

RECENT ADVANCES IN CARDIOLOGY

No. 7

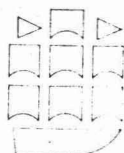
JOHN HAMER

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RECENT ADVANCES IN CARDIOLOGY

EDITED BY
JOHN HAMER

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PREFACE

The situation is still changing so fast in cardiology that we feel no need to apologise for a fresh edition of *Recent Advances in Cardiology* a few years after the previous one. While adhering to the previous policy of dealing with fundamental topics, major clinical problems and advances in therapeutics, the change in the contributors from the previous edition represents the new emphases of recent years. We return to presentation of new material on arrhythmias which have been neglected for several editions, although well covered years ago by East and Bain. Although once said to be the 'cardiological menopause' an interest in arrhythmias is no longer an old man's topic and the many new advances in our understanding of this subject produced by young workers in recent years are shown by Dr Krikler's descriptive chapter and Dr Spurrell's section on surgical treatment. The increased interest in coronary artery surgery has now been digested in Britain and is shown in Dr Rees' chapter on radiology and Mr Rees' section on the surgical aspects.

I have not thought it any part of my editorial duties to produce a uniformity of view among contributors, rather to encourage understanding by allowing overlap between chapters to show different points of view in controversial areas. You will find cardiac surgery in the neonate covered both by Dr Jordan from the physician's point of view and by Mr Rees speaking as a cardiac surgeon. The use of beta-adrenergic blocking agents in hypertension is discussed both by Dr Prichard, as an experienced user of these drugs and in my own chapter from a more theoretical point of view. Dr Aber's comments on arrhythmias in cardiac infarction add clinical flesh to the bones of Dr Krikler's largely diagnostic chapter on arrhythmias and Dr Kumana's contribution on anti-arrhythmic drugs. The appearance of two chapters on heart failure reflects the different research interests of Dr Taylor and myself, Dr Taylor being particularly interested in treatments aimed at reducing the load on the left ventricle and my own work being concerned with myocardial performance and the effects of digitalis and diuretic treatment.

London, 1977

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PAEDIATRIC CARDIOLOGY

S. C. Jordan

Today, paediatric cardiology in the Western World is almost exclusively concerned with the treatment of congenital heart disease, and brings physician and surgeon closer together than in almost any other branch of medicine. This cooperation has established firm principles for the treatment of individual lesions and the understanding of the abnormal anatomy and physiology on which advances in treatment of complex abnormalities can be planned.

SURGICAL TREATMENT

The Beginning

The successful resection of coarctation of the aorta and closure of patent ductus arteriosus in the 1930s and 1940s meant that about 12 per cent of congenital heart disease could be cured. In the 1950s the application of surface cooling ('conventional' hypothermia) with circulatory arrest for 5 to 7 min brought pulmonary stenosis and atrial septal defect within the curable category, making the total 'potentially correctable' group about 35 per cent, although, because of the benign nature of mild pulmonary stenosis and small or moderate sized atrial septal defects, the impact in terms of reducing mortality and morbidity was rather less than the figures imply.

Technical Developments

The end of the sixth and the early part of the seventh decade of the century brought the routine use of cardiopulmonary bypass and profound hypothermia for uncomplicated ventricular septal defect, severe aortic stenosis (with valvotomy or replacement), Fallot's tetralogy and a few less common lesions, notably total anomalous pulmonary venous connection. At this stage surgery was virtually limited to obstructive lesions and shunts, and to many it seemed fanciful to think that more complex lesions could be corrected. However, with Mustard's description of his operation for transposition of the great arteries in 1964 and its subsequent adoption on a large scale, all of the eight common forms of congenital heart disease, accounting for about 80 per cent of the total

number of patients with congenital heart disease could now be cured. To this could be added a further 3 per cent consisting of the uncommon lesions of total anomalous pulmonary venous connection, atrioventricular canal of the less severe (ostium primum) type, left ventricular to right atrial shunt (Gerbode defect), subaortic stenosis, lone infundibular stenosis and cor triatriatum, so that in theory only about 15 per cent of children with congenital heart disease were unable to be cured. Subsequent progress in this group will be described later, but the difference between the potential and actual success rates of the 85 per cent of 'curable' patients needs to be considered.

