

PEDIATRIC CARDIOLOGY

Ventricular Septal Defects: A Surgical Viewpoint

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Seventy-six cardiac autopsy specimens with ventricular septal defects were studied from a surgical viewpoint. The defects were classified as being conoventricular (n = 25), in the right ventricular outlet (n = 21), in the inlet septum (n = 11) or in the trabecular septum (n = 19) with each category having several subcategories. The left ventricular

outflow tract relations of the defects are emphasized. The borders of the ventricular septal defects are described in detail, with use of the prefix "juxta" to indicate the immediate adjacency of the defect to a structure such as the tricuspid valve.

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Ventricular septal defects abutting the anulus of the tricuspid valve in the region of its anteroseptal commissure were found in the late 1950s to require repair in a particular way to avoid damaging the His bundle, and surgical experiences in the beating heart generated the inference that the His bundle coursed along the posteroinferior margin of such defects (1-3). At about this time (4), morphologic studies in autopsy hearts began to categorize the various locations of holes in the ventricular septum, and many such studies have since appeared. Most of these subsequent studies generated new insights for surgeons, new terminologies and new controversies. The relevance of morphologic studies to surgical problems has become greater over the years and, at the same time, the number of surgical studies of morphology and inferences from them have increased. An earlier bias against basing surgical descriptions of morphology on embryology has lessened as both the surgical inferences and embryologic knowledge have become more secure.

This study was undertaken from a surgical point of view but with the intent of providing information that is morphologically correct and relevant to imaging techniques as well as cardiac surgery.

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Methods

Autopsy material. The study material comprised the 76 hearts in the autopsy collection at the University of Alabama with a ventricular septal defect but without other important cardiac anomalies (except 1 with coexisting aortic arch interruption) and with atrial situs solitus, ventricular D-loop and atrioventricular (AV) and ventriculoarterial (VA) concordant connections. These hearts were restudied anatomically.

Normal anatomy. Descriptors of normal anatomy are used for structures that are near the ventricular septal defect, even though these structures are at least subtly abnormal in most hearts with a large defect. To assist in visualizing and understanding the normal anatomy, the right ventricular aspect of the ventricular septum of a normal heart was displayed by two longitudinal transections in the coronal plane. One transection was made just anterior to the aortic root (Fig. 1), and the other was made through the aortic root (Fig. 2). Of course, not all of the internal surface of the right ventricle is ventricular septum. In addition to the inner surface of the obvious free wall anteriorly, that part of the internal surface beneath the posterior tricuspid leaflet is also inner surface of the free wall of the right ventricle (Fig. 2). The inlet septum is between the tricuspid septal and mitral septal (anterior) leaflets and is in the inlet portion of both ventricles. The rather large *trabecular septum* extends to the apex and also to the pulmonary valve anterosuperiorly. The *septum of the outlet portion* of the right ventricle is particularly complex and consists of the conal septum, the left anterior division of the septal band and anterosuperiorly of a tongue of trabecular septum. In its most distal portion, the septum of the right ventricular outlet is not interventric-

