Cardiac surgery in early infancy

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IMPORTANT advances have been made in the diagnosis and treatment of congenital heart disease (CHD) in early infancy. Accurate diagnosis is now possible in the first days or even hours of life. Operation is also possible at this early age and very often it provides the only chance for survival. The risk of conservative treatment often exceeds the risk of the operation. An aggressive approach to the diagnosis and treatment of CHD is dictated by statistics. The estimated incidence of CHD is 6-8/1000 live births. About 50% of the children born with CHD die before their first birthday unless effectively treated. The achievements in the treatment of CHD in infancy are the result of the combined efforts of paediatricians, cardiologists, surgeons, anaesthetists and nurses. Many other specialists and technical staff contribute to the diagnosis and treatment.

The experience of the Thoracic Unit, Hospital for Sick Children at Great Ormond Street, forms the basis of this report (unless otherwise stated). Table 1 shows the overall experience with surgical procedures in infancy and it also indicates the proportion of patients requiring surgery early in life (under the age of 12 months). Although palliative procedures were for a long time preferred in small babies, the total number of corrective procedures under the age of 1 year increases progressively. Figure 1 shows the overall experience with by-pass surgery under the age of 1 year. It demonstrates the improving chances of infants submitted to open-heart surgery in recent years.

TABLE 1. Surgical experience 1946 to August 1971

Diagnosis	Total	Under 1 year
Patent ductus arteriosus	1037	259 (25%)
Systemic-pulmonary shunts	739	283 (38%)
Coarctation of aorta	445	238 (53%)
Pulmonary artery constriction	292	244 (83%)
Transposition of the great arteries		
Atrial septostomy	217	164 (75%)
Mustard operation	177	36 (21%)
Pulmonary stenosis	122	32 (26%)
Total anomalous pulmonary venous drainage	78	57 (73%)
Vascular rings	47	40 (85%)
Total	3154	1353 (43%)

FIG. 1. Open-heart surgery under 1 year of age at the Great Ormond Street Thoracic Unit, February 1963 to September 1971 (115 patients). Open columns, survived; closed columns, died.

Diagnosis

Clinical diagnosis of congenital heart anomalies is not always easy. This is because several anomalies may be present in one patient and the interpretation of physical signs may be difficult. However, cardiac catheterization and angiocardiography can differentiate various defects with great accuracy. Only precise diagnosis can prevent serious errors in indications and operative treatment. No infant should be considered too ill for cardiac catheterization and angiocardiography. We feel that there is no longer a place for exploratory thoracotomies to compensate for inadequate investigation. Furthermore, no infant should be considered too ill for an operation, because very often this is the only chance for survival to a desperately ill baby. As any delay in investigation or operation may prove fatal, teams of investigators and surgeons should be available on a 24-hr basis. To wait until the child demonstrates his will to live is no longer an acceptable concept. This, we feel, merely gives an excuse for not providing adequate diagnostic and surgical facilities.

Indications

Cardiac operations in infancy are performed for two main reasons—severe hypoxemia and/or severe congestive heart failure. The most severe lesions

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present in the first month of life. Because most of the deaths occur very early, operation should be attempted as soon as possible after the diagnosis has been made. It may be a palliative procedure for lesions where correction in early infancy carries too high a risk. Corrective operation may be as simple as ligation of patent ductus arteriosus or it may be an open-heart procedure requiring the heart-lung apparatus (total anomalous pulmonary venous drainage, transposition of the great arteries, etc.). The decision between palliative and corrective procedures is not always an easy one. All factors should be carefully considered. These factors include operative risk of correction early in infancy compared with the combined risk of palliative procedures and correction a few years later; chances of achieving a complete and lasting repair on a very small heart against progressive and possibly permanent damage caused by an uncorrected lesion (pulmonary vascular disease in ventricular septal defect or hemiplegia in cyanotic heart lesions). Other factors include the emotional strain which a staged operation presents for the parents, and last but not least, the experience and ability of the team of surgeons, physicians, anaesthetists and nurses is a very important factor.

Operation

Some of the more frequently used procedures will be described later. The success of the operation does not depend only on surgical skills, but supervision of acid-base balance, monitoring of blood pressure, temperature and fluid output and intake often determine the final outcome.

Postoperative care

In our view, postoperative care is an integral part of the treatment of CHD. It is as important as a precise pre-operative diagnosis and as the operation itself. The importance of good postoperative care should never be underestimated. Details cannot be discussed in this paper: the following remarks are only intended to outline the programme and to point out some of the more important features.

