

# Ventricular Septal Defect Associated with Aortic Insufficiency

## Anatomic Classification and Method of Operation

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### SUMMARY

Thirty-five consecutive patients with ventricular septal defect (VSD) associated with aortic insufficiency (AI) who underwent corrective surgery are presented. There were seven operative and three late deaths among the patients operated upon before 1968. No death, however, was encountered among the most recent 18 consecutive patients. The VSD was closed directly in 14 patients and with a Teflon patch in 21. The aortic valve was repaired in 16 patients, replaced in eight, and no interference was indicated in 11.

They were classified from the surgical viewpoint according to the location of the VSD, the anatomic type of the right ventricular outflow tract, and the severity of the aortic herniation as follows: type Ia, supracristal VSD and AI without aortic cusp herniation; type Ib, supracristal VSD and AI with aortic cusp herniation and conal muscular rim beneath the pulmonic valve; type Ic, supracristal VSD and AI with aortic cusp herniation without conal muscular rim beneath the pulmonic valve; type IIa, infracristal VSD and AI without aortic cusp herniation; type IIb, infracristal VSD and AI with aortic cusp herniation; type III, infracristal VSD and AI with infundibular pulmonic stenosis (PS); type IV, supracristal VSD and AI with infundibular PS.

In type Ia and IIa, VSD was closed directly and the aortic valve was replaced. In most of type Ib, VSD was closed directly and no direct procedure was performed upon the aortic valve. In most of type Ic, VSD was closed with a Teflon patch and the aortic valve was repaired. In type IIb, VSD was closed with a Teflon patch and the procedure upon the aortic valve was not uniform. In type III, VSD was closed with a Teflon patch and the aortic valve was repaired in most of them. In type IV, VSD was closed with a Teflon patch and no direct procedure was performed upon the aortic valve.

The basic policy for repair of this association of anomalies is selected according to the above mentioned anatomic classification.

### Additional Indexing Words:

Aortic cusp herniation

Conal muscular rim

Repair of aortic valve

Tetralogy of Fallot with aortic insufficiency

**K**NOWLEDGE concerning ventricular septal defect (VSD) associated with aortic insufficiency (AI) has advanced greatly in recent years. The clinical, hemodynamic, and pathologic findings have been clarified by many investigators.<sup>1-9</sup> The methods of operation for this association of the anomalies,<sup>4, 8-14</sup> however, are not uniform and are decided upon by the surgeon at the time of operation after cardiectomy.

The purposes of this report are: (1) to review the operative and autopsy findings in 35 patients; (2) to review the types of operations employed; (3) to classify the association of VSD and AI from the surgical viewpoint; and (4) to establish a policy of surgical treatment for this association in relation to the classification.

### Materials

Between September, 1958 and May 1972, 35 patients with the association of VSD and AI were operated upon in Osaka University Hospital and its affiliated hospitals. Twenty-eight males and seven females in this series ranged in age from 4 to 25 years (average 13.4 years). Twenty-three patients were symptomatic and the others were asymptomatic.

Diastolic murmur was noted for the first time during the period of preoperative observation in two patients.

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Received October 26, 1972; revision accepted for publication January 23, 1973.

In 33 other patients, a diastolic decrescendo murmur was present when they were first seen.

The electrocardiogram showed left ventricular hypertrophy in 12, right ventricular hypertrophy in one, and combined hypertrophy in 22 patients. The frontal plane axis varied from  $-4^{\circ}$  to  $+135^{\circ}$ .

Cardiac series X-ray showed left ventricular enlargement in all patients but one, and the cardiothoracic index varied from 47% to 70% (average 57.4%). Aortography was carried out in 21 patients. Grade 1 aortic regurgitation was assessed in four patients, grade 2 in five, grade 3 in seven, and grade 4 in five.

Cardiac catheterization was carried out in all patients except two, who clinically had tetralogy of Fallot associated with AI, and data in 32 patients were available. Mean pulmonary artery pressure ranged from 11 to 38 mmHg (average 21.1 mmHg) and was more than 25 mmHg in eight patients. A systolic pressure gradient of more than 10 mmHg was present in 13 patients at the right ventricular outflow tract. Maximum value of this gradient was 108 mmHg present in a 22-year-old male. Seven patients including two patients with tetralogy of Fallot had anatomic infundibular stenosis. In 27 patients without infundibular stenosis, the ratio of pulmonary-to-systemic flow ranged from 1.0 to 3.7 with an average of 1.78 and the ratio of pulmonary to systemic resistance ranged from 0.01 to 0.22 with an average of 0.09.

