Double-Inlet Single Left Ventricle: Echocardiographic Anatomy With Emphasis on the Morphology of the Atrioventricular Valves and Ventricular Septal Defect

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The echocardiographic anatomy of double-inlet single left ventricle was studied in 57 patients, aged 1 day to 27 years (mean 6 years); the variables examined included morphology, size and function of the atrioventricular (AV) valves and ventricular septal defect and their relation to pulmonary stenosis, aortic stenosis and aortic arch obstruction. The visceroatrial situs was solitus and the heart was in the left side of the chest in all 57 patients. A d-loop ventricle was present in 21 patients and an l-loop ventricle in 36. The great arteries were normally related (Holmes heart) in 8 patients and transposed in 49.

In all hearts, the right AV valve was anterior to the left AV valve. In 53 patients, the tricuspid valve (right valve in d-loop and left valve in l-loop) was closer to and had attachments on the septum. The tricuspid valve straddled the outflow chamber in eight patients. No significant difference was noted in the mean AV valve diameter when comparing mitral and tricuspid valves within the same group or between the groups with a d- or l-loop ventricle. The right AV valve diameter had a significant direct correlation with the aortic valve diameter and the size of the ventricular septal defect regardless of ventricular loop. Both AV valves were functionally normal in 34 patients. Among patients with AV valve dysfunction, the tricuspid valve tended to be stenotic in patients with an l-loop ventricle and regurgitant in patients with a d-loop ventricle. Mitral valve dysfunction was uncommon.

The ventricular septal defect (46 patients) was separated from the semilunar valves in 24 patients (muscular defect) and adjacent to the anterior semilunar valve as a result of hypoplasia or malalignment, or both, of the infundibular septum (subaortic defect) in 19 patients. Multiple defects were present in three patients. The defect was unrestrictive in 26 patients, restrictive in 23 and could not be evaluated in 8. Pulmonary artery banding had been performed in 8 of the 26 patients with an unrestrictive defect and in 10 of the 23 patients with a restrictive defect. Only 4 of 19 subaortic defects compared with 16 of 24 muscular defects were restrictive. The size of the defect was significantly correlated with the measured pressure gradient. Among patients with transposition, only 2 of 13 with pulmonary stenosis had a restrictive ventricular septal defect compared with 15 of 30 without pulmonary stenosis. In patients with transposition, the defect size was significantly smaller when coarctation was present.

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Hearts with a functionally single ventricle are a heterogeneous group and controversy still exists regarding even the nomenclature (1,2). Among these hearts, however, there is a relatively homogeneous subset (3–5) in which the single ventricle is of left ventricular morphology and is connected to both atria by two separate atrioventricular (AV) valves (double-inlet left ventricle). Most often, the great arteries are transposed (the pulmonary artery aligned with the single left ventricle and the aorta arising from an anterior and superior outlet chamber). Occasionally, normally related great arteries are present (Holmes heart).

Previous echocardiographic studies (6–16) have investigated the role of M-mode and two-dimensional echocardiography in diagnosing double-inlet left ventricle and defining the AV connections, the morphology of the main chamber, the position of the accessory chamber and the ventriculo-arterial (VA) alignments and connections. These studies, however, grouped patients with different types of single ventricle and did not address the morphology and function of the AV valves or the nature of the communication between the left ventricle and the outlet chamber.

The purpose of this report is to describe the two-dimensional echocardiographic anatomy of the AV valves and ventricular septal defect in a group of patients with double-inlet single left ventricle. In addition, the relation between the type and size of the ventricular septal defect and aortic outlet and arch obstruction is examined.

Methods

Study patients. All patients with the diagnosis of double-inlet single left ventricle who had undergone two-dimensional and Doppler echocardiography at The Children's Hospital since 1982 were selected. The diagnostic criteria included 1) two patent AV valves; 2) a large ventricular chamber of left ventricular morphology (17,18) that received all or nearly all of the attachments of both AV valves (hearts with minor straddling of one valve into an outlet chamber were included); and 3) a relatively anterior outlet chamber that communicated with the left ventricle by means of one or more ventricular septal defects and that gave rise to at least one great artery.

Echocardiography. All studies were performed with an ATL Mark 600, a Diasonic Cardiovue 100 or a Hewlett-Packard 77020 cardiac imager equipped with a 3, 3.5 or 5 MHz transducer. In each case multiple views of the heart were obtained from the subxiphoid, apical, parasternal and suprasternal positions. In most patients, a complete pulsed Doppler ultrasound examination was also performed in the same echocardiographic views. Sedation with chloral hydrate was used when necessary.

