Total Anomalous Pulmonary Venous Connection

Clinical and Physiologic Observations of 75 Pediatric Patients

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SUMMARY

Experience with 75 cases of proven total anomalous pulmonary venous connection without other significant cardiac malformations treated at the Children's Hospital Medical Center in Boston from January 1950 to June 1968 forms the basis of this report. Clinical observations of patients with this defect presenting in infancy are reviewed in depth, and the few patients presenting after 1 year of age are included to give a spectrum of this disease in the pediatric age group.

Chest x-rays and complete electrocardiograms were available in each case. Sixty-one patients had partial or complete cardiac catheterizations. The anatomic and physiologic data from these studies are presented, and the results of medical management are evaluated.

On the basis of the physiologic data, an approach to management is proposed which includes early operative intervention in those patients with pulmonary vascular obstruction. If pulmonary hypertension is present without vascular obstruction, medical management may be tried. If pulmonary hypertension is not present and particularly if there is a significant gradient across the right ventricular outflow tract, medical management appears to be the treatment of choice until operative intervention can be undertaken with less operative mortality.

Additional Indexing Words:
- Atrial septal defect, surgically created
- Electrocardiographic findings
- Pulmonary vascular obstruction
- Radiographic findings
- Genetic defects
- Pulmonary hypertension
- Right ventricular outflow gradient
- Cardiac catheterization

The diagnosis and management of total anomalous pulmonary venous connection (TAPVC) continues to be one of the most difficult and provocative problems encountered by the pediatric cardiologist and cardiovascular surgeon. The purpose of this paper is to review and correlate the clinical, physiologic, and anatomic spectrum of this cardiac defect.

The poor prognosis of the vast majority of the patients who present in early infancy is emphasized. We propose an aggressive approach to therapy.

Methods

This report includes 75 consecutive patients, representing all the patients with a proven diagnosis of TAPVC without other major cardiac defects admitted to The Children's Hospital Medical Center between January 1950 and June 1968.* The diagnosis was confirmed by autopsy

*Fifteen of these patients have been discussed in two earlier publications from this institution.1,2
examination in 54, at cardiac catheterization and subsequently confirmed at operation in 17, and at cardiac catheterization alone in the remaining four. An atrial septal defect (ASD) or patent foramen ovale (PFO) was considered to be a necessary component of the basic defect. Twenty patients included also had a probe patent or functionally patent ductus arteriosus (PDA); all 20 were less than 4 months of age.

The clinical material was divided by age and level of pulmonary artery pressure (table 1). Sixty-nine patients were first examined when less than 1 year of age. Fifty-two of these had evidence, physiologic or pathologic, or both, of pulmonary artery hypertension (PAH). The pulmonary artery systolic pressure was at least half the systemic pressure in 35 of the patients who were catheterized. Right ventricular hypertension at systemic levels without autopsy evidence of right ventricular outflow obstruction was present in an additional three infants. Indirect evidence of pulmonary hypertension in the remaining 14 cases was based either on gross pathologic pulmonary venous obstruction (11 cases), in seven of which the pulmonary vein drained into the portal vein (PV), or histologic evidence of severe medial and intimal proliferation of the arterioles (three cases).

Forty-two of the 52 infants with PAH also had pulmonary vascular obstruction (PVO). Almost all of these had anatomic or physiologic evidence of pulmonary venous obstruction proven at cardiac catheterization or postmortem examination.

Seventeen infants under 1 year of age had pulmonary artery pressures which were less than half the systemic pressure and were therefore considered not to have significant PAH. None had evidence of PVO either at catheterization or postmortem examination.

Only six patients (8% of our total series) had no cardiac evaluation before they were 1 year of age. All underwent catheterization at a later date (one at 23 months, four between 4 and 6 years, and one at 16 years). Five of the six had pulmonary artery pressures less than half the systemic pressure. In the remaining patient the pulmonary artery was not entered, but the right ventricular pressure was 100 mm Hg. Pressure in the anomalous pulmonary venous trunk was elevated beyond an anatomic narrowing (mean free PV pressure was 32 mm Hg), and we believe his pulmonary artery pressure was comparable to the right ventricular pressure.

Standard 12-lead electrocardiograms and chest x-rays were obtained in each case. Sixty-one patients had a total of 69 preoperative cardiac catheterizations.* The data of these studies will be analyzed.

