Left Heart Volume Characteristics with a Right Ventricular Volume Overload

Total Anomalous Pulmonary Venous Connection and Large Atrial Septal Defect

By Thomas P. Graham, Jr., M.D., Jay M. Jarmakani, M.D., and Ramon V. Canent, Jr., M.D.

SUMMARY

Left heart volume characteristics were evaluated by using biplane cineangiocardiology in 18 studies in 15 patients with total anomalous pulmonary venous connection (TAPVC) and in 37 studies on 35 patients with large secundum or sinus venosus atrial septal defects (ASD). Left ventricular end-diastolic volume (LVEDV) was decreased to less than 67% of normal in five of 15 preoperative TAPVC patients, but the average value for the entire group was not significantly different from normal. In ASD patients, the average LVEDV was 87% of normal which was significantly decreased ($P < 0.001$). The ejection fraction was decreased from normal in patients with TAPVC and ASD who were less than 2 years of age (0.62 vs. 0.68, $P < 0.01$), but was normal in older patients. Left ventricular systolic output was significantly decreased from normal in both TAPVC (75% of normal, $P < 0.001$) and ASD patients (81% of normal, $P < 0.001$). Left atrial maximal volume was significantly decreased from normal in TAPVC patients averaging only 55% of normal ($P < 0.001$), but was normal in eight ASD patients. All volume variables increased following corrective surgery in two TAPVC patients and two ASD patients. These studies document that left heart volumes and outputs in infants and children with TAPVC and large isolated atrial defects can be diminished preoperatively.

Additional Indexing Words:

- Left ventricular end-diastolic volume
- Left atrial maximal volume
- Ejection fraction

TOTAL anomalous pulmonary venous connection and large atrial septal defects share the physiologic abnormality of a large left-to-right shunt which bypasses the left ventricle and produces right ventricular volume overload. With these anatomic lesions there is a potential for underloading the left ventricle with a resultant decrease in left ventricular size and systemic flow. The extent to which left heart size and output may be altered in these conditions in infancy and childhood is not known.

The purpose of this investigation, therefore, was to quantify with biplane cineangiocardiology left ventricular and left atrial volume characteristics in infants and children with total anomalous pulmonary venous connection
Table 1

Vital Statistics, Hemodynamic Data, and Site of Anomalous Connection in Patients with Total Anomalous Pulmonary Venous Connection