(a) Fluid balance

The right amount of fluid to be given in the post-operative period is still being discussed. Because of the 'physiological' postoperative retention, both water and sodium should be restricted for the first few days after the operation. At present we give 5% glucose solution with no added sodium for the first 48 hr. The volume of fluids is usually restricted to about half of the patient's normal daily requirement (calculated on the basis of age and weight). The calculated amount is very strictly adhered to and all additional sources of intake or output should be

considered (loss from gastric aspiration, excessive sweating or gains from flushing the intravenous or intra-arterial catheters). The accuracy of fluid intake is checked regularly by estimating the haematocrit and both plasma and urine osmolality. An inadequate fluid intake can cause thickening of bronchial secretions with subsequent atelectasis, and increased blood viscosity (high haematocrit) presents extra work for the circulatory system. Excessive fluid intake may cause fatal pulmonary oedema. Oral feeding is probably the best prevention of water and electrolyte imbalance. It can often be started 8–12 hr after the operation.

(b) Acid-base balance

Both respiratory and metabolic disturbances of acid-base balance are common. Cyanotic babies may present with severe metabolic acidosis, while respiratory acidosis is seen mainly in infants with a large left-to-right shunt. Metabolic alkalosis, which often follows open-heart procedures in small infants, is not yet fully understood. Respiratory alkalosis can develop in patients on intermittent positive pressure breathing if the PCO_2 is not regularly checked. Early diagnosis is the first step towards effective treatment. Samples of arterial blood for blood gas analysis (pH, PCO_2 , standard bicarbonate) can be obtained by direct puncture of the peripheral arteries, but an indwelling arterial catheter gives a better opportunity to obtain frequent samples and arterial pressure can be continuously monitored at the same time.

Metabolic acidosis is treated with sodium bicarbonate or THAM, but unless adequate peripheral perfusion is established by surgical intervention, these measures are often of only temporary value. Respiratory acidosis is treated with intermittent positive pressure ventilation by a nasotracheal tube or tracheostomy. Volume-controlled respirators are preferred.

(c) Postoperative heart failure

Heart failure may persist from before the operation or it may develop afterwards. Digoxin is given in the usual dose, that is 0.08 mg/kg body weight in a 2–4 kg baby, reducing the dose to 0.04 mg/kg body weight at 10–15 kg. If an immediate inotropic effect is required, catecholamines (isoprenaline or adrenaline) are used in a slow intravenous drip. The smallest effective dose is used to ensure an inotropic and peripheral vasodilating effect. We do not exceed the dose of 1 $\mu\text{g/kg}$ body weight/min. Diuretics (frusemide, chlorothiazide, spironolactone), adequate sedation (morphine 0.02 mg/kg body weight), decompression of the stomach with a nasogastric tube, oxygen and antibiotics are also important in the treatment of postoperative heart failure.

(d) Heat control

Infants with congenital heart lesions tolerate post-operative heat loss poorly. Every effort should be made to maintain a steady environmental temperature. Frequent handling of the patient, together with the number of drains, intravenous and monitoring lines, and connections to the ventilator make the use of an incubator less practical. New infra-red radiant warmers with temperature monitors may greatly improve the temperature control.

(e) Postoperative respiratory care

Pulmonary complications are the commonest cause of death after cardiac operations in infants. Prevention consists of adequate humidification, regular physiotherapy and removing the secretions by nasotracheal or nasopharyngeal suction. If these measures are not adequate, artificial ventilation is required. We prefer a volume-controlled respirator such as the Engstrom. The patient is attached to the respirator with a nasotracheal tube or tracheostomy. We ventilate all infants after open-heart procedures at least for 24–48 hr, and a nasotracheal tube is usually adequate for this. If the secretions become very thick or if ventilation is required for more than 4–5 days, tracheostomy is performed. Elective tracheostomy is used in ill infants with a severe lesion mainly in the first weeks of life, and in patients with abnormal or damaged lungs (pulmonary vascular disease, pulmonary oedema, severe infection). Control of artificial ventilation is achieved by frequent blood gas analysis. This is, in our experience, the only way to avoid serious under or over ventilation. Regular sedation (morphine, diazepam, chloral hydrate) are often used; muscle relaxants are not necessary in this age-group and we do not use them.