#### Classification

Hitherto, anatomic rather than functional classifications have been proposed. The authors divided the 35 patients into four major groups according to the location of the VSD and the stenosis of right ventricular outflow as follows: type I, supracristal VSD and AI; type II, infracristal VSD and AI; type III, infracristal VSD and AI with infundibular pulmonic stenosis (PS);

and type IV, supracristal VSD and AI with infundibular PS. Twenty-one patients (60%) were type I, seven patients (20%) were type II, five patients (14%) were type III, and two patients (6%) were type IV. Types I and II are divided into three and two subgroups, respectively.

*Type Ia.* Supracristal VSD and AI without aortic cusp herniation (fig. 1, left). In this type of defect, the cause of AI was not related to the existence of the VSD. This type was seen in only two patients. The size of the defect were small in these patients (5 mm in diameter). A bicuspid aortic valve was present in one and abnormal shortening of the right coronary cusp (RCC) in the other.

*Type Ib.* Supracristal VSD and AI with aortic cusp herniation and conal muscular rim beneath the pulmonic valve. As shown in figure 1 (middle) there was a thin rim of conal musculature beneath the pulmonic valve when seen from the right side through right ventriculotomy at the time of operation. Herniation of the aortic cusp was usually mild or moderate in this type. No significant pressure gradient was detected across the right ventricular outflow. The herniated cusp appeared to be the right coronary cusp (RCC) in all seven patients through right ventriculotomy and was confirmed through aortotomy in two patients.

*Type Ic.* Supracristal VSD and AI with aortic cusp herniation without a conal muscular rim beneath the pulmonic valve. In this type of defect, there was no conal musculature beneath the pulmonic valve and therefore, the aortic and pulmonic valves were in direct apposition as shown in figure 1 (right). The RCC herniated into the defect in all 12 patients. The degree of cusp herniation was moderate or severe in most of these patients. A systolic pressure gradient of 25 mmHg or more was present in three patients at the right ventricular outflow tract due to the obstruction resulting

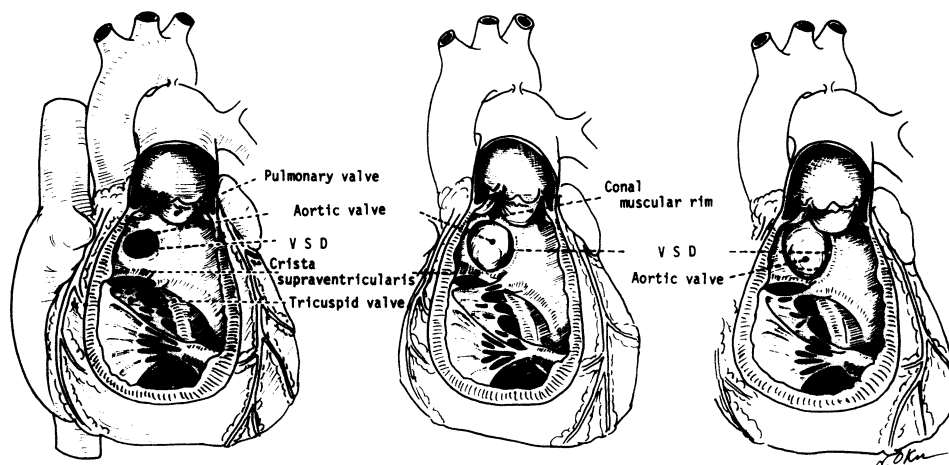
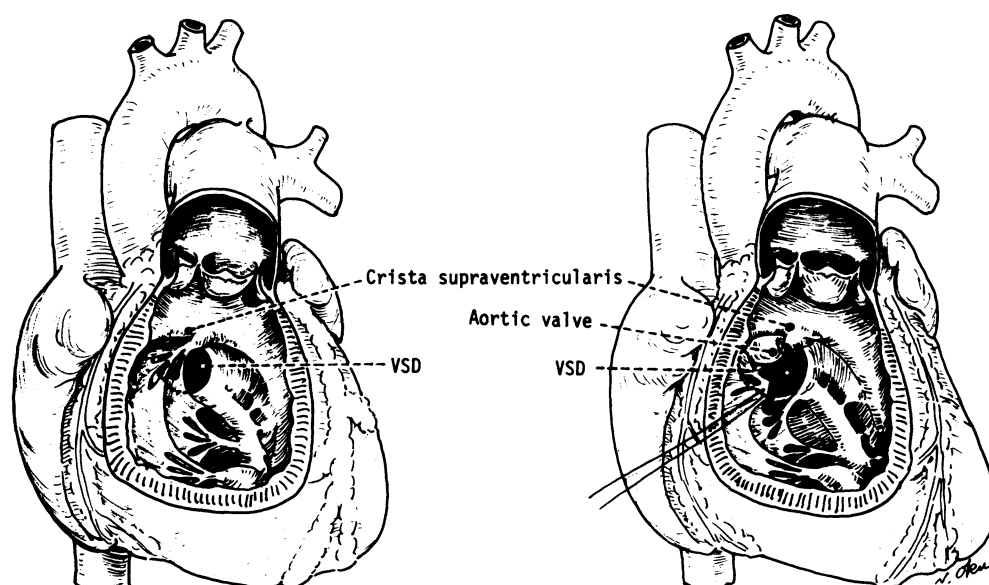


