping and pulsed Doppler interrogation in 10 patients. Only 2 of these patients were suspected to have residual atrial shunts on precordial examination. Thus, transesophageal echocardiography proved to be an extremely sensitive method of detecting a baffle leak.

We would conclude from this initial experience that transesophageal echocardiography has proved to be of great additional value in the definition of a wide range of unsuspected baffle abnormalities after atrial correction procedures. The transesophageal approach routinely allowed a much more detailed insight into abnormal baffle morphology and its related hemodynamic alterations than did the precordial ultrasound approach. Such baffle studies are clearly appropriate in the outpatient clinic as part of the routine follow up in the adolescent and adult age group but their precise place in children in whom heavy sedation or general anaesthesia is required remains to be evaluated.

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Chapter 11

Transesophageal echocardiography in the evaluation and management of the Fontan circulation

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Summary Transesophageal echocardiography was used in 18 patients (age 1.6-34 years (mean age 12.6 years) to assess the immediate (5 patients) or intermediate results (13 patients) following a Fontan-type procedure. The findings were correlated with precordial echocardiographic (all patients) and cardiac catheterization data (11 patients).

Atrial shunting was documented by transesophageal studies in 3 patients (precordial in 1). This was confirmed in two by cardiac catheterization; the third was reoperated based on the transesophageal study alone. Thrombus formation was detected by transesophageal studies in three patients (precordial in 1); repeat studies were used to evaluate thrombolytic therapy in two. Atrioventricular valve regurgitation (11/18) was better defined by transesophageal than by precordial studies (5/18). A coronary artery fistula was identified in two cases (precordial in none). Pulmonary artery obstruction was documented in 3 patients (precordial in 1), and was confirmed by subsequent cardiac catheterization in all. Evaluation of anterior Fontan connections was successful in 5/8 patients (precordial in 6/8), and posterior connections in 10/10 patients (precordial 5/10). Glenn shunts could be evaluated in 8/9 patients (precordial in 3/9). Transesophageal pulsed Doppler interrogation of pulmonary artery and pulmonary vein flow patterns consistently allowed a detailed evaluation of the Fontan circulation.

Transesophageal echocardiography is an important diagnostic and monitoring technique in Fontan patients. In this series, it proved to be greatly superior to precordial ultrasound evaluation and of substantial additional value to cardiac catheterization.

Introduction

Several modifications of the original Fontan procedure (1) are applied for the correction of a wide spectrum of complex congenital heart lesions (2-5). Although the early and late operative results have improved dramatically over the recent decade (6-8), a high proportion of patients have hemodynamic lesions (2-9). Such lesions may not always be clinically

apparent, but may be the cause of impaired functional capacity (10-12). Whereas some lesions are residuae from suboptimal surgical correction and require early reoperation (e.g. atrial shunting), others may develop during the late post operative period (e.g. pulmonary artery obstruction), and may lead to either acute or gradually progressive clinical deterioration (13,14). Thus a close follow-up of all Fontan patients is required. Precordial ultrasound studies are used to assess the functional status of Fontan patients (15,16), but these are often limited in the information that can be obtained, particularly in adolescents and adults. Thus, to effect a complete evaluation of the Fontan circulation, the clinician frequently has to undertake cardiac catheterization.

Transesophageal echocardiography has become an established technique for use in the diagnosis and management of acquired cardiac lesions in the adult patient population (17-19). With the availability of dedicated pediatric probes, this technique is now rapidly evolving to become a diagnostic technique even in small children (20-22). Since the cardiac structures which are in close proximity to the esophagus routinely are better assessed by transesophageal than by precordial ultrasound, the technique might be expected to provide additional insights into the Fontan circulation.

In this report transesophageal echocardiography was prospectively used in a series of patients to determine its diagnostic value in the evaluation of Fontan-type procedures. The results were compared with the findings of correlative precordial ultrasound studies and cardiac catheterization.

Patients and Methods

Patients: Eighteen Fontan patients were studied by transesophageal echocardiography. The age at investigation ranged from 1.6 to 34 years, with a mean age of 12.6 years; the body weight ranged from 9.8 to 82 kilograms (mean 39.7 kilograms). Five patients were studied in the immediate postoperative period and twelve during the early or long term follow-up period. The mean interval between the operation and the transesophageal study was 3.4 years; the longest follow-up interval was 12 years. (see Table I). Eight patients were symptomatic at the time of the transesophageal study.

The Fontan operation had been performed for the correction of tricuspid atresia in 8 patients, for mitral atresia in 1 patient, for double inlet left ventricle in 5, and for other forms of complex congenital heart disease in 4 patients. Eight patients underwent atriopulmonary Fontan connections (anterior in 5, posterior in 3). A valved Hancock conduit was used in 3 of these patients. Seven patients had a total cavopulmonary anastomosis and three had a right atrial to right ventricular connection (valved in 2, non-valved in 1). A Glenn anastomosis, either unilateral or bilateral, was performed in 9 patients (see Table I).

Precordial studies: Complete precordial echocardiographic examinations (16), using the full range of precordial, subcostal and suprasternal scan positions, were attempted in all patients prior to the individual transesophageal studies. Particular attention was paid to fully demonstrating the systemic venous pathways using combined subcostal, parasternal and suprasternal scan positions, and also the connection(s) of the systemic venous return to the pulmonary arterial system. Cross-sectional imaging was followed by color flow mapping studies and pulsed wave Doppler sampling in the relevant areas of interest. In addition, continuous wave Doppler interrogation was carried out for the assessment of atrioventricular

Table I: Patient data

Pt No	age (yr)	weight (kg)	sex	interval	Malformation	Fontan connection	valved	Glenn shunt
1	22	58	f	8 yrs	TA	post RA-PA	yes	/
2	19	53	f	8 yrs	TA	conduit RA-RV	yes	right
3	20	67	m	12 yrs	TA	ant RA-PA	yes	right
4	12.7	49.5	f	1.3 yrs	DILV	ant RA-PA	no	right
5	6.2	22.2	m	periop	DILV	TCPC	/	right
6	23	69	m	10 yrs	TA	conduit RA-RV	yes	/
7	2.4	13.0	f	periop	DILV	TCPC	/	right
8	2.0	9.8	m	periop	TA	post RA-PA	no	/
9	10.3	31	m	7.6 yrs	AVSD,DORV	ant RA-PA	no	/
10	1.6	12.3	f	periop	AVSD,DORV	TCPC	/	/
11	4.0	10.5	m	3 weeks	TA	ant RA-PA	no	/
12	13.6	44	f	2 weeks	TA	conduit RA-RV	no	/
13	3.3	12.9	m	1 week	AVSD,DORV	TCPC	/	bilateral
14	24	72	m	9 yrs	TA	ant RA-PA	yes	/
15	2.8	11.5	f	periop	TGA	TCPC	/	right
16	4.8	18.2	f	1 yr	DILV	TCPC	/	bilateral
17	34	78	m	3 yrs	DILV	post RA-PA	no	/
18	21	82	m	1.4 yrs	DILV	TCPC	/	right

Legend: ant = anterior; AVSD = complete atrioventricular septal defect; DILV = double inlet left ventricle; DORV = double outlet right ventricle; f = female; m = male; periop = perioperative period; post = posterior; RA-PA = atriopulmonary; RA-RV = atrioventricular; TA = tricuspid atresia; TCPC = total cavopulmonary connection;

valve regurgitation and aortic valve velocity waveforms. Precordial contrast echocardiographic studies were not performed routinely.

Transesophageal studies: Informed patient or parental consent was obtained for each study, and hospital ethical approval (Academic Hospital Rotterdam) was granted prior to commencement of the study protocol. All studies were performed using single plane transverse-axis probes.

Nine children were studied under general anesthesia, given either in the early post operative period (5 patients) or during a concurrent cardiac catheterization (4 patients). One child (no 10) was studied on two occasions. In a further two children (no 11,12) transesophageal studies were performed electively under general anesthesia prior to hospital discharge. In ten children a prototype 48 element 5 MHz pediatric transesophageal probe (developed by the Department of Experimental Echocardiography, Thoraxcenter Rotterdam) was used connected to either a Toshiba SSH 160 A or a Hewlett Packard Sonos 500 or 1000 ultrasound system. A 24 element 5 MHz pediatric probe (Aloka Company Japan) was used in one child on an Aloka SSD 870 ultrasound system. The probe dimensions of either probe were comparable with a maximal shaft diameter of 7 mm and a maximal tip circumference of 30 mm (5 x 10 and 7 x 8 mm respectively). Steering facilities were restricted to anterior-posterior tip angulation only.

Seven adult patients were studied on a total of twelve occasions using either a 5 or 5.6 MHz adult transesophageal probe connected to a Toshiba SSH 160 A or a Hewlett Packard Sonos 1000 system. Mild sedation (Midazolam) given intravenously was used for 5 of these studies.

Study protocol A standard transesophageal examination procedure was adhered to in every case. The connection of the inferior caval vein with the right atrium and the hepatic veins were assessed by scanning a series of transverse axis views of the liver with the transducer positioned in the stomach near the esophageal hiatus. By gradually withdrawing the probe a series of views of the right atrial cavity was obtained. The integrity of the atrial septum or the suture line of the intra atrial patch used for total cavopulmonary connections (5) was assessed by additional color flow mapping studies. The communication between the right atrium and the pulmonary arterial system was visualized on cross-sectional imaging and was then evaluated by combined color flow mapping and pulsed wave Doppler interrogation. The right pulmonary artery was searched for in its normal position lying posterior to the superior caval vein, the left pulmonary artery anterior to the descending aorta. Pulsed Doppler sampling was performed within both pulmonary arteries proximal to their first branching. The site and function of a Glenn shunt was documented on combined color flow mapping and pulsed wave Doppler studies. A right Glenn shunt was demonstrated on high short axis views at the level of the right pulmonary artery. A left Glenn shunt was demonstrated when the probe was withdrawn (above the left main bronchus) and rotated minimally counter-clockwise from the location to scan the left atrial appendage. The main pulmonary artery was demonstrated on scan planes which included the proximal ascending aorta, using anterior tip angulation. The site of drainage of all four pulmonary veins was documented by a combination of cross-sectional imaging and color flow mapping. Subsequent pulsed wave Doppler interrogation of pulmonary venous return from right and left sided pulmonary venous return was carried out and traces were recorded on registration paper at

a speed of either 50 or 100 mm/s. The function of the atrioventricular valve(s) was assessed on combined color flow mapping and pulsed wave Doppler studies from transesophageal four-chamber positions. Ventricular function was assessed by scanning transgastric short axis views of the ventricle together with M-mode recordings.

