Total Anomalous Pulmonary Venous Drainage

Results of Open-Heart Correction in Four Infants

During the past 10 years, both diagnosis and surgical treatment of many forms of congenital heart disease have become simplified and are performed routinely and successfully in many centers. Problems associated with management of the less common, more complex cardiac anomalies are, however, still far from solved. One of the most interesting, relatively less common, groups are the patients with total anomalous pulmonary venous drainage. Without treatment, the majority of these patients die in infancy. The lesions appear to be anatomically correctable, and yet the first few years of surgical experience with such patients have been disappointing.

The purposes of this paper are to present 4 infants with total anomalous pulmonary venous return treated surgically, to review the reported surgical results, and to discuss the present status of surgical treatment of this group of patients.

Report of Cases

CASE 1.—This 6-month-old white female was admitted to the Children's Memorial Hospital, University of Oklahoma Medical Center, on Nov. 1, 1959, because of a heart murmur of 3 months. The child was full-term pregnancy, compli-
ANOMALOUS PULMONARY VENOUS DRAINAGE

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minimum, BP (flush) 105 mm. Hg (arms and legs). The patient was a small, poorly nourished, slightly cyanotic white female in no acute distress. There was tachypnea, but the lungs were clear to auscultation and percussion. The precordium was hyperdynamic with a lifting xiphoid impulse. The first heart sound was of slightly increased intensity and was maximal at the apex. The second heart sound was of increased intensity, maximal at the upper left sternal border, and not split. A third heart sound and a Grade 1-2 mid-diastolic rumble were heard at the lower left sternal border. There was a Grade 3, holosystolic blowing murmur at the upper left sternal border. There were no thrills. The liver was palpable 3 cm. below the right costal margin.

Laboratory Data.—Routine blood cell count and urinalysis were normal. Chest radiogram (Fig. 1) revealed moderate cardiac enlargement with a globular cardiac configuration and pulmonary plethora. The electrocardiogram (Fig. 2) revealed right axis deviation, right atrial hypertrophy, incomplete right bundle branch block, and right ventricular hypertrophy. Cardiac catheterization (Table 1) and angiograms (Fig. 3) led to a diagnosis of total anomalous pulmonary venous drainage with all pulmonary veins entering the right atrium via the coronary sinus plus a moderate-sized atrial septal defect.

Clinical Course.—The patient improved slightly after digitalization and was discharged to be followed in the Pediatric Cardiac Clinic. Because of continued growth retardation and progressive cardiac enlargement (Fig. 1), she was readmitted at 1 year of age for corrective surgery. On June 6, 1960, operation was performed with the use of extracorporeal circulation with total cardiopulmonary bypass as previously described. The heart was hyperdynamic, and the right atrium was markedly enlarged. There was a large diameter (2 cm.) fossa ovalis atrial septal defect. The coronary sinus opening was approximately the same size. The septum between the coronary sinus and atrial septal defect was excised and the free edge of the atrial septal defect sutured to the right atrial wall, diverting coronary sinus flow into the left atrium. Total operating time was 2 hours; perfusion time was 20 minutes (Table 2). Her immediate postoperative course was uneventful, and she was discharged on the 19th postoperative day. She gained 7 lb. in the next 5 months. At present there is no cyanosis, no murmur, and no residual electrocardiographic evidence of right ventricular hypertrophy (Fig. 2). Her cardiac size has gradually diminished (Fig. 1).

CASE 2.—This 6½-month-old white male was referred to the Children’s Memorial Hospital, University of Oklahoma Medical Center, because of a heart murmur. He was the product of an uncomplicated pregnancy, labor, and delivery. He had

Fig. 1.—Roentgen findings in Case 1 showing progressive cardiac enlargement from 5 to 10 months of age and return toward normal 7 months after surgery. Pulmonary venous drainage was into the coronary sinus in this patient (Type 2, A). A, 5 months, preoperative; B, 10 months, preoperative; C, 18 months, postoperative.

