Changing role of non-invasive investigation in the preoperative assessment of congenital heart disease: a nine year experience

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Abstract

The total surgical experience of a supraregional paediatric cardiology unit over a nine year period (January 1980 to December 1988) was reviewed to assess the effect of the introduction of the full range of ultrasound techniques. A total of 1517 patients underwent cardiac surgery (955 cardiopulmonary bypass, 562 closed procedures). Of these, 485 patients (32%) did not undergo cardiac catheterisation before operation: 217 bypass (23% of all procedures under cardiopulmonary bypass) and 268 closed procedures (48%). The overall ratio of catheterisations to operations for patients undergoing palliative or corrective surgery fell from 0.97 (1980) to 0.38 (1988). The patients were classified as (a) neonates (0-28 days), (b) infants (one to 12 months), and (c) children (one to 14 years). The main impact of non-invasive surgical referral was in neonates (total catheter:operation ratio 0.38; neonates 0.2 for 1988). The surgical population was further divided according to the principal echocardiographic technique available: (a) 1980-4 cross sectional imaging; (b) 1985-6; imaging plus spectral Doppler ultrasound; (c) 1987-8; imaging plus spectral Doppler ultrasound and colour flow mapping. A fall in the catheter:operation ratio for all age groups was most pronounced in the last four years. This reflects increased familiarity and surgical confidence with non-invasive diagnostic assessment. The introduction of each new echocardiographic technique was associated with a significant fall in the total catheter:operation ratio compared with the preceding period. Six incorrect ultrasound diagnoses were made during the entire period; one of these patients died in the early postoperative period.

The integration of Doppler ultrasound with cross sectional imaging has made non-invasive assessment an increasingly practical alternative to preoperative cardiac catheterisation.

Cross sectional echocardiography has come to be accepted as an excellent non-invasive technique for morphological evaluation of congenital heart defects.^{1,2} In young patients it

provides detailed information about intracardiac structure, and with a sequential approach the precise connections, relations, and morphology can be evaluated in detail.³ With the introduction of spectral Doppler and colour flow mapping it has become possible to derive haemodynamic information even in complex cardiac malformations.⁴⁻⁶ The combined use of these techniques should provide much of the information previously only available from cardiac catheterisation and angiography.

Previous studies of non-invasive preoperative assessment in congenital heart disease have been restricted to small or selected groups of patients. There has not been a study of a total paediatric cardiology practice.⁷⁻⁹ The Wessex region is one of the nine designated supraregional centres in England and Wales for neonatal and infant cardiac surgery. It serves a population of approximately 3.2 million people, and has a static annual number of livebirths of approximately 33 000. The population has remained virtually constant with little migration of the patient population out of or into the Wessex region. Virtually all infants or children with suspected congenital heart disease are referred to the regional centre. Some cases are referred from outside the region. A full range of paediatric surgical services except transplantation is available in the unit. Accurate statistics have been recorded for the Wessex region since 1980.

Cross sectional imaging has been available in the unit from 1980. Spectral Doppler scanning was introduced in 1984 and colour flow mapping in 1987. With each new introduction there has been a significant change in the unit's clinical practice, with increasing surgical referral on the basis of non-invasive investigations alone. We retrospectively analysed the impact of these changes on surgical referral patterns in paediatric heart disease.

Patients and methods

From January 1980 to December 1988, 1517 operations were performed on patients with various congenital heart defects. Initial assessment in all patients included clinical examination, relevant biochemical tests, chest x ray, electrocardiography, and echocardiography. Cross sectional echocardiography was carried out with ATL Mark III, Ultramark 4 (7.5 MHz, 5 MHz short and medium focus, or 3.5 MHz probes), or with Toshiba SSH

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Graph showing the ratios of diagnostic cardiac catheterisations to operations for congenital heart disease from January 1980 to December 1988. The ratios are shown for the total surgical population and for the age group subsets (see text).

60A or SSH 65A ultrasound systems (5 mHz, 3.7 mHz, or 2.5 mHz probes). Since 1984, duplex scanning with the Toshiba system and non-imaging continuous wave Doppler with the Vingmed SD 100 system were also used in evaluation. Colour flow mapping has been routinely used since 1987.

