Two-Dimensional Echocardiographic Spectrum of Univentricular Atrioventricular Connection

JAMES C. HUHTA, MD,* JAMES B. SEWARD, MD, FACC, ABDUL J. TAJIK, MD, FACC, DONALD J. HAGLER, MD, FACC, WILLIAM D. EDWARDS, MD, FACC

Rochester, Minnesota

The spectrum of anomalies in hearts having a univentricular atrioventricular (AV) connection was examined by two-dimensional echocardiography in 183 patients and the anatomic findings were compared with angiography. The mode of AV connection was found to be of three types: 1) *double inlet* via two AV valves; 2) *single inlet* via one AV valve with absence of the other (left or right AV valve atresia); and 3) *common inlet* via a common AV valve. Identification of an accessory chamber by two-dimensional echocardiography was possible with 90% sensitivity, but it was limited compared with angiography in patients with severely hypoplastic anterior chambers and pulmonary valve atresia. All patients with subaortic outlet foramen obstruction were detected. Great artery position and the presence of obstruction to pulmonary flow were correctly predicted in all but one patient. Two-dimensional echocardiography was superior to angiography for the detection of AV valve abnormalities which were present in 27% and included abnormal chordae, hypoplasia or dysplasia of either valve. Two-dimensional echocardiography should play an essential role in the complete preoperative assessment of patients with univentricular AV connection.

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Current concepts of abnormal atrioventricular (AV) connection are based on descriptions of pathologic material (1–4). By definition, a univentricular AV connection is totally or predominantly to one ventricular chamber by way of two AV valves or one AV valve (when only one valve is present) (1). Until recently, angiography was the only available preoperative technique to evaluate patients with abnormal AV connection. Now, two-dimensional echocardiography can define intracardiac structures with sufficient detail to allow confident noninvasive diagnosis (5).

We reviewed our extensive experience in univentricular AV connection to formulate a logical, segmental approach to noninvasive diagnosis. Two questions were considered: 1) what is the spectrum of AV connection which is "univentricular," and 2) how does two-dimensional echocardiography compare with angiography for the diagnosis of associated anatomic abnormalities?

From the Divisions of Pediatric Cardiology, Cardiovascular Diseases and Medical Pathology, Mayo Graduate School of Medicine, Mayo Clinic and Foundation, Rochester, Minnesota. This study was presented in part at the American Heart Association Annual Meeting, Dallas, Texas, 1981. Manuscript received March 12, 1984; revised manuscript received June 25, 1984, accepted July 13, 1984.

*Current address and address for reprints: James C. Huhta, MD, The Lillie Frank Abercrombie Section of Pediatric Cardiology, Texas Children's Hospital, 6621 Fannin, Houston, Texas 77030.

Terms

Atrioventricular connection. Atrioventricular (AV) connection denotes the manner in which the atria anatomically join the ventricles. In univentricular AV connection, the atria join a single main ventricular chamber. There may be two atria or a common atrium connecting to the main chamber by way of two AV valves (double inlet) (Fig. 1), one AV valve (with atresia of the other, single inlet) (Fig. 2) or a common AV valve (common inlet) (Fig. 3). Because valvular morphology is variable in hearts with two AV valves, we do not use the term "mitral" or "tricuspid" to refer to these valves, but use "right" or "left" AV valve to avoid confusion.

Accessory chambers. Accessory chambers are a part of the ventricular mass but do *not* receive at least half of an AV valve orifice. A ventricular accessory chamber that gives rise to one or both great arteries is designated an *outlet chamber*. Accordingly, it may be subaortic, subpulmonary or double outlet and generally occupies the anterosuperior surface of the heart, either to the left or right of the midline. If the accessory chamber does not give rise to a great artery, then it is designated a *pouch*. These usually occupy an inferior or lateral position and often communicate with the main chamber.

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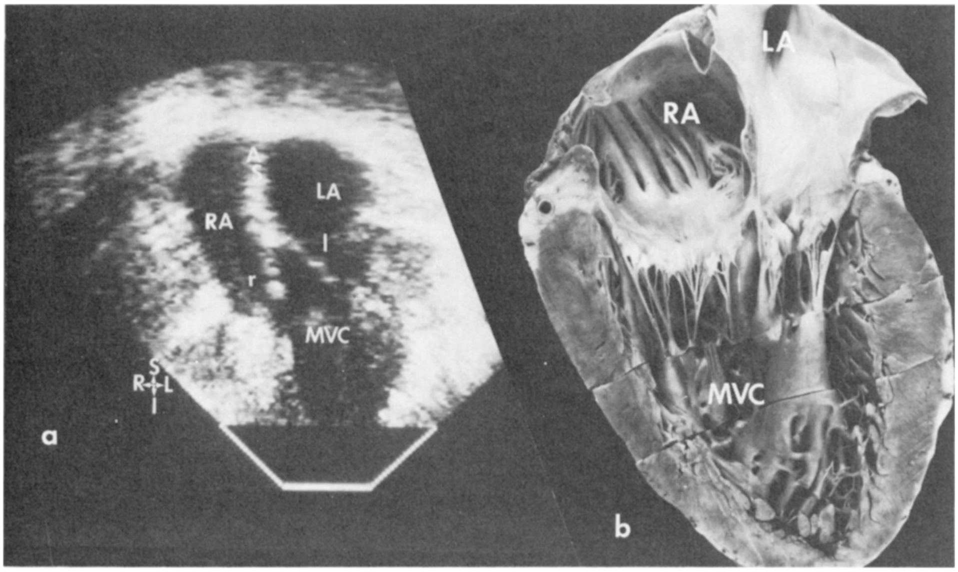