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>BSA (m²)</th>
<th>Hgb (g/100 ml)</th>
<th>HR</th>
<th>RVP (mm Hg)</th>
<th>LVP (mm Hg)</th>
<th>RVP/LVP</th>
<th>O₂ sat (%)</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>C.S.</td>
<td>1 day</td>
<td>0.19</td>
<td>19.3</td>
<td>143</td>
<td>61/5</td>
<td>61/6</td>
<td>1.00</td>
<td>72</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>S.S.</td>
<td>7 days</td>
<td>0.22</td>
<td>16.0</td>
<td>167</td>
<td>130/10</td>
<td>50/2</td>
<td>2.60</td>
<td>81</td>
<td>Portal vein</td>
</tr>
<tr>
<td>J.R.</td>
<td>47 days</td>
<td>0.24</td>
<td>11.9</td>
<td>150</td>
<td>50/2</td>
<td>98/5</td>
<td>0.51</td>
<td>93</td>
<td>Right atrium</td>
</tr>
<tr>
<td>T.T.</td>
<td>62 days</td>
<td>0.25</td>
<td>14.7</td>
<td>143</td>
<td>86/3</td>
<td>74/8</td>
<td>1.16</td>
<td>89</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>E.L.</td>
<td>66 days</td>
<td>0.25</td>
<td>18.0</td>
<td>158</td>
<td>120/18</td>
<td>80/10</td>
<td>1.50</td>
<td>66</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>E.W.</td>
<td>61 days</td>
<td>0.21</td>
<td>14.0</td>
<td>136</td>
<td>80/10</td>
<td>70/5</td>
<td>1.14</td>
<td>82</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>K.B.*</td>
<td>3 mo</td>
<td>0.21</td>
<td>17.6</td>
<td>136</td>
<td>47/8</td>
<td>98/11</td>
<td>0.48</td>
<td>92</td>
<td>Coronary sinus</td>
</tr>
<tr>
<td>P.S.</td>
<td>3 mo</td>
<td>0.22</td>
<td>15.6</td>
<td>143</td>
<td>60/10</td>
<td>80/9</td>
<td>0.75</td>
<td>86</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>D.J.</td>
<td>3 mo</td>
<td>0.24</td>
<td>12.8</td>
<td>125</td>
<td>45/10</td>
<td>78/7</td>
<td>0.58</td>
<td>84</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>D.T.</td>
<td>3 mo</td>
<td>0.26</td>
<td>13.6</td>
<td>125</td>
<td>65/5</td>
<td>57/4</td>
<td>1.14</td>
<td>94</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>M.A.</td>
<td>5 mo</td>
<td>0.25</td>
<td>15.6</td>
<td>152</td>
<td>57/2</td>
<td>64/3</td>
<td>0.89</td>
<td>80</td>
<td>Right atrium</td>
</tr>
<tr>
<td>S.T.†</td>
<td>7 mo</td>
<td>0.35</td>
<td>13.5</td>
<td>107</td>
<td>40/2</td>
<td>98/11</td>
<td>0.41</td>
<td>87</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>K.K.</td>
<td>22 mo</td>
<td>0.40</td>
<td>10.5</td>
<td>115</td>
<td>65/9</td>
<td>83/10</td>
<td>0.77</td>
<td>85</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>J.H.</td>
<td>2 yr</td>
<td>0.47</td>
<td>11.0</td>
<td>136</td>
<td>100/7</td>
<td>105/11</td>
<td>0.95</td>
<td>96</td>
<td>Right atrium</td>
</tr>
<tr>
<td>S.T.†</td>
<td>3 yr</td>
<td>0.54</td>
<td>14.3</td>
<td>120</td>
<td>50/5</td>
<td>82/7</td>
<td>0.61</td>
<td>87</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>C.J.</td>
<td>11 yr</td>
<td>1.04</td>
<td>13.6</td>
<td>83</td>
<td>49/2</td>
<td>104/5</td>
<td>0.47</td>
<td>92</td>
<td>Vertical vein</td>
</tr>
<tr>
<td>Mean ‡</td>
<td>1.4 yr</td>
<td>0.34</td>
<td>14.3</td>
<td>135</td>
<td>69/7.4</td>
<td>84/7.1</td>
<td>0.93</td>
<td>85</td>
<td></td>
</tr>
<tr>
<td>SD</td>
<td>2.8 yr</td>
<td>0.22</td>
<td>2.2</td>
<td>21</td>
<td>29/4.5</td>
<td>16/3.4</td>
<td>0.29</td>
<td>8</td>
<td></td>
</tr>
</tbody>
</table>

Postoperative

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>BSA (m²)</th>
<th>Hgb (g/100 ml)</th>
<th>HR</th>
<th>RVP (mm Hg)</th>
<th>LVP (mm Hg)</th>
<th>RVP/LVP</th>
<th>O₂ sat (%)</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>K.B.</td>
<td>2 yr</td>
<td>0.50</td>
<td>12.5</td>
<td>125</td>
<td>40/10</td>
<td>79/9</td>
<td>0.51</td>
<td>92</td>
<td></td>
</tr>
<tr>
<td>C.J.</td>
<td>14 yr</td>
<td>1.40</td>
<td>13.5</td>
<td>88</td>
<td>30/8</td>
<td>122/15</td>
<td>0.25</td>
<td>96</td>
<td></td>
</tr>
</tbody>
</table>

*Patient also studied after operation.
†Patient S.T. had two studies at 7 mo and 3 yr of age; did not undergo operation.
‡Mean of 16 studies.

Abbreviations: HR = heart rate; RVP = right ventricular pressure; LVP = left ventricular pressure; Systemic O₂ sat = systemic O₂ saturation; site = site of anomalous venous connection.

(TAPVC) and large secundum or sinus venous atrial septal defects (ASD) and to attempt to correlate left heart volume with other hemodynamic data obtained at cardiac catheterization.

Methods

Patient Population

Group I

Sixteen preoperative and two postoperative studies were performed on 15 patients with TAPVC (table 1). Ages ranged from 1 day to 14 years. In five of the 16 preoperative studies, peak right ventricular pressure (RVP) exceeded left ventricular pressure (LVP), and in six studies the RVP/LVP ratio was 0.61 or less (table 1). The average RVP/LVP ratio was 0.93 ± 0.29, and the average peak RVP was 69 ± 29 mm Hg. The RV end-diastolic pressure averaged 7.4 ± 4.5 mm Hg and was not different from normal (P > 0.2). Systemic oxygen saturation was more than 80% in all but two patients and averaged 85%. Hemoglobin concentration averaged 14.3 ± 2.2 g/100 ml.