Thirty-one patients were treated by medical means alone. Twenty-three had operative intervention prior to 1 year of age, and 21 were older than 1 year of age at the time of operation.

Operative intervention in our patients covered a long span (18 years), a variety of operative technics, and frequently changing indications for surgery. Because of this, meaningful evaluation of the results of surgery was not possible. Our recent results have been encouraging, but of insufficient magnitude for analysis.

Autopsy findings will not be discussed in detail, but reference will be made to pathologic findings particularly with respect to drainage site, presence and location of pulmonary venous obstruction, and microscopic evidence of PVO.

Results

Our data do not allow an accurate assessment of the incidence of TAPVC, but others have reported an overall frequency of approximately 1%.3, 4

Our series is in general agreement with the literature as to the site of anomalous venous connection,5 the most frequent being to the left superior vena cava (LSVC) with drainage to the heart via the innominate vein, followed by connection to the coronary sinus (CS), portal vein, and right superior vena cava (RSVC) orazygous vein in that order (table 2). We noted a somewhat more frequent incidence of mixed TAPVC with insertion at more than one site than that reported by others.

Several authors6, 7 have noted a male predominance in the subdiaphragmatic types of anomalous return, but one author has disputed this.8 Our series shows a male predominance in both the supracardiac and

Table 1

<table>
<thead>
<tr>
<th>Age</th>
<th>Total</th>
<th>PAH</th>
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<tr>
<td>1 yr or less</td>
<td>69</td>
<td>52</td>
<td>17</td>
</tr>
<tr>
<td>More than 1 yr</td>
<td>6</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>Total</td>
<td>75</td>
<td>53</td>
<td>22</td>
</tr>
</tbody>
</table>

*Sixty-three of these cardiac catheterizations have been carried out at this institution; the remaining six patients were referred to us after having had cardiac catheterization performed at other institutions.

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Table 2
Anatomic Classification of 75 Cases of Proven Total Anomalous Pulmonary Venous Connection Without Associated Major Intracardiac Defects Treated at The Children's Hospital Medical Center (Modified from Darling and Associates)  

<table>
<thead>
<tr>
<th>Type</th>
<th>Cases</th>
<th>% of total</th>
<th>% of group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supracardiac</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>LSVC (via innominate to right heart)</td>
<td>26</td>
<td>34.7</td>
<td>45.4</td>
</tr>
<tr>
<td>RSVC or Azygos vein</td>
<td>8</td>
<td>10.7</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>34</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cardiac</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Coronary sinus (CS)</td>
<td>14</td>
<td>18.6</td>
<td>22.6</td>
</tr>
<tr>
<td>Right atrium</td>
<td>3</td>
<td>4.0</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>17</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subdiaphragmatic</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Portal vein</td>
<td>13</td>
<td>17.3</td>
<td>21.3</td>
</tr>
<tr>
<td>Ductus venosus</td>
<td>3</td>
<td>4.0</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>16</td>
<td></td>
<td></td>
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<tr>
<td>Mixed</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>RSVC and LSVC</td>
<td>2</td>
<td>2.6</td>
<td></td>
</tr>
<tr>
<td>CS and LSVC</td>
<td>3</td>
<td>4.0</td>
<td></td>
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<tr>
<td>LSVC and RA</td>
<td>1</td>
<td>1.3</td>
<td>10.5</td>
</tr>
<tr>
<td>RSVC and RA</td>
<td>1</td>
<td>1.3</td>
<td></td>
</tr>
<tr>
<td>RSVC and portal vein</td>
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<td>1.3</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>8</td>
<td></td>
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<tr>
<td>Grand total</td>
<td>75</td>
<td>99.8</td>
<td>99.8</td>
</tr>
</tbody>
</table>

the subdiaphragmatic types with an overall male incidence of 65% (P < 0.01) (table 3).

Associated Malformations
Four of our patients had specific syndromes, and an extensive review of the English literature did not reveal any previous reports of TAPVC occurring with these syndromes.