Figure 1. Double inlet univentricular AV connection: **a**, Two-dimensional echocardiographic apical view shows two AV valves connecting the right atrium (RA) and left atrium (LA) to the main ventricular chamber (MVC). The medial leaflets of the right (r) and left (l) AV valves originate at the internal crux. **b**, Heart specimen of a double inlet ventricle from another patient cut in the corresponding tomographic plane. I = inferior; L = left; R = right; S = superior.

Ventriculoarterial connection. Four possibilities exist with univentricular AV connection: 1) the pulmonary artery arises from the main ventricular chamber and the aorta arises from the outlet chamber; 2) the aorta arises from the main ventricular chamber and the pulmonary artery arises from the outlet chamber; 3) one great artery arises from either the main chamber or the outlet chamber, and the other semilunar valve is atretic; and 4) both great arteries arise from either the main ventricular chamber or outlet chamber.

Methods

Patients. Between July 1977 and February 1981, there were 183 patients with univentricular atrioventricular (AV) connection who had angiographic and two-dimensional echocardiographic examination at the Mayo Clinic. Their ages ranged from 1 day to 43 years (mean 9.5 years). Angiographic diagnoses were obtained from roll film angiograms in the anteroposterior and lateral projections.

Echocardiography. Two-dimensional echocardiographic examination was performed with either the Varian 3000 or ATL Mark V instrument with the patient in the supine or left lateral position. A complete examination was performed in each patient utilizing previously published techniques

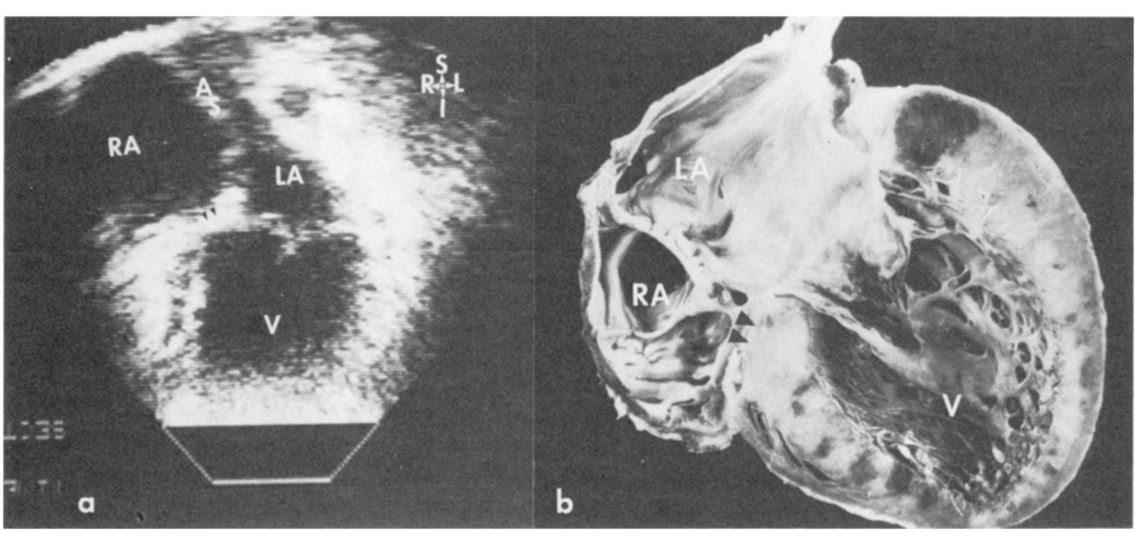

Figure 2. Single inlet univentricular AV connection. **a**, Two-dimensional echocardiographic apical view shows absent right connection (**arrowheads**) and a single inlet via a remaining left AV valve to the main ventricular chamber (V). The commitment of the atretic AV valve remnant cannot be reliably determined by those imaging techniques. **b**, Heart specimen of a single inlet ventricle (tricuspid atresia) cut in a corresponding tomographic plane. AS = atrial septum; I = inferior; L = left; LA = left atrium; R = right; RA = right atrium; S = superior.