Operations in the First Year of Life

Many of the poor results in this group of patients could be related to the early onset of symptoms and the need to carry out surgery within the first year of life. Miller (1974) reporting on the management of 111 infants presenting in the first week of life, found that the overall mortality was 69 per cent, although 50 per cent had curable lesions and only 11 per cent were unsuitable for any form of surgical treatment. There is therefore clearly a need for improvement in diagnosis, surgical techniques and pre- and postoperative care. The high incidence of pulmonary complications after the use of cardiopulmonary bypass in the first year of life and the technical difficulties of carrying out cannulation of right and left sides of the heart (quadruple cannulation) by Drew's technique in small infants have been responsible for the reluctance to advise early corrective surgery for such conditions as large ventricular septal defect, severe Fallot's tetralogy and transposition of the great arteries. This has led to the use of palliative rather than corrective operations in infancy.

Ventricular Septal Defect

Banding of the pulmonary artery was for many years the standard operation for infants with large ventricular septal defects. The operation proved to be simple and safe in experienced hands, but subsequent closure of the ventricular septal defect is made more hazardous. There are few reports of large series of patients, but Coleman et al (1972) had 4 deaths in 15 patients. The result of debanding may not be very satisfactory for Dobell et al (1973) in a study of 18 patients on average of four years after debanding and VSD closure found gradients of up to 72 mmHg (9.6 kPa) across the site of debanding and felt that these gradients would increase with further growth of the child. Girod et al (1974) had better results in 34 patients, all banded before the age of one year. There were four deaths from banding and one from debanding and closure of ventricular septal defect and the highest postoperative gradient across the repaired pulmonary artery was 25 mmHg (3.3 kPa).

Fallot's Tetralogy

Palliative operations for Fallot's tetralogy in the first two years of life have never been very satisfactory. Closed pulmonary valvotomy produces only temporary improvement, infundibular resection is technically difficult and also produces only temporary improvement. Blalock-Taussing shunts are difficult and rarely remain patent, Potts anastomosis is very difficult to close subsequently and the Waterston operation is difficult to judge and may lead to proximal obstruction of the right pulmonary artery and unilateral pulmonary oedema or pulmonary arterial hypertension in the right lung.

This led to something of a vicious circle with patients being operated on only when in extremis, with further worsening of the overall surgical mortality. The vicious circle was broken by the introduction of the technique of extended surface cooling, thus greatly reducing the period of bypass required. Good results were quickly reported for large ventricular septal defects, Fallot's tetralogy and transposition of the great arteries (Barratt Boyes, Simpson and Neutze, 1971; Venugopal et al, 1973). Encouraged by these results operation was extended to the less critically ill infant, and in this situation some surgeons found it possible to revert to conventional cardiopulmonary bypass or bypass-induced profound hypothermia with equally successful results, so avoiding the long period of time required for surface cooling (Stark, 1972). Many surgeons now operate routinely on infants in the first few months of life.

Another cause of high mortality, often linked with the necessity for early operation, was particularly unfavourable anatomy. This particularly applied to Fallot's tetralogy with a very under-developed right ventricular outflow tract and therefore a large right to left shunt. This problem is still largely unresolved, but the ability to use right ventricular outflow tract reconstruction at a later stage has made it more acceptable to carry out a palliative shunt in infancy, without the worry of being unable to correct the lesion later due to further atrophy of the outflow tract.

Total correction in infancy however has the advantage that muscular hypertrophy of the outflow tract of the right ventricle is less than in older patients. Rees and Starr (1973) reported nine patients aged 5 to 11 months with one death and virtually normal postoperative right ventricular pressures (30–40 mmHg, 3.8–5.0 kPa) and in a larger series of 25 patients corrected under two years of age Starr, Bonchek and Sunderland (1973) had only two deaths from total correction compared with five deaths in a similar group of 16 patients treated just with palliative shunts.

Adverse Factors in Infancy

Complicating factors, both physiological and anatomical, have also been a cause of poor surgical results.