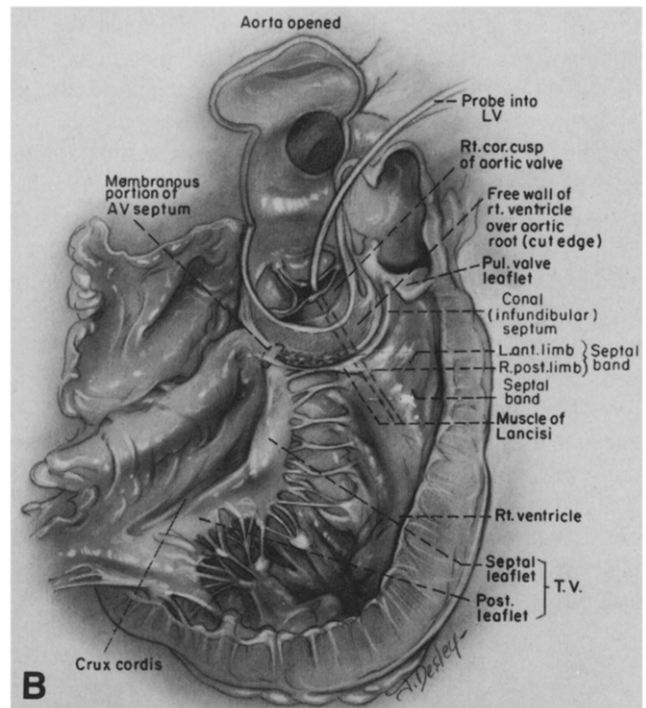
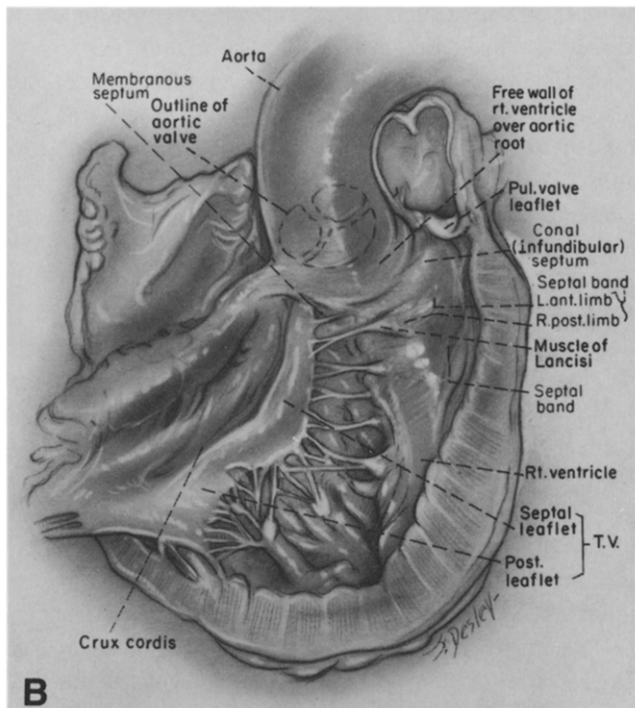
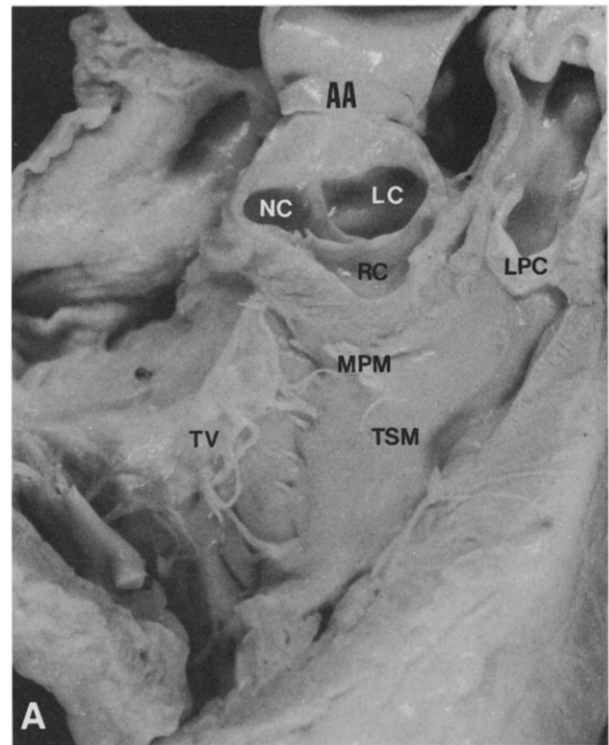
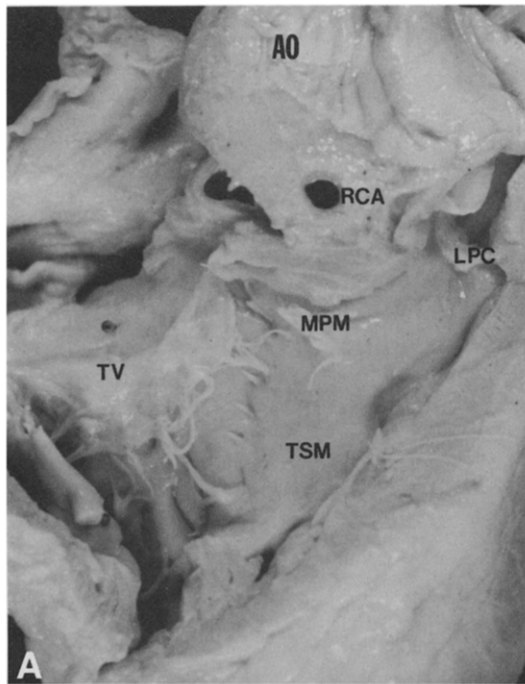