Figure 1

Right ventricular outflow through ventriculotomy in three types of patients. (Left) Type Ia: VSD is supracristal and there is no herniation of the aortic valve cusp. (Middle) Type Ib: VSD is supracristal. There is a thin conal muscular rim just beneath the pulmonic valve and the herniation of the aortic valve cusp is usually mild or moderate. (Right) Type Ic: VSD is supracristal. There is no conal muscle beneath the pulmonic valve. Aortic and pulmonic valves are side by side and the herniation of the aortic valve cusp is usually moderate or severe.



**Figure 2**

*Right ventricular outflow through ventriculotomy in two types of patients. (Left) Type IIa: VSD is infracristal and there is no herniation of the aortic valve cusp. (Right) Type IIb: VSD is infracristal and there is herniation of the aortic valve cusp.*

from herniation of the aortic cusp. In one out of nine patients who underwent aortotomy, a mild degree of commissural fusion was present between the RCC and the left coronary cusp (LCC), and, in two patients the size of the LCC was slightly smaller than that of the other two cusps.

**Type IIa.** Infracristal VSD and AI without aortic cusp herniation (fig. 2, left). In this type of defect, AI was not directly related to the existence of a VSD as it was in type Ia. In one of this group of two patients, the noncoronary cusp (NCC) was abnormally small and retracted. The size of the septal defects were 5 mm and 6 mm in diameter, respectively.

**Type IIb.** Infracristal VSD and AI with aortic cusp herniation (fig. 2, right). There were five patients with this type of defect. The RCC herniated in one and the NCC in two patients. In one patient who did not undergo aortotomy, the specific cusp herniated remained unknown. In the remaining patient with a herniated NCC, there was an anomalous cord between the RCC and LCC and the herniated NCC was short.

**Type III.** Infracristal VSD and AI with infundibular PS (fig. 3, left). There were five such patients. Infundibular stenosis was mild in one and moderate-to-severe in four resulting in a systolic pressure gradient of 56–108 mmHg. One patient had a two-chambered right ventricle. In three of the remaining patients, the anatomic configuration inside the right ventricle was similar to that seen in tetralogy of Fallot. The RCC was prolapsed in three patients, NCC in one, and both RCC and NCC in one.

The degree of cusp herniation was mild in all of these patients and seemed to be barely present in some. In one patient the NCC was particularly small and nodulus Arantii was not found. In the other patient, the

herniated cusp was RCC, while the NCC was extremely small and flat.

**Type IV.** Supracristal VSD and AI with infundibular PS (fig. 3, right). There were two such patients.<sup>15</sup> In both of them, there was severe infundibular and valvular stenosis. The pulmonic valves were bicuspid. The septal defects were 19 and 17 mm in diameter. Both were clinically tetralogy of Fallot with AI. The RCC was herniated into the VSD in both of them, one severely and the other slightly.

Among 25 patients who underwent aortotomy, eight patients were found to have an anomalous aortic cusp. Four of them were found among 14 patients with supracristal VSD and another four among the other 11 patients with infracristal VSD.

