VENTRICULAR septal defect is a relatively common congenital cardiac defect. Rarely, however, is ventricular septal defect associated with major degrees of aortic valvular incompetence. When these two conditions do coexist, they are usually combined in a characteristic malformation. The purpose of this paper is to review our experience with the surgical management of this combination of defects, with special emphasis on the pathological anatomy, the techniques of repair, the results of treatment, and our present concepts regarding selection of patients for operation and choice of operative procedure.

Clinical Material

Up to October 1, 1962, operative repair had been attempted in 19 such patients at the Mayo Clinic. They ranged in age from 8 to 33 years, averaging 17.5 years. The majority of the patients were symptomatic, 10 being in class II and one in class III (New York Heart Association classification). Eight of the patients were asymptomatic. All patients, however, had cardiomegaly, electrocardiographic evidence of ventricular hypertrophy, and clinical and hemodynamic evidence of severe aortic valvular incompetence.

The clinical and hemodynamic aspects of some of these cases have been recorded in detail by Keek and associates; they have emphasized that mild or moderate infundibular pulmonary stenosis is commonly present also and that it may often be suspected on clinical grounds. Clinically, aortic valvular incompetence was usually the predominant hemodynamic defect. The ventricular septal defect was of lesser importance, the left-to-right shunt varying from 24 to 65 per cent and averaging 43 per cent.

Anatomy of the Defect

Ventricular Septal Defect

Special features of the ventricular septal defect in these patients are demonstrated in figure 1. It will be noted that, in contrast to the ordinary type of ventricular septal defect (fig. 1a), the aorta in these cases is not in connection with the ventricular septum at the site of the defect (fig. 1b). The defect characteristically lies beneath the right coronary cusp and adjacent portions of the noncoronary cusp, although some variation exists.

Defects of varying size have been encountered in our experience; they ranged from 0.4 to 2.0 cm in diameter. They were usually somewhat smaller than the more commonly encountered uncomplicated ventricular septal defects. The defects, of course, are high (immediately beneath the aortic valve), but they are often more anteriorly placed than is usual (fig. 2). In fact, four defects were located directly under the pulmonary valve. Only the larger defects extended posteriorly toward the tricuspid valvular ring. Most of the defects were located inferior to the crista supraventricularis. However, those few that were located just beneath the pulmonary valvular ring were so anteriorly placed as to be above the crista.

Aortic Valve

In every instance, the right coronary cusp was thickened, deformed, and prolapsed into and sometimes through the ventricular septal defect. It was an important feature of this condition that the distance from the base to the leading edge of this cusp was shortened in comparison with the distance in the normal cusps (fig. 2). The leading edge itself was elongated. In one instance, there was a perforation at the base of the right coronary cusp. In three cases, the noncoronary cusp was also partially involved; in another, all three cusps were thickened and deformed, probably as a result of the stresses imposed by long-standing aortic valvular incompetence. In one case was calcification present, and this involved the deformed prolapsing right coronary cusp.

Infundibular Pulmonary Stenosis

Varying degrees of hypertrophy of the septal band of the crista supraventricularis were noted.
at operation in nine cases. A pressure gradient ranging from 15 to 60 mm. Hg had been present across this region preoperatively in most of these patients.1 The anatomical defect was not a severe one and, with one possible exception, never approached in degree the severe malformation customarily encountered in cases of tetralogy of Fallot.

Operative Technique

All operations were carried out during whole-body perfusion with a Mayo-Gibbon pump oxygenator. Ischemic cardiome arrest at normothermia was employed in nine cases, potassium citrate arrest in two, ischemic arrest and external cardiome cooling in one, and intermittent coronary arterial perfusion in seven.

A transverse ventriculotomy is now preferred for exposure of the ventricular septal defect, which was closed directly with interrupted silk sutures in 14 instances (fig. 3). In five patients, the defect was closed with a patch (Teflon in three cases, Ivalon in one, and pericardium in one). The hypertrophied crista supraventricularis was excised in seven of the nine cases in which a pressure gradient had been present preoperatively.

