The Reliability and Practicality of Single Crystal Echocardiography in the Evaluation of Single Ventricle

Angiographic and Pathological Correlates

ROBERTA M. BINI, M.D., KENNETH R. BLOOM, M.B., F.R.C.P.(C), J. A. GORDON CULHAM, M.D., F.R.C.P.(C), ROBERT M. FREEDOM, M.D., F.R.C.P.(C), CONSTANCE M. WILLIAMS, R.N., AND RICHARD D. ROWE, M.D., F.R.C.P.(C)

SUMMARY A prospective clinical and echocardiographic diagnosis of single ventricle was made in 42 patients. Each was evaluated for the number of atrioventricular (A/V) valves, presence of an outflow chamber (OC), A/V valve-semilunar continuity, and orientation of the great arteries. Angiographic correlations were subsequently obtained in 40 and autopsies in 12. The overall diagnosis of single ventricle was substantiated in 39. Two other patients diagnosed as single ventricle by angiography were thought to have large ventricular septal defects on echocardiography. Tricuspid valve was interpreted as septum in one. The angiographic diagnosis of single ventricle was incorrect in another, correctly diagnosed by echocardiography and confirmed at pathology. The differential diagnosis also included A-V canal, L-transposition of the great arteries, double outlet right ventricle, and tricuspid atresia. This last condition has to be differentiated on clinical evidence. The echocardiograms were of particular value in determining the number of A/V valves. Two great arteries were demonstrated in 74% of patients and their relationship was correctly determined in 79% of these. Both imaging techniques agreed closely as to A/V valve-semilunar continuity and presence of an OC, but both showed some inaccuracies compared to pathological specimens. The echocardiogram helps both in planning catheterization and in evaluating the overall diagnosis.

ECHOCARDIOGRAPHY IS USEFUL in defining many of the morphological features associated with anatomical intracardiac defects. Most studies have taken patients with known defects and defined the echocardiographic anomalies associated with them.1-4 Prospective application of these criteria to the echocardiograms of children with unknown disease has often led to disappointment in the diagnosis. Hitherto unexpected overlaps between various malformations can lead to confusion in arriving at a definitive diagnosis. In addition, patient selection may bias the observer in the interpretation of the findings. We believe that the echocardiogram is part of the clinical evaluation and as such should be interpreted with knowledge of the clinical features, electrocardiogram and chest X-ray.

We have tried to establish the practicality of single crystal echocardiography used in this manner in the diagnosis of the various forms constituting the pathological entity of single ventricle. The combined clinical and echocardiographic diagnosis of single ventricle formed the basis for patient selection. This was then compared with the subsequent angiographic and pathological diagnoses. We have also evaluated those patients who were diagnosed as having single ventricle on the angiogram but misdiagnosed by echocardiography.

The main purpose of this paper is to define the reliability of the echocardiographic features that are associated with the various anatomical subtypes of single ventricle. The differential diagnoses that emerged from this study will be discussed.

Definition of Terms

Outlet chamber (OC)

The small chamber recognized echocardiographically as lying between the anterior cardiac wall and the anterior surface of the bulbo-ventricular ridge. (There is debate as to whether the septum between the main ventricular chamber and the outlet chamber constitutes a true interventricular septum or is a remnant of the primitive bulbo-ventricular ridge.6,7)

Bulbo-ventricular ridge (BVR)

Septal-like structure that divides the OC from the ventricle.

Anterior and Posterior Roots

Roots were not defined as aorta or pulmonary artery unless systolic time intervals or relative size could be evaluated.6,9 They were then defined as normal (N) or D or L transposed.8,10,11

Atrioventricular (A/V) valves

One or two were identified and called anterior or posterior. The A/V valve ring or anulus (A/VR) was also identified. In the event of an A/V valve being continuous with a semilunar valve, then that A/V valve was considered either a mitral or the mitral portion of a common A/V valve.11

Single Ventricle

There is no unanimity as to the origin, morphology, or terminology of the single ventricle.6,7,12-18 We feel that a practical clinical approach is best served by the hypothesis advanced by Anderson et al.8
We used the term single ventricle to designate functionally univentricular hearts, and then defined the number of A/V valves, relationship of the great arteries, and presence or absence of an outlet chamber.

