
The Morphologic Spectrum of Double Outlet Left Ventricle and Its Surgical Significance

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SUMMARY Double outlet left ventricle (DOLV) is defined in this paper as that condition in which both great arterial vessels emerge completely (complete form) or mostly (incomplete form) from the morphologic left ventricle. This is a study of eight cases diagnosed during surgery and 37 cases diagnosed at autopsy. DOLV was classified as follows: 1) with the aorta to the right, or to the right and posterior to the pulmonary trunk: a) with ventricular septal defect (VSD) confluent with the aorta (10 cases), b) with VSD confluent with the pulmonary trunk (three cases); and c) with VSD confluent with both vessels (two cases), 2) with aorta anterior to, anterior and to the right, or anterior and to the left of the pulmonary trunk: a) with the VSD related to the aorta (10 cases), b) with the VSD related to the pulmonary trunk (one case), and c) with the VSD related to both vessels (two cases). 3) with tricuspid valve abnormalities: a) with tricuspid atresia and stenosis with normally related vessels (four cases), or aorta anterior (10 cases), b) with straddling or displaced tricuspid valve (two cases), or c) with Ebstein's anomaly (one case). Five surgical and four autopsyed cases were of the complete form. From the surgical standpoint, it is important in DOLV to determine the interrelationships and origin of the great arteries, their relationship to the VSD, and the location of the VSD in the ventricular septum. The presence of tricuspid valve abnormalities is emphasized.

DOUBLE OUTLET LEFT VENTRICLE (DOLV) is a condition in which both arterial trunks emerge completely or mostly from the morphologically left ventricle. This heart defect has been known to exist for many years, but has been considered a rarity. In recent years patients with this malformation have been treated surgically, but few pathologic studies have been reported.

We undertook combined pathologic and surgical study of DOLV to further elucidate the anatomic facts which might be surgically important.

Materials and Methods

This study consisted of a gross pathologic examination of 24 hearts from the Congenital Heart Disease Research and Training Center and 13 hearts from the Armed Forces Institute of Pathology. The study also included a review of the morphology of the eight cases (seven of whom survived operation and are doing well) operated on from 1967–1978 at the University of Alabama Medical Center, Birmingham, Alabama. Only hearts with atrioventricular (AV) concordant connections were included in the study.

The atria and viscera were in situ solitus in each case. In the pathologic material all hearts were in levocardia. Two of the surgical cases had dextroversion.

The presence or absence of tricuspid valve anomalies was determined for each heart. The abnormalities included tricuspid stenosis, tricuspid atresia, displaced tricuspid orifice and Ebstein's anomaly. In displaced tricuspid orifice, both the mitral and tricuspid orifices enter the left ventricle completely,
but there is a distinct right ventricle with a sinus and infundibulum. The 45 cases were considered in two groups, those with and those without tricuspid valve abnormalities.

The hearts were also analyzed according to the position and interrelations of the great arteries. Two groups of cases emerged. In one, the aorta was to the right and at the same level as, or posterior and to the right of, the pulmonary artery in the coronal plane. This is the normal or near-normal interrelation of these vessels. In the second group, the aorta was distinctly anterior to the pulmonary artery either directly so, or to the right and anterior, or to the left and anterior. This group includes the positions of the great arteries that can be seen in simple, ordinary (d-loop) transposition of the great arteries.

The location in the septum of the ventricular septal defect (VSD) was determined. In addition to its location in the anterior or posterior septum, the points of reference were 1) the position of the VSD related to the efferent vessels, and 2) the relationship of the VSD to the tricuspid and mitral orifices and valves. There may be a defect beneath, or close to either vessel or both vessels in DOLV, which would be anterior (superior-cranial) to the tricuspid and mitral valves and might or might not be related to these valves. By related, we mean that the defect is close enough to either of these AV orifices, so that there is no, or negligible, muscle between the defect and the orifice; or that any of such defects might be separated from the tricuspid or mitral valves by muscle. This is important from the standpoint of the precise relation of the course of the conduction system to the edges of the VSD.

Results

The anatomic characteristics in relation to the position of vessels and the location of VSD of the 45 cases are summarized in table 1.

