ORIGINAL ARTICLES

Assessment of anomalous systemic and pulmonary venous connections by transoesophageal echocardiography in infants and children

Oliver Stümper, Jesus Vargas-Barron, Maria Rijlaarsdam, Angel Romero, Jos RTC Roelandt, John Hess, George R Sutherland

Abstract

Objective—To assess the value of transoesophageal echocardiography in the preoperative definition of systemic and pulmonary venous connections.

Design—Transoesophageal echocardiographic studies were performed prospectively under general anaesthesia in 76 consecutive unoperated children. Results were compared with those obtained by earlier transthoracic ultrasound studies ($n = 76$), cardiac catheterisation ($n = 62$), and subsequent surgical inspection ($n = 58$).

Setting—Two tertiary referral centres.

Patients—76 unoperated infants and children (age 0.2–14.8 years, mean age 4.1 years) with congenital heart disease.

Main outcome measure—Identification of anomalous systemic and pulmonary venous connections.

Results—Transoesophageal studies showed anomalous venous connections in 14 patients. Two had both anomalous systemic and pulmonary venous connections. Transoesophageal studies showed 12 anomalous systemic venous connections in nine patients. In eight patients these were confirmed at operation or catheterisation: one patient is awaiting operation. Six anomalous systemic venous connections were missed during earlier transthoracic studies. Anomalous pulmonary venous connections (one mixed total, six partial) were shown in seven patients. These were confirmed at operation in six and by cardiac catheterisation in one. Four of these patients were missed during earlier transthoracic ultrasound studies. No patient defined as having normal venous connections by the transoesophageal study was subsequently shown to have anomalous venous connections at operation or angiography.

Conclusions—Transoesophageal echocardiography is a highly sensitive tool for the preoperative definition of systemic and pulmonary venous connections. In this series it was better than transthoracic ultrasound and complemented cardiac catheterisation and angiography.

The range of anomalies of either pulmonary or systemic venous connections to the heart is wide, both in terms of clinical presentation and of underlying morphology.^{1,2} Because of the important implications such anomalies may have for the surgical approach,³ the patterns of the venous connections to the heart have to be defined before surgical correction. Whereas total anomalous pulmonary venous connection in the critically ill neonate is usually readily diagnosed by precordial ultrasound techniques,⁴⁻⁶ other forms of anomalous venous connections may be difficult to diagnose or to rule out.^{7,8} Cardiac catheterisation with appropriate selective angiography is regarded as the best diagnostic technique for the definition of systemic venous connections. However, it can be difficult to define or exclude anomalous pulmonary venous connections.

Transoesophageal echocardiography is a relatively new adjunct to the ultrasound evaluation of children with congenital heart disease.⁹⁻¹¹ Because of the proximity of the transoesophageal ultrasound transducer to the sites of venous return and to the atrial chambers this technique may allow a more accurate evaluation of these cardiac structures than is obtained by the transthoracic ultrasound approach. To determine its value in the preoperative diagnosis or exclusion of anomalous venous connections, transoesophageal studies were performed prospectively and independently in 76 unoperated children. The results were correlated with those obtained by earlier transthoracic ultrasound studies, by cardiac catheterisation and angiography, and by subsequent inspection at operation.

Patients and methods

PATIENTS

Between May 1989 and April 1990 76 consecutive unoperated infants and children with congenital heart disease were prospectively studied by transoesophageal echocardiography. We obtained the approval of the hospital ethical committees of the Academic Hospital, Rotterdam, and the National Institute of Cardiology, Mexico City before the study began. Informed parental consent was obtained for individual transoesophageal

Academic Hospital
Rotterdam—Dijkzigt,
Sophia Children's
Hospital, and
Thoraxcenter,
Rotterdam,
Netherlands, and
National Institute of
Cardiology, Mexico
City, Mexico
O Stümper
J Vargas-Barron
M Rijlaarsdam
A Romero
J R T C Roelandt
J Hess
G R Sutherland

Correspondence to
Dr Oliver Stümper,
Department of Paediatric
Cardiology, Royal Hospital
for Sick Children, Sciennes
Road, Edinburgh EH9 1LF.

