Combining all constants results in:

\[ \ln \sigma = \beta + \alpha C \quad (7) \]

where

\[ \beta = \ln (a/k) - k, \quad (8) \]

and

\[ \alpha = k/C_0. \quad (9) \]

If one assumes that muscle length at zero stress does not vary considerably over a short time in a given muscle, then \( C_0 \) would be constant in a first-order approximation and any change of \( \alpha \) due to an intervention should reflect an alteration in the elastic stiffness constant \( k \) that is identical to \( k \) in equation (3). Further, equation (8) demonstrates that intercept \( \beta \) of the logarithmic stress-circumference relation (equation 7) will be influenced by any change in \( k \) and thereby \( \alpha \).

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Characteristics of Ventricular Function in Single Ventricle

**Soichiro Kitamura, M.D., Yasunaru Kawashima, M.D., Yasuhsa Shimazaki, M.D., Tohru Mori, M.D., Susumu Nakano, M.D., Shintaro Beppu, M.D., and Takahiro Kozuka, M.D.**

**SUMMARY**  Functional characteristics of the single ventricle were studied by means of biplane angiocardiography 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,\(^1\)\(^2\) 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.\(^5\)\(^12\) 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 angiocardiograms in 34 patients with a diagnosis of single ventricle confirmed angiographically or at autopsy.

**Methods**

**Patient Population**

Biplane, large-film angiocardiograms (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 angiocardiograms of good quality was the sole criterion for patient selection. All patients were diagnosed as having a single ventricle by the definition reported previously.\(^1\)\(^13\) 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.\(^13\) In brief, type I is identical to the anatomic type A of Van Praagh et al.,\(^1\) 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 ventriculotruncal 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 ventriculotruncal 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 hemiazygos 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 angiocardiology. 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 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 angiocardiology 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

**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 angiocardiology 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 $147.6 \times BSA$ where BSA is body surface area (m$^2$). 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.

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**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.
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 ± 0.01, 19, 20 0.68 ± 0.0121) or right (0.61 ± 0.01; 19 0.61 ± 0.0121) ventricle previously reported by Jarmakani, Thanopoulos and their co-workers.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](image_url)
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),\(^9\)\(^-\)\(^12\) and in functional correction (right atrial-pulmonary artery bypass surgery),\(^22\)\(^,\)\(^23\) an understanding of the ventricular chamber size and function in single ventricle is important, but few studies have been done.\(^24\)\(^,\)\(^25\)

To make the analysis of ventricular function easier in this complicated cardiac anomaly, all the inflow abnormalities except significant AV valvar 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 valvar or subvalvar 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 preferrable for this purpose.

When a significant angiographic PS was considered, hemodynamic characteristics in single ventricle could

**Figure 4.** The relationship between the end-diastolic volume (EDV) and the patient's age (years) is shown. EDV was expressed as a percentage of normal. The sum of the normal right and left ventricular volumes was considered to be 100% in this study; the normal limit was 75–125%. In group 1 (Gr-1) patients, dilatation of the single ventricle (SV) occurred at an early age. In contrast, in 43% of the patients younger than 2 years of age in group 2 (Gr-2), the ventricular volume was somewhat underdeveloped (less than 75% of normal). The mean EDV of group 1 was significantly larger than that of group 2 (p < 0.001). PS = pulmonary stenosis; PA = pulmonary artery.

**Figure 5.** The ejection fraction (EF) was plotted against the body surface area (BSA). The EF was generally depressed in single ventricle (SV) when compared with a normal left ventricular EF, but there was no obvious difference in EF between the two groups. The area between the dotted lines was considered to be the normal range for a normal left ventricle.\(^27\) PS = pulmonary stenosis; PA = pulmonary artery; Gr = group.
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 oc-
currence 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 con-
centration 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 ven-
tricular 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 sur-
gery was undertaken with a resultant normalized
SaO2. Fibrotic myocardial changes in cyanotic pa-
tients 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 con-
sideration of surgical treatments, particularly when
ventricular septation is considered. A detailed ex-
perimental 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.38, 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.

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