Double-outlet Right Ventricle Associated with Persistent Common Atrioventricular Canal

By Somkid Sridaromont, M.D., Robert H. Feldt, M.D., Donald G. Ritter, M.D., George D. Davis, M.D., Dwight C. McGoon, M.D., and Jesse E. Edwards, M.D.

SUMMARY
A variant of double-outlet right ventricle (DORV) associated with common atrioventricular (A-V) canal was identified. As yet, this syndrome has not been successfully repaired. Of 16 patients with this diagnosis, 14 had the diagnosis confirmed at postmortem examination and two had the diagnosis confirmed by exploratory cardiotomy. The 16 patients were divided into two groups according to the position of the cardiac apex: group 1, ten patients with levocardia, and group 2, six patients with dextrocardia and ventricular inversion. When pulmonary stenosis was present, the clinical presentation was similar to that in tetralogy of Fallot, and when such stenosis was not, the presentation was similar to that in ventricular septal defect with bidirectional shunting. The electrocardiogram had the typical feature of common A-V canal. The right ventricular angiocardiogram had features typical of DORV. The left ventricular angiocardiogram revealed left ventricular outflow deformity typical of A-V canal in patients with levocardia but not in those with dextrocardia. Splenic anomalies were commonly seen (nine of the 16 patients). Asplenia was present in seven, and polysplenia in two. Ten patients had persistent left superior vena cava, seven had common atrium, six had anomalous pulmonary venous connection, and three had subaortic stenosis. Twelve of the 16 patients had pulmonary stenosis. Of the 16, 14 had common A-V canal (Rastelli type C) and two had a ventricular septal defect of the persistent common A-V type.

The uncommon association of double-outlet right ventricle with persistent common atrioventricular canal has been recognized by many authors, including Witham,1 Neufeld et al.,2 Carey and Edwards,3 Ruttenberg et al.,4 and Rastelli et al.5 In this condition, when ventricular inversion is not present, both great arteries arise from the right-sided anatomic right ventricle. If ventricular inversion exists, both great arteries arise from an inverted left-sided anatomic right ventricle. Either of these two anatomic variations may be associated with other variations or anomalies.

We encountered the association of atrioventricular canal in patients who were first diagnosed as having double-outlet right ventricle. This prompted us to review these cases, with emphasis on clinical and angiographic findings and associated cardiovascular anomalies.

Patients and Methods
Sixteen patients with the characteristic anatomic features of double-outlet right ventricle6,7 and persistent atrioventricular canal8 were available for study. In this series of 16 patients, the diagnosis was established at autopsy in 14 and at exploratory cardiotomy in two. Eight patients were seen at the Mayo Clinic from 1960 to 1974, and data on the remaining eight patients were obtained from the pathologic collection of the cardiovascular registry of the Miller Division of United Hospitals from 1958 to 1974. The patients were divided into two groups according to the position of the cardiac apex (table 1). Group 1 consisted of ten patients in whom the cardiac apex was to the left (levocardia); ventricular inversion was not present in any. Group 2 consisted of six patients in whom the cardiac apex was directed to the right (dextrocardia); ventricular inversion, in which the morphologic right ventricle is on the left side of the heart, was present in all six patients. Both groups were subdivided according to whether the patient had a normal spleen, asplenia, or polysplenia. The criteria for asplenia by Ruttenberg et al.8 and for polysplenia by Moller et al.10 were used.

Observations
In 14 patients, the atrioventricular canal was of the complete form. In the other two (cases 3 and 11), the atrial septum was intact whereas the atrioventricular valves were cleft (fig. 1). The common anterior leaflet in each patient who had the complete form (figs. 2 and 3) was undivided and was not attached to the crest of the ventricular septum (type C of Rastelli et al.11). The common posterior leaflet was also undivided but was often attached to the ventricular septum by chordae. Each of two patients (cases 3 and 9) had a muscular
### Table 1