Data analysis. The echocardiographic studies were reviewed with particular attention to the morphology and attachments of the AV valves, the AV valve function on the Doppler examination, the size and location of the ventricular septal defect and the presence of associated lesions, especially pulmonary stenosis, aortic stenosis and coarctation. The following measurements were made from stop-frame images with use of electronic calipers: 1) diameter of the AV valve anuli in early to mid-diastole (apical four-chamber view); 2) diameter of the ventricular septal defect (bulboventricular foramen) in a long-axis (subxiphoid or parasternal long-axis) and short-axis (subxiphoid or parasternal short-axis) view at end-diastole; and 3) systolic diameter of the aortic and pulmonary valve anuli.

The cross-sectional area of the ventricular septal defect was calculated by using the formula for a regular ellipse, with the measured dimensions as the major and minor axes. The peak instantaneous gradient across the ventricular septal defect was estimated from the pulsed or continuous wave Doppler recording obtained in the defect with use of the modified Bernoulli equation: Peak pressure difference = 4 (Peak velocity)^2. The ventricular septal defect was considered restrictive if a pressure gradient of ≥ 15 mm Hg was measured at cardiac catheterization or by Doppler echocardiography or if the major diameter of the defect was <50% of the diameter of the descending aorta at the diaphragm.

Stenosis of an AV valve was diagnosed if the valve anulus diameter was <50% of the diameter of the contralateral AV valve anulus or if a mean gradient ≥ 5 mm Hg was detected by Doppler examination. Atrioventricular valve regurgitation was diagnosed if a pansystolic regurgitant jet could be detected at least half the diameter of the atrium behind the plane of the valve ring with use of either pulsed Doppler recording or Doppler color flow mapping.

The clinical records of the patients were reviewed for the results of cardiac catheterization and the operative reports. In eight patients who died the heart specimens were examined and compared with the echocardiographic findings.

Statistical analysis. Analysis of variance was used to test the significance of differences between group mean values. Regression analysis was used to evaluate relations between continuous variables. A p value ≤ 0.05 was considered significant.

Definitions. *Ventricular septal defect.* The orifice through which the left ventricle communicates with the infundibular outlet chamber has been called a bulboventricular foramen or a ventricular septal defect. Because the wall of the left ventricle that is the ventricular septum in a normal heart is also present in these hearts and because the communication between the left ventricle and the outlet chamber occurs in this wall, ventricular septal defect is an accurate term. The name bulboventricular foramen implies a single communication of uniform location and morphology between the left ventricle and the outflow chamber. Although the residuum of the embryonic bulboventricular foramen appears to be one type of communication between the left ventricle and outlet chamber, our observations in this study and those of others indicate that this is not the only type of communication that can occur. Consequently, the more general term ventricular septal defect is used here because it is accurate and more broadly applicable.

Atrioventricular valves. The identity of the AV valves is defined primarily by the papillary muscle attachments rather than by the number of leaflets (19,20). The tricuspid valve has attachments on the papillary muscle or muscles closest to the septum or on the inferior rim of the ventricular septal defect, or both. In contrast, the mitral valve attaches to the papillary muscles on the left ventricular free wall more distant from the septum. Identification of the AV valves on the basis of the attachments accurately indicated the ventricular loop. A right-sided tricuspid valve was always associated with a d-ventricular loop, whereas a left-sided tricuspid valve occurred only in hearts with an l-ventricular loop.

Results

Fifty-seven patients, aged 1 day to 27 years (mean 6 years), with double-inlet single left ventricle were identified. The data are summarized in Table 1.

Segmental analysis. *Visceroatrial situs,* determined as described previously (21), was solitus in all 57 patients. Systemic and pulmonary venous connections were also normal in all patients.