Surgical procedures in this period were divided into cardiopulmonary bypass and closed (non-bypass) procedures. The records of patients requiring diagnostic preoperative cardiac catheterisation were reviewed, and a ratio of diagnostic catheter procedures to operations was taken to represent patients requiring preoperative invasive assessment (figure). The patients were divided into three age groups: neonates (0–28 days); infants (one to 12 months), and children (one to 14 years). The cut off age of 14 years is the upper age limit for admission to the paediatric unit of the Wessex Cardiothoracic Centre. To define the influence of the echocardiography the patient population was further divided according to the principal non-invasive diagnostic technique(s) available: (a) 1980–4, cross sectional imaging alone; (b) 1985–6, cross sectional imaging plus pulsed and continuous wave Doppler; (c) 1987–8, cross sectional imaging with Doppler colour flow mapping. Although cross sectional echocardiography was available in 1980, the period 1980–81 was taken to represent a “learning period” with this technique because most patients also underwent diagnostic cardiac catheterisation during these two years.

Diagnostic errors (incomplete or incorrect diagnoses) in patients referred for surgery on ultrasound findings alone were identified. We attempted to define the influence of non-

Table 2 Diagnoses in 189 neonates referred for operation without catheterisation

Diagnosis	No
DA	70
Coarctation/interrupted arch	47
Complex cyanotic CHD/shunt	29
Pulmonary stenosis	11
Truncus arteriosus	9
Pulmonary artery banding	5
Ventricular septal defect	5
Aortic stenosis	5
Transposition/anatomic repair	3
TAPVD	2
Atrioventricular septal defect	1
Miscellaneous	2

CHD, congenital heart disease; DA, ductus arteriosus; TAPVD, total anomalous pulmonary venous drainage.

invasive and invasive assessment on the surgical outcome. We compared the proportion of patients undergoing diagnostic cardiac catheterisation in each period with that in the previous period using a χ^2 fourfold table with Yates's continuity correction.

Results

TOTAL POPULATION

As the routine use of cross sectional echocardiography increased there was a highly significant fall in the overall ratio of diagnostic catheterisations to operations (1980/81 v 1983/84; $p < 0.001$). The subsequent introduction of pulsed and continuous wave Doppler and colour flow mapping also produced significant decreases in the ratio ($p < 0.001$ and $p < 0.025$ respectively) compared with the catheter:operation ratio for the preceding period (figure and table 1).

NEONATES

Of 284 operations performed on neonates over nine years (91 bypass, 193 closed), diagnostic cardiac catheterisation was required in 93 (33%) (51 bypass, 42 closed). Interventional catheterisation procedures for conditions that required later surgical correction (for example balloon septostomy for complete transposition) were classified as diagnostic cardiac catheterisation. The number of diagnostic catheterisation procedures continued to fall as confidence grew in the diagnosis of structural malformations by cross sectional echocardiography (catheter:operation ratio for 1980/81 v 1983/84, $p < 0.001$). The addition of pulsed and continuous wave Doppler ultrasound to the routine preoperative assessment produced a further fall in preoperative diagnostic catheterisation

Table 1 Ratio of cardiac catheterisations to operations in all age groups (1980–88)

Year	Op total	Cath total	c:op	Cardiopulmonary bypass			Closed operations		
				Op	Cath	c:op	Op	Cath	c:op
1980	115	111	0.97	68	68	1.0	47	43	0.91
1981	131	129	0.98	82	82	1.0	49	47	0.96
1982	108	101	0.94	63	63	1.0	45	38	0.84
1983	166	141	0.9	108	106	0.98	58	35	0.6
1984	180	134	0.74	109	100	0.92	71	34	0.48
1985	167	107	0.64	98	79	0.81	69	28	0.41
1986	222	126	0.57	137	101	0.74	85	25	0.29
1987	233	108	0.46	157	74	0.47	76	34	0.45
1988	195	75	0.38	133	65	0.49	62	10	0.16
Total	1517	1032		955	738		562	294	

Cath, diagnostic cardiac catheterisations; Op, operations; c:op, ratio of diagnostic catheterisations to operations.