Figure 1. Longitudinal (coronal) section through a normal heart passing just anterior to the aortic root (Ao). **A**, Photograph and **B**, Artist's depiction. L. ant. = left anterior; LPC = left pulmonary valve cusp; MPM = medial papillary muscle (muscle of Lancisi); RCA = central stump of right coronary artery; Rt or rt = right; Post = posterior; Pul = pulmonary; TSM = trabecula septomarginalis (septal band); TV = tricuspid valve.

Figure 2. A more posterior longitudinal coronal section through the same heart shown in Figure 1, this section passing through the mid portion of the aortic root. **A**, Photograph; **B**, Artist's depiction. AA = ascending aorta; AV = atrioventricular; LC = left coronary cusp of aortic valve; LPC = left pulmonary valve cusp; LV = left ventricle; MPM = medial papillary muscle (muscle of Lancisi); NC = noncoronary cusp of aortic valve; RC = right coronary cusp of aortic valve; other abbreviations as in Figure 1.

Table 1. Summary of the Ventricular Septal Defects in 76 Hearts

Category	n
Conoventricular VSD	25
VSD in RV outlet	21
Juxtaaortic	4
Juxtapulmonary	1
Juxtaarterial	5
Muscular in the conal septum	10
Muscular in the trabecular septum (anterosuperior portion)	1
Inlet septal VSD	11
Juxtatricuspid and juxtamitral	6
Juxtacrucial	1
Muscular	4
Trabecular VSD	19
Anterior	13
Midseptal	6
Apical	—

RV = right ventricular; VSD = ventricular septal defect. A complete description of the heart with a ventricular septal defect also includes the size of the defect, any septal or valvular malalignment and the commitment of the defect to the great arteries (subaortic, subpulmonary, doubly committed or noncommitted). Coexisting cardiac anomalies, as well as the atrial and ventricular situs, the positions of the great arteries and the atrioventricular and ventriculoarterial connections are also described.

ular but is bordered on the other side by the transverse sinus of the pericardium.

Categorization of ventricular septal defects. For the purposes of this study, four categories of defects were used (Table 1). *Conoventricular* (infundibuloventricular or junctional) defects are in the junctional area between the trabecular (sinus) and conal portions of the ventricular septum and usually appear to be between the divisions of the septal band (5,6). Such defects are usually, but not always, associated with abnormalities of the conal septum. Conoventricular defects are between the inlet and outlet portion of the