The first patient (G.S., case 8) had the classical family history and physical findings of the Holt-Oram syndrome, associated with TAPVC to the LSVC. Another (D.L., case 51) had classical features of the Klippel-Feil syndrome, with TAPVC to the RSVC. A third (W.T., case 45) had phocomelia, apparently not drug induced, with TAPVC to the LSVC. The fourth patient (T.v.E., case 26) with TAPVC to the coronary sinus had an atrial septal defect, a patent ductus arteriosus, and a ventricular septal defect. A syndrome first reported by Schachenmann and associates.

In addition, we encountered two sibships in which TAPVC occurred. We believe these represent the first cases of TAPVC in siblings.

Five patients had associated genitourinary anomalies ranging from hydronephrosis, secondary to a ureteropelvic junction stricture, to frenular hypospadias. One patient had a small accessory spleen.

Clinical and Laboratory Features
Age at Onset of Symptoms
In 61% of our patients cyanosis was noted before they were 1 month of age; in the majority of these it was noted at birth. This is in marked contrast to other reports. Only two patients were never clinically cyanotic (the arterial O2 saturation at catheterization in these two patients was 88 and 92%, respectively).

Congestive heart failure (CHF), manifested primarily by symptoms of tachypnea, increased sweating, and poor feeding, was present in 41% under 1 month of age. All patients with PAH had CHF, and all but one was cyanotic (table 4). One third of the children without PAH escaped CHF. All but

Table 3
Sex Incidence of Proven Total Anomalous Pulmonary Venous Connection at Various Sites: 75 Patients

<table>
<thead>
<tr>
<th>Type</th>
<th>Males</th>
<th>Females</th>
<th>Significance of male predominance</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>Supracardiac</td>
<td>26</td>
<td>72</td>
<td>10</td>
</tr>
<tr>
<td>Cardiac</td>
<td>6</td>
<td>35</td>
<td>11</td>
</tr>
<tr>
<td>Subdiaphragmatic</td>
<td>12</td>
<td>75</td>
<td>4</td>
</tr>
<tr>
<td>Mixed</td>
<td>5</td>
<td>83</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>49</td>
<td>65</td>
<td>26</td>
</tr>
</tbody>
</table>

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Table 4
Incidence of Cyanosis and Congestive Heart Failure by Age of Patients

<table>
<thead>
<tr>
<th></th>
<th>Cyanosis (73/75)</th>
<th>CHF (68/75)</th>
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<tbody>
<tr>
<td></td>
<td>PAH (53)</td>
<td>PAH (22)</td>
</tr>
<tr>
<td>&lt;1 mo</td>
<td>36</td>
<td>10</td>
</tr>
<tr>
<td>1-3 mo</td>
<td>11</td>
<td>2</td>
</tr>
<tr>
<td>3 mo-1 yr</td>
<td>4</td>
<td>7</td>
</tr>
<tr>
<td>&gt;1 yr</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>52</td>
<td>21</td>
</tr>
</tbody>
</table>

one was cyanotic. The appearance of cyanosis within the first month suggests PAH as does CHF under 3 months.

Physical Findings

The birth weight of our patients ranged from 3 to 9 lbs 11 oz. The average birth weight of males was 7 lbs while that of females was 6 lbs 13 oz, both at the 25th percentile. Nine patients (12%) weighed less than 2,500 g at birth.

All the babies who were seen before they were 1 year of age were scrawny. Most had gained little weight since birth. Of the 39 who survived to 3 months of age, only five, all of them without significant PAH, were above the 10th percentile for weight.

Chest deformities were rare in the infant group, although the right ventricular impulse was hyperkinetic in the majority. Pulmonary rales were noted in 14 patients, all with PAH. Forty-four of the 53 patients with significant PAH, irrespective of the drainage site, had hepatomegaly (liver extended more than 3 cm below the right costal margin). More than half of the non-PAH group also had some enlargement of the liver. The femoral pulses were normal in all but those in extremis.

The majority of these patients were examined by one of us (A.S.N.). The auscultatory findings described below represent the findings of the most senior cardiologist who examined each patient. The first heart sound was usually well audible, often loud. Sixty-two patients had adequate descriptions of the second sound which was split in all but four. Normal respiratory variation of the second sound was noted in 52. An accentuated pulmonary component was noted in most with PAH. The presence of a loud third sound in 43 and a fourth sound in 24 of our patients contributed, together with the split second sound, to the clinical impression of a quadruple rhythm. It should be emphasized that a fourth sound was rarely heard in patients without PAH.

