Echocardiography of primitive ventricle

CARLOS MORTERA, STEWART HUNTER, GORDON TERRY, AND MICHAEL TYNAN

From the Department of Cardiology, Newcastle General Hospital, Westgate Road, Newcastle upon Tyne

The angiocardiographic, echocardiographic, and, where available, the necropsy findings were correlated in 32 cases of primitive ventricle. Single probe echocardiography was shown to be a reliable and accurate technique for diagnosis of primitive ventricle; the ventricular and atrioventricular valve appearances were characteristic, and the outlet chamber was usually recognised when present, though it was not possible to say whether it was right or left sided. Abnormalities of the atrioventricular valves were more accurately shown by echocardiography than by angiocardiography though the two techniques were shown to be complementary in the overall diagnostic process.

The introduction of radical surgery for the treatment of univentricular hearts makes accurate anatomical and functional diagnosis essential. The peculiar ability of the echocardiogram to define intracardiac structures, their relations, and their motion, suggests that the technique should play an important role in this diagnostic process. The definitions and terminology used in this paper are those proposed by Anderson and his colleagues (Anderson et al., 1976). Thus the term primitive ventricle is applied to all hearts with absence of the posterior interventricular septum and includes those in which there is atresia of either atrioventricular valve (Quero 1970, 1972).

By correlating echocardiography and angiocardiography and, where available, necropsy material, we have set out to define the value of echocardiography in the assessment of primitive ventricle.

DEFINITIONS

Primitive ventricle

This exists when there is failure of development of the posterior interventricular septum and both atrioventricular valves are in actual or potential communication with the main ventricular chamber (Anderson et al., 1976). When an outlet chamber is present no complete atrioventricular valve annulus is in actual or potential communication with it (Macartney et al., 1976). When a single atrioventricular valve is present it communicates with the main ventricular chamber and no other atrioventricular valve rudiment potentially communicates with the outlet chamber (Anderson and Becker, 1975).

Outlet chamber

This is a chamber with a trabeculated pouch and an infundibular portion; it communicates with the main ventricular chamber but contains neither a complete atrioventricular valve annulus nor the complete tension apparatus of an atrioventricular valve. Echocardiographically its parietal wall moves towards the bulboventricular septum during ventricular systole and no atrioventricular valve is seen completely within its boundaries.

Atrioventricular valve nomenclature

The terms right and left atrioventricular valves are used to avoid confusion. The anatomical right atrioventricular valve is echocardiographically the anterior and the left atrioventricular valve the posterior one. The terms right and left atrioventricular valve are used in this sense throughout.

Connections of great arteries

In primitive ventricle with outlet chamber the great arteries are considered to be normally connected when the aorta arises from the main chamber and the pulmonary artery from the outlet chamber. When the converse is true transposition exists (Shinebourne et al., 1976). When both great arteries arise from one chamber whether an outlet chamber is present or not, this is a double outlet chamber and the term malposition is used irrespective of the anteroposterior relation of the great arteries (Shinebourne et al., 1976). The prefixes d and l

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indicate only the lateral relation of the great arteries
and have no specific morphogenetic or diagnostic
significance (Carr et al., 1968), though knowledge
of these relations is of obvious surgical importance.
When a single arterial trunk is present the termino-
logy of Shinebourne et al. (1976) is used.

**Straddling atrioventricular valves**

In the context of primitive ventricle, straddling
refers to an atrioventricular valve, part of whose
tension apparatus arises in the outlet chamber.
This atrioventricular valve then straddles the bul-
boventricular septum (Anderson et al., 1976). A
straddling valve is assigned to the ventricular
chamber into which more than 50 per cent of its orifice appears to open. Thus, in primitive ventricle
all straddling valves open at least 50 per cent into
the main chamber.

**Subjects and methods**

Echocardiograms were recorded in 32 patients aged
from 1 day to 28 years with an angiocardiacdiographic
diagnosis of primitive ventricle in 31, necropsy
confirmation of the angiocardiacdiographic diagnosis in
4, and necropsy diagnosis only in 1 case in whom
angiocardiacography was not performed. In all 31
patients who had angiocardiacography main chamber
ventriculograms were recorded using either biplane
Elema or multiple single plane cine projections.
Single probe echocardiograms were obtained using
a 2·25 MHz transducer focused at 7·5 cm with an
Echoview X echocardiograph machine and a
Honeywell ultraviolet recorder. In all cases the
echocardiographic relations were established using
continuous sweeps. In 20 patients the angiograms
had been performed before echocardiacography but
were reviewed by one of us without knowledge of
the echocardiacographic findings. In 12 the echocar-
diacographic assessment and diagnosis had been
made before the angiocardiacographic studies.