Cardiac catheterization: Eleven patients underwent cardiac catheterization for evaluation of the Fontan circulation. These included measurements of pressures and oxygen saturations within the caval veins, the right atrium, the pulmonary arterial system, the aorta and the left ventricle. Pulmonary artery wedge pressures were recorded in all patients. Retrograde left atrial and selective pulmonary venous pressure recordings could be obtained in four patients. Cardiac output was measured by dye dilution technique. For the evaluation of a classical (unidirectional) right sided Glenn shunt an additional internal jugular venous catheterization was required in three patients. Biplane angiocardiograms were performed routinely of the right atrium, the pulmonary arterial system and the left ventricle. Aortic root angiograms were carried out in eight patients.

Results

A total of twenty-four transesophageal studies were performed and completed successfully in the 18 patients. Complications were encountered in only one child (no 9), who underwent simultaneous cardiac catheterization, and had a supraventricular tachycardia of about 2 minutes duration. This was self-terminating and no medical or electrical therapy was required. In the remaining 17 patients no complications occurred.

Residual atrial shunting was documented in three patients during the transesophageal study. In all three a continuous right to left atrial shunt with turbulent flow characteristics was demonstrated on color flow mapping studies. Two of these had this finding subsequently confirmed at cardiac catheterization (no 4,13). The third patient (no 10), in whom significant shunting was detected, underwent early reoperation on the basis of only the transesophageal study. During surgical inspection in the latter case the predicted site of shunting, located at the mid portion of the suture line of the patch with the atrial septum was confirmed (figure 1). A prior precordial contrast study with injection into the right atrial line had revealed atrial shunting, but failed to define the exact site of the leakage. In the remaining 14 patients the transesophageal study excluded residual atrial shunting. This was subsequently confirmed in all 8 of the remaining eight patients in whom cardiac catheterization and angiocardiography were carried out.

Atrioventricular valve incompetence was demonstrated on transesophageal color flow mapping studies in eleven patients. Prior precordial studies detected atrioventricular valve regurgitation in only five of these. Both the extent and the origin of the regurgitant jet were better defined by the transesophageal studies. A central regurgitant jet was documented in nine, an eccentric regurgitant jet in two. One patient with a common atrioventricular valve (no 13) had both central and eccentric regurgitation. Three patients were judged to have moderate valvar regurgitation, no patient had severe regurgitation. The perfect closure of the right sided atrioventricular valve in one patient (no 4) who had an atriopulmonary connection for double inlet left ventricle could be better demonstrated by the transesophageal than by the precordial study.

Coronary artery fistulae were documented on transesophageal color flow mapping

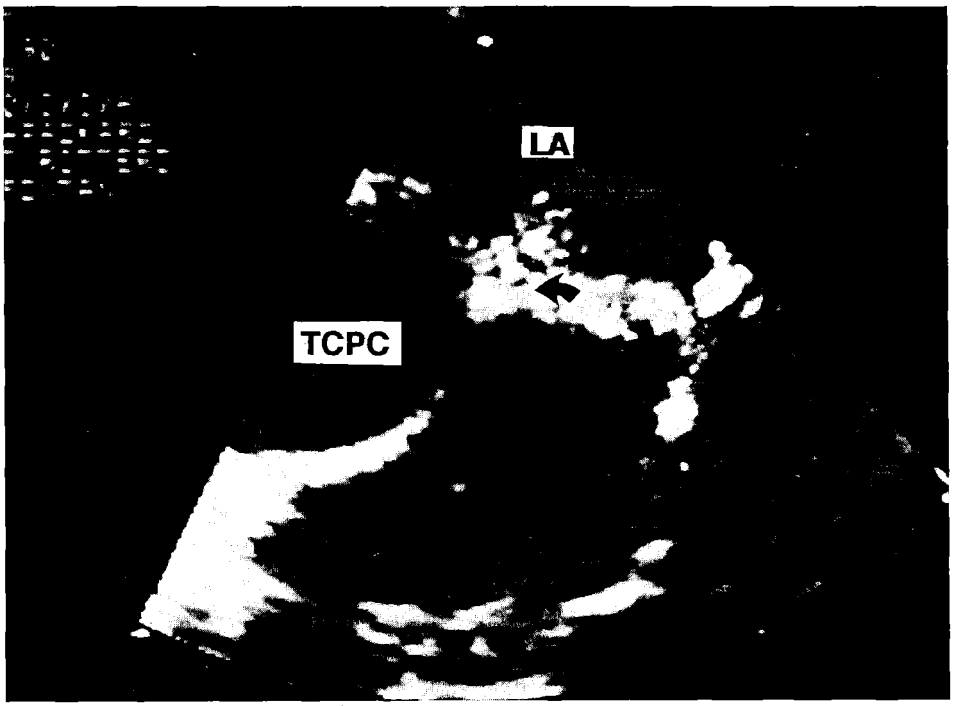

Figure 1: Residual atrial shunting after total cavopulmonary connection (TCPC). The site of the defect at the mid suture line of the atrial baffle with the atrial septum (arrow) was confirmed during subsequent reoperation.
Legend: LA = left atrium.

studies in two patients (no 10,16). In both, the fistula originated from the right coronary artery system and drained into the functional left atrium. In neither patient was the finding judged to represent a significant shunt. Prior precordial ultrasound investigations had not detected these in any patient. The finding was confirmed in one patient who subsequently had an aortic root angiogram (no 10), and was not demonstrated by left ventricular angiography in the other.

Thrombus formation within the right atrial cavity (2 patients) or a total cavopulmonary connection (1 patient) was documented in three patients by the transesophageal study. In one of these (no 3), who presented with acute clinical deterioration associated with atrial fibrillation, the diagnosis had been missed during a prior precordial study and the patient was cardioverted acutely on the intensive care unit. Thereafter his condition deteriorated further and inotropic support and mechanical ventilation was required. The transesophageal study carried out at that time revealed extensive right atrial thrombus formation (figure 2). No flow was recorded over his valved right atrial to pulmonary artery connection nor within the left pulmonary artery. Embolization of the left pulmonary artery or acute thrombotic occlusion of the Hancock conduit was the presumed cause. Pulmonary venous return from the left lung, as assessed by transesophageal pulsed wave Doppler studies, was minimal. The patient was commenced on thrombolytic therapy using intravenous recombinant tissue plasminogen activator for 3 days. A control transesophageal study was carried out to

determine the result of thrombolysis. This was judged to be successful with no evidence of residual thrombus in the atrial cavity and a return to normal in pulmonary artery and pulmonary venous flow patterns. Cardiac catheterization and lung scintigraphy confirmed the transesophageal hemodynamic findings. The patient subsequently underwent elective total cavopulmonary connection, with a perfect result as assessed by a repeat transesophageal study.

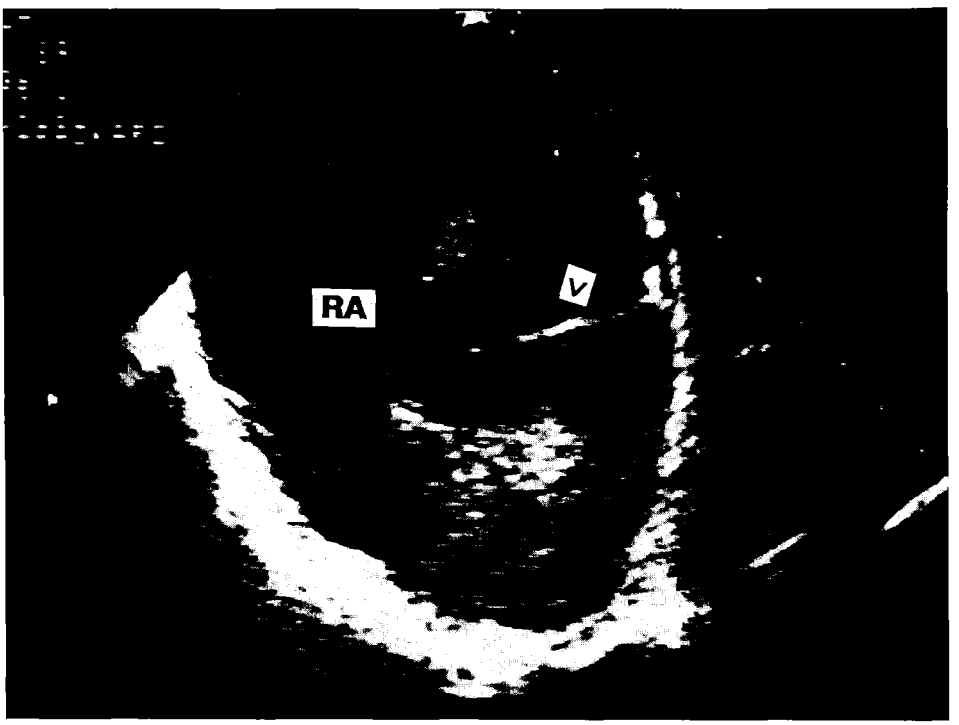

Figure 2: Extensive right atrial thrombus formation, not diagnosed during prior precordial study. Note the prominent Eustachian valve (arrow head).
Legend: RA = right atrium

In the second patient, who was studied on an outpatient basis (no 14), thrombolytic therapy was started on an elective basis after transesophageal studies documented excessive mural thrombus formation. A repeat study was carried out to document the final result, which showed only minimal change in thrombus morphology. The third patient (no 18) was studied during the routine postoperative follow-up. Slow flow, characterized by spontaneous contrast generation, together with small thrombi were documented within his total cavopulmonary connection. These changes were not noted during the prior precordial evaluation. The patient received intravenous heparin for 4 days, and the repeat transesophageal study excluded residual thrombi, although spontaneous contrast was still observed. In an additional two patients (no 9,13) with low cardiac output, as determined by dye dilution, the appearance of spontaneous contrast echos within the right atrial cavity was documented. However, there was no evidence of thrombus formation on a detailed imaging study.