Methods

Patients

Forty-two children aged one day to seventeen years (mean 0.8 yr) were studied by echocardiography and assessed as having single ventricle. Forty patients underwent cardiac catheterization and ten of these, who subsequently died, underwent autopsy examination. Two patients died prior to cardiac catheterization but did have autopsy examinations and have been included. In addition, two patients who had an angiographic diagnosis of single ventricle but in whom the echocardiogram was considered not to be typical of this disease have been evaluated. Patients who had echocardiographic features of single ventricle with single A/V valve, mitral semilunar continuity and who also had the classical electrocardiographic features of tricuspid atresia were excluded.

Echocardiography

Clinical, electrocardiographic, and radiological findings were known to the person interpreting the echocardiogram. All patients were studied prior to cardiac catheterization. The method of performing these examinations is crucial for an accurate diagnosis and must, therefore, be considered in detail.

Commercially available ultrasonoscopes (either Hoffrel 101C or Ekoline 20A), each interfaced to a Cambridge recorder, were used. Transducers were appropriate to patient size varying from 5 to 2.25 MHz with various diameters and focal lengths. Infants and children were studied in the recumbent position or in the shallow right anterior oblique position. Those with dextrocardia were frequently studied in the left anterior oblique position with the transducer to the right of the sternum.

One A/V valve was identified initially and the transducer positioned perpendicular to this. The transducer was then angled inferiorly until the posterior wall endocardium was seen. Some additional medial or lateral angulation was usually required depending on the location of the cardiac apex. The transducer was then angled along the long axis of the ventricle. Ventricular alignment was frequently abnormal. The echo obtained from the A/V valve was, therefore, utilized to facilitate this sweep in the long axis, as A/V valve leaflets tend to open along this axis. This was done by keeping the echo from the valve as undistorted as possible as one angled the transducer along the ventricle. The A/V valve ring or ring-like structure that could represent the interatrial septum was identified and a search made for a second A/V valve on the opposite side of this structure either anterior or posterior to the initial A/V valve that had been found. If only one A/V valve was identified from a single transducer position, the transducer was moved across the precordium to alternative positions. The finding of a second A/V valve at an apparent different depth from the first was then followed by a sweep of the transducer to the position in which the first A/V valve was found. A single A/V valve was defined as an unbroken echo of varying amplitude and might have apparent varying depth. Two separate A/V valves were defined 1) when the anterior A/V valve passed through a ring-like structure (anulus) before the posterior A/V valve was defined (fig. 1) and 2) if the A/V valves could be viewed simultaneously from a single transducer position within the ventricular cavity (fig. 2). A further sweep from each A/V valve was then attempted to try to find a semilunar valve either continuous with it or arising off a conus (figs. 3 and 4). If a root could not be found in this way, the transducer was then repositioned on the chest wall until a root with leaflets was found. The transducer was then angled inferiorly to one or other of the previously demonstrated A/V valves in turn, in order to define continuity. This was established when the A/V valve leaflet merged directly with the posterior wall of either root (fig. 3). Discontinuity was diagnosed when a co-nal structure was demonstrated between the semilunar anulus and A/V valve leaflet (fig. 4). A search was then made for a second root by angling the transducer medially.

FIGURE 1. Two A/V valves are shown on this continuous recording with the anterior A/V valve passing through a ring-like structure separating it from the posterior A/V valve.

FIGURE 2. Two A/V valves recorded simultaneously in the same ventricular chamber, the transducer was kept in a steady position. Compare with figure 9.
or laterally. The position of the other root in relation to the initial one found was then recorded. This formed the anatomical basis for establishing great artery relationship. Systolic time intervals were determined whenever possible to help identify systemic and pulmonary roots. For this purpose recordings of semilunar valve motion were made at 100 mm/sec. A shorter pre-ejection period and longer ejection time characterized the pulmonary valve and helped differentiate it from the aortic valve. Total electromechanical systole was longer for the pulmonary valve measurements. These differences were of particular value in the absence of pulmonary hypertension and were accentuated by pulmonary stenosis.