**Data on 16 Patients with Double-Outlet Right Ventricle with Atroventricular Canal**

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex and age (yr)</th>
<th>Relation of great arteries</th>
<th>Aortic arch</th>
<th>Pulmonary stenosis*</th>
<th>Atrial septum†</th>
<th>Splenic characteristic</th>
<th>Left superior vena cava</th>
<th>Pulmonary venous connection</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M, Newborn</td>
<td>Side by side</td>
<td>Right</td>
<td>Yes (s)</td>
<td>Common atrium</td>
<td>Normal</td>
<td>Joining coronary sinus</td>
<td>Normal</td>
<td>Interrupted aortic arch, PDA, lt. coronary from pulmonary trunk</td>
</tr>
<tr>
<td>2</td>
<td>F, Newborn</td>
<td>Side by side</td>
<td>Left</td>
<td>Yes</td>
<td>Primum defect</td>
<td>Normal</td>
<td>Absent</td>
<td>Normal</td>
<td>Absent pulmonary valve, hypoplastic lt. ventricle VSD AV canal type, muscular VSD</td>
</tr>
<tr>
<td>3</td>
<td>F, 2</td>
<td>Side by side</td>
<td>Left</td>
<td>No</td>
<td>Intact atrial septum</td>
<td>Normal</td>
<td>Absent</td>
<td>Normal</td>
<td>Double-orifice mitral valve, endocardial fibroelastosis of lt. ventricle</td>
</tr>
<tr>
<td>4</td>
<td>M, 3</td>
<td>Side by side</td>
<td>Left</td>
<td>Yes</td>
<td>Common atrium</td>
<td>Normal</td>
<td>Absent</td>
<td>Normal</td>
<td>Hypoplastic lt. ventricle</td>
</tr>
<tr>
<td>5</td>
<td>M, 5</td>
<td>Side by side</td>
<td>Left</td>
<td>No</td>
<td>Common atrium</td>
<td>Normal</td>
<td>Joining lt. side of common atrium</td>
<td>Partial anomalous (RPV to RA)†</td>
<td>Normal</td>
</tr>
<tr>
<td>6</td>
<td>F, 7</td>
<td>Side by side</td>
<td>Left</td>
<td>Yes</td>
<td>Common atrium</td>
<td>Normal</td>
<td>Absent</td>
<td>Normal</td>
<td>Hypoplastic Lt. ventricle, LAD coronary artery from rt. coronary artery</td>
</tr>
<tr>
<td>7</td>
<td>F, Newborn</td>
<td>Side by side</td>
<td>Right</td>
<td>Yes (s)</td>
<td>Common atrium</td>
<td>Asplenia</td>
<td>Joining lt. side of common atrium</td>
<td>Total anomalous to rt. atrium</td>
<td>Hypoplastic Lt. atrium, PDA</td>
</tr>
<tr>
<td>8</td>
<td>M, 3</td>
<td>Side by side</td>
<td>Left</td>
<td>No (s)</td>
<td>Primum defect</td>
<td>Asplenia</td>
<td>Joining coronary sinus</td>
<td>Normal</td>
<td>Tubular hypoplastic aortic arch, PDA, parachute mitral valve</td>
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<td>F, 5</td>
<td>Side by side</td>
<td>Left</td>
<td>Yes</td>
<td>Common atrium</td>
<td>Asplenia</td>
<td>Joining lt. side of common atrium</td>
<td>Total anomalous to rt. atrium</td>
<td>Absent rt. superior vena cava, IVC to rt. atrium, hepatic vein to lt. atrium, right coronary artery from LAD, muscular VSD</td>
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<tr>
<td>10</td>
<td>F, 8</td>
<td>Side by side</td>
<td>Left</td>
<td>Yes</td>
<td>Common atrium</td>
<td>Polyspleenia</td>
<td>Joining lt. side of common atrium</td>
<td>Normal</td>
<td>IVC to lt. superior vena cava viaazygos vein, hepatic vein to rt. atrium</td>
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<tr>
<td>11</td>
<td>M, 25</td>
<td>L-MGA§</td>
<td>Left</td>
<td>Yes</td>
<td>Intact atrial septum</td>
<td>Normal</td>
<td>Absent</td>
<td>Normal</td>
<td>VSD AV canal type, Ebstein deformity of lt. AV valve with double orifice</td>
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<tr>
<td>12</td>
<td>M, Newborn</td>
<td>L-MGA§</td>
<td>Left</td>
<td>Yes</td>
<td>Primum defect</td>
<td>Asplenia</td>
<td>Joining lt. side of common atrium</td>
<td>Total anomalous to rt. superior vena cava</td>
<td>Normal</td>
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<tr>
<td>13</td>
<td>M, 2/12</td>
<td>L-MGA§</td>
<td>Left</td>
<td>Yes</td>
<td>Primum defect</td>
<td>Asplenia</td>
<td>Absent</td>
<td>Total anomalous to portal vein</td>
<td>Normal</td>
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<tr>
<td>14</td>
<td>M, 5/12</td>
<td>L-MGA§</td>
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<td>Yes</td>
<td>Common atrium</td>
<td>Asplenia</td>
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<td>Normal</td>
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<tr>
<td>15</td>
<td>M, 14</td>
<td>D-MGA§</td>
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<td>Yes</td>
<td>Common atrium</td>
<td>Asplenia</td>
<td>Joining lt. side of common atrium</td>
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<td>Normal</td>
</tr>
<tr>
<td>16</td>
<td>F, 10</td>
<td>L-MGA§</td>
<td>Left</td>
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<td>Common atrium</td>
<td>Polyspleenia</td>
<td>Joining coronary sinus</td>
<td>Partial anomalous (RPV to RA)†</td>
<td>IVC to rt. superior vena cava viaazygos vein</td>
</tr>
</tbody>
</table>