Obstruction to pulmonary blood flow was assessed by scanning the Fontan connection

and the central pulmonary arterial system. Anterior connections or anastomoses (direct atriopulmonary or conduit atrioventricular) could be visualized in 5 of 8 patients, posterior connections (direct or conduit atriopulmonary, and total cavopulmonary anastomoses) in 10 of 10 patients. Prior precordial studies allowed an adequate visualization of anterior connections in 6 of 8 patients and of posterior connections in 5 of 10 patients. Of the total of 29 connections, 14 could be evaluated by precordial ultrasound studies (48 %), whereas this was achieved in 25 by transesophageal studies (86 %) (figure 3).

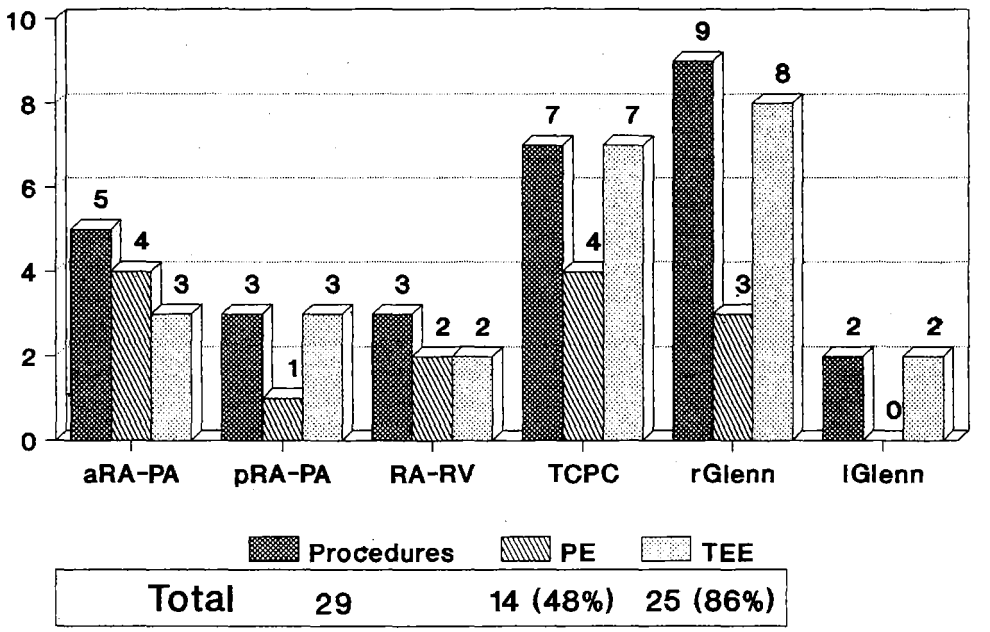

Figure 3: Evaluation of 29 Fontan connections and Glenn shunts by precordial (PE) and transesophageal (TEE) studies.
Legend: aRA-PA = anterior atriopulmonary connection; pRA-RV = posterior atriopulmonary connection; RA-RV = atrioventricular connection; TCPC = total cavopulmonary anastomosis; r Glenn = right sided Glenn shunt; l Glenn = left sided Glenn shunt;

Pulmonary artery obstruction was defined by the transesophageal studies in 3 patients. In only one of these patients this was detected by the prior precordial ultrasound study (no 10). In two patients (no 9,10) the site of obstruction could be demonstrated on cross-sectional imaging and color flow mapping. In the remaining patient (no 3), the documentation of absence of pulmonary venous return from the left lung revealed the diagnosis. Cardiac catheterization confirmed the transesophageal findings in all three patients. Central pulmonary artery obstruction could reliably be excluded by transesophageal assessment of flow patterns within the distal right and left pulmonary arteries (proximal to their first peripheral branching) in 13 of the remaining 15 patients, and was confirmed in all of the remaining eight patients who had subsequent cardiac catheterization.

The function of a Glenn anastomosis could be evaluated in seven of eight patients by the transesophageal color flow mapping and pulsed Doppler study. In one patient (no 4) interposition of the right main bronchus precluded the evaluation. Prior precordial studies allowed

adequate assessment of Glenn shunts in only three patients (no 7,15). A bidirectional left Glenn shunt, present in two patients (no 13,16) could be evaluated in both by the transesophageal study, but in none by the prior precordial study (figure 3). Obstruction of the Glenn shunt was excluded by transesophageal pulsed Doppler interrogation distal to the anastomosis in all patients. This was confirmed in all of the five patients who had subsequent cardiac catheterization. The presence of arteriovenous fistulae was identified on transesophageal color flow mapping studies in two patients (no 4,16), however, the course and the number of fistulae could not be defined.

Pulmonary blood flow was assessed by transesophageal pulsed wave Doppler sampling within the branch pulmonary arteries and within the corresponding pulmonary veins. Subsequent correlation of these traces with those obtained from the contralateral lung revealed total obstruction to right pulmonary artery flow in one (no 9), and occlusion of the left pulmonary artery in a second patient (no 3). In both patients pulmonary venous traces demonstrated an elevated peak retrograde velocity and diminished antegrade systolic and diastolic forward flow, reflecting only to and fro flow within the pulmonary veins of the obstructed side (figure 4). In a further two patients time velocity integrals of the systolic and diastolic phases of pulmonary venous return were largely reduced when compared with the contralateral lung. In one of these (no 10) mild obstruction to the right pulmonary artery at the anastomotic site had been already demonstrated on the transesophageal imaging and color flow mapping study. In the second patient (no 13) no cause for decreased left pulmonary blood flow was detected, but the angiogram revealed a delay and reduction of left pulmonary artery opacification.

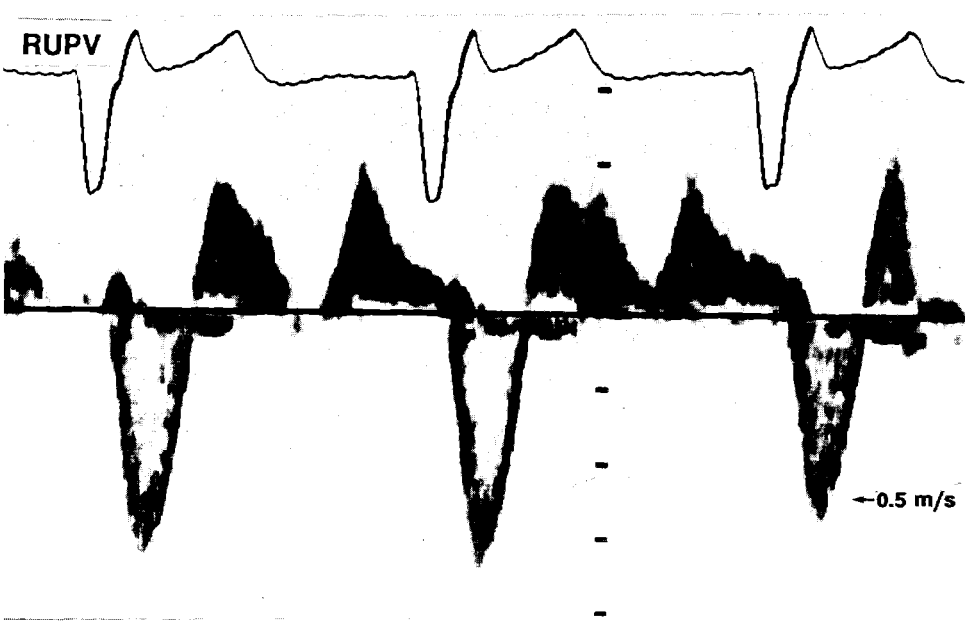

Figure 4: Pulsed wave Doppler tracing of the right upper pulmonary vein (RUPV) in a patient with occlusion of the right pulmonary artery, resulting in to and fro flow within the pulmonary veins of the right lung. The net forward flow approximates zero.

Pulmonary artery flow patterns varied considerably depending on the type of Fontan connection. Following total cavopulmonary anastomosis pulmonary artery flow was continuous and at low velocity but showed marked respiratory changes. The peak velocity of forward flow increased with inspiration and was reduced to almost zero velocity with expiration. Superimposed to these major respiratory changes were minor, but rapid velocity changes during the cardiac cycle. This pattern of flow was found unchanged but largely augmented distal to a left Glenn shunt in one child with interrupted inferior caval vein and hemiazygos continuation (figure 5).

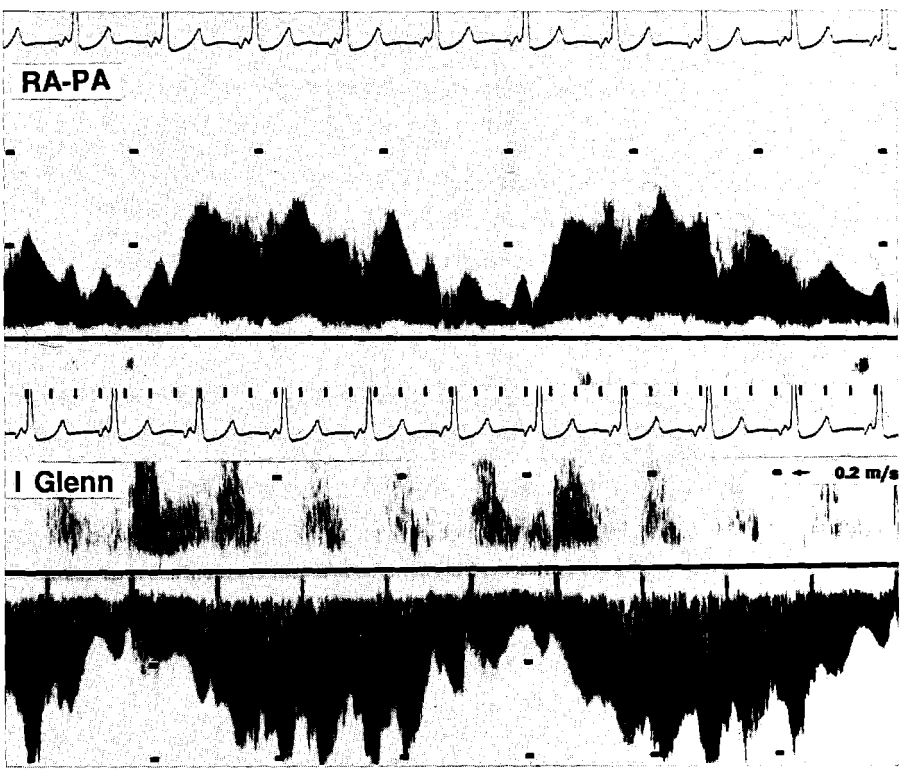

Figure 5: Pulsed wave Doppler study in a patient with a total cavopulmonary connection and bilateral, bidirectional Glenn anastomoses for correction of a complex cardiac lesion associated with hemiazygos continuation of the inferior caval vein. Sampling at the site of the connection towards the left pulmonary artery (RA-PA) reveals low velocity flow with marked respiratory variations. Distal to the anastomosis of the left sided Glenn shunt (l Glenn) the pattern of flow remains comparable, but flow velocities are largely increased (forward flow away from the transducer).

