Total Anomalous Pulmonary Venous Drainage in Infants

I. Clinical and Hemodynamic Findings, Methods, and Results of Operation in 37 Cases


SUMMARY
Thirty-seven infants with total anomalous pulmonary venous drainage have been operated upon at The Hospital for Sick Children, Great Ormond Street, London, with 13 survivors. Most were rapidly deteriorating when admitted. Cardiac catheterization, angiography, and operation were urgently undertaken to achieve the maximum salvage. Physical examination, plain radiography, and electrocardiography were insufficient for accurate diagnosis. Survival was closely related to the degree of pulmonary hypertension, which was dependent on the type of anomalous venous drainage and the presence of pulmonary venous obstruction. The prognosis was good for patients over 3 months of age, especially those without pulmonary venous obstruction, provided that they arrived in the hospital in reasonable condition. The best survival rate, nine of 11 cases, was achieved in those patients between 3 and 12 months of age with supracardiac drainage.

Additional Indexing Words:
Congenital heart disease
Total anomalous pulmonary venous drainage
Infants
Pulmonary hypertension

 TOTAL ANOMALOUS pulmonary venous drainage (TAPVD) is a rare condition among the general population, but it is encountered frequently in hospitals caring for large numbers of infants with heart disease. The embryology and anatomy of TAPVD have been described previously.1-8 Four anatomic patterns have been described.

Supracardiac: Via a vertical vein, usually draining to the left innominate vein. Cardiac: Directly to the right atrium or coronary sinus.

Infrcardiac: Via a vertical vein to the portal vein or inferior vena cava.

Mixed: A combination of two or more of the above types.

Several major hemodynamic abnormalities are common to each of these. There is a volume overload of the right ventricle and pulmonary circulation resulting in congestive cardiac failure, and there is mixing of the systemic and pulmonary venous blood in the right atrium resulting in arterial desaturation. In addition, there is sometimes obstruction to pulmonary venous flow which further increases the burden on the right ventricle and may be severe enough to diminish the pulmonary flow below systemic (figs. 1, 2).3, 4

More than 75% of these patients die within the first year of life,5, 6 often suddenly; therefore, to achieve an appreciable salvage...
Figure 1

Pressure gradients recorded along the anomalous trunk in TAPVD with the corresponding angiogram showing the site of obstruction, a stricture where the anomalous trunk joined the inferior vena cava.
The vertical vein was compressed between the left pulmonary artery and left bronchus.

Our approach to this problem and presents our operative methods for each of the anatomic
types and our results with operations performed upon 37 infants who were part of a larger group of 56 children operated upon for this condition at all ages.

Material and Methods

Clinical Presentation

Thirty-seven infants less than 12 months of age underwent operation for TAPVD at The Hospital for Sick Children, Great Ormond Street, London, from 1963 until January, 1970. The pattern of pulmonary venous drainage in this group was: supracardiac in 22, cardiac in 11, infracardiac in three, and mixed in one (supracardiac and cardiac). The age at operation is shown in figure 3.

These infants presented because of poor feeding, labored breathing, and, often, cyanosis. On examination cyanosis was usually present, but rarely was it prominent. In fact, the average arterial oxygen saturation was 73% (range 36–95), and only in three cases was it below 60%. There was generally a systolic murmur and, in one third of the cases, a diastolic murmur at the left sternal edge. Hepatomegaly was infrequent. Many were in respiratory distress, and seven required endotracheal intubation and ventilation prior to operation.

The chest X-rays generally showed gross cardiomegaly and pulmonary plethora. The wide mediastinum ("snowman"), which is typical of supracardiac TAPVD in older patients, was seen in only six of 22 cases. A "boxlike" configuration of the heart, due to massive right-sided enlargement, was evident in only a few of our cases. Normal heart size was seen in about one quarter of the infants, both with and without obstruction to pulmonary venous drainage.

All 37 patients had right ventricular hypertrophy (RVH) on ECG, and 17 had evidence of right atrial enlargement. The hemoglobin averaged 11.8g% (range 9.9–20.0), and other laboratory tests were noncontributory.