* (s), subaortic stenosis present.
†All had common atroventricular canal except two with intact atrial septum.
‡RPV to RA, right pulmonary vein joining right atrium or right side of common atrium.
§L-MGA, levomalposition of aorta; D-MGA, dextroratmalposition of aorta.
DOUBLE-OUTLET RV AND COMMON A-V CANAL

Figure 1

Case 3. Left) View from right ventricle (RV) demonstrates both aorta (Ao) and pulmonary trunk (PT) arising from right ventricle. Muscular spur (S) (crista) is seen between these two great arteries. Two probes are in the ventricular septal defects (D), upper probe is in ventricular septal defect, persistent common atrioventricular type, and lower probe is in muscular ventricular septal defect. Right) View from left atrium (LA) and left ventricle (LV) demonstrates intact atrial septum. There is a cleft (between arrows) of anterior leaflet of mitral valve (MV).

Figure 2

Case 4. Left) View from right ventricle (RV) demonstrates both aorta (Ao) and stenotic pulmonary trunk (with small probe) arising from right ventricle. Tricuspid valve (TV) is part of common atrioventricular valve, and ventricular septal defect (D) is only outlet of left ventricle. Right) View from right atrium (RA) and right ventricle (RV) demonstrates common atrioventricular canal and undivided common anterior leaflet (A) and undivided common posterior leaflet (P) of common atrioventricular valve. Upper arrow points to small additional atrial septal defect of secundum type. Below is defect of lowermost portion of atrial septum (primum defect). Vertical arrow demonstrates double orifice of mitral valve.

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ventricular septal defect in addition to the ventricular septal defect of the atrioventricular canal.

The ages of the 16 patients (nine males and seven females) ranged from newborn to 25 years; the mean age was 5.1 years.

Seven patients had normal spleens (table 2). Splenic anomalies were common, being observed in nine patients: asplenia in seven and polysplenia in two. Of the ten patients with levocardia (group 1), six had normal spleens, three had asplenia, and one had polysplenia. Of the six patients with dextrocardia (group 2), one had a normal spleen, four had asplenia, and one had polysplenia.

In each patient, the aorta and pulmonary trunk arose completely from the anatomic right ventricle. In each of the ten patients with levocardia, the aorta lay to the right of the pulmonary trunk in a side-by-side relationship as is commonly seen in double-outlet right ventricle. In five of the six patients with dextrocardia and inversion of ventricles, the aorta lay to the left and anterior to the pulmonary trunk (levomalposition, L-MGA). In the one other patient, the aorta was to the right and anterior to the pulmonary trunk (dextromalposition, D-MGA).

Associated Anomalies

Obstruction to Pulmonary and Systemic Flow

Obstruction to pulmonary flow was observed in 12 patients, including six of the seven with asplenia, the obstruction being represented by either valvular or subpulmonary stenosis. Three patients had subaortic stenosis; two of these also had pulmonary stenosis.

Venous Anomalies

Systemic and pulmonary venous anomalies were common (table 2).

Persistent left superior vena cava was present in ten of the 16 patients. In three of the ten, the vena cava joined the coronary sinus. In seven, the left superior vena cava terminated in the left side of the common atrium. In these seven patients, the coronary sinus was absent.

Anomalous pulmonary venous connection was observed in six of the 16 patients. In four of the six patients, the total form was present; in each of the four, asplenia was present. Of these four patients, two had the total anomalous pulmonary venous connection to the right atrium, one to the right superior vena cava, and one to the portal vein.

Two patients had partial anomalous pulmonary venous connection. In each patient, the right pulmonary veins joined the right atrium while the left pulmonary veins joined the left atrium or the left side of a common atrium.

Ten patients had common atrium (this included the seven patients in whom the left superior vena cava joined the left side of the common atrium).

