Surgical management
of total anomalous pulmonary venous drainage
via left vertical anomalous trunk

by

G. RAINNEY WILLIAMS, M.D., WEBB M. THOMPSON, JR., M.D.,
DONALD H. GARRETT, M.D. and LAZAR J. GREENFIELD, M.D.
(from the Departments of Surgery and Pediatrics, University of Oklahoma
Medical Center, Oklahoma City, Oklahoma)

The outlook for patients with total anomalous pulmonary venous drainage via a
vertical anomalous trunk (persistent left superior vena cava) has improved greatly
in the past ten years. During the development of current treatment methods, ques-
tions have been raised regarding the advisability of staged or immediate total
repair of the anomaly. Furthermore, a
number of technical methods of repair
have been described and employed. Fin-
ally, there have been few reports of long-
range follow-up. In an attempt to add
current information this communication
will record our total experience with the
surgical management of this form of
congenital heart disease.

Supported in part by the John and Mary R.
Munkle Foundation.

Material and Methods

Nine patients with total anomalous pulmonary venous drainage via a vertical anomalous trunk have been treated surgically at the University of Oklahoma Medical Center. Pertinent information regarding these patients is included in Table 1. Al-
though the diagnosis may be suspected from the PA chest film exhibiting the
"snowman" configuration of the heart and supported by electrocardiography, car-
diac catheterization and angiography have been employed in all patients
to confirm the diagnosis. In infants with
recognized distress operation has not been
delayed because our experience indicates that prolonged improvement with nonsur-
gical management is unlikely. In older

Table 1.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Date of operation</th>
<th>Age at operation</th>
<th>Weight at operation</th>
<th>Result</th>
<th>Postop. cath.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. BH 303076 WM</td>
<td>11-9-60</td>
<td>8 mos.</td>
<td>13 lbs.</td>
<td>Died 48 hrs</td>
<td>6-8-65</td>
</tr>
<tr>
<td>2. OB 301802 WF</td>
<td>1-4-61</td>
<td>2 yrs.</td>
<td>23 lbs.</td>
<td>Excellent</td>
<td>8-23-67</td>
</tr>
<tr>
<td>3. MC 324086 WF</td>
<td>7-6-62</td>
<td>2 mos.</td>
<td>6½ lbs.</td>
<td>Excellent</td>
<td>2-8-67</td>
</tr>
<tr>
<td>4. EN 329410 IF</td>
<td>12-3-62</td>
<td>3 mos.</td>
<td>8 lbs.</td>
<td>Died 12 hrs</td>
<td>10-8-67</td>
</tr>
<tr>
<td>5. VA 267559 NF</td>
<td>12-15-65</td>
<td>11 yrs.</td>
<td>52 lbs.</td>
<td>Excellent</td>
<td>9-19-67</td>
</tr>
<tr>
<td>6. EG 243068 WF</td>
<td>1-5-66</td>
<td>44 yrs.</td>
<td>116 lbs.</td>
<td>Excellent</td>
<td>9-14-67</td>
</tr>
<tr>
<td>7. DB 378380 WF</td>
<td>9-6-66</td>
<td>3 yrs.</td>
<td>20 lbs.</td>
<td>Excellent</td>
<td>9-14-67</td>
</tr>
<tr>
<td>8. BS 379325 WM</td>
<td>9-28-66</td>
<td>6 mos.</td>
<td>14 lbs.</td>
<td>Excellent</td>
<td>9-14-67</td>
</tr>
<tr>
<td>9. LA 326221 WM</td>
<td>10-7-66</td>
<td>13 yrs.</td>
<td>63 lbs.</td>
<td>Excellent</td>
<td>9-14-67</td>
</tr>
</tbody>
</table>

Cases #1 and 2 have been reported previously⁸ and cases #1, 2, 3 and 4 were mentioned in a previous publication.¹²
Surgical Management of Total Anomalous Pulmonary Venous Drainage

METHODS

Anomalous pulmonary venous return has been surgically treated in a vertical incision, allowing exposure of major thoracic structures. The position of the heart and great vessels is better visualized before the incision. The technique described in 1964 is still applicable.