The ventricular loop was determined by the position of the outlet chamber (22) and the ventricular septum. In a d-loop left ventricle, the smooth septal wall is anterior and rightward, adjacent to the outlet chamber (Fig. 1A). In an l-loop left ventricle, the smooth septal surface is leftward

Table 1. Comparison of Demographic Data, Structural Dimensions and Valve Function Between Patients With an l-Loop or d-Loop Ventricle With and Without Transposition of the Great Arteries (n = 57)

	l-Loop	d-Loop With NRGA	d-Loop With TGA	p Value
Age at study (mo)	76 ± 19	139 ± 58	105 ± 73	NS
Body surface area (m²)	0.65 ± 0.09	0.84 ± 0.21	0.49 ± 0.1	NS
QP/QS	2.10 ± 0.70	4.45 ± 1.35	3.59 ± 0.63	0.0264?
Indexed left AVVD*	1.94 ± 0.39	2.09 ± 0.14	2.26 ± 0.10	NS
Indexed right AVVD*	2.26 ± 0.07	1.97 ± 0.25	2.17 ± 0.10	NS
Indexed AVD*	2.10 ± 3.00	2.10 ± 0.13	1.82 ± 0.08	NS
Indexed VSD area*	10.68 ± 3.84	4.31 ± 2.25	11.32 ± 2.47	0.0018
VSD gradient (mm Hg)	4 ± 1	53 ± 47	8 ± 5	0.0001§
Left AVV regurgitation	2.9% (TV)	0.0% (MV)	0.0% (MV)	NS
Left AVV stenosis	20.6% (TV)	0.0% (MV)	14.3% (MV)	NS
Right AVV regurgitation	8.8% (MV)	27.3% (TV)	16.7% (TV)	NS
Right AVV stenosis	0.0% (MV)	0.0% (TV)	16.7% (TV)	0.0204?
Systemic to pulmonary shunt operation	32.3%	18.8%	25.0%	NS
Coarctation	34.9%	15.4%	12.5%	NS

*Cm-square root of body surface area (BSA). ?cm² body surface area. —comparing l-loop and d-loop transposition; §comparing either transposition group with the normally related great artery group. Unless otherwise indicated, values are mean values ± SD. AVD = aortic valve dimension; AVV = atrioventricular valve; AVVD = AVV dimension; MV = mitral valve; NRGA = normally related great arteries; QP/QS = ratio of pulmonary to systemic flow; TGA = transposition of the great arteries; TV = tricuspid valve; VSD = ventricular septal defect.

and somewhat posterior, adjacent to the outlet chamber (Fig. 1B). In addition to the smooth surface, the ventricular septal defect is a marker for the ..tal wall. A d-loop was present in 21 patients and an l-loop was seen in 36 patients.

The VA alignment was determined by simultaneous imaging of the ventricular and arterial segments as previously described (23). Transposition of the great arteries (aorta aligned with the outlet chamber and pulmonary artery aligned with the single left ventricle) was present in 49 patients (13 with a d-loop ventricle and 36 with an l-loop ventricle). The great arteries were normally related in eight patients, all with a d-loop ventricle (Holmes heart).

The segmental sets (22) were {S,L,L} (35 patients). {S,L,A} (1 patient), {S,D,D} (10 patients), {S,D,A} (2 patients), {S,D,L} (1 patient) and {S,D,S} (8 patients). The first member of the set indicates the visceroatrial situs (S = solitus, I = inversus), the second member the ventricular loop (d- or l-loop) and the third member the position of the aorta (S = solitus normally related great arteries, D, A, L = malposition of the great arteries with the aorta to the right, anterior or left, respectively).

Atrioventricular valve morphology. In all hearts, regardless of ventricular loop, the right AV valve was anterior relative to the left AV valve (Fig. 1). Because the viscero-atrial situs was solitus in all patients, this probably reflects the usual position of the atria in situs solitus, where the right atrium is anterior to the left atrium. In 53 of the 57 patients, the tricuspid valve (right AV valve in d-loop and left AV valve in l-loop) was closer to and had attachments on the inferior and the posterior border of the ventricular septal defect, whereas the mitral valve was more distant from and

had no attachments on the septum (Fig. 1, A and B: 2, A and B and 3, A and B). In the other four patients, both AV valves were equally distant from the septum (Fig. 1C).

Mild straddling of an AV valve into the outlet chamber was noted in eight patients ({S,D,D} in four, {S,L,L} in three and {S,D,S} in one). Straddling was best appreciated in a short-axis subxiphoid or parasternal view (Fig. 4). A straddling valve was consistently associated with a muscular ventricular septal defect that entered the inferior or diaphragmatic aspect of the outlet chamber. The chordal attachments of the valve inserted on the lower rim of the septal defect as well as within the outlet chamber. In each case, the straddling valve was the tricuspid valve (that is, the right-sided AV valve in patients with a d-loop ventricle and the left-sided AV valve in patients with an l-loop ventricle).

Because it was difficult to distinguish deep scallops in the AV valves from commissures, it was not possible to determine consistently the number of leaflets in the AV valves.