Main ventricular chamber size and performance
were assessed echocardiographically using end-
diastolic dimension and the mean rate of internal
dimension shortening (mean V<sub>CP</sub>) respectively
(Cooper et al., 1972; Kaye et al., 1975). These
data were compared with values for the normal
left ventricle (Hunter et al., 1975).

**Results**

Table 1 shows the final diagnoses of all patients;
these diagnoses are a synthesis of all the available
data obtained from angiocardiacograms, echocardiac-
grams, and necropsies.

Thirty patients had primitive ventricle with an

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<td>Primitive ventricle with outlet chamber</td>
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<td>With 2 atrioventricular valves</td>
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<td>Normal great arteries</td>
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<td>Transposition of great arteries (TGA)</td>
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<td>With 1 atrioventricular valve</td>
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<td>Right atrioventricular valve atresia with TGA</td>
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<td>Right atrioventricular valve atresia with normal great arteries</td>
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<td>Left atrioventricular valve atresia with TGA</td>
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<td>Left atrioventricular valve atresia with single arterial trunk (aorta)</td>
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<tr>
<td>Primitive ventricle without outlet chamber</td>
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<tr>
<td>With 2 atrioventricular valves and single arterial trunk (aorta)</td>
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<td>With single (common) atrioventricular valve and l-malposition</td>
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**ECHOCARDIOGRAPHY**

In 28 of the cases with an outlet chamber its
presence was detected by echocardiacography as an
interior echo free chamber (Fig. 1, 3, and 8). How-
ever, it was not possible to say with certainty
whether the outlet chamber was to the right or to
the left. The septal structure separating the outlet
chamber from the main chamber was taken to
represent the bulboventricular septum (Felner
et al., 1976) because of its abnormal anterior posi-
tion and because no complete atrioventricular valve
was detected in front of it. In no case were there
echoes suggesting the presence of a posterior ven-
tricular septum (Assad-Morell et al., 1974; Felner
et al., 1976).
In all 18 patients with 2 atrioventricular valves, both were detected. In each case, both valves lay within the main ventricular chamber (Fig. 1). In 3 patients part of the anterior cusp of the right atrioventricular valve crossed the bulboventricular septum into the outlet chamber. This was taken to indicate straddling, as the major part of the valve, the posterior cusp, was within the main ventricular chamber (see Fig. 3) (Seward et al., 1975; Lacorte et al., 1976). In all patients with 2 atrioventricular valves, these structures could be seen in the same echocardiographic plane without an intervening septal structure (Felner et al., 1976) (Fig. 2, 3, and 4), and in 13 of them both valves were shown simultaneously (Fig. 2). The diastolic movement of the posterior cusp of the right atrioventricular valve was frequently greater than the diastolic excursion of the anterior cusp (Fig. 2), though this finding was not invariable (Fig. 1). The posterior excursion of the right atrioventricular valve and the anterior excursion of the left atrioventricular valve were such that they overlapped in diastole (Fig. 2 and 3). In one patient the right atrioventricular valve was mobile but hypoplastic and unequivocally communicated with the main ventricular chamber. Diastolic vibration of the left atrioventricular valve was present in 7 cases (Fig. 4).

A single fully mobile atrioventricular valve was detected in 14 cases echocardiographically, but in 11 it was not possible to differentiate a common atrioventricular valve (Fig. 5B) from right or left atrioventricular valve atresia (Fig. 5A and 5C). In the remaining 3 cases a structure was seen whose movement and position suggested that it was a rudimentary atrioventricular valve. In all 3, this rudiment appeared potentially to communicate with the ventricular chamber (Anderson and Becker, 1975; Macartney et al., 1976). In 2 it was anterior to the fully formed atrioventricular valve (Fig. 6), and right atrioventricular valve atresia was shown on angiography. In the third case the rudiment was located posteriorly (Fig. 7) and left atrioventricular valve atresia was confirmed angiographically.

In 30 cases the posterior wall of the posterior great artery or the single arterial trunk and the left atrioventricular valve or single atrioventricular valve appeared echocardiographically to be connected (Fig. 8A). In the 30 cases with an outlet chamber there was no consistent relation between the bulboventricular septum and the anterior wall of the
Fig. 2 In this echocardiogram the transducer has been swept from the posterior great artery down into the main ventricular chamber, within which lie 2 atrioventricular valves. The posterior cusp of the right atrioventricular valve shows excessive posterior diastolic movement. There is no evidence of a posterior ventricular septum and the atrioventricular valves touch in diastole. PGA, posterior great artery; RAV, right atrioventricular valve; LAV, left atrioventricular valve.