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studies. The age at investigation ranged from 2.5 months to 14.8 years (mean of 4.1, median 2.8 years). The body weight ranged from 3.7 kg to 48 kg (mean 13.9 kg, median 11.7 kg). Nineteen patients were less than a year old.

All 76 infants and children were studied under general anaesthesia. This was given in 46 consecutive children for scheduled cardiac catheterisation and in a further 18 consecutive children for subsequent surgical correction. In the remaining 12 selected patients with complex congenital heart disease elective transoesophageal studies were performed under general anaesthesia on a day care basis. Transoesophageal studies were performed and interpreted by an investigator who was unaware of any earlier findings obtained by cardiac catheterisation or angiocardiology, which had been carried out in 15 patients.

SCANNING EQUIPMENT

The first 17 patients were studied with an Aloka 24 element paediatric transoesophageal probe connected to an Aloka SSD 870 ultrasound system. The next 59 patients were studied with

a prototype 48 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 both probes was 30 mm (7 × 8 mm and 5 × 10 mm respectively) and the maximal shaft diameter was 7 mm. Probe tip manipulation was restricted to anterior/posterior angulation only.

TECHNIQUE

After sufficient lubrication 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 for the study. In every patient we completed a standardised scheme of investigation (to include cross sectional imaging and colour flow mapping studies) to assess the atrial arrangement, the atrio-ventricular and ventriculoarterial junctions and the intracardiac morphology¹¹ using the standard transverse axis scan planes.^{12,13} The atrial arrangement was defined by direct visualisation of both atrial appendages, according to the criteria previously described by our group.¹⁴ The morphology of the systemic venous and the pulmonary venous connections was thoroughly examined by the techniques described below.

ASSESSMENT OF SYSTEMIC VENOUS CONNECTIONS

The morphology of the inferior vena cava and the hepatic veins was assessed by transgastric views of the liver. A complete scan of the upper abdomen was obtained by rotating the probe clockwise to determine the relation of the infradiaphragmatic great vessels one to another and to the spine. Then the probe was gradually withdrawn to demonstrate the presence of any venous valve within the atrium, the roof of the right sided atrium, and the connection with the superior vena cava (fig 1A). The superior vena cava could routinely be demonstrated anterior to the right pulmonary artery. A more cranial segment of this vessel was demonstrated by withdrawing the probe further within the oesophagus above the right main bronchus. The innominate vein was searched for anterior to the aortic arch. The coronary sinus was demonstrated when low transoesophageal four chamber views were scanned (fig 1C).

The existence of a left persistent superior vena cava was documented by using high left atrial views (fig 1A) and views of the left atrial views (fig 1A) and views of the left pulmonary artery and the left sided mediastinum. An azygos vein was searched for by combined cross sectional imaging and colour flow mapping 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.

Figure 1 Diagram showing the standard transoesophageal sections used in the definition of both systemic and pulmonary venous connections to the heart. (A) High left atrial view demonstrating both atrial appendages, both upper pulmonary veins, and the junction of the superior vena cava with the right atrial chamber (asterisk). (B) Standard transoesophageal four chamber view that allows the demonstration of the connection of the left lower pulmonary vein by counter clockwise rotation of the probe. (C) Low transoesophageal view of the left atrium showing the connection of the right lower pulmonary vein with the left atrial chamber, the coronary sinus at the floor of the left atrium, and the junction of the inferior vena cava with the right atrial chamber. At this level the eustachian valve (asterisk) is also routinely demonstrated. 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; RV, right ventricle; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; SVC, superior vena cava.