Atrioventricular valve size and function. The indexed (diameter/square root of body surface area) diameters of the right and left AV valves are shown in Table 1. Patients with an l-loop ventricle and transposition, with a d-loop ventricle and transposition and with a d-loop ventricle and normally related great arteries are shown separately. The unindexed valve diameters were significantly correlated with age (r = 0.56, p < 0.001), body surface area (r = 0.83, p < 0.0001) and square root of body surface area (r = 0.85, p < 0.0001). Indexed diameters were no longer significantly correlated with age or body size. No significant differences were noted for the mean AV valve size when left and right AV valves were compared within the same group or other groups.
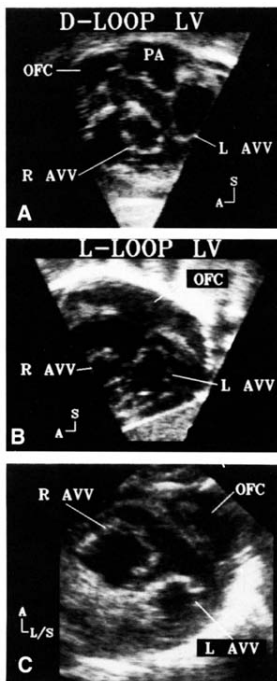
Figure 1. Subxiphoid (A and B) and parasternal short-axis (parasagitally) (C) views in patients with double-inlet single left ventricle. A, d-loop ventricle (LV). The outlet chamber (OFC) is anterior and superior and the atrioventricular valves (AVV) are posterior and inferior with the anterior (right [R]) AVV closer to the septum. B, l-loop ventricle. In contrast to A, the outlet chamber is posterior and superior with the AVV anterior and inferior and the posterior (left) valve closer to the septum. C, l-loop ventricle with the AVV equidistant from the septum. A = anterior; L = left; PA = main pulmonary artery; R = right; S = superior.

Figure 4. The heart from a 2.5-month old boy with double-inlet ventricle and transposition of the great arteries (S.L.L). A. The hypertrophied left ventricle (LV) is right-sided. A slit-like muscular ventricular septal defect (VSD) is present. The left-sided tricuspid valve (TV) is adjacent to the septum and attaches to a single, hypertrophied, paraseptal papillary muscle. The right-sided mitral valve (MV) attaches only to the free wall. The pulmonary valve (PV) is normal and the pulmonary outflow tract unobstructed. B. The left-sided infundibular outlet chamber is underdeveloped. The aortic valve was stenotic with only one well developed commissure; the aortic arch and isthmus were hypoplastic. A conduit was placed from the ascending to the descending aorta to bypass the hypoplastic arch (Conduit) and the main pulmonary artery was anastomosed to the ascending aorta (Stansel) because of the small size of the ventricular septal defect. LCO = left coronary orifice; Underdevel Comm = underdeveloped commissures of the aortic valve; {S.L.L} = atrial situs solitus. l-loop ventricle, malposition of the great arteries with the aorta to the left.

The indexed right (systemic) AV valve diameter had a significant direct correlation with the indexed aortic valve annulus diameter (Table 2) and the indexed ventricular septal defect size (Table 3) regardless of whether the valve was tricuspid or mitral.

An adequate Doppler examination was available in 50 of the 57 patients (32 with an l-loop ventricle and 18 with a
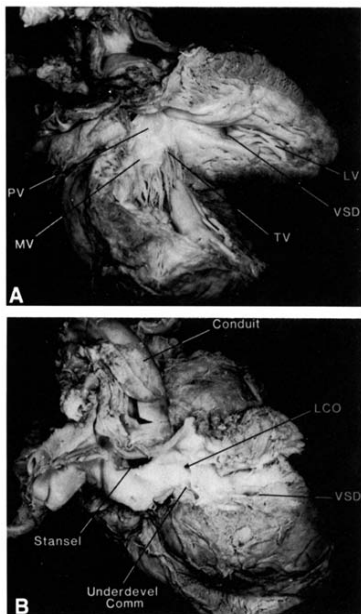
d-loop ventricle). Both AV valves were normal in 34 patients (68%; 23 with an l-loop ventricle and 11 with a d-loop ventricle; Table 1). Among patients with an l-loop ventricle, the mitral valve (right AV valve) was regurgitant in three
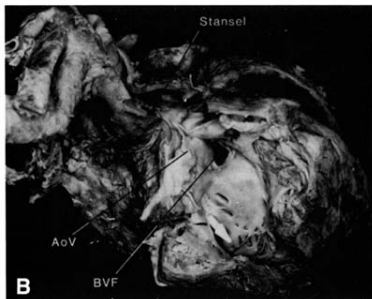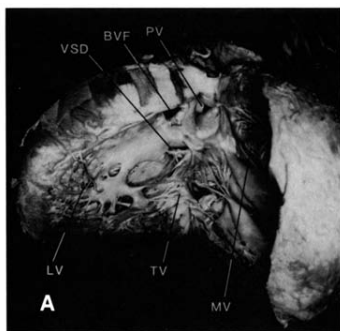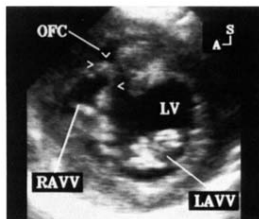
Figure 4. Parasternal short-axis view in a patient with a d-loop ventricle demonstrating straddling of the tricuspid valve (arrowheads) through a large posterior ventricular septal defect. Abbreviations as in Figure 1.