In 10 of the 15 patients the anomalous connection was to a vertical vein, in three to the right atrium, in one to the coronary sinus, and in one to the portal vein. Three of the 10 patients with vertical vein (VV) connections had catheterization of this structure into the common pulmonary vein (CPV), and one (T.T., table 1) had a measurable pressure difference from CPV to VV. The only associated cardiovascular lesions were atrial defects which were present in all but one patient and a ventricular defect present in one patient.

Group II

Thirty-seven studies were performed in 35 patients with atrial septal defects including two patients studied postoperatively. Ages ranged from 83 days to 11 years. Thirty-one patients had secundum defects, and four patients had sinus venous defects with partial anomalous pulmonary venous connection. Patients with mitral insufficiency on left ventricular cineangiocardiography were excluded from the study.
patients had left-to-right shunts $\approx 42\%$ of pulmonary blood flow (table 2). Peak RVP averaged 36 $\pm$ 14 mm Hg in these patients and was not significantly increased ($P > 0.1$). The ratio, RVP/LVP, averaged 0.37 $\pm$ 0.12. Right ventricular end-diastolic pressure averaged 7.2 $\pm$ 2.9 mm Hg and was not different from normal ($P > 0.1$). None of the patients in this group had a hemoglobin level above 15.3 g/100 ml except one infant with a value of 16.3 g/100 ml at age 4 months. All patients were clinically acyanotic with systemic oxygen saturations $\approx 92\%$.

**Data Collection**

All patients were studied during routine diagnostic cardiac catheterization. Patients less than 1 year of age were studied during local anesthesia alone or local anesthesia plus sedation with intramuscularly administered meperidine (1 to 1.5 mg/kg) or promethazine (0.5 to 1.0 mg/kg).
mg/kg) or both. For older patients light general anesthesia with nitrous oxide and small concentrations of halothane (≤0.5%) were used. Left and right heart pressures were recorded before the first cine with NIH catheters (no. 5, 6, or 7) attached to Statham P 23 Gb transducers. Zero pressure was referenced to midchest.

Left heart volume variables were calculated from biplane cineangiograms (AP and lateral, 60 frames/sec) which were filmed after injection of sodium and meglumine diatrizoates (1.0 ml/kg of 75% Hypaque-M) into the left atrium or left ventricle. The electrocardiogram and cine exposure were recorded on photographic paper during the cineangiography.

Correction for X-ray magnification was performed by using a calibrated grid filmed at the end of each study at the midchest position. The details of this system have been reported previously.1,2 Left ventricular end-diastolic volume (LVEDV), end-systolic volume (LVESV), and left atrial maximal volume (LAmax) were calculated in the absence of ectopic beats by the area-length method.3,4 Left ventricular systolic output (LVSO) was derived as LV stroke volume (LVEDV–LVESV) multiplied by heart rate. Left ventricular volumes were corrected by using previously derived regression equations.2 For calculated volumes ≤15 cm³, \( V' = 0.733 \times V \) (where \( V' \) = regressed volume and \( V \) = calculated volume). For calculated volumes >15 cm³, \( V' = 0.974 \times V - 3.1 \).

All volume variables were compared with previously derived normal standards from 56 patients with normal left hearts.2 Predicted normal values were calculated from regression equations relating volume variables to weight, height, and age.2

Because the normal left ventricular volume standards for end-diastolic volume (LVEDV), ejection fraction (LVEF), and systolic output (LVSO) were derived following pulmonary artery (PA) cineangiography (cine) and the data in the present study were derived from left ventricular or left atrial cineangiography, an initial investigation was undertaken to ascertain if these two different modes of data collection produced significantly different volume determinations.

Therefore, comparisons were made of LVEDV, LVEF, and LVSO calculated from 71 patients who had both PA and LV cines at the same catheterization. The time elapsed between the two cines was not less than 15 min in all patients, and the PA cine preceded the LV cine in all but three patients. Patients whose heart rates during the two cines differed by 10 beats/min or more were excluded from this analysis. Ages ranged from 1 week to 17 years, and diagnoses included aortic stenosis, coarctation, ventricular septal defect, pulmonary stenosis, tetralogy of Fallot, transposition of the great arteries, myocardial disease, and normal left heart.