1959, because of a heart murmur noted first at the age of 3 months. The child was the product of a full-term pregnancy complicated by influenza during the first trimester. There was no history of cyanosis. She was seen by her family physician at 3 months of age because of her poor weight gain and was found to have tachycardia, heart murmur, and moderate cardiac enlargement.

Physical Examination.—Temperature was 98.6° F., pulse rate 168 per minute, respiration rate 40 per
grown slowly and had been noted to have tachypnea and intermittent minimal cyanosis.

Phyisical Examination.—Temperature was 98.6°F; pulse rate, 144 per minute; respiration rate, 60 per minute; BP (flush) 70 (arms), 90 (legs). The patient was a poorly nourished white male in no acute distress who did not appear cyanotic at rest. He was tachyemic, but the lungs were clear to auscultation and percussion. There was a left precordial bulge and a hyperdynamic xiphoid impulse. The first sound was of increased intensity, maximal at the apex. The second sound was increased in intensity and heard best at the upper left sternal border. A Grade 3 systolic murmur was heard at the upper left sternal border. A Grade 2 middiastolic murmur was heard at the lower left sternal border and apex. Peripheral pulses were strong and equal bilaterally. The liver was palpable 4 cm. below the right costal margin.

Laboratory Data.—Complete blood cell count and urinalysis were normal. Chest roentgenogram revealed marked cardiac enlargement with a "snowman" configuration and marked pulmonary plethora. The electrocardiogram revealed right axis deviation, right atrial hypertrophy, and right ventricular hypertrophy, Cardiac catheter angiograms led to total anomalous pulmonary the pulmonary veins drain the superior vena cava; (2) at durr type, moderate-to-large artery hypertension with lary vascular resistance; venous obstruction.

Clinical Course.—The pig change significantly after Nov. 11, 1950, operation in use of extracorporeal circuits from both lungs were on the neck which commun left superior vena cava' and entered the right superior vena cava. After tracorporeal circulation, the pericardium and the pulmonary veins trun atrium, and it was possible to open and repair the septal defect. The heart was returned to right atrium opened. It was easily visualized through the atrial wall, the left superior vena cava was opened and filling of the left superior vena cava. The right atrial vein was opened.

Fig. 2.—Pre- and postoperative electrocardiographic findings in Cases 1 and 4. The postoperative tracings in both patients show less right axis deviation and the development of left ventricular dominance (paper speed is 75 mm. per second).

Fig. 3.—Angiogram from Case 1 showing right-to-left streaming of the contrast material across an atrial septal defect after a low right atrial injection.

<table>
<thead>
<tr>
<th>Case</th>
<th>Type*</th>
<th>TAPVD</th>
<th>Qp/Qs</th>
<th>R-L (% Qs)</th>
<th>PA</th>
<th>SA</th>
<th>FY</th>
<th>PA</th>
<th>SA</th>
<th>Elevation</th>
<th>Systolic Pressure</th>
<th>PVR (Units)</th>
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<td>29</td>
<td>90</td>
<td>81</td>
<td>61</td>
<td>67</td>
<td>42</td>
<td>30</td>
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<td>1A</td>
<td>5/2</td>
<td>29</td>
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<td>94</td>
<td>90</td>
<td>20</td>
<td>1.0</td>
<td>4.0</td>
<td>7 RA angio showed filling of PV trunk</td>
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<tr>
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<td>3/1</td>
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<td>62</td>
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<td>92</td>
<td>2.0</td>
<td>4.0</td>
<td>28 RA angio showed filling of PV trunk</td>
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<td>97</td>
<td>90</td>
<td>30</td>
<td>0.3</td>
<td>0.8</td>
<td>8 Left SVC angio showed retrograde filling of the left arm, veins</td>
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</table>