(f) General nursing care

This is one of the most important factors influencing the success or failure of the whole process of treatment. Observations made by an experienced nurse are often more valuable than complicated electrical monitoring. Skin care, feeding, positioning of the baby, administration of drugs, physiotherapy, recording vital signs, are only a few examples of nursing care. It is impossible to go into the details of nursing care but the importance of it must not be underestimated.

It is not possible to give details of diagnosis and surgical treatment of all congenital heart defects in early infancy. Only the more common lesions will be discussed.

Patent ductus arteriosus (PDA)**Indications**

Operation is performed during the first year of life in all patients who remain in congestive heart

failure despite full medical treatment. This happens not infrequently. From a total of 1037 children operated for ductus arteriosus in the Thoracic Unit, 259 (25%) were under the age of 1 year (Table 1).

Operation

The ductus can be ligated or divided. We prefer ligation with several ligatures of heavy plaited silk.

Results

Ligation of PDA is a simple operation in older children with an operative risk of 1.5%, but it carries a considerably higher risk in infancy (21.5%, Stark, 1971). The reason for the high mortality is not age, but the fact that most severe forms, and patients with additional cardiac lesions, present very early in life. Of our patients dying after PDA ligation in the first year of life 91% had additional cardiac lesions.

Aortopulmonary septal defect (window)**Indications**

Patients with this rare malformation present in heart failure in infancy. Operation is indicated to treat the heart failure and to prevent the development of pulmonary vascular disease.

Operation

Various techniques have been described and used. We prefer patch closure of the defect through an aortic incision on cardiopulmonary bypass (Deverall *et al.*, 1969).

Results

Among a total of fifteen patients operated on, six were infants. Five survived the operation and are well.

Ventricular septal defect (VSD)**Indications**

Heart failure which is not adequately controlled by medical therapy is the only indication. Considerable differences in the approach to patients with VSD still exist. These differences are due to several factors: only a small proportion of VSDs ever present in heart failure; spontaneous closure can occur in as many as 50% of patients (Hoffman & Rudolph, 1965). Palliation (pulmonary artery banding) or corrective operation are available.

Operation

Pulmonary artery constriction (banding) produces an increased resistance to pulmonary blood flow and diminishes a left-to-right shunt. It may also prevent the development of pulmonary vascular disease. Closure of VSD can be accomplished either by direct

suture or with a Dacron or Teflon patch. Cardio-pulmonary by-pass or surface-induced hypothermia are used for corrective operations.

Results

Pulmonary artery banding for isolated VSD carries a risk of about 7%, while for patients with VSD and a large PDA the risk is 45% (Stark *et al.*, 1971). Early results of VSD closure, especially in the presence of pulmonary hypertension were discouraging (Hallman, Cooley & Bloodwell, 1966, 42% mortality), but in recent reports Barratt-Boyes, Simpson & Neutze, 1971, reported no deaths in six corrected patients; Ching *et al.*, 1971 (four deaths in eighteen patients operated under the age of 1 year) and our own results (four patients surviving the operation in the first year of life) show an acceptable risk. However, these series are still too small. At present we consider each case individually. Infants with multiple VSDs and patients with complex lesions have the pulmonary artery banded, while early correction is reserved for patients with a single VSD.

Coarctation of the aorta

Indications

Operation is performed to treat heart failure. Medical therapy is always tried but surgery should not be delayed much beyond 24–48 hr. Severe left ventricular hypertension is another indication because it may lead to myocardial ischaemia (Tawes *et al.*, 1969a). Many patients with coarctation of the aorta present in heart failure in the first weeks of life. Of a total of 445 operated children, 238 (53%) were younger than 1 year.

Operation

The coarcted segment is resected and an end-to-end anastomosis using 6–0 silk performed. Continuous suture is interrupted a few times anteriorly to allow for growth. In our series of 238 infants operated for coarctation of the aorta, a synthetic graft has never been required.

Results

The mortality rate of patients treated medically can be high (Mortenson *et al.*, 1959, 64%; Sinha *et al.*, 1969, 88%). At present 60% of infants survive the operation, while in the years 1953–62, survival rate was only 20% in our series. The significance of associated lesions is again obvious in this group. Of eighty-eight infants dying after the operation for coarctation of the aorta, 97% had additional cardiac lesions (Tawes *et al.*, 1969b). The long-term results are good. Persistence of hypertension is rare in patients treated in infancy. Re-stenosis at the site of the anastomosis occasionally occurs, but it does not

cause heart failure and a second operation can be performed with a low risk (one death in fifteen re-operated patients).