The comparison of LVEDV for the two cines is demonstrated in figure 1. There was no significant difference in the mean LVEDV for one cine versus the other, and the correlation between the two values was excellent (\( P < 0.001, r = 0.986 \)). The majority of patients (72%) had slightly larger LVEDV calculated from the LV cine. It averaged only 5.8% higher than the value found for the PA cine.

Similar comparisons were made of the LVEF and the LVSO calculated from the two cines, and no significant differences (\( P > 0.1 \)) were found in these two variables for the two cines.

**Results**

**Left Ventricular End-Diastolic Volume**

Five of 15 preoperative patients (33%) with TAPVC had LVEDV values which were less than the average normal value minus 2 sd (fig. 2). One patient had two preoperative studies separated by 28 months. During this time the LVEDV increased in proportion to her overall growth with the resultant lack of any change in LVEDV expressed as a percentage of predicted normal values. Two patients had LVEDV values which were greater than normal at ages 42 and 27 days, and the mean value for the entire group was not significantly different from normal

![Figure 1](http://circ.ahajournals.org/)

**Figure 1**

Comparison of left ventricular end-diastolic volume calculated from left ventricular and pulmonary artery cineangiography in the same patient. see = standard error of estimate.
Left ventricular end-diastolic volume as a percentage of predicted normal for patients with total anomalous pulmonary venous connection. In this and figures 3 to 6 horizontal broken lines indicate ± 2 SD of normal, and age scale is interrupted.

\( P > 0.4 \). There was no significant correlation of LVEDV expressed as ml/m² with either peak RVP \( P > 0.2 \) or RVEDP \( P > 0.05 \), or with Hgb \( P > 0.5 \).

Two patients had both preoperative and postoperative studies, and both showed an increase in LVEDV following surgery (fig. 2).

Eleven of 35 preoperative ASD patients (31%) had LVEDV values which were less than the average normal value minus 2 SD (fig. 3). The mean value for the entire group was significantly different from normal, averaging 87 ± 21% \( P < 0.001 \). There was no significant correlation of LVEDV expressed as ml/m² with left-to-right shunt \( P > 0.5 \).

Two patients with both preoperative and postoperative studies showed increases in LVEDV following surgery (fig. 3).

**Left Ventricular Ejection Fraction**

The ejection fraction was significantly decreased from normal for TAPVC and ASD patients less than 2 years of age averaging 0.62 ± 0.06 versus a normal value of 0.68 ± 0.06 \( P < 0.01 \). Heart rates were not significantly different \( P > 0.2 \) for this patient group versus the normal group. The patient with the smallest value for LVEDV of the entire group (51% of normal) had an ejection fraction of 43%.

For all patients above 2 years of age, the average ejection fraction was normal. Three older patients in the ASD group had ejection fractions of less than 50%. These values were 0.49, 0.46, and 0.44. None of these three patients had low end-diastolic volumes.

**Left Ventricular Systolic Output**

The LVSO was less than the normal value minus 2 SD, in six of the 15 TAPVC patients before operation (40%) (fig. 4). The average value was significantly different from normal (75 ± 19%, \( P < 0.001 \)). This variable averaged 2.89 ± 0.89 liters/min versus a normal value\(^2\) of 4.42 ± 0.95 \( P < 0.001 \). Again there was no significant correlation of LVSO/BSA with peak RVP \( P > 0.5 \) or Hgb \( P > 0.5 \). The two patients with both preoperative and postoperative LVSO determinations showed increases in this variable.

Eight of 35 patients with atrial defects (23%) had systolic outputs which were less than the average normal value minus 2 SD (fig. 5). The mean value for this group was significantly different from normal, 81 ± 20%, \( P < 0.001 \) or 3.65 ± 1.01 liters/min/m², \( P < 0.01 \). There was no significant correlation of LVSO/BSA with age, but a weak correlation was found with degree of left-to-right shunt: LVSO/BSA = −0.037 (shunt, %) + 5.77, \( P < 0.05 \), \( r = 0.341 \).
difficulties defining the atrial septum in the presence of a large left-to-right shunt. This variable was within the normal range (mean ± 2 SD of predicted volume) in all patients.

**Left Ventricular Pressure**

Both left ventricular peak pressure (LVP) and end-diastolic pressure (LVEDP) were significantly decreased from normal² in TAPVC patients: LVP was 84 ± 16 mm Hg versus a normal value of 98 ± 12 mm Hg (P < 0.01), and LVEDP was 7.1 ± 3.4 mm Hg versus a normal value of 11.4 ± 4.2 mm Hg (P < 0.001).

Both LVP and LVEDP were not different from normal in ASD patients (P > 0.1).

