CONGENITAL AORTIC STENOSIS

A REVIEW ILLUSTRATED BY 18 CASES TREATED BY OPEN-HEART SURGERY

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The subject of this paper is one that until recently was thought to be rare, benign in its course, capable of differentiation between valvular and sub-valvular types, and not amenable to adequate and safe surgical correction. This presentation, on the contrary, will show that it is far from rare, can be malignant in its course, cannot readily be clinically separated into its 2 varieties, and is now satisfactorily and safely cured by direct-vision surgery with the aid of the pump oxygenator.

History

Carolus Rayger in 1672, according to Campbell,[1] first described congenital valvular stenosis in a Parisian cobbler aged 40. Chevers, according to Brock,[2] first described a case of sub-valvular stenosis in 1842. Tuffier[3] in 1913 was probably the first to attempt surgical correction of this malady in humans. In the following year Carrell[4] first suggested direct vision for surgery of the aortic valve and successfully opened the aorta for 2½ - 3 minutes in a series of experiments. Smithy,[5] himself suffering from aortic stenosis, passed a valvulotome through the aortic wall in 22 animal experiments, but haemostasis was inadequate with his methods. In the same year, 1947, Brock[6] used an operating cardioscope which he passed down the right subclavian artery. In 1952 Bailey[7] of Philadelphia first reported his technique of transventricular valvotomy.

The first direct-vision operation recorded was by Clowes[8] in 1954. He employed a pump oxygenator, but the patient did not survive. On 10 October 1955, Julian[9] successfully exposed the aortic valve under hypothermia in man. Lewis[10] and Swan,[11] in the following month, and Brock,[6] in January 1956, had their first successes under hypothermia.

It was left to Lillehei,[12] the pioneer of the bubble oxygenator, to report the first successful treatment of calcific aortic stenosis by direct-vision surgery with cardiopulmonary by-pass.

Incidence

In her classical monograph, first published in 1936, Maud Abbott[13] reported 23 cases in her 1,000 autopsies of congenital heart lesions. Dry,[14] from the Mayo Clinic, in his monograph on 'Congenital anomalies of the heart and great vessels', published in 1948, had no record of congenital aortic stenosis in the 132 cases reviewed. Paul Wood[15] in his book 'Disease of the heart and circulation' devoted half a page to this subject and stated: 'aortic valvular stenosis is rare'. McMahon,[16] in 1953, reviewed all congenital heart lesions in Birmingham over

a 10-year period, and found only 4 patients with congenital aortic stenosis among his 372 cases. On the other hand, Nadas,[17] from his extensive experience at the Children's Hospital in Boston, in 1957 reported 67 cases in 5 years. Campbell[1] reported 40 cases from Guy's Hospital. In his experience, 7% of acyanotic heart lesions and 3% of all congenital lesions are congenital aortic stenosis.

Sex Incidence

Most patients are males. Nadas[17] stated that the condition is 5 times more common in boys. Spencer[18] described 11 males out of 12, while Morrow[19] recorded 20 males out of 30. Of our 18 patients, 12 were males.

Age

Although we are describing a congenital lesion, few cases have been recognized in early infancy. However, together with patent ductus and coarctation the condition should always be considered when cardiac failure occurs in infancy. Cooley,[20] out of a surgical experience of 13 cases, operated upon 3 desperately ill, pulseless infants under the age of 6 months. Our youngest patient was aged 5 years and our oldest 46 years.

Pathogenesis

The bicuspid aortic valves with stenosis are evolved from the proximal ends of the distal bulbar swellings, and fusion could occur at this stage. Brown[21] quoted Farber and Hubbard[22] as showng cases in which foetal endocarditis was responsible for this lesion. Sub-aortic stenosis, on the other hand, according to Keith[23] and quoted by many authors, is always embryological in origin and is caused by imperfect inclusion, or persistence of part of the bulbus cordis into the outflow tract of the left ventricle. This raises an irregular ridge of thickened fibrous endocardium on the prominent muscle of the septum which crosses from there onto the ventricular surface of the aortic cusp of the mitral valve.

