Total anomalous pulmonary venous drainage in infancy¹

D. R. CLARKE,² J. STARK, M. DE LEVAL, J. R. PINCOTT,
AND J. F. N. TAYLOR

From the Thoracic Unit, Hospital for Sick Children, Great Ormond Street, London WC1

Between May 1971 and December 1975, 39 infants had operations for correction of total anomalous pulmonary venous drainage. Fourteen of the 39 patients were under 1 month of age at the time of operation. Twenty-four patients had supracardiac, 7 intracardiac, and 6 infracardiac total anomalous pulmonary venous drainage, and 2 had a mixed type. The overall hospital mortality was 36 per cent. There have been no late deaths. The improvement in survival rate in this series is attributed to: (1) earlier recognition and prompt referral, (2) an aggressive approach to diagnosis involving complete cardiac catheterisation and angiocardiology, (3) vigorous preoperative care, (4) early complete correction including construction of a large anastomosis and enlargement of the left atrium when indicated, and (5) intensive postoperative management paying particular attention to fluid balance and treatment of pulmonary complications. Operative mortality was highest in patients with total anomalous pulmonary venous drainage directly to the superior vena cava, and in those with infradiaphragmatic drainage of whom all had pulmonary venous obstruction. Mortality was not closely related to age, body weight, or severity of pulmonary hypertension.

Total anomalous pulmonary venous drainage was first described by Friedkowsky in 1868 (Brody, 1942). It was not, however, until 1942 when Brody reported 37 cases that it became a recognised clinical entity (Brody, 1942). The incidence of total anomalous pulmonary venous drainage is approximately 1.5 per cent of children born with congenital heart disease (Keith *et al.*, 1967).

The majority of patients with total anomalous pulmonary venous drainage have symptoms during the first year of life (Gathman and Nadas, 1970). The few who remain asymptomatic have a relatively good prognosis and operation can be performed electively later in childhood at low risk (Gomes *et al.*, 1970). Eighty per cent of symptomatic infants, if left untreated, will die before they reach their first birthday (Bonham-Carter *et al.*, 1969).

Medical treatment consisting of antifailure therapy may be beneficial, but in the presence of pulmonary venous obstruction and/or pulmonary hypertension, does not improve survival rate (Gathman and Nadas, 1970). Palliation by balloon

atrial septostomy has been tried (Miller *et al.*, 1967; El-Said *et al.*, 1972), but is helpful only in the few cases where the obstruction to pulmonary venous return is produced by a small foramen ovale.

The first attempt to correct total anomalous pulmonary venous drainage surgically was made by Muller in 1950, who anastomosed the left atrium to the common pulmonary venous channel (Muller, 1951). A semi-open atrial well technique was tried by Burroughs and Kirklin in 1954 (Burroughs and Kirklin, 1956). In 1955, Lewis performed a total correction using hypothermia with inflow occlusion which was successful in a 5-year-old patient (Lewis *et al.*, 1956). In 1957, Cooley performed a successful correction of total anomalous pulmonary venous drainage using cardiopulmonary bypass (Cooley and Ochsner, 1957).

In older children, total correction of anomalous pulmonary venous drainage has been performed successfully, with the use of cardiopulmonary bypass, for several years. Before 1970, however, mortality rates for infants less than 1 year of age remained high (Mustard *et al.*, 1962; Leachman *et al.*, 1969; Gomes *et al.*, 1970; Behrendt *et al.*, 1972; Wukasch *et al.*, 1975). With improved techniques for open cardiac repair in early infancy, the mortality rates for this group have fallen in the past five

¹This work was supported in part by a grant from the British Heart Foundation.

²Present address: Department of Surgery, University of Colorado Medical Center, 4200 East Ninth Avenue, Denver, Colorado 80220, U.S.A.

Received for publication 18 August 1976

Table 1 Total anomalous pulmonary venous drainage in 86 infants

Period	Number	Survived
1963-1970	47	15 (32%)
1971-1975	39	25 (64%)

years (Lemoine, 1975; Barratt-Boyes, 1973; Applebaum *et al.*, 1975; W. M. Gersony, F. O. Bowman, and J. R. Malm, 1975, personal communication). In this unit the operative mortality for total anomalous pulmonary venous drainage in infancy has decreased from 68 per cent in 1963 to 1970 to 36 per cent in 1971 to 1975 (Table 1).

