Characteristics of Ventricular Function in Single Ventricle

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SUMMARY Functional characteristics of the single ventricle were studied by means of biplane angiography in 34 patients. Group 1 consisted of 14 patients with normal or increased pulmonary vascular marking on chest film and no pulmonary stenosis. Group 2 included 20 patients with severe angiographic pulmonary stenosis and decreased pulmonary vascular marking. Ventricular volume parameters were calculated according to Simpson’s rule and were compared with normal values. The sum of the normal left and right ventricular volumes was assumed to be 100%.

In group 1, ventricular end-diastolic volumes averaged 143 ± 11% and were significantly (p < 0.001) larger than end-diastolic volumes in group 2 (81 ± 4%). The presence or absence of severe pulmonary stenosis affecting pulmonary blood flow was a main factor regulating the ventricular chamber size in single ventricle. In both groups, the ejection fraction of a single ventricle was significantly lower than that of a normal left or right ventricle. Ventricular size and function in patients with a single ventricle should be carefully assessed before ventricular septation surgery.

THE ANATOMY OF SINGLE VENTRICLE has been well described, but its functional characteristics have not been reported. Correction of this complicated cardiac anomaly by septation surgery, in which the single ventricle is divided into two chambers, has become a challenge for cardiac surgeons. Therefore, it is important to know the volume and function of the single ventricle before surgical intervention.

We studied the functional characteristics of the single ventricle by analyzing biplane ventricular angiograms of good quality in 34 patients with a diagnosis of single ventricle confirmed angiographically or at autopsy.

Methods

Patient Population

Biplane, large-film angiograms (6 frames/sec) of 34 patients, 20 males and 14 females, ranging in age from 4 months to 12 years, were analyzed. The availability of angiograms of good quality was the sole criterion for patient selection. All patients were diagnosed as having a single ventricle by the definition reported previously. A single ventricle is one ventricle that receives both the tricuspid and mitral valves, or a common atrioventricular (AV) valve. This definition excludes tricuspid and mitral atresia. The angiographic morphology of the single ventricle was classified into three types (I, II and III). Details of this angiographic classification were also previously reported. In brief, type I is identical to the anatomic type A of Van Praagh et al., type II is comparable to type C, and type III is equivalent to Van Praagh’s types B and D.

There were seven, 13 and 14 patients with types I, II and III single ventricle, respectively. Three patients with type I, eight with type II and nine with type III had severe valvular and subvalvular pulmonary stenosis (PS) or pulmonary atresia. In one patient with type I single ventricle, the ventriculoatrial relationship was normal; four other type I patients had a transposition of the great arteries (TGA), and two had a double outlet right ventricle (DORV). Three patients with a type I single ventricle had two AV valves and four patients had a common AV valve. Of
13 patients with a type II single ventricle, six had a normal ventriculotrunical relationship, four had a TGA, and three had a DORV. Ten type II patients had two AV valves and the remaining three had a common AV valve. All 14 type III patients had a DORV and a common AV valve.

Other commonly associated anomalies included absent inferior vena cava with azygos or hemiazysos continuation in four patients, bilateral superior vena cava in six, anomalous pulmonary venous return in three, single atrium or large atrial septal defects in 20, and a right aortic arch in six. Dextrocardia was present in six patients.

Except for the presence of PS, we did not classify the concomitant anomalies present in these patients with single ventricle because, functionally, both systemic and pulmonic venous returns flow into a single ventricle and are ejected into the great arteries, depending on the relative impedance to outflow from the single ventricle into the pulmonary and systemic circulations. In this series, there were no patients with significant AV valve regurgitation or subaortic or aortic stenosis. Thus, the 34 patients were divided into two groups according to the presence or absence of severe PS demonstrated by angiography. Group 1 included 14 patients with no angiographic PS and a normal or increased pulmonary vasculature on plain chest x-ray films. Group 2 included 20 patients who had significant angiographic PS or pulmonary atresia without patent ductus arteriosus, as well as a decreased pulmonary vascular shadow on chest x-ray films. None of the patients had a shunt or corrective surgery. In group 1, there were four, five and five patients with types I, II and III single ventricle, and in group 2, there were three, eight and nine patients of each type, respectively.

Patients in group 1 were 4 months to 12 years (average 4.1 years). In group 2, 16 patients with PS were 5 months to 10 years old (mean 2.5 years); four patients with pulmonary atresia were 4 months to 7 years old (average 2.3 years). The age difference between groups was not statistically significant (p > 0.05).

