the British Pacing and Electrophysiology Group. Within a few years advances in mapping techniques and catheter technology are certain to reduce procedure times and increase success rates. Last but not least, the radiofrequency method is cheaper than surgery.¹⁹

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How well do we manage families with genetic problems?

A national confidential inquiry into counselling for genetic disorders should tell us

Collectively genetic diseases are not rare and nor do they any longer justify automatic pessimism provided doctors are reasonably well informed. Professional ignorance may, however, be the greatest obstacle to allowing couples to use the advances in genetic medicine to make informed choices about their futures and those of their children. To find out how far clinicians are offering the full range of options for those at risk of genetic disease the Department of Health has funded a new confidential inquiry into counselling for genetic disorders, recognising that the prevention of such disorders is acceptable only if it results from informed choices. These include the option of rejecting genetic screening.

Two to three per cent of couples are at high risk of producing offspring with genetic disorders,¹ and 5% of the population will have a genetic disorder by the age of 25. This figure rises to 65% in a lifetime if common diseases with a strong genetic predisposition are included.² Much can now be done to help families to avoid genetic and congenital disorders, although in the absence of population screening for carriers most genetic disease occurs without warning. Nevertheless, ignorance leads to couples having avoidably handicapped children or being so afraid that they remain childless or unnecessarily abort normal fetuses. Because the family history is sometimes ignored adults later develop preventable fatal disease. Since rapid genetic advances affect disease in all specialties most doctors now need some skill in genetics. It is that expertise the new inquiry aims at fostering.

While a maternal or perioperative death is always a matter for regret, the birth of a baby with Down's syndrome must not be regarded as a medical failure when a couple who have been sympathetically and accurately counselled refuses prenatal diagnosis or declines termination of pregnancy. In this respect the new inquiry into counselling for genetic disorders differs from other confidential inquiries, and the inquiry (to be conducted by the department of medical genetics at the University of Manchester and the research unit of the Royal College of Physicians) is starting only after three years of

cautious planning. Advice has been received from the medical royal colleges, other professional groups, lay organisations, and the Department of Health, and the steering committee's* proposals were unanimously accepted at an open forum attended by a wide variety of interested groups.

The inquiry will concentrate on marker disorders† and will cover the potentially avoidable births of affected children or the onset of serious, preventable adult genetic disease, including hereditary cancer. The aim will be to assess what has been written in records about the information and services offered to patients and their relatives, seeking documented evidence of (a) appropriate counselling, (b) the offer of relevant genetic services, and (c) follow up, especially for potentially preventable disorders of late onset. Clinicians will review their own records and will be encouraged to involve their junior staff in a process that should provide a framework of good clinical practice for genetic disorders.

Although clinical records may not always show the real quality of clinical practice, improvement in records probably reflects improvements in practice, and records are expected to improve as a result of the inquiry. Not all clinical staff will be involved, but effective reporting should alert and inform many who have not participated directly and will also highlight deficiencies of resources. The inquiry will be publicised through professional organisations and will emphasise its independence of the government, of management, and of other institutions—and the absolute anonymity and confidentiality of its data.

Means for ascertainment will be different for each disorder but will rely predominantly on existing registers and professional links forged by the conveners. Since the number of events for each disorder will be small and scattered through-

* Steering committee: Professor R Harris (chairman), Dr Anthony Hopkins (Royal College of Physicians research unit), Dr Ian Lister-Cheese (Department of Health), Dame June Lloyd.

†The marker disorders and their conveners are: cystic fibrosis (Professor John Dodge), Down's syndrome (Professor Eva Alberman, Professor Martin Bobrow), neural tube defects (Professor Nicholas Wald), haemophilia (Dr Mark Leyton), thalassaemia (Dr Bernadette Modell), multiple endocrine neoplasia II (Dr Bruce Ponder), and familial adenomatous polyposis (Mr John Northover).

out Britain, completed questionnaires will be sent directly to a coordinator at the Royal College of Physicians, who will remove all identification before transmitting the data to the inquiry team for analysis. After the anonymous data have been extracted the records will be destroyed. As with the inquiries into maternal and perioperative deaths, the Department of Health has confirmed that it supports the total confidentiality of documents prepared for the inquiry. In practice this means that in any court case the court would weigh the public interest in preserving confidentiality in any judgment about disclosure. The inquiry will depend for its success on the cooperation of clinicians, who will take part

only if they are convinced that it is both confidential and non-censorious. The steering committee has taken great pains to ensure that is so.