ASSESSMENT OF PULMONARY VENOUS CONNECTIONS

The right upper pulmonary vein was visualised immediately posterior to the junction of the superior vena cava with the right atrium (fig 1A). 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 because this vessel normally drains into the left atrium in a more posterior-anterior direction (Fig 1C). The left upper pulmonary vein was readily demonstrated posterior and to the left of the left sided atrial appendage, being separated from it by only a crest of atrial tissue (fig 1A). Slight further advancement and anti-clockwise rotation of the probe allowed the visualisation of the left lower pulmonary vein (fig 1B). After the detection of the individual pulmonary veins the scan position was varied to demonstrate the exact course and the site of connection of these vessels to the atrial chambers relative to the atrial septum. Colour flow mapping was used for the rapid identification of venous connections to the atria and pulsed wave Doppler interrogation was carried out to confirm venous flow patterns and to exclude obstruction to venous return.

TRANSTHORACIC ULTRASOUND STUDIES

All 76 infants and children had prior standardised transthoracic ultrasound studies with the full range of parasternal, subcostal, and suprasternal views in an attempt to demonstrate fully the patterns of systemic and pulmonary venous connections. Studies were performed by independent observers using standard 3.75, 5, or 7.5 MHz transducers connected to either a Vingmed CFM 700 or 750 or a Hewlett Packard Sonos 500 ultrasound system. Seventeen children required sedation. Cross sectional imaging was complemented by subsequent colour flow mapping and pulsed wave Doppler interrogation in all patients to define the sites of venous connections. Contrast echocardiographic studies with peripheral

venous injections of hand agitated saline were used in two patients to confirm a persistent left superior vena cava.

CARDIAC CATHETERISATION

Sixty two children had correlative cardiac catheterisation and angiocardiology. Fifteen had had previous catheterisation, 46 patients had concomitant investigations, and one patient was investigated two weeks after the transoesophageal study. Blood samples for oximetry were taken at multiple sites including the high superior vena cava, the inferior vena cava, the right atrium, and the right ventricle. Appropriate selective angiocardiology was performed with biplane equipment and included pulmonary artery injection(s) in all children. Selective contrast injections into the innominate vein were not performed routinely.

EVALUATION

All transoesophageal studies were continuously recorded onto video tape and were analysed both on-line and later off-line at reduced speed by two independent observers who were blinded to the angiocardiology or surgical findings. Thereafter the results of the transoesophageal studies were correlated in all patients with the findings obtained at review of the earlier transthoracic ultrasound studies (n = 76), with the results obtained by either simultaneous or recent cardiac catheterisation (n = 62), and with the findings during subsequent surgical inspection (n = 58). Surgical inspection was used as the reference standard for the definition of venous connections in all patients in whom this information was available. In those 18 patients who did not undergo intracardiac correction, cardiac catheterisation with selective angiocardiology was used as the reference standard.

Results

SYSTEMIC VENOUS CONNECTIONS

The individual patterns of systemic venous

Table 1 Data on 14 patients with anomalous venous connections

Case	Age (yr)	Sex	Weight (kg)	Diagnosis			Systemic venous connections	Pulmonary venous connections
				Situs	AV junction	VA junction		
1	13.7	M	40.2	Solitus	Absent right	Concordant	Normal	RUPV to RSVC
2	14.8	F	42.5	Solitus	Concordant	Concordant	LSVC to CS	RUPV to RSVC
3	7.8	F	20.0	RAI	Ambiguous	DORV	LSVC to left sided atrium multiple HV drainage	All four PVs to left sided atrium
4	2.0	F	9.8	Solitus	Concordant	Concordant	Interrupted IVC azygos continuation	Normal
5	0.7	M	4.7	Solitus	Concordant	Concordant	LSVC 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, multiple HV drainage	All four PVs 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 PVs 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 RSVC, LSVC to CS	Three PVs to left sided atrium LUPV to LSVC
13	5.8	M	15.8	RAI	Absent right	Discordant	RSVC to left sided atrium, LIVC, multiple HV drainage	Three PVs to left sided atrium
14	1.6	M	8.1	Solitus	Absent right	DORV	LSVC to CS	RLPV not identified Normal

CS, coronary sinus; HV, hepatic veins; IVC, inferior vena cava; 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.

