Personal practice

Outpatient paediatric cardiological investigation

An alternative approach

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SUMMARY The use of current technology has allowed an alternative approach for the investigation of children with cardiac disease. A purpose-built unit is described.

Detailed investigation of infants and children with congenital heart disease is traditionally by inpatient admission for invasive study by cardiac catheterisation. This is both expensive for the NHS and traumatic for the patient and parents. In 1976 we wondered whether sufficient knowledge of cardiac anatomy, pathology, and functional disturbance could be obtained in many of our patients by purely outpatient investigation, using new diagnostic techniques. We were fortunate in having the support of the Variety Club of Great Britain to develop this concept. With their help and enthusiasm a purpose built outpatient investigation unit for paediatric cardiology was constructed. The unit (Fig. 1) was opened on 27 July 1977 and this paper gives the reasons for its development and our experience during the initial 18 months of operation.

Indications for considering a new approach

Changing work load. The work of paediatric cardiologists is changing, largely as a result of clinical experience and improved surgical techniques. In addition, a greater general awareness of congenital heart disease has resulted in more newborn infants and young asymptomatic children being referred for cardiological assessment. Investigation is generally conducted so that the child can have corrective surgery (when this is necessary) before he starts school. Nowadays much of the work concerns the long-term follow-up after cardiac surgery,

particularly of children with prosthetic valves and graft material.

New techniques. The most important advances in noninvasive techniques have been echocardiography and other forms of ultrasound, and ambulatory electrocardiographic monitoring. The development of very small soft polyethylene balloon-tipped catheters of the type described by Swan *et al.*¹ has made the previously quite complicated technique of direct recording of intracardiac data a relatively simple technique.

Problems associated with invasive cardiac investigations.

Expense

In our practice paediatric cardiac catheterisation requires a 48-hour hospital admission. Added to this, the expense of the procedure itself, with the staff and equipment, amounts to approximately £300 for a typical study.

Mortality/morbidity

Catheterisation techniques have improved but there is still an inevitable morbidity and, to a small degree, a mortality.²

In view of these developments one must ask whether all the previously accepted indications for inpatient cardiac investigations are now necessary, and if not, what is a suitable alternative. For these reasons our outpatient investigation unit was developed.

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Fig. 1 The purpose built outpatient cardiac investigation unit.

Requirements and design of outpatients investigation unit

In an attempt to gain children's confidence the building was designed in a most unusual style, not at all reminiscent of a hospital (Fig. 1). Briefly, there is a large central area shared by secretarial staff and the patients. Numerous toys and a colour television set are available for patients, and there is a nearby 'open' kitchen for waiting parents to help themselves to refreshments. While not every child requires all the available investigations, the design is such that those who do, progress in a clockwise direction from one laboratory to another. The sequence is an examination/consultation room, followed by the echocardiography laboratory, then to phonocardiography, exercise testing, electrocardiography/vectorcardiography and 24-hour ambulatory monitoring analysis, and finally, to semi-invasive studies (dye dilution curves and right heart pressure studies). The unit has its own photographic processing laboratory and a reporting room in which the computer is kept with its accessories for data analysis.

Techniques of investigation

Echocardiography. This ultrasound technique is available in two forms.

Single crystal or M-mode

Echocardiography, where a hand-held transducer is positioned on the anterior chest wall (usually in the 4th left rib space initially) and 'angled' in order to locate intracardiac structures. In view of the diversity of cardiac positions, several transducer locations on the chest are tried until satisfactory recordings are obtained. There is an in-built scale for 'depth' of structures, and with a known speed of recording, data on both the vertical and horizontal axes can be

calculated. The technique provides a permanent quantifiable record of intracardiac chamber dimensions, wall thicknesses, and rates of valve movements.

Real time echocardiography

A system which produces a moving picture, recording events as they happen is said to be 'real time'. In echocardiography this can be accomplished by having several crystals close together forming a large multi-crystal transducer, the repetitive discharging and recording of each component adding together to 'build up' an image of the heart. An alternative method is for one transducer crystal to be rotated rapidly through an arc, or sector, over the heart thereby building up by repetitive scans a moving cardiac image. In either system, the data are visualised on a screen and recorded on video tape for storage and re-evaluation.

Either M-mode or real time echocardiography can be complemented by using the 'contrast echo technique'. The contrast is not the type used in radiology but is an intravenous bolus of any physiological solution. Microscopic bubbles are present in solutions—for example normal saline or dextrose—because deaerated water is difficult to obtain. Generally, these bubbles cause no problems because they are efficiently filtered out by the lungs when injected intravenously and do not enter the left side of the heart. The bubbles in such a bolus produce a 'shower of echoes' as the solution passes through the right heart (only entering the left atrium or ventricle if there is an intracardiac septal defect). The technique aids the clarification of valves, cardiac chambers, and blood flow patterns within the heart.