#### Method and Result of Operation

Figure 4 illustrates the operative method utilized for the present series of patients and the results of operation. Between 1958 and 1965, six patients were operated upon without opening the aorta except one who underwent aortic valve replacement. Only half of them survived. All of these patients had a mild-to-moderate degree of AI.

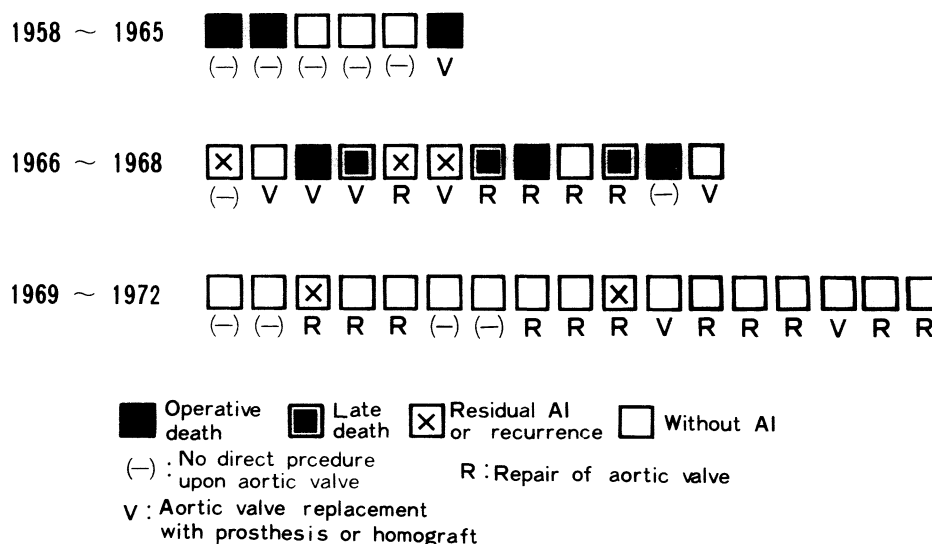
In most of the patients operated upon between 1966 and 1968, aortotomy was performed and either repair or replacement of the aortic valve was carried out. Repair of the aortic valve was performed in five patients with one operative death. AI recurred in three patients who survived operation and two late deaths resulted. Aortic valve replacement was carried out in five patients with one operative and one late death. In two patients without direct surgical procedure upon the aortic valve, one survived the persistent AI and the other succumbed.

**Right ventricular outflow through ventriculotomy in two types of patients. (Left) Type III: VSD is infracrystal and there is infundibular stenosis. (Right) Type IV: VSD is supracrystal and there is infundibular stenosis.**

summarized in figure 5 in relation to the classification described. Postoperative aortic valve competency was evaluated mostly by auscultation.

### Method of Closure of VSD

The VSD in type Ia and IIa was small and was closed directly without patching. Although the VSD found in type Ib patients was larger than that found in type Ia or IIa patients, it was possible to close the defect in most of them without using a patch as a muscular rim was present beneath the pulmonic valve.



*Surgical procedures performed upon the aortic valve and results of operation in 35 patients with association of ventricular septal defect and aortic insufficiency.*

Method of operation Classification		V S D		A I		
		Direct closure	Patch closure	None	Repair	Replace-ment
Ia	Supracristal VSD without aortic herniation	●●				●●
Ib	Supracristal VSD with aortic herniation and conal muscular rim	●●●●●	●	●●●●●	●●	
Ic	Supracristal VSD with aortic herniation without conal muscular rim	●●●●	●●●●●	●●●	●●●●●	●
IIa	Infracristal VSD without aortic herniation	●●				●●
IIb	Infracristal VSD with aortic herniation		●●●●●	●	●●	●●
III	Infracristal VSD and infundibular PS		●●●●●		●●●●	●
IV	supracristal VSD and infundibular PS		●●	●●		

Figure 5

*Anatomic classification and surgical procedures utilized in 35 patients for the closure of ventricular septal defect and for the treatment of insufficient aortic valve.*

In these patients, utilization of a patch for the closure of the VSD was purposefully avoided as the direct closure of the VSD was considered to be much more effective than patch closure in pushing back the herniated RCC to the left ventricular side.