An attempt was made to correct the aortic valvular incompetence in all but one case, and a variety of procedures was employed to this end. In four patients, an attempt was made to lengthen the distance between the leading edge of the right coronary cusp and its base (fig. 3a); in one of these, pericardium was sutured to the leading edge, while in three a patch (pericardium in two cases, Teflon in one case) was inserted after an incision was made near the base of the cusp. The leading edge was also shortened in two of these four patients. Being impressed by the fact that the leading edge of the right coronary cusp was measurably longer than that of its neighbors, we employed several methods in an attempt to shorten this elongated, prolapsing cusp edge in nine patients (fig. 3b). This included the creation, by suturing, of tucks in various portions of the leading edge; wedge excision of part of the leaflet with reapproximation by suturing; and suturing of the leading edge to the aortic wall or to adjacent cusps.

Insertion of a prosthesis was employed in five cases (fig. 3c, d, and e). In one, a tricuspid McGoon-type Teflon prosthesis was used, and in four the deformed right coronary cusp was resected and replaced by a Bahnson knitted-Teflon cusp.

Results

Two patients died after operation (table 1). One died on the day of operation, of severe uncorrected aortic valvular incompetence. An unsuccessful attempt had been made in this patient to restore competence by shortening the leading edge of the right coronary cusp by suturing it to the aortic wall. The second fatality followed replacement of the deformed right aortic leaflet by a Bahnson Teflon cusp. Aortic valvular incompetence was completely
relieved, but the patient died on the night of operation with progressive hyperthermia, restlessness, increasing bronchial secretions and, ultimately, coma. Death was considered to be related to the perfusion.

In addition to the two hospital deaths, there were two late deaths, both patients dying after reoperation for continued severe aortic valvular incompetence. At the first operation in each instance, an attempt had been made to enlarge the right aortic cusp by an advancement procedure. Pericardium was sutured to the leading edge of the cusp in one, and a Teflon patch was inserted into an incision at the base of the right coronary cusp in the other; in both, an attempt had also been made to shorten the leading edge with sutures.

Follow-up information is available on all of the surviving patients; 11 have been re-
examined at the clinic. The interval since operation varies from two months to five years, averaging a little more than two years. Information was sought as to the completeness of the closure of the ventricular septal defect in the 17 operative survivors. Re-examination 10 months after operation in one patient revealed a grade 3 (on the basis of 1 to 6) systolic murmur to the left of the sternum, as well as a slight increase in the pulmonary vascular markings. It is believed that a small left-to-right shunt persists in this patient. Recatheterization four months after operation in another patient disclosed a small residual left-to-right shunt (11 per cent). The ventricular septal defect in the remaining 15 patients is thought to be closed.

Careful postoperative evaluation of aortic valvular competence was made in all patients (table 2). The valve was considered competent when no diastolic murmur could be heard. When a diastolic murmur was audible, the incompetence was regarded as mild when the pulse pressure was less than 60 mm. Hg, moderate when a pulse pressure of 60 to 70 mm. Hg existed, and severe when the pulse pressure exceeded 70 mm. Hg. On this basis, severe aortic insufficiency was present postoperatively in eight patients, moderate insufficiency in three, and mild insufficiency in three. In only five was the aortic valve thought to be totally competent after operation. Severe incompetence persisted in the pa-
patient whose ventricular septal defect was closed but in whom no attempt was made to correct the associated lesion of the aortic valve. Incompetence of a mild degree persisted in one of four patients who underwent a procedure for advancement of the cusp, and of a moderate degree in one other; severe aortic valvular incompetence recurred three and five days after operation in the two remaining patients in whom this procedure was carried out.

Five patients of the nine in whom an attempt was made to shorten the leading edge of the right coronary cusp had severe aortic valvular incompetence after operation. In three of these, the valve seemed competent in the immediate postoperative period, but a diastolic murmur with a widening pulse pressure developed from three to seven days after operation. The incompetence was severe immediately after operation in the other two, and reference has already been made to one of these who died. Two patients in whom this procedure was performed had moderate incompetence after operation and one had mild incompetence. A competent aortic valve was restored in only one patient.

In contrast to the findings just mentioned, none of the five patients who had replacement of the cusp had severe incompetence. Mild incompetence was present in one, and the valve was completely competent after operation in four.

Severe persistent aortic valvular incompetence either directly or indirectly accounted for three of the four deaths; one patient failed to survive the initial operation, and the other two succumbed after reoperation. The other five patients with severe incompetence have remained well with few or no symptoms in spite of their defects. The remaining ten patients also are well without symptoms.