Pathology

Although Campbell[24] and Brown[21] stated that the valve cusps are thickened, irregular and deformed, even in children, this is not our experience, possibly because of the youth of most of our patients. Variations in size and shape of the cusps do occur. Bicuspid valves are a common anomaly and were found in 6 of our cases. A diaphragmatic dome with a central opening, such as is almost invariably found in congenital pulmonic stenosis, has been rare in our cases. Calcification occurs in the older age group, as was shown in one of our patients, aged 32, who was subjected to operation under hypothermia. In almost all our cases commissures could be detected.

Post-stenotic dilatation of the aorta distal to the obstruction is usual, and its absence suggests some degree of hypoplasia of the aortic wall. This we saw in our 12th case, where the aorta was small and its wall was almost cartilaginous in character and associated with narrowing of the valve ring. In our second case performed under hypothermia, the wall of the aorta, itself greatly dilated, was unfortunately paperlike and we lost this patient from reactionary haemorrhage from the aortotomy incision. Autopsy confirmed that she had ovarian agenesis — Turner's syndrome. The left ventricle showed marked concentric hypertrophy with a wall thickness of 3·5 cm. and a minute cavity.

In sub-aortic stenosis there is usually a fibrous diaphragmatic ring or an irregular raised ridge of thickened fibrous endocardium on the prominent muscle of the septum 1 - 3 cm. below the valves, and across from this onto the ventricular surface of the aortic cusp of the mitral valve. This fact must be reiterated — the aortic cusp of the mitral valve forms the left posterior portion of the left ventricular outflow tract and the base of the sub-valvular ring. Marquis and Logan,[25] at autopsy, have confirmed fibrosis of the left ventricular muscle fibres following death of the cells from anoxia and oedema secondary to compression by the hypertrophied muscle mass.

In our series of 18 cases, 6 patients had bicuspid valves, 4 had sub-valvular stenosis, and 4 had hypoplasia of the aortic ring.

Associated Lesions

Probably coarctation of the aorta is the most common lesion associated with congenital aortic stenosis and was present in our first case. Of Downing's[26] 37 cases, 8 had coarctation of the aorta. Pappas[27] reviewed 2 such patients operated upon by closed methods by Bailey through the left chest, and quoted 24 autopsy reports from the literature.

Patent ductus arteriosus is also quite common as an associated lesion. Bonham-Carter[28] reported 8 cases of patent ductus associated with aortic disease, but of these only his last patient had congenital aortic stenosis. We tied off a patent ductus in a child aged 6 years, and 2 years later performed open aortic valvotomy on her. In a second case a patent ductus was ligated in 1949, and at aortotomy 11 years later a hypoplastic aortic valve ring was found.

Hypoplasia of the ascending aorta is not uncommon, and a narrowed aortic ring can be one of the major problems in surgery, as we have found in 4 of our cases.

In our series of 18 cases, 1 was associated with coarctation of the aorta, 2 with patent ductus, and 1 with abnormal thoracic vertebrae.

Prognosis (Table I)

Although until recently it was felt that these lesions were benign in their outlook, this has not been substantiated by more recent critical reviews. Nadas[17] recorded 5 deaths in his 67 patients over a 5-year period. If there is left ventricular hypertrophy on the electrocardiogram the prognosis is bad in his opinion. Helen Taussig[29] stated 'most individuals live at least to adult life. The majority die of subacute bacterial endocarditis. Sudden death may occur'. In Campbell's[1] series of 40 patients 2 died suddenly while under observation. Maud Abbott[13] recorded that the average duration of life in all cases of valvular stenosis was 3¾ years, whereas in 12 cases of sub-aortic stenosis it was 22¾ years. Brock[2] stated that there is no difference in prognosis between the 2 types. Braverman[30] quoted an 8·2% mortality in 73 patients under the age of 20. Kjellberg[31] described 1 death at 14 years of age in his 15 patients.

TABLE I. MORTALITY RATE IN SIX SERIES OF CASES

Author	*No. of cases*	*Mortality* %
Braverman[30]	73	8·2
Nadas[17]	67	7·5*
Campbell[1]	40	5·0
Downing[26]	37	8·0
Marquis and Logan[25]	28	17·8**
Kjellberg[31]	15	6·6

* Over a 5-year period.
** Followed for 4 years.