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- 1 Royal College of Physicians of London. *Purchasers' guidelines to genetic services in the NHS. An aid to assessing the genetic services required by the resident population of an average health district.* London: Royal College of Physicians, 1991.
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Adults with congenital heart disease

A comprehensive specialist service is needed

Over the past 50 years advances in medical treatment and surgical procedures have radically altered the outlook for children with congenital heart disease. In the 1950s only 10-15% of infants with known congenital heart disease survived until puberty,¹ whereas now many survive childhood. This success story has resulted in a growing population of adolescents and young adults with congenital heart disease, an increasing proportion of whom have complex cardiovascular abnormalities. In the United States well over 500 000 such patients have reached adulthood in the past 30 years.² Currently patients who outgrow the paediatric clinic are either lost to follow up or managed as "one off" cases by adult physicians who are more experienced in acquired than congenital heart disease. Specialised centres for this new category of patients are therefore essential to optimise care and to collect follow up data to provide rational advice about lifestyle and future prognosis.

A major requirement for these patients when they grow to adolescence and young adulthood is a facility for excellent cardiac surgery. Many of the patients will have had surgery in childhood, but total correction is rare.³ Most will therefore need long term medical surveillance, and many need reoperation for replacement of bioprosthetic valves or extracardiac conduits inserted in childhood. Further indications for reoperation include residual defects after definitive repair, and staged surgery for complex defects, such as pulmonary atresia with ventricular septal defect. Reoperation is difficult and carries a high risk,⁴ particularly if undertaken by surgeons inexperienced in such problems. Other adults may require their first operation for lesions that were well tolerated during childhood, such as bicuspid aortic valve or Ebstein's anomaly. Finally, despite the best efforts at repair, an increasing number of these patients will eventually become candidates for heart or heart-lung transplantation. They need considerable medical, social, and psychological support.

Arrhythmia is the commonest medical problem encountered in this population. Loss of sinus rhythm and supra-ventricular tachyarrhythmia are seen with increasing frequency during follow up after intra-atrial repair for transposition of the great arteries (Mustard or Senning procedure) and after the Fontan operation.⁵ Awareness of these late complications has contributed to a change in the preferred initial surgery for transposition and a revision in technique for the Fontan procedure.⁶ Ventricular tachyarrhythmia is often observed during ambulatory electrocardiographic monitoring in asymptomatic survivors after

repair of tetralogy of Fallot.⁷ The risk associated with arrhythmia and the need for treatment depend on the underlying haemodynamic condition, so the physician deciding on antiarrhythmic treatment and the need for pacing needs both electrophysiological skill and an understanding of the congenital heart defect itself. The haemodynamic state may be especially vulnerable to either the arrhythmia or the myocardial depressant effects of antiarrhythmic treatment when a right ventricle supplies the systemic circulation after the repair (for example, after the Mustard procedure), or when only one ventricle is functioning (for example, in the Fontan circulation).

Patients with cyanotic congenital heart disease, particularly those with pulmonary vascular disease (Eisenmenger's syndrome), require special care. Erythrocytosis invariably occurs and in some may cause hyperviscosity symptoms such as headache or fatigue. Injudicious phlebotomy may aggravate symptoms by producing iron deficiency.⁸ Stroke is rare in these young adults, but bleeding problems occur (due to defective haemostasis) and renal insufficiency and arthropathy (due to hyperuricaemia) are common complicating features.⁹ Patients with Eisenmenger's syndrome are at particular risk during anaesthesia, exercise, and pregnancy. This is clearly one condition in which counselling against pregnancy is appropriate. In other adults with congenital heart disease considering pregnancy counselling is necessary to explain the potential risks to both mother and fetus.^{10 11}

The most recent confidential enquiry into maternal deaths reported an increase in deaths due to cardiac disease, attributed to deaths in women who had previously had surgery for congenital heart lesions.¹² This emphasises the need for expert attention from obstetricians, cardiologists, and anaesthetists for this group of patients.

Endocarditis is a risk for all patients with congenital heart disease, particularly adolescents, who often do not comply with antibiotic prophylaxis. This is perhaps the main reason for following up patients with simple lesions of minor haemodynamic importance, such as those with a small ventricular septal defect. Any such patient who is unwell with fever may have cardiac infection and should have blood cultures done before empirical antibiotic treatment is prescribed.

Psychosocial problems are also important. Many of the "normal" ordeals of adolescence may be more difficult for this group, in whom chronic illness, embarrassing scars, and excessive limitation of activities may inhibit normal social