Introduction

Cardiac ultrasound and congenital heart disease

Cardiac ultrasound and the study of congenital cardiac malformations seem to be made for each other. The understanding of cardiac malformations has improved in parallel with technological advances in ultrasound. Development of both spheres of interest has been remarkable for the rapidity of change, the increasing complexity of the lesions treated, and the technology applied.

Paediatric cardiologists in the 1960s and '70s depended on long and difficult catheterisation procedures on small blue and sick infants to make accurate diagnoses. These were often performed as an emergency so that the treatable lesions such as transposition of the great arteries and coarctation syndrome could be recognised early and sent for surgery.

It would have been difficult to predict the utility of cardiac ultrasound for diagnosing neonatal congenital heart disease from the early reports by Edler and Gustavson demonstrating echos from the mitral valve with an industrial device originally used for the examination of metal fatigue.[1] Cardiac ultrasound consisted only of M mode with its high resolution, rapid repetition, but very limited view of the heart. As such it was not an obvious diagnostic tool for complex congenital cardiac malformations.

In the paediatric cardiology unit in Newcastle around this time, serendipity took a hand. An echocardiogram on a blue neonate was suddenly and dramatically altered by the flushing of a dextrose infusion as the houseman set up a new drip. A literature review revealed that the ultrasonic effect which we had produced was well recognised following injection of saline, blood, dextrose or even carbon dioxide into the circulation, although nobody seemed to have found a clinical use for the phenomenon. Using the suprasternal approach where the aorta, right pulmonary artery, and left atrium have an almost universal relation, and small injections of 5% dextrose agitated before introduction into the circulation, we were able to produce a cheap and sophisticated method of demonstrating right to left shunts and abnormalities of ventriculo-arterial connections such as transposition.[2]

The usefulness of this technique was short lived because it was overtaken by the arrival of cross sectional echocardiography. The early linear arrays had disadvantages because the ribs got in the way. For cardiac use the newly developed phased array with smaller transducer footprints which allowed the ultrasound to pass between the ribs was the development which made the biggest step forward.

The paediatric cardiological fraternity no longer needed to spend sweaty hours under lead aprons in the middle of the night. The new ultrasonic devices gave good enough information to allow the surgical cases to be recognised without catheterisation and, as important, identified non-structural heart disease which previously had accounted for a significant number of emergency catheterisation procedures. It is ironic that in the 1960s adult cardiologists were seldom out of their beds at night, while today they commonly perform interventions out of hours. On the other hand, cardiac catheterisation as an acute emergency is a rarity in paediatric cardiology.

New nomenclature

In the 1970s, with the improvements in surgical results for congenital cardiac malformations, there was a welcome move towards a new nomenclature. The old naming of congenital cardiac conditions was a horrible mixture of eponyms, bad latin, and embryological probabilities. In Europe and increasingly elsewhere, a descriptive and sequential approach to the malformed heart with a new and better appreciation of cardiac anatomy greatly clarified congenital heart disease.[3] This descriptive approach developed hand in hand with cardiac ultrasound. Particularly in the small infant, most parts of the heart are easily approached with cardiac ultrasound and the anatomical detail to use the descriptive sequential approach is readily available. As the patient with congenital heart disease gets older and bigger, it becomes a little more difficult to achieve the same totality of diagnostic information. Again, technology has radically improved our options in the shape of transoesophageal echocardiography. Interventional closure of atrial septal defects by catheter introduced devices is much easier and less dangerous because of the ability of transoesophageal echocardiography to monitor the procedure and ensure the correct situation of the occluder.

In the early 1970s and '80s, Ronald Pridie and I ran training courses in echocardiography. We invited speakers to talk to the delegates about new techniques. The biggest change in our clinical practice came when Liv Hatle from Norway showed us how to use Doppler to assess haemodynamic events. First, spectral Doppler and then colour flow mapping revolutionised the assessment of congenital heart disease to such an extent that, while carrying out a recent review of the natural history of ventricular septal defects, we concluded that studies before the advent of colour flow mapping and spectral Doppler were no longer worth considering or likely to be accurate.[4] Furthermore, a recent prospective screening programme of normal asymptomatic neonates in Northern Ireland showed that up to 4% of normal infants have small, clinically muscular, insignificant ventricular septal defects.[5]

Assessing intracardiac pressures

A major reason to catheterise children with congenital cardiac malformations was the assessment of intracardiac pressures, particularly in the pulmonary circulation. Doppler studies have shown for some time that tricuspid valvar regurgitation is very common even when not clinically apparent. Application of the Bernoulli equation to this finding allows us to estimate right sided systolic pressures. Similarly, right sided systolic pressures may be predicted with some accuracy in the presence of ventricular septal defects by studying the peak velocity of the

transseptal jet. Cardiac catheterisation for purely haemodynamic reasons is now a relative rarity in paediatric cardiology.

Finally and predictably, the diagnosis of congenital cardiac malformations has become part of prenatal screening for fetal abnormalities. Prenatal diagnosis of non-cardiac abnormalities has been around for some time and has helped to bring about the virtual disappearance of some lesions such as spina bifida. There is now good evidence, based on prospective data, that the early recognition of cardiac malformations before birth can significantly alter the prevalence of complex congenital heart disease.[6] As the technology improves and the skill of the operator follows suit, it is reasonable to expect that more and more cases of congenital heart disease will be recognised in utero. In addition to the effect of termination of pregnancy under these circumstances, there is also evidence that early diagnosis before birth may lead to better outcome in individual lesions, such as transposition of the great arteries after birth.[7]

STEWART HUNTER

Department of Paediatric Cardiology,
Freeman Hospital
Newcastle Upon Tyne, UK

1 Edler I, Gustavson A. Ultrasonic cardiogram in mitral stenosis. *Acta Med Scand* 1957;**159**:85.
2 Mortera C, Hunter S, Tynan M. Contrast echocardiography and the suprasternal approach in infants and children. *Eur J Cardiol* 1979;**9**:437–54.
3 Shinebourne EA, McCartney FJ, Anderson RH. Sequential chamber localisation—logical approach to diagnosis in congenital heart disease. *Br Heart J* 1976;**38**:327–40.
4 Turner SW, Hunter S, Wyllie JP. The natural history of ventricular septal defects. *Arch Dis Child* 1999;**81**:413–6.
5 Sands A, Lynch C, Casey F, *et al*. Ventricular septal defects; the relationship of social class and area of residence to occurrence rate [abstract]. *Fetal Diagn Ther* 1998;**13**(suppl 1):148.
6 Hunter S, Heads, A, Wyllie J P, *et al*. Prenatal diagnosis of congenital heart disease in the northern region of England: benefits of a training programme for obstetric ultrasonographers. *Heart* 2000;**84**: 294–8.
7 Bonnet D, Coltri A, Butera G, *et al*. Fetal detection of transposition of the great arteries reduces morbidity and mortality in newborn infants. *Circulation* 1999;**99**:916–8.