Surgical Considerations of Ventricular Septal Defect Associated with Complete Transposition of the Great Arteries and Pulmonary Stenosis

With Special Reference to the Rastelli Operation

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SUMMARY

Ventricular septal defect (VSD) associated with complete transposition of the great arteries was studied, and each of the typical anatomic varieties was depicted to emphasize the characteristic features of the VSD. Among the 32 specimens, 13 cases were found to have a VSD lying anterosuperior to the origin of the papillary muscle of the conus, indicating that repair of the VSD could be performed without difficulty. Such a defect situated above the crista supraventricularis has proved to be technically most preferable, as shown in the case report. In contrast, the remaining 19 cases appeared to be inappropriate for the technique, as the VSD was overhung by a part of the tricuspid valve. Additional analysis was made of the angiocardiographic appearances of the VSD, demonstrating some characteristic patterns.

Additional Indexing Words:
Papillary muscle of the conus Homograft of the aorta Angiocardiography Intracardiac tunnel

OUR RECENT experience with the Rastelli operation for complete transposition of the great arteries has taught us the fundamental significance of the anatomic features of ventricular septal defect (VSD) associated with this condition. This technique consists of reconstruction of both ventricular outflows: the left ventricular outflow is redirected toward the aortic orifice by repairing the VSD, and the right ventricular outflow is constructed by placing a valve-retaining conduit graft between the right ventricle and pulmonary artery after the pulmonary trunk proximal to the anastomosis is tied off. Usually the latter technique presents no particular issues. However, repair of the VSD—that is, the creation of an intracardiac tunnel—yields some important problems, for it seems to be greatly influenced (compared with the simpler closure of the VSD as in the Mustard technique) by the geometric factors in the right ventricle. Furthermore, if the VSD is significantly smaller than the aortic orifice, the VSD must be enlarged by excising the muscular interventricular septum so as to avoid the left ventricular outflow obstruction.

These surgical problems are yet to be answered, while the hemodynamic and surgical significance of pulmonary stenosis...
has been well documented. The present study attempts to clarify the surgical implications of the positional and spatial relationships between the VSD and adjacent structures, and to discuss the indications for the Rastelli operation from the aspect of its technique.

Methods

Registered in the Heart Institute of Japan are 37 cases of complete transposition of the great arteries with ventricular septal defect. Of these 37 cases, five hearts with either anomalous pulmonary venous connection or atretic atrioventricular valve were excluded from the present series, as these complicated malformations bear different surgical problems from those in the Rastelli repair. This paper is based on a restudy of the remaining 32 patients, analysis of 14 angiocardiograms, and review of a case in which the Rastelli repair was attempted.

Among the 32 hearts, pulmonary stenosis was present in 10 cases (valvular in two, infundibular in five, and both valvular and infundibular stenosis in three). Other main associated lesions were atrial septal defect (or patent foramen ovale) in 12, persistent ductus arteriosus in seven, and coarctation of the aorta in five. The age of the patients ranged from 8 days to 22 years.

Regarding the ventricular septal defects, a comparative study was made between angiocardiographic appearances and anatomic features that were found in the present anatomic study or those that were available from the operative records subsequently made at the time of open-heart surgery. The size of the defect was expressed as a ratio of its anteroposterior diameter to the aortic diameter at the level of the aortic annulus.

Anatomic Considerations (Thirty-Two Cases)

The anatomic characteristics of the ventricular septal defect encountered in this series will be best understood if the specimens are divided into the following three anatomic types, depending on the landmarks suggested by Becu and associates:10 (I) defects anterosuperior to the crista supraventricularis; (II) defects posteroinferior to the crista supraventricularis; and (III) muscular isolated or multiple defects.

Defects Anterosuperior to the Crista

Two cases among the 32 hearts in this study had a defect above the crista supraventricularis. Pulmonary stenosis was present in both cases. When viewed from the right side of the heart, the upper border of the defect was formed by the aortic annulus fibrosus or the base of the aortic cusps. The defect was not closely related to the tricuspid valve, so that there were no obstructive structures between the anterior aspect of the defect and the aortic orifice.