Doppler ultrasound. This is another variation of ultrasound, where a continuous emission of high frequency sound waves pass from a small hand-held probe which is located over a superficial artery or vein. The sound waves pass through the vessel wall

and other static structures and are reflected at the original frequency back to the transducer, but moving structures—for example red blood cells—alter the frequency of reflected sound waves. The degree of frequency change is proportional to the velocity of the moving particles, the frequency shift being the 'doppler signal'. This principle is used in general paediatrics for detecting blood flow for blood pressure recording purposes, but in cardiology the technique can record blood flow patterns in the neck veins or arteries. The traces are recorded simultaneously with the phonocardiogram. In addition to giving improved pulse recordings, some measurable and diagnostic uses have been described³ and there will almost certainly be more applications.

Phonocardiography. Despite being one of the older noninvasive cardiac investigations, this technique still has considerable application not only for recording heart sounds, added sounds, and murmurs but for recording their relationship in time to other events in the cardiac cycle. Other simultaneously recorded parameters include respiration, arterial and venous pulsations, and the cardiac apex pulsation (apexcardiogram).

Clinical correlations from phonocardiography and its allied techniques are useful in certain instances—for example, for estimating the approximate pulmonary arterial systolic pressure,⁴ confirming the movement of the second sounds, and for confirming the presence and timing of 'clicks' and 'snaps'. With progressively more simultaneous recordings to add to the phonocardiogram (such as echocardiograms) it continues to be an interesting clinical aid and research technique.

Vectorcardiography. The principles of this 3-dimensional electrocardiographic technique are well established. In many instances sufficient electrocardiographic data can be obtained from the standard ECG but if doubt or confusion exists the vector cardiogram is often helpful. There are techniques for quantitation of degrees of cardiac hypertrophy, valve gradients, and the magnitude of intracardiac shunts from the vectorcardiogram alone, but we consider the results of this technique in conjunction with other parameters (for example, echoes).

Dye dilution curves. The measurement of an intracardiac shunt, or confirmation of its absence, is often required in paediatric cardiology. We do dye dilution curves using an ear oximeter (which is a photoelectric cell clipped on to an ear lobe) connected to a 'densitometer' and recorder (Fig. 2). The dye (indocyanine green) is injected intravenously via a

Fig. 2 Two dye curves obtained by injecting indocyanine green (5 mg/ml) intravenously, sampling with an ear oximeter in a child of 4 years with a secundum atrial septal defect.

percutaneous needle puncture at the antecubital fossa. With reassurance and in the presence of a parent this can generally be accomplished without too much upset, which, in turn, allows more accurate measurement.

Right heart pressure studies. Through the small cannula already inserted for dye curves (described above) a soft polythelene catheter with an inflatable balloon at its tip is inserted. When advanced sufficiently for the balloon to be in the region of the superior vena cava the balloon is inflated, which has the effect of drawing the catheter along with the 'tide' of blood flow to the right atrium, through to the right ventricle and usually on to the pulmonary artery. The catheter is connected via a pressure transducer to an oscilloscope and hard copy recorder so that the changing wave formats indicate the position of the catheter tip (Fig. 3). The technique is therefore accomplished without radiological

Fig. 3 Directly recorded intracardiac pressure recording from a balloon-tipped catheter advanced percutaneously without radiological control in a child of 3 years with pulmonary valve stenosis. Note the initially normal pulmonary arterial trace followed by a moderately raised right ventricular trace as the catheter is withdrawn from pulmonary artery to right atrium.

screening and is thus eminently suited to an outpatient service.

We find the percutaneous technique for these studies is a practical procedure in children who are more than 15 kg.

Ambulatory ECG monitoring. The development of this technique has revolutionised the investigation and management of children with, or suspected of having, cardiac arrhythmias. Instead of hospital admission and bedside monitoring with its enforced bed rest, the child can now be at home or at school following all normal activities yet with the ECG being constantly recorded. The equipment has a small cassette tape (of the type used for domestic music recording) and a miniature tape recorder. The ECG signal is derived from two chest electrodes, the recorder being supported by a shoulder strap and carrying case. The device is small enough to be worn for long periods (several days) by children of all ages. Each tape (one side only) records continuously for 24 hours. On return to the unit the tape is replayed in an analysing unit at 60 times recording speed—that is it takes approximately 45 minutes to analyse one 24-hour recording, including printing out ECG strips of interesting portions of the tape. The recorder also incorporates a 24-hour clock, enabling precise timing of the occurrence of abnormalities which can therefore be correlated with any symptoms.

