Current status of definitive surgery for congenital heart disease

The past 30 years have witnessed major changes in surgical treatment of congenital heart disease (CHD). The early years of innovation have been superseded by a new era of scientific assessment enabling the surgeon and the cardiologist both to evaluate and develop therapeutic strategies for the different groups of patients with CHD.

In considering the current status of surgery for CHD one should be clear that modern surgical strategies may involve staged procedures in order to achieve a definitive long term outcome. Current treatment protocols therefore demand total integration of the services provided by surgeons, paediatricians, and cardiologists and this 'team approach' remains the cornerstone of any successful surgical programme.

The past decade has witnessed an increasing tendency to advocate 'definitive' operation where possible in neonatal or infant life. Thus, for instance, infants with ventricular septal defect and intractable cardiac failure can now undergo definitive surgical repair with a mortality approaching zero. In this brief review I will address the current status of 'definitive' surgery in this younger age group. In all instances, it must be remembered that growth of the patient will often necessitate some further surgical intervention, most commonly when artificial conduits are employed as part of the original reconstruction. The term definitive must therefore be used with caution and close cardiological supervision is mandatory for all patients who have undergone surgery for CHD in childhood.

Transposition of the great arteries (TGA)

Although it has been possible for many years to achieve good early results in surgery for TGA using an atrial correction in which systemic venous blood is directed by means of an intra-atrial baffle to the left ventricle and pulmonary artery (Senning or Mustard operation), the intermediate and long term results have revealed an unacceptable late morbidity.1 2 The arterial switch operation, in which the aorta and pulmonary arteries are divided and retransposed to the anatomically normal position together with appropriate relocation of the coronary arteries, has thus become preferred treatment of choice for patients with TGA unless there is significant pulmonary or subpulmonary stenosis. Even when TGA is associated with other complex malformations, an arterial switch procedure should normally be undertaken at the time of repair of the associated cardiac or extra cardiac anomalies.

Planche et al recorded a hospital mortality of 8·3% in neonates,3 similar to that reported by Brawn et al for this group of patients.4 With increasing experience, particularly in the management of variations in coronary artery anatomy, the current hospital mortality for patients with TGA and intact ventricular septum at our institution is 1·8%, while that of complex TGA, primarily determined by the nature of the associated lesions, is 5·5%. Most recently, increasing confidence in the arterial switch procedure has led to its application in more complex forms of transposition, for instance those associated with atresia of a single atrioventricular valve, in an attempt to avert the morbidity often previously seen after palliative procedures such as pulmonary artery banding.

Although follow up of patients in most series is relatively short, it appears that the main postoperative problem in patients undergoing an arterial switch repair is the development of pulmonary artery stenosis. This occurs, in our experience, in 10% of patients and may be treated successfully in most cases by catheter balloon dilatation of the obstruction.

In older patients in whom right ventricular dysfunction has developed many years after a previous Senning or Mustard operation, the arterial switch procedure may still be applicable. Before such an operation, however, the left ventricle must be conditioned to develop systemic levels of blood pressure and this may be achieved by pulmonary artery banding. At present the mortality and morbidity of this staged approach remains high.3

Atrioventricular septal defect

While partial atrioventricular septal defect may be treated in a similar way to atrial septal defect (repair before school age), the treatment of complete atrioventricular septal defect poses a major surgical challenge. The risk of obstructive pulmonary vascular disease secondary to pulmonary hypertension is such that primary repair within the first year of life is advocated by most major centres. Current experience suggests that a hospital mortality of 3–10% should be expected.6 7 The reoperation rate, usually for residual ventricular septal defect or left atrioventricular valve incompetence is between 8 and 16%. These results are encouraging when compared with the 20% mortality published in series prior to 1983.

Tetralogy of Fallot

Controversy still remains over the optimal timing for
Aortic valve stenosis

This condition remains one of the more difficult management problems in the young patient. While most patients presenting with critical aortic stenosis will progress well after surgical valvotomy, the longer term outlook for some of these patients and for older patients with aortic valve disease remains uncertain. The morbidity attendant upon anticoagulation treatment in patients who undergo aortic valve replacement, with a mechanical prosthesis at a young age, detracts significantly from the merits of such operations. Experience with aortic homografts as valve substitutes in young children has been encouraging but these patients also face potentially hazardous reoperation as they grow older. More recently, the use of the pulmonary valve as an autograft aortic valve replacement has been recommended by Gerosa et al in the hope that this valve substitute will grow with the child and thereby minimize the longer term morbidity. As yet, however, data on the growth of this valve substitute is available in only a very small number of patients and so the management of patients with aortic valve dysfunction remains problematical.

Conclusion

It will be clear from these observations on a few selected conditions that definitive surgery for congenital heart disease can be considered for most situations with the aim of achieving normal atrial, ventricular, and great arterial connections and relationships. The team approach to the problems posed by patients with congenital heart disease encourages us to look forward to the next decade with optimism as we seek to develop and evaluate the role of definitive surgery in the palliation of entities such as hypoplastic left heart syndrome and other conditions where restitution of normal cardiac relationships remains an impossibility.