The sternal splitting incision has proved quite satisfactory (Fig. 1). The patient is placed on cardiopulmonary bypass utilizing routine cannulation of the venae cavae through the right atrium and either femoral or aortic cannulation for arterial perfusion. After establishment of satisfactory cardiopulmonary bypass, the apex of the heart is lifted anteriorly and held in a vertical position with a padded retractor (Fig. 2). A ligature is passed about the vertical anomalous trunk within the pericardium. Occasionally dissection of this vessel is easier extrapericardially in which case the heart is returned to the pericardial cavity during this aspect of dissection. The common venous channel lying posterior to the parietal pericardium and just behind the left atrium is identified. A long incision is made in this common pulmonary venous chamber and as this incision is made, the ligature around the vertical anomalous vein is tightened. The procedure is done in order to prevent even transient pulmonary venous hypertension. A moderate amount of blood is aspirated, but it has not been found necessary to use a partial occluding clamp on the common pulmonary venous trunk. A corresponding transverse incision is made in the posterior surface of the left atrium. It seems important to have this incision as long as possible and, ordinarily, it extends onto the atrial appendage. Particularly in larger patients, the atrial septal defect can be visualized through this incision. We have not been sufficiently certain of the anatomical relationships to close the atrial septal defect from this distorted approach, but it seems possible to do so. An anastomosis is then performed between the left atrium and the common pulmonary venous trunk using continuous fine cardiovascular suture material (Fig. 2). The heart is returned to the pericardium, the right atrium opened through a standard right atriotomy, and the atrial septal defect closed (Fig. 3). It has not been necessary to shift the atrial septum in order to enlarge the left atrium. The persistent venous trunk is perma-
nently ligated, and the procedure terminated in the routine fashion.

All patients have been followed carefully since operation. The period of follow-up is 18 months to 7 years. Two patients who represent the age extremes in this series will be presented in some detail.

Patient #3, M.C. (U.H. 32-40-86) a two month old white female, was admitted to the Children's Memorial Hospital of the University of Oklahoma because of failure to thrive. The patient was the product of a normal pregnancy, labor and delivery. She was noted to be cyanotic at birth and remained in the hospital where she was delivered for approximately three weeks. A heart murmur was described at three days of life. After leaving the hospital, the patient continued to be slightly cyanotic when crying and did not take feedings well. At the time of admission she weighed 6½ lbs., one

![Diagram](image1)

Fig. 3.—Routine closure of ASD through right atriotomy.

![Image](image2)

Fig. 4.—Pre-(left) and 5 years post-(rt.) operative chest films on Patient #3. There is a 5 year interval between the films.
procedure terminated.
followed care period of fol-
7 years. Two
he age extremes presented in some
32-40-86) a two
was admitted
ma because of
nt was the pro-
 labor and
t to be cyanotic
hospital
approxima-
murmur was
life. After leav-
it continued to
crying and did
t the time of
& 1/2 lbs., one

Fig. 5.—Angiocardiogram of Patient #3. The
catheter is in the pulmonary artery. Pulmonary
venous filling with vertical left anomalous trunk
draining into innominate vein is well visualized.

pound less than delivery weight. On phy-
ical examination the patient was an irri-
table, emaciated white female whoappeared
to be dusky when active. There was
no gross chest abnormality. A Grade
III/IV systolic murmur was present along
the left sternal border, and a soft dia-
stolic murmur was described. The liver
was palpable 3 cm. below the right costal
margin. The spleen was not felt. Peri-
ipheral pulses were full and there was no
clubbing. The preoperative chest x-ray is
shown in Fig. 4. Electrocardiogram
showed right axis deviation and right
ventricular hypertrophy. Catheterization
and angiocardiography confirmed the diag-
nosis of TAPVD and demonstrated the
abnormal drainage route (Fig. 5). Total
 correction was carried out by the techni-
que described above. The post-operative
course was unremarkable; the patient pas-
sed the usual developmental milestones
normally, behaved, and was treated as a
normal child. She was readmitted at five
years of age, at which time she appeared
to be normally developed but remained
at the 84th percentile in both height and
weight. A soft systolic murmur was audi-
ble, and otherwise, physical examination
was within normal limits. The chest x-ray
is reproduced in Fig. 4. Cardiac cathet-
etization revealed no detectable physiologic
abnormalities.

Patient #6, E.G. (U.H. 24-30-68) a 44
year old white female, was admitted to
the University of Oklahoma Hospitals for
correction of previously proven total ano-
malous pulmonary venous drainage. The
patient was told she had rheumatic heart
disease at 17 years of age. She had inter-
mittent episodes of cardiac failure and on
the basis of her chest x-ray was told she
had an aortic aneurysm at 27 years of
age. She led a reasonably active life but was troubled frequently by chest pain and by exertional dyspnea. A single pregnancy was terminated by caesarean section because of cardiac difficulty. Several catheterizations were performed, and the diagnosis of anomalous pulmonary venous return was first made when the patient was 34 years of age.