The finding of an anterior root that was medially situated and a posterior root with a diameter of less than 75% of the anterior root was taken as evidence of transposition with pulmonary stenosis. The relationship of this second root to the atrioventricular valves was also defined. All relationships were considered with knowledge of the position of the cardiac apex as defined on chest X-ray. The minimum information required to define great artery anatomy was the definition of two roots and knowledge of the spatial relationship of each to the other.

Finally, several angulations with high and low positions of the transducer were evaluated across the assumed minor axis.
of the ventricle to attempt to demonstrate the presence of an outlet chamber (fig. 5a).

**Angiography**

Biplane cineangiograms were performed at routine cardiac catheterization. This was usually done within 48 hours of the echocardiographic study. These were evaluated with regard to atrial situs, ventricular morphology, and great vessel relationship.²⁰-²² The OC was assessed²⁴ with regard to size and position (fig. 5b). Vessels were described as to their relationship and the presence and level of stenosis.²⁰,²² The A/V valves were usually seen as negative filling defects within the ventricular cavity (figs. 6 and 7).²⁴ The angiograms were evaluated by one of us without any knowledge of the echocardiographic findings.

**Pathology**

Morphological descriptions of the hearts obtained at necropsy were evaluated by another author without knowing either the echocardiographic or angiographic diagnoses.

The results of all these techniques were then compared.

**Results**

Three patients were shown to have been misdiagnosed by echocardiography. These patients together with two patients diagnosed angiographically as single ventricle but who had different echocardiographic diagnoses, will be considered separately. All data in tables 1 through 4 refer to the remaining 39 patients, and include patient 2 who was not thought to have a single ventricle angiographically but was shown to have this on autopsy.

The clinical features are shown in table 1.²⁶-²⁸ The low mean age reflects the fact that a large number of patients were neonates (22 patients were less than one month of age).

The echocardiographic features of the 39 patients are defined in table 2. The relationship of the roots is marked as

---

**FIGURE 5.** Echocardiogram (left) and angiogram (right) showing outlet chamber (OC). The bulbo-ventricular foramen is seen between the ventricle (V) and OC on the angiogram and the bulbo-ventricular ridge (BVR) is seen on the echo.

**FIGURE 6.** Two A/V valves with separate orifices outlined by non-opacified blood in diastole.
indeterminate where only one root was recorded or where identification was uncertain. Twenty-four patients (62%) had only one A/V valve. This is in agreement with previously published pathological reports.\textsuperscript{13, 29} Discontinuity was more commonly diagnosed where no OC was defined, and these patients also had a high incidence of pulmonary atresia.

Angiographic and echocardiographic findings were compared. For this purpose, patients were grouped depending on the number of A/V valves present as defined by the echocardiogram (table 3). Autopsy correlates are shown in table 4.

A/V valves

Good correlation was found with agreement in 85% of patients. One patient (\#21) diagnosed echocardiographically as having only one A/V valve was though to have two A/V valves on angiography, and this was confirmed at autopsy. Two other patients (\#2, 31) had two A/V valves demonstrated echocardiographically, were thought to have only one on angiography but were found to have two discrete A/V valves at autopsy. The other three cases remain unsubstantiated.

Continuity

Agreement was less (79%). None of the patients in whom continuity was argued came to autopsy, but both imaging techniques were found to be unreliable in 40% of the patients at autopsy.

Relationship of Great Arteries

Sufficient information was available to attempt to define this in 29 patients (74%). There was agreement in all five patients with normally related great arteries, six of eleven patients (54%) with D-TGA and twelve of thirteen patients (92%) with L-TGA. Eight of the remaining patients, all of whom had one A/V valve and no OC, had indeterminate great artery relationships by echocardiography, as neither