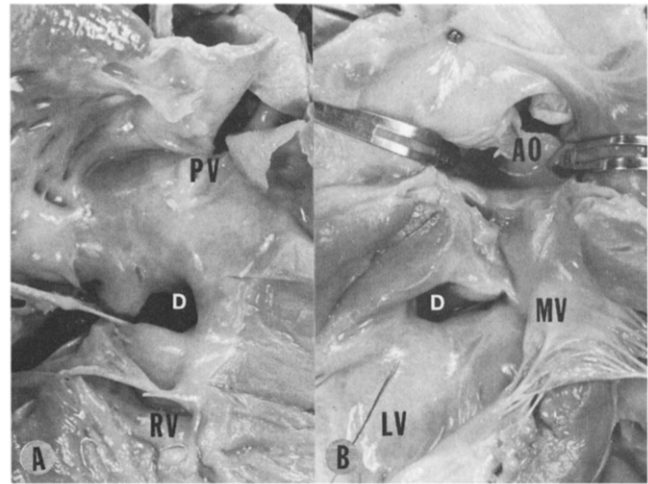
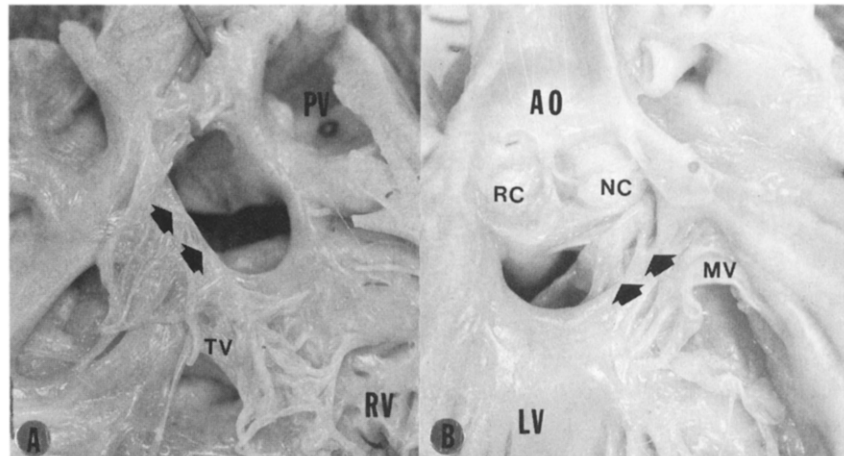


Figure 4. Moderate-sized conoventricular ventricular septal defect (D), viewed from the right ventricle (RV) in (A) and the left ventricle (LV) in (B). The defect is juxtatricuspid and almost juxtamitral but is not juxtaaortic. The conal septum is not hypoplastic but is malaligned posteriorly and to the left and is obvious beneath the aortic valve when viewed from the left ventricular aspect. The defect is not perimembranous because it is completely separate from the membranous septum and is not juxtaaortic. AV = aortic valve; MV = mitral valve; PV = pulmonary valve; other abbreviations as in Figures 2 and 3.

right ventricle but are in the outlet portion of the left ventricle. *Defects in the right ventricular outlet* are either immediately adjacent to a semilunar valve or have muscular borders and are in the conal septum or in the anterosuperior tongue of the trabecular septum. Such defects are in the outlet portion of both the right and the left ventricle. *Inlet septal defects* are either immediately adjacent to the AV valves or are muscular defects. Inlet septal defects are in the

Figure 3. Large conoventricular perimembranous ventricular septal defect, viewed from the right ventricle (RV) in (A) and from the left ventricle (LV) in (B). In (A), the septal tricuspid leaflet was folded back into the right atrium after its chordae were cut. The arrows indicate the areas of tricuspid-mitral continuity, and the defect is juxtatricuspid, juxtamitral, and juxtaaortic. This defect, viewed from the right ventricle, lies far to the right in the aortic margin of the ventricular septum (see **stippled area** in B) and thus is beneath the commissure between noncoronary (NC) and right coronary (RC) cusps. The defect is then termed perimembranous, and no muscle is between it and the anterior-septal tricuspid commissure. The conal septum is hypoplastic but not malaligned. MV = mitral valve; PV = pulmonary valve; TV = tricuspid valve; other abbreviations as in Figure 1.



