Double-outlet Right Ventricle: Wide-angle Two-dimensional Echocardiographic Observations

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SUMMARY M-mode echocardiographic delineation of double-outlet right ventricle (DORV) has relied primarily on the demonstration of mitral–semilunar valve discontinuity—a feature that requires an interpretation of spatial anatomic relationships. Thirty-six patients with DORV were examined by wide-angle real-time two-dimensional echocardiography. Anatomic diagnosis was established by surgery in 28 patients and by angiography alone in eight. Typical two-dimensional echocardiographic features were (1) parallel orientation and origin of both great arteries from the anterior right ventricle, (2) mitral–semilunar valve discontinuity demonstrated on parasternal long-axis scans by the presence of muscular conus separation, and (3) absence of left ventricular outflow other than a ventricular septal defect. This technique also allows better recognition of spatial orientation of the great arteries and the position of the ventricular septal defect relative to the great arteries. An unexpected finding was a high incidence of atrioventricular valvar anomalies, particularly annular override or abnormal chordal attachments (straddling) (eight patients), isolated cleft of the mitral valve (two patients), and complete atrioventricular canal (five patients). Thus, these two-dimensional echocardiographic findings have allowed improved noninvasive recognition of DORV and the demonstration of associated anomalies that previously were unrecognized preoperatively.

DOUBLE-OUTLET RIGHT VENTRICLE is an uncommon congenital cardiac defect characterized by the origin of both great arteries from the morphologic right ventricle. M-mode echocardiographic delineation of double-outlet right ventricle has relied primarily on the demonstration of mitral–semilunar valve discontinuity. However, this M-mode feature inherently has been limited by its dependence on operator interpretation of spatial anatomic relationships. Forbidable obstacles have been the complex semilunar valve and atrioventricular valve relationships frequently associated with double-outlet right ventricle, the inability to localize clearly the position of the ventricular septal defect, and the poor definition of the conus that produces the semilunar and atrioventricular valve separation. Because of the lack of adequate spatial orientation, even in experienced hands, the rapidity of an apex-to-base scan and the angle of beam projection could produce false-positive and false-negative results on M-mode echocardiography. Early two-dimensional real-time studies have suggested improved recognition of great vessel spatial relationships to diagnose double-outlet right ventricle more reliably.

We analyzed the two-dimensional wide-angle sector echocardiographic features of 36 patients with double-outlet right ventricle to determine distinctive features that would permit the noninvasive recognition of this complex congenital cardiac defect.

Definitions

For this report, we relied primarily on the definitions and classifications reported by Sridaromont et al. and the angiographic observations of Carey and Edwards and Hallermann et al. Double-outlet right ventricle was defined as a congenital cardiac defect in which both great arteries originate exclusively from the morphologic right ventricle and neither semilunar valve is in fibrous continuity with an atrioventricular valve. The definition proposed by Lev et al. and by Anderson et al., however, does not require mitral–semilunar valve discontinuity. The two-dimensional echocardiographic diagnosis has relied primarily on the orientation of the great vessels with reference to the plane of the ventricular septum and very small degrees of mitral–semilunar valve discontinuity may not be readily appreciated, so the criteria required for diagnosis have not rigidly included the presence of a large conus for mitral–semilunar valve discontinuity. Thus, for this study, two-dimensional echocardiography has relied primarily on the predominant or nearly exclusive origin of both great arteries from the morphologic right ventricle and the presence of some mitral–semilunar valve separation.

We also evaluated the great-artery relationships and the position of the ventricular septal defect in reference to the great arteries. Great-artery relationships were designated as (1) normal (aorta to the right and posterior to the pulmonary trunk), (2) side-by-side, (3) aorta anterior and to the right of the pulmonary trunk, (4) aorta directly anterior to the pulmonary trunk, and (5) aorta anterior and to the left of the pulmonary trunk. The location of the ventricular septal defect was designated as subpulmonary, subaortic, doubly committed or remote.