Ventricular Muscle Disease

In severe obstructive lesions there may be hypoplastic ventricular cavities, gross ventricular hypertrophy or fibroelastosis. Freed et al (1973) found that of 19 patients presenting with cyanosis due to severe pulmonary stenosis in the neonatal period 13 had some degree of hypoplasia of the right ventricular cavity. However, all of the 12 who were treated by pulmonary valvotomy survived (although two required a further valvotomy subsequently). One patient was treated with a palliative shunt and died. Similarly Miller et al (1973) reported five patients with severe pulmonary stenosis treated by pulmonary valvotomy and all survived. However, of their 15 patients with pulmonary atresia, all of whom had small right ventricular cavities, seven died during or immediately following operation and five later, the only three survivors being those treated by systemic-pulmonary artery shunts. Until a few years ago pulmonary valvotomy was attempted in all cases of pulmonary atresia as it provided the only hope of ultimate correction. The development of the Fontan operation (see later), with its greater potential, now makes palliation by some form of shunt a better prospect when the right ventricular cavity is very small. In severe aortic stenosis presenting with symptoms in infancy the problems are even worse. Lakier et al (1974) found that of ten such patients only two had a small left ventricular cavity. Nevertheless three died prior to surgery and only one survived surgery. They attribute these poor results to a very hypertrophied, non-compliant left ventricle.

Hypertensive pulmonary vascular disease associated with intracardiac shunts, particularly ventricular septal defect, has been the most important physiological problem. It is likely that most patients with high pulmonary vascular resistance, including those who end up with the Einsemler syndrome, have during early infancy gone through a stage when pulmonary blood flow is relatively high and hypertensive pulmonary vascular disease is reversible. Now that corrective surgery in infancy has an acceptable mortality, and provided these patients can be detected at the operable stage, the problem should be overcome.

Right Ventricular Outflow Reconstruction

Considerable progress has been made in treating the 15 per cent of patients with the more complex lesions. Right ventricular outflow tract reconstruction (Rastelli operation) has made curable pulmonary valve atresia with ventricular septal defect, persistent truncus arteriosus and transposition of the great arteries with ventricular septal defect and pulmonary or subpulmonary obstruction (the group having in common a discontinuity of pulmonary artery and right ventricle). In the least complicated of these lesions, pulmonary atresia with ventricular septal defect, the results of reconstruction have been good using a homograft aorta (with aortic valve in situ) and heterogenous

valves sewn into a Dacron tube, but the use of fascia lata has proved unsatisfactory as there is shrinkage with recurrence of right ventricular outflow obstruction (Macartney, Scott and Ionescu, 1975). In the more complicated lesions problems have occurred in locating the graft so that kinking and obstruction do not occur. In order to ensure a free exit from the right ventricle into the conduit the latter must be angulated forwards and, particularly in small children, this has then become squashed when the sternum is closed over it. To avoid this some surgeons advise leaving a window in the sternum and McGoon, Wallace and Danielson (1973) suggest using a conduit directed more to the left and carefully measured for size. When the pulmonary artery lies behind the aorta, in transposition, greater care has also to be used to place the distal end of the conduit around the aorta without obstruction or tension. McGoon et al (1973) reported on their results in 111 operations, all using homografts of aorta, including the aortic valve. The overall mortality was 32 per cent and was similar in the three main lesions treated, persistent truncus arteriosus, transposition with ventricular septal defect and pulmonary stenosis and pulmonary atresia with ventricular septal defect. As might be expected the worst results were in patients under the age of two years with a mortality of 80 per cent and the best results (18 per cent mortality) were in the 5 to 12 year group. Over 12 years the mortality was again higher (32 per cent) mainly due to the presence of hypertensive pulmonary vascular disease and multiple previous operations.

The particular problems in the overall management of persistent truncus arteriosus (the lesion for which the Rastelli operation was originally designed) are highlighted by a report of Poirier, Berman and Stansel (1975). From the literature they estimated that in untreated patients the mean age at death was two and a half months. Twenty-two reports of banding of the pulmonary artery in 76 patients gave an overall mortality of 50 per cent, although in the most propitious, type I cases (where a common pulmonary trunk arises from the base of the truncus) it was 33 per cent. Against this must be set the known high mortality, untreated, in infancy and the virtual certainty of hypertensive pulmonary vascular disease in unbanded survivors. In eight reports of the Rastelli procedure, including the 58 in Rastelli's own group, mortality varied from 29 to 100 per cent.