The characteristics of the important subsets without tricuspid valve abnormalities are as follows:

With the aorta directly to the right or to the right and posterior to the pulmonary trunk with the VSD related to the aorta (fig. 1). There were seven autopsied and three surgical cases. In the pathologic material, the pulmonary trunk emerged completely from the left ventricle. The aorta overrode the VSD with about one-and-one-half aortic cusps in each ventricle (that is, the aorta emerged about 50% from the left ventricle), or in some cases with a greater proportion in left ventricle. In the three surgical cases both vessels emerged completely from the left ventricle. In all 10 cases the pulmonary valve was stenotic and lay anterior (superior-cranial) to and removed from the VSD. In all the pathologic cases there was aortic-mitral continuity in and some cases tenuous mitral-pulmonic continuity. The position of the VSD was as follows: As seen from the left side, the VSD faced the aortic orifice in an oblique manner and the defect was situated posteriorly adjacent to the mitral valve. There was very little if any pars membranacea detected grossly. In four cases in the pathologic material, the VSD was adjacent to the tricuspid valve. In two, a small amount of muscle separated the defect from the tricuspid valve, and in one there was a considerable amount of muscle between the two. In two of the surgical cases there was a slight muscle separation between the VSD and the tricuspid valve and in one no

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**Table 1. Anatomic Characteristics of the Heart of 45 Cases of Double Outlet Left Ventricle with Concordant Atroventricular Connection**

<table>
<thead>
<tr>
<th>Position of Great Arteries</th>
<th>Subaortic Related to tricuspid</th>
<th>Subpulmonic Related to tricuspid</th>
<th>Muscular septum unrelated to either vessel</th>
<th>No VSD</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Separated from the tricuspid by muscle or absent tricuspid</td>
<td>Separated from the tricuspid by muscle</td>
<td>Separated from the tricuspid by muscle</td>
<td>Separated from the tricuspid valve</td>
</tr>
<tr>
<td>Without tricuspid valve abnormalities</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aorta to right of PT</td>
<td>15 cases</td>
<td>5</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Aorta anterior to PT (directly, or anterior to right, or anterior and to the left)</td>
<td>13 cases</td>
<td>6</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>With tricuspid valve abnormalities</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aorta to right of PT</td>
<td>4 cases</td>
<td>0</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Aorta anterior to PT (directly, or anterior and to right, or anterior and to the left)</td>
<td>13 cases</td>
<td>2</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>45 cases</td>
<td>13</td>
<td>19</td>
<td>2</td>
</tr>
</tbody>
</table>

Abbreviations: PT = pulmonary trunk; VSD = ventricular septal defect.
The architecture of the musculature of the right ventricle was as follows: The parietal band in all cases but one did not form an arch with the septal band, but it was anchored on the right side of the septal band adjacent to the VSD. The septal band in these cases consisted of a typical Y-shaped structure as seen in truncus arteriosus communis and pulmonary atresia with VSD. In the one exceptional case a slightly excavated arch was formed between the septal and parietal bands. Associated cardiac abnormalities in this group were bicuspid pulmonic valve (two cases), unicuspid pulmonic valve (one case) and subpulmonic stenosis (one case).

With the aorta to the right and posterior to the pulmonary trunk with the VSD confluent with the pulmonary trunk (fig. 2). There were three cases of which two were obtained at autopsy. The one surgical case of this type was the only death among the surgical cases. In all three cases the aorta emerged completely from the left ventricle related to the mitral valve, while the pulmonary trunk overrode the septum over the VSD. There was no pulmonary stenosis in any case. The defect was present in the anterior septum. It excavated the arch (crista) formed by the septal and parietal bands and was anterior (cranial) to the mitral valve and the pars membranacea. A small amount of muscle in one case, and a considerable amount in the other two cases, separated the tricuspid valve from the defect. A small part of the parietal band separated the two arterial trunks. Associated cardiac abnormalities in this group were interrupted aortic arch (one case), bicuspid aortic valve (one case) and aortic stenosis (one case).

With the aorta to the right and posterior to the pulmonary trunk with the VSD related to both arterial trunks (fig. 3). There were two autopsied cases. Here the aorta emerged completely or almost completely from the left ventricle while the pulmonary trunk emerged partially from the left ventricle. The VSD was anterior and separated from the mitral valve and
Figure 2. Double outlet left ventricle with arterial trunks in relatively normal position with the pulmonary trunk related to the ventricular septal defect. Left ventricular view. A = aorta; PT = pulmonary trunk; VSD = ventricular septal defect; MV = mitral valve.