Cardiac Catheterization and Angiography

In all patients the diagnosis was established by cardiac catheterization and angiography. In 22 patients complete left and right heart catheterization was carried out. In all instances an increase in oxygen saturation was recorded at the site appropriate to the type of anomalous pulmonary venous drainage, i.e., the superior vena cava (SVC), inferior vena cava (IVC), or right atrium (RA). In six patients the RA mean pressure substantially exceeded the left atrial (LA), suggesting that the interatrial communication was small. In four of these a balloon atrial septostomy was performed. Right ventricular (RV) peak systolic pressure was elevated in each patient, in 10 instances to a level considerably in excess of systemic arterial pressure. An RV systolic pressure in excess of systemic proved to be associated with angiographic evidence of obstruction to pulmonary venous drainage in nine of 10 such cases in which the required measurements were made (fig. 4). In 14 of 23 cases with supracardiac drainage, pressure recordings were made along the path of the anomalous vein from pulmonary vein wedge to RA to ascertain the presence or absence of obstruction to venous return. In the nine cases of supracardiac drainage with obstruction, gradients of 7–40 mm Hg were recorded (fig. 2).

Relationship of pulmonary hypertension to angiographic evidence of obstruction of the anomalous pulmonary venous trunk. (Compare also with fig. 7.)

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Twenty-three patients had both right and left heart angiocardiograms, whereas in 14 instances the study was limited to a demonstration of the pattern of pulmonary drainage by injection of contrast media into the pulmonary artery or into the anomalous vein itself. These angiograms were all carefully reviewed by two observers. Even accepting the fact that biplane angiography provides only a very approximate assessment of ventricular size, the capacity of the LV appeared definitely small in only six of 22 instances. (Only one of these survived operation.)

Seventeen patients were found to have other cardiovascular malformations (table 1), the most common being a patent ductus arteriosus (PDA) of medium or large diameter.

### Table 1

TAPVD in 37 Infants

<table>
<thead>
<tr>
<th>Associated anomalies</th>
<th>No.</th>
</tr>
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<tbody>
<tr>
<td>PDA (medium large)</td>
<td>16</td>
</tr>
<tr>
<td>Obstructed ascending vertical vein</td>
<td>9</td>
</tr>
<tr>
<td>Preductal coarctation</td>
<td>1</td>
</tr>
<tr>
<td>Subaortic stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Persistent left SVC</td>
<td>1</td>
</tr>
<tr>
<td>A-V canal with double IVC</td>
<td>1</td>
</tr>
<tr>
<td>Alimentary tract</td>
<td>5</td>
</tr>
<tr>
<td>No other lesions</td>
<td>20</td>
</tr>
<tr>
<td>PDA only</td>
<td>14</td>
</tr>
</tbody>
</table>

Abbreviations: PDA = patent ductus arteriosus; SVC = superior vena cava; IVC = inferior vena cava.

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**Figure 5**

Drawings of stages in operative correction of supracardiac TAPVD (see text). (Top, left) Incisions and placement of pulmonary artery and right ventricular vent. (Top, right) Anastomosis of anomalous vein to left atrium half completed. (Bottom) Anastomosis completed and common anomalous vein ligated.

Supracardiac and Infracardiac (Figure 5)

A left thoracotomy (crossing the sternum) was used to correct drainage via a vertical vein up to the left innominate vein or down to the portal system or IVC. This approach gave good exposure of the common trunk and LA and also allowed ligation of the PDA, if present. Cardiopulmonary bypass was begun, using the left common iliac artery for arterial input and a single line in the RA appendage for venous drainage.

The left common iliac artery was cannulated prior to thoracotomy. With this cannula in place, hypotension could often be reversed by a small intraarterial transfusion from the pump oxygenator, and cardiopulmonary bypass could be rapidly begun in the event of circulatory collapse. Dissection of the PDA was deferred until after all connections to the pump oxygenator had been made. To decompress the pulmonary circulation and prevent pulmonary edema, a vent line was placed through the RV infundibulum into the PA with one side hole left within the RV (fig. 5, top left).
The confluent vein was dissected free, and a side-to-side anastomosis was constructed between it and the overlying LA using straight thin-jawed vascular clamps for partial occlusion (fig. 5, top right). The anastomosis was carefully made as long as possible, and the smallest anastomosis was 17 mm in length. If the patient's condition was stable, a trial occlusion of the anomalous vertical vein was next performed while the pulmonary venous pressure was recorded. If the pulmonary venous pressure remained low, the vertical vein was ligated (fig. 5, bottom), completely diverting the pulmonary drainage into the LA. This was done in half the cases. No attempt was made to close the interatrial communication, since this might have provided a safety valve for the left side of the heart should it have proved unable to handle the new load imposed on it.

Cardiac (Figure 6)

TAPVD to the coronary sinus or RA was approached by a right thoracotomy or, in recent years, by a median sternotomy. The RA was opened, and a portion of the atrial septum was excised (fig. 6, top left and right). Then a Dacron patch was placed to direct the pulmonary venous return to the LA (fig. 6, bottom). Since the coronary sinus receives the anomalous drainage in some cases, the coronary venous blood is diverted to the left atrium by this technic; but, as coronary venous return comprises only about 4% of the cardiac output, the resulting right-to-left shunt is unimportant.