Figure 3. The heart of a 3-year 11-month old boy with double-inlet left ventricle and transposition {S.D-D}. A. The right-sided tricuspid valve (TV) has thickened leaflets and attaches to paraseptal papillary muscles and the lower margin of a small inlet muscular ventricular septal defect (VSD). Fibrous tissue accumulation around the subaortic ventricular septal defect (BVF) after banding of the pulmonary artery was associated with suprasystemic left ventricular pressure. The left-sided mitral valve (MV) has attachments only to the free wall of the left ventricle (LV). The pulmonary valve (PV) is well formed. B. The right-sided outflow chamber communicates with the left ventricle by means of the subaortic ventricular septal defect (BVF) and the small inlet defect (thick white arrow). The aortic valve (AoV) is normal. The proximal main pulmonary artery has been connected with the ascending aorta (Stansel). {S.D-D.} = atrial situs solitus, d-loop ventricle, malposition of the great arteries with the aorta located anteriorly.

patients and stenotic in none, whereas the tricuspid valve (left AV valve) was stenotic in seven patients and regurgitant in one patient (Fig. 5 and 6, A and B). Among the patients with a d-loop ventricle, the tricuspid valve (right AV valve) was stenotic in one patient and regurgitant in four patients.

whereas the mitral valve (left AV valve) was stenotic in one patient and regurgitant in none. Among the eight patients with a straddling tricuspid valve (Fig. 4), the straddling valve was mildly regurgitant in three patients, stenotic in one patient and functionally normal in the other four patients.

Ventricular septal defect. The morphology of the ventricular septal defect could be evaluated in 46 patients. In 24 patients, the defect was separated from the semilunar valves and appeared to be completely surrounded by muscle (muscular defect) (Fig. 2, A and B and 7A); it tended to enter the outflow chamber inferiorly and apically. In 19 patients the ventricular septal defect was adjacent to the anterior semilunar valve (subaortic defect) and was associated with hypoplasia of the infundibular septum (Fig. 6, A and B and 7B) in 5 patients and posterior malalignment of the infundibular septum with (Fig. 7C) or without (Fig. 7D) hypoplasia in the other 14 patients. The remaining three patients had multiple muscular ventricular septal defects (Fig. 3, A and B and 7E).

The size of the ventricular septal defect did not differ significantly between patients with a d-loop or an l-loop ventricle if transposition was present (Table 1). However, the patients with a d-loop ventricle and normally related great arteries (Holmes heart) had a significantly smaller ventricular septal defect than that of the patients in the other two groups.

The ventricular septal defect was unrestrictive in 26 (53%) and restrictive in 23 (47%) of the 49 patients in whom analysis was possible. Pulmonary artery banding had been performed in 8 (30%) of the 26 patients with an unrestrictive defect and 10 (43%) of the 23 patients with a restrictive defect. When examined by type of defect, only 4 (21%) of the 19 subaortic defects were restrictive, whereas 16 (67%) of the 24 muscular defects were restrictive. In the six patients with Holmes heart, all six defects were muscular and five were restrictive. The size of the ventricular septal defect was significantly correlated with the pressure gradient measured across the defect (Table 3) with use of an exponential regression (Fig. 8).