**Data Acquisition**

Biplane, large-film angiography was performed at 6 frames/sec in all patients, who were sedated with intramuscular injections of meperidine or ketamine; lidocaine was used for local anesthesia. Frontal and lateral films were obtained. In the majority of patients, two catheters were inserted through the femoral vein and artery, and ventriculography was performed twice by the injection of contrast material through a retrograde transaortic valve catheter and an

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**Figure 1.** Simultaneous recording of filming time and ECG. The film taken during the PR interval was considered to be an end-diastolic view (EDV) and the one taken at the end of the T wave an end-systolic view (ESV). With large-film angiography at 6 frames/sec, analysis of the EDVs and ESVs from a single beat was unfeasible; therefore, a composite analysis from several beats was made.
antegrade trans-right atrial catheter to confirm the diagnosis.

The best angiograms were selected for ventriculographic analysis, regardless of the site of injection of contrast material. The intraventricular pressure was measured using a Statham P23Db transducer connected to a cardiac catheter by means of a short, debubbled, water-filled teflon tube. The end-diastolic pressure was measured before the angiographic studies.

ECGs were recorded during filming and were used to select end-diastolic and end-systolic films (fig. 1). The end-diastolic film selected was obtained during the PR interval and the end-systolic film was obtained at the end of the T wave. Using this method, we could not obtain end-diastolic and end-systolic views in a single beat; therefore, the views were composed from several beats. Only the films taken during normal sinus rhythm were analyzed. It was harder to obtain a good quality end-systolic image than to obtain an end-diastolic image. The end-systolic image was suitable for evaluation in only 26 of 34 patients (77%), but the end-diastolic views were of good quality in all patients. The representative biplane angiocardiograms in one 2-year-old patient from each group are shown in figure 2.

Ventricular volumes were computed according to Simpson’s rule using an ultrasound graf-pen connected to a Hitac 10 computer system. The magnification of the film was corrected in each patient. End-diastolic volume (EDV), end-systolic volume (ESV) and ejection fraction (EF) were calculated and compared with the normal values reported by Graham, Nakazawa and their co-workers. Because most single ventricles could not be angiographically separated into the right and left ventricular compartments, the ventricular volumes were calculated as a “single ventricle.” Therefore, the sum of the normal right and left ventricular volumes was assumed to be 100%; this can be expressed as 147.6 × BSA where BSA is body surface area (m²). The calculated volume was corrected by a regression equation. In types II and III single ventricle, the calculated volumes were corrected by the equation Va = 0.856Vc + 2.5 (r = 0.995; p < 0.001), where Va is the actual volume and Vc the calculated volume. This equation was made primarily for the biplane analysis of the ventricle with right ventricular morphology. For type I single ventricle with left ventricular morphology, we used the regression equation Va = 0.928Vc − 3.8. Although there was an associated small outflow tract chamber in type I single ventricle, its volume was small and therefore excluded from the total volume. Ventricular stroke volumes were calculated from the difference between EDV and ESV. The EF was calculated as the ventricular stroke volume divided by EDV.

Figure 2. Representative biplane angiocardiograms from one patient with single ventricle from each group. Both patients were 2 years old and had a type III single ventricle with (group 2) or without (group 1) pulmonary stenosis (PS). Top) Group 1 (Gr-1): no PS; dilatation of the ventricular chamber was obvious. Bottom) Group 2 (Gr-2): pulmonary atresia; the ventricular chamber was small. A-P = anteroposterior view; Lat = lateral view.
Table 1. Ventricular Function in Single Ventricle

<table>
<thead>
<tr>
<th>Group</th>
<th>n</th>
<th>Age (years)</th>
<th>HR (beats/min)</th>
<th>SaO₂ (%)</th>
<th>Hgb (g%)</th>
<th>EDP (mm Hg)</th>
<th>EDV (%)</th>
<th>SVI (ml/m²)</th>
<th>Ventricular output (l/min/m²)</th>
<th>EF</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1</td>
<td>14</td>
<td>4.1</td>
<td>118 ± 7</td>
<td>76.7 ± 3.0</td>
<td>15.8 ± 0.4</td>
<td>8.4 ± 0.8</td>
<td>143 ± 11</td>
<td>80 ± 9</td>
<td>9.44 ± 0.91</td>
<td>0.49 ± 0.02</td>
</tr>
<tr>
<td>Group 2</td>
<td>20</td>
<td>2.5</td>
<td>127 ± 6</td>
<td>71.4 ± 3.7</td>
<td>17.6 ± 0.6</td>
<td>6.8 ± 0.6</td>
<td>81 ± 4</td>
<td>47 ± 3</td>
<td>5.97 ± 0.48</td>
<td>0.54 ± 0.02</td>
</tr>
</tbody>
</table>

Significance: NS p <0.05 p <0.001 p <0.005 p <0.005 NS

Values are mean ± SEM.

