Total anomalous pulmonary venous drainage in infancy

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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 angiography, (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 few
Table 1  Total anomalous pulmonary venous drainage in 86 infants

<table>
<thead>
<tr>
<th>Period</th>
<th>Number</th>
<th>Survived</th>
</tr>
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<tbody>
<tr>
<td>1963-1970</td>
<td>47</td>
<td>15 (32%)</td>
</tr>
<tr>
<td>1971-1975</td>
<td>39</td>
<td>25 (64%)</td>
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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 retrosternal 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 angiocardiography 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 angiocardiograms 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*

<table>
<thead>
<tr>
<th></th>
<th>Range</th>
<th>Mean</th>
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<tbody>
<tr>
<td>Age at operation (d)</td>
<td>1-352</td>
<td>83</td>
</tr>
<tr>
<td>Body weight at operation (kg)</td>
<td>2-6-6-4</td>
<td>4-0</td>
</tr>
<tr>
<td>Arterial O(_2) saturation (%)</td>
<td>33-92</td>
<td>75</td>
</tr>
<tr>
<td>Pulmonary/systemic systolic pressure ratio</td>
<td>0-27-1-83</td>
<td>0-98</td>
</tr>
<tr>
<td>PV to RA mean gradient (mmHg) (17 patients)</td>
<td>0-34</td>
<td>11</td>
</tr>
</tbody>
</table>

**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 achieved.
TAPVD in infancy

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°C 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 conventional 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; Gerseny 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
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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.

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. 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.

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.
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

<table>
<thead>
<tr>
<th>Type</th>
<th>No.</th>
<th>Died</th>
</tr>
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<tbody>
<tr>
<td>Innominate vein</td>
<td>20</td>
<td>7 (35%)</td>
</tr>
<tr>
<td>Coronary sinus or right atrium</td>
<td>7</td>
<td>1 (14%)</td>
</tr>
<tr>
<td>Infradiaphragmatic</td>
<td>6</td>
<td>3 (50%)</td>
</tr>
<tr>
<td>Right superior vena cava</td>
<td>4</td>
<td>3 (75%)</td>
</tr>
<tr>
<td>Mixed</td>
<td>2</td>
<td>0 (0%)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>39</td>
<td>14 (36%)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th></th>
<th>Survivors</th>
<th>Non-survivors</th>
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<tbody>
<tr>
<td>Mean age at operation (d)</td>
<td>89</td>
<td>80</td>
</tr>
<tr>
<td>Mean body weight at operation (kg)</td>
<td>4.1</td>
<td>3.8</td>
</tr>
<tr>
<td>Mean pulmonary/systemic systolic pressure ratio</td>
<td>0.95</td>
<td>1.00</td>
</tr>
</tbody>
</table>
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

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Fig. 8 Survival rate related to age at operation.

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

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