Figure 2 Transoesophageal echocardiogram showing a left persistent superior vena cava draining into the left sided atrium in a patient with left atrial isomerism (case 11). On this cross sectional image at the level of the pulmonary arteries the vessel (asterisk) is shown to the left of the (dilated) pulmonary trunk near its bifurcation. Slight advancement of the probe within the oesophagus identified the connection to the left sided atrium. Ao, ascending aorta; PT, pulmonary trunk.

Figure 3 Persistent left superior vena cava draining to the coronary sinus in a patient with dextrocardia, atrial situs solitus, and absent right atrioventricular connection (case 14). On this transoesophageal "four chamber" view the connection with the dilated and abnormally positioned coronary sinus (arrow) is visualised. See legend to figure 1 for abbreviations.

connections were readily documented by transoesophageal studies in all 76 patients studied from the views described above. Systemic venous return was defined as normal in 67 children.

Transoesophageal studies showed a left persistent superior vena cava (table 1) in seven patients. The course of the vessel, running anterior to the left pulmonary artery (fig 2) and interposed between the left sided pulmonary veins and the left sided atrial appendage, was demonstrated in all seven patients by combined cross sectional imaging and colour flow mapping. The left persistent superior vena cava was demonstrated draining into the roof of the left sided atrium in three patients (cases 3, 8 and 11). In none of these patients had this anomaly been identified during earlier transthoracic ultrasound studies (table 2). The anomaly was confirmed at operation in two patients and by selective angiocardiology in the remaining patient. In four patients (cases 2, 5, 12 and 14) the left persistent superior vena cava was shown to be connected to the coronary sinus (fig 3). This had been identified during earlier transthoracic studies in three patients, two of whom had additional confirmatory contrast echocardiographic studies. An unroofed

coronary sinus in patient 5 (table 1) was not recognised during either the transoesophageal or the transthoracic ultrasound study; the lesion was only identified at surgical correction. Connection of a left persistent superior vena cava to the coronary sinus was subsequently confirmed by surgical inspection in both patients who underwent correction and by cardiac catheterisation in all four.

Anomalies of the right superior vena cava were shown by transoesophageal studies in two patients. In one child with atrial situs solitus (case 12) an interrupted right superior vena cava was detected. The persistent left superior vena cava in this child was shown to be connected to the coronary sinus. The earlier transthoracic study was ambiguous about the presence of a right superior vena cava, which was definitively excluded at subsequent cardiac catheterisation. In the second patient, who had right atrial isomerism (case 13), the right sided superior vena cava was shown to be connected to the left side of the remnant of the atrial septum (fig 4). Neither transthoracic ultrasound studies nor angiocardiology defined the site of drainage of the superior vena cava in relation to the largely deficient septum. This finding awaits surgical confirmation.

The morphology and connection of the inferior vena cava (interruption in three patients) were equally well documented by subcostal and transoesophageal studies. An azygos vein was identified by both techniques in all three patients with an interrupted inferior vena cava. Two of these patients were shown by direct transoesophageal visualisation of atrial appendage morphology to have left atrial isomerism (cases 8 and 11). The third patient had atrial situs solitus (case 4). The pattern of hepatic venous drainage in four patients with atrial isomerism (three at multiple sites, one with common drainage) was better documented by the transoesophageal technique in one patient with multiple atrial drainage sites,

Table 2 Assessment of anomalous venous connections by transthoracic and transoesophageal echocardiography

Anomalous venous connection	Transthoracic	Transoesophageal
Systemic venous:		
LSVC to LA	0/3	3/3
LSVC to CS	3/4	4/4
Absent RSVC	0/1	1/1
RSVC to LA	0/1	1/1
Interrupted IVC	3/3	3/3
Pulmonary venous:		
PAPVC	2/6	6/6
Mixed TAPVC	0/1	1/1

LA, left atrium; PAPVC, partial anomalous pulmonary venous connection. See footnote to table 2 for other abbreviations.