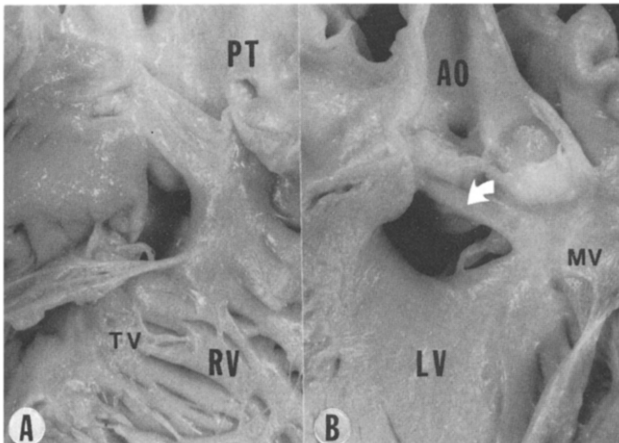


Figure 5. Conoventricular septal defect, which coexisted with interrupted aortic (Ao) arch, viewed from the right ventricular (RV) (A) and left ventricular (LV) aspects (B). The defect is juxtatricuspid, in the region of the commissure between the septal and anterior leaflets. It is close to the mitral valve (MV) but is not juxtamitral. It is not juxtaaortic, related to the bifid configuration of the somewhat hypoplastic conal septum (indicated by arrows), one portion of which is malaligned and beneath the aortic valve (B). The defect lies a little more to the left (as viewed from the right ventricle) in the aortic margin of the ventricular septum, and thus is mostly beneath the right coronary cusp. PT = pulmonary trunk; other abbreviations as in Figures 1 to 3.

inlet portion of both the right and the left ventricle. *Trabecular defects* are muscular and bordered entirely by trabecular septum or by trabecular septum and anterior free wall. Ventricular septal defects that are so large as to be in several areas are termed *confluent*, but such defects were not found in the present study.

Terminology. The prefix "juxta" was used to indicate the defect's immediate adjacency to a structure; in other words, immediately adjacent to or bordered by the structure. The adjective "perimembranous" was used for defects that 1) were immediately adjacent to and bordered by the tricuspid anulus in the region of the commissure between septal and anterior leaflets, and 2) were also juxtaaortic and juxtamitral and adjacent to the central fibrous body and membranous septum. The penetrating portion of the His bundle is along the posteroinferior margin of perimembranous defects (7), but it is also along this margin in other defects that are juxtatricuspid but not perimembranous.

The aortic margin of the ventricular septum is that part of the septum immediately upstream to the aortic valve as viewed from the left ventricle. As viewed from the right ventricle, it stretches from the region of the membranous septum (and thus from the tricuspid anulus in the region of the anteroseptal leaflet commissure) to the leftward and superior aspect of the conal septum (Fig. 2).

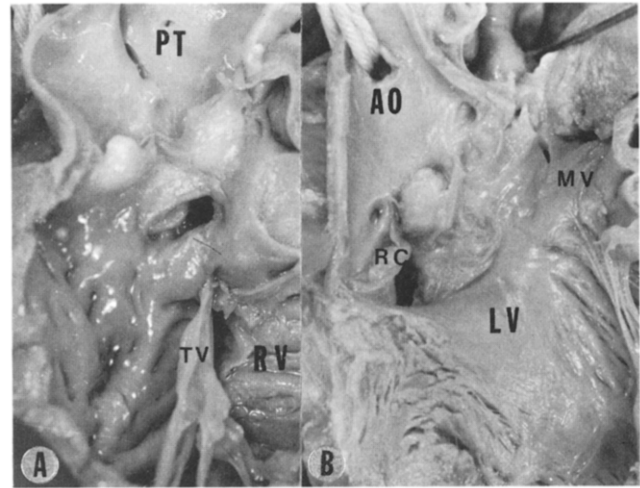


Figure 6. Conal (infundibular) septal defect in the right ventricular outlet, in the middle third of the conal septum, viewed from right ventricular (RV) (A) and left ventricular (LV) (B) aspects. The defect is not juxtatricuspid (and not perimembranous) nor is it juxtapulmonary, being separated from the pulmonary valve by the distal remnant of conal septum. The defect is juxtaaortic and lies beneath the right coronary cusp (RC), which can be seen to be somewhat prolapsing behind the remnant of conal septum (A). Abbreviations as in Figures 1 to 3 and 5.

