THE SCOPE OF CARDIAC SURGERY—INCLUDING NEONATAL SURGERY

BY

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The rapid advancement of cardiac surgery over the last 20 years has transformed the practice of cardiology and has markedly improved the outlook for the patient with heart disease. Accurate prognosis as well as complete haemodynamic diagnosis have become the main tasks of the modern cardiologist because open heart surgery has now reached a stage when nearly all patients can be radically improved by surgery.

Severe pulmonary vascular obstruction in congenital heart disease and poor myocardial function in acquired heart disease remain the major problems. In general the following are the minimum basic ingredients for the successful outcome of an operation.

In congenital heart disease:
1. absence of too severe pulmonary vascular obstruction;
2. the presence of two ventricles.

In acquired heart disease:
1. reasonable left ventricular contractile function:
2. absence of failure of other organ systems—particularly the lungs and liver.

These considerations apply to all ages from birth even up to the 7th and 8th decades. The ability to correct radically, or to palliate, nearly all forms of heart disease (except for primary myocardial disease) has placed a heavy responsibility upon the physician. He must accurately assess the current status and medical prognosis of his patients as well as the operative risk and morbidity, the amount of improvement to be anticipated and the alteration in prognosis (including new hazards) after surgery. Since the advent of open heart surgery provided the impetus for accurate anatomical and haemodynamic diagnosis, new diagnostic and surgical techniques have developed pari passu with each other. These have revealed a yawning gap in our knowledge of the natural history of many forms of heart disease, particularly since such information as we have applies to large numbers of patients treated as a single entity rather than to individuals with their own particular variation of a disorder.

CONGENITAL HEART DISEASE

The surgery of congenital heart disease began in 1939 when Gross and Hubbard ligated the first patent ductus. Five years later Blalock created the first artificial ductus for the palliation of Fallot's tetralogy after Helen Taussig had observed the benefit of a natural ductus in children with this disorder (Blalock and Taussig, 1945). In 1948 Brock and Holmes Sellors first carried out closed pulmonary valvotomy and infundibulectomy in severe closed-septum pulmonary stenosis, as well as in Fallot's tetralogy. Atrial septal defects were first closed in the early 1950s and the later development of open heart surgery using hypothermic techniques allowed them to be dealt with at low risk. The late 1950s and 1960s saw the development and application of safe open heart surgery using heart-lung bypass, whilst the main development in the last few years has been in the successful surgical management of the neonate with congenital heart disease (Aberdeen, 1968) and in the radical treatment of transposition of the great arteries.

Almost 1 per cent of newborn babies have congenital heart disease: nearly half of these fail to survive childhood. Since most of the deaths occur in the neonatal period, the newborn infant has been regarded as poor surgical material. This is untrue. The deaths occur because the defects are severe. The abnormalities are frequently multiple and highly complex and are simply incompatible with growth and activity unless something is done to improve the perfusion of vital organs. Experience with other congenital intrathoracic abnormalities, such as oesophageal atresia, has proved that the neonate is a good

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surgical subject provided he is not allowed to deteriorate too far before surgery is attempted and provided a highly skilled team is available. Unfortunately, congenital heart disease is all too often first recognized relatively late or the infant permitted to go downhill before diagnosis and treatment is attempted. Subdiaphragmatic total anomalous pulmonary venous drainage (TAPVD) and complete simple transposition of the great arteries (TGA) are two very good examples of this (Gathman and Nadas, 1970; Plauth, Nadas and Bernard, 1968). In both conditions cyanosis may be wrongly attributed to a respiratory cause because murmurs are absent and heart failure and cardiomegaly are not early features.

In subdiaphragmatic TAPVD the pulmonary veins pass down through the diaphragm to join the inferior vena cava or portal venous system. Pulmonary venous obstruction is inevitable both because of the actual length of the common pulmonary vein as it courses down below the diaphragm to join a branch of the portal system and because its blood has to traverse the capillary bed of the liver before it gets back to the right atrium. The ground-glass appearance of the congested lungs is diagnostic but often mistaken for the respiratory distress syndrome. The common pulmonary vein actually crosses the back of the left atrium as it passes to its anomalous destination so that correction is not a very difficult procedure technically. The exceedingly high mortality of this condition is due almost entirely to the fact that it is incompatible with life beyond the neonatal period and is operated upon too late.