Figure 4 Transoesophageal echocardiogram showing a right superior vena cava connected to the left sided atrium in a patient with right atrial isomerism (case 13). The crest of atrial tissue of the cavoatrial junction was continuous with the remnant of the atrial septum (arrow). In addition the right upper pulmonary vein (asterisk) connected with the left sided, morphologically right atrium (RA). Trabeculations of the right atrial appendage caused the echoes seen at its base. See legends to figures 1 and 2 for abbreviations.

in whom more sites were identified from within the oesophagus.

PULMONARY VENOUS CONNECTIONS

The site of drainage of all four pulmonary veins was identified by cross sectional transoesophageal imaging or colour flow mapping in 69 (91%) of the 76 patients studied. Both the right upper and left upper pulmonary veins were identified in all patients: the left lower pulmonary vein in 74 patients (97%) and the right lower in 70 patients (92%).

The transoesophageal study showed partial anomalous pulmonary venous drainage in six children. Only two of these had been identified by earlier transthoracic ultrasound studies (table 2). Partial anomalous connection of the right upper pulmonary vein to the superior vena cava was shown in two (cases 1 and 2) by the transoesophageal study and was suggested in only one (case 2) by the earlier transthoracic study. The site of drainage was correctly identified in one of these from within the oesophagus (fig 5) but in the second patient it was not because of interposition of the right bronchus. The site of drainage could not be documented by the transthoracic study. In two patients (cases 6 and 9) the right upper pulmonary vein and in one patient (case 7) the right lower pulmonary vein were shown by the transoesophageal study to be connected to the

right sided atrium (fig 6). Transthoracic studies had shown this abnormality in only one patient (case 6). All three patients had a sinus venosus atrial septal defect. Additional defects in the secundum septum were present in two of these (cases 7 and 9), in whom earlier transthoracic studies had not defined partial anomalous pulmonary venous drainage. Anomalous connection of the left upper pulmonary vein to a left persistent superior vena cava was documented by the transoesophageal study in one patient (case 12). The earlier transthoracic study had suggested normal venous connections. In five of these six children in whom transoesophageal ultrasound defined a partial anomalous pulmonary venous connection the findings were later confirmed by subsequent surgical inspection. In the sixth patient (case 12) it was confirmed by cardiac catheterisation.

One child with total anomalous pulmonary venous connection (case 10), as defined by transthoracic ultrasound scans, was shown to have a mixed type of connection during the transoesophageal study. 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 earlier suprasternal study led to this finding being misinterpreted as total anomalous supracardiac pulmonary venous connection. On a subsequent review of the transthoracic study mixed total anomalous drainage was suggested; however, the individual site of drainage of all four pulmonary veins was not demonstrated. The transoesophageal finding was confirmed by subsequent cardiac catheterisation and later at surgical inspection.

NORMAL VENOUS CONNECTIONS

In fourteen of the 76 children studied anomalous venous connection was identified by transoesophageal studies. In the remaining 62 children the transoesophageal ultrasound studies showed normal systemic and pulmonary venous connections. In all of these patients the findings were subsequently confirmed by surgical inspection or cardiac catheterisation.

Complications related to the transoesophageal studies were encountered only one child (4.7 kg) with severe pulmonary hypertension, who had a hypertensive crisis after the introduction of the probe. With appropriate anaesthesia the study was successfully completed. None of the patient's showed signs of oesophageal bleeding or trauma.