Clinical diagnosis

Infants with total anomalous pulmonary venous drainage may present with signs of heart failure and/or cyanosis. The development of severe heart failure is often rapid: cyanosis is usually moderate on examination. Peripheral pulses are poor. Tachypnoea, tachycardia, gallop rhythm, and enlarged liver complete the picture of heart failure. On auscultation there are usually no murmurs. A closely split second sound may be a clue to the diagnosis.

The appearances on the chest x-ray film depend on the type of total anomalous pulmonary venous drainage and its physiological consequences. When there is no obstruction to pulmonary venous return, the heart tends to be large and the pulmonary vascularity is increased. If the pulmonary venous return is obstructed, the heart is usually not enlarged and the lungs are oedematous because of pulmonary venous hypertension; indeed, the combination of a heart which is not enlarged and pulmonary oedema is characteristic of obstructed total anomalous pulmonary venous drainage, though it also occurs in cor triatriatum. In younger infants, pulmonary oedema is localised to the perihilar regions, with septal lines often visible in the retro-sternal space or costophrenic angles. Pulmonary oedema must be distinguished from hyaline membrane disease where the changes are usually diffused throughout all lung areas. In later infancy when total anomalous pulmonary venous drainage is to the left innominate vein or to the right superior vena cava the diagnosis may be suspected from widening of the upper mediastinal shadow.

The electrocardiogram shows right axis deviation, and right ventricular hypertrophy is a constant feature; right atrial hypertrophy usually develops

during the first 3 months of life and is therefore uncommon in the neonatal period.

The presence of pulmonary oedema, enlarged liver, and a normal sized heart in a sick, slightly cyanosed baby is very suggestive of obstructed total anomalous pulmonary venous drainage. When the diagnosis is suspected, the infant should be transferred immediately to a specialised unit, where cardiac catheterisation and angiocardiology can be performed on an emergency basis. We believe that it is safer to investigate more infants suspected of having an obstructed total anomalous pulmonary venous drainage rather than to miss the diagnosis and lose the patient.

DEFINITIVE INVESTIGATION

The importance of complete cardiac catheterisation and of high quality selective angiocardiology cannot be overemphasised. It is important to outline the drainage from both lungs clearly so that the mixed type of total anomalous pulmonary venous drainage is not overlooked and the presence of pulmonary venous obstruction recognised. It is equally important to exclude additional cardiac malformations particularly persistent ductus arteriosus, ventricular septal defect, and coarctation of the aorta. If these are present, they should be corrected at the time of operation for redirection of pulmonary venous inflow.

PREOPERATIVE MANAGEMENT

Intensive supportive care is of utmost importance to neonates and small infants with total anomalous pulmonary venous drainage. We believe that all the techniques used in intensive care should be administered from the time the diagnosis is suspected, through transport, investigation, operation, and postoperative recovery, if circumstances warrant such measures. Insertion of arterial and central venous catheters is essential if arterial blood pressure, central venous pressure, and arterial blood gases are to be monitored. A central venous line is also useful for the administration of drugs, including sodium bicarbonate, calcium, and catecholamines, if required. A urinary catheter is inserted. The baby's temperature is controlled in an incubator or with an infrared warmer. All metabolic imbalances are corrected. Oxygen should be administered, and if hypercapnia or respiratory distress is present, the child should be intubated and ventilated. Though these measures are not always necessary, the use of some or all of these techniques in the preoperative period will allow transport and time for the adequate investigation of extremely ill infants.