Table 3 Diagnoses in 119 infants referred for operation without catheterisation

Diagnosis	No
DA	33
Ventricular septal defect	16
Coarctation	14
Complex cyanotic CHD/shunt	13
Atrioventricular septal defect	9
Secundum atrial septal defect	8
Aortic stenosis	5
TAPVD	5
Pulmonary stenosis	3
Truncus arteriosus	3
Transposition/anatomical repair	2
Tetralogy of Fallot	1
Aortopulmonary window	1
Miscellaneous	6 (vascular ring, arteriovenous fistula etc)

See footnote to table 2 for abbreviations.

(catheter:operation ratio for 1983/84 *v* 1985/86, $p < 0.001$). The trend towards reduced catheterisation was especially pronounced in neonates requiring closed cardiac surgery. In 1987/88, 16/20 neonates undergoing shunt procedures, 15/15 patients with coarctation, and 16/16 neonates requiring duct ligation did not have preoperative catheterisation. This trend has been followed to a lesser degree for procedures requiring cardiopulmonary bypass. Table 2 categorises the individual diagnoses in those who had an operation without prior catheterisation.

INFANTS

Of 509 operations carried out in patients aged 1–12 months (328 bypass, 181 closed), 391 patients (274 bypass, 117 closed) required diagnostic cardiac catheterisation (total catheter:operation ratio 0.77) (figure). The ratio of diagnostic catheterisations to operation fell significantly with the introduction of each new non-invasive diagnostic technique (1980/81 *v* 1983/84, $p < 0.005$; 1983/84 *v* 1985/86, $p < 0.005$; 1985/86 *v* 1987/88, $p < 0.05$). Separate analysis of the bypass and closed groups showed that the effect of cross sectional imaging alone was not statistically significant in either group. The introduction of spectral Doppler and colour flow mapping techniques, however, was associated with a significant decrease in preoperative catheterisation in infants undergoing surgical repair with cardiopulmonary bypass (1983/84 *v* 1985/86, $p < 0.025$; 1985/86 *v* 1987/88, $p < 0.005$). Table 3 lists the individual diagnoses for all infants undergoing surgery without prior catheterisation.

CHILDREN

Of 724 operations performed on children aged one to 14 years (536 bypass, 188 closed), diagnostic catheterisation was required in 548 patients (catheter:operation ratio 0.68) (figure). The catheter:operation ratio was similar for both bypass (0.77) and closed (0.72) procedures. Although the catheter:operation ratio decreased with the introduction of cross sectional imaging, the fall was not statistically significant for bypass or closed groups. The introduction of spectral Doppler and colour flow mapping, however, was associated with a significant fall in the catheter:operation ratio

Table 4 Diagnoses in 177 children referred for operation without catheterisation

Diagnosis	No
Secundum atrial septal defect	72
DA	37
Subaortic stenosis	11
Aortic stenosis/valvotomy	8
Ventricular septal defect	7
Coarctation	7
Tetralogy of Fallot	6
Atrioventricular septal defect	5
Aortic/pulmonary valve replacement	5
Aortopulmonary shunts	3
Pulmonary stenosis/valvotomy	3
PAPVD/TAPVD	2
Cor triatriatum	2
Miscellaneous	9

PAPVD, partial anomalous pulmonary venous drainage. See footnote to table 2 for other abbreviations.

for the whole group compared with the preceding period ($p < 0.001$). Table 4 lists the individual diagnoses for children referred to surgery without catheterisation.