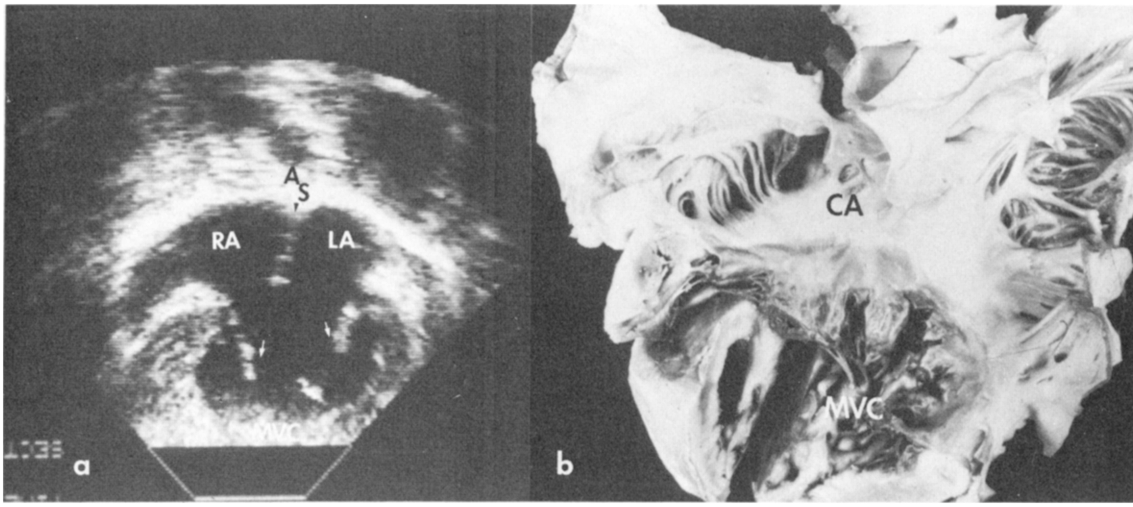

Figure 3. Common inlet univentricular AV connection. **a**, Two-dimensional echocardiographic apical view shows both right (RA) and left (LA) atria connecting to one main ventricular chamber. There is no atrial septum primum or internal crux. **b**, Heart specimen with right atrial isomerism, a common atrium (CA) and a common inlet. Abbreviations and orientation as in Figures 1 and 2.

(5–7). Apical and subcostal four chamber views provided information concerning the number of AV valves and their commitment (8,9). Parasternal long- and short-axis views were also used to obtain information concerning the presence or absence of an accessory chamber, the origin of the great arteries, the ventriculoarterial connection and the morphology of the AV valve or valves. Apical long-axis views provided visualization of the outlet foramen when an anterior outlet chamber was present. The presence or absence of pulmonary obstruction also could be assessed in this view. Suprasternal views gave information on the side of the aortic arch and the position of the great arteries. Discrete outlet foramen obstruction was predicted to be present when the size of the foramen in diastole was less than two-thirds the diameter of the corresponding semilunar valve anulus.

This approach to the patient with univentricular AV connection was designed to obtain information concerning seven important anatomic categories including: 1) mode of AV connection, 2) position and type of accessory chamber, 3) outlet foramen obstruction, 4) AV valve abnormalities, 5) great artery position and connections, 6) pulmonary obstruction, and 7) ventricular morphology.

Cardiac catheterization. Angiographic and hemodynamic data were available in all 183 patients. Identification of the accessory chamber by angiography was based on standard angiographic criteria (10–12). Obstruction of the outlet foramen was defined as a discrete peak systolic pressure gradient of greater than 30 mm Hg between the main ventricular chamber and a well developed outlet chamber. Hemodynamically significant obstruction to pulmonary flow was defined as a main chamber to pulmonary artery gradient of greater than 30 mm Hg. Great artery position was described as the position of the aorta with respect to the pulmonary valve or main pulmonary trunk.

Surgery/autopsy. Surgical confirmation was available in 120 patients (66%). The surgical approach in the vast majority of these patients was a Fontan-type operation and, therefore, surgical confirmation of the ventricular anatomy was limited to that which could be easily visualized through the AV valve or valves. Autopsy confirmation of the anatomic diagnosis was available in seven patients.

Results

Diagnosis (Table 1, Fig. 1 to 3). Using angiography as a diagnostic reference standard, the diagnosis of univentricular atrioventricular (AV) connection was made correctly by two-dimensional echocardiography in all 183 patients. Three subgroups of univentricular AV connection were: 1) double inlet (100 patients), 2) single inlet (67 patients), and 3) common inlet (16 patients). There was no significant difference between the type of connection found by angiography or at operation and that predicted noninvasively. Two-dimensional echocardiography and angiography were complementary. Angiography was superior for appreciation of extracardiac vascular anatomy. However, two-dimensional echocardiography was superior for visualization of AV valve number, visualization of chordal support apparatus and differentiation of large muscle bands from septa. Fourteen patients with dextrocardia did not present a diagnostic problem for two-dimensional echocardiography.