In TGA it is the infants without intracardiac anomalies who succumb most quickly, yet these are the very children with the best prognosis for radical correction. In TGA the pulmonary and systemic circulations are separate systems. Since the aorta arises from the right ventricle access of any oxygenated blood to this vessel depends on shunting at either atrial, ventricular or aorta-pulmonary level. Similarly the return of deoxygenated blood to the lungs depends on a shunt in the reverse direction. Maintenance of a constant volume of blood in either circulation depends on these opposing shunts being equal in magnitude. Since the pulmonary artery pressure falls, as in the normal newborn after birth, a small ductus or small ventricular septal defect (VSD) will not help matters as blood can only go one way—into the lungs. A big VSD induces pulmonary hypertension and a tendency to pulmonary oedema from overfilling of the left atrium but will allow bidirectional shunting. A big atrial septal defect is ideal because it allows bidirectional shunting without allowing free transmission of pressure from the high pressure systemic circuit and right ventricle to the low pressure pulmonary circuit and left ventricle. In the majority of cases an ASD can now be created without thoracotomy by dragging a balloon-tipped catheter across the foramen ovale using the technique developed by Rashkind (Rashkind and Miller, 1968). This technique occasionally fails to provide an adequate-sized ASD and cannot usually be carried out after the neonatal period because the septum becomes too tough to tear. In older children surgical creation of an ASD using a variation of the closed operation developed by Blalock and Hanlon (1950) still has a place, but radical operation for TGA using the Mustard procedure is now being applied increasingly to very young children (Mustard, 1964). The Mustard procedure is essentially a vein switch. It leaves the great arteries transposed and therefore the left ventricle as the low pressure venous ventricle supplying the lungs. Earlier attempts to retranspose the great arteries and connect them normally failed in such cases, largely because the thin left ventricle in simple TGA cannot cope with the systemic pressure. In the Mustard operation a pericardial baffle is placed within the atrial cavity (after removal of the natural septum) in such a way as to leave the pulmonary orifices in communication with the tricuspid valve and to channel the caval and coronary sinus blood towards the mitral valve. Although this technique has been successfully applied to neonates most surgeons prefer to wait until the child is over 5 or 6 kg at least, survival until this stage having been permitted by the Rashkind procedure. In TGA with a big VSD, heart failure dominates the scene and palliation can be achieved by banding the pulmonary artery. This lowers the torrential pulmonary blood flow, reduces the pulmonary artery pressure and retards the development of pulmonary vascular obstructive changes. Atrial
septostomy is also carried out as the first step in these babies, since it vents the overfilled left atrium, reduces its pressure and encourages mixing at this level. Later, a radical procedure can be carried out.

For babies with TGA, a big VSD and severe pulmonary stenosis, neither heart failure nor profound hypoxia is necessarily a problem because mixing occurs at ventricular level. The pulmonary stenosis is frequently uncorrectable because of participation of the mitral valve in a subvalvar stenosis but benefit can be expected from a Blalock systemic-pulmonary shunt if cyanosis is severe. They may then wait until an increase in their size permits a Rastelli operation (Rastelli, 1969). In this operation the great arteries are repositioned so that an anatomical correction is achieved. This is done by transecting and oversewing the stenotic outflow tract to the lungs as it leaves the left ventricle. A window is then cut in the anterior wall of the right ventricle and a pulmonary artery prosthesis and pulmonary homograft are sewn in. The other end of the homograft is sutured to the distal cut end of the transected pulmonary artery. The aorta is directed to the left ventricle via the ventricular septal defect by means of a patch which excludes its orifice from the right ventricle.

In truncus arteriosus the same technique can be used: the trunk vessel becomes the aorta and the ventricular septal defect is patched so as to prohibit access of blood to it from the right ventricle. A new right ventricular outflow tract is constructed and connected to the pulmonary arteries which are detached from the truncus.