Figure 3. Double outlet left ventricle with arterial trunks in relatively normal position with both vessels related to the ventricular septal defect. A) Right ventricular view; B) left ventricular view. D = ventricular septal defect; PT = pulmonary trunk; P = parietal band; S = septal band; TV = tricuspid valve; A = aorta; MV = mitral valve.

Associated cardiac abnormalities in this group were absent transverse arch (one case), bicuspid aortic valve (one case) and aortic stenosis (one case).

With the aorta anterior and to the right, directly anterior, or anterior and to the left of the pulmonary trunk, with the VSD confluent with aorta (fig. 4). There were eight autopsied and two surgical cases. Here the pulmonary trunk emerged completely from the left ventricle with the aorta emerging mostly from the left ventricle. The aorta overrode the septum over the VSD often with a straddling conus. The aorta was anterior and to the left of the pulmonary trunk in seven, directly anterior, or anterior and slightly to the right in three, and in one the vessels were side by side with the aorta to the left. Mitral-pulmonic continuity was present in six of the autopsied cases, and both mitral-pulmonic and mitral-aortic continuity in two. Nevertheless, there was a distinct infundibulum for the pulmonary trunk. The orifice of the latter was stenotic in six cases, and situated posteriorly or postero-distally (downstream) to the VSD. The ventricular septal defect was anterior to the tricuspid valve and separated from it by no, or a varying amount of, muscle. It was thus related to the tricuspid valve in five cases, and related to both tricuspid and mitral valves in one. It was not related to either AV valve in the remainder, and was anterior to both. We noted the parietal band proceeding into the left ventricle in six autopsied cases so that an arch was not formed by the septal and parietal bands. In two cases a partial arch was formed. The septal band was Y- or U-shaped, and held the VSD in its arms. One surgical case was in dextroversion (situs solitus atria and viscera and d-loop). Associated cardiac abnormalities in this group were bicuspid pulmonic valve (five cases), patent ductus arteriosus (PDA) (one case), and juxtaposition of the atrial appendages (one case).
With the aorta anterior and to the right of the pulmonary trunk with VSD confluent with the pulmonary trunk. There was one case in this group. The aorta overrode the septum over a straddling conus, while the pulmonary trunk arose from the left ventricle. Pulmonary stenosis was present. The VSD lay anterior and adjacent to the tricuspid valve and removed by muscle from the mitral valve. There was a bicuspid pulmonic valve.

With the aorta anterior and to the right of the pulmonary trunk with VSD confluent with both vessels. There were two autopsied cases. These cases were identical with the group with the VSD confluent with the aorta, but there was no muscle separating the aorta and the pulmonary trunk. The aorta was to the right and anterior and the pulmonary trunk to the left and posterior in both. Both were associated with pulmonary stenosis. In both cases the VSD was in the anterior septum. It was related to the tricuspid valve and separated by muscle from the mitral valve. An associated cardiac abnormality in this group was bicuspid pulmonic valve (one case).

The characteristics of the subsets with tricuspid valve abnormalities are as follows:

With tricuspid stenosis or atresia with the aorta to the right and posterior to the pulmonary trunk.* There were four hearts, two with tricuspid stenosis. In all cases the aorta was to the right and posterior and the pulmonary trunk to the left and anterior. In those with atresia, the aorta came off completely from the left ventricle unrelated to the defect with aortic-mitral continuity. The pulmonary trunk overrode the septum over the VSD emerging about 50% from each ventricle. There was an overriding conus, with the parietal band proceeding into the left ventricle. The VSD was large and anterior. It was anterior to and unrelated to the mitral valve.

In one case with tricuspid stenosis the aorta emerged mostly from the left ventricle with a straddling conus. The pulmonary trunk overrode the septum coming off the left ventricle by 50–60%. Here again, as in tricuspid atresia, there was an overriding conus with the parietal band proceeding into the left ventricle. The VSD was separated from the tricuspid and mitral valves by considerable muscle. It was related to both the aorta and pulmonary trunk. In the other case with tricuspid stenosis, there was no VSD.† Both the aorta and the pulmonary trunk emerged completely from the left ventricle. The aorta and the pulmonary trunk were side by side with the aorta to the right of the pulmonary trunk. Both vessels were related to the mitral valve.