Discussion

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

TRANSTHORACIC ULTRASOUND

Transthoracic ultrasound is a sensitive method of detecting anomalous venous connections in small children, who have good transthoracic

Figure 5 Transoesophageal echocardiogram showing partial anomalous pulmonary venous connection of the right upper pulmonary vein to the high right atrium. (arrow) RPA, right pulmonary artery. See legends to figures 1 and 2 for abbreviations.

Figure 6 Transoesophageal colour flow mapping showing an anomalous connection of the right upper pulmonary vein (arrow) to the right atrium. RA, right atrium; RPA, right pulmonary artery.

ultrasound windows.^{4,6} However, in older children or in those with complex cardiac malformations the transthoracic approach is frequently limited in clinical practice. In this series of patients the relatively poor results in the transthoracic identification of anomalous systemic venous connections were presumably in part related to the older age of these patients (mean age 9.9 years).

The definition of the site of connection of all four pulmonary veins by transthoracic ultrasound studies is difficult in children beyond infancy. Thus the correct identification 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 the venous connections is often impossible. In this series both the nature of the lesions encountered together with the age of the patients (mean age 6.7 years) may explain the low incidence with which partial anomalous pulmonary venous drainage was identified by the transthoracic approach. Finally, transthoracic pulsed wave Doppler interrogation of individual pulmonary vein flow patterns, which can detect pulmonary vein obstruction^{15,16} is not always possible in the older child.

Figure 7 (A) Cross sectional transoesophageal image showing the left superior vena cava anterior to the dilated left pulmonary artery. (B) Subsequent colour flow mapping and pulsed wave Doppler studies identified the connection between the left upper pulmonary vein (arrow) and the left persistent superior vena cava. LPA, left pulmonary artery; LSVC, left superior vena cava.

CARDIAC CATHETERISATION AND ANGIOCARDIOGRAPHY

Cardiac catheterisation with angiocardio-graphy is a highly sensitive technique in the preoperative definition of the systemic venous connections to the heart. However, the assessment of the pulmonary venous connections by selective pulmonary artery contrast injections may not provide diagnostic information. This is particularly true of patients in whom there is slight obstruction of the abnormal connection. Subtle morphological details such as the relation of the site of the venous connections to the interatrial septum in patients with sinus venosus atrial septal defects or in those with complex congenital heart disease with a largely deficient interatrial septum are difficult to assess by angiocardio-graphy.

TRANSOESOPHAGEAL ECHOCARDIOGRAPHY

Transoesophageal echocardiography is a newer method of assessing congenital heart disease. With the availability of dedicated paediatric transoesophageal transducers it has now become a feasible and safe technique even in small children.^{10,11} The proximity of the transducer to the atrial chambers and to the sites of venous return potentially allows for an improved evaluation of these structures. During a prospective study protocol the value of this technique in the definition of both systemic and pulmonary venous connections was assessed. All transoesophageal studies were performed and interpreted by investigators who were blinded to the results of cardiac catheterisation. The study population consisted of two groups. The larger group consisted of consecutive patients being studied while undergoing cardiac catheterisation or total surgical correction and a smaller group consisted of those with complex congenital heart disease who were selected for elective transoesophageal studies carried out on a day care basis.

SYSTEMIC VENOUS CONNECTIONS

A left persistent superior vena cava draining to the coronary sinus was readily identified on both cross sectional imaging and colour flow mapping in all patients. Such vessels were found to be interposed between the left sided

atrial appendage and the left sided pulmonary veins. Their course, cranially and in front of the left pulmonary artery could then be followed by slight changes in the level of probe insertion. Drainage of a left superior vena cava to the roof of the left sided atrium was reliably established. Transoesophageal imaging allowed the identification of very subtle morphological details of the relation of the systemic venous connections to the interatrial septum, which could not be delineated by either transthoracic ultrasound or angiocardigraphic studies. The limitations of single plane transoesophageal imaging included difficulties in the documentation of the presence or absence of an innominate vein. The probe had to be positioned high within the oesophagus and a careful search had to be undertaken to visualise this vessel just anterior to the aortic arch. Interposition of the bronchial tree may preclude adequate visualisation of this area. Although we were able to image the vessel appropriately in 18 of 23 patients later in this series, we thought that this could be performed more readily from the suprasternal transthoracic scan positions. In this series azygos and hemiazygos veins could be demonstrated only in cases where these vessels were enlarged. In only one patient was the definition of inferior vena cava or hepatic venous connections enhanced by the transoesophageal study.