Following atriopulmonary connections a biphasic pulmonary artery flow pattern was observed. Transient retrograde flow during early systole was documented in three patients at the site of the connection. In another patient (no 9), a marked retrograde flow was noted in the distal third of the left branch pulmonary artery. Maximal antegrade flow velocities were documented following atrial contraction (0.5 m/s), with a second (smaller) peak of

forward flow occurring at the end of systole. There were marked respiratory changes of the traces (figure 6). The finding of retrograde flow within the distal third of the branch pulmonary artery was interpreted to be related to a high pulmonary artery pressure. At subsequent cardiac catheterization a right atrial a wave of 36 mm Hg and a mean atrial pressure of 28 mm Hg were documented. There was no obstruction to left pulmonary artery flow (mean pressure 28 mm Hg). Left atrial mean pressure measured 7 mm Hg, with an a wave of 10 mm Hg.

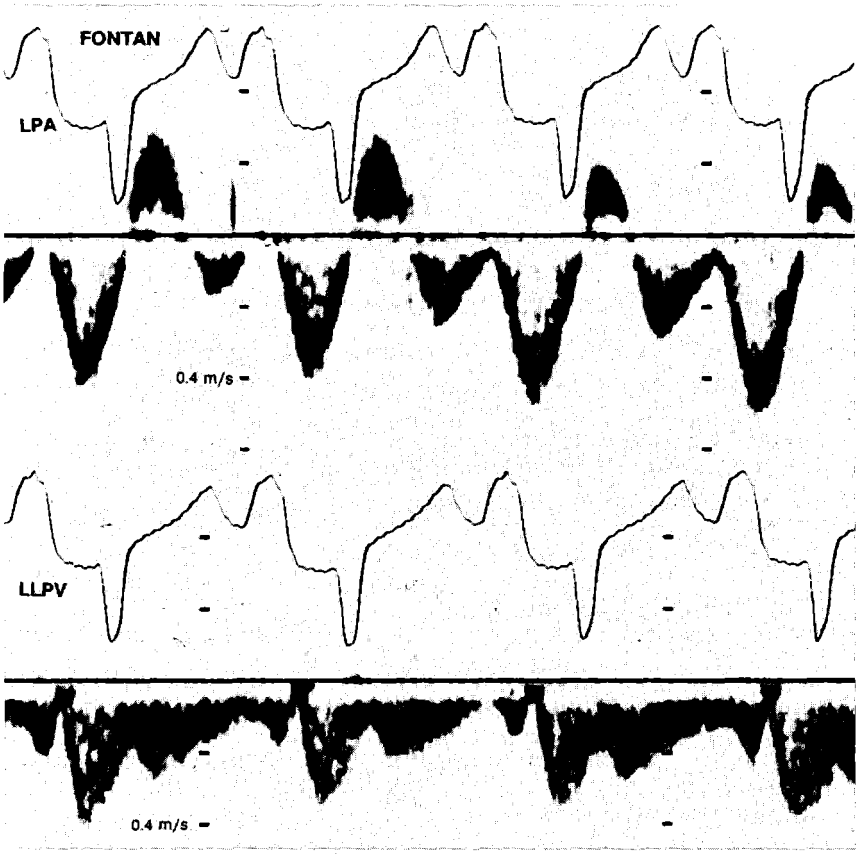

Figure 6: Pulsed wave Doppler study in a patient with atriopulmonary connection in the presence of raised pulmonary artery pressures. Forward flow (away from the transducer) in the distal left pulmonary artery (LPA) peaks following right atrial contraction, and reverses during atrial relaxation. End systolic forward flow is presumably passive. The corresponding flow pattern within the left lower pulmonary vein (LLPV) demonstrates a biphasic low velocity profile. Due to cardiac rotation antegrade flow is away from the transducer. Note the absence of retrograde flow following atrial contraction.

In two patients with atrioventricular connections filling of the conduit was noted in both diastole and systole, whereas forward flow in the pulmonary arteries was noted only during systole with retrograde flow throughout diastole. The corresponding pulmonary vein flow pattern demonstrated near normal biphasic flow patterns (figure 7). These findings suggested that the right ventricular chamber incorporated in the Fontan circulation in these two patients

contributed to systolic forward flow, but functioned as a reservoir without pulmonary valve closure, during diastole.

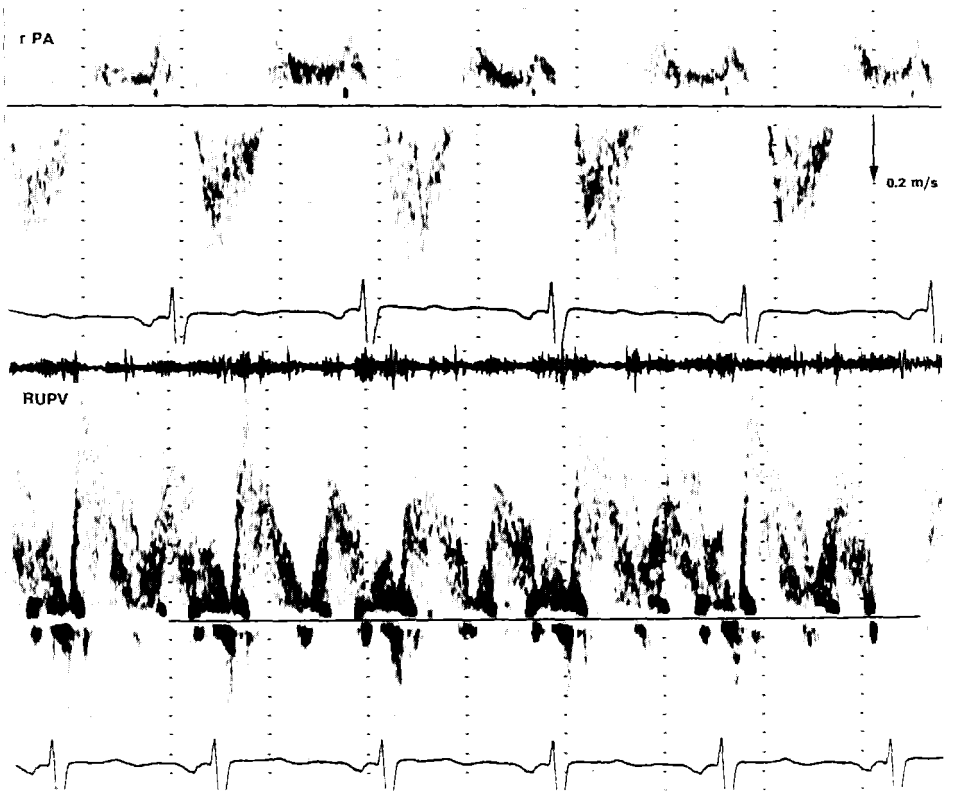

Figure 7: Right pulmonary artery (RPA) and right upper pulmonary vein (RUPV) flow patterns in a patient with valved atrioventricular Fontan connection. Note the retrograde flow (towards the transducer) within the pulmonary artery throughout ventricular diastole occurring simultaneously with antegrade flow in the pulmonary vein.

Ventricular function was studied using transgastric short axis scans of the ventricle(s) together with M-mode studies. High quality images of the endocardium could be obtained in only 13 of the 18 patients studied. This was partly related to marked liver enlargement in the majority of cases. No further analysis was undertaken. The precordial approach allowed a better assessment of ventricular function in ten patients.

Discussion

A high proportion of the patients who have undergone a Fontan-type procedure have residual or acquired hemodynamic lesions such as persisting atrial shunting or obstruction to pulmonary blood flow. The latter abnormality is in most cases acquired, and is found particularly frequent following Fontan procedures involving an extracardiac conduit (6,13,23). Obstruction to pulmonary blood flow may occur insidiously in the asymptomatic patient.

However, a high proportion of these patients will not present until the onset of secondary sequalae such as acute atrial rhythm problems or right atrial thrombus formation (9,24).

Precordial ultrasound studies in the follow-up of such patients are frequently incomplete, especially in older patients or when the Fontan connection lies posteriorly. The extensive use of prosthetic material in total cavopulmonary anastomoses (5), frequently limits a reliable assessment of the connection even in younger children. Contrast echocardiographic studies, used for the detection of residual atrial shunting, are difficult to interpret in the presence of Glenn shunts or arteriovenous fistulae. Irrespective of the surgical procedure employed, a reduced precordial image quality is often encountered in Fontan patients. This limits the detection of spontaneous contrast and thrombi within the right atrium. The distances between the chest wall and the pulmonary arteries and pulmonary veins, together with the poor alignment of the precordial Doppler beam to flow within these structures, limits the acquisition of high quality pulsed wave Doppler signals, which would allow the non-invasive evaluation of the Fontan circulation. Thus, the majority of Fontan patients who are suspected of having residual or acquired lesions undergo (repeat) cardiac catheterization in order to effect a complete assessment of the hemodynamic results.