Total anomalous pulmonary venous drainage (TAPVD) Indications

Patients often present in severe heart failure and deterioration can be very rapid. The risks are higher if the pulmonary venous return is obstructed. Five of our patients died while awaiting surgery, therefore operation should be performed as soon as the diagnosis is established. Our present policy is to investigate and operate on patients with TAPVD within 24 hr of admission. In the absence of heart failure and pulmonary hypertension, operation can be delayed until the age of 3–5 years.

Operation

Operative technique depends on the anatomy of the lesion. If the veins enter the right atrium directly or via the coronary sinus a patch of pericardium or Dacron directs the blood to the left atrium through the atrial septal defect. In supra- or infradiaphragmatic drainage, anastomosis between the horizontal limb of the common pulmonary vein and the posterior aspect of the left atrium is made. The communicating vein may be ligated and the atrial septal defect closed or this may be left for a second stage operation. Partial cardiopulmonary by-pass is used in a two-stage procedure (Mustard, Keith & Trusler, 1962; Behrendt *et al.*, 1972). Total by-pass under normal temperature (Gersony *et al.*, 1971) or deep hypothermia (Barratt-Boyes *et al.*, 1971) are used for one stage correction.

Results

The risk is very high in the first weeks of life. Gomez *et al.* (1970) reported a 46% survival rate among fifteen operated infants. Gersony *et al.* (1971) had seven survivors in a group of ten infants younger than 4 months. In our series of fifty-three infants operated under the age of 1 year, twenty survived the operation (37%). Long-term results are good—only occasionally does the anastomosis shrink a few weeks after the operation.

Fallot's tetralogy

Indications

Patients with Fallot's tetralogy usually do not present in the first days of life as patients with transposition of the great arteries. However, a number of patients develop cyanotic or anoxic spells in the early months of life. A shunt operation has been preferred by most centres in this situation. Some use trans-ventricular valvotomy with an infundibular resection and recently total correction has been suggested and successfully performed in very young infants.

Operation

Shunt operations. A Blalock-Taussig operation (subclavian artery to pulmonary artery shunt) preferably on the side of an innominate artery, can be performed even in neonates but both the early and the late occlusion rate is high (Bonham-Carter, 1971). A Waterston shunt (ascending aorta to right pulmonary artery) is therefore preferred by most surgeons in infants. Shunts to the descending aorta or Dacron grafts are rarely used in Fallot's tetralogy.

Total correction. This consists of pulmonary valvotomy, resection of hypertrophied muscular bands and fibrous tissue from the infundibulum and closure of the ventricular septal defect. If the pulmonary valve ring is small, an outflow tract patch or homograft valve can be inserted. Correction is done on cardiopulmonary by-pass with or without mild hypothermia or using deep hypothermia and circulatory arrest. Although successful corrections have been achieved in infancy, most centres prefer to do total correction between the ages of 2 and 5 years. Early correction is mostly reserved for anatomically favourable cases with a localized infundibular obstruction, while patients with a severely hypoplastic infundibulum are usually corrected later.

Results

Even a palliative procedure—shunt operation—carries a considerable risk in early infancy. In our series of 125 shunt operations under the age of 1 year, ninety-two survived the operation (74%). The risk was highest in the first 6 weeks of life (40%), while after the age of 1 year we have lost only nine of 286 patients (3%). Bernhard *et al.* (1971) reported a 21% mortality rate in forty-seven infants and Pickering *et al.* (1971), 50% mortality in a group of seventeen infants under 6 months of age.

The survival rate after total correction in older children has much improved in recent years. Mortality under 3% has been reported by Kirklin & Karp (1970), Hawe *et al.* (1969), Vathayanon *et al.* (1968) and others. The indications for and results of corrective surgery in infancy have yet to be determined. Recently Barratt-Boyes *et al.* (1971) corrected nine patients under 19 months of age without a death.

The late results after successful correction are good. Residual infundibular obstruction, pulmonary valve incompetence, heart block and residual ventricular septal defects are the possible complications.

Transposition of the great arteries (TGA)

Indications

The natural history of patients with TGA shows a very high mortality in the first weeks and months of life. Survival beyond a few months of age is unlikely unless an effective communication between the

systemic and pulmonary circulations is present or established. Palliative treatment is therefore required as soon as the diagnosis is made. Total correction can be performed later but the delay should not be too long. This is because increasing cyanosis and polycythaemia are associated with appreciable morbidity and mortality. We now prefer to perform total correction between the age of 6 and 12 months.