\begin{table}
\centering
\caption{Clinical Features}
\begin{tabular}{l|c}
\hline Age & 1d–17 yrs \\
\hline ECG & \\
QRS axis & 0–90 \\
& 91–180 \\
& 181–270 \\
& 270–359 \\
P axis & 0–90 \\
& 91–180 \\
& 270–359 \\
RVH & \\
LVH & \\
CVH & \\
No hypertrophy & \\
Cyanosis & 23 \\
CHF & 16 \\
CXR & \\
\hline
\end{tabular}
\end{table}

\begin{table}
\centering
\caption{Echocardiographic Findings}
\begin{tabular}{l|c}
\hline One A/V Valve Present & 24 \\
Continuity & \\
D-TGA & 6 \\
L-TGA & 2 \\
N & 1 \\
Ind. & 3 \\
Discontinuity & \\
D-TGA & 18 \\
L-TGA & 6 \\
N & 3 \\
Ind. & 1 \\
\hline Two A/V Valves Present & 15 \\
Continuity & \\
D-TGA & 5 \\
L-TGA & 2 \\
N & 2 \\
Ind. & 1 \\
Discontinuity & \\
D-TGA & 10 \\
L-TGA & 2 \\
N & 7 \\
Ind. & 0 \\
\hline
\end{tabular}
\end{table}

their anatomical relationships nor systolic time intervals were adequately defined. Four were shown on angiography to have D-TGA, three A/P type of transposition and one L-TGA. Only one root was recorded in five patients, four of whom had pulmonary atresia and one pulmonary stenosis. Both imaging techniques were found to be fallible in patient

Abbreviations: CHF = congestive heart failure; RVH = right ventricular hypertrophy; LVH = left ventricular hypertrophy; CVH = combined ventricular hypertrophy; CXR = chest X-ray; Vasc. = vascularity; Pul. = pulmonary; L. Juxta. A.A. = left juxtaposition of atrial appendages.

Abbreviations: Ind. = indeterminate; TGA = transposition of the great arteries; N = normally related great arteries.
#7 where pathological examination showed A/P transposed rather than L malposed great arteries.

Outlet Chamber

The definition of an outlet chamber by the echocardiogram was supported by angiography in all patients. All were confirmed in the four patients who came to autopsy.

Absence of an OC by echocardiography was not a reliable feature. Nine of nineteen patients (47%) had an OC demonstrated angiographically. However, angiography also underestimated the incidence of an OC as shown by the autopsy examinations in patients 26 and 36.

Diagnosis

The echocardiographic diagnosis of single ventricle was confirmed in 36 of the 40 patients (90%) who had had angiography. The angiographic diagnoses in the four patients in whom there was no agreement were: 1) double outlet right ventricle with L-ventricular loop; 2) double outlet right ventricle of the Taussig-Bing type; 3) A/V canal with a large ventricular septal defect in a patient with dextrocardia; 4) multiple ventricular septal defects, anatomically corrected malposition and dextrocardia (Table 5). This patient (2) was shown at autopsy to have single ventricle with a large high outlet chamber and normally related great arteries.

Malposition of the cardiac apex did not alter the reliability of the echocardiographic assessment.

Two patients who had angiographic diagnoses of single ventricle were diagnosed as ventricular septal defects by echocardiography. These echocardiograms have been reviewed retrospectively, and the tricuspid valve echo was confused as septum in one case (fig. 8). The other diagnosis remains a matter of dispute.

Overall, single ventricle was correctly diagnosed by echocardiography in 39 of 44 patients (89%). The diagnosis was incorrect in three patients and was missed in two. The angiographic diagnosis was shown by autopsy to be incorrect in one patient.

**Discussion**

This study differs in some aspects from recently published accounts. The majority of patients in those studies were older, had two A/V valves and normal visceral situs. Chesler’s earlier series was also selected from those patients in whom an angiographic diagnosis had been made. None of these authors have attempted to define great artery relationship.

