Total Anomalous Pulmonary Venous Drainage

II. Spontaneous Functional Closure of Interatrial Communication after Surgical Correction in Infancy

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SUMMARY

The spontaneous functional closure of the interatrial communication in total anomalous pulmonary venous drainage (TAPVD) subsequent to surgical correction of the TAPVD in infants is reported. Three cases are documented with descriptions of clinical features, cardiac catheterization data, thermodilution curves, and angiography. This evidence suggests that it might be unnecessary in infants to close the interatrial communication at the time of correction of the TAPVD. It also suggests that balloon atrial septostomy may sometimes be unwise before the first-stage correction.

Additional Indexing Words:
Atrial septal defect, spontaneous closure of
Atrial septal defect, size in total anomalous pulmonary venous drainage
Thermodilution determination of shunts

In patients with total anomalous pulmonary venous drainage (TAPVD) the systemic cardiac output is limited to the volume which can pass through a defect in the interatrial septum. It has been our practice not to close this interatrial communication during the first-stage repair of supracardiac TAPVD in infants. We prefer a later closure as a second-stage operation. On reevaluating our earliest patients who are now approaching school age, we have found clinical evidence that four of these interatrial communications have closed spontaneously. We have reinvigotated three of these children by cardiac catheterization and angiography.

The spontaneous closure of ventricular septal defects and patent ductus arteriosus has been well documented. There have been few recorded cases of spontaneous closure of atrial septal defects. Mustard et al. mentioned one case of TAPVD which, after a first-stage corrective operation, was found subsequently to have no shunt at atrial level. No other data were provided. This report appears to be the first clear documentation of spontaneous functional closure of an interatrial communication following surgical correction of TAPVD.

Case Reports

Case 1

This girl was born of a normal pregnancy and delivery, weighing 3.12 kg. In the first few months of life she developed a dry cough, was difficult to feed, and failed to thrive. When seen at this hospital, at the age of 10 months, feeding produced dyspnea and cyanosis. The respiratory rate was about 70-80/min. The liver edge was palpable 4 cm below the costal margin. The cardiac impulse suggested right ventricular hypertrophy (RVH). A long systolic ejection murmur was heard at the second left intercostal space. The second sound was clearly split, and
Electrocardiograms of case 1 taken (top) prior to corrective surgery at 10 months of age, and (bottom) at 4½ years of age. (Standardization is 1.0 mv = 1.0-cm deflection.)
Case 2

This boy was seen at the age of 4 months with a history of cyanosis at birth and subsequent cyanotic attacks. His respiration had always been labored, particularly during feeding, but he had developed normally and weighed 5.22 kg when seen here. His respiratory rate was approximately 60/min, and the liver was palpable 2 cm below the costal margin. The cardiac impulse suggested RVH. There was a systolic ejection murmur at the second left intercostal space, and the second sound was clearly split. The split did vary with respiration. A diastolic murmur was heard along the left sternal edge. The electrocardiogram showed right-axis deviation and moderate RVH with an R wave of 13 mm in V1. The chest roentgenogram showed enlargement of the heart, increased pulmonary vascular markings, and pulmonary edema. Cardiac catheterization (table

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>PA pressures (mm Hg)</th>
<th>LV pressures (mm Hg)</th>
<th>Pw</th>
<th>Pj</th>
<th>Pp</th>
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<tr>
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<td>90/45</td>
<td>100/60</td>
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</table>

Abbreviations: PA = pulmonary artery; RV = right ventricle; LV = left ventricle; Pp = pulmonary artery wedge; RA = right atrium; RVH = right ventricular hypertrophy; Pw = pulmonary wedge pressure; Ps = systemic arterial pressure. *Samples were taken well distal to the junction of the superior vena cava.
Thermodilution curves obtained from thermistor in pulmonary artery following right ventricular injection of room-temperature isotonic solution. Curves A, B, and C were obtained from cases 3, 2, and 1, respectively. Note that the downslope of the curve returns to the baseline in the absence of a left-to-right shunt compared with the interruption of the downslope in curve D, obtained from a patient with an atrial septal defect.

Figure 2

1) showed the presence of TAPVD into the left innominate vein. Severe pulmonary hypertension was present. The right atrial pressure was 2 mm Hg higher than the left. Angiocardiography confirmed that the pulmonary venous trunk drained into the left innominate vein via a vertical vein, without any apparent obstruction to venous flow. A week later the anomalous pulmonary venous drainage was corrected, and the vertical vein was ligated. A routine tracheostomy was performed, and ventilation was maintained postoperatively. He made an uneventful recovery and, when seen 2 months later, was essentially normal. Two years postoperatively he was thought to have normal heart sounds.