Double Outlet Right Ventricle

On a slightly less dramatic note, progress has been made in dealing with varieties of malposition of the great arteries other than complete transposition. Much of the lack of success in the earliest attempts to cure these lesions resulted from a lack of definition of the exact anatomy. Both great arteries may arise from the right ventricle, with or without pulmonary stenosis. This may be associated with malposition of the aorta and pulmonary artery with respect to each other, as well as with respect to the two ventricles (Lincoln et al,

1975). When the ventricular septal defect is immediately below the aorta, repair is relatively straightforward as a patch can be inserted from the ventricular septal defect to the anterior margin of the aorta so that the left ventricular outflow through the ventricular septal defect is channelled exclusively into the aorta. Attempts to treat the other variety in which the ventricular septal defect lies below the pulmonary valve, by producing a much longer tunnel within the right ventricle from ventricular septal defect to aorta, have seldom been successful and a more promising approach is to direct left ventricular outflow into the pulmonary artery producing physiologically complete transposition and to make an intra-atrial baffle (Mustard procedure) to divert systemic venous return to the left ventricle and pulmonary venous return to the right.

Double Outlet Left Ventricle

Comparatively little has been written about the condition in which both great arteries arise from the left ventricle. Pacifico et al (1973) report on four cases (three with associated pulmonary stenosis). The same operation can be carried out irrespective of the relationship of the ventricular septal defect to the two great arteries, for the ventricular septal defect is closed, the outflow to the pulmonary artery from the left ventricle is also closed and a conduit containing a homograft or heterograft aortic valve is placed between the right ventricle and the pulmonary artery.

Since many operations on complex lesions, from severe Fallot's tetralogy to double outlet left ventricle, now require construction of a right ventricular outflow tract, the fate of this reconstruction is of considerable importance. Kaplan et al (1973) reviewed 150 patients, many with severe Fallot's tetralogy, treated by patching the outflow tract or by homograft reconstruction. They found that homografts continued to function very well (although the actual conduit may calcify) whereas patch reconstruction led to aneurysm formation and embolism into the lungs.

Single Ventricle

A significant advance in terms of lesions previously regarded as uncorrectable has occurred in the treatment of single or common ventricle. (The two terms are interchangeable in terms of physiology although their use is disputed on anatomic and embryologic grounds.) Previously the best that could be hoped for was to produce a circulation with adequate but not excessive pulmonary blood flow by either banding the pulmonary artery if there was no pulmonary stenosis or carrying out a Blalock shunt or Glenn operation for those with pulmonary stenosis and much reduced pulmonary blood flow. An optimal situation would be a pulmonary blood flow about twice normal, since this load is reasonably well tolerated, cyanosis is minimal and there is no significant elevation of pulmonary artery pressure. (Patients without pulmon-

ary stenosis or pulmonary artery banding usually develop pulmonary hypertensive vascular disease which by the age of two to three years makes them inoperable.) Sporadic attempts have been made to divide the single ventricle in the past, and the use of the Fontan operation will be mentioned later but it is only recently that the principles on which successful septation of the single ventricle are based have been determined.

The first principle is to determine the site of the conducting tissue which, in single ventricle, is usually posteriorly situated (Anderson et al, 1974) but its position cannot always be predicted. Edie et al (1973) advocate the use of a probe to detect the His bundle activity. They reported four patients treated successfully by inserting a patch in the common ventricle, placing all sutures well away from areas in which His bundle activity was detected.

The second principle is to place the patch to give the most direct pathway between each AV valve and its nearest semilunar valve. This usually results functionally in the production of a situation of physiological transposition and it is then necessary to carry out an interatrial baffle procedure (Mustard operation) to provide physiological correlation (Edie et al, 1973). If there is an inadequate outflow to the pulmonary artery it may be necessary to reconstruct this in the same way as for pulmonary atresia or severe Fallot's tetralogy (Ionescù, Macartney and Wooler, 1973).

The third principle is to correct associated abnormalities which may include abnormalities of pulmonary venous drainage.

