

The surgical relief of transposition of the great vessels in infancy

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The necessity for palliative surgery in transposition of the great vessels is indicated and the basic haemodynamics of the condition are outlined. The clinical picture of infants with transposition is divided into three types, and a method of surgical treatment is suggested for the two most common types. Our technique of investigation and treatment is described and the reasons for our choice of therapy are discussed. The results of cases operated upon are presented.

To the cardiac surgeon embarking upon the treatment of infants with congenital heart disease, transposition of the great vessels is the lesion which assumes pride of place. It is the most common cause of death from heart disease in infancy, and without surgery only 14% survive to the age of 6 months (Keith, Neill, Vlad, Rowe, and Chute, 1953). Yet this condition is rarely associated with extracardiac anomalies, and since the development of the Mustard procedure (Mustard, Keith, Trusler, Fowler, and Kidd, 1964), complete correction can be carried out with a low operative mortality and a good chance of permanent cure.

Complete transposition of the great vessels, as defined by Edwards (Fontana and Edwards, 1962), embodies a fundamental deviation from the normal pattern of haemodynamics (Fig. 1).

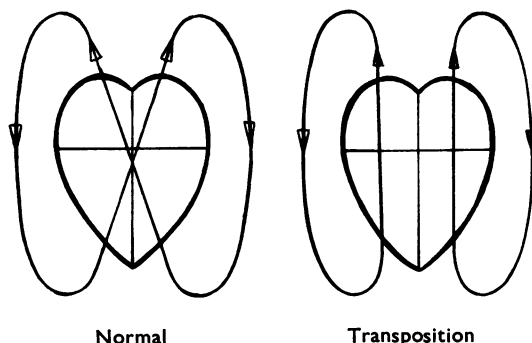


FIG. 1. *Haemodynamics.*

Systemic venous blood can reach the lungs only via extra- or intra-cardiac communications between the systemic and pulmonary circulations. Moreover, any flow of blood from the systemic to the pulmonary circulation must be balanced by an equal flow in the opposite direction because of the independence of the two circulations (Shaher, 1963). The adequacy of mixing between these two circulations determines the level of arterial oxygen saturation in any particular case. Therefore patients with good communications have the best arterial oxygen saturations and should have the best prognosis. Unfortunately, a very good communication with an excessive pulmonary blood flow often reduces the possibility of a favourable outcome. Because of the natural post-natal fall in pulmonary vascular resistance, the flow through the lungs progressively increases until pulmonary engorgement embarrasses normal lung function, and pulmonary infection and cardiac failure supervene.

Clinically, infants with this condition are cyanosed from birth and present in three ways. First, those with inadequate communications generally present in cardiac failure within the first few weeks of life when the ductus closes. In this large group of infants, mixing can be improved appreciably by the creation of an atrial septal defect.

Secondly, patients with adequate communications develop pulmonary engorgement and congestive cardiac failure within a few months of birth. The lungs may be protected in these cases by banding the pulmonary artery and creating an artificial pulmonary stenosis. This creates a favourable situation since a combination of a

ventricular septal defect and pulmonary stenosis confers the best protection on patients with transposition of the great vessels.

Thirdly, a small group of patients with complex malformations, often including interruption of the aortic arch, present with severe cardiac failure within a few days of birth and constitute a formidable and largely unsolved problem.

The onset of cardiac failure is usually followed by rapid deterioration, so that urgent investigation and treatment are needed if these children are to survive. Digitalis, diuretics, and oxygen are often ineffective, and it is important to establish an accurate diagnosis as a prelude to appropriate urgent surgery. It is our policy to investigate all infants thought to have transposition of the great vessels as the clinical findings can be misleading, especially in the neonatal period. Accurate assessment of the anatomical defects is of great assistance in choosing the correct surgical approach, and in our hands these investigations carry a very low risk.

THE TECHNIQUE OF INVESTIGATION

Investigation is undertaken in all infants in whom this diagnosis is suspected. Occasionally, the diagnosis may be difficult or misleading. These infants are fragile and often in poor clinical condition and will not tolerate extensive intracardiac manipulation (Grainger, Taylor, and Verel, 1966). The investigation, therefore, is limited and a carefully planned study is safe and rewarding in our experience. Angiocardiography provides sufficient anatomical detail and forms the major part of the study. We prefer a selective right ventriculogram in the antero-posterior and lateral positions using a large film serial changer. This shows the abnormally situated aorta, defects of the ventricular septum, the ductus arteriosus, and abnormalities of the aortic arch. Selective left ventriculography demonstrates pulmonary or sub-pulmonary stenosis. We do not prolong the procedure by attempting to enter the pulmonary artery. The arterial oxygen saturation is the best guide to the degree of mixing between the systemic and the pulmonary circulations: thus, if it is less than 65%, there is an inadequate communication and a shunt procedure is indicated. It is important to replace any blood loss, correct the acidosis, and prevent hypothermia by swathing the infant in gamgee and warming the theatre (Gotsman, 1967; Gotsman, Beck, Klein, O'Donovan, Barnard, and Schrire, 1967). Once the diagnosis has been established, the patient is transferred to the operating theatre with assisted manual ventilation if required. Residual acidosis is corrected by the intravenous administration of sodium bicarbonate, and a venous cannula is inserted via the long saphenous vein to lie in the inferior vena cava.