* Darling's classification.7

Legend: L.V. indicates left ventricle; PA, pulmonary artery; Pre Cop., pre-capillary; PV, pulmonary vein; PVR, pulmonary vascular resistance; Qp, pulmonary flow; Qs, systemic flow; RA, right atrium; R-L, right-to-left shunt; RV, right ventricle; SA, systemic artery; SV, systemic vein; Sys., systolic; TAPVD, total anomalous pulmonary venous drainage.
The patient's status was evaluated daily after the operation, and the pulmonary vasodilator was continued. The patient was discharged from the hospital on Nov. 11, 1960, with the diagnosis of an anomalous pulmonary venous connection, type moderate-to-large size. The patient was asymptomatic at the time of discharge. The postoperative course was uneventful, and the patient was discharged in good condition.

Case 4—The 15-year-old male was admitted to the Children's Memorial Hospital, Chicago, Ill., on Oct. 1, 1960, with the diagnosis of an anomalous pulmonary venous connection, type moderate-to-large size. The patient was asymptomatic at the time of admission. The postoperative course was uneventful, and the patient was discharged in good condition.

Table 2—Surgical Data and Results

<table>
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<tr>
<th>Patient No.</th>
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<td>PA STTO</td>
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<tr>
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<td>18</td>
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<td>12 years</td>
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<td>PA STTO</td>
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<tr>
<td>3</td>
<td>19</td>
<td>M</td>
<td>14 years</td>
<td>AVJMA</td>
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<td>4</td>
<td>20</td>
<td>M</td>
<td>15 years</td>
<td>AVJMA</td>
<td>PA STTO</td>
</tr>
</tbody>
</table>

Legend: AVJMA—atrioventricular septal defect, AV—atrioventricular, ST—ventricular septal defect, TO—transposition of the great arteries.
directly into the right atrium; (2) atrial septal defect, large; (3) pulmonary artery hypertension with normal pulmonary precapillary vascular resistance; (4) moderate pulmonary venous obstruction.

Clinical Course.—In spite of medical therapy the patient's course in the hospital was one of steady deterioration. Surgical correction of his anomaly was undertaken on Nov. 21, 1960. At operation marked enlargement of the right atrium and right ventricle was noted. The right atrium was opened, and 2 septal defects were noted. The most cephalic opening was 1 cm. in diameter and represented the atrial septal defect. The caudal opening, which was 2 cm. in diameter, was thought to be the ostium of the pulmonary venous trunk. The septum between these 2 openings was excised. The margin of the atrial septal defect was then sutured to the right atrial wall anterior to the opening of the pulmonary venous trunk. During the procedure the patient developed complete heart block. An internal pacemaker was implanted. The right ventricular systolic pressure was 20 mm. Hg after surgery. Saturation data indicated the presence of a persistent small left-to-right shunt. Operative time was 2 hours, 45 minutes, and perfusion time was 44 minutes (Table 2). The patient's immediate postoperative course was satisfactory. His heart rate was sustained between 100 and 120 by the pacemaker. He showed no evidence of congestive heart failure. His condition remained satisfactory until the third postoperative day when he had an apneic episode during a feeding. He responded to resuscitative measures but after this was noted to have crepitant rales over the right lung posteriorly. On the fourth postoperative day, dyspnea became more severe and a chest radiogram revealed a right upper lobe infiltrate. A tracheostomy was performed. A precordial isotope dilution curve revealed evidence of a large left-to-right shunt. On the fifth postoperative day he developed sudden cardiac arrest and died despite resuscitative measures. Postmortem examination revealed: (1) total anomalous pulmonary venous drainage into the right atrium via the coronary sinus, incompletely repaired; (2) atrial septal defect; (3) possible hypoplasia of left atrium and ventricle; (4) pulmonary atelectasis, bilateral.

At the time of operation it was erroneously thought that the pulmonary veins entered the right atrium separately from the coronary sinus; therefore, the entire coronary sinus flow had not been redirected into the left atrium.