All the patients without PAH had systolic ejection murmurs (grade II to IV/VI in intensity); 10 of the 53 with PAH, all with pulmonary venous obstruction, had no systolic murmur. Nineteen of 22 patients with relatively normal pulmonary artery pressure and 20 of 53 with significant PAH had mid-diastolic-presystolic flow rumbles. We suspect that these rumbles represent increased flow across the tricuspid valve. Fifteen patients, four of them with subdiaphragmatic drainage, had continuous murmurs over the anomalous venous trunk.

Electrocardiographic Findings

Right axis deviation is a characteristic finding regardless of age at initial evaluation or the site of drainage. Seventy-four of our 75 patients had a frontal plane axis between +90° and +210° with an average of +140°. The remaining patient with TAPVR to the portal vein had a type B Wolff-Parkinson-White conduction pattern with a QRS axis of -120°. A P-wave axis between 0° and +90° was present in 74 patients with an average axis of +60°. The remaining patient with drainage to the RA had an ectopic atrial pacemaker with a P axis of -90°.

Only three of 22 patients (including 13 with subdiaphragmatic drainage) with complete electrocardiograms prior to 1 month of age had right atrial hypertrophy (RAH) (peaked P waves of 3 mm or more in lead II, V1R, or V1). By contrast, of the 23 patients who had electrocardiograms while between 1 and 3 months of age, 17 had RAH. Of the remaining 30 patients, 3 months of age and older, 27 had RAH. In the majority of instances in which serial electrocardiograms were obtained, RAH generally developed in the first 3 months of life.
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Figure 1

G. S. TAPVC to the LSVC at age 3 days showing "snowman" appearance.

Right ventricular hypertrophy (RVH) was present in all patients, either by voltage criteria (R in VqR or V1 greater than normal for age, or S in Vq greater than normal for age), by persistence of an upright T wave in V1 beyond the first week of life, or because of complete reversal of R/S progression in the precordial leads. Complete R/S reversal was present in 67 of our 75 cases; in the remaining eight, there was partial reversal. There has been discussion in the literature about the diagnostic significance of a qR pattern in VqR or V1. This finding was present in 30 of our 75 cases, in patients with and without PAH and in patients with each site of connection.

Disturbances of conduction are rare. Two of our patients had first degree A-V block (prior to the use of digitalis), one had type B Wolff-Parkinson-White conduction, and another developed a complete right bundle-branch block at age 3.

Radiologic Findings

All but two of the patients without significant PAH had at least moderate cardiac enlargement. By contrast 20 of the 53 patients with PAH, all with PVO, including 12 with portal vein drainage, had hearts of normal size. Pulmonary venous congestion alone or associated with active pulmonary vascular engorgement was observed in 48 of the 53 with PAH and 13 of the 22 without PAH. All the others showed pure active congestion.

Recognition of the site of drainage by plain films and fluoroscopy is possible in cases with LSVC drainage (via the innominate) and to the azygos. Those with LSVC drainage are difficult to recognize under 4 months of age, but we have seen one patient whose "snowman" appearance was recognized at 3 days of age (fig. 1). RSVC insertion may be suspected within the first few weeks of life by blurring of the RSVC-RA junction.

Patients with severe pulmonary venous obstruction can be recognized by marked passive congestion giving rise to a ground glass appearance on chest film. We know that this picture, with a normal-sized heart, is compatible with obstructive drainage at any level, not only subdiaphragmatic (figs. 2 to 4).

Cardiac Catheterization

Sixty-one patients underwent a total of 69 preoperative cardiac catheterizations.

Saturations. The relationship of simultaneous pulmonary arterial (PA) saturation and systemic arterial (SA) saturation at catheterization is presented in table 5. It is important
to recognize that pulmonary artery saturation and systemic artery saturation in TAPVC may not be equal (in our most extreme case the PA saturation was 90% compared with a systemic saturation of 60%). PA saturation was lower than SA saturation in almost all cases with portal vein drainage and significantly higher than SA saturation in all with mixed drainage sites. Other drainage sites may have PA saturations equal to, or in a significant percentage, higher than SA saturation. This is a reflection of whether the venous stream is...
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Figure 4

B. g. B. Obstructed TAPVC to the RSVC at age 5 days.

directed toward the tricuspid valve or across the atrial septum.

