Long term outlook in treated congenital heart disease

Without treatment, 85–95% of the 5–10 per 1000 live births affected by congenital heart disease (CHD) will die before adolescence. However, the situation has been transformed by spectacular advances in medical and surgical care during childhood, so that now the majority can expect to survive into adulthood. Echocardiography has become a key diagnostic tool, refinements in surgical procedures and myocardial protection allow longer and more complex operations to be carried out, and transcatheter interventions obviate the need for surgery altogether in some conditions. Further advances can be expected, so that an increasing number of survivors with complex CHD will require continuing expert care into adolescence and adulthood.

This new and growing population of adolescents and adults represent a challenge to the health service. Most adult cardiologists, who in the past might have been comfortable dealing with the small numbers of survivors with relatively simple lesions, do not have the training and experience to manage the very abnormal and complex circulations with which many patients are left, even after their definitive surgical repair. However, paediatricians may also be ill equipped to deal with the acquired medical conditions of adulthood or the psychosocial issues for which these patients require so much assistance. Even patients with ‘simple lesions’ such as aortic coarctation may run into problems without vigilant surveillance: while the risks of recoarctation and aneurysm formation may be appreciated by paediatric cardiologists, the management of their systemic hypertension and acquired premature coronary artery disease may be better managed in the setting of adult cardiology.

It is crucial not only to maintain continuity of high level medical and surgical care, but also to provide feedback about late results in order to improve initial management in infancy and childhood. For example, as a result of such long term follow up information, the favoured surgical approach for transposition is now the arterial switch operation because of the significant late problems that have been encountered in adolescence and adulthood by patients who had undergone interstitial repair.

Surgical needs

Some patients may not need their first operation until adolescence or adulthood, either because a complex lesion, such as Ebstein’s anomaly, was well balanced in early life, or because a simple lesion was missed, or only became haemodynamically significant in later life. This may be the case for atrial septal defect, the dilating aortic root in Marfan’s syndrome, and bicuspid aortic valve disease.

Reoperations, however, are the major surgical need for patients in this age group, some are inevitable, either as part of a staged approach such as in complex pulmonary atresia, or as valvar prostheses and conduits degenerate. Reoperations may also be unexpected and needed as a result of endocarditis, prosthetic valve failure, or thrombosis within an area of low flow, such as across a Fontan conduit. The risks of such surgery are further increased if the patient is haemodynamically compromised or has uncontrolled sepsis. Eventually, if myocardial failure or pulmonary vascular disease develop, transplantation may be the only option.

Operations in these patients are particularly challenging and should not be undertaken by inexperienced surgeons, as previous surgery may make re-entry to the chest difficult, cyanosis increasing bleeding problems and myocardial depression and pulmonary vascular disease adding to the risks of anaesthesia and cardiopulmonary bypass.

Finally, non-cardiac surgery may also represent a danger to the cyanosed patient in whom the severity of complex CHD is not fully appreciated. For example, induction for a general anaesthetic in a patient with Eisenmenger’s syndrome may produce catastrophic vasodilation from which it may not be possible to resuscitate the patient. All too often, the medical records that may guide the surgeon, physician, or anaesthetist are missing because of inappropriate destruction of old notes, adding an avoidable and inexcusable difficulty to these patients’ safe management.

Medical needs

Although some of the more simple conditions may have been definitively corrected in early life, infrequent long term follow up remains important as they may remain at risk of endocarditis or develop complications such as arrhythmia or pulmonary vascular disease in adulthood.

ARRHYTHMIAS AND CONDUCTION DEFECTS

These are the most frequent problems encountered and need to be considered in the context of the patients’ underlying circulation. Some, such as such as atrioventricular dissociation and accessory pathways in corrected transposition, may develop as a consequence of the cardiac lesion itself, others may arise as a complication of surgery. Atrial arrhythmias are particularly common after atrial surgery, especially in the presence of atrial distention after the Fontan operation.

Understanding the clinical significance and optimal approach to the treatment of an arrhythmia depends on the cardiologist understanding the underlying cardiac defect. If myocardial function is depressed, the onset of arrhythmia or the inappropriate use of negatively ionotropic...
agents may result in rapid haemodynamic deterioration or sudden death. It is only with the centralisation of care that risk factor stratification for sudden death can be achieved for infrequently encountered complex CHD.4

CYANOSIS
Despite careful management, some patients will remain cyanosed or become cyanosed with the development of pulmonary vascular disease. Assiduous attention to the packed cell volume should help to prevent disastrous thromboembolic events,4 and a heightened awareness of renal impairment (angiographic contrast media may precipitate renal failure)10 and specific complications such as cerebral abscess can improve both quality and length of life.

INFECTIVE ENDOCARDITIS
Endocarditis is often badly managed in young adults with CHD. This may be because of a lack of appreciation of which patients are at risk, or because of a failure to search thoroughly for the causative organism before administering antibiotics, or because of the physicians' naivety about the social behaviour of young adult and adolescents.

