Double-outlet right ventricle

A collective review with a surgical viewpoint

Double-outlet right ventricle (DORV) should be classified according to its various subsets. Van Praagh's symbolic terminology provides an appropriate mechanism for doing that. Each of these subsets is described in terms of its pathology, clinical characteristics, cardiac catheterization findings, and operative repair. The operative repair for each is different. The presence or absence of continuity between the semilunar and atrioventricular valves is unimportant from the standpoint of operation and should not be a criterion for classification.

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The classic definition of double-outlet right ventricle (DORV) requires that (1) both the aorta and pulmonary artery take origin from the right ventricle (RV), (2) a ventricular septal defect (VSD) provide the only outlet from the left ventricle (LV), and (3) the semilunar valves be separated from the atrioventricular valves by conal tissue. This latter requirement permits a precise anatomic distinction between such entities as tetralogy of Fallot with severe dextroposition of the aorta and DORV with subaortic VSD and pulmonic stenosis or between d-transposition of the great vessels with VSD and an overriding pulmonary artery and DORV with subpulmonic VSD. The exactness of this definition excludes some hearts which should otherwise be classified as DORV. Neufeld and others have pointed out that semilunar-atrioventricular valve continuity occasionally may be preserved in DORV when the anterior leaflet of the mitral valve elongates and remains contiguous with the dextroposed, rotated aortic root. Lev believes that these conotruncal malformations represent a spectrum as one merges into another, rather than well-defined separate and distinct entities. Both he and Pacifico have suggested that the definition of DORV be more liberal. They would require only that one great vessel and the majority of the other (in relationship to the ventricular septum) originate from the RV. This concept is more useful to the surgeon since the type of operation done in each subset of DORV is dependent upon the relationship of the great vessels to the ventricular septum and VSD and not upon the presence or absence of conal tissue between the semilunar and atrioventricular valves.

Nomenclature

The nomenclature of DORV has suffered from a lack of standardization. This has produced considerable confusion in the literature. Van Praagh's symbolic terminology to describe segmental sets (situs, loop, and aortic position relative to the pulmonary artery), when combined with precise words to indicate the relationship of the VSD to the great vessels and the presence or absence of pulmonic stenosis, provides a very accurate and meaningful classification. Each subset of DORV will be described in these terms.

DORV (S,D,D), VSD related to aorta, PS absent

Pathology. The atria and ventricles are concordant and the aorta is to the right of the pulmonary artery (Fig. 1). The external appearance of the great vessels may be normal, or the aorta may be more to the right and anterior than normal. The pulmonary artery and most, if not all, of the aorta emerge from the RV chamber. The orifices of both vessels are usually in a side-by-side position in approximately the same coronal plane. The aortic orifice is to the right. The normal continuity between the aortic annulus and the anterior leaflet of the mitral valve is usually interrupted by the parietal band of the crista supraventricularis. Rarely, that continuity may be maintained by an elongated anterior leaflet. When this occurs,
that leaflet is usually more contiguous with the base of the left coronary cusp sinus rather than with the noncoronary cusp sinus. The VSD lies below the crista and is related to the aorta and remote from the pulmonary artery. The VSD is bound dorsally by the junction of the anterior leaflet of the mitral valve and the septal leaflet of the tricuspid valve. The caudad, ventral, and cephalad edges are bound by muscle. The VSD is usually large, but it may be small and restrictive to LV emptying. Associated anomalies include secundum atrial septal defect, persistent left superior vena cava, patent ductus arteriosus, absent pulmonary valve, coarctation, cor triatriatum, and Ebstein-like deformity of the mitral valve. With the exception of patent ductus arteriosus, coarctation, and VSD of the atroventricular canal type, other anomalies are extremely rare.