Postoperative Care

Each of these infants was cared for in the intensive care unit for several days. Arterial and venous pressures were continuously monitored via cannulae. The ECG, urine output, temperature, and serum electrolytes were monitored.

Tracheostomy was performed after bypass in each of these infants in anticipation of a prolonged need for ventilatory support during a difficult postoperative course. In 11 of the 13 infants who survived operation, ventilation was required for 1–4 weeks postoperatively.

Positive-pressure ventilation with an Engström ventilator was employed in all patients and was adjusted in accordance with the arterial blood gases. Several of our patients with TAPVD deteriorated quite suddenly on the second or third postoperative day after initially progressing well. Therefore, we now make no attempt to wean these patients from the ventilator until this period has elapsed. Digoxin was resumed on the day after operation. Furosemide was often given, and other drugs such as isoproterenol, epinephrine, calcium chloride, and sodium bicarbonate were sometimes used.

Feeding was generally resumed the first day after operation by means of a fine nasogastric tube.
Results

Survival

Of the 37 patients, 13 survived operation and were discharged from the hospital (fig. 7). The survivors consisted of: supracardiac, 11 of 22; cardiac, one of 11; infracardiac, none of three; and mixed cardiac and supracardiac, one of one.

Of the nine patients with obstructed supracardiac drainage, three survived (fig. 4). In one, the RV pressure was 70/9, the left ventricular pressure 74/8, and the gradient along the anomalous vein 7 mm Hg. In the second, the RV pressure was 80/5, the systemic arterial 75/45, and the venous gradient 16-20 mm Hg. In the third, the RV pressure was 70/20 and the arterial pressure 45/35. The venous gradient was not measured. Thus, even severe venous obstruction did not preclude survival.

Twelve patients failed to come off bypass. Another four patients remained in a state of low cardiac output and died within a few hours. Five patients appeared to be doing well for 1 to 3 days before suddenly deteriorating. Two others died as a result of massive atelectasis and one of an intracranial hemorrhage. No patient who required endotracheal intubation and ventilation preoperatively survived operation. The presence of a PDA did not adversely affect survival, but no patient survived who had a more complex associated malformation. Only two patients under 3 months of age survived, whereas nine patients of 11 over 3 months of age with supracardiac drainage survived.

Complications

Only six of the 13 survivors convalesced without a complication. The complications experienced by the remainder are listed in table 2. The one patient with the mixed type of drainage was incorrectly diagnosed preoperatively and had an anastomosis made between the LA and the vertical vein. She deteriorated 3 weeks postoperatively, and upon recatheterization it was found that only the left pulmonary veins drained through the vertical vein, the right veins draining into the coronary sinus. At reoperation 6 weeks after the first operation, the coronary sinus was diverted to the left with a Dacron patch; and the previous anastomosis, which appeared small, was enlarged. This patient survived with no further complications.

Postmortem Findings

Invariably, at autopsy the RV was massively enlarged, making the LV appear relatively small. The LV was thick walled, but in most instances was judged to be of relatively normal cavity size for age. The RA was large and the LA of normal size or somewhat small. The interatrial communications were between 4 and 7 mm in diameter, that is the same approximate size as the aortic valve orifice.9 Two appeared to be actual secundum ASDs, and the remainder appeared to be greatly

Table 2

<table>
<thead>
<tr>
<th>Complications in 37 Infants</th>
<th>No.</th>
<th>No. fatal</th>
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</thead>
<tbody>
<tr>
<td>Severe CHF</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Massive atelectasis</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Tracheostomy complications</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Seizures</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Late stricture of anastomosis</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Severe bleeding diathesis</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Aspiration with arrest</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Plugged endotracheal tube with arrest</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Hyperkalemia with arrest</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Hypocalcemia with seizures</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Survivors without complication</td>
<td>6/13</td>
<td></td>
</tr>
</tbody>
</table>

Abbreviation: CHF = congestive heart failure.
enlarged foramina ovalae. One patient had an atrioventricular canal defect as well as a secundum ASD. No other patient had a VSD. In one patient the anastomosis was inadequate, but in all others it appeared to be technically satisfactory.

Sections were taken from right and left ventricles and examined with conventional and special staining techniques for evidence of ischemic damage. In only six instances was there evidence of severe ischemic damage. These six infants were of varying ages and patterns of venous drainage. One had pulmonary venous obstruction, and none had angiographic findings of severe RVH or a small LV. Only one appeared critically ill preoperatively.

Survival was closely related to the degree of pulmonary hypertension. When this was less than 75% of the systolic pressure, six cases of seven survived. When this was greater than systolic, only two of 10 cases survived (fig. 7).