DIAGNOSTIC ERRORS WITH ECHOCARDIOGRAPHY

There were six important incorrect ultrasound diagnoses in the nine years (0.01% of the number investigated non-invasively). One patient had multiple levels of coarctation. In a neonate with situs ambiguus, univentricular atrioventricular connection, and coarctation, the laterality of the aortic arch was misdiagnosed. In a patient with pulmonary atresia, ventricular septal defect, and bilateral arterial ducts, the absence of central pulmonary arteries was missed. In a fourth case, coarctation of the aorta was incorrectly diagnosed in a patient with a thrombosed descending aorta. In the fifth patient, despite apparent clinical and colour flow mapping evidence of a secundum atrial septal defect, no interatrial shunt was found at operation. Finally, in one patient with transposition of the great arteries and a ventricular septal defect, additional muscular ventricular septal defects were missed both by echocardiography and at the initial preoperative angiographic review. These were discovered on review of the angiogram in the immediate postoperative period. This patient underwent a Mustard procedure and had closure of a ventricular septal defect but died soon after operation.¹⁰ Surgical mortality could not be directly attributed to errors in preoperative diagnosis in the remainder.

OPERATIVE MORTALITY

Operative or early mortality was defined as death occurring during the operation, within 30 days of operation, or during the period of hospital stay if this was longer than 30 post-operative days. During 1987 and 1988, when the surgical referral based on non-invasive diagnosis was at its highest, there were 11 (4%) early deaths among 245 patients referred for operation without cardiac catheterisation: neonates 7/69, infants 4/51, children 0/125. These figures compare favourably with the mortality figures for the same two year period for patients who underwent preoperative catheterisation: neonates 4/23, infants 6/65, and children 2/96.

The higher mortality for the neonates was

not related to the primary diagnostic method (ultrasound or cardiac catheterisation) because the diagnosis in all 11 neonates who died was found to be complete and correct at operation. The individual diagnoses for the seven neonates who died after operation without cardiac catheterisation were: type I or type II common arterial trunk (three patients), coarctation with ventricular septal defect (two patients), pulmonary atresia with intact septum requiring pulmonary valvotomy and a shunt procedure (one patient), and cerebral arteriovenous fistula (one patient).

Discussion

Cross sectional echocardiography is a reliable method for identification of intracardiac morphology, and it can provide better information than cardiac catheterisation and angiography.¹¹ In most children extracardiac vascular structures can also be identified reliably.¹²⁻¹⁴ With the current improvements in echocardiographic machines it is possible to categorise completely and systematically most congenital cardiac malformations. These echocardiographic findings have been extensively validated both at operation and necropsy and numerous studies have testified to the benefits of cross sectional echocardiography in the morphological assessment of congenital heart disease.

NEONATES AND INFANTS

In our experience cross sectional echocardiography had the biggest impact on the diagnostic assessment of sick neonates and infants. Cardiac catheterisation in this group is associated with considerable morbidity.^{15,16} In most neonates with symptoms and an obstructive lesion of the outflow tract (for example critical aortic or pulmonary valve stenosis, coarctation of the aorta, or aortic arch interruption) operation can be based on morphology. For neonates with more complex malformations who require palliative procedures to regulate pulmonary blood flow (pulmonary artery banding or an aortopulmonary shunt) the surgical referral can be made on echocardiographic findings alone.¹⁷ In 1987 and 1988, only 4/20 neonates who required an emergency shunt procedure and 1/5 who required pulmonary artery banding had prior catheterisation. They needed angiography for better definition of the size and distribution of the pulmonary vasculature.

Cross sectional imaging may be inadequate in several situations. Haemodynamic information such as the estimation of outflow tract gradients, valve regurgitation, or multiple sites of intracardiac shunting require additional techniques to assess blood flow. Pulsed and continuous wave Doppler echocardiography are complementary to cross sectional imaging in this assessment and have also aided identification of extracardiac abnormalities including coarctation of the aorta, interruption of the aortic arch, anomalous pulmonary venous drainage, or a ductus arteriosus.¹⁸⁻²⁰ It has become possible reliably to derive the pulmonary artery systolic pressure in a large propor-

tion of patients from Doppler echocardiography by measuring the peak velocity of tricuspid regurgitation; for ventricular septal defects similar estimates can be made by measuring the velocity of the jet of the ventricular septal defect.^{21,22}