Clinical. These patients usually present in a manner similar to those with a large VSD and pulmonary artery hypertension. They have signs of increased pulmonary blood flow, left ventricular dysfunction, repeated respiratory infections, and failure to thrive. Cyanosis is rare. The chest x-ray film usually shows an enlarged heart and evidence of increased pulmonary blood flow. The electrocardiogram usually shows RV hypertrophy. There may also be LV hypertrophy and in some patients a superiorly oriented vector and counterclockwise loop in the frontal plane. Prolongation of the P-R interval and an intraventricular conduction delay are not common.

Cardiac catheterization. The hemodynamic data may be indistinguishable from that of a large VSD and pulmonary artery hypertension. The pressures in the RV and LV are equal unless the VSD is restrictive, in which case the LV pressure will exceed both the RV and aortic pressures. The pulmonary artery pressure is usually at systemic level. Pulmonary vascular resistance may range from normal to severely elevated. An oxygen saturation step-up is present in the RV. The oxygen saturation in the aorta exceeds that in the pulmonary artery because of differential streaming within the RV as blood is ejected from the LV across the VSD into the aorta. Angiographic visualization of the RV and LV is paramount to a correct preoperative diagnosis. The RV angiogram in the posteroanterior projection usually shows simultaneous visualization of the aorta and pulmonary artery. The aorta and pulmonary valves are often, but not always, on the same coronal plane. The aorta is displaced anteriorly to varying degrees. The discontinuity between the aortic annulus and anterior leaflet of the mitral valve is usually best visualized on a cineangiogram in the lateral projection. The presence of fibrous continuity between these structures, although rare, does not discount the possibility of DORV.

Operative management. Thirty patients have undergone complete correction of this subset and 20 have survived. The combined operative mortality rate for all published series is 27 per cent. Many of the operative deaths occurred more than 10 years ago and were avoidable. Some were related to submitting patients to operation who had excessive pulmonary vascular disease, technical errors with cardiopulmonary bypass, or a simple closure of the VSD because the true nature of the intracardiac anatomy was not recognized. Kirklin was first to repair this subset in 1957. He appreciated the intracardiac anatomy at operation and constructed an internal conduit which permitted the LV to eject through the VSD and into the aorta. Both he and McGoon have described clearly...
the appropriate operative technique. A "D" or diamond-shaped patch is designed to provide the roof and lateral walls of an internal conduit connecting the VSD at one end with the aortic orifice at the other. The floor of the conduit is the parietal band of the crista lying between the aortic annulus and upper margin of the VSD. Blood is ejected through this conduit from the LV to the aorta. The specialized cardiac conduction tissue is avoided by placing the suture line well back from the lower muscular edge of the VSD and into the base of the septal leaflet of the tricuspid valve along its dorsal aspect. If the VSD is small and restrictive to LV emptying, it must be enlarged by excising a portion of the ventral muscular edge. The VSD should be as large as the aortic valve annulus. It is possible to perform the repair through the right atrium, although the operation is generally done through a right ventriculotomy.

In general, primary repair is advisable except perhaps in the infant. The complex geometry of complete repair within a very small RV may result in obstruction to the flow of blood into the pulmonary artery or a restrictive internal conduit. The mortality rate of complete repair in the infant exceeds 50 percent. In this setting pulmonary artery banding and later repair is preferable.

**DORV (S,D,D), VSD related to aorta, PS present**

**Pathology.** The atria and ventricles are concordant and the aorta is to the right of the pulmonary artery (Fig. 2). The external appearance of the great vessels may be normal or the aorta may be more to the right and anterior than normal. The pulmonary artery and most, if not all, of the aorta emerge from the RV. The orifices of both vessels are usually in a side-by-side position on approximately the same coronal plane. The aortic orifice is to the right. There may or may not be fibrous continuity between the anterior leaflet of the mitral valve and the aortic annulus. When the continuity is maintained, the mitral leaflet is usually more contiguous with the base of the left coronary cusp sinus than with the noncoronary cusp sinus. When the valves are not contiguous, they are separated by the horizontal parietal limb of the crista supraventricularis. The position and boundaries of the VSD are similar to those of the previous subset. Hypoplasia of the infundibulum, pulmonary valve stenosis, or a combination of the two produce RV outflow tract obstruction. Lev believes that there is a tendency for the anatomic characteristics of hearts having the severe form of tetralogy of Fallot to overlap with the characteristics of hearts having DORV. He reviewed his specimens of these two entities and did find some subtle differences. In tetralogy of Fallot the aortic orifice often was more posterior than the pulmonary orifice and the mitral valve more contiguous with the base of the noncoronary cusp sinus than with the left coronary cusp sinus.