At 6½ years of age he was reinvestigated. The cardiac impulse appeared to be normal, there were no murmurs, and there was normal splitting of the second sound. The electrocardiogram showed a RsR’ pattern in V1, and the initial frontal-plane loop was counterclockwise (fig. 4). These features had first been apparent at a follow-up examination when he was 1 year old. Chest roentgenogram was normal. At cardiac catheterization (table 1), performed from the right femoral vein, the left atrium was entered through what was presumed to be a patent foramen ovale. The left atrial pressure was 4 mm Hg higher than the right. A mean pressure gradient of 4 mm Hg was present across the anastomosis of the pulmonary venous trunk to the left atrium. The pulmonary arterial pressure was normal. No shunts were demonstrated by oximetry (table 1), thermodilution curves (fig. 2), or angiocardiography (fig. 5).

Case 3

This girl weighed 3.19 kg at birth, was difficult to feed, and thrived poorly. When seen at 10 months of age, she weighed 5.00 kg, had become progressively more dyspneic on feeding, and was occasionally cyanosed when crying or feeding. Her respiratory rate was 50–70/min. The liver was not clinically enlarged. The cardiac impulse suggested RVH. The second heart sound was accentuated in the left second intercostal space, but no murmurs were heard. The chest roentgenogram showed some cardiac enlargement with a “figure-of-eight” configuration, and the pulmonary vascular markings were increased (fig. 6). The electrocardiogram showed RVH with an R wave in V1 of 34 mm. The P waves were tall and peaked, suggesting right atrial enlargement. Cardiac catheterization (table 1) demonstrated the presence of total anomalous pulmonary venous drainage to the left innominate vein. A moderate degree of pulmonary hypertension was present. The mean right atrial pressure was 2 mm Hg higher than the left. Angiocardiography confirmed the presence of TAPVD with no stenosis of the large common trunk (fig. 7). Surgical anastomosis of this pulmonary venous trunk to the left atrium was performed under cardiopulmonary bypass, and the vertical vein was ligated. With routine postoperative care her recovery was uneventful, and she subsequently thrived normally.

She was reinvestigated at 5 years 3 months of age. The cardiac impulse was normal. A short systolic ejection murmur was present at the left second intercostal space, and the second sound

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Figure 3
Anteroposterior angiogram with diagrammatic representation of case 1, nearly 4 years after correction of TAPVD. Contrast was injected into the pulmonary artery. This late film shows right and left pulmonary veins (RPV's and LPV's) draining high into the left atrium and a sharply defined left atrial (LA) border with no visualization of right atrium on this or subsequent frames.

split normally. The electrocardiogram was normal. Chest roentgenogram showed a normal heart size and normal pulmonary vascular markings (fig. 6). Cardiac catheterization (table 1) showed the pulmonary artery pressure to be slightly elevated and the pulmonary arterial wedge pressure to be within normal limits. No shunt was demonstrated by oxygen saturations, thermodilution curves (fig. 2), or angiocardiography (fig. 8). The catheter, introduced from the right femoral vein, could not be passed through an interatrial communication. Although the pressures did not suggest the presence of significant obstruction to pulmonary venous flow, contrast material which was injected into the main pulmonary artery had not cleared from the pulmonary arteries after 6 sec (fig. 8).

Thermodilution Method
To perform thermodilution curves, a 6F Lehman catheter was placed in the pulmonary artery. A radiopaque thermistor catheter (Yellow Springs Instrument Co., no. 530) was passed through its lumen until its tip protruded. The Lehman catheter was then withdrawn into the right ventricle. An injection of 3-5 ml of 5% dextrose in water at room temperature was made into the right ventricle, and the thermodilution curves obtained from the pulmonary arterial thermistor are shown in figure 2. If cold indicator recirculates through the pulmonary circulation due to a left-to-right shunt, the downslope of the
thermodilution curve is interrupted in a similar fashion to the downslope of a conventional dye-dilution curve (fig. 2).