Operation

Balloon septostomy (Rashkind & Miller, 1966) gives a better result (Tynan, 1971) than surgical septostomy (Deverall *et al.*, 1969) and it also makes the subsequent corrective procedure easier. Balloon septostomy is performed during cardiac catheterization by passing a special catheter through the foramen ovale to the left atrium. The balloon at the tip of the catheter is then inflated and pulled back to the right atrium. The septum is torn and equilibration of atrial pressures with increased atrial shunting is achieved. Various techniques for surgical atrial septectomy were described and used, but now we prefer balloon septostomy.

Total correction (Mustard, 1964) is an 'inflow correction'. This is a physiological correction redirecting the systemic venous blood to the mitral valve, left ventricle and pulmonary artery and pulmonary venous blood to the tricuspid valve, right ventricle and aorta. Pericardium or Dacron is used for the separation of the two 'atria'.

Results

Danielson *et al.* (1971) and Aberdeen (1971) reported a 12% hospital mortality. Recently more patients are being corrected in infancy. We have had twenty-nine survivors in a group of thirty-one infants with TGA plus ASD corrected under the age of 1 year (93% survival rate).

In the late results, arrhythmias, SVC and pulmonary venous obstruction, defects in the patch and tricuspid incompetence are the possible complications. In their absence children lead a normal life.

Transposition of the great arteries complicated by additional lesions

TGA + PDA

Soon after birth PDA presents a useful source for blood mixing but pulmonary vascular disease can develop early. This is why it has been our policy to ligate the PDA as soon as an adequate atrial septal defect is established.

TGA + VSD

Because pulmonary vascular disease usually develops early in patients with TGA plus VSD, pulmonary artery banding should be performed before

the age of 1 year (Stark *et al.*, 1970). Mustard's procedure and closure of the VSD is possible but in the first year of life this procedure presents a higher risk.

TGA + VSD + PS

Patients with this combination of lesions may require a shunt operation (Blalock-Taussig or Waterston) in infancy. Anatomical correction using a homograft valve (Rastelli, McGoon & Wallace, 1969) is usually delayed until the age of 5–7 years. Improvement after a shunt operation is often striking and lasts for a number of years.

Tricuspid atresia (TA)

Indications

A shunt operation is indicated in cases of severe cyanosis while pulmonary artery banding may help in the presence of a large ventricular septal defect and congestive heart failure.

Operation

Corrective surgery is not available for these patients but palliation offers good improvement and many patients live a reasonable life until their 20s or 30s. We prefer an arterial shunt (Waterston or Blalock-Taussig) in infants. The risk of Glenn operation (superior vena cava to right pulmonary artery anastomosis) is too high under the age of 6 months. If the atrial septal defect is too small, balloon septostomy or surgical septectomy may be performed.

Results

The improvement after a shunt operation may last several years. Later a second shunt may be required. The operative risk is higher in younger infants. We have lost sixteen of thirty-four infants operated under the age of 6 months. Hunt *et al.* (1970) reported ten deaths in a group of eighteen infants under the age of 6 months.

Pulmonary stenosis with intact ventricular septum (PS)

Indications

Severe hypoxemia is an indication for emergency treatment. Cardiac catheterization and angiocardiology is the basis for surgical intervention. If the child is not acutely ill operation is deferred until the age of 3–6 years.

Operation

This depends on the anatomical findings. Patients with small but thick-walled right ventricles are best treated with a shunt procedure or a combination of valvotomy and a shunt. In patients with an adequate right ventricular cavity, valvotomy alone may suffice. Closed transventricular valvotomy or transarterial

valvotomy using inflow occlusion at normal temperature, or under hypothermia or deep hypothermia with circulatory arrest are being used. A large right-to-left shunt across an ASD or patent foramen ovale may be present, therefore closure of this communication may be required.

Results

Excellent results were achieved by Mustard, Jain & Trusler (1968), with only one death in a series of twenty-six patients. Anderson & Nouri-Moghaddam (1969) reported 38% mortality under the age of 2 years (eight of twenty-one). In our series, only five of seventeen infants with severe pulmonary stenosis survived the operation.

Pulmonary atresia with intact ventricular septum

Indications

This is a very severe lesion which often requires emergency surgery in early infancy. Severe hypoxemia is the indication for surgery.