Associated cardiac anomalies include secundum atrial septal defect, patent ductus arteriosus, absence of the pulmonary valve, atrioventricular canal defect, drainage of a persistent left superior vena cava into the left atrium, total anomalous pulmonary venous return, cor triatriatum, subvalvular aortic stenosis, and vascular ring. Only atrioventricular canal defect and persistent left superior vena cava occur with any frequency.

**Clinical.** These patients exhibit signs and symptoms similar to those of patients with tetralogy of Fallot. A clinical differentiation between the two types is usually not possible. They are all cyanotic from the first few months of life, and hypoxic episodes and squatting may occur. Growth retardation is usual and clubbing of the fingers and toes may be present.

The chest roentgenogram usually shows decreased pulmonary blood flow. However, in contrast to tetralogy of Fallot, the heart is often slightly enlarged.

The electrocardiogram has been reviewed in
patients by Mirowski. Almost all had evidence of right atrial enlargement, severe RV hypertrophy, and intraventricular conduction disturbances. Many patients also had signs of LV hypertrophy and a delay in the atrioventricular conduction time. The high incidence of LV hypertrophy, first-degree atrioventricular block, and complete right bundle branch block seen in this group of patients is in contrast to the usual electrocardiographic findings in those patients with tetralogy of Fallot. Similar observations have been noted by others. A superiorly oriented vector has been described several times, but not a counterclockwise loop in the frontal plane, as in the previous subset.

Cardiac catheterization. The cardiac catheterization data are very similar to those seen in tetralogy of Fallot. The RV and LV pressures are equal unless the VSD is restrictive, in which case the LV pressure will exceed both the RV and aortic pressures. There is a pressure gradient between the RV and the pulmonary artery. Unlike tetralogy of Fallot, there is an oxygen saturation step-up between the right atrium and the RV since oxygenated blood from the LV must pass through the RV before entering the aorta.

The RV and LV angiograms are similar to the previous subset but in addition show varying degrees of infundibular and pulmonary valve obstruction. The operative mortality rate for all published cases is 39 per cent. Many of these deaths occurred prior to 1965 and were associated with other complicating cardiac anomalies such as anomalous distribution of the right coronary artery, complete atroventricular canal, unrecognized cor triatriatum, persistent left superior vena cava to the left atrium, and restrictive VSD. The technique of creating an internal conduit to shunt blood from the LV through the VSD to the aorta is similar to that described in the previous subset. The RV outflow tract obstruction is relieved by the same techniques used in tetralogy of Fallot. When the VSD is restrictive it is enlarged in the manner previously described.

Anomalous origin and distribution of the coronary arteries are not infrequent in DORV and may present a particularly difficult problem when the RV outflow tract obstruction cannot be adequately relieved in the usual manner. The anomalous coronary circulation seems to be related to the counterclockwise rotation of the coronary ostia as the aorta moves more to the right and anterior. The embryonic left coronary ostium frequently comes to lie in the noncoronary cusp sinus while the right coronary ostium lies in the left coronary cusp sinus. In this manner the left coronary ostium may give off the right coronary artery and the right coronary ostium may give off the left anterior descending and circumflex coronary arteries. This transfer may be complete or incomplete. When this results in an anomalous major coronary artery transversing the RV outflow tract, it may be technically impossible to place a patch across the RV outflow tract and onto the pulmonary artery or even to perform an adequate right ventriculotomy. The difficulty may be resolved in the previous subset without RV outflow tract obstruction by performing the repair through the right atrium. In those cases which require an outflow tract patch to adequately relieve RV outflow tract obstruction, the anomalous coronary artery has been dissected out of its bed and the patch positioned beneath it. A preferable technique would be the use of an external valved conduit. The presence of a single coronary ostium has also been reported.

