Editorial

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Cross sectional echocardiographic diagnosis of congenital heart disease in infants

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The goal of any method of medical diagnosis is that it should obtain the maximum possible accurate, relevant information at the minimum cost in time, expense, discomfort, and risk. To what extent has cross sectional echocardiography assisted achieving this goal in infants? Two factors immediately stand out in the technique's favour. First, infants have superb subcostal and suprasternal windows. Second, conventional clinical diagnosis at this age using physical examination, chest radiography, and electrocardiography is notoriously unreliable, particularly in the first two months of life. For this reason, cardiac catheterisation and angiocardiography have for many years been the standard diagnostic methods in this age group.

Bearing in mind the goal already set forth for a diagnostic method, the time and money spent on cross sectional echocardiography can be justified on only three grounds, (a) that it reduces the risk of invasive investigation, (b) that it supplements invasive investigation by supplying some information which cardiac catheterisation and angiocardiography cannot do, and (c) that it can replace cardiac catheterisation and angiocardiography, at least in certain situations.

Reduction of risk of cardiac catheterisation

As yet, we have no unequivocal evidence that cross sectional echocardiography achieves this. There are, however, several good reasons why it might. During catheterisation of infants, and sick neonates in particular, time is precious, because throughout the cardiac catheterisation the infant usually becomes progressively more acidotic and hypothermic, and suffers progressively more side effects from the contrast medium. Cross sectional echocardiography should save time in cardiac catheterisation because of prior demonstration both of the route of access to the heart and of the presence or absence of normal or abnormal communications within the heart or between the heart and great arteries. For example, interruption of the inferior vena cava with azygos continuation makes catheterisation of the patient from the usual saphenous or femoral approach technically tedious. This abnormality is easily recognised by cross sectional echocardiography[1] before cardiac catheterisation and, if found, argues strongly for catheterisation from the axilla instead. Again, some of the most serious problems occurring during cardiac catheterisation arise from attempts to pass a catheter across a communication that does not exist. Misguided attempts to enter the pulmonary artery from the right ventricle in the presence of pulmonary atresia may produce severe bradycardia, heart block, and myocardial trauma or even perforation. If cross sectional echocardiography has already shown that there is no pulmonary trunk, such futile manoeuvres will not be attempted. Contrariwise, prior knowledge of the presence of an abnormal communication will encourage the investigator to make use of it. Demonstration of a ventricular septal defect in the subaortic position[2] when the cardiac connections are normal will mean that retrograde arterial catheterisation to reach the ascending aorta is unlikely to be required. The investigator, knowing that there is a hole there, will feel much more confident about manoeuvring the catheter so as to go through it. Cross sectional echocardiography can also reduce the amount of contrast medium given to the patient, both by judicious use of contrast echocardiography, and by enabling better planning of the angiocardiographic projections required. Contrast echocardiography is probably a better method than angiocardiography for showing atrioventricular valve regurgitation, as it is just as sensitive. It is also probably more selective, since spurious regurgitation resulting from high pressure injection and induction of ectopic beats can be avoided. The planning of angiocardiographic projections is particularly important in conditions where the orientation of the ventricular septum is not predictable, as for example in hearts with univentricular atrioventricular connections, and in cardiac malpositions. Finally, there is some evidence that limitation of the complexity of the initial investigation in patients with complete trans-

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position of the great arteries is beneficial. Nowadays, cross sectional echocardiographic diagnosis of complete transposition of the great arteries is so reliable that the first catheter inserted into the patient is a balloon catheter for atrial septostomy. The dramatic immediate improvement this produces in the state of the patient must surely reduce the overall risk of the procedure.

Supplementation of information provided by angiocardiography

Any structure within the cardiac cavities whose width is less than about 2 mm creates severe problems for angiocardiography. If the structure is thin in one plane only, it can be satisfactorily imaged only if the projection is absolutely right, or if the injection of contrast medium can be arranged in such a way that there is a difference in opacification of blood on the two sides of the structure. By contrast, a thin structure within the heart usually produces superb reflections of the echo beam, such that there is no difficulty at all in seeing it. Thus, its nature ensures that cross sectional echocardiography far outperforms angiocardiography in showing structures such as the atrial septum,[3] subaortic diaphragms,[4 5] membranes within the left atrium as in cor triatriatum,[6 7] valve leaflets,[8 9] and the Eustachian valve and Chiari network.[10] For the same reason, abnormalities in these structures are also much more reliably demonstrated, for example isolated cleft of the anterior mitral leaflet,[11] and Ebstein's anomaly of the tricuspid valve.[9 12]

If the structure is thin in two planes, it becomes impossible to image it with angiocardiography. This is why echocardiography is the only technique currently capable of showing the tension apparatus of atrioventricular valves. This explains its superiority in demonstrating straddling atrioventricular valves,[13 14] the detailed nature of atrioventricular septal defects,[15 16] anomalous insertion of the tricuspid valve into the underside of the infundibular septum,[17] and anomalous attachment of the mitral valve into the ventricular septum creating left ventricular outflow tract obstruction (JF Smallhorn, 1983, personal communication).

Angiocardiography produces a projection, whereas echocardiography produces a cross section. This fundamental difference in what is imaged also makes the two techniques complementary. We have found the combination of echocardiography and angiocardiography particularly helpful in sorting out the maximally complex problems presented by hearts with criss-cross atrioventricular connections or superoinferior ventricles. In order to identify the atrioventricular connection in these hearts, it is necessary not only to recognise the morphology of the atria and ventricles, but also to recognise their precise connection. At present, angiocardiography is probably superior in identifying the ventricular morphology, but its ability to demonstrate the connection is limited by the overlapping of structures that is almost bound to occur in a projection. In a cross section, such overlapping cannot occur.

