SURGICAL PROSPECTS IN COMPLEX CYANOTIC CONGENITAL HEART DISEASE

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"Complex" implies "made up of parts, intricate, complicated involved, entangled". In cyanotic congenital heart disease, the complex varieties may be characterised either simply by the number of individual components or by the nature and severity of some individual components. Additional abnormalities may compensate physiologically for others, but may greatly increase the risks of surgery, as when a large ventricular septal defect is associated with complete transposition of the great arteries. The severity of one component of abnormality may change significantly the appropriate surgical approach, as in pulmonary atresia with ventricular septal defect in contrast to the pulmonary stenosis of the Tetralogy of Fallot.

The commonest category of complex cyanotic congenital heart disease is complete arterial transposition with additional serious lesions, such as ventricular septal defect, pulmonary stenosis and aortic coarctation. In our experience, this constitutes about 20% of the group. Other major components of the group include persistent truncus arteriosus, the various forms of pulmonary atresia, single ventricle and tricuspid atresia and hypoplastic left heart syndrome.

Surgical prospects in the group may be looked at in several ways. The first aspect concerns the benefits already available or likely to become available to appropriate individuals as a result of anatomically or physiologically reparative procedures that have come from recent developments in surgery and supportive measures. The second aspect concerns the impact that surgery may be expected to have on the total group, and the limiting problems that will be associated with its performance.

There is no doubt of the effect, immediately at any rate, of successful reparative surgery in suitable conditions such as complex transposition with ventricular septal defect and severe pulmonary stenosis, where either a Mustard operation with direct attack on the additional lesions, or, if that is not feasible, a Rastelli re-constructive procedure, may be possible. The individual who has such surgery successfully performed benefits enormously, as for example, a 16 year old patient of our own unit who was converted several years ago by Sir Brian Barratt Boyes from a severely disabled invalid pensioner to a person with full capacity for life and work. The long term prospects for such individuals, however, will only really become clear as the nature and incidence of late post-operative complications are ascertained by detailed and prolonged follow-up. In transposition, problems from possible complications such as contraction of the inter-atrial baffle, arrhythmia, tricuspid incompetence, progressive pulmonary vascular obstruction, and, in the Rastelli procedure, homograft changes, will all need evaluation before the excitement experienced with patient and family over the immediate transformation can be translated into a realistic evaluation of the consequences of such operation. In other words, a careful study of long term progress after such surgery is needed to place accurate facts about future prospects beside the clear cut short term reward.

Consideration of total surgical prospects for the group, including both palliation and repair, requires an initial assessment of its numerical importance. Cyanotic congenital heart disease of all types probably accounts for between 20 and 25% of all congenital heart disease, as indicated by various figures of incidence, subject as they are to varying problems of selection of intake. The Tetralogy of Fallot accounts for approximately 10% of congenital heart disease and the forms of complete arterial transposition without other significant defect probably about 4% of all congenital heart disease. There is therefore a substantial residue of cyanotic cases which are in fact largely complex in nature.

Another source of information about the magnitude of the problem, and therefore its potential yield, may be obtained from actual current working loads of active units rather than from accumulated incidence figures. In the years 1969 through 1971, a total of 1019 cardiac catheterisations were performed in the Department of Cardiology at the Royal Children's Hospital, Melbourne (Table I). Of these 488 (48%) were in infants under one year of age and 220 (21.5%) in infants less than one month of age. During that period there were 101 infants who could be classified as having complex cyanotic lesions (Table II). These included 17 cases of hypoplastic left heart syndrome which have so far been accepted as inevitably lethal and inoperable problems. The remainder have had conditions that in the main could at least theoretically be regarded as suitable for salvage. There were therefore approximately 30 new infants per year with complex cyanotic congenital heart disease in the paediatric age group, and certainly in infancy, from a population group exceeding 3 million with a birth rate of over 70,000 per year.