Case 4.—This 2-year-old white female was admitted to the Children's Memorial Hospital, University of Oklahoma Medical Center, on Aug. 29, 1960, because of syncope and cyanosis on exertion. The child was the product of a full-term, uncomplicated pregnancy, labor, and delivery. She did well until 9 months of age when she had a sudden acute episode of syncope associated with cyanosis. During the next 12 months she had several similar episodes usually associated with exertion. She developed marked exertional dyspnea and tired easily. Her growth and development were poor, and she had recently lost weight.

Physical Examination.—Temperature was 98.8 F; pulse rate, 90 per minute; respiration rate, 24 per minute; BP 70/50 mm. Hg (arms), 80/60 mm. Hg (left leg). The patient was a poorly developed and nourished 2-year-old white female who appeared chronically ill. There was slight cyanosis and early clubbing of the fingers. There was a lifting xyploïd cardiac impulse. The first heart sound was slightly increased in intensity and was maximal at the apex. The second heart sound was split, of increased intensity, and maximal at the upper left sternal border. A third heart sound was present. A 2 mid-diastolic rattle was noted over the left sternal border.

Laboratory Data.—C. Urinalysis was normal. Moderate cardiac enlargement. The cardiac silhouette was an enlarged right atrium and right ventricle. The right atrial and right ventricular catheterization (Fig. 4) were performed with the patient in the supine position. The right atrial pressure was 12 mm. Hg, and the right ventricular pressure was 30/10 mm. Hg. The pulmonary artery pressure was 22/12 mm. Hg. The pulmonary capillary wedge pressure was 12 mm. Hg. The electrocardiogram showed right axis deviation and right atrial hypertrophy.

Clinical Course.—The patient was transferred to the operating room on Jan. 4, 1961, under intracorporeal circulation bypass. The pulmonary venous drainage was then anastomosed to the left atrium. The operation was completed successfully.

Fig. 4 (Case 4 [Type 1, A]) A, showing the typical “snowman” configuration radiogram of the pulmonary veins. The aorta is outlined by contrast material. B, the angiographic study of the pulmonary veins. The normal pulmonary veins drain into the left atrium. The abnormal drainage into the right atrium is seen through the coronary sinus.
revealed evidence of a large left-to-right shunt postoperatively. He developed arrhythmias and died despite resuscitative efforts.

Postmortem examination revealed: (1) a malacic pulmonary venous drainage into atrium via the coronary sinus, incompletely obliterating it; (2) atrial septal defect; (3) possible aortic stenosis and ventricular hypertrophy; (4) pulmonary stenosis, bilateral.

The patient was a 2-year-old white female who appeared ill. There was slight cyanosis and early apical gallop. There was a soft systolic murmur. The first heart sound was slightly split and maximal at the apex. The second heart sound was split, of increased intensity, and maximal at the lower left sternal border. A third heart sound was heard at the lower left sternal border. A Grade 3 systolic blowing murmur was present at the base, and a Grade 2 mid-diastolic rumble was heard at the lower left sternal border.

Laboratory Data.—Complete blood cell count and urinalysis were normal. Chest radiogram revealed moderate cardiac enlargement and pulmonary plethora. The cardiac silhouette exhibited a typical "snowman" configuration (Fig. 4). The electrocardiogram (Fig. 2) revealed right axis deviation, right atrial and ventricular hypertrophy. Cardiac catheterization (Table 1) and angiograms (Fig. 4) were performed, and the conclusions reached were: (1) total anomalous pulmonary venous drainage with the pulmonary veins draining into a persistent left superior vena cava; (2) atrial septal defect, moderate to large size; (3) slight pulmonary venous obstruction.