Marquis and Logan[25] had 5 deaths in their 28 patients followed for 4 years. They, however, stated that 'sub-aortic stenosis as an isolated lesion appears to be very uncommon and rarely of sufficient severity to require operative treatment'. Downing[26] summed up the position thus: 'This is the only congenital heart disease compatible with a relatively long life that requires restriction of activity. Aortic stenosis constantly threatens acute coronary insufficiency. The margin between adequate and inadequate coronary flow is small and any activity may quickly exhaust the reserve and lead to failure and sudden death'. He reported 3 sudden deaths (from either sudden diminution in left ventricular output or carotid sinus reflexes) in 37 cases.

Symptomatology (Fig. 1)

Most patients are asymptomatic during infancy and childhood, but develop symptoms in adolescence. Rarely, marked cardiac failure occurs in infancy and sweats are a pronounced symptom. According to Marquis and Logan[25] even slight disability is significant. Fatigue is a common and early symptom, is often associated with palpitations and an awareness of forceful heart action, and is related to inadequate systemic blood flow. These symptoms indicate that the myocardium is beginning to feel the effects of an unyielding obstruction at the aortic-valve level.

Angina results from the discrepancy between coronary flow and myocardial oxygen demands. Of Campbell's[1] 40 patients, 11 suffered from angina, but all started complaining in their

teens. His youngest patient with angina was 13 years old. Of Downing's[26] 37 patients, only 1 had angina. Marquis and Logan[25] stated that angina usually occurs in older patients; it presented in only 1 of their patients, aged 24 years. One of the youngest recorded patients with angina was case 4 of Spencer,[15] who developed angina at 4 years of age. Thirteen of our patients complained of angina or some form of chest pain, not necessarily related to exercise — the youngest was a boy of 5 years of age (case 7). In case 2 — a girl of 17 — angina was incapacitating. Dizziness, probably from relative cerebral ischaemia, is a common symptom and occurred in 20% of Campbell's[1] 40 cases. Cardiac syncope is ominous, and again is related to inability of the left ventricle to increase cardiac output and cerebral blood flow during exercise.

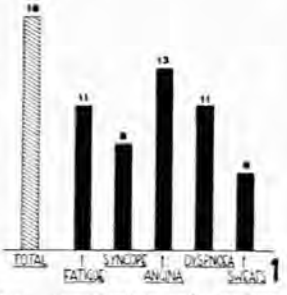

Fig. 1. Number of patients showing various symptoms (present series).

The increased requirements of the left ventricle can be met only by higher ventricular filling pressure, which is associated with elevated pulmonary venous pressure resulting in left ventricular failure and its symptoms. Dyspnoea usually occurs over the age of 30.

Physical Signs

1. General development is satisfactory except in those patients with other associated anomalies.

2. The pulse is often normal, sometimes diminished in amplitude, and occasionally virtually absent. Marquis and Logan[25] stated that the aortic orifice has to be reduced to less than a quarter of its normal size before the form of the arterial pulse is altered.

3. Blood pressure is often within normal limits. With associated aortic incompetence or coarctation of the aorta, it is abnormal.

4. There is a left ventricular lift to the apex beat which is forceful.

5. There is a palpable coarse systolic thrill over the aortic area, best felt on expiration and on leaning forwards. This thrill is felt over the carotids and in the suprasternal notch.

6. There is a loud harsh systolic stenotic murmur, best heard at the second right interspace, with wide transmission to the neck, back and chest. In very young infants it is stated by Spencer[15] that the murmur is best heard to the left of the sternum.

7. There is often an early systolic ejection click and the first sound is often loud and split.

8. The second heart sound over the aorta may be diminished or absent.

9. The first sound at the apex is often accentuated.

10. There is rarely an apical mid-diastolic murmur which disappears after successful valvotomy.

11. There is often (perhaps in 20 - 30% of cases) an early diastolic murmur along the left sternal border indicative of some aortic incompetence caused by dilatation of the ring from post-stenotic dilatation.