The VSD in type Ic patients was sometimes functionally small as it was partially occluded by the herniated aortic cusp. The anatomic defect, however, was usually large. There was no firm tissue in which to put the sutures above the defect except that of the

pulmonic valve ring. It seemed dangerous to approximate the pulmonic ring to the lower margin of the defect as it might have resulted in later tearing of the tissue and subsequent recurrence of the defect. The defect was, therefore, closed with a Teflon patch except in four patients who had small defects. The method of closure is illustrated in figure 6. Interrupted mattress sutures with small pledgets were placed all around the defect. Along the upper margin of the defect, these sutures were placed from the inside of the pulmonic

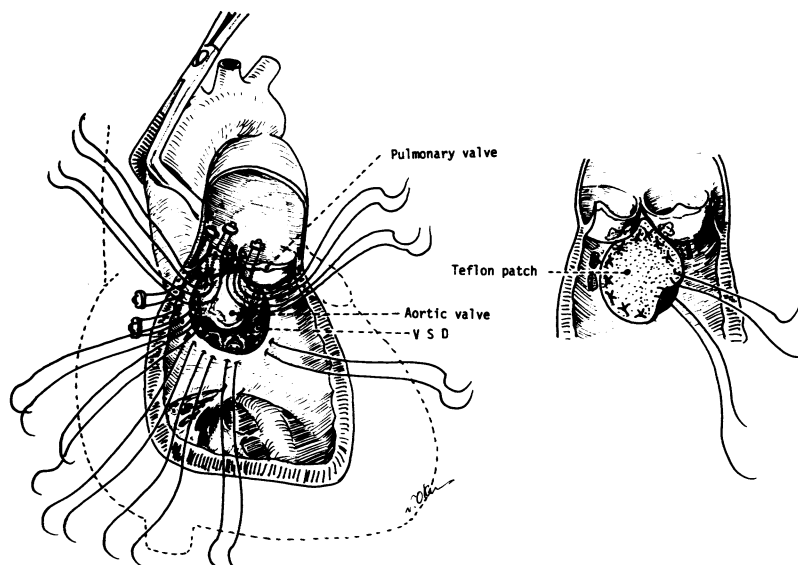


Figure 6

*Surgical repair utilized for the closure of ventricular septal defect in type Ic patients. Along the upper margin of the defect, sutures were placed from inside of the pulmonic valve cusps.*

cusps. The anatomic defect in the septum was then closed utilizing a patch of Teflon.

#### *Management of the Insufficient Aortic Valve*

As there was no aortic cusp herniation in type Ia and IIa patients before surgery, closure of the VSD did not result in any beneficial effect upon these insufficient aortic valves. In one of these four patients, the aortic valve was bicuspid and in the other the NCC was small and retracted. Repair of these valves was obviously impossible and they were replaced. The remaining two patients were operated upon in the early period of this study and the aortic valves were replaced. It may be feasible to repair these aortic valves at the present time.

In type Ib patients AI was usually mild and could be controlled by closing the defect without a patch, as the herniated cusp was pushed back to the left ventricular side. In two patients with large VSD of 15 and 16 mm in diameter, respectively, repair of the aortic valve was necessary although the defect was closed directly. In the remaining five patients, AI disappeared without a direct surgical procedure upon the valve.

In most of type Ic patients AI was severe. As it was necessary to use a patch of Teflon for the closure of the VSD, AI did not disappear unless a direct surgical procedure was performed upon the valve. Among eight patients who underwent patch closure of the VSD, the aortic valve was repaired in seven and replaced in one. On the other hand, among four patients who underwent direct closure of the VSD, the aortic valve was repaired in one and was left untreated in three. In two of the latter insufficiency disappeared, but in the third residual insufficiency persisted. As aortotomy was not performed in this patient, it was not clear whether there was a deformity of the valve which might have resulted in residual insufficiency.

In type IIb, III, and IV patients, the VSD was large in almost all of them but the degree of aortic cusp herniation was mild. The role of cusp herniation in the development of AI seems to be less important in these types of patients than in type Ic patients. It seems, therefore, difficult to reduce or eliminate the AI by closure of the defect. Among five type IIb patients, repair was done in three and replacement in one. In another with small VSD of 7 mm in diameter, no direct procedure on the aortic valve was necessary.

Among five type III patients, repair was done in four and replacement in one. In one patient, the aortic valve was so severely deformed, as mentioned before, that it was impossible to eliminate the insufficiency with conservative means. This patient was a 10-year-old girl and her aortic ring was too narrow for the aortic valve replacement that repair was performed, even though it was obviously inadequate.