Table 2. Relation of Indexed Aortic Valve Anulus Size to the Occurrence of Other Structural Defects, Ventricular Loop and the Size of Related Cardiac Structures in 49 Patients With Transposition

	Indexed Aortic Valve Diameter (cm √ BSA)			
	Abnormality Present	Abnormality Absent	p Value	r Value
Structural abnormality				
Left AVV stenosis	2.4 ± 0.2	1.9 ± 0.1	0.0227	
Right AVV regurgitation	2.1 ± 0.8	2.0 ± 0.1	NS	
Systemic to pulmonary shunt operation	2.3 ± 0.1	1.9 ± 0.1	0.0215	
Coarctation	1.8 ± 0.1	2.2 ± 0.1	0.0096	
Ventricular loop				
l-loop	2.1 ± 1.0		NS	
d-loop		1.9 ± 0.1	NS	
Correlation between indexed aortic valve diameter and:				
Age			NS	0.26
Indexed left AVVD			NS	0.19
Indexed right AVVD			0.001	0.47
VSD gradient			NS	0.08
Indexed VSD area			NS	0.11
QP/QS			NS	0.04

BSA = body surface area; other abbreviations as in Table 1.

The ventricular septal defect size could be determined in 43 of 49 patients with transposition. Thirteen of these 49 patients had pulmonary stenosis and 30 had a normal pulmonary outflow tract. Only 2 (15%) of the 13 patients with pulmonary stenosis had a restrictive ventricular septal defect compared with 15 (50%) of the 30 patients without pulmonary stenosis. The size of the ventricular septal defect was significantly smaller in patients with transposition and coarctation of the aorta than in patients without coarctation (Table 3).

Aortic valve size. The aortic valve diameter did not differ significantly among the three groups of patients (Table 1). The aortic valve diameter was highly correlated with age (r = 0.52, p = 0.0002), body surface area (r = 0.84, p < 0.0001) and the square root of body surface area (r = 0.87, p < 0.0001). Indexing the aortic valve diameter for the square root of body surface area eliminated any significant correlation with age or body size.

The indexed aortic valve diameter was significantly smaller in patients with coarctation than in patients without

Table 3. Relation of Indexed Ventricular Septal Defect Size to the Occurrence of Other Structural Defects, Ventricular Loop, Gradient Through the Septal Defect and Size of Other Related Structures in 49 Patients With Transposition

	Indexed VSD Area (cm² BSA)			
	Abnormality Present	Abnormality Absent	p Value	r Value
Structural abnormality				
Left AVV stenosis	6.9 ± 2.2	10.6 ± 1.6	NS	
Right AVV regurgitation	13.7 ± 5.9	9.8 ± 1.5	NS	
Systemic to pulmonary shunt operation	12.4 ± 3.8	9.0 ± 1.3	NS	
Coarctation	4.7 ± 1.0	12.8 ± 1.8	0.0038	
Ventricular loop				
l-loop	10.6 ± 1.8		NS	
d-loop		9.2 ± 1.8	NS	
Correlation between indexed VSD area and:				
Age			NS	0.12
Indexed left AVVD			NS	0.22
Indexed right AVVD			0.013	0.37
VSD gradient			0.010	−0.41
Indexed AVD			NS	0.26
QP/QS			NS	0.15

Abbreviations as in Tables 1 and 2.

Figure 5. Subxiphoid short-axis view in an infant with double-inlet single left ventricle and transposition (S.L.L) illustrating hypoplasia of the left atrioventricular valve (L AVV). Abbreviations as in Figures 1 and 2.

coarctation (Table 2). It was not significantly correlated with the size of the ventricular septal defect (Table 2).

Associated anomalies. Coarctation of the aorta was present in 14 patients and interruption of the aortic arch in 2. Pulmonary stenosis was noted in 17 patients and pulmonary atresia in 6.

Necropsy findings. In eight of the patients who died, the heart specimen was available for examination. The necropsy findings confirmed the echocardiographic findings in all but three patients in whom additional small ventricular septal defects (two defects in two patients and one defect in one patient) were discovered that had been missed on both echocardiography and angiography.