Clinical Course.—The patient was digitalized on Dec. 1, 1961. He underwent operation utilizing extracorporeal circulation with total cardiopulmonary bypass. The pulmonary veins were noted to join posteriorly as a large trunk which passed upward into a persistent left superior vena cava to reach the left innominate vein. The left atrium appeared small. After the establishment of cardiopulmonary bypass, the left superior vena cava was occluded by a tape. The heart was rotated out of the pericardium and held in place by the apex. The apex was then excised, and the heart was exposed as a whole. The pulmonary veins were then anastomosed to the left atrium, and a 1.5 cm. anastomosis was carried out. Marked electrocardiographic changes occurred during dislocation of the heart (Fig. 5) with a rapid return to normal after replacement. An incision was then made in the right atrium. Through the 1.5 cm. atrial septal defect, the new anastomosis of the pulmonary venous channel to the left atrium was easily visualized and appeared to be widely patent and of sufficient size. The atrial septal defect and atrioventricular canal were closed. After the termination of perfusion, the heart beat was vigorous, and the systemic pressure returned to the preoperative level. After loosening of the tape occluding the left superior vena cava, the heart dilated and the systolic pressure fell. The left superior vena cava was again ligated, with prompt return of normal cardiac action. The total operating time was 3 hours and 15 minutes.

Comment

The 4 cases of total anomalous pulmonary venous drainage in infants presented represent approximately 0.4% of the total number of patients with congenital heart disease seen at the Children's Memorial Hospital during the past 3 years. In a review of the patients studied in cardiac catheterization laboratory, 4% of the children who have had open-heart surgery. In addition to these cases, our experience with infants and children with total anomalous pulmonary venous drainage has included: (1) a 6-month-old male who died in congestive heart failure; (2) a 6-year-old male in severe chronic congestive heart failure, whose parents refused operation; (3) a 10-year-old male with Eisenmenger physiology; (4) a male with associated severe tetralogy of Fallot who died at 4 weeks of age; (5) a male with total pulmonary venous drainage into the portal veins, dextrocardia, transposition of the great vessels, and cor pulmonale, who died at 15 weeks of age (Fig. 6); (6) a male with total pulmonary venous drainage into the portal vein who died at age 10 weeks. We have also performed successful atrial septoplasty on a 12-year-old girl with all the features of the left superior vena cava draining anomalously.

Brodys' case, for which the tracing of his description of total anomalous pulmonary venous return and collected a series of 37 autopsy cases. Darling et al., in 1957, pre-
Fig. 6.—Angiocardiogram of a patient with total anomalous pulmonary venous drainage below the diaphragm into the portal circulation. This patient also had dextrocardia, transposition of the great vessels, and cor bicornuare. The arrows outline the pulmonary and portal veins.

Presented data from 43 additional cases and suggested the classification listed below:

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

The figures given in parenthesis are the incidences of the various types determined by Guntheroth et al. in a review of 159 reported cases.

For clinical purposes total anomalous pulmonary venous drainage cases may be grouped simply as supracardiac and infradiaphragmatic based on drainage of the veins above or below the diaphragm. The prognosis for both groups without surgical correction is quite poor. Approximately 85% of patients with the supradiaphragmatic type died before 2 years. Patients with pulmonary venous drainage below the diaphragm do even more poorly; none has survived early infancy. Of the 31 patients reported by Guntheroth et al., only 4 (12%) drained below the diaphragm, and all had died by 7 weeks. None of the 12 patients with supradiaphragmatic drainage died this early. The typical clinical picture of the infant with infradiaphragmatic drainage is an early onset of congestive heart failure with marked hepatomegaly, minimal or no cardiac enlargement on x-ray, and the roentgen appearance of passive, not active, pulmonary congestion. We know of only one reported case of the infradiaphragmatic type having had successful surgical correction.