The presence or absence of PAH is reflected in the O₂ saturation to the extent that the SA saturations among the group with PAH averaged 75% (23 to 92%), whereas among those with normal PA pressure it averaged 86% (69 to 93%).

Pressures. There was no difference between the average RA pressures of the groups with and without significant PAH. Both averaged 5.8 mm Hg with a range of 1 to 14 mm.

Thirty-nine (38 infants and one child) of the 61 patients catheterized had significant PAH. In 15 the PA pressure was higher than the SA pressure (in two it was double the SA pressure), in 22 others it ranged from half to systemic levels, in one child PA pressure equalled systemic pressure, and in the remaining patient, the SA pressure was not measured at catheterization. None of the patients with significant PAH had a pressure differential of greater than 12 mm Hg across the RV outflow tract.

There were 22 patients (17 infants and five children) without significant PAH and 18 of

Table 5

<table>
<thead>
<tr>
<th>Site of drainage</th>
<th>Total</th>
<th>Not known</th>
<th>PA &lt; SA</th>
<th>PA = SA*</th>
<th>PA &gt; SA†</th>
</tr>
</thead>
<tbody>
<tr>
<td>LSVC</td>
<td>24</td>
<td>2</td>
<td>11</td>
<td>11</td>
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</tr>
<tr>
<td>RSVC</td>
<td>7</td>
<td>4</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Coronary sinus</td>
<td>13</td>
<td>8</td>
<td>5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>3</td>
<td>3</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mixed</td>
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<td>PV</td>
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<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>61</td>
<td>1</td>
<td>8</td>
<td>27</td>
<td>25</td>
</tr>
</tbody>
</table>

*PA saturation identical with or no more than 3% greater than SA.
†PA saturation more than 3% greater than SA.
these had PA-RV pressure differentials ranging from 6 to 45 mm Hg (mean, 21 mm Hg).

The pulmonary capillary wedge pressure or free pulmonary venous pressure was measured in 20 of the 39 patients with PAH and averaged 22.7 mm Hg (range, 10 to 34 mm Hg). In 17 of the 22 patients without significant PAH, these pressures were significantly lower with an average of 10.5 mm Hg (range, 7 to 17 mm Hg).

In 27 of the patients with PAH, a mean withdrawal tracing was recorded from left atrium to right atrium. There was no pressure difference whatsoever in seven. A difference of 2 mm Hg or less was present in 11. In only five was the right atrial mean pressure 3 mm Hg more than the left atrial mean (maximal, 7 mm Hg in one) and in four the left atrial mean pressure as measured was greater than the right atrial pressure.

Sixteen of the patients without significant PAH also had mean withdrawal tracings recorded from left to right atrium. In eight there was no difference, in another four the difference was 2 mm Hg or less, and in a single patient it was 6 mm Hg. In three the left atrial mean pressure was 1 to 2 mm Hg greater than the right atrial mean.

These data suggest that significant obstruction at the atrial level is rare. However, the presence of a large capacitance RA-venous system may modify the pressure generated by the RA. The fact that the compliance of both ventricles is similar and that they require equivalent filling pressures should also be considered in evaluating these pressure data.

Flow Ratios and Resistances. It is extremely difficult to measure the flow ratios and resistances accurately in patients with TAPVC because of the difficulty in obtaining accurate samples of mixed venous blood. This is virtually impossible in patients with supracardiac drainage and only somewhat less difficult in patients with drainage at other levels. Determinations of dye output are impractical in critically ill infants but are certainly of value in older patients.

For 19 patients in our group with PAH we felt that calculated pulmonary flows and resistances were reasonably accurate based on an adequate mixed venous sample and measurement of pulmonary capillary wedge pressure or free pulmonary venous pressure. Thirteen of these (average Qp/Qs, 1.6/1) had pulmonary venous obstruction and six (average Qp/Qs, 4.2/1) had hyperkinetic hypertension.

In 18 patients without significant PAH in whom outputs could be determined, the average Qp/Qs ratio was 3.8 to 1. It is clear from these data that patients with large pulmonary flows may or may not have PAH. The factors influencing the magnitude of pulmonary blood flow include the presence or absence of obstruction of the pulmonary outflow tract and the presence or absence of significant PVO.