Clinical applications of outpatient investigation

Paediatric cardiological problems can be divided into the following:

Uncomplicated defects. These are the more common conditions such as pulmonary stenosis, atrial or ventricular septal defect, or mitral prolapse (Fig. 4). In the past, if any of these lesions was thought to be significant the patients would have been admitted for cardiac catheterisation. Now they first attend for

Fig. 4 Two M-mode echocardiograph traces of the mitral valve area. On the left, the valve is normal, on the right, the arrows indicate systolic prolapse.

outpatient investigation and only children in whom results suggest that angiography would be of value are subsequently admitted for conventional cardiac catheterisation. While the investigations performed depend on the clinical problems, most children in this group are studied by phono- and echocardiography, vectorcardiography, dye curves, and right heart pressure studies. These outpatient investigations combine to clarify the situation so that many patients can be referred directly to surgery without further study, or alternatively reassured that surgery is not required. An example is shown (Fig. 5) of a child with slight pulmonary stenosis and no complicating septal defect indicating that surgery is unnecessary.

Complicated defects. Complex derangements of cardiac anatomy have to be clarified by angiography, but a surprisingly accurate assessment can be made by echocardiography. Real time echocardiography is of particular benefit in this group, so we generally start investigations with this technique, progressing to the M-mode echocardiograph and later, if necessary, to contrast echocardiography before

Fig. 5 A normal dye curve with the directly recorded pressures added demonstrating a systolic pressure gradient of 14 mmHg between right ventricle (RV) and pulmonary artery (PA), confirming the diagnosis of pulmonary stenosis but indicating no intervention required.

proceeding with cardiac catheterisation. Echocardiography helps considerably to shorten the duration of cardiac catheterisation, principally by suggesting the choice and projection of angiography. The echocardiogram often additionally helps as an adjunct to angiography in clarifying details of the ventricular septum and of the atrioventricular valves.

In this group are also included all types of left ventricular outflow tract obstructions. The severity of obstruction is difficult to assess without cardiac catheterisation. However the combination of several noninvasive parameters, principally the electro- and vector-cardiograms and M-mode echocardiogram with the apex cardiogram, indicate the approximate category—slight, moderate, or severe left ventricular obstruction. More definite quantitation still requires invasive study, particularly when assessing not only the resting haemodynamics, but the response to exercise (which is important when planning the timing of surgical intervention), but the timing of the invasive study can be optimally arranged by prior outpatient studies.

Whether the defect is of complex anatomy or left-sided obstruction, once the problem has been clarified by catheterisation and angiography, the progression of the disease can be followed by repeated outpatient noninvasive assessment, and further invasive studies before surgery avoided unless there is some unexpected development.

Neonates.

Babies with obvious congenital heart disease

These are all investigated initially by echocardiography as inpatients, avoiding subjection to invasive investigation if an adequate diagnosis can be made (for example, hypoplastic left heart). In others, the probable diagnosis can be established by the echo—for example total anomalous pulmonary venous drainage, single ventricle, truncus, etc.—although invasive study may still be required, either because it is essential to confirm the anatomy or, as in the usual type of transposition of the great vessels, to perform a balloon atrial septostomy.

Ill babies

Any baby in whom the paediatrician is anxious to exclude heart disease is usually studied in the outpatient investigation unit and, if heart disease is excluded, the baby is directly returned without admission.

Postoperative. One of the most interesting aspects in the management of congenital heart disease is the follow-up after surgery. The long-term function of the myocardium, of prosthetic valves, conduits, and arrhythmic problems will not be fully appreciated for

many years. A continuous follow-up programme is necessary and here the outpatient investigation service is very important. As repeated invasive catheterisation studies are unacceptable to the patient and the physician, a follow-up based on regular non- or semi-invasive techniques is ideal. Changes in cardiac wall thickness or chamber size, with significant changes in ventricular or valve function can be appreciated by echocardiography. Flow-guided catheters readily check on postoperative pulmonary arterial and right ventricular pressures, with residual shunts being excluded or quantitated by dye curves.