In one case the defect was so large as to extend inferiorly to the septal limb of the crista, having a ratio of 1.4 (fig. 1). The anterior edge of the defect was at the junction between the anterior wall of the ventricle and the ventricular septum. Posteriorly, a large part of the membranous portion of the ventricular septum (or membranous septum) was defective, but there was no continuity between the tricuspid and mitral valve leaflets.

In the other case, the region involved by the defect was restricted to the upper portion of the crista supraventricularis. When viewed from the left ventricle, the defect was located anterior to the intact membranous septum just beneath the pulmonary cusps. The anterior edge of the defect was separated with a thin rim of muscular tissue from the conjunctional area between the anterior wall of the left ventricle and the ventricular septum. This defect showed a ratio of 0.7, and if necessary it should be enlarged by careful excision of the anterior rim of the defect.

Defects Posteroinferior to the Crista

A defect was present in this region in 26 of the 32 hearts. These 26 cases could be subdivided into two groups. Group A comprised eight patients in whom the papillary muscle of the conus (Lancisi muscle) was adherent to the posterior or lower border of the defect, so that the center of the defect was well visualized from the anterior aspect of the right ventricle. When viewed from the left ventricle, the defect was situated in the region involving the membranous septum posteriorly and the muscular septum anteriorly, lying some distance from the pulmonary cusps. These anatomic situations would make it possible to excise a part of the crista supraventricularis forming the upper ridge of

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Figure 1

Ventricular septal defect (VSD) anterosuperior to the crista supraventricularis in a 22-year-old man. (Upper left) Right ventricular view. A large VSD is present just beneath the aortic cusps. The papillary muscle of the conus (P.M.C.) arises on the lower border of the defect. In this heart, the anterior tricuspid leaflet is split. (Upper right) Left ventricular view. VSD occupies the anterosuperior quadrant of the ventricular septum. (Lower left and right) Lateral frame and its diagram of right ventricular injection from the same patient as above. Pulmonary valvular and subvalvular stenosis is evident. Note that the crista supraventricularis is lacking. Ao = aorta; PT = pulmonary trunk; RA = right atrium; MV = mitral valve; LV = left ventricle; RV = right ventricle.
Venricular septal defect (VSD) posteroinferior to the crista supraventricularis (CS) in a 5-year-old boy. (Upper left) Right ventricular view. The papillary muscle of the conus (P.M.C.) arises on the lower border of the defect. Dotted line indicates the region corresponding to that involved by the pulmonary valve and artery at the left side of the heart. Hatched area denotes the region that can be excised without serious postoperative complication. (Upper right) Left ventricular view. Through the defect is seen a part of the well-developed crista which can be removed (hatched area). (Lower left and right) Right ventricular injection in the same patient. The dome-shaped pulmonary valve and infundibular stenosis is clearly visible. VSD is delineated by contrast medium streaming into the pulmonary trunk (PT) from the right ventricle (RV). Ao = aorta; LV = left ventricle; TV = tricuspid valve.
the defect, as shown in figure 2. Three patients in this category were found to have pulmonary stenosis. In two of these three cases the defects needed to be enlarged, while in the third the defect was large enough, with a ratio of 1.0. The remaining five patients without pulmonary stenosis showed a range of ratios from 0.2 to 0.9 which averaged 0.6 (table 1).

Table 1

<table>
<thead>
<tr>
<th>Type of VSD</th>
<th>With pulmonary stenosis</th>
<th>Without pulmonary stenosis</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No.  Size (average)</td>
<td>No.  Size (average)</td>
<td>No.  Average size</td>
</tr>
<tr>
<td>(I) Anterior defect</td>
<td>2  0.7 - 1.4 (1.1)</td>
<td>0  (--)</td>
<td>2  1.1</td>
</tr>
<tr>
<td>(II) Posterior defect</td>
<td>3  0.5 - 1.0 (0.7)</td>
<td>5  0.2 - 0.9 (0.6)</td>
<td>8  0.7</td>
</tr>
<tr>
<td>Group A (not overhung)</td>
<td>5  0.4 - 1.5 (0.7)</td>
<td>13  0.3 - 1.2 (0.7)</td>
<td>18  0.7</td>
</tr>
<tr>
<td>Group B (overhung)</td>
<td>5  (--)</td>
<td>4  0.3 - 2.2 (1.0)</td>
<td>4  1.0</td>
</tr>
<tr>
<td>(III) Muscular defect</td>
<td>0  (--)</td>
<td>22  (0.71)</td>
<td>32  0.73</td>
</tr>
</tbody>
</table>