Accessory chambers (Table 2). An *outlet chamber* was present by angiography in 135 patients and was correctly identified by two-dimensional echocardiography in 125

Table 1. Anatomic Data: Univentricular Atrioventricular Connection (183 patients)

	Double Inlet		Single Inlet		Common Inlet		Total	
No. of patients	100		67 (5L)		16		183	
Male/female	66/34		40/27		10/6		116/67	
Age (yr):mean (range)	11.9 (1 mo to 43 yr)		7.0 (2 wk to 33 yr)		4.7 (1 day to 25 yr)		9.5 (1 day to 43 yr)	
Situs								
Solitus/inversus/ambiguous	100/0/0		67/0/0		11/0/5		178/0/5	
Dextrocardia	7		3		4		14	
Accessory chambers								
Outlet chamber	72		61 (2L)		2		135	
Subaortic		59		13 (1L)		1		73
Subpulmonary		13		47		1		61
Double outlet		0		1 (1L)		0		1
Outlet foramen obstruction	9		16		0		25	
Subaortic		3		2		0		5
Subpulmonary		6		14		0		20
Pouch	6		2 (2L)		4		12	
None	22		4 (1L)		10		36	
Great artery position								
Aorta position (with respect to pulmonary trunk) RP/RA/LA/LP	17/27/56/0		48/10/9/0 (1/2/2/0L)		2/11/2/1		67/48/67/1	
Great artery connections								
Aorta from OC	56		10 (1L)		1		67	
Aorta from MC	10		30		1		41	
Single outlet aorta (all pulmonary atresia)	8		20		2		30	
From OC		2		3		0		
From MC		6		17 (1L)		2		
Double outlet OC	0		1 (1L)		0		1	
Double outlet MC	26		6 (3L)		12		44	
Obstruction to pulmonary flow								
Valvular-subvalvular	69		30 (1L)		7		106	
Atresia	8		20		2		30	
Pulmonary trunk band	16		4 (2L)		2		22	
None	7		13 (2L)		5		25	
Main chamber morphology								
LV/RV/?	75/14/11		61/2/4		0/6/10		136/22/25	

AV = atrioventricular; L = absent left connection; LA = left anterior; LP = left posterior; LV = left ventricular; MC = main ventricular chamber; OC = outlet chamber; R = right; RA = right anterior; RP = right posterior; RV = right ventricular; ? = indeterminate.

(sensitivity 93%)(Fig. 4). All subaortic chambers were visualized. Outlet chambers difficult to identify by echocardiography were angiographically small, slit-like chambers associated with severe pulmonary valve stenosis or pulmonary atresia. Two patients with muscular subpulmonary obstruction were predicted to have an outlet chamber when there was none identified by angiography (two false positive diagnoses, specificity 96%). A *pouch* was present angiographically in 12 patients and was detected by two-dimensional echocardiography in 11 patients (sensitivity 92%) (Fig. 5).

Table 2. Detection of Accessory Chambers and Obstruction to Pulmonary Flow: Two-Dimensional Echocardiography Versus Angiography

	True Positive	True Negative	False Positive	False Negative	Sensitivity	Specificity	Predictive Accuracy
Outlet chamber detection	125	46	2	10	93%	96%	93%
Outlet foramen obstruction detection	19	110	0	6	76%	100%	96%
Pouch detection	11	171	0	1	92%	100%	99%
Obstruction to pulmonary flow detection	158*	24	1	0	100%	96%	99%

*Includes 30 patients with pulmonary atresia and 22 patients with previous pulmonary trunk band.

In all, the pouch was posterior in location and appeared to communicate with the main chamber. There were no cases where echocardiography identified a pouch that was not visualized angiographically.

Outlet foramen obstruction (Table 2, Fig. 4). Discrete outlet foramen obstruction was subaortic in 5 patients and subpulmonary in 20 patients. Two-dimensional echocardiography recognized subaortic outlet foramen obstruction in all 5 patients and discrete subpulmonary obstruction in 14 patients (overall sensitivity 76%). There were no false positive diagnoses. Outlet foramen obstruction resulted from abnormal chordal insertions in two patients. Two patients with subaortic obstruction had previous pulmonary trunk banding.