Methods

Our series involved 36 patients with double-outlet right ventricle who were examined by an 80° phased-array real-time sector scanner (Varian V-3000) from April 1977 to November 1979. The ages of the 36 patients (21 males and 15 females) ranged from 10 days to 39 years (mean 10.1 years). Three patients...
were younger than 1 year of age. Anatomic diagnosis was confirmed by catheterization and angiography in all patients and by surgery in 28 patients. All but four patients were studied immediately before and during cardiac catheterization. Patients were examined by two-dimensional echocardiography, and the diagnosis of double-outlet right ventricle was established prospectively. Great-artery orientation, ventricular septal defect location in reference to the great arteries, and associated anomalies also were identified. During cardiac catheterization, the cardiac chambers and great artery identification by two-dimensional scan was confirmed by catheter position and contrast echocardiographic studies. In the other four patients, previous cardiac catheterization and angiography allowed confirmation of the two-dimensional echocardiographic observations. Associated conditions detected at cardiac catheterization and surgery are summarized in Table 1. Pulmonary stenosis, pulmonary artery band and atrioventricular canal defects also were detected by two-dimensional echocardiography in these patients. The two-dimensional echocardiographic findings were reviewed independently of the surgical findings, and the patients were categorized with respect to the orientation of the great arteries and the position of the ventricular septal defect relative to the great arteries. The two-dimensional echocardiographic observations were subsequently correlated with the findings noted at surgery in 28 patients.

Studies were performed using a 2.25- or 3.5-MHz transducer, and the patients were examined while in the supine or left lateral decubitus position. One-inch videotape recordings were obtained and 35-mm stop-action photographs were taken. All measurements determined for this study were obtained from stop-action video images using the depth markers generated by the sector scanner. In all patients, we attempted to obtain parasternal, apical, and subcostal long- and short-axis and four-chamber views. Long-axis views (in reference to the long axis of the left ventricle and its outflow tract) were used primarily to assess atrioventricular valve and semilunar valve relationships in reference to the plane of the ventricular septum, while short-axis scans provided an analysis of great-artery orientation and relationships. Apical four-chamber views allowed demonstration of atrioventricular valve and ventricular chamber relationships. A complete description of methods of examination and anatomic validation has been previously discussed.15

Findings

Two-dimensional echocardiographic findings for the diagnosis of double-outlet right ventricle were established. Three basic observations were noted for the diagnosis: (1) origin of both great arteries from the anterior right ventricle; (2) mitral–semilunar valve discontinuity; and (3) absence of left ventricular outflow other than the ventricular septal defect.

Origin and Spatial Relationships of the Great Arteries

In all 36 patients, both great arteries were observed to originate predominantly from the anterior right ventricle. If one great artery overrode the ventricular septal defect, we required at least 80% commitment to the right ventricle. An associated observation was a parallel course of the great arteries at their origin, which was recognized by simultaneous observation of both semilunar valves originating from the right ventricle, as noted in the same parasternal or subcostal long-axis scan in 12 patients (10 with the pulmonary artery posterior to the aorta). In the other 24 patients, both great arteries (without clear visualization of both semilunar valves) could be observed originating anteriorly from the right ventricle on the same long-axis scan or with slight right or left transducer angulation (fig. 1). In four patients, all cardiac chambers, atrioventricular valves, and semilunar valves could be observed in the same parasternal long-axis scan (fig. 2). In one other patient, all four cardiac chambers and one or the other great artery originating from the right ventricle could be observed in the same parasternal long-axis scan.

Short-axis scans from the apex to base also were helpful in demonstrating the primary commitment of both great arteries to the right ventricular cavity, as observed by their anterior position with reference to the plane of the ventricular septum (fig. 3). These findings were similar to those described by Henry et al.7,8 By using a short-axis scan at the cardiac base, a double-circle appearance of the great arteries was consistent with a parallel orientation of the great arteries. In addition, the great-artery position relative to the ventricular septal defect and the great-artery spatial relationships could be demonstrated.