Clearly these complex operations still require considerable further experience before they can be regarded as standard procedures, but the results so far are promising, and although operation cannot usually be attempted before the age of 8 to 10 years, there is now a considerable stimulus to preserving infants with single ventricle by early palliative operations designed to produce a reasonable growth and exercise tolerance and to prevent the development of hypertensive pulmonary vascular disease. When there is no pulmonary stenosis this will usually entail banding of the pulmonary artery in infancy, and in the presence of severe pulmonary stenosis a palliative Blalock-Taussig shunt will be required to relieve cyanosis.

Tricuspid Atresia

Tricuspid atresia has until just recently been regarded by most surgeons with little enthusiasm. Although arterial to pulmonary artery shunt or Glenn (superior vena cava to right pulmonary artery) anastomosis provides palliation, the long-term outlook for affected children has been poor.

In the normal heart, and in cyanotic lesions with right heart obstruction and reduced pulmonary blood flow, it is known that the pulmonary artery pressure is only slightly higher than that in the right atrium. In the Glenn operation the additional pressure is provided by the hydrostatic pressure in the superior

vena cava, especially when the patient is upright, but no similar way could be found to allow inferior vena caval blood to perfuse the lungs.

Fontan and Baudet (1971) realised that the right atrium could provide this small extra pressure (particularly when, as commonly occurs in tricuspid atresia, it is hypertrophied) provided that reflux down the inferior vena cava could be prevented during atrial contraction. The solution was to provide a conduit, from the right atrium to the pulmonary artery and place a homograft aortic valve at the junction of inferior vena cava and right atrium. In their first patient, aged 12, Fontan and Baudet carried out a Glenn procedure and then anastomosed the right atrial appendage to the proximal stump of the right pulmonary artery (Fig. 1.1A). In their second patient they used a conduit containing a homograft aortic valve from the right atrium to the main pulmon-

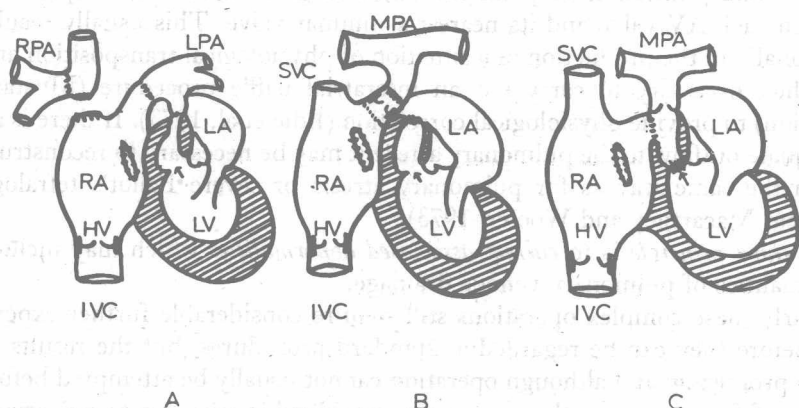


Figure 1.1 Fontan operation. (For description see text.) HV, homograft aortic valve; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery; RA, right atrium. The arrow points to the ventricular septal defect, which is closed in C

ary artery (Fig. 1.1B) and used the right atrium to perfuse the whole pulmonary bed. Both these patients had a homograft inserted at the IVC-right atrial junction and both survived, although a third died. Kreutzer et al (1973) introduced a modification for patients in whom the pulmonary valve was normal, in which the conduit from the right atrium is inserted below the pulmonary valve into the rudimentary right ventricle and the ventricular septal defect closed, so avoiding the use of two homografts (Fig. 1.1C). Both their two patients survived.

Several groups have now reported their results with this operation (Stanford et al, 1973; Walker, Sbokos and Lennox, 1975) and it seems clear that dramatic improvement can be obtained. Most of the reports include patients who have previously had a Glenn operation and since in these the right atrium has only to handle a little over half the venous return these are a particularly favourable group. Most reports, though, are of only a few patients and it is

equally clear that many problems need to be clarified. In most instances the immediate postoperative period is complicated by a considerable rise in right atrial pressure. One frequent consequence of this is that atrial fibrillation occurs and with the lack of atrial transport the right side of the heart acts only as a conduit. Long-term survival with atrial fibrillation has been reported but only at the price of a permanently raised venous pressure. In the few days following operation problems in fluid balance are almost invariable (Walker et al, 1975). Large amounts of intravenous fluids are required to maintain an adequate cardiac output and some of these problems are related to the effects of distension of the right atrium in stimulating stretch reflexes and producing both reflex cardiac acceleration and reflex suppression of antidiuretic hormone release.