Twelve patients had the aortic arch on the left side, and three on the right side. In one, the position was not determined. In addition, one patient had interrup-
DOUBLE-OUTLET RV AND COMMON A-V CANAL

Table 2

Relationships of Various Factors to Condition of Spleen in 16 Cases of Double-Outlet Right Ventricle with Atrioventricular Canal

<table>
<thead>
<tr>
<th>No. of patients</th>
<th>Splenic characteristics</th>
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<tr>
<td></td>
<td>Normal</td>
</tr>
<tr>
<td>Distribution of patients</td>
<td>16</td>
</tr>
<tr>
<td>Ages (yr)</td>
<td>Newborn, Newborn, 2, Newborn, Newborn</td>
</tr>
<tr>
<td>Cardiac apex</td>
<td>Levocardi</td>
</tr>
<tr>
<td></td>
<td>Dextrocardia</td>
</tr>
<tr>
<td>Relation of great arteries</td>
<td>Side by side</td>
</tr>
<tr>
<td></td>
<td>D-MGA*</td>
</tr>
<tr>
<td></td>
<td>L-MGA*</td>
</tr>
<tr>
<td>Obstruction to pulmonary flow</td>
<td>12</td>
</tr>
<tr>
<td>Subaortic stenosis</td>
<td>3</td>
</tr>
<tr>
<td>Systemic venous anomalies</td>
<td>Persistent L SVC†</td>
</tr>
<tr>
<td>Anomalous pulmonary venous connection</td>
<td>Total</td>
</tr>
<tr>
<td></td>
<td>Partial</td>
</tr>
<tr>
<td></td>
<td>Common atrium</td>
</tr>
<tr>
<td></td>
<td>Aortic arch</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Coronary arterial anomalies</td>
</tr>
</tbody>
</table>

*D-MGA, dextromalposition of great arteries; L-MGA, levomalposition of great arteries.
†Left superior vena cava.

...tion of the arch and another had tubular hypoplasia of the arch. This latter patient also had a parachute mitral valve as part of Shone's syndrome.12

Coronary Arterial Anomalies

Four patients had coronary arterial anomalies. Three had normal spleens and one had asplenia. The left anterior descending coronary artery arose from the pulmonary trunk in one patient and from the right coronary artery in another. In each of two additional patients, a single coronary artery gave origin to all branches.

Clinical Findings

The predominant clinical findings depended on the presence or absence of pulmonary stenosis. When pulmonary stenosis was present, the clinical presentation was like that in tetralogy of Fallot. Each patient had cyanosis of various degrees, but squatting and anoxic spells were infrequent. When pulmonary stenosis was absent, the presentation was similar to that in ventricular septal defect with bidirectional shunting. All patients who did not have pulmonary stenosis had congestive heart failure and minimal cyanosis.

Electrocardiographic Findings

Electrocardiograms were available for six patients with levocardia. Superior QRS axis deviation in the frontal plane was found in each of five patients with the complete form of atroventricular canal who had electrocardiograms available for study; the QRS frontal plane loop was directed counterclockwise and superiorly. One patient (case 3) who had an intact atrial septum and a ventricular septal defect of the persistent common atrioventricular canal type had right axis deviation and a clockwise QRS frontal plane loop inferiorly oriented.

One patient with dextrocardia and common atroventricular canal had a clockwise superior QRS loop in the frontal plane. Another patient (case 11) with dextrocardia and an intact atrial septum with a ventricular septal defect of the persistent common atrioventricular canal type had right axis deviation and a clockwise inferior QRS frontal plane loop.

Roentgenographic Findings

The thoracic roentgenogram revealed mild-to-moderate cardiomegaly. The pulmonary vascularity was decreased or increased, depending on the presence or absence of pulmonary stenosis, and this vascularity also reflected the volume of left-to-right shunt. There was no specific diagnostic feature referable to double-outlet right ventricle or atrioventricular canal.
Angiographic Findings

Angiocardiograms from seven patients were available for review, five from patients with levocardia (group 1) and two with dextrocardia (group 2). In group 1, the right ventricular angiocardiogram had features typical of double-outlet right ventricle. Both great arteries arose from the morphologic right ventricle in a side-by-side relationship in which the aorta was to the right of the pulmonary trunk (figs. 4 and 5). The left ventricular angiocardiogram revealed a deformity of the left ventricular outflow tract produced by the abnormal insertion of the left atrioventricular valve and the ventricular septal defect. The pattern is suggestive of that seen in ordinary persistent common atrioventricular canal (figs. 4, 5 and 6).

In group 2, dextrocardia with ventricular inversion, the angiocardiograms from the left-sided, morphologic right ventricle demonstrated that both great arteries had their origins from this ventricle. The angiocardiogram from the right-sided, morphologic left ventricle revealed the ventricular septal defect, but an angiographic diagnosis of atrioventricular canal could not be made (fig. 7).