Wide-angle sector echocardiography allowed better recognition of spatial orientation of the great arteries and demonstration of the ventricular septal defect

### Table 1. Associated Conditions of 36 Patients With Double-outlet Right Ventricle

<table>
<thead>
<tr>
<th>Condition</th>
<th>No.*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valvular or subvalvular (or both) pulmonary stenosis</td>
<td>21</td>
</tr>
<tr>
<td>Pulmonary artery band</td>
<td>3</td>
</tr>
<tr>
<td>Complete atrioventricular canal</td>
<td>5</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous connection, right atrium</td>
<td>2</td>
</tr>
<tr>
<td>Asplenia</td>
<td>2</td>
</tr>
<tr>
<td>Polysplenia</td>
<td>2</td>
</tr>
<tr>
<td>Left juxtaposed atrial appendages</td>
<td>5</td>
</tr>
<tr>
<td>Severe pulmonary vascular</td>
<td>3</td>
</tr>
<tr>
<td>obstructive disease</td>
<td></td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>1</td>
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</tbody>
</table>

*Some patients had more than one associated defect.
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Arteries originate entirely from the pulmonary artery (PA). Both great arteries are committed to the right ventricular cavity and are observed in parallel orientation originating from the right ventricle (RV). In this standard long-axis scan, all four cardiac chambers and both great arteries are observed simultaneously. As identified, the cardiac chambers and great artery locations were verified at cardiac catheterization with two-dimensional echocardiographic contrast studies. The pulmonary valve appeared slightly thickened, and in real time the valve domed during systole, consistent with pulmonary valve stenosis. The mitral valve (MV) is markedly separated from the semilunar valves. LV = left ventricle; VS = ventricular septum; AS = atrial septum; LA = left atrium; RA = right atrium; TV = tricuspid valve.

Figure 1. (top) Parasternal long-axis scan in a patient who had double-outlet right ventricle, with the aorta (AO) anterior and to the left of the pulmonary artery (PA). Both great arteries originate entirely from the anterior right ventricular cavity. (bottom) A parallel orientation of the great arteries is observed with simultaneous visualization of both semilunar valves in the same long-axis plane. The PA is recognized by its posterior course to the lungs, and pulmonary stenosis is apparent. A large subpulmonary ventricular septal defect is observed. Arrowheads point to moderate subpulmonary conus tissue that separates the mitral valve (MV) from the pulmonary artery. Depth markers are present across the top and down the left side of the figures. RV = right ventricle; LV = left ventricle; LA = left atrium.

Figure 2. Parasternal long-axis scan in a patient who had double-outlet right ventricle, with the aorta (AO) anterior and to the left of the pulmonary artery (PA). Both great arteries are committed to the right ventricular cavity and are observed in parallel orientation originating from the right ventricle (RV). In this standard long-axis scan, all four cardiac chambers and both great arteries are observed simultaneously. As identified, the cardiac chambers and great artery locations were verified at cardiac catheterization with two-dimensional echocardiographic contrast studies. The pulmonary valve appeared slightly thickened, and in real time the valve domed during systole, consistent with pulmonary valve stenosis. The mitral valve (MV) is markedly separated from the semilunar valves. LV = left ventricle; VS = ventricular septum; AS = atrial septum; LA = left atrium; RA = right atrium; TV = tricuspid valve.

Location with reference to the great arteries. Parasternal long-axis scans (often from a slightly more superior position at the left sternal edge) demonstrated the initial parallel course of the great arteries. The pulmonary artery was recognized by its posterior course to the lungs, as observed in the long-axis scan (fig. 4), and its bifurcation into right and left pulmonary artery branches, as noted with short-axis scans. A more anterior and superior course of the great artery allowed its identification as the aorta. Short-axis scans at the cardiac base allowed recognition of the great-artery relationships (figs. 3 and 5). Orientation of the great arteries was observed by sector echocardiography in 32 of the 36 patients (table 2). The aorta was anterior to the pulmonary artery in 20 (63%) of the 32 patients. These great-artery positions were confirmed at surgery in 26 patients and by angiography alone in six.
Figure 3. Short-axis scan of heart from apex (A) to base (E) in a patient with double-outlet right ventricle. As scan progresses up through the atroventricular valves, the ventricular septal defect is observed. At the cardiac base, it is apparent that the ventricular septal defect is the only outlet from the left ventricular cavity and that both great arteries originate entirely from the anterior right ventricular cavity. The pulmonary artery (PA) is to left and is separated from the ventricular septal defect by a bridge of muscular tissue. Thus, the ventricular septal defect is noted to be subaortic at the cardiac base. The aorta (AO) is anterior and to the right of the PA, and a large conus is observed posterior to both great arteries. The pulmonary valve appears bicuspid and small in this patient with associated pulmonary stenosis. Ventricular septal defect is indicated by arrow. RV = right ventricle; VS = ventricular septum; LV = left ventricle; RA = right atrium; LA = left atrium; MV = mitral valve.