**Necropsy Studies**

In 10 postmortem formalin-fixed specimens of patients with TAPVD who died postoperatively, we used a graduated obturator to measure the diameters of the interatrial communication (ASD) and the aortic and pulmonic valve rings. In none was the ASD smaller than the aortic valve ring (table 2). It was in the secundum position, and in seven of the 10 specimens it had the appearance of a "stretched" foramen ovale. In three specimens there was an absence of tissue suggesting the presence of a true atrial septal defect. The average age at death of the patients studied was 3 months, and there was no relationship between the age and the size of the ASD.

**Discussion**

There have been few well-documented cases of spontaneous closure of an interatrial communication (ASD).\(^3\)\(^-\)\(^6\) None of these had associated TAPVD. In TAPVD the interatrial communication is, of necessity, large because the entire systemic cardiac output passes through it.

**Table 2**

<table>
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<tr>
<th>Specimen</th>
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Abbreviations: FO = the presence of a flap which suggests that the interatrial communication is a "stretched" foramen ovale; ASD = absence of tissue suggesting a true atrial septal defect.

Figure 4

Electrocardiogram of case 2, 6 years after correction of TAPVD showing RsR' pattern in V1, despite spontaneous closure of interatrial communication. (Standardization is 1.0 mv = 1.0-cm deflection.)
Figure 5

Left heart phase of anteroposterior pulmonary arterial angiogram with diagrammatic representation in case 2. Pulmonary venous drainage (RPV's and LPV's) into upper portion of left atrium is demonstrated. The left atrial (LA) border is sharply defined with no evidence of a left-to-right shunt on this or subsequent frames. Ao represents aorta.

through it. Our necropsy studies have demonstrated that no ASD was smaller than the aortic valve ring. Our necropsy specimens also suggested that in most cases the ASD was a “stretched” foramen ovale but that a true deficiency of tissue was present in some. Indeed, in one surviving patient a residual ASD has been closed surgically 4 years after the first-stage correction. This ASD was large, requiring a patch to close it. At the time of the initial investigation of this patient, the right atrial pressure was 9 mm Hg and the left atrial pressure 2 mm Hg. This was an even greater pressure differential than that found at the initial investigations of our three patients in whom the ASD subsequently closed spontaneously. Thus, the atrial pressure differential may not be reliable in predicting either the size of the ASD or whether it will later close spontaneously.

Of the three patients described in this report, one had the auscultatory signs of an atrial septal defect (case 1), and another had an electrocardiogram compatible with an
Figure 7
Anteroposterior angiogram with diagrammatic representation obtained in case 3. Initial investigation. Contrast material, injected into the pulmonary venous trunk (vertical vein) fills pulmonary veins (PV's) retrograde, and drains via the left innominate vein (L Innom. V) and superior vena cava into the right atrium. There is no evidence of stenosis of this trunk.

atrial septal defect (case 2, fig. 4). We felt that it was important to determine whether the ASD had closed in all our patients. Our data suggest that a minor degree of stenosis of the anastomosis might have occurred in cases 2 and 3; cardiac catheterization in early adolescence should determine whether this is important.

Balloon atrial septostomy has been advocated prior to corrective surgery of the total anomalous pulmonary venous drainage.8,9

Figure 6
Chest roentgenogram of case 3. (Top) At 10 months of age the heart has a "figure-of-eight" configuration. (Bottom) At 5 years of age the heart has a normal size and configuration.
Anteroposterior angiogram with diagrammatic representation obtained in case 3. Investigation 4½ years after correction of TAPVD. Pulmonary artery injection of contrast medium shows left atrium filling from pulmonary veins (PV's) with sharply defined left atrial (LA) border and no right atrial visualization on this or subsequent frames. Contrast is still present in the pulmonary artery (PA) 6 sec after injection (see text).

Gathman and Nadas\(^\text{10}\) observed that in 40 of 43 cases of pulmonary venous obstruction, a narrow site on the pulmonary venous trunk was demonstrable. The hemodynamic evidence which they presented showed that there was no obstruction at interatrial level in their series. They concluded that atrial septostomy would be of little value in the presence of pulmonary venous obstruction. Our data support this view and suggest that, in many cases of TAPVD, septostomy may be better avoided. As we have already discussed, all of our patients who have had autopsies have had a large atrial septal defect, even those who died at a young age. Also, since functional closure of the interatrial communication may occur following correction of the TAPVD, an atrial septal defect induced by trauma might later make an otherwise unnecessary operation become essential. However, we recognize that if an infant with TAPVD has a small interatrial communication, and as a consequence is desperately ill, balloon atrial septostomy could be lifesaving.

Acknowledgments

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