The method of aortic valve repair the authors have been using since 1969 is that described by Frater<sup>16</sup> in 1967. However, it is not the purpose of this report to describe the method of aortic valve repair in detail.

#### **Discussion**

Keck and his co-workers<sup>2</sup> divided the patients with this association of anomalies into two groups

according to the presence or absence of infundibular PS. Nadas,<sup>3</sup> Van Praagh,<sup>6</sup> and their co-workers proposed a classification of this association of anomalies according to the location of the VSD. The latter further divided the patients with infracristal VSD into two subtypes according to the presence or absence of infundibular PS. Sakakibara and Konno<sup>17</sup> placed this association in the category of aneurysm of the sinus of Valsalva and divided patients with these anomalies into four types regardless of the presence or absence of infundibular PS. Their type I-VSD and type II-VSD correspond to type Ic and Ib, respectively. Their type III-VSD and IV-VSD seem to be included in type IIb according to the present classification.

It is well known that the defect of the ventricular septum is more frequently found above the crista supraventricularis in Japanese literature<sup>18</sup> than is described in the English literature. It is the same among patients with VSD associated with AI.<sup>17</sup>

Van Praagh and McNamara<sup>6</sup> reported three cases with infracristal VSD and AI without infundibular PS. It is quite different from the present series of patients in that all three of their cases seem to have belonged to type IIa and none to type IIb. All these three cases had been reported to have had bicuspid valves but none of type IIa patients in the present series had it.

Two type IV patients had tetralogy of Fallot associated with AI<sup>15</sup> and these two cases were encountered among 243 cases of tetralogy of Fallot who underwent corrective surgery.

It has been the author's policy in the surgical treatment of VSD and AI to close the defect directly when possible and this seemed to reduce or eliminate the AI by pushing back the herniated cusp to the left ventricular side. When it was impossible to do so, the authors did repair the valve but avoided replacing the valve as far as possible.

Up to the present time, there has been no clarification as to which kind of valve should be repaired and which kind of valve should be replaced. It was not possible, therefore, to decide on the specific operative procedure before cardiectomy. As the present anatomic classification the authors proposed was made from the surgical viewpoint, the operative procedures to be undertaken could roughly be predicted before operation. Table 1 summarizes the proposed surgical procedure for the respective types of VSD and AI based on the present classification of anatomic type.

The method used to close the VSD in types Ia and IIa is quite the same as that used for simple small

**Table 1**

*Proposed Surgical Procedures for the Respective Types of Ventricular Septal Defect and Aortic Insufficiency Based on the Present Classification of Anatomic Type*

Type	Ventricular septal defect (closure)	Aortic insufficiency
Ia	Direct	Repair or replacement
Ib	Direct	None
Ic	Patch	Repair
IIa	Direct	Repair or replacement
IIb	Patch	Repair or replacement
III	Patch	Repair or replacement
IV	Patch	None, repair or replacement

VSD. For types Ib and Ic, the authors are convinced that the closure of the VSD without patching is much more apt to push back the herniated aortic cusp into the left ventricle as described by Van Praagh and McNamara.<sup>6</sup> In most cases of types Ib and Ic with relatively small VSD, AI disappears after direct closure of VSD. In cases where patch closure is necessary, the technic of closing the VSD in type IIb and in type III is not of particular importance. In patients with type Ic and in some patients with type IV, however, particular caution is usually necessary, as mentioned above, in affixing the patch to the cranial margin of the defect. The method described is the one that the authors utilize for the closure of VSD located immediately beneath the pulmonic valve regardless of the presence of AI.

Recurrence of AI after repair of the prolapsed cusps was encountered frequently in the early cases of the present series. However, since utilizing the method described by Frater<sup>16</sup> the incidence of recurrence has decreased significantly. When aortic valve replacement is inevitable, it is important to prepare a firm foundation for anchoring the prosthetic or homograft valve. Twisting of the suture line should be meticulously avoided particularly when a homograft aortic valve is implanted without a mounting frame. Some investigators<sup>8,14</sup> recommended closure of the VSD with the aortic cusp or reinforcing the closure with the aortic cusp. The authors also prefer to use the remnant of the excised cusp for reinforcement of the closure of the VSD. This facilitates a sound implanting of the aortic valve.