Mitral–Semilunar Valve Discontinuity

On long-axis scans, mitral-semilunar valve discontinuity was demonstrated by the presence of muscular conus separation (fig. 6). In 13 of the 29 patients (45%) for whom this observation was recorded, the conus separation was small (less than 1 cm) (fig. 6). In the other 16 patients, a large conus or separation was
Figure 4. Parasternal long-axis scan from slightly more superior position at left sternal edge in same patient as in figure 1 demonstrates posterior angulation of pulmonary artery (PA) to lungs, which allows its identification as the PA. Arrowheads indicate the posterior course of the PA. Both great arteries are entirely committed to right ventricular cavity (RV). Subpulmonary conus is demonstrated by the large arrow. AO = aorta; VS = ventricular septum; LV = left ventricle; LA = left atrium.

Figure 5. Short-axis scan at cardiac base in patient with double-outlet right ventricle. A double-circle appearance of the great arteries is noted. Scan shows aorta (AO) anterior and slightly to left of pulmonary artery (PA). LA = left atrium; LAA = left atrial appendage.

Figure 6. (A) Parasternal long-axis scan in same patient as in figure 3 demonstrates large conus (c) separating mitral valve (MV) from aortic valve (AV), which originates entirely from right ventricular cavity (RV) and is superiorly displaced by conus tissue. A ventricular septal defect is the only outlet from left ventricular cavity (LV), and is subaortic in position. VS = ventricular septum. (B) Parasternal long-axis scan in another patient with double-outlet right ventricle and subaortic ventricular septal defect. Aorta (AO) is nearly entirely committed to anterior RV. Ventricular septal defect is sole outlet from the LV. Smaller subaortic conus (arrowhead, approximately 1 cm) separates mitral valve (MV) from aortic valve. LA = left atrium; VS = ventricular septum.
Table 2. Associated Two-dimensional Echocardiographic Findings Compared With Angiographic and Surgical Findings in Patients With Double-outlet Right Ventricle

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<th>Finding</th>
<th>2-D echo</th>
<th>Angiography</th>
<th>Surgery</th>
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<tr>
<td>Aorta position</td>
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<tr>
<td>Right and posterior to pulmonary trunk (normal)</td>
<td>7</td>
<td>9</td>
<td>6</td>
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<tr>
<td>Side-by-side with pulmonary trunk</td>
<td>5</td>
<td>6</td>
<td>3</td>
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<tr>
<td>Right and anterior to pulmonary trunk</td>
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<td>2</td>
</tr>
<tr>
<td>Left and anterior to pulmonary trunk</td>
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</tr>
<tr>
<td>Total</td>
<td>32</td>
<td>36</td>
<td>26</td>
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<td>Ventricular septal defect position</td>
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<tr>
<td>Subaortic</td>
<td>12*</td>
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<tr>
<td>Subpulmonary</td>
<td>14</td>
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<td>Doubly committed</td>
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</tr>
<tr>
<td>Remote</td>
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<td>Total</td>
<td>35</td>
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<tr>
<td>Associated defects</td>
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<td>Atrial and ventricular septal malignment (annular override)</td>
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<td>2</td>
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<tr>
<td>Type A straddling mitral valve (minor straddling)†</td>
<td>3</td>
<td>—</td>
<td>3</td>
</tr>
<tr>
<td>Type C straddling mitral valve (major straddling)‡</td>
<td>1</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Type A straddling tricuspid valve (minor straddling)‡</td>
<td>2</td>
<td>—</td>
<td>2</td>
</tr>
<tr>
<td>Mitral valve eleft</td>
<td>2</td>
<td>—</td>
<td>2</td>
</tr>
</tbody>
</table>