In general, primary repair seems advisable. As in the previous subset, the symptomatic infant may present a special problem. In addition to the complex geometry of the internal conduit within a small RV, adequate relief of the RV outflow tract obstruction may require more than just the usual mobilization of the infundibulum. An external conduit is not practical in the small infant. It is advisable in this group to offer initial early palliation with a systemic-pulmonary artery shunt and later complete repair.

DORV (S,D,D), VSD related to PA, PS absent

Pathology. The atria and ventricles are concordant and the aorta is to the right of the pulmonary artery (Fig. 3). Externally, the aorta and pulmonary artery may be side by side or else the aorta may be more anterior but still to the right of the pulmonary artery. The pulmonary artery is usually dilated. The aorta and the majority or all of the pulmonary artery emerge from the RV. The aortic orifice is to the right of the pulmonary orifice and either on the same coronal plane or slightly above it. The aorta is separated from the pulmonary artery by a segment of the parietal band. The VSD is located in the posterior part of the anterior portion of the base of the ventricular septum. It may or may not involve the membranous portion of the septum. It is usually quite large, but it can occasionally be restrictive. The VSD is related to the pulmonary artery and remote from the aorta. The continuity
between the pulmonary and mitral valves is usually, but not always, interrupted by an arch of muscle formed by the fusion of the septal and parietal bands. Van Praagh believes that this arch is composed of parietal band alone. Lev has described this subset occurring with persistent continuity between the pulmonary and mitral valves. Van Praagh considers that continuity between these structures absolutely precludes against the diagnosis of DORV. This situation seems analogous to the problem of continuity between the aortic and mitral valves in the previous two subsets. There appears to be a gradual merging of DORV into transposition with VSD as the pulmonary artery moves to a more posterior position. There is no useful purpose in excluding cases with persistent pulmonary artery–mitral valve continuity from this subset of DORV if the majority of the pulmonary orifice still emerges from the RV, since the operation remains the same. Associated anomalies have included coarctation of the aorta and cleft anterior leaflet of the mitral valve.

**Clinical.** These patients often present in infancy with mild-to-moderate cyanosis, dyspnea, and congestive heart failure. Digital clubbing is common and growth retardation usual. Hypoxic episodes are rare. The chest x-ray film shows an enlarged cardiac silhouette and increased pulmonary blood flow. The electrocardiogram shows right ventricular hypertrophy, usually right axis deviation, and frequently left ventricular hypertrophy. A counterclockwise frontal loop in the vector cardiogram is not common. In contrast to the previous subsets, conduction abnormalities are rare.

**Cardiac catheterization.** The pressures in the RV and LV are equal and similar to the aortic and pulmonary artery pressures. Pulmonary vascular changes may occur at an early age. There is an oxygen saturation step-up in the RV and usually a further oxygen step-up in the pulmonary artery as blood streams from the LV across the VSD into that vessel. The magnitude of each of these oxygen step-ups is dependent upon the sampling site within the RV cavity. Blood sampled near the VSD will have a higher oxygen saturation than that sampled elsewhere.

The diagnosis is established by RV angiography. The aorta and the majority or all of the pulmonary artery emerge from the RV. The aorta is to the right of the pulmonary artery and anterior to it to varying degrees. The pulmonary artery is usually dilated. The pulmonary and aortic valves are on the same coronal plane in the lateral projection. When the aorta is considerably anterior to the pulmonary artery, its valve may be on a slightly higher plane.