The discovery that, even with the right atrium acting merely as a conduit, survival is possible has led some surgeons to perform the operation without the use of an IVC-RA homograft valve (Kreutzer et al, 1973). (The insertion of the valve is made difficult by the need to maintain cannulation of the IVC during cardiopulmonary bypass.) Although there have been some survivors using this simpler technique it has been necessary at times to insert the valve at a later stage in order to control severe right heart failure. Because of the need to insert a conduit and valve which will remain of adequate size into adult life the optimum age for operation is not before about seven to eight years although a few surgeons have reported success in younger children.

Since the success of the operation depends partly on the degree of the hypertrophy of the right atrium, the long-term possibility of the operation needs to be considered in the early palliative management of the condition. Formerly it was usual to perform balloon septostomy or surgical septectomy to ensure free drainage of blood from right atrium to left atrium, but now it may be better to leave the right atrium with a little obstruction to ensure adequate hypertrophy.

Fontan Operation in Other Lesions

Although the operation of right atrial to pulmonary artery conduit was originally designed for tricuspid atresia its potential use is more widespread. Because of the low pumping pressure possible it can only be applied where the pulmonary vascular resistance is normal. In pulmonary atresia with intact interventricular septum and hypoplastic right ventricle the haemodynamics are almost identical to those of tricuspid atresia. Although in some patients early pulmonary valvotomy may allow the right ventricle to grow this is not always the case and where the right ventricle retains its minute cavity and thick non-compliant walls, Fontan's operation represents a big improvement in prognosis. In single ventricle with pulmonary stenosis, or where the pulmonary artery has been banded early enough in life to prevent hypertensive

pulmonary vascular disease, the operation is carried out as for tricuspid atresia, the tricuspid valve being closed over from the right atrium by a patch.

Stanley and Kolff (1973) made one interesting observation from their experience of this operation. It has been known that in severe cyanotic congenital heart disease there is a considerable collateral circulation to the lungs, but whether this joined the pulmonary circulation at the precapillary or capillary level was uncertain. However, during perfusion of two patients for Fontan operations for tricuspid atresia (both had had previous Glenn operation) they found that blood passed from the collaterals through the alveolar circulation to the pulmonary arteries, establishing the capillaries as the point of entry of the collateral vessels.

Valve Replacement in Children

One of the major disadvantages that confront the paediatric cardiac surgeon, which does not trouble his counterpart operating only on adults, is the impracticability of valve replacement during the years of rapid growth of the child. However, Klint et al (1972) have now reported their results in 14 children aged $4\frac{1}{2}$ to 15 years, all given prosthetic valves (including one triple valve replacement). There were five deaths and nine survivors. They also reviewed a total of 166 paediatric patients treated by valve replacement (176 valves in all). One hundred and eighteen of these patients were given anticoagulants, in general with no more problems than in adults and the overall rate of systemic embolism in the whole group was 5 per cent. In patients of 10 years and over requiring valve replacement it was generally found that the degree of cardiac enlargement was great enough to allow adult sized prostheses to be used. Cartmill et al (1974) also reported the use of tilting-disc prostheses in right ventricular outflow reconstruction. These results should certainly encourage wider use of valve replacement in life-threatening situations in children.

INVESTIGATION OF CONGENITAL HEART DISEASE

More complex operations call for more detailed and meticulous investigation and cardiac catheterisation and angiocardiography are still the mainstay of preoperative diagnosis. Advances have been mainly in the accurate and more detailed anatomical and physiological diagnosis of uncommon and complex lesions.

Blood supply in pulmonary atresia

In pulmonary atresia with ventricular septal defect it is necessary to know not only the anatomy of the blood vessels carrying blood to the lungs but also the pressures in the different parts of the pulmonary arterial tree. Macartney, Scott and Deverall (1974) demonstrated that the blood supply to the lungs