The typical clinical picture associated with supradiaphragmatic total anomalous pulmonary venous drainage has been well outlined by DuShane, Keith et al., and Guntheroth et al., and the physiological changes have been discussed by Burchell, Friedlich et al., and Swan et al. An associated communication between the pulmonary and systemic circulation is necessary for survival in patients with total anomalous pulmonary venous drainage. Based on Brody’s original classification, an atrial septal defect is not considered as a complication but as a necessary part of the anomaly. Variations in the clinical picture, hemodynamics, and prognosis depend on several factors including:

1. The size and number of communications between the systemic and pulmonary circulation—generally the smaller the atrial communication, the lower is the systemic flow and the poorer the prognosis.
2. The adequacy of the left atrium and left ventricle—we believe true left-sided hypoplasia contributed to the unsuccessful result in Case 3. It is possible that hypoplasia is more common in patients with the infradiaphragmatic drainage. Angiocardiograms may be helpful in the preoperative assessments of the left atrium and left ventricle (Fig. 3).
3. The presence of precapillary pulmonary vascular obstruction—we have seen one 10-year-old boy with total anomalous pulmonary venous drainage (Type 1, A) with Eisenmenger physiology and a pulmonary-to-systemic flow ratio of less than one. At the present time this type patient is not a surgical candidate.
4. Pulmonary venous obstruction—this is usually caused by stenosis of the pulmonary and systemic venous connection or by a long, relatively small tubular connection between the pulmonary and systemic venous circuits. Case 3 showed a marked pressure gradient from the pulmonary to systemic veins. Passive pulmonary artery hypertension was so marked in this patient that the pulmonary systolic pressure exceeded the systemic. Pulmonary venous obstruction probably occurs in all infradiaphragmatic cases and accounts for the distinctive clinical picture and course of these patients. Symptoms of venous obstruction may be accentuated during feeding.

The first successful repair of the infradiaphragmatic type was reported in discussion of a paper at the Academy of Pediatrics Meeting, October, 1961.

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Table 3.—Surgical Results in 68 Collected Reported Cases of Total Anomalous Pulmonary Venous Drainage *

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<tr>
<th>Type</th>
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* The cases reported by Nichols et al. 11 and one of the cases of Muller 13 are not included in this tabulation, since the exact site of drainage was not clear. Figures in parenthesis indicate survivors. 
† According to Darling’s anatomical classification. 

ings when the pulmonary venous trunk parallels the course of the esophagus.

Successful surgical treatment of total anomalous pulmonary venous drainage was first reported by Muller in 1951.10 The operation performed was anastomosis of the left auricular appendage to the left superior pulmonary vein. Anastomosis of the left auricu-
lar appendage to the common pulmonary venous trunk with ligation or constriction of the persistently left superior vena cava was reported by Mustard in 1957.20 The first successful total corrections of total anomalous pulmonary venous drainage were reported by Burroughs and Kirklin 4 and Lewis.47 A total of 70 cases of total anomalous pulmonary venous drainage have now been reported, and these results are tabulated in Table 3.3,4,6,8,10,12,13,16,22,24,27

Total correction has been achieved by (1) atrioseptoplasty or (2) transplantation of the pulmonary venous trunk. The most commonly performed and most successful procedure appears to be shifting of the atrial septum to repair completely the anomalous drainage in the right superior vena cava and right atrial forms of this anomaly. The same operation is successful in the coronary sinus type drainage except that the coronary flow is also diverted to the left atrium resulting in a persistent small right-to-left shunting of blood. In the persistent left superior vena cava type, anastomosis of the left atrium to the common pulmonary venous trunk: closure of the atrial septal defect, and interruption of the persistently left superior vena cava is necessary for total correction. It seems clear that the use of extracorporeal circulation has added to the precision and safety of these operations, and the trend towards its use is increasing. At present, it appears that patients requiring only atrioseptoplasty for correction (Types 1, B; 2, A; 2, B) can be treated surgically with low risk; however, correction of the left superior vena cava type (Type 1, A), which is the most common and which requires transplantation of the pulmonary venous trunk, still involves a high mortality rate. It seems likely that several factors are involved in surgical failures in these patients. The greatest difficulty is probably failure to achieve a technically adequate anastomosis between the common pulmonary trunk and left atrium. Cooley 8 has preferred an approach through the right atrium; however, we feel a more satisfactory anastomosis can be performed from an extracardiac posterior exposure accomplished

