Radionuclide investigation of congenital heart disease

In a previous issue of Heart, Verttukattil and colleagues presented a study of intrapulmonary arteriovenous shunting after a superior cavopulmonary anastomosis using radionuclide labelled albumin microspheres. Their study is unusual. Despite there being long established indications for radionuclide imaging in congenital heart disease, such investigations are not often performed. Equally they are not often used in research studies. Why are they not more widely used? There are many reasons. Some are shared with acquired heart disease, where the use of nuclear cardiology varies greatly between different centres. Cultural and economic reasons have been used to explain these differences in use. There are certainly differences between use in Europe and North American centres. There are particular factors that apply to congenital heart disease. Perhaps most importantly investigation of congenital heart disease is focused much more on structural rather than functional abnormalities. Radionuclide imaging has little to contribute in this regard. Cardiologists looking after patients with congenital heart disease use structural imaging, chiefly echocardiography and x-ray angiography, and now increasingly magnetic resonance imaging, on a daily basis. When functional information is needed they tend to stick to the techniques they know. The cardiologist who feels that M mode (one dimensional) echocardiography tells him or her all they need to know about a patient’s ventricular function is unlikely to consider radionuclide imaging despite the evidence of its value.1

There are other factors at play. The majority of diagnosis of congenital heart disease takes place in childhood, predominantly in very young children. Radionuclide imaging is, albeit minimally, invasive and uses ionising radiation. Paediatric cardiologists are naturally and properly reluctant to expose children to investigations involving intravenous cannulation and radiation without good reasons. The radiation dose from paediatric nuclear medicine procedures is low, certainly compared to cineangiography, but radionuclide investigations are only justified in children if they produce clinically useful information. For established, validated, and clinically important indications, such as the measurement of left-to-right shunts and the measurement of ventricular function, there should be no problem. But there is real concern about the use of new radionuclide techniques and new indications of established techniques, particularly as part of research protocols.

The problems of research

Research workers using radionuclide techniques to investigate congenital heart disease face formidable problems. Firstly there are the ethical issues about the use of radionuclides in children and secondly the problems of identifying normal controls. Take the example of the study of myocardial perfusion in patients who have had surgical relocation in patients who have had the arterial switch operation. In the former group right ventricular function can be measured by radionuclide techniques and right ventricular perfusion can be imaged using widely available radionuclides such as thallium-201 or technetium-99m labelled agents. While we can usefully measure the spectrum of right ventricular function in these circumstances, we have no idea of what constitutes normal function for a hypertrophied, systemic right ventricle coupled to a low pressure left ventricle. The situation of what constitutes normal myocardial perfusion for the right ventricle under these conditions is even more problematic. It might be imagined that the situation is more straightforward for imaging the left ventricle after the arterial switch. Not really. We do not know what normal left ventricular function or perfusion looks like in children. Nor can we ethically find out. Controls often have to be from groups of patients who are having radionuclide studies for other reasons such as shunt measurement. Their left ventricles may be devoid of obvious reasons for abnormal myocardial perfusion, but they are certainly not normal.

Lack of day to day experience and problems such as these have limited the amount of research that can be undertaken with radionuclide techniques in children with congenital heart disease. As the population grows up and reaches adulthood the ethical problems with informed consent are less, but the problems of interpretation largely remain. This should not put us off, however, for it is in adult patients with congenital heart disease, particularly long term survivors of surgery, that radionuclide techniques are most clinically valuable.

Clinical indications

While the structural abnormality of the heart itself is probably the most important determinant of immediate prognosis for patients with congenital heart disease, long term prognosis following surgery often depends upon cardiac function. Radionuclide investigations are valuable in the follow up of many of these patients. The value of gated studies using radionuclide labelling of the blood pool in the routine clinical measurement of ventricular function is well established. This technique needs to be used with care in hearts with very abnormal anatomy, since it requires the acquisition of views of the ventricles that exclude counts from other chambers. In practice this can usually be achieved for the left ventricle, but often not satisfactorily with the right ventricle. In either case, the projection used for analysis of ventricular function needs to be chosen carefully, taking into account the abnormal anatomy.

It is perhaps for the measurement of intracardiac shunts that radionuclide imaging is most often called on in the study of congenital heart defects. First pass radionuclide angiography is certainly an accurate way of measuring left-to-right shunts. The preferred method is based upon curve analysis of the time–activity curve acquired over the lung fields. Acquisition of first pass angiograms of sufficient quality for curve analysis and accurate analysis of the time–activity curve requires experience, so this is not a technique suitable for occasional use. Here we find exemplified one of the other problems with nuclear cardiology
for congenital heart disease. If a centre only uses it occasionally it will not yield accurate or reproducible results.

It is myocardial perfusion studies that have stood out as nuclear cardiology’s unique contribution in acquired heart disease. As we have seen, attempts have been made to extend this use in research studies in congenital heart disease. Is there ever a routine clinical indication? There is a paucity of evidence to support clinical decision making based on myocardial perfusion studies in congenital heart disease. They should be used with caution. Thallium is not suitable for children, the resolution is poorer and the radiation dose higher than with one of the variety of technetium labelled agents. The introduction of these agents and the improvement in tomographic imaging have greatly enhanced the quality of myocardial perfusion imaging that can be achieved. Interpretation is difficult and needs to take into account the anatomy and preceding clinical history. For instance, perfusion defects in these patients are often related to previous surgery rather than to the structural heart disease. We do not know what the implications of this for prognosis are. There is a pressing need for more research in this area, particularly in the long term survivors of surgery. It may be that positron emission tomography will be of value to study blood flow and other metabolic processes of the myocardium, but its limited availability is a major additional handicap to progress.10

Pulmonary perfusion studies

Vertukattil and colleagues’ paper describes the use of technetium labelled human albumin microspheres.1 These particles are typically 20 µm in diameter and are trapped in either the pulmonary or systemic capillaries. They can be expected to lodge in about 0.1% of the capillary bed. Such microspheres, or a similar preparation of macroaggregated albumin, have long been used for pulmonary perfusion studies. Where there is a right-to-left shunt some of the microspheres escape from the pulmonary circulation and lodge in the systemic capillaries. By estimating the counts in the lungs and in the systemic circulation, it is possible after an intravenous injection of microspheres to estimate the size of the right-to-left shunt. This is a well established, although little used, technique. The escape of microspheres into the systemic circulation has been shown to be safe; indeed one of the first indications for such microspheres was for cerebral scanning.12

A more frequent and growing use of radionuclide labelled microspheres is for pulmonary perfusion scanning in patients with congenital or postoperative obstructions to the pulmonary arteries. The use of intravascular stents has had a major impact in the treatment of these patients. Assessment of the severity of stenosis and of the effect of intervention will depend upon several factors such as the angiographic appearances and the pressure gradient, but must include an assessment of the blood flow through the stenosed segment. Radionuclide scanning provides a reproducible assessment of regional pulmonary perfusion and is an important adjunct to these procedures.13 Indeed, it is difficult to see how these patients can be satisfactorily followed up without lung perfusion imaging.

Conclusion

There is a limited role for nuclear cardiology in the study of congenital heart disease. This is changing as clinical practice changes. It will continue to change as functional imaging by other techniques, principally magnetic resonance imaging, becomes more widely available. That having been said, radionuclide imaging has unrealised potential for the research into and the clinical follow up of cardiac function in long term survivors of surgery for congenital heart disease.

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