*Associated muscular ventricular septal defects in one patient.
†Five with atrioventricular canal.
‡Type A = chordal attachments to the crest or within 1 cm of the crest of the ventricular septum; type B = chordal attachments to the opposite side of the ventricular septum; type C = chordal attachments to the free wall or papillary muscle of the opposite ventricle.

Present. Also, because of the superior displacement of the semilunar valves, both atrioventricular valves and all four cardiac chambers could be observed simultaneously in the standard parasternal view that ordinarily would show a long-axis scan (fig. 7).

Absence of Left Ventricular Outflow Tract Other Than the Ventricular Septal Defect

Although similar in concept to the findings of commitment of both great arteries to the right ventricle, this observation was striking in the long-axis view,

Figure 7. Parasternal long-axis scan in patient with double-outlet right ventricle demonstrates foreshortened four-chamber view with simultaneous visualization of both atrioventricular valves. A large papillary muscle and chordal attachment of the tricuspid valve (tv) are also noted in the right ventricle. The left atrium is posterior to the atrial septum (AS). Semilunar valves are not observed because of their superior displacement from the atrioventricular valves by conus tissue. RV = right ventricle; LV = left ventricle; VS = ventricular septum; mv = mitral valve; RA = right atrium.
both from the parasternal and subcostal positions (fig. 6). All 36 patients had associated ventricular septal defect, and in 35 of the 36 patients, sector echocardiography accurately predicted the position of the ventricular septal defect in reference to the great arteries (28 surgically confirmed and seven confirmed angiographically) (table 2). In most patients, typical subaortic or subpulmonary defects could be demonstrated by parasternal or subcostal long-axis scans (figs. 2 and 6). Short-axis scans of the heart were helpful in confirming this relationship (fig. 3). Very large ventricular septal defects or doubly committed defects were recognized by parasternal or apical long-axis views. Remote defects (noncommitted or posterior) were usually complete atrioventricular canal-type defects and were best recognized by apical four-chamber views. Two patients had a posterior or remote ventricular septal defect, but not a complete atrioventricular canal defect. In six patients, two-dimensional echocardiography revealed small or restrictive ventricular septal defects (all surgically confirmed). In one patient examined during cardiac catheterization, left ventricular contrast sector echocardiography using indocyanine green dye revealed additional muscular ventricular septal defects, as was observed with a parasternal four-chamber view. This observation was confirmed by subsequent angiography and surgery. In one patient, the ventricular septal defect position was not adequately defined in reference to the great arteries.

Three patients had two-dimensional echocardiographic features consistent with double-outlet right ventricle, which at surgery were found to be tetralogy of Fallot in one patient and transposition of the great arteries in two patients. In these, only a small mitral–semilunar valve separation was present; however, primary commitment of both great arteries to the right ventricle led to the two-dimensional echocardiographic diagnosis of double-outlet right ventricle. In two of these patients, the angiographic and two-dimensional echocardiographic diagnoses were the same, while in one patient, the angiographic diagnosis was transposition of the great arteries.