Discussion

Extension of the use of the Fontan principle to the surgical management of patients with single left ventricle

Figure 6. The heart of a 2.5-year old boy with double-inlet left ventricle and transposition of the great arteries (S.L.L). **A.** The left-sided tricuspid valve (TV) is abnormal with thickened leaflets, a stenotic orifice and hypoplastic papillary muscles. Its attachments are paraseptal and at the lower margins of the subaortic ventricular septal defect (VSD). Fibrous tissue below the pulmonary valve produced severe subpulmonary stenosis (Sub PS). The right-sided mitral valve (MV) attaches only to the free wall of the hypertrophied left ventricle (LV). **B.** The left-sided infundibular outlet chamber has a very small cavity and hypertrophied free wall. The subaortic ventricular septal defect (VSD) is immediately below the aortic valve. The central aortopulmonary shunt (Ao-P Shunt) is seen exiting the side of the ascending aorta (Ao).
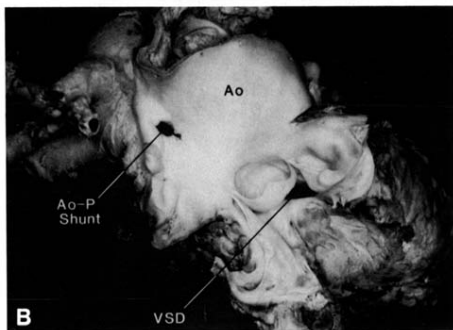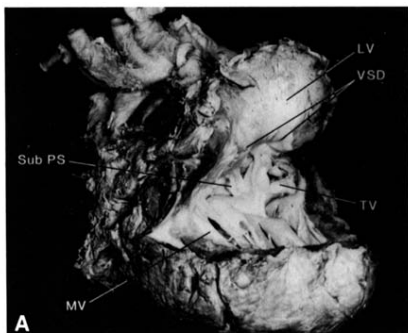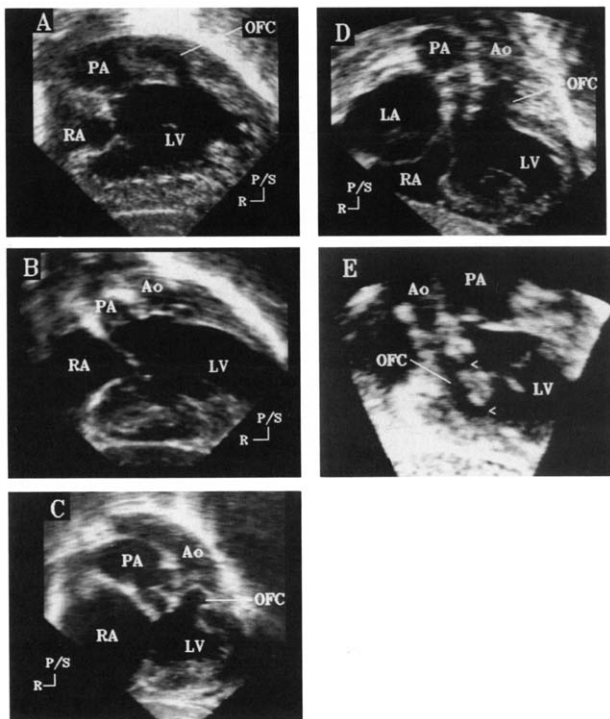
Figure 7. Types of ventricular septal defect seen in double-inlet single left ventricle as delineated by two-dimensional echocardiography. A, Muscular defect near the apex of the outlet chamber (OFC) and separated from the semilunar valves. B, Subaortic defect associated with extreme hypoplasia of the infundibular septum. C, Subaortic defect associated with posterior malalignment and hypoplasia of the infundibular septum. D, Subaortic defect associated with marked posterior deviation of the infundibular septum. Here the infundibular septum is well developed and produces significant subpulmonary stenosis. E, Multiple defects (arrowheads) in an infant with a d-loop single left ventricle. Ao = ascending aorta; P = posterior; R = right; RA = right atrium; other abbreviations as in Figure 1.

(24,25) mandates accurate assessment of the anatomy and function of these hearts. The morphology of the single ventricle, the morphology and function of the AV valves and

the size and location of the ventricular septal defect may all be important determinants of outcome in such patients.

Ventricular morphology. The ability of two-dimensional echocardiography to display the morphologic characteristics of the right and left ventricles has been demonstrated previously (17,18). Similarly, pathologic studies (26,27) in double-inlet left ventricle have described the single ventricular chamber as a typical left ventricle. Surprisingly, previous studies have found two-dimensional echocardiography to be unreliable in defining the morphology of the single ventricle. Rather, in these studies (11), the echocardiographic diagnosis of single left ventricle has generally been based on finding an outlet chamber or infundibulum located anterosuperiorly with respect to the single ventricle. In our series, the diagnosis of single left ventricle could be made on the basis of ventricular morphology. The difference between our

Figure 8. Correlation between the size of the ventricular septal defect (VSD Area) and the measured gradient across the defect (VSD Gradient) in 24 patients with no ductus arteriosus (closed circles). The 13 patients with a significant ductus arteriosus (open circles) were not included in the correlation analysis.