THE TECHNIQUE OF SURGERY

The operation is performed under hypothermia at 30° C., which is induced by surface cooling using a Thermo-Rite blanket. The chest is opened through an anterolateral thoracotomy in the fourth interspace with the shoulders positioned obliquely to ensure that the right atrium will be higher than the aorta (Fig. 2a). The incision is begun and rewarming is started when the rectal temperature reaches 33° C. The patient continues to cool during surgery and the lowest temperature of 30° C. is reached at the time of inflow occlusion.

After the chest has been entered, the lung is retracted posteriorly and the pericardium is split longitudinally, 3 mm. anterior to the phrenic nerve, in order to spare the pericardium for the subsequent second-stage correction. No. 2 silk snares are passed around the superior and inferior venae cavae and led through lengths of rubber catheter (Fig. 2b). A small atrial clamp is applied to the right atrium obliquely over the site of the foramen ovale. The atrium is then opened distal to this clamp and stay sutures are inserted in the lips of this incision to facilitate re-application of the clamp (Fig. 2c). The caval snares are now snugged down and after seven or eight beats the atrium empties, the atrial clamp is removed, and respiration is stopped. If the atrium has been allowed to empty adequately, little or no suction is required to visualize the atrial septum, which is then lifted with fine-toothed forceps to bring the foramen ovale into view, which emits small jets of bright red blood with each cardiac contraction. The limbus is now grasped (Fig. 2d) with the fine-toothed forceps and is divided with scissors for some 5 mm. in a cephalad direction to enlarge the opening (Fig. 2e). The valve of the foramen ovale is excised (Fig. 2f) and the atrium is filled with blood by re-expanding the lungs, with the lips of the incision elevated by stay-sutures. The inferior vena caval snare is released to flush out air trapped near the tricuspid valve, and the atrial clamp is reapplied (Fig. 2g). The superior vena caval snare is released and respiration is started again.

Within a few beats the colour of the myocardium improves greatly and the heart beat exhibits the benefit provided by the improved atrial mixing. The atriotomy is closed by a running 4-0 silk suture and the caval snares are removed (Fig. 2h). If the patient has a ventricular septal defect, the pulmonary artery is now banded to protect the lungs from a torrential pulmonary blood flow and increased pulmonary arterial pressure. Umbilical tape is passed around the pulmonary artery and a needle is inserted into the artery distal to this point to record the intraluminal pressure. The tape is now tightened using curved artery forceps, and the tightening is increased repeatedly until the pressure is reduced by half and remains stable at this level for 5 minutes. The tape is now fixed in this position by silk sutures and, after repeating the pressure measurements, the pericardium is loosely approximated with interrupted silk