Operation

Systemic to pulmonary artery shunt, transventricular opening of the atretic valve or a combination of both can be used. Total correction using a homograft valve is possible, but this is usually deferred until the age of 6–8 years.

Results

Hypoplasia of the right ventricle is the most important factor influencing survival. Operative risk is higher than in patients with pulmonary stenosis. (Gersony *et al.* reported in 1967 thirteen deaths in fifteen patients (86%), Bowman *et al.*, 1971, eight in twelve (66%) and Murphy *et al.*, 1971, twelve in twenty operated infants (60%).)

Congenital aortic stenosis (AS)

Indications

Heart failure is an indication for operation in infancy. The gradient from the left ventricle to the aorta may be lower than in older children or adults because of poor cardiac output in a very ill infant.

Operation

The aortic valve has to be visualized and commissures opened. Postoperative valve incompetence is a very serious and often fatal complication, therefore there is no place for a blind procedure. Cardiopulmonary by-pass, circulatory arrest under deep hypothermia or inflow occlusion in a hyperbaric chamber have been used. Patients with a subvalvular stenosis require resection of the obstructing membrane. Great care must be taken not to injure the conduction mechanism. The supra-aortic type of stenosis is relieved with a Dacron patch.

Results

Most of the operations have to be done on an emergency basis with a high operative risk. Coran & Bernhard (1969) reported a 50% mortality rate under the age of 2 years (eleven deaths in twenty-one patients). Recent changes in by-pass technique may improve the results but the numbers of reported cases are small for statistical analysis. Endocardial fibroelastosis or a severely malformed aortic valve have a bad prognosis. The abnormal aortic valve, especially if bicuspid, may calcify and valve replacement may be required later.

Vascular rings

Indications

Operation is performed in infancy to relieve stridor and prevent repeated upper respiratory infections. Severe feeding difficulties are rarely present.

Operation

Diagnosis is best made by barium swallow. The operative technique is determined by the type of anomaly. Patent ductus arteriosus or ligamentum arteriosum are always divided and the fibrous bands between the trachea and oesophagus dissected (Lincoln *et al.*, 1969).

Results

Relief from stridor is rarely immediate. It often takes a few weeks and in some patients the post-operative period may be difficult. Operative risk can be low—we have had thirty-eight survivors among forty-one operated infants.

Rare lesions and lesions presenting rarely in the first year of life

Atrial septal defect—secundum type (ASD)

Very few patients with ASD present early in life. If so, heart failure from a large left-to-right shunt dominates the clinical picture. ASD can be closed on cardiopulmonary by-pass or under circulatory arrest in deep hypothermia.

Endocardial cushion defects

Atrial septal defect—primum type. This relatively uncommon defect may present in infancy. Mitral valve incompetence is often present. Heart failure from a left-to-right shunt and mitral incompetence may require an early operation. Cardiopulmonary by-pass or deep hypothermia can be used. The cleft in the mitral valve is repaired and the atrial septal defect closed with a patch.

Common atrio-ventricular canal. Severe heart failure with episodes of pulmonary oedema is a

common presentation of this rare malformation. When the shunt is predominantly at ventricular level, pulmonary artery banding is a useful palliative procedure. Eleven of our fifteen patients survived pulmonary artery banding. Complete repair can be thus delayed until later. Immediate complete repair gives the only chance for survival in infants with a shunt predominantly at atrial level and with a severe mitral incompetence. Long-term results depend on the degree of residual mitral incompetence. Arrhythmias present another serious complication.

Truncus arteriosus

Total correction is now available at a later age (Rastelli, Ongley & McGoon, 1969). Because pulmonary vascular disease develops early, pulmonary arteries should be banded in infancy. The risk of banding is high but the risk of total correction in patients surviving without pulmonary vascular disease is very acceptable. (McGoon, Rastelli & Wallace, 1970, reported twenty-one survivors of twenty-eight patients.)

Double outlet right ventricle (DORV)

Pulmonary artery banding is indicated for patients in heart failure. If pulmonary stenosis is present a systemic to pulmonary artery shunt is performed and total correction usually delayed until after the age of 5–7 years.

Other rare malformations, such as Ebstein's anomaly, congenital mitral stenosis, congenital mitral incompetence, triatrial heart, anomalous origin of a coronary artery, tumours of the heart and others, will not be discussed in this paper.

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