Circumstances where cardiac catheterisation can be replaced

There is now no variety of congenital heart disease which cannot be diagnosed with considerable accuracy by cross sectional echocardiography. We have recently reviewed the field in infants in detail.[18] Conditions not already mentioned that are of particular importance in newborns and young infants are atrial situs,[19] total anomalous pulmonary venous connection,[20 21] absent left and right atrioventricular connection,[22] left heart hypoplasia,[23] double inlet ventricle,[22 24–26] ventricular septal defect (in particular precise localisation of the defect),[2] tetralogy of Fallot,[27] pulmonary atresia with intact ventricular septum,[28] double outlet right ventricle,[27 29] complete[30 31] and corrected[32] transposition, aortopulmonary window,[33 34] truncus arteriosus,[33 35] interrupted aortic arch,[36] anomalous origin of one pulmonary artery from the aorta,[33] or the left coronary artery from the pulmonary trunk,[37] persistent ductus arteriosus,[38] and coarctation of the aorta.[39] The true answer to the question of when it is justifiable to replace invasive investigation with non-invasive investigation based upon cross-sectional echocardiography depends upon formal application of decision theory to the problem (FJ Macartney, J Douglas, and D Spiegelhalter, unpublished observations). Unfortunately, the complete information necessary to apply this approach is not available for any condition, including as it does the sensitivity and specificity of both invasive and non-invasive investigation, and the incrementation of risk associated with cardiac surgery produced by prior invasive investigation. Such advances as there have been have therefore relied heavily on intuition. We have recently reported 70 patients operated on for congenital heart disease without prior invasive investigation, who according to our previous policy, would have all required it.[40] The majority of these patients were infants, and the conditions involved ranged from coarctation of the aorta through critical aortic stenosis to such complex problems as total anomalous pulmonary venous connection and persistent truncus arteriosus. Though some seven errors in diagnosis were made, in no case did the error result in the death of the patient, and in only one case did it result in any morbidity, in that an unnecessary thoracotomy was carried out for a patient with abdominal coarctation.

It is instructive to compare this experience with that at the Mayo Clinic, recently reported.[41] Almost all our patients had obstructions to the left heart or predominantly left to right shunts. By contrast, they carried out shunt operations on a number of infants with reduced pulmonary blood flow on the basis of clinical examination and cross sectional echocardiography alone. These differences probably reflect variations in confidence in particular diagnoses, as well as in surgical approach to the problems involved. Put together, however, they span most of congenital heart disease in infancy.

These results are clearly preliminary, and the number of patients submitted to operation without cardiac catheterisation is still a small fraction of the total. There seems, however, little doubt that considerable progress in this area will be made, provided that certain principles are borne in mind.

Sending a patient for operation without prior cardiac catheterisation concentrates the mind wonderfully. It reinforces very strongly the need for cross sectional echocardiography to be viewed as a part of the whole clinical investigation of the child. Unless everything is typical for the condition concerned, one should not hesitate to carry out cardiac catheterisation and angiocardiography. The objective of cross sectional echocardiography must be far more than simply that of identifying the principal abnormality present. The goal must be to identify every cardiac structure and every major blood vessel in the thorax. Sometimes it may be necessary to sedate the child for the cross sectional echocardiogram in order to do this, because, unfortunately, the two most important windows for examination, the suprasternal and subcostal, are also those that cause most discomfort to the infant. It is vital that the limitations of the technique should be understood both by surgeons and physicians. For example, it is impossible to rule out a patent foramen ovale, ventricular septal defect, or persistent ductus arteriosus, if any of them are too small to be within the resolution of the instrument used. The problems of near field resolution in some instruments produce problems with adequate demonstration of the right ventricular outflow tract, though these can usually be solved by obtaining a high parasternal long axis cut running through the right ventricular outflow tract to the anterior great artery. Though criteria have been proposed for distinguishing between dropout and defect, in many instances this is a matter of art rather than science. Finally, there are some patients, particularly those who have been on prolonged ventilation, who simply do not have an adequate window for echocardiography. If the echocardiographic windows are poor, cardiac catheterisation will be unavoidable.

Because of advances in ultrasound examination of the brain and abdomen, ultrasound scanners are slowly becoming standard equipment in special care baby units throughout the country. This may have profound implications for the pattern of referral of neonates with congenital heart disease in the future. The arguments for supraregional centres for infant cardiac surgery[42,43] remain as strong as ever, because of the need for concentration of facilities for infant heart surgery. In the past, however, such specialist centres were also necessary because they were the only ones where an accurate diagnosis of congenital heart disease in infants could be obtained. Of necessity, the price paid for early referral of patients with critical heart disease was the unnecessary transfer of patients with normal hearts or non-critical heart disease to these specialist centres. Does the advent of cross sectional echocardiography mean that there is a role in the district general hospital for a paediatrician or cardiologist with particular experience of congenital heart disease in newborns and young infants? Our initial experience has been discouraging. Far from expediting the transfer of infants who need urgent treatment, cross sectional echocardiography in unskilled hands has on occasion delayed referral because of failure to recognise even such obvious abnormalities as a large ventricular septal defect. This emphasises how much technical success depends on the quality of the operator. It demands not only considerable manual dexterity, but a profound knowledge of both normal and abnormal cardiac anatomy. Without these, the unskilled operator is likely to fail to record what is not recognised as significant. These problems are compounded if machines are used which have been selected for their suitability for scanning adult patients. The key to successful imaging in infants is high resolution, not penetration.

These problems will probably be solved by providing adequate training in the technique in infants, but this will not be achieved overnight. Though there is still plenty of room for improvement both in instrumentation and in operating skills, cross sectional echocardiography has undoubtedly caused the biggest revolution in the management of congenital heart disease since the advent of open heart surgery.

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