These are the most dramatic developments in congenital heart disease but steady progress in the common defects has both made their correction safer and further defined the indications. If a child with a simple ostium secundum atrial septal defect (ASD) has the probability of good health for another 10 or 20 years and the possibility of the good health continuing for a further 10 or 20 years or more, then the risk of operation must be very small in order that it may be justified. Not only the risk to life but the risk of serious postoperative morbidity has to be taken into consideration. In major centres uncomplicated ASDs can be closed with a risk to life of under 1 per cent and a morbid risk of under 5 per cent (Cleland et al., 1968). A higher risk than this is intolerable.

In ventricular septal defect (VSD) the commonest cause of death in the first two decades after infancy was surgical repair—a chastening thought. It is now known that the one pressing indication for surgery in childhood is pulmonary hypertension and that when the pulmonary artery pressure is normal the prognosis is probably extremely good without operation, whatever the measured size of the shunt. When the pulmonary artery pressure is at systemic level it means that the VSD is of a size which offers no impediment to left ventricular emptying through it. Survival depends on a resistance to flow at another site, otherwise the left ventricle discharges most of its blood into the lungs with resultant pulmonary oedema and low output failure. This is what happens in the infants who die. The resistance can be a man-made band which narrows the pulmonary artery. Such a banding operation has saved many babies who later need the band removed and the VSD closed (Goldblatt et al., 1965). Although VSDs are being closed at a younger age there are few centres recommending primary VSD closure in the first year of life because the risk is higher than in later life and because some defects become smaller or close spontaneously (Hoffman and Rudolph, 1965).

ACQUIRED HEART DISEASE

Valve replacement.

In rheumatic valve disease the risk of valve replacement has steadily dropped. In the best centres mortality is now around 10 per cent for mitral valve replacement, 5–10 per cent for aortic valve replacement and 15–25 per cent for mitral and tricuspid or triple valve replacement (Braunwald, 1969). Mitral valvotomy continues to have a place in the repertoire of most British cardiologists and cardiac surgeons although the indications for closed mitral valvotomy have been restricted and there are now some surgeons who advocate open mitral valvotomy for almost all cases. Those who believe in closed mitral valvotomy emphasize the low risk, the low operative mortality (which is only one-tenth that of valve replacement) and the excellent results which are free from the potentially disastrous late risks which face the owners of prosthetic valves.
Excellent results from closed mitral valvotomy can be forecast with confidence only when the valve shows obstruction without any regurgitation, and when it is mobile and pliable without calcification. Less propitious valves should be replaced, the decision being aided by mitral echocardiography which is now a reliable non-invasive means of determining severity of obstruction, anterior cusp mobility and presence or absence of calcification.

The Starr-Edwards valve continues to be the most popular prosthesis for the mitral valve although inverted aortic homografts mounted on a rigid metal stent, and fascia lata similarly mounted, are gaining some ground. In 1968 the Starr-Edwards valve was modified by substituting a stellite ball for the silastic one and by covering the cage with knitted dacron. This had been shown in dog experiments to reduce the risk of late thromboembolism once the cloth became covered with neo-intima. Unfortunately the cloth covering reduced the effective orifice of the valve by a critical amount which rendered the early extended cloth-covered mitral valves (series 6300) unacceptably obstructive in some instances. This problem has now been corrected by further modifications and the newest mitral Starr valves (series 6310) seem to be haemodynamically acceptable as well as less liable to thrombus formation than the early models. So-called “biological” mitral prostheses have the advantage that the risk of thromboembolism is negligible and the risk of fatal or crippling late complications is much less than with the Starr valve. They also have the advantage of a full flow orifice without the varying amount of stenosis which is built into the artificial valves on account of their central obturators. Nevertheless, early or late valve failure is still a problem with biological valves and infection seems to be more common than with the Starr valve. The possibility of late valve failure resides with the suture line inside the stent which, together with the sewing ring, effectively prevents ingrowth of reinforcing fibrous tissue from the natural annulus.