**Typical Echocardiographic Features of Single Ventricle**

**A/V Valves**

The echocardiographic feature said to be typical of single ventricle is the presence of two A/V valves without an intervening interventricular septum (fig. 2). This alone can lead to misinterpretation as shown in figure 9 where both A/V valves were recorded simultaneously in a patient with a moderate ventricular septal defect. We have found the sweep
along the long axis of the ventricle to be of great value in defining two A/V valves when these are not visualized simultaneously. An A/V valve ring or ring-like structure separating one valve from the other should be demonstrated (fig. 1). This is particularly important as a single A/V valve may have a large amplitude. A single A/V valve viewed from different angles can appear to be two A/V valves (fig. 10). Some patients with single ventricle may have a large common A/V valve or single A/V valve ring with atresia of the other A/V valve. We do not believe that these can be distinguished echocardiographically as the amplitude of an A/V valve may depend on hemodynamic as well as anatomical factors.

In this series the echocardiographic appearance of straddling A/V valve was noted occasionally. Definition of the anterior leaflet going through the bulbo-ventricular ridge into the OC and evaluation of the degree of straddling are important features. The use of contrast techniques may add accuracy to the evaluation of the A/V valves.

Presence of an OC

We found this to be an unreliable feature by either technique. Seward et al. and Williams et al. both stress a high transducer position with inferomedial direction for dis-

**FIGURE 8.** A structure showing septal-like motion (arrow). The middle and right panels show that this is probably the tricuspid valve viewed from different angles.

**FIGURE 9.** Left) Two separate A/V valves, without intervening ventricular septum, recorded with the transducer in a steady, single position in a patient with moderate ventricular septal defect. Right) By scanning along the ventricular axis the interventricular septum has been demonstrated clearly.
play of this chamber. We have used this and still have found it difficult to detect this chamber in almost half of our patients where it was small and unusually situated. This agrees with the findings of Beardshaw et al.23 All patients who did not have an OC by echocardiography and who came to autopsy were found to have an OC. The echocardiographic demonstration of a small anterior chamber (fig. 5) raised several diagnostic possibilities. The echocardiogram of the patient with tricuspid atresia, VSD, and hypoplastic right ventricle was indistinguishable from that of single ventricle with OC. Classical clinical features of tricuspid atresia help in the diagnosis, and we have not had problems differentiating the two conditions on the basis of a combined clinical and echocardiographic approach.

A small outlet chamber may be seen in patients with otherwise uncomplicated ventricular inversion due to the abnormal orientation of the septum.4 Severe forms of A/V canal have been recognized previously as a diagnostic dilemma in this condition.4, 36, 37 and we confirm this. Double outlet right ventricle may also present a problem.

**Great Vessel Relationship**

Using a combination of anatomical relationships and systolic time intervals, we attempted to define this relationship in 74% of patients. We were correct in 79% of these. This adds another feature to the total assessment due to the high incidence of malposition associated with this malformation. Difficulties in assessing great vessel relationships occurred in patients with associated pulmonic stenosis or atresia and, in our experience, with one A/V valve and no echocardiographic OC.

**Continuity and Discontinuity**

The demonstration of continuity was correct in 73% of patients and supported by pathology. Discontinuity was less reliable when predicted by either imaging technique. The variability of continuity as described by Rosenquist et al.38 and the frequently unusual orientation of the outflow tract and great arteries explains the difficulty in demonstrating continuity in some patients.

**Practical Use**

An echocardiogram should be performed in all patients suspected of having single ventricle prior to cardiac catheterization and should be considered part of the general clinical evaluation. By combining echocardiographic and clinical features, a higher degree of accuracy may be achieved both in making the overall diagnosis of single ventricle and in defining many of the anatomical features. This information can then be utilized in planning the catheterization. This has great value particularly in neonates as one can plan specific angiographic views and so make maximum use of the limited amount of contrast medium that can safely be used for angiography in this age group.

**Acknowledgment**

We wish to thank Ms. A. Di Giacomo for her secretarial help.

---

**Figure 10.** The same large, common A/V valve recorded from different transducer positions. An intervening A/V valve ring is not seen. Compare with figure 1.
References

The reliability and practicality of single crystal echocardiography in the evaluation of single ventricle. Angiographic and pathological correlates.
R M Bini, K R Bloom, J A Culham, R M Freedom, C M Williams and R D Rowe

Circulation. 1978;57:269-277
doi: 10.1161/01.CIR.57.2.269

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/57/2/269