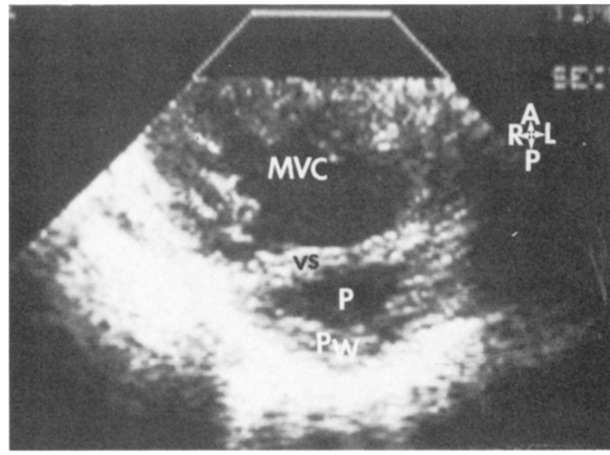

Figure 5. Parasternal short-axis view showing a posterior pouch (P) in a patient with a double inlet AV connection with "right ventricular morphology" of the main ventricular chamber (MVC). L = left; R = right; other abbreviations as in Figure 4.

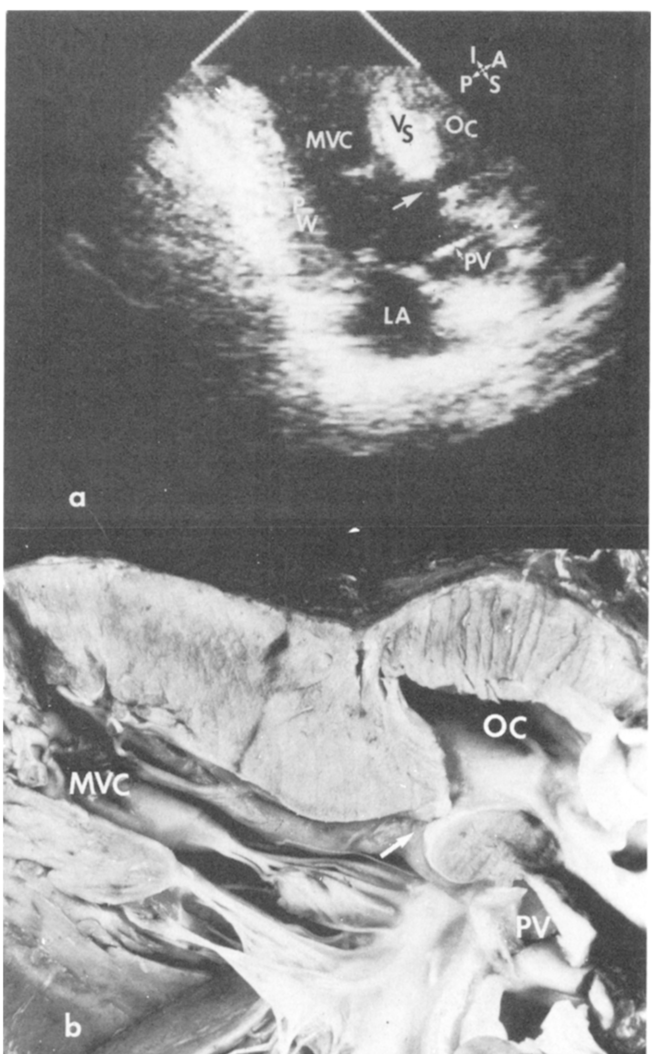

Figure 4. a, Two-dimensional echocardiogram (apical long-axis view) of a patient with a double inlet ventricle and anterior subaortic outlet chamber (OC). The outlet foramen (**arrow**) connecting the main ventricular chamber (MVC) and outflow chamber is stenotic. Obstruction of the foramen was confirmed at catheterization. **b**, A pathologic specimen sectioned in the corresponding plane demonstrates an anterior outlet chamber (OC) and stenotic outlet foramen. A = anterior; I = inferior; LA = left atrium; MVC = main ventricular chamber; P = posterior; PV = pulmonary valve; PW = posterior wall; S = superior; VS = ventricular septal remnant.

AV valve abnormalities (Table 3, Fig. 6). Atrioventricular valve abnormalities were present in 49 patients (27%). In 19 patients with univentricular AV connection and two AV valves, the chordae inserted into the outlet chamber (right in 12 patients; left in 7 patients) (Fig. 7). In five of these patients, the right AV valve chordae inserted into the left anterior subaortic outlet chamber. In four patients, AV valve chordae inserted in the contralateral portion of the main ventricular chamber (interdigitating AV valve chordae).

Significant AV valve abnormalities were uncommon in patients with either a single or common inlet. Prolapse of the left AV valve was present in seven patients with absent right AV connection (tricuspid atresia). The anterior leaflet of this AV valve was cleft in two additional patients, causing moderate left AV valve insufficiency in one.

Great artery position. The great artery position was correctly predicted in all 183 patients. In 35 patients in whom pulmonary cusp tissue was not visualized, the aortic position and the proximal pulmonary arteries were imaged.