Thoracic roentgenograms were available for review in eight patients, and a decrease in the size of the cardiac silhouette was demonstrable in seven of these (fig. 4).

Comment

Ventricular septal defect, when associated with severe aortic valvular incompetence can be closed by direct suture, with rare exceptions. Postoperative heart block has not been seen. Although closure from above through the opened aortic valve, as reported by Garamella and associates,2 may rarely be feasible when the defect is small, the right ventricular approach is a more direct one and should permit more accurate closure. This approach will also permit accurate resection of the hypertrophied septal band of the crista supraventricularis when significant infundibular pulmonary stenosis coexists. At present, a transverse incision into the ventricle would be used.

Repair of the associated lesion of the aortic valve is a more difficult problem, as evidenced by residual incompetence of moderate or severe degree in 11 of 19 patients operated on by us by a variety of techniques. We have not found that simple closure of the ventricular septal defect, as advocated by some,3 will relieve the aortic incompetence; nor have cusp advancement procedures been successful in our hands.

The nature of the deformity of the right coronary cusp suggested to us, as it did to

Table 2

<table>
<thead>
<tr>
<th>Procedure on aortic valve</th>
<th>Number of patients</th>
<th>Postoperative aortic valve incompetence</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>None</td>
<td>Mild</td>
</tr>
<tr>
<td>None</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Cusp advancement</td>
<td>4</td>
<td>0</td>
</tr>
<tr>
<td>Leading edge shortened</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>Cusp replacement</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
<td>5</td>
</tr>
</tbody>
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Spencer and his associates,\(^4\) that a procedure designed to shorten the leading edge of this cusp might prevent prolapse and restore competency. In most instances this did not prove to be the case, whether the procedure consisted of taking tucks in the leading edge\(^4\) or suturing it to the aortic wall or to adjacent cusps.\(^5\) Either aortic valvular incompetence persisted after these procedures or it recurred early in the postoperative period, probably because the sutures tore out of the involved tissues.

The most effective means of correcting the

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Figure 4

Thoracic roentgenograms of two patients with ventricular septal defect and aortic valvular incompetence treated by direct suture of ventricular septal defect and prosthetic replacement of right coronary cusp. a. Before operation, and b. one and one-half months after operation in a 28-year-old man. c. Before operation, and d. 13 months after operation in a 19-year-old girl.
lesion of the aortic valve was by excision of the deformed cusp and its replacement with a prosthetic cusp.

Because of the apparent need for a prosthetic cusp in the repair of this combination of defects, it is now the policy at the Mayo Clinic to defer such an operation in growing children unless significant symptoms are present. When the patient is past the age of about 12 to 14 years, operation is advised if symptoms are present or if there is progressing or pronounced enlargement of the heart in the absence of symptoms. When operation is performed, it now consists of direct closure of the ventricular septal defect, resection of the hypertrophied septal band of the crista when indicated, and replacement of the deformed right coronary cusp of the aortic valve with a Bahnson Teflon cusp.

Summary
Nineteen patients with ventricular septal defect and severe aortic valvular incompetence have been operated on at the Mayo Clinic. There were two early deaths and two late deaths. The ventricular septal defect was located high and anteriorly in the septum and was often small. Aortic valvular incompetence was due, in most cases, to a deformed, prolapsing right coronary cusp. Mild to moderate degrees of infundibular pulmonary stenosis coexisted in nine patients.

Repair of the ventricular septal defect was readily accomplished in most cases by direct suture, and relief of obstruction to right ventricular outflow was effected, when required, by resection of the crista supraventricularis. Repair of the aortic incompetence was attempted by a variety of means, but the incidence of persistent significant regurgitation was high, except when prosthetic cusp replacement was used.

It is currently our policy to defer operation in children unless there are significant symptoms. When the patient is past the age of about 12 to 14 years, operation is advised under proper circumstances, and it consists generally of suturing of the ventricular septal defect and replacement of the right coronary cusp of the aortic valve.

References
Ventricular Septal Defect with Aortic Valvular Incompetence: Surgical Considerations
F. HENRY ELLIS, JR., PATRICK A. ONGLEY and JOHN W. KIRKLIN

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