Surgical Results

This complex cardiac anomaly has not yet been successfully repaired. Eight of the 16 patients un-
Case 6. Top panels) Right ventricular angiocardiograms in frontal (left) and lateral (right) views demonstrate both great arteries arising from morphologic right ventricle (RV). Left ventricle (LV) and ventricular septal defect (VSD) are opacified. Bottom panels) Left ventricular angiocardiograms in frontal (left) and lateral (right) views. Frontal view demonstrates deformity of left ventricular (LV) outflow tract caused by abnormal insertion of left atrioventricular valve and deficient ventricular septum. Left atrioventricular valve (arrow) also demonstrates discontinuity with semilunar valves.

Three patients underwent unsuccessful operation at other institutions. In none of the three cases was the diagnosis of double-outlet right ventricle or persistent common atrioventricular canal made clinically or at operation. In one case, there was an associated infradiaphragmatic complete anomalous pulmonary venous connection; correction of this anomalous connection was attempted. In the second case, the clinical
Discussion

Double-outlet right ventricle associated with atrioventricular canal is a rare cardiac anomaly. Its presence can be suspected before operation in patients who have double-outlet right ventricle either with levocardia or with dextrocardia and ventricular inversion. If there is an associated splenic anomaly, the condition is especially likely to be found. In our series, however, six patients who had levocardia did not have a splenic anomaly. In addition, the electrocardiogram and vectorcardiogram in five of six patients with levocardia had the characteristic features of atrioventricular canal, in which the QRS frontal plane loop is directed counterclockwise and superiorly. Krongrad et al.\textsuperscript{15} recognized, however, that five of 31 patients with double-outlet right ventricle without associated atrioventricular canal had counterclockwise superiorly directed frontal plane QRS loops. The selective left-sided left ventricular angiogram reveals a deformity in the left ventricular outflow tract produced by the abnormal insertion of the left atrioventricular valve and the ventricular septal defect, and the angiographic pattern is suggestive of that seen in ordinary persistent atrioventricular canal. Patients with ventricular inversion and dextrocardia, however, had no angiographic features suggestive of an associated atrioventricular canal.

Clinical and hemodynamic features are dependent on the presence or absence of pulmonary stenosis. When there is pulmonary stenosis, the clinical presentation is similar to that in tetralogy of Fallot, and when there is no pulmonary stenosis, the presentation is similar to that in ventricular septal defect with bidirectional shunting.

The ventricular septal defect of common atrioventricular canal in double-outlet right ventricle is remote from both semilunar valves. This complicates closure of the ventricular defect in a tunnel fashion, which would direct blood from the left ventricle into the aorta.\textsuperscript{16-19} The lack of success in the surgical treatment of this condition is noteworthy; however, the present report is weighed with postmortem material selectively, and increasing success with other forms of highly complex anomalies offers promise that these associated anomalies will be correctable. One of various approaches ultimately may be feasible. For ex-

Figure 6

Case 5. Left ventricular angiogram. Frontal (left) view demonstrates deformity of left ventricular outflow tract. Lateral (right) view demonstrates hypoplastic left ventricle (LV) and large common atrioventricular valve (CAVV). A, common atrium.
Case 15. Dextrocardia, asplenia, common atrium, and ventricular inversion. Top panels) Angiocardiograms from inverted left-sided morphologic right ventricle, frontal (left) and lateral (right) views. Both great arteries arise from this ventricle. Great arteries are dextrotransposed, with aorta (AO) lying directly anterior to pulmonary trunk. There is valvar and subvalvar pulmonary stenosis. Catheter passed through right superior vena cava, common atrium, and left atrioventricular valve into morphologic right ventricle (RV). Bottom panels) Angiocardiograms from inverted right-sided morphologic left ventricle, frontal (left) and lateral (right) views. Frontal view demonstrates ventricular septal defect (VSD) as only outlet of left ventricle (LV). There is severe atrioventricular valve regurgitation into large common atrium (A). Aorta (AO) and pulmonary artery opacified via ventricular septal defect.

Example, the ventricular septal defect could be enlarged and standard repairs for double-outlet right ventricle and for complete atrioventricular canal could be combined; or the standard repair for complete atrioventricular canal could be modified to accomplish intratrial transposition of venous return (Mustard operation), combined with insertion of an external conduit from the left (noninverted) ventricle to the distal end of the divided pulmonary artery; or in inversion of ventricles, a simpler repair may be possible by accomplishing a standard repair of the atrioventricular canal, combined with insertion of an external conduit from the right-sided left ventricle to the distal end of the divided pulmonary artery.
References

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