In our series of 18 cases, all 18 had a systolic murmur and thrill, 13 had a forceful left ventricular apex beat, and 4 had an early diastolic murmur.

Electrocardiography

The electrocardiogram is normal in most asymptomatic cases. The presence of left ventricular hypertrophy is always associated with moderate to severe outflow-tract obstruction according to Morrow.[32] In the 37 cases recorded by Downing,[26] 22 showed left ventricular hypertrophy.

Radiological Examination (Fig. 2)

The heart is often of normal size in asymptomatic cases. There is usually post-stenotic dilatation of the ascending aorta, best seen as an anterior prominence in the left anterior oblique view. Calcification in the ring or aortic cusps is exceptional under the age of 40. There is occasionally some prominence of the left atrium, but the pulmonary vasculature is invariably normal in the uncomplicated case. In the presence of associated lesions the radiograph may show other signs — e.g. notching of the ribs in coarctation.

Haemodynamic Studies

Brock and his associates[3,9] have shown the safety of transventricular left-heart catheterization. A needle is inserted through the anterior chest wall into the left ventricle. A polythene catheter is threaded through the needle and is passed up into the aorta. A withdrawal tracing may help to differentiate between valvular and sub-valvular stenosis. We have found this easy and safe in cases of acquired aortic stenosis, but we have had great difficulty in finding the left ventricular cavity in cases of congenital aortic stenosis, even at open operation. This we have shown to be caused by the marked concentric hypertrophy of the left ventricle, which leaves a minute chamber. When the heart is enlarged the procedure is simple, since the cardiac enlargement indicates cardiac dilatation and an enlarged chamber. This procedure can also be carried out transbronchially, but in the absence of an enlarged left atrium and in young children it is more dangerous. We have, therefore, performed few pre-operative left-heart catheterizations in this series, but have taken pressures at cardiotomy where indicated.

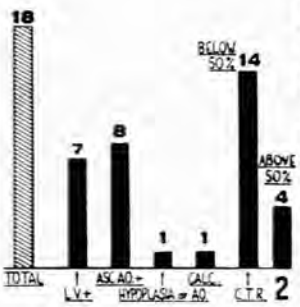

Fig. 2. Radiological signs found in this series. (LV + = enlarged left ventricle, Asc. Ao. + = dilatation of the ascending aorta, calc = calcification. CTR = cardiothoracic ratio.

When hypothermia was used, because of the limitations of operating time, the distinction between valvular and subvalvular stenosis was of paramount importance. However, now that we use the pump oxygenator, the differentiation is academic, since there is ample time, not only for full assessment, but for reparative surgery. Spencer[15] agreed with this opinion, saying: 'left-heart catheterization is not advised as a routine', because the clinical diagnosis was often clear and the electrocardiogram was a good guide to the severity of the stenosis. Morrow[32] agreed that left-heart catheterization is the most helpful procedure for the evaluation of congenital aortic stenosis. However, for real value, not only the pressure, but also the flow across the valve, must be measured.

Angiocardiography

Selective left-heart angiocardiography will show the stenosis very beautifully, as demonstrated by a case reported by Bjorck.[34] We have not used this method in our cases.

Indication for Operation

Until a few years ago the prognosis was thought to be uniformly good and, in any case, no adequate surgery was available. Although we had used hypothermia, its limitations in this field were such that we operated upon urgent cases only. Now with dry-heart visual surgery with the pump oxygenator, the need for surgery can be matched with safety and adequacy in technique.

We advise surgery in all symptomatic cases and in all cases where there is left ventricular hypertrophy on electrocardiogram or on radiological investigation. Morrow[32] advised operation in all symptomatic cases and in asymptomatic cases where the pressure gradient is in excess of 70 mm.Hg or where the orifice is less than 0·3 sq. cm. per sq. metre of body surface. Nadas[17] advised surgery if the valve area is less than 0·6 sq. cm., or where the gradient is above 40 mm.Hg. Braverman[30] stated that patients with syncope, easy fatiguability, or left ventricular hypertrophy should be referred for surgery. Spencer[15] and Downing[26] supported these criteria. Campbell[24] stated: 'The outlook for these congenital cases without operation is, however, worse and more uncertain than we thought at one time. Sudden death, sometimes with no

obvious warning or after attacks of syncope or angina is not rare'.