Fig. 3 This echocardiogram shows 2 atrioventricular valves and an outlet chamber. The left atrioventricular valve lies wholly within the main chamber. The right atrioventricular valve lies mainly within the main chamber but its anterior cusp crosses the bulboventricular septum into the outlet chamber. OC, outlet chamber; RAV, right atrioventricular valve; LAV, left atrioventricular valve.
posterior great artery (Fig. 8A and 8B). Though the right atrioventricular valve and the posterior great artery were seen frequently at the same echocardiographic depth, we failed to show a consistent pattern of connection between these two structures. In 21 cases 2 great arteries were definitely identified though it was not possible to differentiate aorta from pulmonary artery by echocardiography. In all the other cases only one great artery was definitely seen and in 2 of these the diagnosis by angiocardio-

ography was single arterial trunk (aorta). There was no obvious reason why only one great artery could be identified echocardiographically in the remaining 9 cases. However, 7 of these had right atrioventricular valve atresia with normally related great arteries suggesting that in this variant of primitive ventricle it is difficult to show the anterior great artery echocardiographically.

Echocardiographic estimates of main ventricular chamber size and performance

The dimensions and mean rates of internal di-

Fig. 4 This echocardiogram shows 2 atrioventricular valves lying within the main ventricular chamber. The right valve is only partially seen but its posterior cusp touches the anterior cusp of the left valve in diastole. The left valve shows conspicuous fine vibration of both cusps in diastole. RAV, right atrioventricular valve; LAV, left atrioventricular valve.

Fig. 5 This figure shows 3 examples of single atrioventricular valve lying within a primitive ventricle. The appearances are closely similar, and are recorded in (A) a case of right atrioventricular valve atresia; (C) a case of left atrioventricular valve atresia; (B) a case of common atrioventricular valve.
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Fig. 6 A large mobile posterior left atrioventricular valve is seen lying within a primitive ventricle. Anterior to it and within the main ventricular chamber a small less mobile atrioventricular valve structure is seen. Catheterisation and angiocardiography confirmed that this patient had right atrioventricular valve atresia. OC, outlet chamber; LAV, left atrioventricular valve; R, rudimentary right atrioventricular valve.

Fig. 7 This patient was shown by angiocardiography to have primitive ventricle with left atrioventricular valve atresia. The echocardiogram shows a right atrioventricular valve with excessive posterior diastolic excursion. Posteriorly a poorly formed left atrioventricular valve structure is seen within the primitive ventricle and touches the anterior valve in diastole. RAV, right atrioventricular valve; R, rudimentary left atrioventricular valve.
mension shortening are given in Table 2. These measurements were made in 30 of the 32 patients. The end-diastolic dimensions were compared with normal values for the left ventricle obtained from 409 normal subjects and were found to be greater than 2 standard deviations larger than the mean normal for age in 20, greater than 1 standard deviation larger in 5, and less than the mean normal for age in only 1 case. The mean rate of internal dimension shortening was less than 1.0-1 in 8, but in only 1 case was it less than 0.8-1. When compared with our normal data, mean $V_{CR}$ was within 2 standard deviations of the mean in 21 patients, more than 2 standard deviations above the mean in 5, and more than 2 standard deviations below the mean in 4.

Discussion

The echocardiographic approach in primitive ventricle does not differ radically from that in other forms of congenital heart disease. The complexity of the lesions encountered necessitates a logical approach to the diagnosis, a picture being built up using continuous sweeps based on structures with recognisable echocardiographic features. After the standard scans have been performed it is possible to show the relations of the atrioventricular valves to each other, to the ventricular cavities, and to the great arteries by appropriate modifications of the angle of sweep. Careful attention to the near gain

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EDD, end-diastolic dimension; ESD, end-systolic dimension; $V_{CR}$, mean rate of internal dimension shortening.

*Fig. 8 These echocardiograms show the variability of the relation between the posterior great artery, the bulboventricular septum, and the left atrioventricular valve. The upper picture (A) shows echocardiographic connection of the posterior wall of the posterior great artery and the left atrioventricular valve. In the lower picture (B) there is straddling of the bulboventricular septum by the right atrioventricular valve. These examples also show the variable relation between the posterior great artery and the right atrioventricular valve. OC, outlet chamber; PGA, posterior great artery; LAV, left atrioventricular valve; RAV, right atrioventricular valve.*
control is necessary to identify the presence or absence of superficial structures such as the outlet chamber. We emphasise, therefore, that in general no single echocardiographic slice can be diagnostic in congenital heart disease.