Obstruction to pulmonary flow (Table 3). Valvular, subvalvular or supravalvular pulmonary stenosis or pulmonary atresia was present in 158 (86%) of the 183 patients. Of the 25 patients without obstruction to the pulmonary flow, there was 1 patient predicted to have significant pulmonary stenosis who had reached adulthood with pulmonary vascular disease and no significant obstruction (one false positive diagnosis). In the 22 patients with a previous pulmonary trunk band, the site of banding was visualized by two-dimensional echocardiography in all. Of the 30 patients

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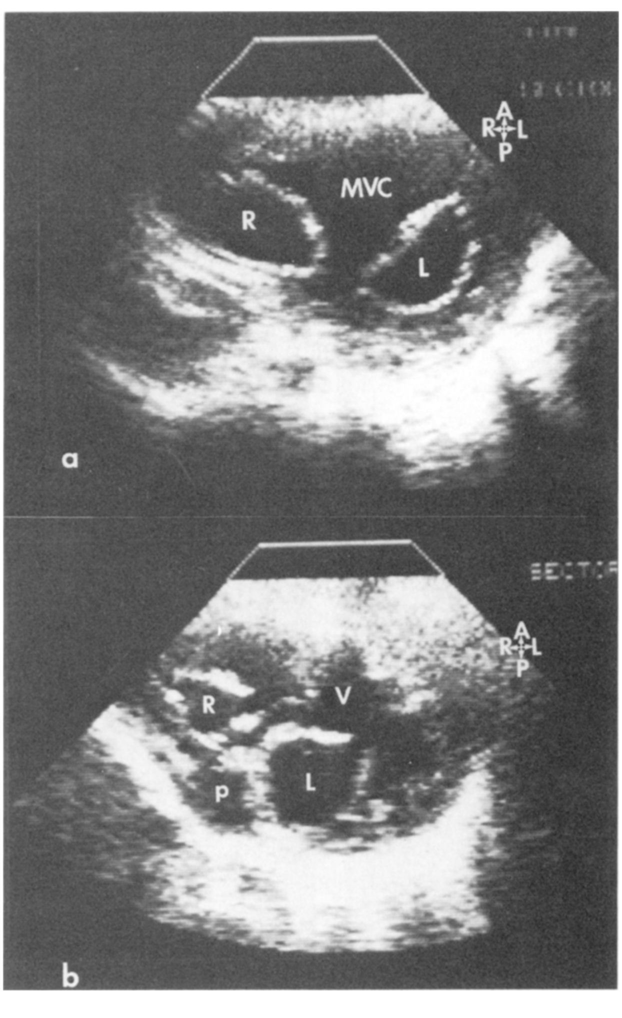
Table 3. Two-Dimensional Echocardiographic Assessment of Atrioventricular Valve Abnormalities

	Chordae Into Outlet Chamber	Chordae Into Pouch	AV Valve Hypoplasia	Other	Total
Two AV valves (100 patients)	19	3	5	8*	35 (35%)
Left	7	3	4		
Right	12	0	1		
One AV valve (83 patients)	1	1	0	12†	14 (17%)
Total	20	4	5	10	49 (27%)

*Interdigitating atrioventricular (AV) valve chordae in four patients, bilateral tricommissural valves in two, single papillary muscle in one and thickened left atrioventricular valve in one; †prolapse in seven thickened leaflets in three patients and cleft anterior valve leaflet in two.

Figure 6. a, Parasternal short-axis view of a patient with a double inlet AV connection illustrating bileaflet right (R) and left (L) AV valves both entering the main ventricular chamber (MVC). There was an anteriorly located outlet chamber indirectly suggesting a main ventricular chamber of left ventricular morphology. **b**, Parasternal short-axis view of another patient with a double inlet AV connection with relative hypoplasia of the right AV valve. A posterior pouch (p) suggests a main ventricular chamber (V) of right ventricular morphology. Abbreviations as in Figure 4.



with pulmonary atresia, no pulmonary valve tissue could be identified by either angiography or two-dimensional echocardiography. Among the remaining 108 patients with valvular or subvalvular pulmonary stenosis at catheterization, the pulmonary valve in 6 was not visualized by two-dimensional echocardiography; however, the presence of pulmonary obstruction was correctly predicted. Therefore, obstruction to pulmonary flow was correctly predicted by two-dimensional echocardiography in 157 (99%) of 158 patients. No cases were encountered of valvular aortic stenosis or aortic hypoplasia or atresia.