The introduction of colour flow mapping in 1987 provided several additional advantages in diagnosis. In ventricular septal defects, colour flow mapping has proved more sensitive than angiography in the diagnosis of multiple defects.²³ With isolated defects, direct visualisation of the ventricular septal defect jet has enabled easier alignment of the jet for continuous wave Doppler interrogation. Colour flow mapping aided the assessment of valve regurgitation and helped in the recognition of extracardiac shunts such as patent ducts (25/25 neonates and infants had duct ligation without catheterisation in 1987/88), particularly in the sick preterm infant in whom the characteristic clinical signs are often absent and imaging alone is often unsatisfactory.²⁴ Pulmonary vascular structures are more easily identified in complex defects such as pulmonary atresia.²⁵ Other lesions for which corrective surgery is now being regularly performed without prior catheterisation include total anomalous pulmonary venous connection (7/7 patients in 1987/88), complete atrioventricular septal defect (7/10 patients), and common arterial trunk (10/11 patients).

The routine use of these techniques has therefore resulted in an important change in our clinical practice, shifting the emphasis from cardiac catheterisation and angiography towards ultrasound assessment. This is particularly true for palliative or corrective surgical procedures in the neonate that do not require cardiopulmonary bypass.

OLDER CHILDREN

Older children with simple lesions such as uncomplicated secundum atrial septal defect are referred for operation on the clinical and ultrasound findings alone. For 1987/88, 58/59 patients with secundum atrial septal defects were referred for operation without catheterisation. Similarly, all arterial ducts were ligated without additional investigation.

The relatively high catheterisation rate in the older children is related to several factors. In 1980 to 1984 after the introduction of cross sectional imaging, virtually every child who underwent surgery had preoperative cardiac catheterisation. First, this was the period during which physicians and surgeons assessed the value of the technique. It has been the policy of the paediatric cardiologists to measure the haemodynamic variables at catheterisation in most patients over the age of one year with moderate sized left to right shunts. Cardiac catheterisation was undertaken to measure the shunt and assess the pulmonary vascular resistance in 6/11 children who underwent closure of an isolated septal defect in 1987/88. Additionally, the relative values of echocardiography and cardiac catheterisation for the identification and characterisation of specific lesions were compared in several studies during

this period; this accounted for some of the cardiac catheterisation procedures.²³⁻²⁵ The catheterisation rate is also related to the problems inherent in the currently available methods for non-invasive diagnosis. With increasing age the available echocardiographic window may be reduced as the chest wall and lungs become bigger. Many of the older patients had also undergone palliative or corrective procedures at a younger age, with further reduction in echocardiographic visualisation because of acquired adhesions or implanted prosthetic material within the heart.²⁶ Thus in older patients with coarctation of the aorta suprasternal imaging often provides less satisfactory images of the arch. During this study, balloon angioplasty of coarctation was not undertaken and all patients with native coarctation or recoarctation underwent surgical repair. Preoperative cardiac catheterisation was performed in 12/14 children in 1987/88. For complex cyanotic heart disease requiring corrective surgery cardiac catheterisation was invariably required to assess the pulmonary vasculature and to calculate the pulmonary vascular resistance. All children undergoing a Fontan procedure, repair for tetralogy of Fallot, or correction of pulmonary atresia had cardiac catheterisation in the six months before operation.

In summary, the aim of any diagnostic technique is to provide the information necessary for correct management decisions. Although there has been a progressive fall in the ratio of diagnostic catheterisations to operations for all age groups over the nine years of review, this is the result of a combination of factors, and it is difficult to isolate the role of any individual diagnostic technique. Thus while significant falls in diagnostic catheterisation were associated with the introduction of pulsed/continuous wave Doppler and subsequently of colour flow mapping, the increasing experience with cross sectional echocardiography, improvements in image resolution, and the surgeons' confidence with the technique may all have contributed. Areas of inaccuracies with echocardiography, such as with multiple ventricular septal defects or complex pulmonary atresia, are also sometimes poorly defined by cardiac catheterisation and angiography. It is also reassuring that the avoidance of cardiac catheterisation did not significantly affect surgical mortality. On the basis of our experience, we conclude that with good quality imaging and Doppler echocardiography it is both feasible and safe to avoid cardiac catheterisation in selected patients with congenital heart disease. The growing acceptance of these techniques by surgeons is reflected in the increasing number of patients of all age groups undergoing cardiac operation on the basis of ultrasound findings alone.

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