Marquis and Logan[25] assessed the severity of the lesion and the basis for surgery as follows:

1. By increased force of contraction, compensation is maintained and symptoms are minimal. The presence of symptoms suggests that the stenosis is severe and compensation inadequate.
2. Hypertrophy alone seldom leads to sufficient cardiac enlargement for clinical or radiological detection. Clinical enlargement of the heart is usually caused by dilatation, and its presence, when there is no associated aortic incompetence, suggests that hypertrophy alone has proved inadequate and that the stenosis must be severe.
3. There is no evidence that a congenitally stenosed orifice gets larger with the increasing demands of a growing child. With an orifice of fixed size, compensation that is adequate in early childhood may become inadequate as body growth proceeds. Stenosis that appears mild in early childhood may become severe before adult life is reached.

OPERATIVE CORRECTION

The surgical procedures available before safe surgical by-pass were: closed transventricular or transaortic valvotomy and open-vision correction under hypothermia.

A. Closed 'Blind' Methods

1. *Brock's Transventricular Approach*

Although this approach through the left ventricle was introduced by Bailey in 1952 for acquired aortic valvular stenosis, it was popularized by Sir Russell Brock, whose technique was safer and simpler. It was also used for congenital cases. Marquis and Logan[25] operated on 6 patients, 5 of whom had postoperative aortic incompetence. Downing[26] reported 19 cases with only 1 operative death and improvement in 14. We have not used this technique at all, since we believed that incompetence was produced in valvular stenosis, and that the obstruction was insufficiently removed in the sub-valvular cases. From what we have seen at open operations, this method has no place in our modern armamentarium, but is of historic interest only.

2. *Retrograde through the Aorta*

This was said to have the advantage that patients died of haemorrhage instead of ventricular fibrillation!! Its limitations are similar to those of the transventricular approach.

B. 'Open' Vision Methods

1. *Under Hypothermia*

This was a great advance, since the valve now could be clearly visualized, and adequate and controlled valvotomy performed. However, in our experience, the time is so limited that there is no margin of safety if anything untoward is found or occurs. This method was first successfully employed by Julian[9] in 1955 and by Brock on 26 January 1956. As recently as 1957 Brock[9] stated 'for non-calcified cases open aortic valvotomy under direct vision and with hypothermia is used'. Brock[2] lost case 5 (a girl aged 7 years from Johannesburg) whose aortotomy incision tore, and 12 minutes of occlusion were necessary for closure. Dye[32] had no mortality in 5 cases, of which 2 were sub-valvular. Morrow[39] had 2 deaths in 10 cases. Swan[36] had 3 deaths in 11 cases.

In 1957 and early 1958 we operated on 4 patients under hypothermia, using Swan's[36] technique as modified by Bedford, Sellors and Sellick.[37] The first patient was a woman of 43 (referred by Dr. S. J. Fleishman) who had an associated coarctation of the aorta, and who developed irreversible ventricular fibrillation after valvotomy. The second patient, a girl of 16, was referred by Dr. G. R. McLeish with intractable disabling angina. She also had primary amenorrhoea. An excellent valvotomy was obtained, but the lower end of the incision extended beyond the clamp. A further period of venous occlusion allowed control of the incision. She collapsed 3 hours after operation from reactionary haemorrhage. Autopsy confirmed a Turner's syndrome with ovarian agenesis, and haemorrhage from the aortic incision.

The third patient, a male of 32 (referred by Dr. N. Segal of the Cardiac Clinic of the Johannesburg General Hospital), known to have had a murmur all his life, had a very successful valvotomy. Operation confirmed grossly calcified valves. These 3 patients were all incapacitated. The fourth was a woman of 21 years, referred by Dr. B. van Lingen, with a forceful apex beat clinically and left ventricular hypertrophy on electrocardiogram. Aortotomy showed normal valve cusps with a sub-valvular diaphragm which was readily incised and dilated. Soon after closure of the chest a reactionary haemorrhage was suspected and the bilateral thoracotomy incision was re-opened. A bleeding point on the aorta was controlled and she recovered from this, but not without incident. She developed anuria which was successfully combated.

