Echocardiographic Diagnosis of Double-Outlet Right Ventricle*

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An echocardiographic technique (V scan) for confirming the diagnosis of double-outlet right ventricle is described. The technique demonstrates a frequent pathologic feature of double-outlet right ventricle, i.e., origin of both great arteries side by side and anterior to the ventricular septum. The V scan offers greater diagnostic accuracy than the mere demonstration of mitral-semilunar valvular discontinuity.

Double-outlet right ventricle is a rare anomaly that is now amenable to complete surgical correction.\(^{1,4}\) Disagreement exists as to the exact pathologic criteria for its diagnosis,\(^{5-9}\) but the usual feature is that both great arteries arise anterior to the ventricular septum. In the usual situation, there is a bilateral conus and fibrous discontinuity between the semilunar valves and the atrioventricular valves. Previous papers, in which the echocardiographic diagnosis of double-outlet right ventricle was discussed, stressed the necessity of demonstrating mitral-semilunar valvular discontinuity to make this diagnosis;\(^{10-14}\) however, it has recently been pointed out that this criterion can be subject to error\(^{10,15}\) due to position and differences in the angulation of the transducer. The purpose of this report is to describe an echocardiographic method (V scan) which demonstrates a frequent anatomic feature of double-outlet right ventricle, i.e., the origin of both great arteries anterior to the ventricular septum. This method, when used as part of a complete echocardiographic examination in conjunction with the demonstration of mitral-semilunar valvular discontinuity, enhances the accuracy of the echocardiogram in the diagnosis of double-outlet right ventricle.

**Materials and Methods**

The echocardiographic and angiographic findings of three cases of double-outlet right ventricle were reviewed and form the basis of this report. Patient 1 was a girl with a double-outlet right ventricle and a large subpulmonic ventricular septal defect who had undergone pulmonary artery banding in infancy. Patients 2 and 3 had double-outlet right ventricle of the more common variety with subaortic ventricular septal defects; these two patients had also undergone pulmonary artery banding (Table 1).

Echocardiograms were obtained with an ultrasonic scope (Ultrasoundcope, Smith-Kline Instruments), using a 2.25-MHz and 5-MHz nonfocused transducer of 0.25-inch diameter. The records were made on strip chart recorder (Honeywell) at a paper speed of 50 mm/sec. The time lines indicated 0.5 second intervals. The examination was begun with the transducer at the left sternal border, from which position all four cardiac valves were located in the left-right as well as superior-inferior axis by previously described techniques.\(^{16-21}\) Scanning between the semilunar valves and atrioventricular valves was then done to define the relationship among

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Case 1 (7 yr, F)</th>
<th>Case 2 (6 mo, F)</th>
<th>Case 3 (6 mo, M)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pressure, mm Hg (mean)</td>
<td>AO 88/50 PA 20/6</td>
<td>AO 90/50 PA 55/20</td>
<td>AO 95/50 PA 60/20</td>
</tr>
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<td>(69) (10)</td>
<td>(67) (35)</td>
<td>(62) (40)</td>
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<tr>
<td>Oxygen saturation, percent</td>
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<td>92</td>
<td>85</td>
</tr>
<tr>
<td>R-C interval, seconds</td>
<td>0.36</td>
<td>0.24</td>
<td>0.20</td>
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<tr>
<td>Outside diameter, mm</td>
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<td>14</td>
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<tr>
<td>Depth, mm**</td>
<td>39</td>
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*AO, Aorta; and PA, pulmonary artery.
**Distance from anterior chest wall to anterior wall of great artery.