Associated cardiac abnormalities in this group with tricuspid stenosis and atresia were fetal coarctation with PDA (two cases), subaortic and aortic stenosis (one case), atrial septal defect (one case), left superior vena cava entering the coronary sinus (one case) and single coronary artery (one case).

With tricuspid stenosis or atresia with the aorta anterior, or anterior and to the right, or anterior and to the left of the pulmonary trunk.‡ There were eight autopsied (seven with tricuspid atresia and one with tricuspid stenosis) and two surgical cases. The aorta was anterior and to the left and the pulmonary trunk posterior and to the right in all of the autopsied cases. In two surgical cases the aorta was anterior and to the right and the pulmonary trunk posterior and to the left. In both surgical cases both vessels emerged completely from left ventricle. In the eight autopsied cases the aorta straddled the ventricular septum over a defect. Six autopsied cases were with pulmonary stenosis and two without pulmonary stenosis. One surgical case was in dextroversion (situs solitus atria and viscera) with mild pulmonary stenosis, small stenotic tricuspid valve, and small right ventricle. In addition, there was a muscular VSD. The other surgical case had severe pulmonary stenosis with an abnormal attachment of the tricuspid valve chordae in the right ventricle. The defect was usually anterior, separated

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*Pictures of these hearts have been previously published.*
†This case was previously reported.†
‡Pictures of these hearts have been previously published.*
FIGURE 5. Double outlet left ventricle, complete type with Ebstein's anomaly. A) Right atrial and right ventricular view; B) view of distal chamber of right ventricle; C) view of separate infundibulum beneath the aorta leading into the left ventricle. D) left ventricular view. L = limbus fossae ovalis; TV = downward displacement of the tricuspid valve; RVP = proximal chamber of right ventricle; MV = mitral valve; VSD = ventricular septal defect; A = aorta; PT = pulmonary trunk; E = entry of infundibulum beneath aorta into left ventricle; RVD = distal chamber of right ventricle; I = infundibulum beneath aorta; AD = anterior descending coronary artery.
by muscle from the mitral valve. Where the defect extended more posteriorly, there was a straddling conus with the parietal band passing into the left ventricle. Thus the aorta emerged from both ventricles but came out mostly from the left, while the pulmonary trunk arose from the left ventricle related to the mitral valve. In the case with tricuspid stenosis, the defect was not related to the tricuspid but related to the mitral valve. Associated cardiac abnormalities were bicuspid pulmonic valve (one case), subpulmonic stenosis (one case), juxtaposed atrial appendages (one case) and an additional VSD (one case) located in the muscular septum towards the apex.

With displaced tricuspid orifice, with the aorta anterior, or anterior and to the left of the pulmonary trunk. There were two autopsied hearts. The aorta straddled the ventricular septum over the defect. The pulmonary trunk emerged from the left ventricle related to the mitral valve. Pulmonary stenosis was present in one. The VSD was large and anterior. It was related to the tricuspid valve in one case, but not related to this valve in the other. Associated cardiac abnormalities were: PDA (two cases), subaortic stenosis (one case), cleft aortic leaflet of the mitral (one case), fetal coarctation (one case) and subaortic stenosis (one case).

With Ebstein's anomaly. There was one autopsied case (fig. 3).* In this remarkable case the aorta emerged completely from the left ventricle over a well developed conus. The pulmonary trunk overrode the septum over a posteriorly placed VSD, but emerged mostly from the left ventricle. The aorta was anterior and to the right of the pulmonary trunk and the latter was related to the mitral valve. The VSD was related to the tricuspid but not the mitral valve. This case was associated with fetal coarctation and PDA.

Discussion

These 45 cases have been analyzed with particular attention to features which may be important surgically, and we believe the findings complement previous anatomic studies.1, 2, 7, 8 Eight of the 45 cases have been operated upon at the University of Alabama Medical Center, Birmingham, Alabama. Seven of these patients survived operation and are well. Four of these have been reported previously.9, 10 The present analysis indicates the great variability of DOLV and the need to evaluate each aspect of the malformation in individual patients.

The surgical problem appears simple, since the aorta presumably already is connected to the left ventricle completely or in part. Thus, the VSD is repaired, and then the pathway from left ventricle to pulmonary artery is closed off and a new pathway from the right ventricle to pulmonary artery is constructed. Usually this is accomplished by placing a valved external conduit from the right ventricle to pulmonary artery. However, detailed knowledge of each aspect of the malformation is necessary.