2. *Open Vision with Cardiac By-pass*

There is no doubt from our experience that this is the only safe and adequate method. Cooley[20] has operated upon 13 patients of whom 3 were under 6 months of age.

TABLE II. RESULTS IN FIVE SERIES OF CASES—OPEN AORTIC VALVOTOMY WITH THE PUMP OXYGENATOR

Author	*No. of cases*	*Operative mortality %*	*Sub-valvular*	*Aortic incompetence*
Spencer[15]	12	0·0	4	2
Cooley[20]	13	7·5		
Morrow[32]	3	33·3		
Gross[38]	16	12·5		
Adler and Fuller (present series) ..	14	0·0	4	4*

* Increased postoperatively in 2 patients.

We operated on 4 patients under hypothermia, of whom 2 died, while there were no deaths in our 14 patients operated on under open vision with cardiac by-pass (Table II).

OPEN AORTIC VALVOTOMY WITH CARDIAC BY-PASS

The particular variety of pump oxygenator is not as important as the efficiency of its operation, the prudence of the surgery, the maintenance of normal blood pressure and pH during operation, and postoperative supervision. We believe, however, that the disc oxygenator combined with de Bakey pump heads in a heart-lung machine of the Gross type[37] is slightly better than the others currently available.[45]

Pre-operative Treatment

The patient is admitted a week before surgery and is again fully assessed clinically and haematologically. Our patients are digitalized, given diuretics if necessary,

schooled in physiotherapy, and introduced to the 'special' nurses who will be in charge of them when they return from the theatre. Three days before the proposed operation the whole Unit meets to discuss every aspect of preoperative therapy and operative technique. Blood is also taken on this day for grouping, to exclude any rare group which might entail extra work for the blood transfusion service. Blood is again sent off the day before surgery for compatibility tests.

We have found no problem with blood collected the evening before surgery. The South African Blood Transfusion Service has never yet failed to supply us with our blood transfusion requirements. The details of this service have recently been reviewed by Shapiro.[39] A glucose drink is given, 3 - 4 hours before surgery, to obviate any hypoglycaemia which might be present because of enforced starvation and the low sugar content of stored blood.

The patient is premedicated, and cannulations for venous and arterial pressure readings are performed under local anaesthesia in adults and older children. In infants and young children we prefer having this done under general anaesthesia.

Incision

We now use a vertical sternal split which gives excellent access to the heart. This incision is preferred to the bilateral 'suitcase' incision which crosses from one axilla to the other. It takes much less time to open and to close the vertical split and, since it is much less traumatizing to the chest wall, the thoracotomy is virtually painless postoperatively. It also has the advantage of not opening either pleural cavity.

Cannulation

For our arterial return we now employ the right femoral artery, using metal cannulae made from tracheotomy tubes. These have the advantage of being curved, with a thin wall and a large lumen. For venous return we use McGill's portex endotracheal catheters, inserted through separate incisions in the wall of the right atrium, and controlled with purse strings passed through a short length of rubber tubing. The inferior catheter is passed from above downwards through the side of the atrial appendage into the inferior vena cava. The superior is passed from below upwards through a separate incision in the side wall of the atrium. In left-heart surgery we also pass a coronary sinus sucker through the tip of the left atrial appendage into the left atrium for decompression of the left ventricle; this is especially necessary after by-pass. The venous line is now fitted from the machine to the nylon 'Y' connection, care being taken not to capture any air bubbles which could act as an air lock and interfere with siphonage. Anoxic or cold arrest is now used in preference to the potassium arrest of earlier cases.

Phase of Partial By-pass

Immediately before going on to partial by-pass, a mixture of 98% oxygen and 2% CO_2 is fed into the oxygenator, and the anaesthetist prepares for further 'scoline', which is usually necessary because of dilution of the anaesthetic when by-pass is instituted. Every connection, as well as the additional sucker which has been attached to the coronary sinus sucker, and which will be used to keep the intracardiac operative field dry, is tested. Venous pressure, arterial pressure, and electrocardiogram are all checked. When the surgeon is satisfied on these points partial by-pass is instituted.