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RESULTS

All three patients had echocardiographic evidence of mitral-semilunar valvular discontinuity. The echocardiographic diagnosis of double-outlet right ventricle was confirmed by the V-scan maneuver. As can be seen (Fig 4), the aortic and pulmonic valves are shown anterior to the ventricular septum approximately equidistant from the anterior chest wall. In Figure 5, the ventricular septum is not seen in the V scan, probably because of malalignment of the septum or the presence of the ventricular septal defect. In addition, both semilunar valves were located at the same level on the superior-inferior axis. In the case with tetralogy of Fallot (Fig 2) and in the normal heart (Fig 3), the posterior wall of the aorta is always seen before the tricuspid valve during the V scan. This reflects the fact that the pulmonary artery normally overlaps the aorta. None of the cases of double-outlet right ventricle showed this finding during the V scan, which indicates that both great arteries are side by side. In all of our three cases of double-outlet right ventricle, the aorta was to the right of the pulmonary artery with a concordant D-loop. The semilunar valves were in close relation to the tricuspid valve. In Figure 5, the bilateral outflow tracts are clearly demonstrated.

DISCUSSION

Previous echocardiographic reports have stressed the necessity of demonstrating mitral-semilunar valvular discontinuity for the diagnosis of double-outlet right ventricle. However, mitral-semilunar discontinuity may be a variable sign and susceptible to technical error, due to position and angulation of the...
transducer. The V scan described in this report avoids these errors by clearly showing that aortic and pulmonary valves lie side by side, anterior to the ventricular septum and apart from the posterior atrioventricular valves in true double-outlet right ventricle (Fig 4 and 5). Although our experience is limited, this technique, when properly executed, adds accuracy to the echocardiographic diagnosis of double-outlet right ventricle. Important steps in the V scan are (1) locating the semilunar valves, (2) obtaining an echocardiogram of good quality showing the closure of the semilunar valves, and (3) identifying the great arteries by the R-C interval and outside diameter at the valvular level. Measurements of the R-C interval are obviously very important. The paper speed of 50 mm/sec used in our report has been found to be satisfactory in the determination of R-C interval by other works, although a paper speed of 100 mm/second is preferable.

Identification of the pulmonary artery by the R-C interval is more difficult in pulmonary hypertension because the pulmonary diastolic pressure is approximately that of the aorta; however, in our cases the diastolic pressure differences were great enough to allow identification of the two great arteries by the criterion of the R-C interval (Table 1). In addition, in the absence of pulmonic stenosis, the greater diameter of the pulmonary artery also helps in the identification of this vessel. With pulmonic stenosis the diameter of the pulmonary artery may be less than that of the aorta, but in this case the R-C interval, when measurable, is prolonged. In the presence of an effective pulmonary artery band, the R-C interval is also prolonged.

The V scan alone may not differentiate true double-outlet right ventricle from a single ventricle with bilateral conus or l-transposition with a malaligned septum. A differentiating feature of single ventricle with bilateral conus not seen in double-outlet right ventricle is that both atrioventricular valves open into a ventricular chamber. In our cases of double-outlet right ventricle, the atrioventricular
Figure 5. Echocardiogram (case 2) showing sequential record of V scan from pulmonic to tricuspid (TV) to aortic valves. Note features similar to case 3 (Fig 4). Ventricular septum is not seen. Outflow tract (OT) separates both semilunar valves from tricuspid valve (TV). During examination, semilunar valve of larger great vessel (PA) is found to left of semilunar valve of smaller great vessel (AO); both valves are at same level in superior-inferior axis.

Valves are on different sides of the ventricular septum. Furthermore, in the most common form of single ventricle, the aorta arises anteriorly from the outflow chamber and lies to the left side of the pulmonary artery. In l-transposition with a malaligned septum, in which bilateral conus is sometimes present, the aorta is always to the left and usually anterior to the pulmonary artery; however, in double-outlet right ventricle with d-loop, the aorta is always to the right and at the same depth as the pulmonary artery. In cases of tetralogy of Fallot with overriding of the aorta, the close relationship between the aorta and mitral valve is easily visualized, and, in addition, the posterior aortic wall is seen posterior to the ventricular septum. The V-scan echocardiogram of tetralogy of Fallot is different from that of double-outlet right ventricle in that the posterior aortic wall is clearly seen posterior to the posterior wall of the pulmonary artery (Fig 2). In the normal heart the V-scan echocardiogram shows aortic-mitral continuity and the continuity between the anterior aortic wall and ventricular septum (Fig 3).

