Double-Outlet Right Ventricle
with Pulmonic Stenosis

Surgical Considerations and Results of Operation

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SUMMARY
The results of complete repair of double-outlet right ventricle with pulmonic stenosis in 22 Mayo Clinic patients were analyzed. The overall mortality rate was 32%. Since 1960, the mortality rate has been reduced to 16%. Anomalies of the distribution of the right coronary artery, a second ventricular septal defect of the atrioventricular canal type, and significant residual pulmonic stenosis were associated with a higher mortality rate. The difficulties in the diagnosis of this uncommon condition are emphasized.

Complete repair is best carried out after the patient is 4 years old. A systemic-to-pulmonary arterial shunt is the procedure of choice for those who are younger than this. In the occasional patient with anomalous distribution of the right coronary artery which precludes adequate relief of the pulmonic stenosis, use of an aortic homograft from the right ventricle to the pulmonary artery should be considered.

Additional Indexing Words:
Ventricular septal defect Cardiac surgery Cardiac anomalies

DOUBLE-OUTLET right ventricle is a rare cardiac malformation characterized by a rightward transposition of the aorta so that its orifice originates entirely from the right ventricle and lies in the same coronal plane as the pulmonic valve. The aortic ring lies entirely to the right of the ventricular septal defect, which is posterior and infracristal and is the only exit from the left ventricle. The usual continuity between the aortic valve and the anterior or aortic leaflet of the mitral valve is lacking. Pulmonic stenosis, when present, may be valvular, infundibular, or both.

Little information has been reported about the surgical treatment of this malformation. Our experience with double-outlet right ventricle without pulmonic stenosis has been recently reported. The presence of pulmonic stenosis makes a subgroup of double-outlet right ventricle with distinct clinical and hemodynamic characteristics. This study presents data on all patients with double-outlet right ventricle who were operated on at the Mayo Clinic in whom significant pulmonic stenosis was an associated finding.

Clinical Studies
From 1957 through 1969, 22 patients with double-outlet right ventricle and pulmonic stenosis underwent surgical repair at the Mayo Clinic. (Data on the first eight patients of this series have been reported previously.) Ten patients were males and 12 were females; the median age at the time of operation was 10 years (range, 5 to 33 years).

All patients were cyanotic. The clinical histories and auscultatory findings were similar to those of patients with tetralogy of Fallot. The single second sound in many
patients, however, was loudest along the upper left sternal border rather than along the lower left sternal border, as is usual in tetralogy of Fallot. The cardiothoracic (C/T) ratio on chest roentgenograms varied from 0.40 to 0.64, with a mean of 0.48. In most cases, pulmonary vasculature was decreased.

The electrocardiograms (ECG) of all but four patients showed a mean QRS axis directed inferiorly in the frontal plane, with a clockwise QRS loop. In the four exceptions, the loop was counterclockwise, with a mean axis directed superiorly. All ECGs showed evidence of right ventricular hypertrophy. Four ECGs showed evidence of left ventricular hypertrophy as well, but since only two patients had a small ventricular septal defect, the ECG was not helpful in differentiating double-outlet right ventricle with pulmonic stenosis from tetralogy of Fallot or in predicting whether the ventricular septal defect was small. Right atrial hypertrophy was common. In two patients, the QRS interval was greater than 0.10 second.

Seven patients had undergone previous palliative procedures: four had a Blalock-Taussig anastomosis, two had a Brock valvotomy, and one had both a Potts and a Blalock-Taussig shunt. The pulmonic stenosis was valvular in four patients, infundibular in nine, and combined valvular and infundibular in nine.

Preoperative hemodynamic data were available for 16 patients. In 12, the pulmonary artery was entered, and the peak systolic pressure gradient between the right ventricle and the pulmonary artery varied from 40 to 152 mm Hg, with a mean of 73 mm. Data on peak systolic pressures in the right and left ventricles were available on eight patients; only two had a pressure gradient between the ventricles (15 and 20 mm Hg, respectively).

Associated cardiac anomalies were frequent (table 1). Six patients had a large branch of the right coronary artery that crossed the outflow tract of the right ventricle and gave origin to the left anterior descending coronary artery. An additional patient had no main left coronary artery; both the left anterior descending and the left circumflex arteries originated from a branch of the right coronary artery that crossed the outflow tract of the right ventricle.