Ventricular morphology. The usual two-dimensional echocardiographic criteria for the recognition of ventricular morphology when the ventricular septum is present (6) were not applicable in all cases of patients with univentricular AV connection. Ventricular morphology could be inferred by the presence or absence of an accessory chamber as described in the pathology literature. Therefore, the only two-dimensional echocardiographic criteria *consistently* available for determination of ventricular morphology of the main ventricular chamber were shape and trabecular pattern. Semilunar-AV valve continuity was noted, but was not considered to be a strong morphologic signate in this entity. Of the 100 patients with a double inlet, the papillary muscle and trabecular pattern of the main chamber by echocardiography resembled that of a left ventricle (two distinct papillary muscles and smooth walled pattern) in 12 (16%) of 75 and a morphologic right ventricle in 8 (57%) of 14. Of the 67 patients with a single inlet, main chamber ventricular morphology was correctly predicted to be left in 58 (95%) of 61, right in 2 of 2 and indeterminate in 4 of 4. In this group, the presence and type of accessory chamber and the presumed main chamber morphology on this basis alone corresponded exactly with the ventricular morphology determinations by both echocardiography and angiography. Of the 16 patients with a common inlet, ventricular morphology was that of a left ventricle in none, a right ventricle in 2 of 6 and indeterminate in 10 of 10. Two-dimensional

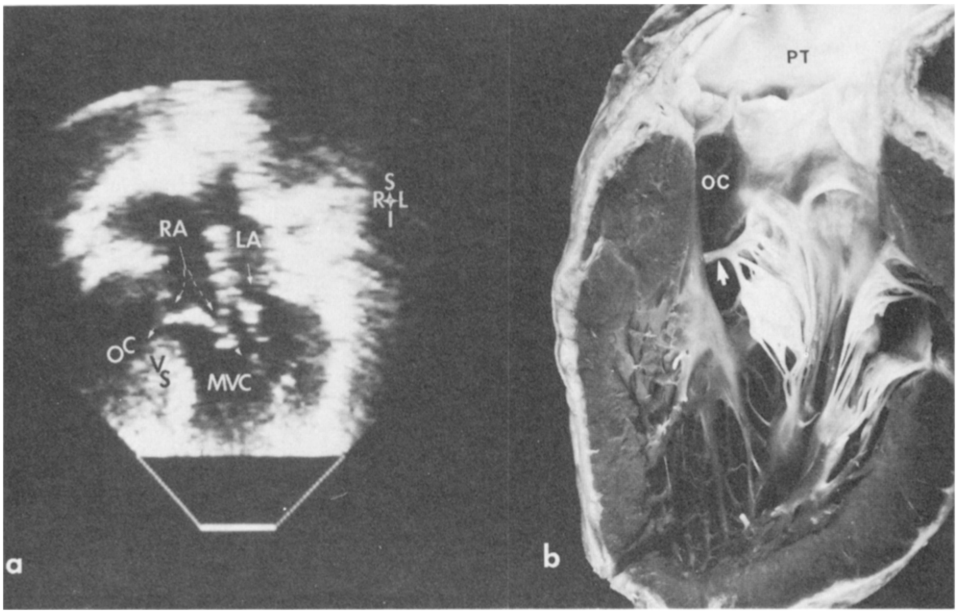


Figure 7. Double inlet ventricle with abnormal insertions of portions of the right AV valve chordae into the outlet chamber (OC). Two-dimensional echocardiogram (**a**) and corresponding specimen cut in a similar tomographic plane (**b**). Note the abnormal right AV valve and chordae (**white arrow**). PT = pulmonary trunk; other abbreviations as in Figure 4.

echocardiography was poor in assessing the trabecular pattern of the accessory chamber.

Discussion

Univentricular atrioventricular connections. Angiography has been successfully used to diagnose univentricular atrioventricular (AV) connection (10–12). Two-dimensional echocardiography has provided comparable information noninvasively and has been a superior means of examining the region of the AV junction ("internal crux") (13). M-mode echocardiography has previously been used to assess the number of AV valves and suggest associated abnormalities (14–16), but it lacked the spatial orientation necessary for confident diagnosis. The two-dimensional echocardiographic recognition of subgroups of univentricular AV connection was possible noninvasively and was based on the mode of the AV connection. In the category of univentricular AV connection were three broad types of ventricular and AV valve anatomy: 1) double inlet, 2) single inlet, and 3) common inlet. These entities are presently managed clinically in a similar fashion. Previous echocardiographic and angiographic studies (13) attempted to correlate images with what would be expected based on the results of pathologic studies. We have emphasized a descriptive anatomic approach and purposely avoided terminology based on the ventricular morphology (2).

In patients with univentricular AV connection and two discrete AV valves (double inlet), the valve anular sizes and relations and the chordal insertion patterns could be readily visualized. Two AV valves were visualized in short- and long-axis views, and when one AV valve was hypoplastic or stenotic, M-mode echocardiography was a complementary modality for assessing valve opening. Patients with a common inlet were not included in this group.