**Discussion**

This study documents the low left atrial volumes in all patients studied with TAPVC and the low LV end-diastolic volumes in 33% of these patients. In addition, the LV ejection fractions and LV systolic outputs were decreased in this group of infants with total anomalous pulmonary venous connection.

Multiple hemodynamic factors may contribute to these findings. The size of the atrial defect may be one determinant of left heart size with small communications associated with small volumes. In this regard, there was no significant inverse correlation (P > 0.3) of LAmax and LV end-diastolic volume or LV

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**Figure 4**

Left ventricular systolic output as a percentage of predicted normal for patients with total anomalous pulmonary venous connection.

**Figure 5**

Left ventricular systolic output as a percentage of predicted normal for patients with large atrial septal defects: L-R shunt ≥ 50%.

**Figure 6**

Left atrial maximal volume as a percentage of predicted normal for patients with total anomalous pulmonary venous connection.
systolic output with either mean atrial pressure difference across the ASD or with the height of the right atrial a wave.

Another factor that may contribute to small LV size in patients with TAPVC could be increased RV diastolic pressure which may alter the LV diastolic pressure-volume relationship resulting in a less distensible LV. This phenomenon has been demonstrated in open-chest dogs by Taylor and co-workers. In the small group of TAPVC patients studied, however, there was no statistically significant relationship between LVEDV/BSA and RVEDP ($P > 0.2$).

The decrease in ejection fraction in infants with TAPVC was modest, may well be related to a decrease in LVEDV, and may not reflect any basic alteration in myocardial contractile state.

Of the 15 patients with TAPVC who had preoperative studies, seven underwent attempted corrective surgery, and two have survived with good results 3 and 4 years following surgery. These two patients were 1 year and 12 years old at operation. Both had peak RV pressures of 51% or less of peak LV pressure preoperatively and postoperatively and both showed increases in normalized LA and LV volumes when studied 1 and 2 years following operation. One of these patients (K.B.) had a small LV volume preoperatively (57% of normal) at a time when there was a 2-mm mean difference in pressure across the atrial septum prior to balloon atrial septostomy. He demonstrated remarkable clinical improvement following septostomy and corrective surgery was delayed for 8 months after this procedure. Unfortunately, no data were obtained immediately preoperatively on this patient to assess the effect of the septostomy on left heart size.

Of the five patients who died after attempted corrective surgery, only one had a decreased LVEDV to below the lower limit of normal (74% of predicted). The one patient (J.H.) with an abnormally low LV volume (51% of normal) had the unique situation of TAPVC to the right atrium with an intact atrial septum, normal mitral valve, and a ventricular septal defect. It was felt that this patient's left ventricle failed acutely in the operating room following attempted correction at age 2 years. This severe degree of LV hypoplasia may provide a significant obstacle to successful correction as a one-stage procedure.

Three of the remaining four patients who died following surgery had peak RVP $\geq$ LVP indicating that pulmonary hypertension may play a large role in surgical mortality, as has been suggested by other investigators. Gersony and associates recently have reported successful corrective surgery in seven of 10 infants with TAPVC whose ratio of RVP/LVP averaged 0.66. One patient in their series operated on successfully had an RVP/LVP ratio of 1.20 which indicates that severe pulmonary artery hypertension in infancy does not preclude successful surgery.

Patients with isolated atrial septal defects showed small decreases from normal in LVEDV and LV systolic output. These findings could not be attributed to an effect of anesthesia since the normal values were obtained under identical conditions. The two ASD patients with preoperative and postoperative studies showed increases in the variables following corrective surgery.

Only one of the ASD patients studied had clinical signs of heart failure or required digitalis therapy. This infant, C.R., age 21 months, had a decreased LVEDV to only 64% of normal with the LV systolic output only 66% of normal. She has not undergone surgery.

Twenty-eight of the 35 ASD patients have undergone corrective surgery to date with no deaths. None of these patients has shown signs of left heart failure following surgery. Thus, left heart dysfunction which has been reported in adults with large atrial defects does not appear to be a factor in children with this lesion.

In summary, these findings indicate that left heart size and output can be decreased to abnormally low values in infants and children with TAPVC and isolated atrial septal defects with large shunts. The contribution of small left heart size to surgical morbidity and
mortality in infants with TAPVC is unclear. Future studies correlating preoperative left atrial and left ventricular size with postoperative monitoring of left atrial pressure and systemic output should help to resolve this important question.

Acknowledgment

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References


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