TABLE I

CARDIAC CATHETERISATIONS PERFORMED 1969-71

Age under one month	220
Age one month to 11 months	268
Age one year and over	531
TOTAL	1019

TABLE II

NEW CASES OF COMPLEX CYANOTIC CONGENITAL HEART DISEASE

	Neonates (aged less than one month)	Tot al infants (aged less than one year)
Complex transposition	15	22
Truncus	5	14
Pulmonary atresia/VSD	13	14
Single ventricle etc.	7	11
Pulmonary valve atresia (no VSD)	6	8
Tricuspid atresia etc.	4	8
Others	2	7
Hypoplastic left heart syndrome	15	17
TOTAL	67	101

The numbers of older children studied with complex cyanotic congenital heart disease in the same period have much less significance. There were 38 of whom 33 had already been under follow-up in the unit, nearly all having previously been investigated. Only 5 had been referred for the first time, either from interstate or from overseas. Various factors influence the timing of follow-up studies and these patients do not truly represent the yield from any earlier definable intake.

The nature of the conditions is shown in Table II. Approximately 2/3rds of the infants presented and

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were studied in the neonatal period. One striking factor is the delay in presentation of cases of persistent truncus arteriosus.

These infants presented usually with hypoxia or with heart failure and were frequently severely ill. The natural prognosis of the group was clearly poor. Even in spite of active treatment where feasible the mortality for those with conditions other than hypoplastic left heart syndrome was approximately 50%.

Prompt referral, investigation and management are essential to obtaining maximum salvage, and the amount of wastage that would occur just from dropping out the neonatal component is clear. Only a limited proportion have a capacity for continuing survival in a state suitable for ultimate repair without palliation, and even with active intervention the prospects are very poor in some conditions. For instance, our experience of three individuals with complete transposition complicated by ventricular septal defect and aortic coarctation or interruption has yielded one unsatisfactory survivor, while only three of eight patients who have had pulmonary artery banding for persistent truncus are alive. None of these four patients have as yet had any attempts at final definitive correction. Consideration must be given therefore to whether intervention in such conditions will really give a greater yield than the natural course.

In some conditions, the question of early primary repair under deep hypothermia now arises as an alternative to palliation and later secondary repair. When this approach is technically feasible it offers the prospect of mortality rates from one procedure that may appear better than the estimated summated rate of several procedures, together with the removal of problems from such inter-current complications as cerebrovascular accidents. Long term follow up will nevertheless be required to define the situation clearly and to assist in determining optimal surgical management.

In other conditions, in which there are as yet no satisfactory reparative procedures, surgery can only offer at present a purely palliative solution. Such palliation is acceptable when it offers reasonably long term relief as with satisfactory shunt procedures. It is less acceptable when it induces further problems itself. Pulmonary artery banding, in itself somewhat unreliable and unpredictable, imposes an inevitable need for later secondary surgery to relieve the effects of increasing pulmonary artery obstruction, and can hardly be regarded as providing acceptable permanent palliation. Other palliative procedures may change the haemodynamic situation unfavourably even more rapidly. For instance, creation of an atrial septal defect may be indicated in the uncommon syndrome of mitral atresia and single ventricle, in order to reduce left atrial pressure and thereby increase pulmonary flow. This will decrease dyspnoea and cyanosis. The procedure may decompress the left atrium so effectively that it leads to excess pulmonary flow and heart failure, so that care must be given to deciding whether it is in fact indicated. Although some individuals may obtain clear benefit, the overall prospects of surgery in such conditions may be uncertain and difficult to predict.

The group of complex cyanotic congenital wastage rate in spite of surgical progress and increased experience. This is by virtue of the predominant early presentation with severe trouble, and the fundamental nature of many of the problems. Improvement in salvage will require increasing effort in proportion to the gains obtained. It is important to point out that surgical management of this group of patients requires extensive and expensive supporting facilities in various areas. It provides much emotional strain, particularly for the families of the patients. It imposes large demands on general paediatric and cardiological care, and on special investigatory facilities, together with other hospital departments. For example, in the year 1971-72 at the Royal Children's Hospital in Melbourne, the total biochemical tests performed in the hospital reached the figure of 120,000. Of these, 30,000 that is 25%, were performed on cardiac patients. In considering the surgical prospects of this group as a whole we must all take into account the limits that such requirements may place on our endeavours.