Muscular Isolated or Multiple Defects

Four patients had this type of defect. Although none of the defects was associated...
with pulmonary stenosis, the anatomic findings in these cases deserve special description. In three of the four cases a defect was present below the crista supraventricularis, but the center of the defect was more anteriorly located than is usually found in defects posteroinferior to the crista supraventricularis. When viewed from the left ventricle, the defect was situated comparatively anteriorly, lying some distance from the intact membranous septum as well as the pulmonary cusps. In the right ventricle the chordae tendineae to the septal leaflet and in part to the anterior leaflet of the tricuspid valve were directly inserted into the posteroinferior angle of the defect in each of the three cases. The surgical implications of these anatomic findings proved to be identical to those of group A in type II VSD. Two of these three defects had a ratio of 0.3 and 0.7, respectively, whereas in the third the ratio was undecided because the defect had been surgically closed.

In the remaining patient, multiple defects were present below the crista supraventricularis; two large defects were separated by a column of muscular tissue which vertically bridged the center of the defect. The anterior defect was located in the muscular septum, while another defect lay in the region involving the membranous septum. Since the papillary muscle of the conus arose nearly on the midpoint of the muscular column in

Figure 4

Lateral projections from the patient undergoing the Rastelli operation. (Left) Right ventricular injection; angiogram taken before operation. A large ventricular septal defect is visible (arrow indicates lower margin of the VSD). The pulmonary valvular and infundibular stenosis can be noted. (Right) Right atrial injection; angiogram taken after operation; homograft indicates a homograft of the ascending aorta with aortic valve used for a new pulmonary valve and artery. Abbreviations as in figures 1 through 3.
question, the posterior defect was partially overhung by the tricuspid valve. In this instance it would be difficult to determine whether the upper half of the column could be excised without resulting in serious tricuspid insufficiency.

**Angiocardiographic Patterns**

In general, ventricular septal defects can be seen in the lateral projection of a frame taken when the contrast medium injected into the right ventricle streams into the pulmonary trunk. Consequently, the intensity of opacification in the defect must be almost equal to that of the right ventricle; in contrast, the left ventricle is less opacified. The crista supraventricularis, the origins of both great arteries, and the ventricular septum serve as valuable landmarks. It must be emphasized, however, that the position of the ventricular septal defect is reasonably identifiable when the outflow tracts of the two ventricles or both great arteries are not superimposed but are clearly outlined.

It is also essential to study a series of successive frames (not a single film) so as to avoid a false interpretation. This is particularly important when complete transposition with VSD must be differentiated from a single ventricle with transposed great arteries. Angiocardiograms of a single ventricle with well-developed trabeculations may present figures resembling the ventricular septum, which often confuses the diagnosis. In early films there is usually no obvious difference in density of opacification between “two” ventricular cavities bordered by the false ventricular septum.

The observations from the 14 angiocardiograms in this study demonstrated two major patterns regarding the position of a ventricular septal defect: either the absence or the presence of the crista supraventricularis. The crista supraventricularis usually appears, if present, in the lateral projection as a translucent area just beneath the conjoint region of the aortic and pulmonary cusps. In two of the 14 angiograms the crista was absent (fig. 1, lower). The anatomic study of these two cases revealed that a large defect was located above the crista supraventricularis. In 11 of the 14 angiograms the crista was evident, suggesting that the defect was positioned between the crista above and the ventricular septum below (fig. 2, lower). This interpretation was positively confirmed by anatomic findings that the defect was located below the crista supraventricularis. In all patients except one, the defect was posteroinferior to the crista, involving the membranous septum, while in the other it was of the muscular isolated type with intact membranous septum. In the latter patient, none of the films provided useful information since both great arteries were