In conclusion, V-scan echocardiography is a useful technique in the diagnosis of double-outlet right ventricle, when used in conjunction with the standard echocardiographic examination.

REFERENCES

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Marfan's Syndrome

The eponymic title of this subject originates from a report published eighty years ago [Marfan, AB (Bull Soc Med Hop Paris 13:220, 1896)]. It is within the domain of interest of the chest physician because of involvement of the cardiovascular and respiratory systems. The syndrome is known also as arachnodactyly, and dolicostenomelia. This inherited autosomal dominant trait is associated with generalized premature dystrophy of mesodermal tissues, with a tendency to fragility and with a host of anatomic anomalies. According to Priest, RE et al (Nature 245:264, 1973) collagen produced by fibroblasts is more soluble than normal in solvents used to extract it. The syndrome may be observed in adults and in children. Hohn, AR et al (Am J Dis Child 122:526, 1971) recorded its occurrence in 5-month-old twins. Bowers, D (Canad M A J 89:337, 1963) reported on a family of which 42 members were afflicted by Marfan's syndrome. Typical cases occur in both sexes, are likely to be underweight, show unusual height, slender habitus, excessively long extremities, disproportionately elongated digits, dolichocephaly, prominent supraorbital ridge, enlarged paranasal sinuses, long, thin face, protuberant jaw with long narrow teeth, highly arched palate, large pointed ears, sparse muscles and subcutaneous tissue, tall vertebral bodies, low position of the sacrum relative to the iliac crest, pectus excavatum or pectus carinatum, striae distensae of the skin on the upper thorax, lower abdomen and buttocks. Also, there may be kyphoscoliosis, spina bifida, widening of the spinal canal, calcaneal spurs, weakening of muscles, relaxation of ligaments, weakness of joint capsules, hyperflexibility and recurrent dislocation of joints. Family history may reveal similar traits in other siblings, parents and relatives. Ocular anomalies, such as dislocation of the optic lenses due to weakened suspensory ligaments, nyctagynus, and tendency to cataracts occur in from 50 to 75 percent of these patients. Retinal detachment is frequent. Occasionally, cornes frustes may be encountered. Cardiovascular disease may be seen in from 30 to 60 percent of subjects with this syndrome, with about one-third of this contingent having dissecting aneurysm of the ascending segment or arch of the aorta, with underlying cystic medial degeneration, and fragmentation of elastic fibers. Such occurrence in the abdominal aorta is very rare, although fusiform aneurysm has been reported. There may be dilatation of the aortic sinuses of Valsalva. Mitral insufficiency is frequent. Phornputkul, C et al (Circulation 47:537, 1973) found that out of 36 infants and children with this syndrome, 47 percent had mitral regurgitation and 11 percent aortic regurgitation. Also, one may encounter atrial and interventricular septal defects, slight coarctation, patent ductus arteriosus and hypoplastic aorta. James, TN et al (Arch Int Med 114:339, 1964) noted arrhythmias and conduction disturbances associated with pathologic changes in the sinus node and A-V node in two young men with this syndrome. Alarcon-Segovia, D et al (Chest 54:73, 1968) observed a 19-year-old Mexican Indian with this syndrome who had nephrotic manifestations due to renal vein thrombosis probably secondary to nearby large dissecting aneurysm of the abdominal aorta. In addition to the usual diagnostic means, Spangler, RD et al (Chest 69:72, 1976) emphasized the advantages of echocardiography for the detection of isolated dysfunction of the mitral valve and of the involvement of the aortic root. Inadequacy of pulmonary connective tissue may result in cystic formation (polycystosis, congenital bronchiectasis) with honeycomb appearance in the chest x-ray. It may lead to recurrent spontaneous pneumothorax. Also, one may find diffuse chronic emphysema or bullous emphysema. Lung function tests may be normal or reveal dysfunction proportionate to respective thoracic deformity and/or pulmonary abnormalities. As a corollary, attention may be called to the article, entitled "Abraham Lincoln and the Marfan Syndrome," by Schwartz, H (JAMA 187:473, 1964).

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