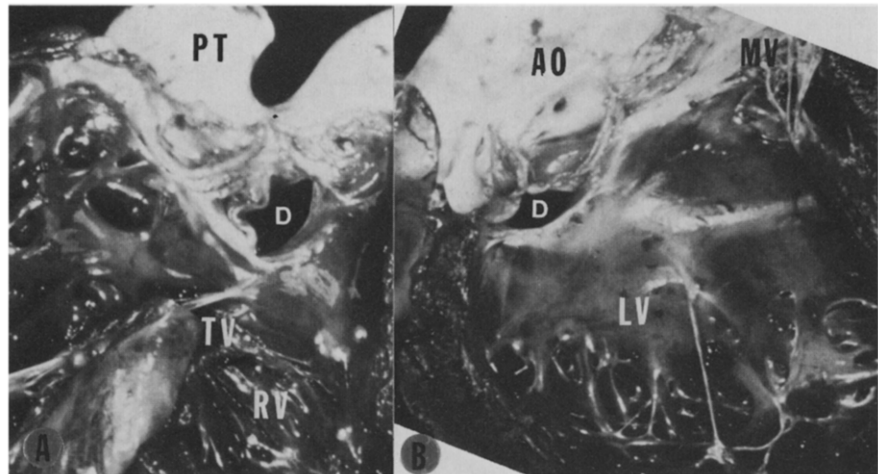
Results

Conoventricular ventricular septal defects. Twenty-five hearts had a conoventricular defect lying in the midportion of the right ventricular aspect of the ventricular septum and in the outflow portion of the left ventricular aspect of the septum. Some, but by no means all, conoventricular defects were juxtatricuspid, juxtamitral and juxtaaortic (Fig. 3), and hypoplasia of the conal septum was frequently associated with such defects. Such defects were also perimembranous. Some conoventricular defects were not juxtatricuspid, but instead had a bar of muscle (the right posterior division of the septal band) of variable width forming the posterior border of the defect.

Some conoventricular ventricular septal defects were juxtatricuspid, nearly but not quite juxtamitral and not juxtaaortic and, therefore, were not perimembranous defects. Some of these were associated with posterior and leftward malalignment of a portion of the conal septum (Fig. 4). At operation, it may not be possible to determine that such a defect is not perimembranous, but it can easily be determined that it is juxtatricuspid and thus that the His bundle is along the posteroinferior margin of the defect.

Conoventricular defects often were beneath but not necessarily juxtaposed to the commissure between noncoronary and right coronary cusps of the aortic valve; but some were slightly more leftward (when viewed from the right ventricle) and were more beneath the right aortic cusp than beneath the

Figure 7. Conal septal juxtaarterial ventricular septal defect (D), viewed in (A) and (B) as before. The defect is juxtaarterial. It is immediately beneath the pulmonary valve (A) and the aortic valve (B) and these are separated by only a fibrous raphe. The defect is neither juxtatricuspid nor juxtamitral and is far anterior to the membranous septum, central fibrous body and His bundle. Note that two artifacts in the pulmonary trunk (PT) (A) mimic coronary ostia. Abbreviations as in previous figures.



commissure between it and the noncoronary cusp (when viewed from the left ventricle) (Fig. 5).

Ventricular septal defects in the right ventricular outlet. Twenty-one hearts had a defect that was considered to lie in the right ventricular outlet and in the outlet portion of the septum when viewed from the left ventricular side. Some were *juxtaaortic* and beneath the right aortic cusp but separated from the pulmonary valve by the distal remnant of the conal septum (Fig. 6). The right aortic cusp prolapsed through some ventricular septal defects in this location.

Some defects in this category were both *juxtaaortic* and *juxtapulmonary* and, therefore, were termed *juxtaarterial* (8) (Fig. 7). The two valves were generally separated by a narrow fibrous band, and the aorta was a little more anteriorly situated than normal. Purely *juxtapulmonary* defects occurred but were uncommon.