Associated Abnormalities

Another unexpected and previously unrecognized observation in these patients was the high incidence of atrioventricular valve abnormalities (15 of 36 patients) that included (1) complete atrioventricular canal defect, (2) isolated cleft of the anterior mitral valve leaflet, (3) straddling left and right atrioventricular valves, and (4) atrioventricular septal malalignment (annular override) (table 2). These abnormalities were diagnosed primarily on two-dimensional echocardiography and confirmed surgically or pathologically in all except two patients with associated complete atrioventricular canal and one patient with severe straddling mitral valve. Most were detected using the apical four-chamber view. The apical four-chamber view demonstrated a type C (Rastelli classification) complete atrioventricular canal defect in five patients. In two patients, short-axis scans of the mitral valve demonstrated a cleft of the anterior leaflet (associated with a posteriorly located ventricular septal defect but with intact atrial septum). With the apical four-chamber view, two patients had pronounced atrial and ventricular septal malalignment without actual valve straddling or abnormal chordal attachments (annular override). Three patients had minor type A straddling of the left-sided mitral valve with chordal attachment to the crest of the ventricular septum (fig. 8). One patient had major (type C) straddling of the left-sided mitral valve, and two patients had a minor (type A) straddling right-sided tricuspid valve with chordal attachments to the crest of the ventricular septum.

Discussion

Wide-angle sector echocardiography has greatly improved the noninvasive recognition of double-outlet right ventricle and its differentiation from other conotruncal abnormalities. The recognition of double-outlet right ventricle has improved primarily because of better assessment by two-dimensional echocardiography of the primary commitment of both great arteries to the right ventricle and because of improved recognition of mitral–semilunar valve discontinuity. Mitral–semilunar valve discontinuity has been more clearly established by the recognition of the conus that

Figure 8. Apical four-chamber view in patient with double-outlet right ventricle demonstrates severe atrial and ventricular septal malalignment (valve annular override). Abnormal chordal attachment of left (mitral) atrioventricular valve to crest of ventricular septum, representing type A straddling valve. RV = right ventricle; VS = ventricular septum; LV = left ventricle; ch = chordae; mv = mitral valve; tv = tricuspid valve; RA = right atrium; as = atrial septum; LA = left atrium.
produced physical separation and a more superior position of the semilunar valves relative to the atrioventricular valves. The spatial recognition of the more superior position of the semilunar valve was not possible by M-mode echocardiography. Very small amounts of subvalvular conus (less than 4–5 mm) approached the resolving limits (lateral resolution) of current two-dimensional instrumentation. Thus, consistent recognition of such small degrees of mitral–semilunar valve discontinuity remained difficult echocardiographically. It should also be noted that such small separation is difficult to demonstrate by angiography.

In four patients with double-outlet right ventricle, all cardiac chambers, atrioventricular valves and semilunar valves could be observed in the same parasternal long-axis scan (fig. 2). This observation, in our experience, has been observed only in patients with double-outlet right ventricle. In addition, the observation of a four-chamber type view (all four cardiac chambers and both atrioventricular valves) from a standard position for a left parasternal long-axis scan was a frequent finding in patients with double-outlet right ventricle, although it was not one that is diagnostic for this entity (fig. 7). We believe that the four-chamber view is frequently observed from this standard position for a parasternal long-axis scan because of the more superior position of the semilunar valves as a result of their physical separation by conus tissue. In addition, enlargement of the right atrial and right ventricular cavities as an associated finding in double-outlet right ventricle may produce some cardiac rotation, allowing better observation of these structures in a standard parasternal long-axis scan.

Three patients had two-dimensional echocardiographic features consistent with double-outlet right ventricle, but surgery revealed tetralogy of Fallot in one and transposition of the great arteries in two. Two of these patients had angiographic diagnosis of double-outlet right ventricle, and one had angiographic diagnosis of transposition of the great arteries. When only a small conus separation was present, the two-dimensional echocardiographic diagnosis relied primarily on the spatial orientation (primary commitment) of both great arteries to the right ventricle, so a retrospective review of the two-dimensional echocardiographic findings in these patients did not resolve these differences between the two-dimensional echocardiographic, angiographic, and surgical diagnoses. These differences in diagnosis should not be considered false-positive echocardiographic diagnoses, but rather, they reflect the differences in the interpretation of the same anatomic features by three methods of observation. The surgical observations are considered the most reliable, but the surgical view is limited and cannot be considered the same as one observed pathologically, angiographically or echocardiographically. Thus, the degree of arterial override and mitral–semilunar valve separation will vary with each method of observation and, in instances such as these, may result in variable diagnoses. Because of the subjectivity of lesser degrees of arterial override, we required at least 80% or primary commitment of the overriding great artery to the right ventricle.