For replacement of the aortic valve Starr valves and free homografts are the main alternatives, the choice depending on individual beliefs, skill and the availability of cadaver aortic homograft valves. As with the mitral valve, the free aortic homograft has the advantage of negligible late thromboembolic risk but the disadvantage of possible early or late valve failure.

Late failure of the Starr aortic valve from variance of the silastic ball is now prevented by substitution of a stellite ball for the previously used silastic one so that swelling and deformation of the ball from imbibition of lipid from the blood stream is no longer a possibility.

Early failure of free homograft function usually results from imperfect selection or placement of the homograft. Late failure may occur from degenerative changes in the grafted valve with distortion or even splitting of the cusps. Infection may destroy graft function at any stage. The best method of preparation of homograft valves is still hotly debated. Fresh valves, removed under sterile precautions and stored in antibiotic/fungicidal solutions, are almost certainly the best, but logistical difficulties prohibit their use in most centres. Irradiation and freeze-drying have largely been abandoned because of induced alterations in the collagen and elastic tissues of the valves. Deep freezing seems to be acceptable but simple storage in beta-propiolactone solution in a refrigerator appears to be the easiest method and also seems to give satisfactory function with preservation of tensile strength. So far, and with follow-up extending only for a maximum of 5 years in the leading centres, the free aortic homografts are on the whole still doing well. However, as with a leaking natural valve, mild early regurgitation may necessitate second replacements eventually. Calcification extending into the hinge mechanism of the valve may impair cusp movement but this is uncommon so far in our experience and has not yet been seen in the beta propiolactone preserved homografts. In the best hands the results of free aortic homograft valve replacement are now exceedingly good (Barratt-Boytes, 1967).

The indications for valve replacement vary widely in single valve disease. At the present time the mitral valve is usually replaced only when disability becomes intolerable, but the aortic valve is replaced when real cardiac symptoms develop. This is because the prognosis in aortic valve disease is usually under 2 years once any disability has developed, whilst the patient with lone mitral disease may still have
10–20 years of life ahead after the first symptoms appear.

In rheumatic valve disease tricuspid valve replacement should be carried out in addition to replacement of the mitral valve (+ aortic valve) wherever there is rheumatic deformity of this valve. This is usually recognized by the detection of a gradient across this valve when the right atrial and right ventricular pressures are measured simultaneously with a double-lumen catheter. Tricuspid regurgitation without stenosis poses a still unanswered problem. If it is left alone will it regress after successful mitral valve replacement with subsequent fall in right ventricular pressure? Although some earlier reports suggested that it did, recent experience has been less happy and opinion has swung towards regarding “functional” tricuspid regurgitation as synonymous with right ventricular failure. When the right ventricular systolic pressure is high, as in severe pulmonary hypertension, functional tricuspid incompetence may well regress postoperatively, whereas when the right ventricular systolic pressure is low but the venous pressure is high then right ventricular dysfunction may be more likely and may need to be aided by insertion of a prosthetic valve to correct the associated tricuspid reflux. Although triple valve replacement carries an operative mortality of up to 25 per cent the risk is less and the results better than when an incompetent tricuspid valve is left, whether or not this is secondary to an incompetent right ventricle.

Thromboembolism, infection and mechanical failure are not the only late risks. Myocardial failure is now being recognized with increasing frequency, probably because, until recently, patients with poor muscle function as well as poor valve function were not surviving operation. Now they survive the actual operation but may fail to benefit or improve only marginally.

Intensive work is being put into the construction of other “biological” valves for the replacement of the mitral and tricuspid valves. Valves which are constructed from fascia lata removed at the time of operation and sewn on to a stent (like the inverted aortic homografts) have the advantage of being living autografts, although whether they can acquire a new blood supply via the metal stent and sewing ring is doubtful. Early results with these valves are encouraging but the long-term results are still awaited (Ionescu et al., 1968).

DEVELOPMENTS IN THE SURGERY OF CORONARY ARTERY DISEASE

First, the correction of disastrous mechanical complications of cardiac infarction—ventricular aneurysm, ventricular septal defect and mitral regurgitation—must be considered.