On physical examination the patient was a well developed, well nourished white female. There was slight left chest deformity and a parasternal lift was present. A Grade III blowing systolic murmur was audible over the precordium. The liver was not palpable and the remainder of the physical examination was unremarkable. The chest x-ray is reproduced in Fig. 6. Electrocardiogram showed incomplete right bundle branch block with RVH. The previous cardiac catheterization and angiocardio gram were reviewed and the diagnosis of TAPVD via a persistent left superior cava was obvious. The operation was carried out exactly as described previously. The anastomosis was approximately 4 cm. in length and the time of total bypass was 80 minutes. The postoperative condition of the patient was entirely unremarkable. 15 months after the operation she was admitted for elective cholecystectomy. Her exercise tolerance became normal, and she had no further signs or symptoms of cardiac failure. At the time of readmission for cardiac catheterization 18 months after operation she was asymptomatic. The postoperative chest x-ray is shown in Fig. 7. There was no audible cardiac murmur and cardiac catheterization revealed all values to be within normal limits.

RESULTS

All of the nine patients survived operation but two died in the immediate postoperative period. There have been no late deaths. The first death occurred in the first patient in whom operation was carried out. This was an eight month old white male who died 48 hours after an initial satisfactory course. The cause of death was interstitial pneumonitis and the repair appeared to be anatomically adequate at the time of autopsy. The second death occurred in Patient #4, a three month old female who died 12 hours after operation in cardiorespiratory failure for which no cause was obvious at autopsy. The course in surviving patients was highly satisfactory although close attention to pulmonary problems was required in the immediate postoperative period. Resumption of normal weight gain and growth was observed in infants; functional activity levels improved in older children and adults. Representative pre- and postoperative chest x-rays are reproduced in Figs. 8 and 9. Postoperative cardiac catheterization was carried out on all surviving patients except the most recent two and the findings are
iously. The anat-
ely 4 cm. in
total bypass was
tive condition
unremarkable.
ration she was
holecystectomy.
became normal,
as or symptoms
ime of readmis-
ion 18 months
asymptomatic.
x-ray is shown
 audible cardiac
etization reveals
ormal limits.

Fig. 8.—Patient #7, pre-(rt.) and 1 year postoperative chest films.

Table 3 shows the immediate occurrences and the survival rates of patients. The incidence of death occurred among patients after an eight month period. The cause of death was neumonitis and sepsis. The patient #4, who died 12 hours postoperatively, was one of the patients who continued to improve postoperatively. The chest x-rays are normal.

The condition of total anomalous pulmonary venous drainage proposed by Darling has found wide acceptance and this classification is reproduced in Table 2. Patients under discussion in this paper fall entirely into Group 1.A. The condition has been termed total anomalous pulmonary venous drainage by a per-

Tabulated in Table 3. There is no evidence of shunting and when pulmonary wedge pressures have been measured, they are normal. Right ventricular pressures have returned to normal.

DISCUSSION

The classification of total anomalous pulmonary venous drainage proposed by Darling has found wide acceptance and this classification is reproduced in Table 2. Patients under discussion in this paper fall entirely into Group 1.A. The condition has been termed total anomalous pulmonary venous drainage by a per-

Table 2.—Darling’s classification.

1. Supracardiac (supradiaphragmatic)
   A. Left superior cava (48%)
   B. Right superior cava (9%)
2. Cardiac level (supradiaphragmatic)
   A. Coronary sinus (14%)
   B. Right atrium (18%)
3. Infracardiac (infradiaphragmatic)
   (9%)
4. Mixed (27%)

Fig. 9.—Angiocardiogram of Patient #7 showing common venous trunk and vertical trunk emptying into innominate vein.