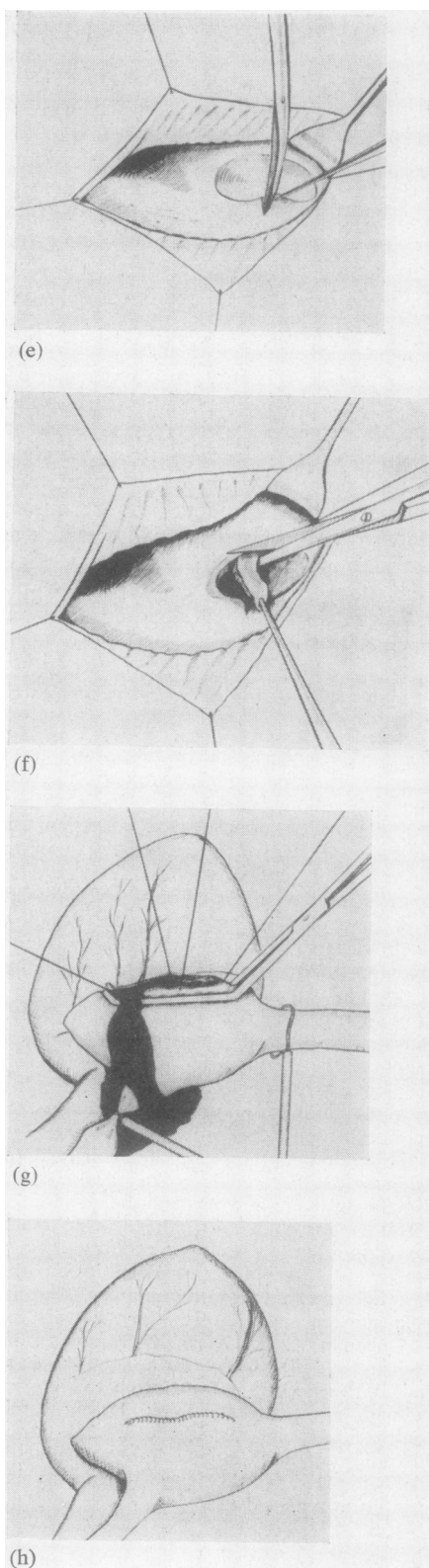
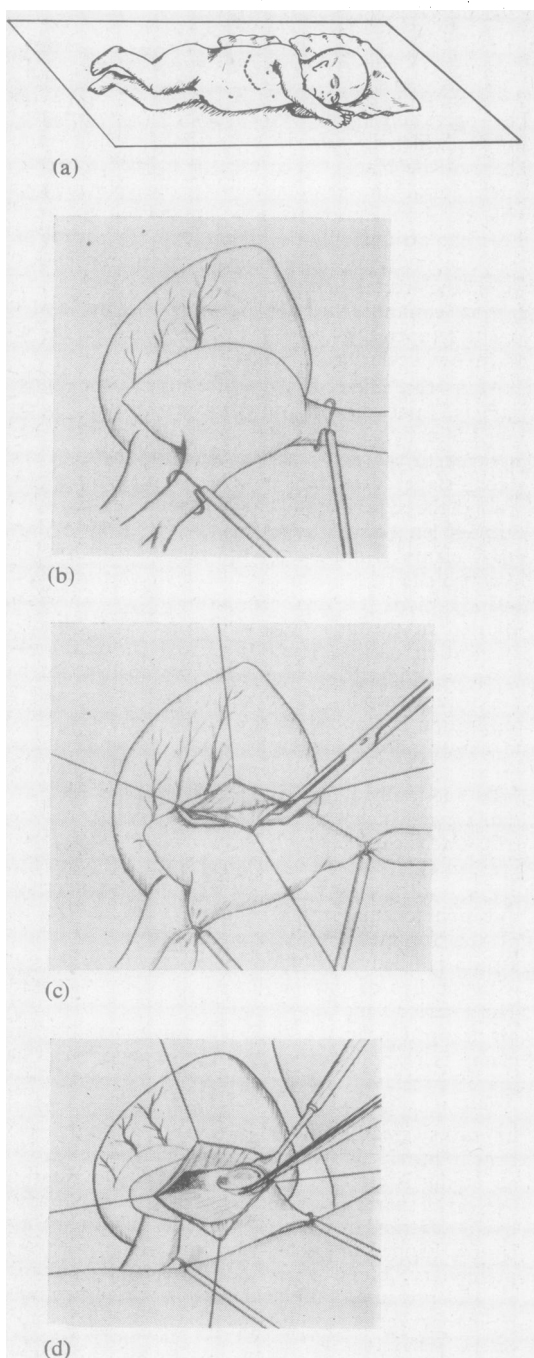


FIG. 2. Details of operative technique. (a) Position on operating table. (b) Caval snares in position. (c) Atrium opened, stay sutures in position, cavae occluded. (d) Clamp removed, limbus grasped. (e) Limbus incised. (f) Valve excised. (g) Atrium filled, inferior vena cava snare released, clamp reapplied. (h) Atrium closed.

sutures and the chest is closed after inserting one intercostal drainage tube.

POST-OPERATIVE CARE

Post-operatively the patient receives assisted ventilation via a Portex endotracheal tube and a Bird Mark 10 respirator, which pumps pure oxygen through a warm air humidifier. Intermittent positive pressure respiration is used for at least 24 hours post-operatively and for longer if the patient has pneumonia.

Digitalis therapy is continued and isoprenaline hydrochloride (1:800,000) is given intravenously to maintain a good cardiac output, peripheral circulation, and urine output. Fluid requirements are calculated on the basis of 120 ml./kg. on the first day, 135 ml./kg. on the second day, and 150 ml./kg. daily thereafter. Intravenous fluid only is given for the first day, and this is usually supplemented by oral feeds of half-strength S26 and 20 ml./kg. plasma on the second day. The intravenous fluid consists of half-strength Darrow's solution in 7.5% dextrose to promote diuresis. Antibiotic cover is used in all cases.

RESULTS

The results are shown in the Table.