**Results**

The repair of this anomaly is basically the same as that described previously\(^1,2\) (fig. 1), and the pulmonic stenosis is relieved by the same technique as in the repair of tetralogy of Fallot.\(^4,5\) An especially challenging problem is created when a large anomalous branch of the right coronary artery crosses the outflow tract.

The ventricular septal defect was enlarged in four patients. Two of them had a preoperative peak systolic gradient between the left and right ventricles. The patch used in the construction of the left ventricular conduit was made of knitted Teflon (16 patients) or pericardium (6 patients).

**Early Results**

Seven patients died postoperatively (overall operative mortality rate, 32%). The first four patients to be operated on failed to survive operation; the operative mortality rate since 1960 has been 16%. Four of the seven patients who died had anomalous distribution of the right coronary artery, one had a complete form of atrioventricular canal (type C),\(^3\) one had ventricular septal defect of the atrioventricular canal type, and one had undiagnosed cor triatriatum.
Figure 1

Anatomy and repair as seen through a right ventriculotomy. The atrioventricular node is seen through the tricuspid valve orifice, with the bundle of His coursing through the tricuspid annulus onto the ventricular septum along the postero-inferior margin of the ventricular septal defect (VSD).

Three crucial sutures, labeled 1, 2, and 3, attach the large oval patch to the base of the septal leaflet of the tricuspid valve (between asterisks). Penetration of the tricuspid annulus by these sutures is avoided, but complete closure of this posterior angle of the VSD is achieved by approximating septum, patch, and the base of the septal leaflet of the tricuspid valve (TV) with suture 3. Injury to the main bundle of His is thus avoided, and injury to the left bundle branches is avoided by grasping (with suture 3 and subsequent adjacent sutures) only the right ventricular surface of the ventricular septum. After these crucial sutures are tied, the patch is attached (inset A) to the ventricular septum caudally (***) and to the muscle leading from the tricuspid annulus to the orifice of the aortic valve (****). The final suturing (inset B) continues the attachment of the patch about the aortic orifice, then across the crista supraventricularis to the previously placed sutures along the margin of the VSD. A tunnel or conduit thus has been created, connecting the left ventricle via the VSD with the aortic orifice.

Right ventricular ($P_{RV}$) and left ventricular ($P_{LV}$) pressures were measured in the operating room, after repair, in 19 of the 22 patients. In the nine patients with infundibular pulmonary stenosis, the $P_{RV}/P_{LV}$ ratio was lower (varied from 0.33 to 0.85, with a mean of...
Table 2

Relationship Between Operative Mortality and Postoperative PRV/PLV in 19 Patients With Double-Outlet Right Ventricle and Pulmonic Stenosis

<table>
<thead>
<tr>
<th>PRV/PLV*</th>
<th>No. of patients</th>
<th>Hospital deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;0.6</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>0.6-0.75</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>&gt;0.75</td>
<td>4</td>
<td>3</td>
</tr>
</tbody>
</table>

*PRV/PLV = right ventricular-left ventricular peak systolic pressure ratio after repair in the operating room.

0.51) than in the 13 patients with valvular or combined pulmonic stenosis (varied from 0.5 to 1.0, with a mean of 0.71), and none of the nine required a pericardial patch to enlarge the outflow tract of the right ventricle. Eight of the nine survived operation.

Of the 13 patients with valvular stenosis or combined valvular and infundibular pulmonic stenosis, six died postoperatively. In six of the 13 patients, the outflow tract of the right ventricle had been enlarged with a pericardial patch; the mean PRV/PLV after repair was 0.61 (range, 0.5 to 0.88). In the other seven patients, no enlargement of the outflow tract had been used, and the mean PRV/PLV was 0.77 (range, 0.6 to 1.0).

Three of the four patients with residual PRV/PLV higher than 0.75 died postoperatively (table 2). Of the five patients with a residual peak systolic pressure in the right ventricle of more than 75 mm Hg, four failed to survive operation. Only one patient had a residual pressure gradient between the left ventricle and the aorta after repair (25 mm Hg).

In addition to being related to inadequate relief of the pulmonic stenosis, operative death was also related to the presence of an anomalous distribution of the right coronary artery (four of seven patients died) and to a ventricular septal defect of the atroventricular canal type (both patients died—one of whom had a complete atroventricular canal).

Two patients had complete heart block, and both died. In one of them, heart block developed at the time of closure of a second ventricular septal defect of the atroventricular canal type. Repair of the ventricular septal defect in one patient resulted in aortic insufficiency, and it was necessary to replace the valve with a Starr-Edwards prosthesis 1 week later. Reexploration for excessive bleeding was necessary in three patients, and three others required tracheostomy for assisted ventilation.