Transoesophageal studies defined anomalous systemic venous connections in nine patients and excluded these in the remaining patients. Eight of these nine children had subsequent surgical or angiocardigraphic confirmation of the findings. One patient awaits surgical confirmation. None of the remaining 67 patients, who were defined having normal systemic venous connections by the transoesophageal study, was shown by either surgical inspection or cardiac catheterisation and angiocardigraphy to have anomalous systemic venous connections. Thus in this series the sensitivity of transoesophageal echocardiography in the detection of anomalous systemic venous connections was 100%.

PULMONARY VENOUS CONNECTIONS

The site of connection of all four pulmonary veins could be demonstrated in most cases (91%). Only the connection of the right lower pulmonary vein, because of its rather posterior/anterior course on the right lateral aspect of the heart, was difficult to demonstrate. However, with increasing experience during the study the detection of this vessel became less difficult. Thus anomalous venous connections should be assumed in every patient in whom all four pulmonary veins cannot be demonstrated at their normal position. In these patients subsequent detailed imaging and colour flow mapping of the adjacent structures showed the site of drainage in most patients. Problems were only encountered in this series in those patients in whom the site of connection was at the level of the bronchial tree and thus inaccessible for transoesophageal ultrasound.

Seven of the 76 children studied were shown to have anomalous pulmonary venous connec-

tions. The findings were confirmed by surgical inspection or cardiac catheterisation in all of these. Such connections were excluded in the remaining 69 patients, in whom the transoesophageal study showed normal pulmonary venous connections. Thus in this series the sensitivity of transoesophageal ultrasound in detecting anomalous pulmonary venous connections was 100%.

LIMITATIONS

Interposition of the trachea and the main bronchi precluded the visualisation of a distal segment of the superior vena cava and limited the definition of the site of anomalous pulmonary venous connections at these levels. The near-field artefact present in the first generation of paediatric transoesophageal probes has now been much reduced allowing the reliable identification by transoesophageal colour flow mapping studies of all four pulmonary veins even in small children. Single plane transverse axis transoesophageal imaging of the venae cavae produces short axis cuts. So to display fully the course and 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 a substantial additional value in the evaluation of systemic venous connections. However, it will contribute only little in the assessment of the pulmonary venous connections.¹⁷ The relative values of transoesophageal ultrasound studies and magnetic resonance imaging^{8,18} in the definition of anomalous venous connections needs to be assessed. So far our experience with magnetic resonance imaging is limited.

POTENTIAL INDICATIONS

Because transoesophageal echocardiographic studies can only be performed in children under general anaesthesia or heavy sedation, the indications for elective studies should be strict. However, these examinations can be performed safely during simultaneous cardiac catheterisation or immediately before surgical correction without the requirement of additional procedure or anaesthesia time. The results of this study protocol suggest that transoesophageal studies may be indicated in children with complex congenital heart disease, those with abnormal atrial arrangement, or in older children with poor transthoracic ultrasound windows in whom anomalous venous connections to the heart are either suspected or in whom their existence would alter the surgical approach.

Transoesophageal echocardiographic studies seem to be a most valuable adjunct in the preoperative definition or exclusion of anomalous venous connections to the heart. In this series the technique was better than the transthoracic ultrasound approach. It equalled cardiac catheterisation and angiocardigraphy in the definition of systemic venous connections and was of additional value in the definition of pulmonary venous connections.