Table 2 Clinical values in 39 infants, 1971 to 1975

	Range	Mean
Age at operation (d)	1-352	83
Body weight at operation (kg)	2.6-6.4	4.0
Arterial O ₂ saturation (%)	33-92	75
Pulmonary/systemic systolic pressure ratio	0.27-1.83	0.98
PV to RA mean gradient (mmHg) (17 patients)	0-34	11

Patients

Thirty-nine infants had operations for correction of total anomalous pulmonary venous drainage between May 1971 and December 1975. The age of the infants at the time of operation ranged from 1 day to 352 days (mean 83 days). The weight at the time of operation ranged from 2.6 kg to 6.4 kg (mean 4.00 kg). Twenty-six were boys and 13 girls. All infants were symptomatic and 31 had a pulmonary arterial systolic pressure which was

more than 60 per cent of the systemic arterial pressure. Pulmonary venous obstruction was difficult to measure, but was probably present to some degree in most cases. The gradient between pulmonary vein and right atrium gradient averaged 11 mmHg in 17 patients in whom it was measured (Table 2).

Seven infants had intracardiac, 24 supracardiac type (4 with drainage into the right superior vena cava), and 6 had infracardiac total anomalous pulmonary venous drainage; there were 2 infants with mixed drainage. The most common types encountered are illustrated in Fig. 1.

Surgical techniques

A two-stage correction for total anomalous pulmonary venous drainage has been tried with limited success (Mustard *et al.*, 1962; Behrendt *et al.*, 1972). At present, complete repair at a single operation is

Fig. 1 Types of total anomalous pulmonary venous drainage in 39 infants, 1971 to 1975.

generally preferred. Correction can be carried out either on cardiopulmonary bypass or using deep hypothermia with circulatory arrest. Results reported so far in various series do not clearly indicate superiority of either technique, but most surgeons prefer the latter in small infants. We began to use circulatory arrest with deep hypothermia in 1971 and now use this technique for infants under the age of 6 months.

When preparations for total correction have been completed, babies are taken to theatre where, under general endotracheal anaesthesia, arterial, venous, and urinary catheters are inserted if this has not already been done. Surface cooling to 27° to 28°C nasopharyngeal temperature is accomplished and a midline sternotomy incision is then performed. The ascending aorta is cannulated with a thin walled metal cannula and a single plastic cannula is placed through the right atrial appendage. The ductus arteriosus is ligated if patent and cardiopulmonary bypass is instituted with further cooling to 19°C nasopharyngeal temperature. The aorta is cross-clamped and the patient is exsanguinated into the oxygenator. The superior and inferior venae cavae are occluded and the repair is carried out. The technical details of the repair vary with the type of anomalous drainage.

INTRACARDIAC TYPE OF TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

When the pulmonary veins drain into the right atrium, either directly or by way of the coronary sinus, a good surgical result can be achieved with a relatively simple repair. When the drainage is into the coronary sinus, the coronary sinus is opened back into the left atrium through the patent foramen ovale (Fig. 2). A patch of pericardium or 'dacron' is then placed over the resulting defect to direct the flow into the left atrium. Anteriorly, the patch is sutured into the floor of the coronary sinus thus avoiding the tail of the atrioventricular node (Fig. 3).

SUPRACARDIAC TYPE OF TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

When the pulmonary veins drain through a left-sided vertical vein into the innominate vein, the first step in the repair is the ligation of the ascending vein. The dissection of this vein is carried out outside the pericardium and care is taken to avoid the phrenic nerve and to ligate well above the entry of the left upper pulmonary vein. When circulatory arrest is used, the anomalous vein should not be ligated until the circulation is stopped. When con-

Fig. 2 Operative technique for total anomalous pulmonary venous drainage to the coronary sinus. The coronary sinus is cut back into the left atrium and the septum between the coronary sinus and foramen ovale excised. PFO, patent foramen ovale; CS, coronary sinus.

ventional bypass is used, the common pulmonary vein should be incised immediately after ligation of the ascending vein. Observance of these rules will prevent severe pulmonary congestion which occurs if the anomalous vein is ligated too early in the procedure.

We now prefer the right-sided transatrial approach for the repair of this type of total anomalous pulmonary venous drainage (Shumaker and King, 1961; Gersony *et al.*, 1971). An incision is made transversely in the right atrium, continuing posteriorly through the patent foramen ovale, across the interatrial septum, and horizontally across the posterior wall of the left atrium. Care should be taken to make this incision parallel to and adjacent to the pulmonary vein. The horizontal collecting vein is incised on its anterior surface and an anastomosis is constructed between the posterior left

Fig. 3 A patch is used to redirect pulmonary venous blood to the left atrium. Anteriorly, the suture is placed through the floor of the coronary sinus to avoid its anterior rim. Abbreviations as in Fig. 2.