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**Table 2**

<table>
<thead>
<tr>
<th>No.</th>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Pulmonary stenosis</th>
<th>Type of VSD*</th>
<th>Defect size</th>
<th>Indications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>H.A.</td>
<td>22 yr</td>
<td>M</td>
<td>Valvular and infund.</td>
<td>I</td>
<td>1.4</td>
<td>Ideal candidate</td>
</tr>
<tr>
<td>2</td>
<td>Y.N.</td>
<td>12 yr</td>
<td>F</td>
<td>Infundibular</td>
<td>II – B</td>
<td>1.5</td>
<td>Difficult to repair</td>
</tr>
<tr>
<td>3</td>
<td>T.Y.</td>
<td>3 yr</td>
<td>M</td>
<td>Infundibular</td>
<td>II – B</td>
<td>0.6</td>
<td>Difficult to repair</td>
</tr>
<tr>
<td>4</td>
<td>T.N.</td>
<td>5 yr</td>
<td>M</td>
<td>Valvular and infund.</td>
<td>II – A</td>
<td>0.6</td>
<td>Good candidate with VSD to be enlarged</td>
</tr>
<tr>
<td>5</td>
<td>K.M.</td>
<td>7 mo</td>
<td>F</td>
<td>Valvular and infund.</td>
<td>II – B</td>
<td>0.5</td>
<td>Difficult to repair</td>
</tr>
<tr>
<td>6</td>
<td>U.T.</td>
<td>2 yr</td>
<td>F</td>
<td>Infundibular</td>
<td>II – B</td>
<td>0.4</td>
<td>Difficult to repair</td>
</tr>
<tr>
<td>7</td>
<td>Y.T.</td>
<td>12 mo</td>
<td>M</td>
<td>Valvular</td>
<td>II – B</td>
<td>0.4</td>
<td>Difficult to repair</td>
</tr>
<tr>
<td>8</td>
<td>A.Y.</td>
<td>5 yr</td>
<td>M</td>
<td>Valvular</td>
<td>II – A</td>
<td>0.5</td>
<td>Good candidate with VSD to be enlarged</td>
</tr>
<tr>
<td>9</td>
<td>M.K.</td>
<td>5 yr</td>
<td>M</td>
<td>Infundibular</td>
<td>II – A</td>
<td>1.0</td>
<td>Ideal candidate</td>
</tr>
<tr>
<td>10</td>
<td>A.U.</td>
<td>11 mo</td>
<td>F</td>
<td>Infundibular</td>
<td>I</td>
<td>0.7</td>
<td>Ideal candidate</td>
</tr>
</tbody>
</table>

*I = anterior defect; II = posterior defect; A = ventricular septal defect (VSD) situated above the papillary muscle of the conus; B = VSD overhung by the papillary muscle of the conus.*
entirely superimposed in the lateral frame and in the anteroposterior frame as well.

Report of a Case
A 12-year-old boy evaluated as having complete transposition of the great arteries, VSD, and pulmonary stenosis was admitted for surgical correction. A preoperative cardiac catheterization showed the peak systolic gradient between the left ventricle and the pulmonary artery to be 60 mm Hg. The preoperative right-heart angiograms demonstrated moderate infundibular stenosis with domed pulmonic valve and a large VSD without evidence of the crista supraventricularis (fig. 4).

The operation was performed on January 19, 1970. The intracardiac anatomy was as follows. The VSD was located above the crista supraventricularis, but the upper border of the defect was separated from the base of the aortic cusps by a thin rim of muscular tissue. The defect extended inferiorly to the septal limb of the crista, and the size of the defect was assessed as 3.0 cm, while the aortic diameter was 3.5 cm, showing a ratio of 0.9. The papillary muscle of the conus was adherent to the lower border of the defect, so that repair of the defect was easily accomplished with the use of a semicylindrical Teflon tube. A homograft of aorta, including the aortic valve, was used to reconstruct the pulmonary valve and artery. The patient is now doing well 15 months after the operation.