Transesophageal echocardiography was used in this report in a series of patients to determine its diagnostic value in the evaluation of the Fontan circulation. The findings were compared with those obtained by precordial ultrasound studies and the data derived from cardiac catheterization and angiocardiography. When compared to precordial ultrasound studies transesophageal studies provided a much improved image quality. This was irrespective of the patient's size or the presence of entrapped air or suction tubes in the early perioperative period. The atrial chambers and the morphology and function of the atrioventricular valve(s) could be assessed in more detail from within the esophagus in all patients studied. This allowed the detection of a variety of unsuspected hemodynamic lesions, such as atrial shunting in two, atrioventricular valve regurgitation in six and a coronary artery fistula in two patients. In all those patients in whom the prior precordial studies suggested atrial shunting or mitral regurgitation, the transesophageal color flow mapping study provided superior information, allowing for an accurate localization of the origin of these flow disturbances. Good hemodynamic results with only minimal pathologic changes, such as minimal mitral regurgitation, were documented in ten of the eighteen patients. This was confirmed in all three of these who underwent scheduled cardiac catheterization.

Thrombus formation was detected by the transesophageal study in three of the eighteen patients. This suggests that thrombus formation after a Fontan procedure may be more frequent than previously reported (24,25). Spontaneous contrast within the Fontan connection, which is indicative for an increased thromboembolic risk (26), was more readily diagnosed by the transesophageal approach. Because of the deleterious effects that thromboembolic events will cause in the Fontan circulation, anticoagulation should be considered in patients who show spontaneous contrast, and thrombolytic therapy may be attempted in those with evident thrombi or embolization (25).

Posterior Fontan connections could be evaluated in all ten such patients studied, thus making the transesophageal approach clearly superior to the precordial approach in this patient group. In addition, it was only the transesophageal approach that routinely allowed the visualization of distal segments of the right and left pulmonary arteries, the evaluation

of Glenn anastomoses, and the visualization of all four pulmonary veins. This in turn allowed for a detailed Doppler evaluation of the Fontan circulation in fifteen of the eighteen patients studied. Anterior connections could not be visualized and evaluated adequately in three of the eight patients in whom these were performed. The distance of these connections from the esophagus, together with the restrictions in imaging related to interposition of the bronchial tree between the esophagus and anteriorly placed conduits, were the major limiting factors for single plane transesophageal studies. The recent adjunct of longitudinal or biplane transesophageal imaging (27,28) may reduce these difficulties, but they are unlikely to be completely eliminated.

Pulsed wave Doppler studies could be performed at multiple sites for the exclusion of any obstruction to pulmonary blood flow and to determine the precise hemodynamics of the various types of Fontan circulation. Good angles of incidence for the transesophageal Doppler beam to flow could be obtained for posterior connections, both branch pulmonary arteries and the corresponding pulmonary veins. Obstructions at the site of the Fontan anastomosis were documented by the finding of continuous turbulent flow throughout the cardiac and respiratory cycle. Total occlusion of one pulmonary artery was demonstrated by only to and fro flow within the corresponding pulmonary vein(s). Although these patterns of flow would appear to be diagnostic, care has to be employed during their interpretation since they provide only information on blood flow velocity but not volumetric data.

Pulmonary venous flow patterns, which reflect left heart pressure events (29), were found to be biphasic in all patients studied, irrespective of the type of surgical procedure used. In patients with atriopulmonary connections, similar biphasic flow patterns were documented in the branch pulmonary arteries. The velocities of forward flow may peak following atrial systole. Retrograde flow within the pulmonary arteries, in these patients, is an inconstant finding, that is probably related to the relaxation of the right atrium in the presence of raised pulmonary artery pressures (see figure 6).

Following total cavopulmonary anastomosis, which result in an almost complete exclusion of the right atrium, near normal biphasic pulmonary vein flow patterns were documented. However, the pulmonary artery flow patterns in these patients demonstrated only minor variations in flow velocities during the cardiac cycle. These were superimposed over the marked changes caused by respiration (figure 5,8).

The comparison of the observations in patients with either atriopulmonary or total cavopulmonary connections support the theory that the principal determinant factor for the Fontan circulation lies in the systemic venous pressure, which, among others, is related to systemic ventricular function (30-33). The finding of retrograde flow within the pulmonary arteries occurring simultaneously with antegrade flow within the pulmonary veins (see figures 6,7) excludes a "suction effect" of the left ventricle to drive the Fontan circulation. Variations in pulmonary artery flow patterns are reflections of, firstly, changes of left atrial pressure events, which are transmitted retrogradely through the pulmonary capillary bed, secondly, the presence and function of a right atrial or ventricular chamber, and thirdly, of intrathoracic pressure changes occurring during respiration.

Several recent studies (34,35) have indicated that assessment of pulmonary vein flow traces together with the assessment of mitral valve inflow patterns may allow a more detailed insight into the (diastolic) function of the systemic ventricle. In this respect, transesophageal

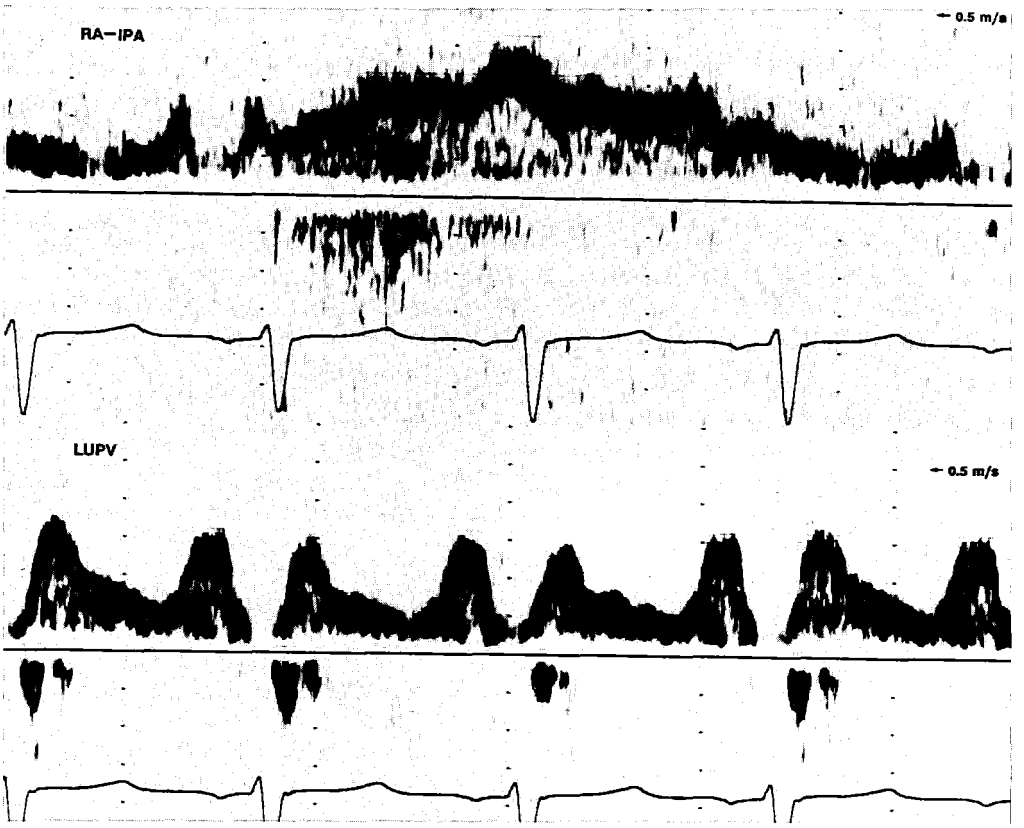

Figure 8: Pulsed wave Doppler study in a patient with total cavopulmonary connection. Sampling at the site of connection between the right atrium and the left pulmonary artery (RA-lPA) documented a continuous low velocity flow pattern with marked respiratory changes, however, the flow pattern in the left upper pulmonary vein (LUPV) is biphasic.

Doppler echocardiography may provide new insights into the ventricular function of Fontan patients, which is one of the key determinants in the long term result.

Conclusion Transesophageal echocardiography provides a detailed insight into the Fontan circulation. The information derived from these studies is of great value both in the surgical and medical management of early or late post operative sequelae. In this series the technique was superior to precordial ultrasound and was of substantial additional value to cardiac catheterization. The results indicate that transesophageal echocardiography may become the diagnostic technique of first choice in the follow-up of Fontan patients.

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Chapter 12

- General Discussion -

Transesophageal echocardiography recently has become available for investigation of even small children (1). Over a one year period we used this new diagnostic modality in a series of 127 children with congenital heart disease to determine its value in firstly, the primary diagnosis, secondly, as a perioperative monitoring technique, and thirdly, in the follow-up of congenital heart disease. The principal diagnoses of the children studied are listed in figure 1. The age at investigation ranged from 0.2 to 14.8 years (mean age 5.1 years); the weight from 3.7 to 52 kilograms (mean weight 18.7 kilograms). Seventy-nine children weighed less than 20 kilograms.