Fig. 4 Operative technique used for supracardiac total anomalous pulmonary venous drainage. SVC and IVC are occluded, venous cannula removed and ascending vein ligated. Right atrium is then opened across the septum into the left atrium. Horizontal collecting vein is incised and anastomosis to left atrium constructed. LPV, left pulmonary veins; RPV, right pulmonary veins; SVC, superior vena cava; IVC, inferior vena cava; AVV, ascending vertical vein.

atrium and the common vein (Fig. 4). To ensure an adequate opening, the anastomosis can be carried across the posterior interatrial septum, using a portion of the posterior wall of the right atrium to complete the connection. The cavity of the left atrium can then be enlarged either by shifting the interatrial septum to the right or by enlarging the

Fig. 5 Anastomosis is completed. The cavity of the left atrium is being enlarged by closing the atrial septal defect with a patch. Abbreviations as in Fig. 4.

atrial septal defect and closing it with a generous patch (Fig. 5). This technique has the distinct advantage of allowing the anastomosis to be constructed with the heart and the common pulmonary vein lying in their natural position, thus avoiding kinking or traction at the anastomosis.

In a variant of supracardiac total anomalous pulmonary venous drainage, drainage is via an anomalous ascending vein on the right side which enters the back of the right superior vena cava at the level of the confluence of the innominate veins. In all 4 patients with this anatomical variant, the vein was varicose and very friable; extreme care in dissection is therefore advisable.

Construction of the anastomosis with this variant can also be difficult since the horizontal collecting vein is usually high and to the right, causing difficulty in approximating it to the posterior wall of the left atrium.

INFRACARDIAC TYPE OF TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

In the repair of infracardiac type of total anomalous pulmonary venous drainage, we prefer the retrocardiac approach (Fig. 6 and 7). This allows better exposure of the common vein which is orientated vertically. The first step is again ligation of the anomalous vein immediately before beginning the repair. If there is a long distance between the con-

Fig. 6 Operative technique for infracardiac type. Heart is lifted upwards, pulmonary veins and descending vertical vein are dissected. The incision runs from the left upper pulmonary vein obliquely into the vertical vein. Corresponding incision is made in the left atrium. LV, left ventricle; LA, left atrium; DVV, descending vertical vein; PV, pulmonary vein.

Fig. 7 The vertical vein is ligated and divided. Anastomosis is performed with continuous suture. LA, left atrium; LV, left ventricle; DV, descending vein; LPV, left pulmonary vein.

fluence of the pulmonary veins and the left atrium, the descending vertical vein can be doubly ligated and divided, thus allowing the site of the anastomosis to come into closer apposition to the left atrium (Stark *et al.*, 1977). Oblique parallel incisions are then made in the confluence of pulmonary veins and in the posterior wall of the left atrium (Fig. 6). The incision in the common pulmonary vein can be carried down into the upper portion of the dilated ascending vein, allowing a larger anastomosis to be constructed (Fig. 7). The atrial septal defect or patent foramen ovale is then closed through a separate right atriotomy.

MIXED TYPES OF TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

Anomalous drainage does not always follow the same pathways from both lungs or from all lobes. Almost any combination of the standard types of drainage can exist in one patient. Operative techniques must be individualised in this small group of patients, but in general, when an anomalous connection, different from the main drainage, involves only one lobe, it can be left uncorrected without great physiological consequences. In the most common mixed type the left upper lobe vein drains directly into a vertical vein entering the left innominate vein, and there is infracardiac or intracardiac drainage of the remaining pulmonary veins.