Systemic flow in all three groups of our patients was within normal limits.

Table 6
Pulmonary Artery Pressure, Pulmonary Vascular Obstruction, and Pulmonary Venous Obstruction Related to Drainage Site

<table>
<thead>
<tr>
<th>PAH</th>
<th>Total</th>
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<th>Pulmonary vascular obstruction</th>
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</thead>
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<td>LSVC</td>
<td>26</td>
<td>1</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>RSVC</td>
<td>8</td>
<td>2</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>CS</td>
<td>14</td>
<td>4</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>RA</td>
<td>3</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subdiaphragmatic</td>
<td>16</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Mixed</td>
<td>8</td>
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</tr>
<tr>
<td>Total</td>
<td>75</td>
<td>1</td>
<td>22</td>
<td>9</td>
</tr>
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</table>
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Pulmonary Vascular Obstruction and Pulmonary Venous Obstruction. The vast majority of patients with PAH had pulmonary vascular obstruction (PVO) (43/52; table 6), and of those with PVO, 40 had pulmonary venous obstruction. Those with LSVC drainage most frequently had a definite narrowing of the trunk as it crossed the left bronchus. With RVSC drainage the site of obstruction uniformly occurred at the junction of the anomalous trunk with the RSVC and in one of our cases this junction was only 1 mm in diameter. One of our patients with CS drainage had stenosis of the individual pulmonary veins prior to their entrance into the CS.

Of those 16 patients with subdiaphragmatic drainage six had stenosis at the diaphragm, and 10 had stenosis as the trunk entered the portal vein or ductus venosus which was uniformly closed in our postmortem specimens. Of the three patients with PVO who did not have pulmonary venous obstruction demonstrated at autopsy, by pressure data, or by angiogram, all had PAH, low pulmonary blood flow, and marked medial and intimal arteriolar proliferation (in these cases the arterioles were described as “angiomatous”).

Angiography

Fifty-three of the 61 patients who were catheterized had either cineangiograms or biplane angiograms at the time of their initial catheterization. The angiogram was diagnostic.

Figure 5

P. C. Angiogram of patient with mixed obstructed drainage. The right upper pulmonary vein drains into the RSVC and the remainder of the pulmonary veins drain into the obstructed portal vein.
Results of Medical Management in Patients Treated in the First Year of Life According to Drainage Site and PA Pressure

![Graph showing results of medical management in patients treated in the first year of life according to drainage site and PA pressure.]

Figure 6

Results of medical management in patients treated in the first year of life according to drainage site and PA pressure.

of the drainage site in all but one patient with subdiaphragmatic drainage and supra-systemic pulmonary artery pressure because the pulmonary venous filling phase was not well seen in this one individual.

In eight of the 28 who had PVO, a discrete area of pulmonary venous obstruction was visualized. Four had a definite narrowing of the LSVC as it crossed the left bronchus, one had obstruction at the junction of the venous trunk with the RSVC, and three had a discrete area of narrowing as the trunk entered the portal vein (fig. 5). In an additional four patients, obstruction was suggested, but not localized by extremely slow filling of the anomalous trunk.

Clinical Course

The usual clinical course of patients with TAPVC is that of death within the first year of life. However, there are significant differences between the clinical courses of patients with PAH and those with relatively normal PA pressures. It is hoped that by reviewing the natural history of these patients, that some rational guide lines for the management of TAPVC can be established.

Patients with PAH and Pulmonary Vascular Obstruction

Forty-three (57%) of our patients had evidence of PAH and PVO at catheterization or autopsy, or both. Forty of these patients had pulmonary venous obstruction. These patients uniformly had evidence of early severe congestive heart failure, marked growth retardation, and cyanosis. Of the 42 with pulmonary vascular obstruction who presented before 1 year of age, 25 were treated by medical means alone (fig. 6). The majority of these 25 patients died in the first 3 months of life, and only two survived their first year.

Patients with PAH but without Pulmonary Vascular Obstruction (PVO)

Nine patients (12%) had PA systolic pressures greater than half the systemic pressure, without catheterization or postmortem evidence of PVO. Six of the nine had CS drainage; in the other three the drainage was supracardiac. CHF developed slightly later in this group than in the patients with PVO. Six were treated medically (fig. 6), one expired before the age of 3 months, and two others at 5 and 6 months, respectively. Three survived past 1 year with delicate medical management and later had surgical repair.