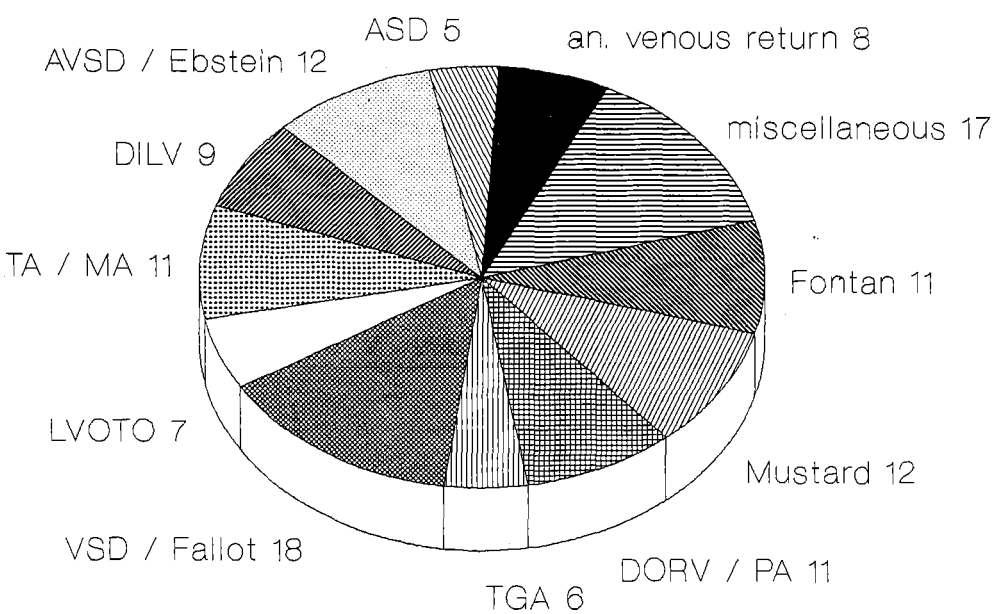

Figure 1: Principal diagnoses of 127 children studied during a one-year period.
Legend: an. = anomalous; ASD = atrial septal defect; AVSD = atrioventricular septal defect; DILV = double inlet left ventricle; DORV = double outlet right ventricle; LVOTO = left ventricular outflow obstruction; MA = mitral atresia; PA = pulmonary atresia; TA = tricuspid atresia; TGA = complete transposition.

Dedicated pediatric single-plane (transverse axis) transesophageal probes were used during the entire series. The information obtained was correlated with the results obtained by precordial ultrasound studies (all patients), cardiac catheterization and angiocardiography (in 109 patients), epicardial ultrasound (in 31 patients) and surgical inspection (in 72 patients).

Safety of transesophageal studies in children Complications were encountered in 3 of the 127 children, of whom two weight less than 5 kilograms. In one child with pulmonary hypertension a hypertensive crisis was induced following probe introduction. Appropriate anesthetic management resolved the crisis and the study could be completed uneventful. The second child, studied in the follow-up after a Fontan procedure, underwent a supraventricular tachycardia of about 2 minutes duration. This resolved spontaneously and did not require any medical management. The third child, a heavily cyanosed baby of 3.7 kilograms, underwent electro-mechanical dissociation, and required resuscitation. No patient died. No signs of esophageal bleeding or trauma were encountered in any patient. Thus, transesophageal echocardiography, using the current equipment, should be considered a safe technique in the anesthetized child above 5 kilograms of weight. Studies in children below this weight limit should only be undertaken, in cases where the information likely to be obtained by such studies is mandatory for patient management.

The value of transesophageal echocardiography

I. in the primary diagnosis of congenital heart disease. The technique, for the first time, allows the direct visualization of both atrial appendages. Thus, by observing their unique morphologic characteristics (2), the direct diagnosis of atrial situs is feasible. Neither cardiac malrotation nor malposition of the atrial appendages, such as juxtaposition, are limiting factors. Although indirect methods used in the definition of atrial situs have been shown to be accurate in the vast majority of cases (3,4), there are cases with discord between the bronchial and the atrial arrangement (5,6). In addition, the documentation of (hemi)azygos continuation of the inferior caval vein on subcostal ultrasound scans is not necessarily indicative of left atrial isomerism (3), thus requires further investigation. The definitive documentation of the atrial arrangement is of major relevance to the clinician for the exclusion of associated lesions and for the surgeon for optimal planning of the surgical repair (6,7). Finally, juxtaposition of the atrial appendages, which may go unrecognized during precordial ultrasound investigations, may alter the surgical approach in patients scheduled for either a Fontan or a Mustard procedure (8,9). Transesophageal studies in children with these suspected conditions, in our opinion, should be considered the ultimate diagnostic test. However, the indication for routine preoperative investigation should be based on the exclusion of associated lesions.

Transesophageal echocardiography proved to be a highly sensitive technique in the documentation of anomalous venous connections to the heart. Both anomalous systemic venous and pulmonary venous connections are readily identified. Thus, the technique may influence the need and the practice of cardiac catheterization in selected patients. Nonetheless, the major contribution lies in the definitive exclusion of this range of lesions in cases where these are suspected or would alter the surgical approach considerably. All four pulmonary veins can be documented either on cross-sectional imaging or color flow mapping in 91 % of the cases, whereas this is routinely not possible by precordial ultrasound techniques (10,11). Fine

morphologic details of the atrial septum relative to the site of venous connections, which are of relevance to the surgeon in planning the repair, can be documented with ease. In addition, individual pulmonary vein stenosis can be rapidly excluded by pulsed wave Doppler interrogation. Thus, it would appear that transesophageal studies used in the definition of the venous connections to the heart are a valuable diagnostic adjunct to both precordial ultrasound studies and cardiac catheterization.

Abnormalities of the atrioventricular junction can be assessed in great detail by the transesophageal approach. In particular, the morphology of the atrioventricular valve(s) and their subvalve apparatus frequently can be evaluated in greater detail than by the precordial approach. The technique would appear to be the diagnostic modality of choice in the exclusion of chordal straddling. In cases with a double inlet ventricle, studies may be used in the preoperative evaluation when a septation procedure is considered. The underlying morphology of an incompetent atrioventricular valve frequently is better documented by transesophageal studies. However, transesophageal imaging remains inadequate in delineating fine morphologic details in those cases with gross enlargement of the atrial chambers, such as in Ebstein's malformation of the tricuspid valve or severe mitral stenosis. The hemodynamic evaluation of the atrioventricular valves by pulsed wave Doppler interrogation is limited to the assessment of the ventricular inflow patterns. The mitral valve flow can be examined under optimal angles of incidence, whereas this often is difficult for the tricuspid valve. The potential of transesophageal pulsed Doppler studies to acquire both atrioventricular valve and pulmonary venous flow patterns is likely to provide new insights into the ventricular (diastolic) function of children with congenital heart disease (12,13).

Left ventricular outflow obstruction in children with congenital heart disease can be produced by a variety of lesions (14). Although fibromuscular membranes are most often the cause of these lesions, abnormal chordal insertions of the subvalvar apparatus of the mitral valve, contributing to the obstruction, are a frequent finding. Transesophageal cross-sectional imaging is a superb technique to delineate these fine morphologic details, which are of crucial importance to the surgical approach. In addition, the technique is far superior to precordial ultrasound in the evaluation of patients who have undergone a Mustard or Senning procedure, in whom residual or progressive left ventricular outflow obstruction is a frequent finding. Nonetheless, the resulting pressure gradient can only be determined by precordial continuous wave Doppler studies or cardiac catheterization.

Indications for transesophageal studies in unoperated children would appear to be in the assessment of the venous connections to the heart, in the definition of the arrangement and the morphology of the atrial chambers and the atrial septum, in the evaluation of atrioventricular junction abnormalities or in the delineation of left ventricular outflow obstruction. Taken into consideration the safety of this new technique in children with more than 5 kilograms of body weight, transesophageal studies, in our opinion, should become an integral part during the preoperative cardiac catheterization of children with complex congenital heart disease involving these cardiac levels. Performed as such, these studies do not require for additional anesthesia, they do not prolong cardiac catheterization. They can contribute additional valuable information, that may influence the catheterization procedure (e.g. left heart catheterization via a patent foramen ovale), and be of substantial value for patient management and the planning of surgical correction.

II. as a monitoring technique. When compared to epicardial ultrasound techniques in surgery for congenital heart disease (15-17), transesophageal echocardiography potentially has several advantages. Firstly, it does not interfere with the surgical procedure, secondly, there is no potential risk to induce bacterial pericarditis or mediastinitis, and thirdly, the technique would allow for a continuous monitoring during the early post operative period. However, the image quality obtained intraoperatively is currently still inferior to that obtained by direct epicardial scanning. In addition, the range of scan planes provided by single-plane transesophageal imaging is strictly limited. In particular the assessment of the right ventricular outflow tract and the great arteries is largely limited when compared to the epicardial approach. The post bypass exclusion of residual outflow tract obstruction or residual ventricular shunting following prosthetic patch closure of ventricular septal defects by transesophageal studies, in our opinion, remains inadequate. Although recently there has been much interest in the use of the technique in a wide range of surgery of congenital heart disease (18-20), in our opinion the epicardial approach will remain the standard intraoperative monitoring technique. It is possible in virtually all children, and the majority of studies can be performed by the cardiac surgeon, thus reducing manpower requirements. In selected cases, such as after a Fontan procedure, a combination of both epicardial and transesophageal monitoring may be advantageous. The major impact of perioperative transesophageal echocardiography would appear to be in the diagnosis and management of early post operative complications.

The monitoring of interventional cardiac catheterization appears to be a further indication for transesophageal studies in children. Although, to date, no studies have described the value of the technique in the wide range of catheter interventions currently used in the pediatric patient population, our own initial experience reflects that the technique is of additional value in the guidance of these procedures, in the immediate exclusion of related complications and in the assessment of the hemodynamic results [unpublished data]. It provides real-time imaging and hemodynamic information without interference or prolongation of the procedure itself. The technique proved to be of major value in the monitoring of balloon dilatations of venous pathway obstructions, such as after a Mustard procedure, in the monitoring of guidewire and catheter position relative to the atrioventricular valves and their subvalvar apparatus, and in the immediate exclusion of aortic regurgitation after each balloon inflation during aortic valvuloplasties. The monitoring of duct device occlusion was not enhanced by transverse-axis transesophageal imaging. Recently its value in the monitoring of device closure of atrial septal defects in an adult population has been described by Hellenbrandt and associates (21). It may be anticipated that both further developments in the imaging technology and more detailed studies on the atrial septal morphology will make transesophageal studies an integral part both in the selection of patients and the monitoring of atrial septal defect device closure.

III. in the follow-up of congenital heart disease. Precordial ultrasound studies in the operated child frequently are cumbersome and often remain incomplete in the information that can be obtained. Fibrous tissue adhesions together with the natural reduction in the size of the precordial and subcostal ultrasound windows with age frequently preclude a detailed assessment of the morphology and function of surgical repair at atrial level. A majority of children who have been operated for complex congenital cardiac lesions have to undergo repeat

cardiac catheterization for a detailed follow-up evaluation.