The differentiation of these anatomic types is usually possible by means of aortography and left ventriculography except in those cases between types Ib and Ic with mild or moderate herniation of the RCC. The details of the differentiation will be

discussed in another paper. There are different opinions on the timing of operation for this association of the anomalies as the repair of the aortic valve is not always successful. However, the pathologic change of the aortic cusp seems to be progressive and the aortic regurgitation is possible to be repaired without the aortic valve replacement when operated upon in early stage though it also depends on the anatomic type. Preoperative diagnosis of the anatomic types of this association of anomalies is, therefore, of practical importance in deciding the timing of operation.

### References

1. SCOTT PC, MCGUIRE T, KAPLAN S, FOROLER NO, GREEN RS, GORDON LZ, SHAHETAI R, DAVOLOS DD: The syndrome of ventricular septal defect, with aortic insufficiency. *Amer J Cardiol* 2: 530, 1958
2. KECK EWO, ONGLEY PA, KINCAID OW, SWAN HJC: Ventricular septal defect with aortic insufficiency: A clinical and hemodynamic study of 18 proved cases. *Circulation* 27: 203, 1963
3. NADAS AS, THILENIUS OG, LA FARGE CG, HAUCK AJ: Ventricular septal defect with aortic regurgitation: Medical and pathological aspects. *Circulation* 29: 862, 1964
4. PLAUTH WH, BRAUNWALD E, ROCKOFF SD, MASON DT, MORROW AG: Ventricular septal defect and aortic regurgitation: Clinical, hemodynamic and surgical considerations. *Amer J Med* 39: 552, 1965
5. HALLORAN KH, TALNER NS, BROWNE MJ: A study of ventricular septal defect associated with aortic insufficiency. *Amer Heart J* 69: 320, 1965
6. VAN PRAAGH R, MCNAMARA JJ: Anatomic types of ventricular septal defect with aortic insufficiency: A diagnostic and surgical consideration. *Amer Heart J* 75: 604, 1968
7. BLUMENTHAL S, KANGOS JK, BOWMAN FO, MALM JR: Ventricular septal defect with prolapse of the aortic cusp. (Abstr) *Circulation* 28: 691, 1963
8. SOMERVILLE J, BRANDO A, ROSS DN: Aortic regurgitation with ventricular septal defect: Surgical management and clinical features. *Circulation* 41: 317, 1970
9. HALLIDIE-SMITH KA, OLSEN EGJ, OAKLEY CM, GOODWIN JF, CLELAND WP: Ventricular septal defect and aortic regurgitation. *Thorax* 24: 257, 1969
10. STARR A, MENASHE V, DOTTER C: Surgical correction of aortic insufficiency associated with ventricular septal defect. *Surg Gyn Obstet* 111: 71, 1960
11. SPENCER FC, BALMSON HT, NEILL CA: The treatment of aortic regurgitation associated with ventricular septal defect. *J Thorac Cardiovasc Surg* 43: 222, 1962
12. ROBINSON G, FELL SC, JACOBSON BE: Ventricular septal defect with aortic insufficiency: A method of management. *J Thorac Cardiovasc Surg* 43: 785, 1962

13. ELLIS H JR, ONGLEY PA, KIRKLIN JW: Ventricular septal defect with aortic valvular incompetence: Surgical considerations. *Circulation* 27: 789, 1963
14. GONZALEZ-LAVIN L, BARRATT-BOYES BG: Surgical considerations in the treatment of ventricular septal defect associated with aortic valve incompetence. *J Thorac Cardiovasc Surg* 57: 422, 1969
15. MATSUDA H, KAWASHIMA Y, FUJITA T, MIYAMOTO T, NAITO Y, MORI T, NOMURA N, HORIGUCHI Y, MANABE H: Tetralogy of Fallot associated with aortic insufficiency. *Heart* 4: 437, 1972 (In Japanese)
16. FRATER RWM: The prolapsing aortic cusp: Experimental and clinical observations. *Ann Thorac Surg* 3: 63, 1967
17. SAKAKIBARA S, KONNO S: Congenital aneurysm of the sinus of Valsalva associated with ventricular septal defect. *Amer Heart J* 75: 595, 1968
18. KAWASHIMA Y, FUJITA T, UEDA T, CHIKAMORI J, NAITO Y, MORI M, NISHIZAKI K, MANABE H: Anatomical classification of Tetralogy of Fallot. *J Jap Assoc Thorac Surg* 17: 1006, 1969 (In Japanese)