Group 1 consisted of patients without pulmonary stenosis and with a normal or increased pulmonary vascular shadow. Group 2 consisted of patients with pulmonary stenosis or pulmonary atresia and with a decreased pulmonary vascular shadow.

Abbreviations: HR = heart rate; SaO₂ = arterial oxygen saturation; Hgb = blood hemoglobin; EDP = end-diastolic pressure; EDV = end-diastolic volume expressed as a percentage of the sum of the normal right and left ventricular volumes (147.6(BSA)¹⁴³); SVI = ventricular stroke volume index of single ventricle; EF = ejection fraction of single ventricle.

Results

The mean values of hemodynamic variables in single ventricle are presented in table 1. The mean heart rates for groups 1 and 2 at the time of the study were 118 ± 7 (SEM) and 127 ± 6 beats/min, respectively. The arterial oxygen saturation (SaO₂) in room air averaged 76.7 ± 3.0% (range 60.0–89.0%) in group 1, and 71.4 ± 3.7% (range 49.0–89.5%) in group 2. The SaO₂ was higher by approximately 5% in group 1 (NS). The blood hemoglobin content differed (p < 0.05) between groups, although the data were available in only eight patients in group 1 and 13 in group 2. The hemoglobin averaged 15.8 ± 0.4 g%, range 12.8–18.8 g%, in group 1, and 17.6 ± 0.6 g%, range 14.7–23.2 g%, in group 2. In group 1, the end-diastolic pressure was 2–16 mm Hg, average 8.4 ± 0.8 mm Hg, and in group 2 it was 0–13 mm Hg, average 6.8 ± 0.6 mm Hg.

EDVs of the single ventricle of both groups were plotted against BSA (fig. 3). The EDV of single ventricle in group 2 was always larger than that of the normal right or left ventricle, but generally smaller than the combined volumes of the normal right and left ventricles. In contrast, the EDV of single ventricle in group 1 was generally larger than that in group 2, and also larger than the sum of the normal right and left ventricular EDVs. When the EDV of single ventricle was expressed as percentage of the combined volumes of the normal right and left ventricular EDVs (100%), EDVs in groups 1 and 2 averaged 143 ± 11% (73–209%) and 81 ± 4% (54–119%), respectively. The difference in EDVs between two groups was statistically significant (p < 0.001).

The %EDV was plotted against the patient’s age in figure 4. In group 1, 11 of 14 patients (79%) had an increase in EDV to more than 125% of normal. This increase was apparent even in patients younger than 2 years of age. In contrast, in group 2, six of the 14 patients (43%) younger than 2 years of age had a ventricular chamber size of less than 75% of normal.

The average ventricular stroke volume index was 80 ± 9 ml/m² in group 1 and 47 ± 3 ml/m² in group 2 (p < 0.005). The EF averaged 0.49 ± 0.02 in group 1 and was not significantly different from that in group 2 (0.54 ± 0.02). In both groups, the average EF of a single ventricle was significantly (p < 0.001) less than that of a normal left (0.63 ± 0.01⁹, ¹⁹ 0.68 ± 0.01²¹) or right (0.61 ± 0.01⁹ 0.61 ± 0.01²¹) ventricle previously reported by Jarmakani, Thanopoulos and their co-workers.⁹, ¹⁹–²¹ Figure 5 shows the relationship between EF and BSA compared with a normal left ventricular EF. The EF of a single ventricle was generally lower than that of a normal left ventricle, even when corrected for body size.

![Figure 3](http://circ.ahajournals.org/)

**Figure 3. The distribution of end-diastolic volume (EDV) in single ventricle (SV) in groups 1 and 2 is plotted against body surface area (BSA). The formula 72.5 (BSA)¹⁴³ represents a normal left ventricular volume curve, and the formula 147.6 (BSA)¹⁴³ represents the sum of normal right and left ventricular volumes. PS = pulmonary stenosis; PA = pulmonary artery.)
Discussion

Single ventricle is one of the most complicated cardiac anomalies and is usually associated with other abnormalities. With the increasing interest in total correction (ventricular septation, ventricular septum creation surgery), and in functional correction (right atrial-pulmonary artery bypass surgery), an understanding of the ventricular chamber size and function in single ventricle is important, but few studies have been done.

To make the analysis of ventricular function easier in this complicated cardiac anomaly, all the inflow abnormalities except significant AV valvular insufficiency were neglected in this study because a single ventricle receives blood from both atria. Among the factors affecting after-load of the single ventricle, such as pulmonary and systemic vascular resistance and anatomic stenosis or obstruction, only significant angiographic valvular or subvalvular PS was considered. The patients with subaortic or aortic stenosis were not included in this series.