Table 3 Results of operation related to anatomical type of total anomalous pulmonary venous drainage

Type	No.	Died
Innominate vein	20	7 (35%)
Coronary sinus or right atrium	7	1 (14%)
Infradiaphragmatic	6	3 (50%)
Right superior vena cava	4	3 (75%)
Mixed	2	0 (0%)
Total	39	14 (36%)

POSTOPERATIVE CARE

Intensive postoperative management of infants with total anomalous pulmonary venous drainage is crucial for survival. Sophisticated monitoring techniques referred to earlier, ventilatory support, and constant attendance by competent personnel are essential. Prevention and treatment of fluid overload and its complications is vital in the management of these infants. Lungs of infants who have had pulmonary oedema are particularly prone to accumulation of interstitial water. Avoidance of volume overload and maintenance of optimal cardiac output are necessary to prevent recurrent pulmonary oedema.

Atelectasis often occurs as a complication in the postoperative period. Ventilatory support either with intermittent positive pressure breathing or continuous positive airway pressure may be needed for a few days. Intensive physiotherapy, starting immediately after operation, is continued throughout the postoperative period. If the proper operation has been performed and these principles are followed, reasonable results can be expected.

Results

Of 39 infants, 14 died in hospital giving an overall mortality rate of 36 per cent. There have been no late deaths. The factor with the most striking effect on mortality was the anatomical type of total anomalous pulmonary venous drainage (Table 3). The mean age, body weight, and degree of pulmonary hypertension present in survivors was roughly equal to that of non-survivors (Table 4).

Table 4 Comparison of 25 survivors and 14 non-survivors.

	Survivors	Non-survivors
Mean age at operation (d)	89	80
Mean body weight at operation (kg)	4.1	3.8
Mean pulmonary/systemic systolic pressure ratio	0.95	1.00

Fig. 8 Survival rate related to age at operation.

Survival related to age at operation and to pulmonary/systemic systolic pressure ratio at cardiac catheterisation is illustrated in Fig. 8 and 9.

Pulmonary histology was examined in all but two of the infants who died, and particular attention was paid to the pulmonary vascular changes. Seven of the 12 showed the arterial changes of grade 2 pulmonary vascular disease, by Heath and Edwards' classification (1958). One showed grade 1 changes, while in the remaining 4, who were less than 1 month old, fetal arterial appearances persisted. Similar arterial changes were seen in the different lobes of each case.

The venous changes consisted of endothelial hyperplasia in 9 cases; 2 of these also showed

medial hypertrophy. The venous changes tended to be uniform throughout all lobes in 5, but were more severe in the lower lobes than upper lobes in 3; in 1 patient, the changes were restricted to the left lung. Some lymphatic dilatation was seen in 7 of the 12. None had bronchial venous dilatation. There was no constant correlation between the severity of arterial changes and the venous or lymphatic changes.

Discussion

Lemoine, Gersony, and Applebaum and their colleagues have also observed recent improvements in results of surgical correction of total anomalous

Fig. 9 Survival rate related to preoperative pulmonary/systemic systolic pressure ratio.

Table 5 Results of operation for total anomalous pulmonary venous drainage in infants in four large series

Author	No.	Died No.	%
Appelbaum <i>et al.</i> (1975) 1968–1974	31	15	48
Lemoine (1975) 1967–1974	45	15	33
Gersony <i>et al.</i> (1975, personal communication) 1969–1975	29	14	48
This study (1976) 1971–1975	39	14	36
Total	144	58	40

pulmonary venous drainage in infancy (Table 5) (Lemoine, 1975; Gersony *et al.*, 1975, personal communication; Applebaum *et al.*, 1975). The most significant improvement has been the increased survival of the younger infants. Before 1971, no infant less than 3 months of age had survived operation for total anomalous pulmonary venous drainage in this unit. In the current series, 16 of 26 (62%) survived. The success rate is still slightly lower in the group less than 1 month of age (Fig. 8). This could be related to the fact that the more severe lesions are the ones that present shortly after birth; it is significant that the infants in this series with the infracardiac type of total anomalous pulmonary venous drainage had a mean age of 3 weeks at operation, compared with the mean age of 12 weeks for the whole group.