Using this approach we have been able to show good agreement between the echocardiographic findings and the anatomical features assessed by angiography and necropsy. Angiography is in general superior to echocardiography in the definition of ventricular anatomy and the ventriculo-arterial connections. However, in the examination of the atrioventricular valves, the two methods are complementary. For example, the detection of hypoplastic or atretic atrioventricular valve rudiments is possible with the echocardiogram but, if valve rudiments are not recognised, the angiogram is necessary to decide which atrioventricular valve is atretic. In our group of patients the complementary roles of these two methods are well shown by the finding of a range of right and left atrioventricular valve abnormalities from hypoplasia to rudimentary valve structures in actual or potential communication with the main ventricular chamber, providing further support for the concept of Anderson and Becker (1975) that the majority of cases of ‘tricuspid atresia’ do in fact have a primitive ventricle with right atrioventricular valve atresia.

The echocardiographic diagnosis of primitive ventricle may be summarised as follows. When there are 2 atrioventricular valves and an outlet chamber the following echocardiographic features are diagnostic: (i) Both atrioventricular valves occupy one ventricular cavity with their diastolic motions overlapping. (ii) There is an anterior chamber showing a ventricular type of free wall motion and containing no atrioventricular valve completely within its boundaries. (iii) There is absence of echoes suggesting a posterior interventricular septum. Additionally the posterior diastolic motion of the right atrioventricular valve may appear to be excessive but this is by no means a universal finding. If no outlet chamber is present the demonstration of criteria (i) and (iii) is strongly suggestive of the diagnosis of primitive ventricle.

If only 1 atrioventricular valve is present then the demonstration of an outlet chamber as defined above is strongly suggestive but not diagnostic. If, however, rudiments of an atretic valve potentially communicating with the main chamber are found, then this together with criteria (ii) and (iii) is diagnostic. When one atrioventricular valve is shown but no echoes are seen from an outlet chamber or atretic valve rudiments, then the diagnosis cannot be made with certainty by echocardiography alone.

These diagnostic criteria may appear rather cautious but suggestive evidence such as the increased excursion of the posterior cusp of the right atrioventricular valve may be misleading because a wide range of atrioventricular valve amplitude of motion can be seen in primitive ventricle. Furthermore, negative echocardiographic findings such as the absence of posterior septal echoes or the absence of atrioventricular valve echoes in the outlet chamber are potentially misleading when taken in isolation.

Straddling of the right atrioventricular valve appears to occur more frequently in association with primitive ventricle (Anderson et al., 1976) than with other abnormalities (Liberthson et al., 1971). Thus, if echocardiographic straddling of this valve is demonstrated, primitive ventricle must be definitely excluded before an alternative diagnosis is made. In our experience, however, in the presence of straddling right atrioventricular valve, there still exists an area of diagnostic difficulty between primitive ventricle and other conditions associated with straddling, e.g. transposition of the great arteries with ventricular septal defect (Liberthson et al., 1971). Though valuable clinical information about the ventriculoarterial connections and the great arteries can be obtained echocardiographically, this information is not essential for the diagnosis of primitive ventricle which depends on examination of the intraventricular structures. Furthermore, angiocardiography can still more reliably define ventriculoarterial connections.

Not surprisingly the majority of patients in this series had main ventricular chamber end diastolic dimensions which were considerably larger than the normal left ventricular dimensions. In spite of this, the performance of the main chamber was found to be at least comparable to that of the normal left ventricle in the majority of cases, and in none was severely impaired.

In the diagnosis of primitive ventricle, as in other forms of complex congenital heart disease, echocardiography does not supplant or replace cardiac catheterisation and angiocardiography. However, it has a very valuable part to play in the overall diagnostic process. Echocardiograms provide information about atrioventricular valve structure and function, which is more accurately and easily obtained in this way than by other techniques. In addition, information about the performance of the main ventricular chamber may well be of great importance in the preoperative assessment of primitive ventricle (F. Fontan, 1976, personal communication).
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References


Requests for reprints to Dr. Stewart Hunter, Department of Cardiology, Newcastle General Hospital, Westgate Road, Newcastle upon Tyne NE4 6BE.