A defect with completely muscular borders was sometimes present in the midportion of the conal septum. Rarely, a defect in the right ventricular outlet had muscular borders

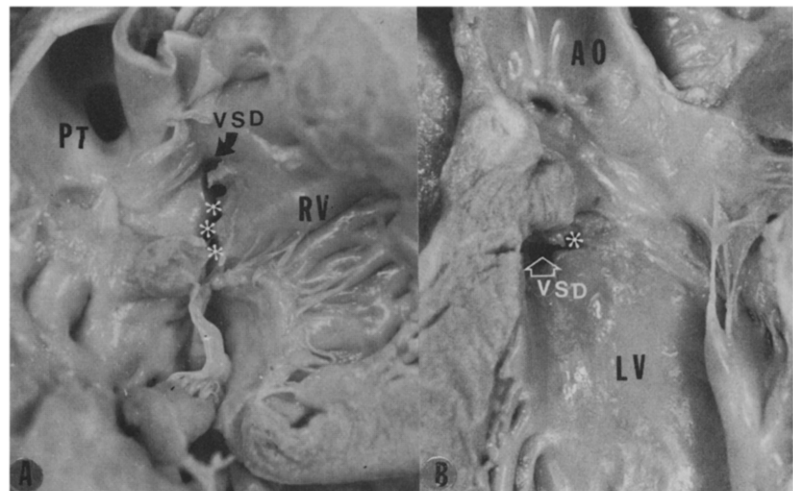
but lay very anteriorly in the anterosuperior tongue of the trabecular septum (Fig. 8).

Inlet septal ventricular septal defects. Less commonly, in the group of hearts studied, there was a defect in the inlet portion of the ventricular septum. Usually it was juxtatricuspid and juxtamitral (Fig. 9) and sometimes perimembranous. The phrase "AV canal type of ventricular septal defect" has been avoided because of possible confusion between such a defect and some subsets of AV canal defects, specifically those with only an interventricular communication (9). A few defects in the inlet septum had purely muscular borders.

Rarely, an inlet septal defect extended to the crux cordis and, therefore, was juxtacrucial as well as juxtamitral and juxtatricuspid (Fig. 10). Malalignment of the atrial to the ventricular septum accompanied such a defect.

Trabecular ventricular septal defects. These were found in the usual locations (Table 1).

Figure 8. A muscular ventricular septal defect (VSD) in the anterior superior tongue of the trabecular septum. This small defect is almost juxtapulmonary but not quite and is not juxtaaortic. The jagged opening extending down toward the tricuspid valve (covered by the asterisks) is artifactual. The very anterior position of the defect is particularly evident in the view from the left ventricle (LV) (B). Abbreviations as in previous figures.