The degree of pulmonary hypertension before operation had very little effect on ultimate survival. The mean value of the pulmonary/systemic systolic pressure ratio was similar in the survivors and non-survivors (Table 4). Further scrutiny of this relation shows slightly decreased survival in the 5 patients with pulmonary/systemic systolic pressure ratios greater than 1.4. This difference is not statistically significant (Fig. 9). In the relative lack of correlation between the degree of pulmonary hypertension and survival, our experience contrasts with that from other centres and indeed with our own past experience (Wukasch *et al.*, 1975; Behrendt *et al.*, 1972).

Mortality is most closely related to the anatomical type of total anomalous pulmonary venous drainage (Table 3). The coronary sinus or right atrial type had the lowest operative mortality. Only 1 of the 7 infants died; this patient failed to come off bypass and at necropsy was found to have a severely hypoplastic left ventricle. A high survival rate with this type has also been observed by others. All 15 patients in Lemoine's series (1975) and all 7 patients in Gersony *et al.*'s (1975) survived operation. Pulmonary venous obstruction is uncommon in this type and the repair is technically easy.

The mortality rate for the supracardiac type, which accounted for over 50 per cent of the patients in our series, was similar to the mortality for the overall group (Table 3). Four of our patients had drainage to the right superior vena cava, and 3 of these died. The reason for this high mortality is not entirely clear. Bleeding from the very friable ascending vein and difficulties in approximating the common pulmonary vein to the posterior wall of the left atrium may be important factors.

The infracardiac type was associated with a 50 per cent mortality (Table 3). All in this group had obstructed pulmonary venous drainage and were very ill at the time of admission and operation.

When the causes of death in 14 patients were examined, pulmonary oedema was found in 12. Arrhythmias were responsible in 2 patients, fluid overload in 1, and stenosis of the anastomosis in 1. The common vein, left atrium, or left ventricle was hypoplastic in 4, and no cause was found for persistent fatal pulmonary congestion in 4 cases. It was possible that the anastomosis was kinked, or that an adequate sized left atrial reservoir had not been constructed (Parr *et al.*, 1974); precise assessment of these factors is very difficult at necropsy.

There are obviously problems with the management of total anomalous pulmonary venous drainage in infancy which remain unsolved, but the substantial recent increase in survival rate is encouraging. We attribute the improved results to: (1) early recognition and prompt referral, (2) an aggressive approach to diagnosis involving complete cardiac catheterisation and angiocardiology to show the exact site of drainage of all pulmonary veins and to exclude any additional cardiac lesions, (3) vigorous preoperative care, (4) early complete correction including construction of a large anastomosis and enlargement of the left atrium when indicated, and (5) intensive postoperative management, paying particular attention to fluid balance and treatment of pulmonary complications.

We thank the Henry Ford Hospital and Appleton-Century-Crofts for the use of Fig. 2, 3, 4, 5, 6, and 7 from the article 'Surgical treatment of total anomalous pulmonary venous drainage in infants' by J. Stark, D. Clarke, M. de Leval, and J. F. N. Taylor, to be published in the Proceedings of 2nd Henry Ford International Symposium on Cardiac Surgery, held in Detroit, U.S.A., in October 1975.

References

- Applebaum, A., Kirklin, J. W., Pacifico, A. D., and Barger, L. M., Jr. (1975). The surgical treatment of total anomalous pulmonary venous connection. *Israel Journal of Medical Sciences*, **11**, 89–96.