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by displacing the heart anteriorly and superiorly above the pericardium, a technique which will be illustrated in detail in a subsequent publication. A second factor in surgical failure in these patients is the size of the left atrium and ventricle. Inadequacy of the left cardiac chambers is difficult to assess in infants at autopsy, and it is possible that the repeated statements that the left side of the heart is hypoplastic in the presence of total anomalous pulmonary venous drainage may not be valid. In cases where the left atrium appears obviously small at surgery, transfer of the atrial septum anteriorly in the right atrium is advisable and technically simple. Inadequacy of the left ventricle may be present in a certain number of these infants, and this may represent a presently hopeless group in terms of total correction. Provided that the left atrium is of adequate size and the anastomosis between the left atrium and common pulmonary veins is sufficiently large, subsequent closure of the atrial septal defect and interruption of the persistent left superior vena cava will complete repair of this anomaly. We have preferred to perform final occlusion of the left superior vena cava only after:

1. The cardiac beat is regular and forceful.
2. The systemic arterial pressure is maintained at preoperative levels.
3. The measured right ventricular systolic pressure is below 30 mm. Hg.

If the right ventricular pressure is high, the pulmonary venous and left atrial pressures should then be measured. If the mean pulmonary venous pressure is greater than 20 mm. Hg and there is a 10 mm. Hg or greater pulmonary venous to left atrial gradient, it is likely that the anastomosis is too small and requires revision. If the pulmonary venous pressure is elevated but there is no pulmonary venous to left atrial gradient, the left atrium and/or the left ventricle are probably not adequate to handle the flow. The left atrium in such a case could then be further enlarged by atrial septoplasty. It may be necessary to leave the left superior vena cava partially patent in order to obtain a normal pulmonary venous pressure. Postoperative hemodynamic adjustments hopefully may allow complete occlusion several months later. In patients requiring atrial septoplasty it may be necessary to leave a small atrial septal opening in order to avoid significant left atrial and pulmonary venous hypertension in patients with small left-sided chambers.

Although promising, experience in complete correction of total anomalous pulmonary venous drainage in infants is still quite limited, and selection of patients for early operation should remain very critical in spite of the usual dismal medical prognosis. Of the patients, 10% to 20% will survive to the age of 2 to 5 years, and at present it appears advisable to defer surgery when the clinical condition and course of the patient indicate that survival through infancy is likely. Infants selected for early surgical correction are those who exhibited marked growth retardation, acute and chronic congestive heart failure, and marked or progressive cardiac enlargement. If surgery is deferred, these infants should have very careful medical supervision, since clinical deterioration and progressive cardiomegaly may occur rapidly (Fig. 1). Prior to operation patients should be maximally digitalized. Diuretics and low-salt formula are often necessary to control edema and congestive hepatomegaly. An accurate catheterization and angiographic appraisal of the anatomy and physiology is, of course, very helpful in planning surgical therapy. We believe that total correction using extracorporeal circulation is the operation of choice even for infants. Although it is generally considered that surgery using extracorporeal circulation is of increased risk in infants, in our experience in carefully selected cases the mortality has not been increased in patients 6 months to 3 years of age compared to older children. Except for Case 2 of this report, all our patients aged 6 months to 3 years in whom total correction of their defect has been achieved have survived surgery (total of 22 patients). Comparatively high perfusion rates have been used—100 ml. per kilogram per minute or greater. Such perfusion rates are possible in small patients by the use of maximal-sized arterial (Nos. 10 to 14) and venous (Nos. 22 to 28) cannulae. A pump or a low priming volume permits blood volume control during surgery. Accurate, rapid postoperative adjustments have been possible of changes of continuously rec arterial pressure.