Discussion
The observations from the present study indicate that greater skill and more accurate evaluation of the intracardiac anatomy, especially in regard to the ventricular septal defect as well as pulmonary stenosis, are required to achieve successful Rastelli repairs. This procedure, in many respects, contrasts with the Mustard technique which attempts redirection of the venous inflows. In the latter technique the repair of a VSD may be much easier, but relief of the pulmonary stenosis, particularly of the infundibular type, poses some serious problems. In contrast, the presence of pulmonary stenosis is rather preferred in the Rastelli technique, because this method does not require relief of the pulmonary stenosis and, in addition, associated pulmonary stenosis has been known to protect the pulmonary vascular bed. Hence, the ideal candidate for this procedure is considered to be a patient with complete transposition of the great arteries, a large VSD, and pulmonary stenosis. Table 2 summarizes such cases encountered in the present series.

Rastelli stated that when there was no pulmonary stenosis present, repair could be carried out in children at about 1 year of age before the pulmonary obstructive disease progressed to 75% or higher of the pulmonary-systemic resistance ratio. In the present series five of the 22 patients without pulmonary stenosis survived 1 year or longer, ranging from 12 months to 11 years; the remaining 17 patients died earlier, most commonly by age 6 months or before.

It is also important to take into account the technical aspects of this procedure. It is clear that success of this procedure depends mainly on adequate construction of the left ventricular outflow. On the other hand, reconstruction of the right ventricular outflow with a homograft of aorta including the aortic valve has been, to date, routinely carried out for the treatment of certain cardiac anomalies. The present anatomic restudy showed that a VSD lying above the crista supraventricularis seemed to be most favorable for the technique, as it was not intimately related to the tricuspid valvar tissue which might otherwise interfere with the procedure. This type of defect is, however, very rarely encountered in the complete transposition complex. Elliott and associates exhibited a variety of anatomic arrangements of complete transposition of the great arteries with VSD, showing that the defect was located above the crista in only three of the 22 cases. Shafer and colleagues also reported three patients with the same type of defect who were found among 11 cases of complete transposition with pulmonary stenosis. This tendency was further emphasized in the present series.

The most common type of defect in this series was the defect posteroinferior to the crista supraventricularis including, in a larger sense, the muscular isolated or multiple defects. As a matter of fact, the technique is greatly concerned about the location of the defect as well as its adjacent structures, particularly the tricuspid valve. Therefore, it

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seems pertinent to distinguish the defects that were overhung by the papillary muscle of the conus from the defects that were not. The latter variety showed similar surgical merit to that observed in the defect lying above the crista supraventricularis. On the other hand, the tunnel formation appeared to be difficult or really impossible when the defect was overhung by the papillary muscle of the conus adherent to the anterior or superior border of the defect, unless it could be properly treated; this technique remains to be established.

On the basis of the theoretic concept, the following conclusions can be drawn from the present anatomic study of the 32 cases:

1. The technique seemed to be feasible in 13 cases; in particular, five of these would be ideal candidates because of the associated pulmonary stenosis. Despite the absence of pulmonary stenosis, two of the remaining eight patients were likely to be possible candidates, as the age of the patients was 1 year or so.

2. The other 19 patients were noticed to be poorly indicated for this technique because the VSD was obstructed by the valvular tissue or because there was no pulmonary stenosis and the patients were aged less than 1 year.

Angiocardiographic study of this pathologic entity has been fully performed, and quite reliable information is now available about its intracardiac anatomy.6,17 Shafer17 stated that selective (either right or left ventricular) angiocardiography can demonstrate the site and size of the defect reasonably well. The angiocardiographic evaluation of the site of the defect has certainly proved to be accurate, but the estimation of the defect size by angiography was not attempted in this study because (1) the lower margin of the defect is prone to be obscure, while its upper border is clearly defined in terms of the crista supraventricularis; and (2) McGoon and associates18 reported two patients in whom a small defect had been enlarged or an artificial defect had been created in the intact ventricular septum. Despite the unsuccessful outcome of those attempts, the technique is still believed to be worthwhile.

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