**Long-Term Follow-Up**

One of the 13 survivors died 7 months postoperatively of congestive heart failure after an initially good result. At autopsy his anastomosis measured only 5 mm in diameter; therefore, pulmonary venous obstruction may have been the cause of his death. Two patients, operated upon recently, have been followed for less than 1 year. The remaining 10 patients have no cardiovascular symptoms 3–6 years postoperatively (average follow-up 55 months). One is mentally retarded, but the others are normal mentally and physically.

Ten have residual interatrial communications, and four have unligated vertical veins. One of these successfully underwent ligation of the anomalous vein and patch closure of a moderate-size ASD at age 5 years. However, we have suspected on clinical grounds that some of the other interatrial communications have closed spontaneously, and this was proven by cardiac catheterization in three patients 3–5 years postoperatively. The only survivors with the coronary sinus and mixed types of drainage were, of course, completely corrected at the initial procedure.

**Discussion**

Infants with TAPVD remain a difficult problem. Without treatment at least 75% will die within the first year of life, the majority within the first 6 months. A diagnosis based upon clinical findings alone is often misleading. However, with an accurate diagnosis based upon cardiac catheterization and angiography and with active surgical treatment, many of these can be salvaged. We have learned that when an infant with TAPVD deteriorates sufficiently to require hospitalization, he should be treated as an acute emergency, because his downhill course may be rapid. In our earlier experience four patients died while in the hospital awaiting operation, and even now many require resuscitation in the few hours between admission and operation. Nevertheless, cardiac catheterization is mandatory before operation in order to establish the diagnosis, the detailed anatomy of the anomalous drainage, and the presence of associated lesions. We adopted an operative technic which allowed the rapid commencement of cardiopulmonary bypass if the infant deteriorated during thoractomy. Tracheostomy was performed at the completion of bypass in all patients, since we have found that infants with TAPVD generally require prolonged respiratory support. All are closely attended for several days in the intensive care unit.

The cause of death after operation in this condition remains uncertain. As in the case of other congenital cardiac defects, there is a high mortality rate among infants in the first 3 months of life. Angiographic or pathologic evidence of underdevelopment of the left ventricle was found in only a minority. The morbid changes of chronic ischemia that have been seen in other situations were not impressive. Those who required preoperative intubation and ventilation invariably succumbed. Late referral possibly contributed to their deaths.

Cooley et al. have suggested that increased pulmonary resistance may be correlated with a fatal outcome. We found the calculation of relative pulmonary and systemic
blood flow from oxygen saturation data unreliable in this group of sick infants. Certainly, only three of our nine patients with preoperative pulmonary venous obstruction survived, and the prognosis was clearly worse in those patients with severe pulmonary hypertension.

The pulmonary artery or right ventricular pressure was related not only to survival but also to the type of anatomic derangement in each case. Of the favorable group whose RV pressure was less than 75% of the systemic, six of seven cases had drainage of the supracardiac type (fig. 8), whereas of those with RV pressures greater than systemic nine of 10 were known to have obstruction of the pulmonary vein (fig. 4). The finding of an RV pressure close to the systemic level in almost all the cases with drainage at cardiac level was not explained by finding a small atrial septal defect. In many the atrial septal defect was large. Whatever the explanation of this degree of hypertension, it probably explains why this group had a poor prognosis. Gathman and Nadas also noted the occurrence of extreme pulmonary hypertension in the cardiac type of TAPVD, without obstruction to pulmonary venous flow or a small ASD.

Balloon atrial septostomy has been performed to decrease pulmonary and increase systemic blood flow in these patients and thus improve the patient sufficiently to allow operation to be deferred. Our present policy is not to do this, because each of our patients who has come to autopsy has a large communication already. Furthermore, we believe operation should not be deferred because, in the past, four patients of ours died suddenly while awaiting operation. However, if the atrial communication is small, as indicated by a pressure gradient between the atria, the flow to the left heart may be severely limited, and the pulmonary flow and hypertension increased. Under these circumstances balloon atrial septostomy and delay of the corrective operation may help a critically ill infant. This is a most important question in the management of these infants which will be answered by further experiences.

At present we prefer to correct extracardiac TAPVD in two stages, as suggested by Mustard, ligating the vertical vein at the first stage, if this is well tolerated, but leaving the interatrial communication for a subsequent operation if necessary. This simplifies the initial procedure in these very sick infants and also allows a left thoracotomy approach to be used. We prefer a left thoracotomy because the frequently associated PDA can be ligated easily and because the anastomosis is more readily done than from a median sternotomy.

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