- Barratt-Boyes, B. G. (1973). *Advances in Cardiovascular Surgery*, p. 127. Ed. by J. W. Kirklin. Grune and Stratton, New York.
- Behrendt, D. M., Aberdeen, E., Waterston, D. J., and Bonham-Carter, R. E. (1972). Total anomalous pulmonary venous drainage in infants. *Circulation*, **46**, 347-356.
- Bonham-Carter, R. E., Capriles, M., and Noe, Y. (1969). Total anomalous pulmonary venous drainage: a clinical and anatomical study of 75 children. *British Heart Journal*, **31**, 45-51.
- Brody, H. (1942). Drainage of the pulmonary veins into the right side of the heart. *Archives of Pathology*, **33**, 221-240.
- Burroughs, J. T., and Kirklin, J. W. (1956). Complete surgical correction of total anomalous pulmonary venous connection: report of three cases. *Proceedings of the Staff Meetings of the Mayo Clinic*, **31**, 182-188.
- Cooley, D. A., and Ochsner, A., Jr. (1957). Correction of total anomalous pulmonary venous drainage. Technical considerations. *Surgery*, **42**, 1014-1021.
- El-Said, G., Mullins, C. E., and McNamara, D. G. (1972). Management of total anomalous pulmonary venous return. *Circulation*, **45**, 1240-1250.
- Gathman, G. E., and Nadas, A. S. (1970). Total anomalous pulmonary venous connection: clinical and physiologic observations of 75 pediatric patients. *Circulation*, **42**, 143-154.
- Gersony, W. M., Bowman, F. O., Steeg, C. N., Hayes, C. J., Jesse, M. J., and Malm, J. R. (1971). Management of total anomalous pulmonary venous drainage in early infancy. *Circulation*, **43**, Suppl. 1, 19-24.
- Gomes, M. R., Feldt, R. H., McGoon, D. C., and Danielson, G. K. (1970). Total anomalous pulmonary venous connection. *Journal of Thoracic and Cardiovascular Surgery*, **60**, 116-122.
- Heath, D., and Edwards, J. E. (1958). The pathology of hypertensive pulmonary vascular disease. *Circulation*, **18**, 533-547.
- Keith, J. D., Rowe, R. D., and Vlad, P. (1967). *Heart Disease in Infancy and Childhood*, 2nd ed., p. 512. MacMillan, New York.
- Leachman, R. D., Cooley, D. A., Hallman, G. L., Simpson, J. W., and Dear, W. E. (1969). Total anomalous pulmonary venous return. *Annals of Thoracic Surgery*, **7**, 5-12.
- Lemoine, F. (1975). Retour veineux pulmonaire total anormal chez le nourisson. Traitement chirurgical. Presented at 7th Riunioni Medico-Chirurgiche Internazionali, Torino.
- Lewis, F. J., Varco, R. L., Taufic, M., and Niazi, S. A. (1956). Direct vision repair of triatrial heart and total anomalous pulmonary venous drainage. *Surgery, Gynecology and Obstetrics*, **102**, 713-720.
- Miller, W. W., Rashkind, W. J., Miller, R. A., Hastreiter, A. R., Green, E. W., Golinko, R. J., and Young, D. (1967). Total anomalous pulmonary venous return: effective palliation of critically ill infants by balloon atrial septostomy. *Circulation (Abstract) Suppl. II*, vol. **35-36**, 189.
- Muller, W. H., Jr. (1951). The surgical treatment of transposition of the pulmonary veins. *Annals of Surgery*, **134**, 683.
- Mustard, W. T., and Dolan, F. G. (1957). The surgical treatment of total anomalous pulmonary venous drainage. *Annals of Surgery*, **145**, 379-387.
- Mustard, W. T., Keith, J. D., and Trusler, G. A. (1962). Two-stage correction for total anomalous pulmonary venous drainage in childhood. *Journal of Thoracic and Cardiovascular Surgery*, **44**, 477-484.
- Parr, G. V. S., Kirklin, J. W., Pacifico, A. D., Blackstone, E. H., and Lauridsen, P. (1974). Cardiac performance in infants after repair of total anomalous pulmonary venous connection. *Annals of Thoracic Surgery*, **17**, 561-572.
- Shumacker, J. B., and King, H. (1961). A modified procedure for complete repair of total anomalous pulmonary venous drainage. *Surgery, Gynecology and Obstetrics*, **112**, 736-765.
- Stark, J., Clarke, D., de Leval, M., and Taylor, J. F. N. (1977). Surgical treatment of total anomalous pulmonary venous drainage in infants. In *Proceedings of 2nd Henry Ford International Symposium on Cardiac Surgery, Detroit, U.S.A., October 1975*. In the press.
- Wukasch, D. C., Deutsch, M., Reul, G. J., Hallman, G. L., and Cooley, D. A. (1975). Total anomalous pulmonary venous return. Review of 125 patients treated surgically. *Annals of Thoracic Surgery*, **19**, 622-633.

Requests for reprints to Dr. J. Stark, Thoracic Unit, The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.