Patients without Significant PAH

Twenty-two patients (29%) had PA systolic pressures less than half the systemic; none had any evidence of PVO either at catheterization or postmortem examination. Seventeen were evaluated when less than 1 year of age. CHF under 3 months was present in only four and seven never had clinical heart failure. Growth retardation was less marked (five of the 17 evaluated early were above the 10th percentile for weight at 1 year). Only three expired before 1 year of age with progressive CHF, a fourth with RA drainage died at 20 months.
with failure and atelectasis. This death was the only death among patients managed medically which occurred beyond 1 year of age.

The four patients who expired with medical management had minimal gradients across the pulmonary outflow tract (mean, 8 mm Hg; range, 6 to 11 mm Hg). On the other hand 14 of the 18 survivors had a mean gradient across the pulmonary outflow tract of 21 mm Hg. In general, patients who survived longest had larger gradients across the right ventricular outflow tract. We feel that this is a significant prognostic observation in this group of patients.

Follow-Up Catheterizations in Patients without PAH

Seven patients without PAH underwent serial catheterizations at intervals of 1 to 8 years. The youngest patient was initially catheterized at 4½ months; the oldest, at 16 years. Six showed no change in the magnitude of the shunt, and no significant change in pulmonary artery pressure, resistances, or oxygen saturations at the time of the second study. One patient with CS drainage had a 15-mm Hg gradient across the right ventricular outflow tract at 17 months of age. At 9½ years this gradient was no longer present, and her pulmonary flow had increased somewhat. There was no significant change in total pulmonary resistance between the two cardiac catheterizations.

Thus patients without PAH appear to have a stable hemodynamic pattern and a comparatively benign course.

Surgical Management of Infants Under 1 Year of Age

Most reports in the literature discuss one or two operative survivors and give no indications of the overall mortality in this age group. Two groups of authors with larger series have reported mortalities with attempted total correction of 54 and 100%, respectively. Various staged procedures were devised in an attempt to improve this high mortality; however, most surgeons now favor attempted total correction as the procedure of choice. Earlier diagnosis, improved surgical technics, and more careful postoperative respiratory care will undoubtedly improve these discouraging statistics.

Discussion

The majority of patients with TAPVC in the pediatric age group will present with cyanosis and CHF within the first 3 months of life. Only an occasional patient (8% in our series) will survive to 1 year of age without significant signs or symptoms pointing to the presence of congenital heart disease.

An aggressive diagnostic approach has led to the early recognition of this defect, and at this institution more than 80% of the patients with proven TAPVC have had catheterization in the period since 1950. We now understand that PVO is an important feature of this defect and also that anatomic obstruction to pulmonary venous return can occur with anomalous return to any site. Pulmonary venous obstruction can be diagnosed by evaluation of pressure and flow data or by the actual visualization of a site of narrowing in the pulmonary venous trunk in the angiogram. We have not been able to document significant obstruction to venous return at the atrial septal level. In 40 of 43 patients with PVO a hemodynamically significant point of pulmonary venous obstruction was visualized on angiogram, proven by pressure data at catheterization, or defined at gross postmortem examination. These observations suggest that creation of an atrial septal defect (Rashkind or Blalock-Hanlon) in these patients would result in no significant hemodynamic improvement and may only postpone to a later, less favorable time, the inevitable curative surgical approach. The inherent additional hazard of systemic embolization with the use of the balloon catheter has been described elsewhere.

On the basis of our observations, we suggest the following approach to the management of this defect.

1. All patients with PAH and PVO should have an attempt at total correction on an urgent basis; and if TAPVC below the diaphragm is present, operative intervention on an emergency basis is indicated.
2. Patients with PAH without PVO should be managed medically. If decongestive measures are unsuccessful in a relatively short period of time, or if CHF suddenly worsens, operative intervention is indicated.

3. If PAH is not present, and particularly if there is a significant pressure gradient across the right ventricular outflow tract, then medical management is the treatment of choice. Operative intervention in those patients who respond to medical management can be undertaken more successfully at a later date, most optimally after the patient is 4 years of age.

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Total Anomalous Pulmonary Venous Connection: Clinical and Physiologic Observations of 75 Pediatric Patients

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