There is considerable controversy surrounding the nomenclature of univentricular AV connection via one AV valve (17–18). We describe the mode of AV connection, separating those with absent AV connection (single inlet) and intact inferior atrial septum from those with a common AV valve (common inlet) and no inferior atrial septum. Debate has centered around whether patients with a single inlet have one or two ventricles. From the echocardiographic and surgical point of view, these patients have one main ventricular chamber and one usable AV valve. It is notable that patients with right AV valve atresia have a main chamber that frequently has echocardiographic morphologic features consistent with a left ventricle (two distinct papillary muscles and AV-semilunar valve continuity). Often, the converse is true of left AV valve atresia (that is, a posterior pouch and main ventricular chamber with increased trabeculation). Patients with an imperforate AV valve were not encountered in this series, but belong in the subgroup of single inlet because a patent AV connection exists with only one ventricular chamber via one valve.

In univentricular AV connection via a common AV valve (common inlet), two-dimensional echocardiography was as capable as angiography in describing the ventricular and AV valve anatomy. There is a spectrum between 1) common inlet, and 2) common AV valve orifice with a biventricular heart with dominance of one ventricle. Occasionally, it can be difficult to confidently separate these two types of common AV valve orifice by two-dimensional echocardiography alone, as pointed out by Smallhorn et al.(19)

Associated anomalies. *Identification of outlet chambers* by two-dimensional echocardiography was highly specific and sensitive. All subaortic outlet chambers were detected and subaortic obstruction was accurately predicted. The development of subaortic obstruction at the outlet foramen after a pulmonary trunk banding procedure is becoming a well recognized entity (20), and early detection of this complication by two-dimensional echocardiography is now possible. Detection of very small anterior chambers by two-dimensional echocardiography required attention to the anterior mediastinum, either from the parasternal or subcostal approach utilizing higher frequency transducers. When pulmonary valve atresia was present in older patients, the accessory chamber was not always detected.

Pouches were reliably detected by two-dimensional echocardiography. Some ventricular pouches may not communicate with the main ventricular chamber and may be recognized only at autopsy (21). The importance of two-dimensional echocardiographic recognition of outlet chambers and pouches is not entirely academic. Inferences can be made about the ventricular morphology and conduction tissues based on the type and position of the rudimentary chamber. For example, a patient with univentricular AV connection by way of two AV valves and a posterior pouch could be presumed to have a main chamber of right ventricular morphology and conduction tissue located posteriorly. These considerations may have importance in the postoperative hemodynamics after a modified Fontan operation and require further investigation (22).

We found two-dimensional echocardiography to be superior to angiography in defining AV valve leaflet and chordal abnormalities. The incidence and spectrum of abnormalities found in this study were similar to those in previous pathologic studies (23). These abnormalities often have importance with regard to the surgical management and should be excluded in all patients before operation. For example, the septation operation (24,25) is contraindicated in patients who have AV valve chordae connecting to the contralateral outlet chamber. Surgically significant AV valve abnormalities were uncommon in patients with single inlet.

Great artery position was accurately diagnosed by two-dimensional echocardiography. Great artery connections had a wide spectrum and were correctly predicted in each patient.

Although pulmonary obstruction was detected with high specificity and sensitivity, two-dimensional echocardiography was limited in predicting the severity of pulmonary obstruction and differentiating between severe pulmonary valve hypoplasia and pulmonary valve atresia. In the future, Doppler echocardiography may be helpful in this regard.

There was great variety in the two-dimensional echocardiographic appearance of the ventricular morphology of the main ventricular chamber in patients with univentricular AV connection. In those with two AV valves, assignment of ventricular morphology was speculative in the majority and could be inferred only in those patients with an identifiable accessory chamber. In contrast, patients with a single inlet frequently had a main ventricular chamber that appeared similar to a left ventricle. Sahn et al. (26) described a posterior muscle bundle in patients with a double inlet as a criterion for left ventricular morphology. However, a coarse trabecular pattern may coexist with an anterior outlet chamber, leading us to doubt the accuracy of any single echocardiographic criterion.

Diagnostic approach. The strength of two-dimensional echocardiography was the ability to assess the AV junction. In patients with a double inlet, the valve size, valve relation and valve support apparatus could be readily visualized. Using a step by step segmental approach by two-dimensional echocardiography and examining all seven categories of anatomic information, the appropriate surgical treatment could be individualized. Angiography and hemodynamic information complemented noninvasive data in the assessment of the atrial septum, pulmonary vascular bed and aorta.

Limitations. Univentricular AV connection via two AV valves that align and connect predominantly to one ventricular chamber in the presence of a second ventricular chamber of *comparable* size was not encountered in this study. Such hearts, with straddling or overriding AV valves, are a transition group (27). Patients were included when the other chamber was considerably smaller than the main chamber and lacked an inlet portion. In the unlikely possibility of ventricular chambers of equal size and greater than 50% overriding of one AV valve anulus without straddling, is the AV connection univentricular? By definition this may be true. However, a label of univentricular AV connection may not communicate the appropriate surgical treatment and, therefore, the size of the chambers must be specified.

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