Patients believed to have DOLV must be carefully investigated for possible associated tricuspid valve abnormalities. These may be associated with underdevelopment of the right ventricle.9, 10 Their presence may complicate the surgical procedure, or sometimes make it impossible.11, 12 In the preoperative study, particular attention should be devoted to the possibility of straddling tricuspid valve, which, if present, complicates the repair.13

Most patients with DOLV are cyanotic, either because of coexisting pulmonary stenosis or because the relations of the VSD and the great arteries result in the pulmonary and systemic circulations being mostly in parallel, as in ordinary transposition of the great arteries. Some patients with DOLV with the aorta to the right of the pulmonary trunk may seem to have tetralogy of Fallot; a correct diagnosis can be made only by angiocardiography with special views or at operation. Other patients with DOLV and anterior aorta may seem to have transposition of the great arteries with VSD, but a special study or operation shows that not only does the pulmonary artery arise completely from left ventricle, but the aorta also arises completely or in large part from this chamber.

This study supports the idea that often one and sometimes both great arteries override DOLV and therefore originate in part from the right ventricle. The basic problems of diagnosis and treatment are no different when this occurs than when both great arteries arise solely from the left ventricle. The details of these arrangements are, however, of utmost surgical importance, particularly in planning and executing the repair of the VSD so that subaortic stenosis does not result.

The localization of the defect in the ventricular septum is also important to the surgeon.13 All the VSDs which we have described, with the exception of one additional defect further apicalward in one case, lie anterior to the junction of the medial and anterior leaflets of the tricuspid valve. However, they differ as to whether muscle separates the defect from the tricuspid valve. If there is no significant muscle separation, the defect lies more or less against the tricuspid valve ring. It may or may not be separated from the mitral valve and pars membranacea, if present, by muscle. The bundle of His may then lie along the left side of the inferior margin of the defect as in the usual "perimembranous type" of VSD, or somewhat more posterior to this rim on the left side of the septum as in complete transposition.14 In making a repair in such cases, stitches must be placed well back from the inferior margin of the defect and on the right ventricular side. If there is a significant amount of muscle between the defect and the tricuspid valve, the bundle of His is usually not at the inferior margin of the defect, but posterior to this. Thus, we close such a defect by placing the stitches right along the edge of the defect. These suppositions are supported by the fact that heart block has not occurred in our operated

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*This case has previously been diagrammatically presented and briefly discussed.1
cases of DOLV. Since DOLV does not require that a VSD be beneath the efferent vessels, we expect that a VSD in every possible position will probably sometime be seen. Complete AV canal with DOLV is theoretically possible. The absence of a large VSD beneath the efferent vessels does not pose the same problem as it does in double outlet right ventricle with AV concordant connection because in DOLV, the repair of the VSD does not also have to construct a pathway for left ventricular-aortic blood flow. Furthermore, a right ventricular-pulmonary artery pathway is easily provided by closure of the orifice of the pulmonary artery and insertion of a valved external conduit. Thus the VSD repair does not have to be done so as to provide for this pathway. Rarely, as in Sakakibara’s case, the location and size of the VSD and its relations to the great arteries allow repair of the VSD which also provide internally for right ventricular-pulmonary artery blood flow.

Just as in double outlet right ventricle, there are cases with mitral stenosis and atresia (25 of 133 cases), 15, 16 so DOLV is seen with tricuspid stenosis and atresia (17 of 45 cases). Thus, when mitral atresia or stenosis is present, there is a tendency in some cases for the posterior efferent vessel to move anteriorly and to the right. Where there is tricuspid atresia or stenosis there is a tendency for the anterior efferent vessel, whichever it is, to move posteriorly and to the left.

The complete form of DOLV is rare. Usually there is an incomplete form with one or both vessels overriding. In general, there is a spectrum of positional variations of the efferent vessels, including tetralogy of Fallot, double outlet right ventricle and complete regular (d-) transposition. Within this spectrum lies DOLV. Speculatively, all these may be subsumed under the concept of abnormal absorption of the bulbus embryologically, and its concomitant abnormal right deviation of the atrial canal. 17

Finally, in all our cases the atria and viscera were in situs solitus. Others 4, 12 have reported patients with situs inversus totalis and concordant AV connections.

References

The morphologic spectrum of double outlet left ventricle and its surgical significance.
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