Twenty-one patients had pulmonary stenosis, occurring most frequently in patients with the aorta anterior to the pulmonary artery. This great-artery relationship allowed easier recognition of the pulmonary stenosis. Only three patients with normally related great arteries and none with side-by-side great arteries had associated pulmonary stenosis. The presence or absence of pulmonary stenosis was also successfully recognized in this small subgroup. However, more severe distortion and obstruction of the pulmonary outflow, similar to that observed in patients with tetralogy of Fallot, may impair the ability to recognize the pulmonary valve by two-dimensional echocardiography. In such cases, a positive diagnosis of double-outlet right ventricle may not be possible.

Two-dimensional echocardiography allowed better recognition of the great-artery relationships and the relative position of the ventricular septal defect in reference to the great arteries. In our series, a very high correlation was found between the two-dimensional echocardiographic description and the angiographic and surgical observations. Accurately described by two-dimensional echocardiography were the great-artery relationships in 32 of the 36 patients and the ventricular septal defect position in 35 of the 36 patients. Two-dimensional echocardiographic assessment of the ventricular septal defect position relative to the great arteries seems at least as accurate as the angiographic determination. One patient with remote ventricular septal defect by two-dimensional echocardiography and surgery was considered to have subaortic ventricular septal defect by angiography.

An unexpectedly high incidence of atrioventricular valve abnormalities was demonstrated by two-dimensional echocardiography. The abnormalities had not been previously suspected by clinical or angiographic observations. The association of complete atrioventricular canal and double-outlet right ventricle has been described, but it represents a difficult angiographic diagnosis because of a lack of a typical gooseneck deformity of the left ventricular outflow tract. However, the presence of complete atrioventricular canal can be assessed very readily by two-dimensional echocardiography using the apical four-chamber view. In addition, previously described mitral valve abnormalities (cleft mitral valve and straddling mitral valve), as observed in our series of patients with double-outlet right ventricle, have been rare. Lev and associates briefly described the association of normally formed or cleft anterior mitral leaflet in double-outlet right ventricle with subpulmonary ventricular septal defect. Tandon et al. first reported the association of straddling mitral valve with double-outlet right ventricle. Van Praagh et al. described the pathologic findings in one patient with mitral valve abnormalities. In a series of 62 patients, Sridaromont et al. described one patient with straddling mitral valve and
two patients with parachute mitral valve. The patient with straddling mitral valve died after attempted surgical correction, as did the two patients with straddling mitral valve in a series of 87 patients with double-outlet right ventricle reported by Sondheimer and associates.21 We found that the anatomic features of straddling atrioventricular valve can be more clearly defined by two-dimensional echocardiography than by previous angiographic methods. In our series, preoperative two-dimensional echocardiographic recognition of this associated anomaly led to successful surgery in two patients who had minor straddling of the mitral valve and in one patient who had minor straddling of the tricuspid valve. For the patient with major straddling (type C) of the mitral valve, surgery was not advised because mitral valve replacement would have been necessary.

Previously, we reported the two-dimensional echocardiographic features of other forms of conotruncal abnormalities, such as truncus arteriosus, pulmonary atresia with ventricular septal defect, tetralogy of Fallot and complete transposition of the great arteries.22-24 The two-dimensional findings in these abnormalities, as also reported by other investigators,7, 8, 25-28 are different from those we describe with double-outlet right ventricle. In three patients, discrepancies among the two-dimensional echocardiographic, angiographic and the surgical interpretations revealed the range of these observations in double-outlet right ventricle, tetralogy of Fallot and complete transposition of the great arteries.

We conclude that wide-angle two-dimensional echocardiography has allowed improved diagnostic recognition of double-outlet right ventricle, its distinction from other conotruncal abnormalities, and demonstration of associated anomalies previously unrecognized preoperatively.

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Double-outlet right ventricle: wide-angle two-dimensional echocardiographic observations.

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