Following a Mustard or Senning procedure for complete transposition, the precordial assessment of the systemic venous pathways is strictly limited. In contrast, transesophageal studies allow for a complete assessment of the entire atrial baffle (both the systemic and pulmonary venous pathways), the exclusion of individual pulmonary vein obstruction and a detailed insight into associated lesions such as left ventricular outflow obstruction. When compared to cardiac catheterization and angiocardiography in this group of patients, transesophageal echocardiography provides the same sensitivity in detecting pathway obstruction, and is superior in the detection of small or multiple baffle leaks (22). Thus, the technique would appear to be the diagnostic technique of choice in the assessment of atrial baffle morphology and function. Thus, it may obviate the need for cardiac catheterization in selected cases and reoperation may be performed based on the transesophageal findings alone. When compared to magnetic resonance imaging in this group of patients (23) several advantages of the transesophageal approach become apparent. Firstly, it provides real-time imaging and hemodynamic information, and secondly high quality studies can be performed in virtually all patients including those with a pacemaker.

Following a Fontan-type procedure in children with complex congenital heart disease, the transesophageal studies provide a much improved evaluation of the morphology and function of posterior Fontan connections and Glenn shunts, when compared to precordial ultrasound studies. Anterior Fontan connections are equally well assessed. Residual hemodynamic lesions on atrial level are more readily detected by transesophageal studies, as is the appearance of spontaneous contrast within the right heart chambers or the presence of evident right heart thrombi. Thus, such studies may be expected to become a routine part in the follow-up of Fontan procedures and will help to reduce the need for serial cardiac catheterization. Transesophageal pulsed wave Doppler studies provide a detailed insight into the pulmonary circulation after various modifications of the original Fontan procedure. Flow patterns of pulmonary venous return is found to be biphasic in all patients. This was found to be independent of the type of Fontan connection, a fact hitherto unobserved. Pulmonary artery flow patterns vary widely dependent on the presence and function of an incorporated right atrial or ventricular chamber, major pressure changes within the left atrium, and the effects or respiration. The comparison of different pulmonary artery flow patterns after various modifications of the Fontan circulation suggest that the systemic venous pressure is the principal determinant force in driving the Fontan circulation. The comparison of pulmonary artery and pulmonary vein flow patterns with those of the contralateral lung provides clues in the detection of abnormal lung perfusion. Together with atrioventricular valve inflow patterns, pulmonary vein flow profiles may give additional information on ventricular function in Fontan patients.

Additional information: When compared to precordial ultrasound studies and cardiac catheterization, transesophageal echocardiographic studies provided new morphologic or hemodynamic information in 68 of the 127 children studied (53 %) (figure 2). Important new information for patient management was provided by these studies in 26 children (20 %). Surgery was carried out on the basis of the transesophageal findings alone in 3 patients. In those who subsequently underwent surgical correction the additional morphologic information

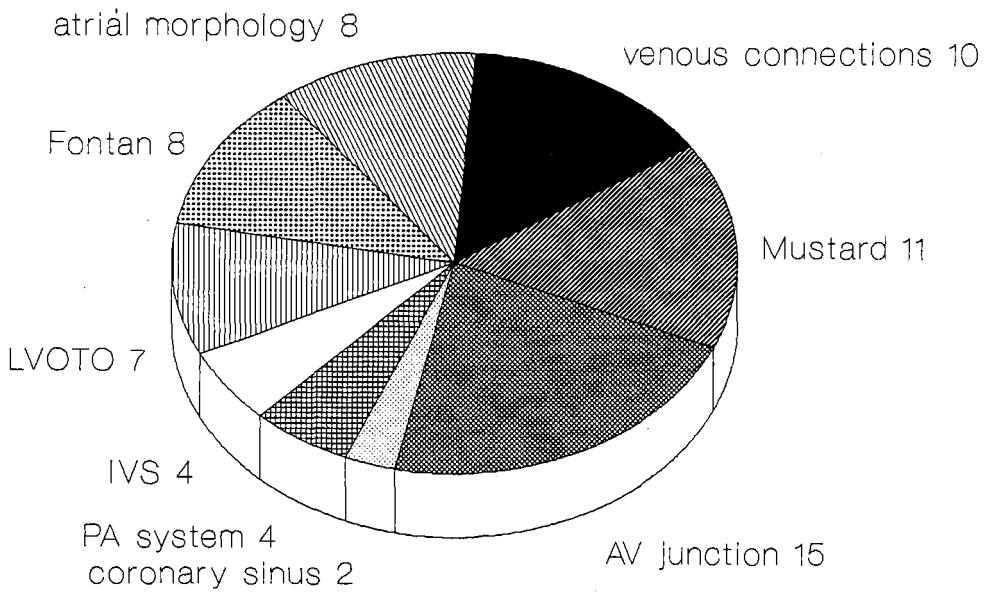

Figure 2: Additional hemodynamic and morphologic information derived from transesophageal studies, when compared to precordial ultrasound.
Legend: AV = atrioventricular; IVS = interventricular septum; LVOTO = left ventricular outflow obstruction; PA = pulmonary artery.

that was obtained was of value to surgical decision making. Relatively little information was derived on such cardiac structures as the ventricular chambers, the ventriculo-arterial junction and the morphology and function of the great arteries. These findings should largely influence the indications for such studies (Table).

Limitations: Although transesophageal echocardiography provides a new ultrasound window to the heart which provides excellent insight into the cardiac structure closest to the esophagus, there are several limitations of the technique in the assessment of congenital cardiac lesions. These include, firstly, the poor demonstration of the cardiac structures at great distance from the esophagus, secondly, ultrasound masking by interposition of the bronchial tree, and, thirdly, the limited hemodynamic information that can be derived from such studies. The integrity of the apical interventricular septum frequently cannot be assessed accurately. Pathologic changes of the right ventricular outflow tract such as infundibular stenosis are not well demonstrated by transverse-axis imaging. In children with marked ventricular hypertrophy, or left atrial dilatation, the pulmonary artery is often elevated in respect to the bronchial tree, thus limiting the range of transesophageal ultrasound information to be obtained. A wide segment of the ascending aorta cannot be visualized from within the esophagus. Finally, transverse axis scan

Table: Indications for TEE studies in children with congenital heart disease

Primary diagnosis	anomalous pulmonary venous connections anomalous systemic venous connections atrial arrangement juxtaposition of the atrial appendages sinus venosus atrial septal defects multiple atrial septal defects atrioventricular junction abnormalities left ventricular outflow tract obstruction
Monitoring	
- *perioperative*	surgical repair at venous and atrial level atrioventricular valve repair / replacement Mustard / Senning procedure Fontan-type procedures, Glenn shunts continuous monitoring during early post op period diagnosis and management of early post op complications
- *interventional catheterization*	guidance guidewire and catheter position exclusion of immediate complications assessment of immediate haemodynamic results / changes balloon dilatation of venous pathways device closure of atrial septal defects monitoring of aortic valvuloplasty
Follow-up	Mustard / Senning procedures Fontan-type procedures atrioventricular valve repair / replacement residual or acquired atrial lesions

planes are strictly limited in the evaluation of coarctation of the descending aorta or in the definition of arterial override above a ventricular septal defect. The hemodynamic information that can be derived from transesophageal imaging is limited to, firstly, pulmonary vein flow patterns, secondly, atrioventricular valve flow patterns, and, thirdly, the flow patterns within the branch pulmonary arteries. The orientation of either ventricular outflow tract relative to the esophagus precludes any meaningful evaluation of these structures by transesophageal Doppler techniques. This is particularly true since, to date, only pulsed wave Doppler facilities are provided on pediatric probes.

Future directions: Further improvements in transducer design have to focus on an improved image quality. The development of 48 element 5 MHz transducers, in this respect, was a definitive advancement compared to the first generation 24 element pediatric probe (1). An increase of the transducer frequency to 7.5 MHz would appear to be beneficial for study of small children, although a decrease in ultrasound penetration may be anticipated. One of the major limitations of transesophageal imaging, to date, are the fixed transverse axis scan planes, which result in a much compromised assessment of the ventriculo-arterial junction and the

great arteries. The adjunct of biplane or, ideally, multiplane imaging thus may be expected to be of substantial additional value (24,25). In particular the assessment of the systemic venous return, the right ventricular outflow tract and the descending aorta will then be much improved. In addition, only the advent of additional scan planes will provide a complete evaluation of atrial septal defects.

The value of transesophageal echocardiography as a monitoring technique during interventional cardiac catheterization deserves further studies. It might be expected that the technique will prove to be the monitoring technique of choice during device closure of atrial septal defects, or during balloon dilatation of obstructed venous pathways. Transesophageal pulsed wave Doppler studies open up a new field of hemodynamic investigations in primary or corrected congenital heart disease. New insights may be anticipated for instance in the assessment of diastolic ventricular function or the hemodynamics of surgically created circulations.

Finally, detailed studies are required to determine the value of transesophageal echocardiography in relation to magnetic resonance imaging in the assessment of congenital heart disease in children.

Pediatric transesophageal echocardiography is a new diagnostic modality in pediatric cardiology. It is a safe, albeit semi-invasive technique, that requires general anesthesia or heavy sedation. On the basis of the results of the study protocols described above, transesophageal studies would appear to be indicated in both in the primary diagnosis and in the follow-up of congenital cardiac lesions which involve the atrial chambers or atrioventricular junction of the heart and in those with suspected anomalies of either venous return. In addition, its use as a monitoring technique during either the perioperative period and during interventional cardiac catheterization is highly rewarding.

The technique is of complementary value to precordial ultrasound studies, cardiac catheterization and angiocardiography. It has well defined and important role in modern pediatric cardiological practice which is rapidly increasing.

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Chapter 13

- Summary -

Transesophageal echocardiography, using dedicated pediatric single-plane transesophageal probes, was carried out in a series of children with congenital heart disease, to determine its value, firstly, in the primary diagnosis of congenital heart disease, secondly, to assess its relative value in the monitoring of surgical repair, and, thirdly, to define its contribution in the follow-up of congenital heart disease.