Ambulatory monitoring. Symptoms—for example of syncope, or episodes of pallor or quietness—may be related to an intermittently abnormal cardiac rhythm. Wearing a tape recorder at the time of symptoms can easily establish the diagnosis. If however the child does not have typical symptoms during the time the tape is applied, changes may be seen which suggest an underlying cardiac reason for the symptoms. Of equal importance in children with symptoms, particularly of syncope, is the documentation of a normal cardiac rhythm throughout the period of symptoms, thereby excluding cardiac disease as the cause. It can be seen that it is very important to correlate changes on the tape with the occurrence of symptoms. To establish or refute a cardiac rhythm problem it is occasionally necessary to continue taping for several days, but generally sufficient information is available after three consecutive 24-hour tapes.

There is still uncertainty about the range of heart rates and transient rhythm variations which occur in normal children. This information is clearly required for correct interpretation, and therefore a study of ECG tape recordings in normal school-children is now well advanced as one of the research activities of this unit. Rhythm disturbances may, of

Fig. 6 An extract from a 24-hour ambulatory ECG tape recording in an 11-year-old girl with a history of syncopal episodes. Asystole occurred after the 4th QRS complex and was accompanied by loss of consciousness, with spontaneous recovery.

course, be naturally occurring or they can be induced by cardiac surgery, thus the monitoring of postoperative asymptomatic children is necessary and continues as another research project.

The drug treatment of symptomatic arrhythmias is often on an empirical basis as far as dosage and drug combination is concerned. Serum drug levels if available, are difficult to interpret, but of practical value is the ability to assess the efficiency of treatment throughout 24 hours by ambulatory 24-hour tapes.

Similarly, pacemakers in children can be difficult to assess in that often, when reviewed, the child's spontaneous heart rate is such that the pacemaker is inhibited. Although most pacemakers can be made to function temporarily in the fixed rate mode for the purposes of generator evaluation, it is not a satisfactory assessment of its natural function as a demand pacemaker. Rather than slowing the heart rate pharmacologically thereby permitting the pacemaker to function, a 24-hour tape can be used. This is because at some stage during a 24-hour period (usually at night) the heart rate will slow sufficiently for the pacemaker to be required. Clearly this is not necessary in the absence of symptoms, but can occasionally be applied to clarify normal or abnormal pacemaker function.

Exercise testing. Children with a history of a syncopal episode related to physical activity have this investigation in addition to 24-hour ambulatory monitoring.

Operational organisation

There is a daily appointment system and investigations are conducted by medical or technical staff working within the unit.

Patients attend in the following ways. New patients to the hospital attending for their first outpatient clinic visit have the usual standard ECG, chest x-ray, and M-mode echocardiogram. When seen for clinical assessment the attending paediatric cardiologist has the results of these. If further investigations are required, an appointment is made for a specific outpatient attendance at the unit.

When attending specifically for investigations (which altogether take about one hour) one parent is usually present. Children are not given any premedication or sedative and can return home or to school immediately after the investigation.

The investigation unit is linked to the main hospital so that inpatients are studied within the unit, and only occasionally is equipment transported within the hospital (the exception being to the intensive care unit).

The echocardiograms are analysed by a digitising device linked to a computer. Data and parameters are calculated and printed out with a complete record of all data being stored by the computer.

When all the investigations (dye curves, pressures, echoes etc.) have been analysed and reported for each patient a report summary, with a conclusion, is printed and forwarded to the referring paediatrician and general practitioner.

Future trends

The system has the major advantages of: minimal domestic disturbance to parents; minimal distress to the patient; general efficiency of investigation (for the medical staff); and financial saving for the NHS.

As a clinical service it will expand, particularly as technical developments give even greater accuracy of diagnosis by noninvasive means. There are other techniques currently available but their value has yet to be clarified, particularly within paediatric cardiology. I refer to the pulsed depth-gated doppler, impedance cardiograph, and the improved resolution now available in nuclear medicine technology. The resolution of ultrasound continues to improve resulting in more widespread application—for example, echocardiography is already helping to identify the areas of myocardium responsible for ectopic rhythm activity. Computer data analysis, particularly by integrating several noninvasive signals, will help the noninvasive predictions of cardiac overload to improve so that the flow-guided studies may then be avoided.

While computerised axial tomography (body scanning) is not yet being used in cardiology, refinements enabling sufficiently rapid filming will probably overcome the present difficulties so that we can look forward to this technique helping with details of intracardiac anatomy.

All these developing techniques will be used in outpatients departments and there is an exciting future for an outpatient paediatric cardiology investigation service. Clearly there will also remain for many years a requirement for cardiac catheterisation and angiography to clarify the anatomy in some complex defects (for example, for unusual types of pulmonary blood supply or coronary artery distribution).

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