$$Y = 56 (1/X) + 0.1$$
$$SEE = 19$$
$$N = 24$$
$$r = 0.69$$
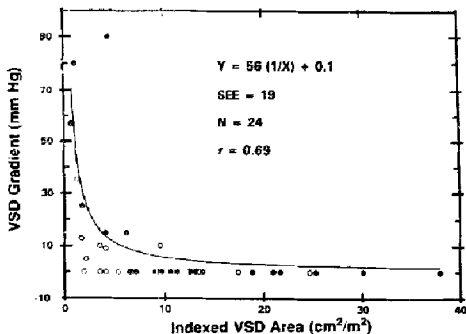
results and those of previous studies may be at least partly explained by improved echocardiographic images due to the young age of our patients and technologic advances.

Atrioventricular valves. As noted previously, the AV valve that is concordant with the ventricular loop (the right AV valve in d-loop and the left AV valve in l-loop) is commonly abnormal (5,12,26,28–32). In our series, it was abnormal in 25% to 30% of patients. This valve exhibits the characteristics associated with the tricuspid valve of the normal heart: it is closer to the septum (generally the posterior part of the septum), it may have septal attachments and in some cases it straddles the septum through a posterior ventricular septal defect (33). Hence, we think it is justified to identify it as the tricuspid valve. In our series as well as in previous reports (5,28–32) of double-inlet left ventricle, there was a tendency for the left-sided tricuspid valve to be stenotic in an l-loop ventricle and for the right-sided tricuspid valve to be regurgitant in a d-loop ventricle. These findings are of practical importance now that the Fontan principle is used commonly in patients with double-inlet left ventricle. Of interest, the straddling tricuspid valve functioned normally or was mildly regurgitant but was not stenotic.

Ventricular septal defect. The location and characteristics of the ventricular septal defect seen in single left ventricle are not uniform (34). At least two types of defects can be identified by echocardiography: subaortic defect, which is associated with hypoplasia or malalignment of the infundibular septum, or both, and muscular defect. In our series, only about 22% of subaortic defects were stenotic whereas 66% of muscular defects were obstructive. On pathologic examination, it may be possible to discriminate among subsets of these basic types (for example, muscular defects may be divided into mid-muscular and AV canal types), but we were unable to recognize the subtypes reliably by echocardiography.

Stenosis of the ventricular septal defect in patients with

transposition of the great arteries results in subaortic obstruction and adversely affects the outcome of patients with single left ventricle (35,36). The defect may be stenotic at birth or may become restrictive later in life (35,37–40). Although the mechanism by which it becomes restrictive is not known, the association between stenosis of the ventricular septal defect and banding of the pulmonary artery has been emphasized (35,37,39,40). Because a small ventricular septal defect was significantly more common in our patients without pulmonary stenosis, it may be that patients who undergo pulmonary artery banding are simply at higher risk for developing obstruction of the ventricular septal defect.

In our series, a significant inverse correlation was noted between the size of the defect and the pressure gradient measured across it. Nonetheless, many small defects were not associated with a detectable pressure gradient. Most likely, this is due to low flow across the defect in the presence of a large ductus arteriosus that carries most of the cardiac output. Consequently, absence of a pressure gradient does not exclude a restrictive ventricular septal defect. The size of the defect must be considered.

Conclusions. Double-inlet single left ventricle can be reliably diagnosed on the basis of ventricular morphology with use of two-dimensional echocardiography. The annular diameter and function of the AV valves were normal in about 70% of our patients. The AV valve on the side of ventricular looping exhibited characteristics usually associated with the tricuspid valve in the normal heart and was abnormal more often than the mitral valve. The abnormalities commonly seen were stenosis of the left-sided tricuspid valve in an l-loop ventricle and regurgitation of the right-sided tricuspid valve in a d-loop ventricle.

The ventricular septal defect was variable in size, location and morphology. Subarterial defects were associated with hypoplasia or malalignment, or both, of the infundibular septum and were less likely to be restrictive. Muscular

defects were commonly restrictive and the only type associated with a straddling tricuspid valve. In patients with transposition, a strong direct correlation was noted between both the size of the ventricular septal defect and the aortic anulus diameter and the presence of coarctation or arch interruption. A restrictive ventricular septal defect was present in five of six patients with normally related great arteries. Pulmonary stenosis was present in about 25% of patients with transposition and was usually not associated with a restrictive ventricular septal defect.

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