The VSD is located below the pulmonary artery and is usually separated from it by conal muscle. The aorta often opacifies prior to the pulmonary artery and more densely because the preferential streaming of blood from the LV to the pulmonary artery dilutes the contrast within that vessel. The VSD is located below the pulmonary artery and is usually separated from it by conal muscle. The aorta often opacifies prior to the pulmonary artery and more densely because the preferential streaming of blood from the LV to the pulmonary artery dilutes the contrast within that vessel.

**Operative management.** Seven patients have undergone complete correction and 6 have survived. The combined operative mortality rate for all published series is 14 per cent. In 4 patients in internal conduit was constructed with the VSD at one end and the pulmonary artery at the other. In 2 patients this was done through the intact tricuspid valve to avoid an incision in the RV. This established LV–pulmonary artery continuity and thereby created physiological transposition of the great vessels. Systemic venous return was then transposed by the Mustard intra-atrial baffle. In one case the VSD was restrictive and was enlarged in the usual manner. Two cases have been repaired with an internal conduit between the VSD and the aorta. This would appear to be the preferred technique if the internal geometry of the RV permits it. However, this is unlikely in most instances since the spatial configuration of such a conduit would
be obstructive to the flow of blood from the body of the RV to the pulmonary artery.

Early operation may be necessary for the small child or infant with congestive heart failure from excessive pulmonary blood flow. Although one child of 2 years has survived complete correction, all other children surviving complete repair have been older than 5 years. The relatively high mortality rate for complete repair of transposition with VSD in infancy might also be expected in this subset of DORV since the operation is similar. Palliation in infancy would seem to be a rational approach. Appropriate palliation should include reduction of pulmonary blood flow and improved mixing at the atrial level. The streaming effect at the ventricular level, in contrast to transposition, generally precludes adequate mixing in these chambers. Pulmonary artery banding alone has been effective for the symptoms of congestive heart failure but has resulted in slight increase in cyanosis. Creation of an atrial septal defect may improve arterial desaturation but will not protect against excessive pulmonary blood flow. It would seem reasonable that symptomatic infants of this subset should benefit from a combination of these procedures with complete correction done at an older age.

**DORV (S,D,D), VSD related to PA, PS present**

Pathology. The atria and ventricles are concordant and the aorta is anterior and to the right of the pulmonary artery (Fig. 4). The pulmonary artery is smaller than the aorta. The aorta and the majority or all of the pulmonary artery emerge from the RV. The aortic orifice is to the right of the pulmonary orifice and is separated from it by a segment of the parietal band. The VSD is located high in the ventricular septum. It is related to the pulmonary artery and remote from the aorta. There is infundibular subpulmonic stenosis created by the approximation of the parietal and septal bands. There may be pulmonic valve stenosis as well. The pulmonary and mitral valves may or may not be contiguous.

Clinical. These patients are cyanotic as infants, their growth is retarded, and they may have hypoxic episodes. Digital clubbing may be present. The chest x-ray film usually shows slight cardiac enlargement and a normal or reduced pulmonary blood flow. The electrocardiogram shows right axis deviation and right ventricular hypertrophy.

Cardiac catheterization. Only a few patients have undergone cardiac catheterization studies. The RV and LV pressures have been equal and there has
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Fig. 6 Double-outlet right ventricle (S,D,L), ventricular septal defect related to the aorta, pulmonary stenosis present.

been an oxygen saturation step-up in the RV. The pulmonary artery has been entered in only one patient.68

Few angiographic studies have been performed.20, 47, 62, 68 The aorta appears anterior to the pulmonary artery and may opacify first during an RV injection. The posteriorly positioned pulmonary artery overrides the VSD and fills predominately from the LV.