Chapter 1 summarizes the recent development of pediatric transesophageal echocardiography, which evolved only after the technique became an established diagnostic and monitoring technique in the adult patient with acquired cardiac disease. Several limitations of precordial ultrasound techniques are discussed, suggesting that the transesophageal approach may be of substantial additional value in certain diagnostic areas of pediatric cardiology.

Chapter 2 describes the technology of pediatric transesophageal transducers and outlines a standard examination protocol used in the assessment of congenital heart disease. The standard imaging planes are illustrated by anatomic sections of a normal heart obtained at autopsy.

Chapter 3 reports our initial experience with this technique in a group of twenty-five children with congenital heart disease. The results were correlated with the information derived from precordial ultrasound studies and cardiac catheterization. Additional information was obtained in 60 % of the cases. Complications related to such studies were not encountered.

Chapter 4 describes the definition of atrial appendage morphology by transesophageal cross-sectional imaging. The direct visualization of the unique morphologic characteristics of either atrial appendage provides the direct diagnosis of atrial situs in every patient studied. Thus, transesophageal studies would appear to be the ultimate diagnostic test in the determination of atrial situs.

Chapter 5 outlines the improved sensitivity of transesophageal echocardiography in the documentation of juxtaposition of the atrial appendages. The morphologic characteristics of left juxtaposition, which involves an abnormal orientation of the atrial septum, were described in detail.

Chapter 6 reports the additional value of transesophageal studies in the definition of systemic and pulmonary venous connections in a large series of children with congenital heart disease. The technique was found to be largely superior to the precordial ultrasound approach in the detection of anomalous venous connections or in their definitive exclusion. In addition, it provided the assessment of fine morphologic details, not appreciated by angiocardiography.

Chapter 7 describes our initial experience with transesophageal studies in the assessment of complex atrioventricular junction abnormalities. The results indicated that the technique may be expected to be the diagnostic technique of choice in the exclusion of chordal straddling and in the determination of ventricular morphology.

Chapter 8 summarizes the additional value of transesophageal imaging in the assessment

of left ventricular outflow tract obstruction in childhood. The major advantages were found in the definition of multiple insertions of so-called fibromuscular membranes, or in the ultimate exclusion of involvement of the subvalvar mitral valve apparatus.

Chapter 9 compares the intraoperative transesophageal with the epicardial ultrasound approach in the surgery of congenital heart disease. Whereas the information provided by either approach on the atrial chambers or the atrioventricular junction was found to be comparable, it was only the epicardial approach which allowed for the definitive post bypass exclusion of residual ventricular outflow obstruction or residual ventricular shunting. However, transesophageal studies appeared to be the technique of choice in the diagnosis and management of early post operative complications.

Chapter 10 describes the transesophageal assessment of atrial baffle morphology and function after atrial correction procedures for complete transposition. The technique was largely superior to precordial ultrasound studies, and equal to cardiac catheterization in the documentation of venous pathway obstructions. It was superior to angiocardiography in the detection of small or multiple baffle leaks.

Chapter 11 reports the value of transesophageal studies in the follow-up of patients who have undergone a Fontan-type procedure. Hemodynamic lesions on atrial level were more readily identified than by precordial studies, as were right heart thrombi. Transesophageal studies, in contrast to precordial studies, allowed the assessment of the majority of Fontan connections and Glenn shunts. Pulsed wave Doppler studies provided a much improved insight into the hemodynamics after different types of Fontan procedures, and provided clues in the determination of the driving forces of this type of circulation.

Chapter 12 discusses the value of transesophageal echocardiography in children with congenital heart disease. Based on a one-year experience, the major contribution of this new diagnostic modality in pediatric cardiology were described, its limitations were dicussed, as were the anticipated future directions. Finally, guidelines for the indication of such studies in children were outlined.

-Samenvatting-

Transoesofageale echocardiografie werd met behulp van speciale pediatrische oesofagustransducers toegepast bij een groep kinderen met een aangeboren hartafwijking om in de eerste plaats, de betekenis ervan bij de primaire diagnostiek van aangeboren hartafwijkingen vast te stellen en, in de tweede plaats, na te gaan wat de waarde ervan is tijdens en na chirurgische behandeling. In de derde plaats werd onderzocht in hoeverre transoesofageale echocardiografie van belang is bij de follow-up van patiënten die geopereerd zijn voor een aangeboren hartafwijking.

In hoofdstuk 1 wordt een samenvatting gegeven van de recente ontwikkelingen van transoesofageale echocardiografie bij kinderen. Deze ontwikkelingen kwamen pas op gang nadat deze techniek voor diagnostiek en bewaking van volwassenen met verkregen hartafwijkingen zijn waarde had bewezen.

De beperkingen van precordiale ultrageluidstechnieken worden besproken, daarmee vooruitlopend op het feit dat de transoesofageale techniek, hiermee vergeleken, een belangrijke aanwinst betekent voor het diagnostisch areaal binnen de kindercardiologie.

In hoofdstuk 2 worden de technische aspecten van de pediatrische transoesofageale transducers besproken. Tevens wordt het standaard onderzoeksprotocol dat gebruikt wordt bij de diagnostiek van aangeboren hartafwijkingen toegelicht. De standaard echoopnamedoorsneden worden geïllustreerd aan de hand van doorsneden van anatomische preparaten van het normale hart.

In hoofdstuk 3 worden onze eerste ervaringen met transoesofageale echocardiografie bij 25 kinderen met een aangeboren hartafwijking besproken. De resultaten hiervan worden vergeleken met die, welke met behulp van precordiaal ultrageluidsonderzoek en hartcatheterisatie werden verkregen. Hieruit kwam naar voren dat in 60% van de gevallen via de transoesofageale methode additionele informatie werd verkregen. In deze groep deden zich geen complicaties voor.

In hoofdstuk 4 wordt de morfologie van de hartoren met behulp van de transoesofageale methode gedefinieerd, resulterend in een directe diagnose van de atriale situs bij iedere onderzochte patiënt. Hieruit wordt geconcludeerd dat de transoesofageale methode de beste diagnostische techniek is voor het vaststellen van de atriale situs.

In hoofdstuk 5 wordt de grotere sensibiliteit van transoesofageale echocardiografie ten aanzien van de identificatie van juxtapositie van de hartoren beschreven. De karakteristieke morfologie van juxtapositie van links wordt in detail toegelicht.

In hoofdstuk 6 wordt ingegaan op de additionele waarde van transoesofageale studies bij het vastleggen van de systeem- en longveneuze connecties bij een groot aantal kinderen met een aangeboren hartafwijking. De transoesofageale methode bleek superieur te zijn ten opzichte van de precordiale ten aanzien van het aantonen of uitsluiten van abnormale veneuze connecties. Bovendien bleek het mogelijk morfologische details te visualizeren, die niet met behulp van angiocardiografie konden worden aangetoond.

In hoofdstuk 7 worden onze eerste ervaringen met transoesofageale echocardiografie bij het vaststellen van complexe, abnormale atrioventriculaire connecties beschreven. De resultaten geven aan, dat deze techniek waarschijnlijk het beste diagnosticum is bij het uitsluiten van "straddling" van de chordae en bij de determinatie van de morfologie van de ventrikels.

In hoofdstuk 8 komt de waarde van transoesofageale beeldvorming bij het aantonen van obstructies in de uitstroombaan van de linker ventrikel naar voren. De belangrijkste voordelen blijken te liggen in het gedetailleerd vastleggen van multiple inserties van de zogenaamde fibromusculaire membranen, of juist in het uitsluiten van betrokkenheid van het mitralis-klepapparaat bij de membranen.

In hoofdstuk 9 worden de transoesofageale en epicardiale echocardiografie tijdens operatie van aangeboren hartafwijkingen met elkaar vergeleken. Hieruit komt naar voren dat de resultaten van beide technieken ten aanzien van de beeldvorming van de atria en de atrioventriculaire connecties gelijkwaardig zijn, maar dat na cardiopulmonale bypass alleen door middel van de epicardiale techniek definitieve uitsluiting van residuële obstructies in de uitstroombaan van de ventrikels of van residuële intraventriculaire shunting mogelijk is. Daarentegen is het juist de transoesofageale methode die naar voren komt bij de diagnostiek en behandeling van vroeg-post-operatieve complicaties.

In hoofdstuk 10 wordt de transoesofageale evaluatie van de morfologie en functie van de atriale baffle na atriale correctie van transpositie van de grote vaten beschreven. De techniek is duidelijk superieur ten opzichte van de precordiale techniek en gelijkwaardig ten opzichte van hartcatheterisatie bij het aantonen van veneuze obstructies. Voor de detectie van kleine of multiple baffle lekkages is de transoesofageale methode superieur aan angiocardiografie.

In hoofdstuk 11 wordt de waarde van transoesofageale echocardiografie bij de follow-up van patiënten die een Fontan-type operatie hebben ondergaan, aangetoond. Hemodynamische afwijkingen op atriumniveau en thrombi in het rechter hart worden beter geïdentificeerd dan met behulp van precordiale echocardiografie. Visualizatie van de Fontan connecties en Glenn shunts is meestal goed mogelijk met behulp van transoesofageale technieken, in tegenstelling tot precordiale technieken. Met behulp van pulsed wave Doppler onderzoek vanuit de transoesofageale positie wordt meer inzicht verkregen in de hemodynamiek bij verschillende typen Fontan connecties en in de drijvende krachten die dit type circulatie bepalen.

Hoofdstuk 12 omvat de discussie over de waarde van transoesofageale echocardiografie bij kinderen met een aangeboren hartafwijking. De belangrijkste contributie van deze nieuwe techniek aan de diagnostiek van aangeboren hartafwijkingen enerzijds en de beperkingen ervan anderzijds, worden, gebaseerd op de ervaringen van één jaar, besproken, evenals de te verwachten ontwikkelingen voor de toekomst. Tenslotte worden de indicaties voor deze techniek bij kinderen aangegeven.

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