Left ventricular aneurysm is usually anterolateral or apical and results from occlusion of the anterior descending branch of the left coronary artery. Less often the aneurysm is inferior or even posterior; these sites are less common because large infarcts resulting from occlusion of the right coronary or its posterior descending branch or the circumflex branch of the left coronary are apt also to involve the mitral valve apparatus—a lethal combination. The other reason is that inferior and posterior aneurysms are infrequently diagnosed because they are not obvious on the chest radiograph. Left ventricular aneurysms can be resected at remarkably low risk because the surgery is swift and the perfusion time short. The indications for resection are persistent left ventricular failure, intractable angina, systemic embolization and repetitive ventricular tachycardia, in that order of frequency. Failure and angina are often dramatically alleviated because an aneurysm increases left ventricular metabolic need by increasing its size and wall tension, it reduces the stroke output because blood is directed into the aneurysm, and it increases the end-diastolic pressure because a compliant aneurysm adds to ventricular filling when it recoils and returns some of its systolic volume to the body of the ventricle in diastole. Repeated tachycardias and angina may also stem from ischaemic muscle at the perimeter of the aneurysm which may be resected at operation. However, this is probably only of importance in a minority of cases.

Ischaemic rupture of the ventricular septum may develop 2–7 days after infarction and the event is usually followed by the rapid development of congestive heart failure and pulmonary oedema following upon the appearance of a loud pansystolic murmur and thrill. It is a singularly fatal complication but many patients
have been restored to apparent good health by surgical closure of the defect.

Ischaemic fibrosis of a mitral papillary muscle can result in severe mitral regurgitation, but much more commonly the mitral valve dysfunction is mild and associated with poor left ventricular pump function. A few patients develop mitral regurgitation after localized and even subendocardial infarction which paralyzes the posterior papillary muscle. The coronary supply of this muscle derives from a single source, the circumflex branch of the left coronary artery, in about 30 per cent of individuals. In such circumstances widespread coronary disease and consequent pump failure are not necessarily present and great symptomatic benefit can follow mitral valve replacement.

The snag about this sort of coronary surgery is that operation on soft necrotic heart muscle is somewhat unrewarding and whenever possible the patient should be maintained by medical means until at least 6 weeks and preferably 3 months have elapsed after the infarction.

The surgery of intractable angina has only become established in the last few years. Before the development of successful saphenous vein coronary bypass grafts, surgery for angina had gone through an era of pericardial phenolization and poudrage, of sympathectomy and even of omental implantation. Vineberg’s operation of implantation of the internal mammary artery into a tunnel in the left ventricular myocardium only achieved popularity through the work of the Cleveland Clinic group (Vineberg, 1946; Favaloro et al., 1967). They put the operation on a more scientific basis by using coronary angiography to localize the ischaemic area. Unfortunately, long-term evaluation of the results of the Vineberg operation has been disappointing. Direct attack on the coronary arteries is now at last proving safer and more successful than in past years, thanks again to the pioneer efforts of the Cleveland Clinic group. The use of saphenous vein grafts to provide a bypass between the aorta above the coronary ostium to the coronary artery beyond the obstruction brings immediate benefit to the patient and it looks as though these grafts are going to remain patent in the majority of instances (Favaloro, 1963). The right coronary and the anterior descending branch of the left coronary are both accessible for this operation but surgery on the circumflex branch remains difficult and therefore risky.

The recent phase of un criticised enthusiasm for cardiac transplantation has fortunately receded to be left, properly, in the hands of a few committed centres who will no doubt eventually succeed in overcoming the immunological problems to a sufficient extent to make survival of the transplant operation worth while for the patient. In the meantime artificial hearts may make the faint-hearted harder-hearted: these prosthetic hearts would probably have succeeded already had the problems of a suitable power supply been solved. No doubt a source of nuclear power small enough for implantation will soon be developed and it is by no means impossible that artificial hearts will make their clinical debut well before the transplant problems are better understood.

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PRIMARY FELLOWSHIP EXAMINATION

The last occasion on which Anatomy and Pathology will be accepted is May, 1971. As and from that date the subjects of the Primary Examination will be Physiology, Pharmacology, Physics, Clinical Measurement and Clinical Chemistry.