Operative management. One patient has undergone complete correction.68 The infundibulum was mobilized and an internal conduit constructed between the VSD and the aorta. An attempt at palliation has been made with a systemic-pulmonary artery shunt in 4 patients. Each patient died in the early postoperative period.47, 67

DORV (S,D,L), VSD related to PA, PS absent

Pathology. The atria and ventricles are concordant and the aorta is anterior and to the left of the pulmonary artery (Fig. 5). The pulmonary artery is dilated. The aorta and the majority of the pulmonary artery emerge from the RV. The aortic orifice is to the left of the pulmonary orifice and separated from it by a segment of the parietal band. The VSD is located in the anterior portion of the base of the septum. It is related to the pulmonary artery and remote from the aorta.48, 49

Clinical. Only limited clinical information is available for this subset. The symptoms have been those of a large VSD with excessive pulmonary blood flow and minimal cyanosis. The chest x-ray film has shown cardiac enlargement and increased pulmonary blood flow. The electrocardiogram showed left axis deviation with biventricular hypertrophy in the only report available.48

Cardiac catheterization. Only limited data are available from a single patient. The RV and systemic pressures were equal and there was an oxygen saturation step-up in the RV. An RV angiogram demonstrated the aorta anterior to the pulmonary artery. The aorta arose entirely from the RV and the pulmonary artery did so predominantly. The pulmonary artery overrode the VSD.48

Operative management. Only one patient has undergone complete repair, and a satisfactory result was obtained.48 Muscle was resected from the cephalad roof of the left ventricle and the septum to establish free communication between the LV and the aorta. An oval patch was then positioned within the RV as a conduit between the VSD and the aorta.

DORV (S,D,L), VSD related to aorta, PS present

Pathology. The atria and ventricles are concordant (Fig. 6). The aorta is to the left of the pulmonary artery and on the same plane or anterior. The pulmonary artery is smaller than the aorta. The pulmonary artery and most or all of the aorta emerge from the RV. The aortic orifice is to the left of the pulmonary orifice and on the same coronal plane or slightly higher. There is infundibular pulmonary stenosis created in part by the parietal band. There may be pulmonic valve stenosis as well. The VSD is located high in the ventricular septum. It is related to the aorta and remote from the pulmonary artery. The aortic and mitral valves are not contiguous. The right coronary artery passes across the RV outflow tract in front of the pulmonary artery.31, 50, 51, 65

Clinical. These patients may be cyanotic from birth or else the onset of cyanosis may be delayed. Hypoxic episodes may occur. Digital clubbing is present. Growth retardation has not been prominent.31, 50, 51

The chest x-ray film shows a normal or small heart and decreased pulmonary blood flow. The electrocardiogram shows right axis deviation and RV hypertrophy.31, 50, 51 Left axis deviation and a counterclockwise loop in the frontal plane of the vectorcardiogram have also been noted.65

Cardiac catheterization. The RV and LV pressures were equal. The pulmonary artery was only rarely entered, and in each instance a reduced pressure
reflected the subpulmonic stenosis. There was an oxygen saturation step-up in the RV in some cases and not in others. Failure to identify an oxygen step-up in these cases was most likely related to a low sampling site within the inflow portion of the RV.

RV angiography demonstrates the aorta to be anterior and to the left of the pulmonary artery. There is infundibular pulmonary stenosis. LV angiography shows the dye to cross the VSD and opacify an overriding aorta. There is discontinuity between the aortic and mitral valves.65

Operative management. Four patients have undergone complete correction with one operative death.50, 51, 65, 66 The obstructing infundibulum was resected in each and an internal conduit constructed between the VSD and the aorta. In 2 cases adequate relief of the RV outflow obstruction was obtained by mobilizing the infundibulum.51 In another it was necessary to place a patch across the RV outflow tract under the mobilized coronary artery.50 Although this technique achieved a satisfactory result, an external valved conduit, as constructed in the fourth case, would be preferrable.51, 65

One patient has been palliated with a systemic—pulmonary artery shunt.31 The initial result was satisfactory but the child later died with congestive heart failure.

DORV (S,L,L, S,L,A, S,L,D), VSD related to aorta or PA, PS present

Since these subsets are represented by limited numbers and have atrioventricular discordance in common, they will be described together (Fig. 7).