In this study, biplane, large-film angiocardiograms were used to assess ventricular size and function. Selection of the films at end-diastole and end-systole was based on the simultaneously recorded ECGs. This method had certain limitations. First, end-diastolic and end-systolic views could not be obtained in a single beat, and they had to be composed from the different beats. Second, end-diastolic frames were chosen during the PR interval and end-systolic frames were selected at the end of the T wave. End-diastole usually occurs at the end of the PR interval and end-systole usually occurs before the end of the T wave. Thus, it is likely that some end-diastolic volumes were underestimated and some end-systolic volumes overestimated, which might have resulted in an erroneously low EF in some patients. Cineangiographic evaluation is certainly preferable for this purpose.

When a significant angiographic PS was considered, hemodynamic characteristics in single ventricle could
be roughly separated into two groups. The EDV in group 1 averaged 143 ± 11% of normal, and increased chamber size was apparent, even in children younger than 2 years of age. This finding explains the occurrence of congestive heart failure in early childhood. In group 1, ventricular EDV in older children was relatively smaller than that in younger children (fig. 4), which may result from a relative decrease in pulmonary blood flow secondary to an increased pulmonary vascular resistance. Also, improvement of congestive heart failure with age might contribute to a decrease in the relative size of the ventricular chamber in surviving older children. In group 2, 43% of patients had a somewhat underdeveloped ventricle of less than 75% of normal.

In this study, neither pulmonary blood flow nor a pulmonary-systemic flow ratio were measured because of the difficulty in obtaining blood samples from the stenosed or obstructed pulmonary artery. However, because the ventricular stroke volume of group 1 was nearly twice that of group 2, and because hemoglobin concentration in group 1 was significantly less than that in group 2, it is quite likely that the pulmonary blood flow was normal or increased in group 1 patients. The increased pulmonary blood flow seemed to be related to the increased ventricular size in group 1 patients. This relationship has been reported in patients with common AV canal.20, 21

The SaO₂ depends on the pulmonary and systemic blood flows, the mixed venous and pulmonary venous oxygen saturations, and the degree of mixing and streaming in the single ventricular chamber.24 The SaO₂ was approximately 5% higher in group 1 (NS). The pulmonary blood flow is usually limited in patients with severe PS, but blood mixing is usually better in patients with PS than in patients without it.25 Also, the SaO₂ may not differ significantly between the patients with and without PS under sedation during cardiac catheterization. However, the difference in SaO₂ may become quite apparent when the patient is exercised, as suggested by the higher hemoglobin concentration in group 2 than in group 1.

In our study, patients who had severe angiographic PS associated with a higher blood hemoglobin content had a significantly smaller ventricle than patients without PS. Jarmakani et al.19 showed that right ventricular size was significantly less than normal in patients with severe tetralogy and a hemoglobin level > 16 g%. The EF of a single ventricle was slightly but significantly depressed regardless of the presence or absence of PS. Again, however, our ECG-based method of film selection might yield a somewhat lower EF than actually present. The depressed EF was also reported in patients with tetralogy of Fallot26, 27 and tricuspid atresia.28 The cause of a depressed EF in cyanotic patients is not entirely clear, but chronic hypoxemia seems to play an important role. Graham26 and Jarmakani27 and their co-workers reported that the depressed EF persisted even after corrective surgery was undertaken with a resultant normalized SaO₂. Fibrotic myocardial changes in cyanotic patients may also play at least a partial role.29

The assessment of ventricular volume and function in single ventricle may be of great importance in consideration of surgical treatments, particularly when ventricular septation is considered. A detailed experimental study of ventricular septation in dogs has been reported.30 but characteristics of the human single ventricle are different from those of dogs with a normal ventricular volume. Ventricular septation may be indicated in patients with a good EF and an enlarged, normal or nearly normal ventricular volumes compared with combined normal right and left ventricular volumes. Patients with PS and a small ventricular volume may not be suitable candidates for ventricular septation but may be good candidates for a systemic artery-pulmonary artery shunt to increase pulmonary blood flow, with a resultant increase in chamber size.30, 31 Ventricular septation may become feasible after the shunt operation in these patients. We conclude that ventricular size and function should be carefully assessed before surgery in patients with single ventricle.

References
15. Graham TP Jr, Jarmakani JM, Canent RV Jr, Morrow MN:
Left heart volume estimations in infancy and childhood; reevaluation of methodology and normal values. Circulation 43: 895, 1971
Characteristics of ventricular function in single ventricle.
S Kitamura, Y Kawashima, Y Shimazaki, T Mori, S Nakano, S Beppu and T Kozuka

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