- 1 Lucas RV, Schmidt RE. Anomalous venous connections, pulmonary and systemic. In: Moss AJ, Adams FH, Emmanouilides GC, eds. *Heart disease in infants, children and adolescents*. 2nd ed. Baltimore: Williams and Wilkins, 1973;437-70.
- 2 Bharati S, Lev M. Congenital anomalies of the pulmonary veins. *Cardiol Clin* 1973;5:23-41.
- 3 deLeval MR, Ritter DG, McGoon DC, Danielson GK. Anomalous systemic venous connection. Surgical considerations. *Mayo Clin Proc* 1975;50:599-610.
- 4 Sahn DJ, Allen HD, Lange LW, Goldberg SJ. Cross-sectional echocardiographic diagnosis of the sites of total anomalous pulmonary venous drainage. *Circulation* 1979;60:1317-25.
- 5 Smallhorn JF, Sutherland GR, Tommasini G, Hunter S, Anderson RH, Macartney FJ. Assessment of total anomalous pulmonary venous connection by two-dimensional echocardiography. *Br Heart J* 1981;46:613-23.
- 6 Huhta JC, Gutgesell HP, Nihill MR. Cross sectional echocardiographic diagnosis of total anomalous pulmonary venous connection. *Br Heart J* 1985;53:525-34.
- 7 Huhta JC, Smallhorn JF, Macartney FJ, Anderson RH, de Leval M. Cross-sectional echocardiographic diagnosis of systemic venous return. *Br Heart J* 1982;48:388-403.
- 8 Julsrud PR, Ehman RL. The "broken ring" sign in magnetic resonance imaging of partial anomalous pulmonary venous connection to the superior vena cava. *Mayo Clin Proc* 1985;60:874-9.
- 9 Cyran SE, Kimball TR, Meyer RA, et al. Efficacy of intraoperative transoesophageal echocardiography in children with congenital heart disease. *Am J Cardiol* 1989;63:594-8.
- 10 Kyo S, Koike K, Takanawa E, et al. Impact of transoesophageal Doppler echocardiography on paediatric cardiac surgery. *Int J Card Imaging* 1989;4:41-2.
- 11 Stümper O, Elzenga NJ, Hess J, Sutherland GR. Transoesophageal echocardiography in children with congenital heart disease—an initial experience. *J Am Coll Cardiol* 1990;16:433-41.
- 12 Schlüter M, Hinrichs A, Thier W, et al. Transoesophageal two-dimensional echocardiography: comparison of ultrasonic and anatomic sections. *Am J Cardiol* 1984;53:1173-8.
- 13 Seward JB, Khandheria BK, Oh JK, et al. Transoesophageal echocardiography: technique, anatomic correlations, implementation, and clinical applications. *Mayo Clin Proc* 1988;63:649-68.
- 14 Stümper O, Sreeram N, Elzenga NJ, Sutherland GR. The diagnosis of atrial situs by transoesophageal echocardiography. *J Am Coll Cardiol* 1990;16:442-6.
- 15 Smallhorn JF, Pauperio H, Benson L, Freedom RM, Rowe RD. Pulsed wave Doppler assessment of pulmonary vein obstruction. *Am Heart J* 1985;110:483-6.
- 16 Smallhorn JF, Freedom RM. Pulsed Doppler echocardiography in the preoperative evaluation of total anomalous pulmonary venous connection. *J Am Coll Cardiol* 1986;8:1413-20.
- 17 Stümper O, Fraser AG, Ho SY, et al. Transoesophageal echocardiography in the longitudinal axis: correlation between anatomy and images and its clinical implications. *Br Heart J* 1990;64:282-8.
- 18 Didier D, Higgins CB, Fischer MR, Osaki L, Silverman NH, Cheitlin MD. Congenital heart disease: gated MR imaging in 72 patients. *Radiology* 1986;158:227-35.