Pathology. The heart is in dextroversion. The atria and ventricles are discordant. The aorta is anterior and usually to the left of the pulmonary artery. Rarely, these two vessels may be in a direct anteroposterior relationship of the aorta may be slightly to the right. The pulmonary artery is small. The RV is anterior and to the left of the LV. The aorta and the pulmonary artery emerge from the RV. The aortic orifice is usually to the left of the pulmonary orifice, and the VSD usually underlies the pulmonary artery. The VSD is frequently of the atrioventricular canal type. If the pulmonary orifice is to the left of the aorta and posterior, the VSD is more closely related to the aorta. There is infundibular pulmonic stenosis and there may be valvular stenosis as well. There is discontinuity between the semilunar and atrioventricular valve tissue.31, 52, 53

Clinical. These patients are cyanotic from infancy. Growth retardation and hypoxic episodes are rare. The chest x-ray film shows the heart to be in dextroversion and normal in size. The stomach bubble is to the left. The pulmonary vasculature is normal or reduced. The electrocardiogram has shown right ventricular hypertrophy and occasionally left ventricular hypertrophy. A counterclockwise loop in the frontal vector cardiogram is not unusual and may reflect the high incidence of atrioventricular canal type defects.52, 53

Cardiac catheterization. The RV and LV pressures are equal and similar to systemic pressure. The pulmonary artery was not catheterized in any instance.52 Pulmonary artery sampling has been obtained by a suprasternal puncture. There was always an oxygen saturation step-up in the pulmonary artery, and the pressure within this vessel reduced.

RV angiography demonstrates that both the aorta and pulmonary artery arise from this chamber. The aorta is anterior and usually to the left of the pulmonary artery. The VSD lies below the crista supraven-
tricularis. There is subpulmonic stenosis and discontinuity between the semilunar and atrioventricular valves.\textsuperscript{52, 53}

**Operative management.** Three patients have undergone complete correction with one death. In 2 patients the VSD was more closely related to the pulmonary artery, which was to right of the aortic orifice. A right ventriculotomy was made, the body of the crista was resected, and the infundibulum mobilized. An internal conduit was constructed between the VSD and the pulmonary artery.

In one case the pulmonary artery orifice was to the left of the aortic orifice, and the VSD therefore was more closely related to the aorta. The VSD was simply closed through the LV, the origin of the pulmonary artery ligated, and an external valved conduit inserted between the LV and the pulmonary artery. An internal conduit would most likely have been obstructed by the systemic pressure within the RV pressing on that prosthesis. Four patients have been palliated with systemic–pulmonary artery shunts.\textsuperscript{31, 52}

Infants and small children who are severely symptomatic should be palliated with a shunt. The older child should undergo complete correction. Although an internal conduit would seem preferable, it is probably not possible to determine accurately if this can be constructed prior to making a right ventriculotomy. If it is not possible, then a second incision in the LV is necessary to position a conduit. To avoid an unnecessary incision in what may become the systemic ventricle, the surgeon should employ the technique of a left ventriculotomy, simple closure of the VSD, and an external valved conduit between the LV and pulmonary artery in all cases.

**Summary**

In general, the operative management of DORV is straightforward if each case is evaluated in an organized manner. When the VSD is related to the aorta, a simple internal conduit is used. In the presence of pulmonary stenosis, the obstructing infundibulum and pulmonary valve are mobilized in the usual manner. If this does not adequately relieve the obstruction, an external valved conduit is preferred to an outflow tract patch. When the VSD is related to the pulmonary artery, an internal conduit is positioned to create physiological transposition, and the repair is completed with an intra-atrial baffle. Rarely, an internal conduit between the VSD and aorta may be possible.

When the atria and ventricles are in a discordant relationship, the VSD is closed through the LV, the pulmonary artery ligated, and LV–pulmonary artery continuity established with an external valved conduit. No cases of noncommitted VSD have been repaired.

The question of continuity or discontinuity of the semilunar and atrioventricular valves is unimportant from the standpoint of operation. It is necessary that the surgeon identify at operation the exact nature of the anomaly so that the appropriate repair be done.

**REFERENCES**


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