Late Results

Of the 15 patients with double-outlet right ventricle and pulmonic stenosis who survived operation, one died 1 year after operation of cardiac failure. His PRV/PLV was 0.66 after the repair. The other 14 patients were doing well 1 to 8 years after operation (mean follow-up, 4 years), with no need for cardiac drugs and no restrictions of activity.

Comment

Double-outlet right ventricle or origin of both vessels from the right ventricle is a rare cardiac malformation with many variations. The lack of a clear pathologic classification of the different forms of double-outlet right ventricle has been an impediment to the development of a rational plan of surgical treatment. Recently, we have presented a surgical classification of double-outlet right ventricle.6

Pulmonic stenosis and single ventricle can be associated with any of these types of double-outlet right ventricle. Neufeld and co-workers,7,8 Hazell,8a and Krongrad and his associates (unpublished data) have made detailed studies of the pathologic anatomy and clinical findings, the hemodynamics, and the electrocardiographic patterns, respectively, in cases encountered at the Mayo Clinic.

Kirklin9 was the first to correct the classic type of double-outlet right ventricle. Results of the surgical repair of other types6,10,11 at our institution have been reported. A few cases of surgical repair of double-outlet right ventricle with pulmonic stenosis have been reported by others.12–14

The differential diagnosis between double-outlet right ventricle with pulmonic stenosis and severe forms of tetralogy of Fallot is
DOUBLE-OUTLET RIGHT VENTRICLE

difficult to make clinically. Electrocardiographic evidence of left ventricular hypertrophy may be a clue to double-outlet right ventricle, but this was not a common finding in our patients. Cardiac catheterization may not establish the diagnosis unless the position of the aorta and pulmonary artery relative to each other is demonstrated with catheters in each of these arteries. The diagnosis can sometimes be made with confidence by the use of angiocardiology; the typical findings are simultaneous opacification of both great arteries from the right ventricle, infracristal ventricular septal defect, aortic and pulmonic valves at the same transverse level, a variable degree of transposition of the aorta, separation of the aortic valve from the aortic leaflet of the mitral valve by the crista, and valvular or infundibular pulmonic stenosis, or both.

It has not been our practice to obtain preoperative angiograms of all patients with tetralogy of Fallot; surgeons who follow this practice, however, must be able to recognize the intracardiac pathologic changes of double-outlet right ventricle with pulmonic stenosis in order to accomplish a successful repair when this lesion is encountered unexpectedly at operation.

The features that suggest the presence of double-outlet right ventricle at the time the heart is exposed at operation are (1) the side-by-side position of the aorta and pulmonary arteries in the same coronal plane, as the result of pronounced dextroposition of the aorta, and (2) the location of both semilunar valves in the same transverse plane. This appearance is far from diagnostic, however. In our experience, often we have been suspicious in cases of severe tetralogy of Fallot that we were dealing with a double-outlet right ventricle, only to find after ventriculotomy that there was continuity between the mitral and aortic valves. The same has been true (but less often) in ventricular septal defect without pulmonic stenosis.

In four patients in whom the ventricular septal defect was smaller than the aortic valve, the defect was enlarged; only one patient had a significant residual gradient across the conduit.

The presence of atrioventricular canal defect is not rare and is associated with a higher operative mortality rate and a higher incidence of complete heart block. A residual $P_{RV}/P_{LV}$ more than 0.75 is also associated with significant operative mortality and indicates the need for enlargement of the outflow tract of the right ventricle with a pericardial patch. In most patients, this patch needs to be extended across the pulmonic valve. The frequent association of anomalous distribution of the right coronary artery to the left anterior descending coronary artery is often incompatible with good relief of the pulmonic stenosis; the use of an aortic homograft as in repair of truncus arteriosus or in transposition of the great vessels (Rastelli's operation) should then be considered.

In our practice, patients with double-outlet right ventricle and pulmonic stenosis who require surgical intervention before the age of 4 years receive a systemic-to-pulmonary arterial shunt, preferably a Blalock-Taussig anastomosis if they are older than 6 months, or a Waterston anastomosis if they are younger than 6 months. Complete intracardiac repair is performed in older patients.

The results presented in this study represent the entire experience of this institution since the early days of open-heart surgery. A better understanding of the management of the pulmonic stenosis has improved the recent hospital survival significantly. Late results in patients having satisfactory relief of associated pulmonic stenosis have been gratifying.

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