TABLE

Patient	Age	Operation	Result
P.H. ..	3 weeks	Blalock-Hanlon	Good
S.v.W. ..	12 weeks	A.S.D. under inflow occlusion	Good
	14 months	Subsequent banding of P.A.	Died
J.v.T. ..	4 weeks	A.S.D. under inflow occlusion	Good
B.R. ..	1 week	A.S.D. under inflow occlusion	Good
F.v.Z. ..	6 weeks	A.S.D. under inflow occlusion	Good
T.L. ..	12 weeks	A.S.D. under inflow occlusion and banding of P.A.	Good
E.w. ..	4 days	A.S.D. under inflow occlusion	Good
P.P. ..	13 weeks	A.S.D. under inflow occlusion	Good

DISCUSSION

Several different surgical procedures have been used to create a large atrial communication. The Blalock-Hanlon technique seems to carry a high operative risk in the first three months of life—at the time when the majority of these infants need palliative surgery. The major cause of mortality probably is produced by occlusion of the right pulmonary artery, and Hallman and Cooley (1966) modified the technique to occlude the superior division only and hence avoided this. Our first infant to undergo palliative surgery had a Blalock-Hanlon type procedure but the right pulmonary artery was not occluded. The child survived operation but developed consolidation of the right upper lobe post-operatively, presumably

because the atrial clamp obstructed the venous return from this lobe. Any trauma to the lungs of these infants is poorly tolerated, and following this experience we decided to use mild hypothermia and inflow stasis and to excise part of the atrial septum under direct vision (Gallaher, Fyler, and Lindesmith, 1966). Hanlon and Blalock (1948) felt that it 'involved loss of blood, was poorly tolerated, and necessitated hurry, the defects were of indeterminate size and their persistence problematical, and the technique did not allow of direct vision'. Lindesmith, Gallaher, Durnin, Meyer, and Jones (1966) and Barratt-Boyes (1965), however, have found this to be a reasonable alternative procedure, and we feel convinced that a large defect can be created under direct vision without hurry, the procedure is well tolerated, little blood is lost, and the defect persists.

Recently, Rashkind and Miller (1966) described a technique of atrial septostomy using a balloon catheter, and many enthusiastic reports suggest that this may replace open cardiomy (Jordan and McCarthy, 1967; Watson and Rashkind, 1967). The technique may not be successful in all infants and open cardiomy may still be required in some. We failed in two infants, who subsequently needed surgical creation of an atrial septal defect.

Banding of the pulmonary artery has not been popular in infants with transposition, but it is well known that children with an A.S.D. or V.S.D. of moderate size and mild sub-pulmonary obstruction have the best prognosis (Noonan, Nadas, Rudolph, and Harris, 1960). Infants with normally disposed great vessels and a large ventricular septal defect, who have a torrential pulmonary blood flow and repeated respiratory infections, often show a dramatic clinical improvement after pulmonary artery banding (Muller and Dammann, 1952; Albert, Fowler, Craighead, Glass, and Atik, 1961; Grainger, Nagle, Pawidapha, Robertson, Taylor, Thornton, Verel, and Zachary, 1967). Infants with transposition and a large pulmonary blood flow are also prone to repeated respiratory infections, and reactive and organic changes in the muscular pulmonary arterioles occur in children with a V.S.D. or A.S.D. (Ferencz, 1966). Banding of the pulmonary artery, therefore, is a logical procedure in children without sub-pulmonary stenosis (Cornell, Maxwell, Haller, and Sabiston, 1966). Creation of the A.S.D. alone may also flood the low-resistance pulmonary circulation, lower the systemic output, and predispose to respiratory infections, and some of these patients may also require pulmonary

artery banding. These infants have a stormy post-operative course and their subsequent childhood is punctuated by repeated episodes of respiratory infection. Our best result was obtained in the infant in whom the pulmonary artery was banded when the A.S.D. was created. Another infant had repeated respiratory infections after palliative surgery and improved greatly after subsequent banding. Two other children without a band have had some chest infections and one has since died.

Pulmonary arterial banding requires open thoracotomy and cannot, therefore, be undertaken at the same time as balloon catheter atrial septostomy. The following therapeutic approach seems logical. In patients without a significant ventricular septal defect balloon atrial septostomy is attempted at the time of cardiac catheterization. If the arterial oxygen saturation is increased by 10 or 15%, indicating that an effective communication has been created, then further procedures are not indicated unless the child has repeated respiratory infections. If the balloon septostomy fails to create an adequate communication, or the child has a moderate-sized ventricular septal defect, then thoracotomy is indicated to enlarge the atrial communication and to band the pulmonary artery at the same time. This will create an adequate communication between the two circuits and yet protect the pulmonary arteries from torrential pulmonary blood flow so that the child has a smoother post-operative course, fewer subsequent respiratory infections, and protected pulmonary vessels. Complete correction is not feasible in infants with pulmonary vascular obstruction as they are left with significant residual pulmonary arterial hypertension. Banding protects the muscular pulmonary arterioles and prevents these deleterious effects.

Two of our patients had pneumonia at the time of operation, but we do not regard this as a contra-indication to surgery, although it necessitates prolonged respirator therapy after operation.

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