Phase of Total By-pass

The superior vena caval catheter is now introduced into the superior vena cava from its position in the right atrium. Momentary occlusion of the inferior vena caval catheter ensures patency and drainage of the superior catheter. When this has been shown to be satisfactory, the caval encircling ligatures are tightened and secured in position by clamping the ligatures distal to their rubber shrouds.

Phase of Total Cardiac Occlusion

After full by-pass has been established for several seconds, and when monitoring of arterial and venous pressure is satisfactory, the anaesthetist keeps the lungs inflated with helium, but does not inflate them rhythmically. A transverse sinus clamp is now placed across the aorta just below the innominate artery.

Open Cardiotomy Phase

The aorta is now incised with a No. 15 Bard-Parker blade and the incision is extended for 2 inches with a right-angled Pott's scissors. This extends down almost to the origin of the right coronary artery. Blood is now sucked away by the 'coronary sinus sucker' and the aorta and its valves are carefully inspected. Blunt nerve hooks are used to assess the valves and the stenosis. Again, a sharp scalpel blade is used to divide the fused commissures most carefully to within 2 mm. of the ring. In several of our earlier cases we cut only 2 commissures for fear of causing incompetence, in accordance with Spencer's[18] recommendations. This, we believe, is wrong, and we now carefully divide all 3 commissures. Six of our 18 cases have been bicuspid. At that time we were not aware of Austen, Shaw and Scannell's[40] work at the Massachusetts General Hospital where, with autopsy perfusion, they showed that 'division of only 2 of the commissures results in moderate aortic stenosis'.

After adequate valvotomy (aided if necessary, as in case 12, by gradual controlled stretching with a Brock's 3-bladed dilator), or if no valvular stenosis is found, the sub-valvular area of the outflow tract of the left ventricle is most carefully inspected for sub-valvular obstruction. In 3 of our 18 cases this was found. The diaphragm was either carefully incised and dilated, or partially excised and dilated. A satisfactory opening was accepted when an index finger and the largest Brock's bougie passed in readily.

Closure of Aortotomy

The left atrial suction is reduced sufficiently to allow blood to spill up from the left ventricle through the valves into the aorta. A finger is inserted down through the cusps into the left ventricle to empty it of any trapped air. The incision is then carefully closed with a running mattress suture. The first and last stitches are placed beyond the line of the incision and blood is allowed to fill the aorta before tying the last 2 stitches. When the aorta is filled, any retained air is aspirated by needle

puncture. Firm pressure is now applied to the aortotomy incision and the clamp across the aorta is slowly released. Coronary-artery flow now commences immediately and cardiac action gradually returns. In case 5 ventricular fibrillation appeared, but the heart defibrillated readily.

Cessation of By-pass

When cardiac action is seen to be effective, as judged by maintenance of arterial pressure, satisfactory electrocardiogram, and normal venous pressure, the caval ligatures are loosened, inflation of the lungs with 100% oxygen is commenced, and the superior vena caval catheter is withdrawn into the body of the right atrium. The inferior vena caval catheter is then completely withdrawn and its incision controlled by tying the purse-string suture. Left atrial suction is still being gently employed to assist the left ventricle. After a few minutes, when blood balances have been carefully assessed, by-pass is stopped, but the remaining superior vena caval catheter is withdrawn only when the electrocardiogram and arterial pressures are completely satisfactory. Immediately thereafter our attending haematologist gives the calculated dose of polybrene (previously protamine sulphate) slowly, to reverse the heparin. Pressures are now taken again in the left ventricle and from the central arterial pressure line to see if the gradient has been abolished.

Closure

The left atrial catheter is now withdrawn. All sutures are carefully inspected, catheters to drain the pericardium and mediastinum are placed, and the anterior pericardium loosely approximated. In cases operated on subsequent to this initial series, the pericardium has been widely fenestrated into the right pleural cavity, which has been drained by a large underwater seal catheter. The sternum is held together firmly by wiring, and the incision carefully closed in layers. The patient is X-rayed at the end of the operation, in the theatre, and bronchoscopy performed if necessary. In case 11 an unsuspected left pneumothorax was demonstrated and decompressed.