sistent left superior vena cava or via an anomalous vertical trunk. Use of the term “persistent left superior vena cava” is open to criticism as this is not strictly
TABLE 3.—Catheterization findings.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at</th>
<th>SVC sat.</th>
<th>RV pres.</th>
<th>Pul. wedge</th>
<th>Shunt post</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. BH</td>
<td>8 mos.</td>
<td>Op. death</td>
<td>92</td>
<td></td>
<td>66</td>
</tr>
<tr>
<td>2. OB</td>
<td>2 yrs.</td>
<td>7 yrs.</td>
<td>96</td>
<td>62</td>
<td>30</td>
</tr>
<tr>
<td>3. MC</td>
<td>2 mos.</td>
<td>5 yrs.</td>
<td>97.8</td>
<td>75</td>
<td>50</td>
</tr>
<tr>
<td>4. EN*</td>
<td>3 mos.</td>
<td>Op. death</td>
<td>89</td>
<td>83</td>
<td>70</td>
</tr>
<tr>
<td>5. VA</td>
<td>11 yrs.</td>
<td>13 yrs.</td>
<td>92</td>
<td>77</td>
<td>52</td>
</tr>
<tr>
<td>6. EG</td>
<td>44 yrs.</td>
<td>46 yrs.</td>
<td>90</td>
<td>.76</td>
<td>62</td>
</tr>
<tr>
<td>7. DB</td>
<td>3 yrs.</td>
<td>4 yrs.</td>
<td>90</td>
<td>76</td>
<td>62</td>
</tr>
<tr>
<td>8. BS**</td>
<td>6 mos.</td>
<td>not done</td>
<td>90</td>
<td>68</td>
<td>68</td>
</tr>
<tr>
<td>9. LA</td>
<td>13 yrs.</td>
<td>not done</td>
<td>88</td>
<td></td>
<td>63</td>
</tr>
</tbody>
</table>

*Only angiocardiology done.
**Preoperative data from Children’s Medical Center, Dallas, Texas.

Patients #8 and 9 have not had postoperative catheterization.

embryologically accurate. For brevity, the condition is sometimes referred to only as “supracardiac total anomalous venous drainage” or the “snowman” type deformity. Surgical management of this particular sub-type of total anomalous pulmonary venous drainage was first performed by Muller who constructed an anastomosis between the left atrial appendage and the common pulmonary venous trunk from outside the heart without using cardiopulmonary bypass. Similar operations were described by Mustard. Ultimately, Senning performed complete correction of this defect in an older patient using an anastomosis between the left atrium and the common pulmonary trunk, ligating the pulmonary trunk and subsequently closing the atrial septal defect by a closed technique.

The first successful one-stage total correction of this anomaly using cardiopulmonary bypass was reported by Cooley. The technique described by Cooley consisted of opening the right atrium, incising the atrial septum, opening the posterior wall of the left atrium, and performing the anastomosis with the common pulmonary vein from an intracardiac exposure. It is a credit to Cooley and his associates that successful repair in infants could be performed using this technique. The approach was modified by Shumacker to allow an easier posterior anastomosis but the internal route was preserved. The use of cardiopulmonary bypass and an external approach for the posterior anastomosis and subsequently, complete correction of the anomaly was described in 1964. This procedure appears to be commonly employed at the present time and has the obvious advantages of allowing better exposure and a larger anastomosis, particularly in very small infants.

The question of performing a one-stage total correction or of leaving either the vertical anomalous trunk or atrial septal defect open for a second stage has been raised, particularly by Mustard. A two-stage operation would be necessary if the left side of the heart is not capable of resumption of adequate systemic output. Autopsy examination of the hearts of infants dying with this anomaly indeed reveal a very large right ventricle and a seemingly small and thin-walled left ventricle. This question is not completely answered, but our experience and that of Cooley's group suggests that this approach is reasonable.
of Cooley and others would suggest that the left side of the heart is adequate to support systemic output and that one-stage total correction is the procedure of choice.\textsuperscript{11}

The fate of vascular anastomoses performed in growing individuals has been studied and discussed at length.\textsuperscript{3} The incidence of late failures of such operations as the subclavian pulmonary anastomosis (Blalock) when performed in small infants is clear evidence that originally adequate anastomoses do not always grow at the same rate as the individual. Since adequacy of repair of total anomalous pulmonary venous drainage via a vertical anomalous trunk is dependent upon a circular anastomosis in a low pressure system frequently performed in very small infants, this question is of great importance. The data presented indicating normal postoperative hemodynamics, particularly pulmonary arterial wedge pressures, over the period of observation are highly encouraging. Observation of a larger number of patients over longer periods of time, of course, is necessary to make final observations of the adequacy of this technical repair.

**CONCLUSIONS**

Nine patients with total anomalous pulmonary venous drainage via a vertical anomalous trunk have been treated surgically at the University of Oklahoma Medical Center. One-stage total correction using cardiopulmonary bypass and a posterior anastomosis performed from outside the heart is technically feasible at any age.

Seven of the nine patients survived operation and have been followed from 18 months to 5 years. Clinical improvement has been marked in all survivors. Cardiac catheterizations from 1 to 5 years after operation indicate excellent hemodynamic results.

**REFERENCES**