Recommendations for antibiotic prophylaxis differ, but it is currently widely prescribed for all CHD, whether surgically repaired or not, except for conditions such as divided patent ductus arteriosus and secundum atrial septal defect closed by direct suture.11 While it is usually recognised that dental, urogenital, and obstetric procedures should be covered by antibiotic prophylaxis, it is often not appreciated that patients may also be at risk from indulging in intravenous drug abuse, tattooing and body piercing, all of which can cause bacteraemia. Patients are unlikely to volunteer this kind of information unless their physician is both understanding and perceptive.

MYOCARDIAL DYSFUNCTION
Ventricular failure is the commonest cause of decline and death in patients with CHD. Even after repair, ventricular deterioration may eventually occur if the correction has been "physiological" rather than "anatomical", as is the case after interatrial repair for transposition or the Fontan operation.12 Poor intraoperative myocardial preservation in the early days of CHD surgery may also contribute to myocardial damage and late clinical problems.

Pregnancy/contraception
Adults with CHD have all the same expectations as the general public, and want advice about contraception, pregnancy, and risk of recurrence.

Oestrogen containing oral contraceptives increase the risk of thrombosis in those with polycythaemia due to cyanosis and those with low flow circulations. In these women, either the progesterone only pill, or a barrier method should be used. Intrauterine devices are associated with endocarditis in the presence of pelvic infection, and menorrhagia may be problematic for those with cyanosis or who are anticoagulated; in general therefore they are not recommended for women with CHD.

In the 1990s the only absolute contraindication to pregnancy because of maternal risk is the Eisenmenger's syndrome.13 However, the haemodynamic load that pregnancy imposes may also compromise those with a Fontan-type circulation, and the woman with Marfan's disease and a dilating aortic root may also be at risk. Pregnancy tends to be tolerated badly in women with stenotic lesions and reasonably well in those with regurgitant lesions. Joint cardiac and obstetric antenatal care in an 'at risk' clinic will further the reduce risk to both mother and fetus.

Although many women with CHD can attempt pregnancy without undue risk to themselves, fetal hazards mean that many pregnancies will not be successful. Cyanosis and polycythaemia have particularly adverse effects with a 50% risk of fetal death if systemic arterial oxygen saturation is below 85%, or the packed cell volume above 0.65.14 The fetus may also be at risk from maternal anticoagulant treatment,15 and there is a 20% chance of fetal death if the mother has to undergo cardiac bypass surgery during pregnancy.

Although there are well documented data on recurrence risk when a sibling is affected (1–8%),16 information is limited in the offspring of a mother or father with CHD. It is only now that sufficient numbers of patients are surviving to start a family that information is accumulating. The specific genetic risk to the fetus depends on the maternal or paternal cardiac lesion. For many lesions the risk is now considered to be higher for the offspring when a parent rather than a sibling has CHD, and when the mother rather than the father is the affected parent.17

Psychosocial issues
These issues need to be appreciated sympathetically by the cardiologist. Questions may be difficult to answer, ranging from embarrassment about physical scars to fear of what surgery and what life may lie ahead, particularly when the prognosis after new operations is not known. Furthermore, long spells in hospital result in strain on the patient and their family, and on the patient's education.

Although the ability to find work relates to clinical well-being,18 patients may be unfairly discriminated against when they apply for jobs, mortgages and life insurance, because of lack of data about the outcome of modern treatments.19 Cardiologists are thus often called upon to act as advocates for their patients as they reach adulthood and try to live independently, and so it must therefore be an absolute priority for them to gather and publish modern outcome data.

Patients often want advice on taking part in sports, and restraining advice does need to be given to some individuals who are relatively asymptomatic.20 The risk of sudden death should rule out competitive sports and weightlifting in individuals with obstructive lesions such as aortic stenosis and hypertrophied cardiomyopathy, or in those with aortic root dilatation, and contact sports should be avoided in those who are anticoagulated or who have conduits underlying the sternum. Most patients with complex CHD are however, symptomatically limited by their cardiac lesion.

Who should care, and where?
The debate as to who should care for long term survivors of CHD, and where, continues to be controversial. We believe that patients receive the best care when resources are centralised in a multidisciplinary environment, and although the solution may differ from region to region, the adult cardiologist with training in CHD should, in conjunction with paediatric colleagues, be best placed to allow a smooth transition from paediatric to expert adult care.

The high cost of such specialised practice is under threat with the reorganisation of the NHS, and funding issues are fractionating rather than rationalising resources; disasters happen when doctors are discouraged from seeking expert care.
advice and making extracontractual referrals. The specific needs of groups such as those with CHD must be addressed within the context of the NHS reforms if the same high quality of care that helped them survive childhood against the odds, and which they surely deserve to receive in adult life, is to continue.

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