Medical and Biochemical Control

Dr. M. Zion and Dr. L. Braudo have been responsible for constant monitoring of the arterial pressure and electrocardiogram during operation, and have personally supervised the early postoperative care. Mr. L. du Plessis, later Mr. D. Evans and Mr. W. du Plessis, have managed the pump oxygenator and have been guided by the arterial pressure (we believe cerebral circulation is adequate in the presence of a satisfactory systemic pressure), the venous pressure and the venous and arterial oxygen saturation. Drs. H. B. W. Greig and A. Walker of the South African Institute for Medical Research have, with their mobile laboratory attached to the theatre, estimated heparin and protamine (later polybrene) requirements, and monitored both venous and arterial oxygen saturations. Their frequent pH estimations during operation and especially after by-pass, and in the critical 8 hours following operation, have been invaluable. They have also performed a large number of haematological and biochemical estimations which have confirmed the safety of the Gross type of pump oxygenator.

Dr. W. Scott has made fluid balance his concern and has been responsible for simple venous monitoring, a valuable index of blood depletion or of overloading.

Postoperative Complications (Table III)

A. *Special to perfusion cases:*

1. Post-perfusion syndrome with hypotension, peripheral cyanosis and irritability. This is not common where high flow rates have been employed and oxygenation has been satisfactory; it has not been seen in this series. However, case 5 showed marked postoperative restlessness.

TABLE III. OPERATIVE AND POSTOPERATIVE COMPLICATIONS IN PRESENT SERIES

Case no.	*Complication*
4	Reactionary haemorrhage
4	Aggravation of aortic incompetence (subvalvular)
5	Ventricular fibrillation
7, 9, 11 and 12	Post-pericardiotomy syndrome
8	Cerebral oedema
10	Mild wound disruption
11	Aggravation of aortic incompetence
7	Mild aortic incompetence induced

Case 8 became comatose with elevated blood pressure 36 hours postoperatively, and a lumbar puncture showed a pressure of 300 mm. H_2O, relieved by slow decompression.

2. Immediate acidosis is usually caused by inadequate perfusion with a low mean pressure. Delayed acidosis is usually caused by inadequate postoperative ventilation following pain, pleural haemorrhage or bronchial secretions.

3. Reactionary haemorrhage, of sufficient degree to necessitate thoracotomy, has not occurred in our pump cases. The patient, case 4, operated on under hypothermia, was re-opened while being re-warmed.

4. Pulmonary complications are much less common now that we employ the sternal split, though case 8 had a left-lower-lobe atelectasis. The aetiology and prevention of chest complications has been fully described by Kolff and Effler.[43]

5. Thirst is a most distressing symptom, both for patient and attending staff. It is caused by hyperosmolarity of the blood and is not assuaged by drinking fluids. Fluid control has been very carefully supervised. This has been fully reviewed by Sturtz *et al.*[42] from the Mayo Clinic and du Plessis and Scott[42] from early experience with our patients.

6. Transient cardiac failure when arrest has been prolonged can occur, but, in our series, was only seen in case 7.

7. Prolonged postoperative pyrexia, as in the post-commissurotomy syndrome described by Papp and Zion,[44] occurred in cases 7, 9, 11 and 12.

8. Septicaemia from infection during by-pass has been described, but fortunately we have not yet encountered this dread complication.

9. Postoperative haematological changes after a few days are minimal — mild anaemia and leucocytosis. (In case 13 two transfusions were necessary 14 days postoperatively.)

B. *Normal postoperative complications* common to any major transthoracic procedure.

Postoperative Results

1. The systolic murmur decreased, but never vanished.

2. The pressure gradient was diminished in all, but not often abolished except in case 8.

3. Mild aortic incompetence. We produced this in case 7 and aggravated a pre-existent aortic incompetence in case 11.

4. All patients seen postoperatively were relieved of their symptoms. Case 7, aged 5, with disabling angina, has been relieved completely and is very active and well.

These operations of necessity required team-work, and we should like to thank our many medical colleagues, the technicians, and the nursing staff, who all formed part of the team. Also, we thank those colleagues who referred the patients to us for operation.

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