Summary

Four patients with total anomalous pulmonary venous drainage were operated upon by surgical correction utilizing circulation was performed at 4 months and 11 in transplantation of the pulmonary veins into the pulmonary trunk in the other 2 (7 in months). The 2 younger patients survive—those who had not been selected and the 2 died from insufficiency due to extension of the condition. The 2 older patients survived and underwent successful corrections. Patent ductus arteriosus was present in 2 patients, but was ligated in the remaining 2. The surgical experience further confirmed that our patients were not unusual. The median survival time was 2 years. We report surgical experience during infancy using extracorporeal circulation is as yet small, the surgical risk of 40%-50% is the medical risk of 80%-90%.

Addendum

Since submission of this report, 1 infant and 1 small child with pulmonary venous drainage were censored. Although no April 11, 1962, a 4-month-old 5.4 kg. male with Type A with pulmonary veins entering the right atrium (April 11, 1962, a 36-month-old, 23 lb. male with pulmonary veins entering the right atrium); (2) July 6, 1962, 1962, A; (3) on July 6, 1962, 1962, A; (3) on July 6, 1962, a 36-month-old, 25 lb. male with pulmonary veins entering the right atrium).—Ryan et al.
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22 to 28) cannulae. A pump oxygenator with a low priming volume permits more precise blood volume control during the perfusion. Accurate, rapid postoperative blood volume adjustments have been possible on the basis of changes of continuously recorded systemic arterial pressure.

Summary

Four patients with total anomalous pulmonary venous drainage are presented. Total surgical correction utilizing extracorporeal circulation was performed at the ages of 4 months (10 lb. [about 4,536 gm.]), 7 months (13½ lb. [about 5,911 gm.]), 11 months (12 lb. [about 5,443 gm.]), and 24 months (24 lb. [about 10,886 gm.]). Correction was accomplished by atrioseptoplasty in 2 of the patients (4 months and 11 months) and by transplantation of the pulmonary venous trunk in the other 2 (7 months and 24 months). The 2 younger patients did not survive—the first had not been totally corrected and the second died from pulmonary insufficiency due to extensive pneumonitis. The 2 older patients survived and have had successful corrections. Patient selection for operation during infancy should remain quite critical; however, because of the high infant mortality associated with total anomalous pulmonary venous drainage, survival is often dependent on early surgery. Although the reported surgical experience for total correction during infancy using extracorporeal circulation is as yet small, the reported surgical risk of 40%-50% is lower than the medical risk of 80%-90%.

Addendum

Since submission of this paper, 2 further infants and 1 small child with total anomalous pulmonary venous drainage have been successfully operated on at our hospital: (1) On April 11, 1962, a 4-month-old, 12 lb. (about 5,443 gm.) male with Type 4 (mixed type—with pulmonary veins entering the coronary sinus and right atrium); (2) on April 20, 1962, a 36-month-old, 25 lb. male with Type 2, A; (3) on July 6, 1962, a 2-month-old, 7 lb. (about 3,175 gm.) female with Type 1, A. The 2 patients with pulmonary venous drainage into the coronary sinus who required atrial septral septoplasty both developed third-degree atroventricular (A.V.) block as the septum was sutured. Both patients survived ventricular rate above 30. The block persisted for 48 hours in one patient and for 3 weeks in the other. Thus, 3 of the total of 4 patients of our experience who underwent atrial septoplasty developed third-degree A.V. block. This complication is apparently related to unavoidable surgical manipulation and suturing in the region of the sinoatrial (S.A.) node. In our last 2 patients who developed block, the ventricular rate remained adequate, was not treated, reverted spontaneously, and did not appear to have a deleterious effect on the patients during the postoperative period.

References


On rare occasions the right artery arises from the ascending immediate branches. This comprises an account of 3 patients embryological error. Includ of previous similar reports, a the aberrant evolution of this artery, the resultant signs and the changes produced in the small lungs.

**Right Artery A Ascend**

**Literature Review**

We are aware of the apparent writing of 11 patients with ao-

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