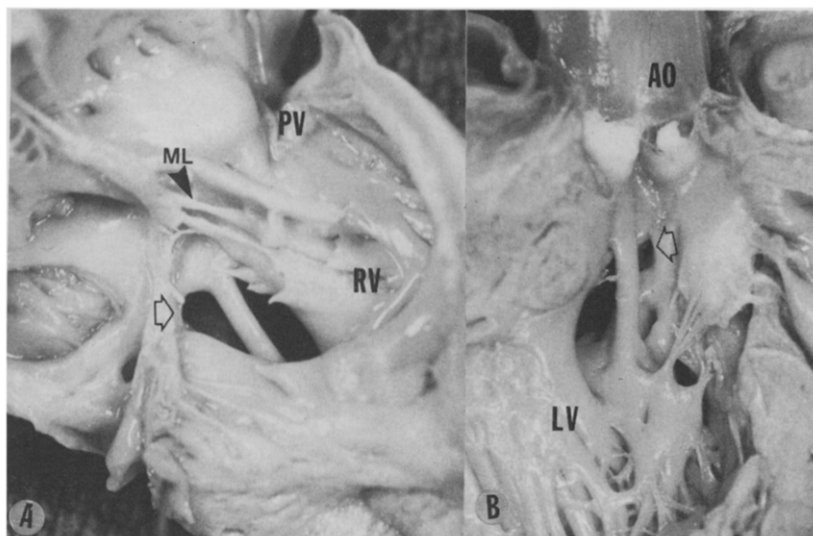
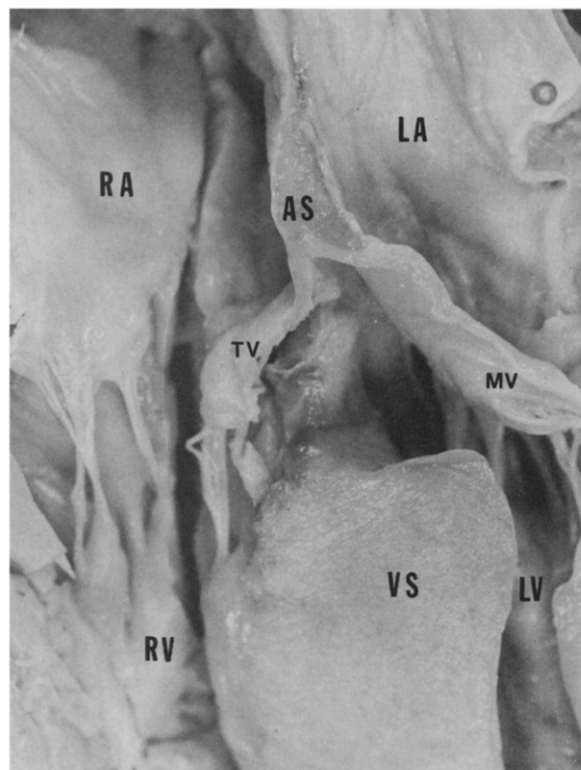


Figure 9. Inlet septal defect, which is inferior to the right posterior division of the septal band and the muscle of Lancisi (ML). The defect is juxtamiltral and juxtaticuspid in the areas indicated by the arrows but is neither juxtataortic (nor perimembranous) nor juxtacrucial. Presumably the atrioventricular (AV) node is displaced toward the crux cordis, and the His bundle traverses the posteroinferior border of the defect. The defect can also be termed an "AV canal type of ventricular septal defect." Abbreviations as in Figures 1 and 3.

Discussion

Categorization of ventricular septal defects. This study represents a serious effort to reconcile the inferences from a large and long surgical experience with ventricular septal defects (both isolated and combined with other cardiac anomalies) with those from the detailed anatomic study of 76 hearts with essentially an isolated defect and those from similar previously reported studies. Although the anatomy is unchanging, from a surgical standpoint, some descriptors of the defect are more useful than others, some relations are more important than others and a categorization that includes, to as great a degree as possible, all types of defects that may be seen surgically is important. The study emphasized the border-forming structures around the defect because, in closing the defect, sutures must be placed around it. The categorization of the defects, by no means original but rather an adaptation of that of Van Praagh et al. (5) and Smolinsky et al. (6) is applicable generally, not just to isolated ventricular septal defects. Therefore, the most common type of ventricular septal defect in tetralogy of Fallot is a large conoventricular defect that is characterized by anterior and rightward malalignment of the conal septum and that is juxtataortic and usually juxtaticuspid and thus perimembranous. Conoventricular defects can be identified echocardiographically in part by their proximity to the central fibrous body, as emphasized in concept by Baker et al. (10), but the echocardiographer cannot differentiate between perimembranous defects and those that are not perimembranous because of a sometimes narrow band of muscle separating the ventricular septal defect from the tricuspid anulus and central fibrous body.

Figure 10. Inlet septal juxtacrucial ventricular septal defect. The defect is also juxtamiltral and juxtaticuspid. The specimen has been photographed by an observer looking toward the crux cordis (the free wall junction of right ventricle [RV], left ventricle [LV], right atrium [RA] and left atrium [LA]). Thus, the right ventricle and right atrium are to the observer's left side. AS = atrial septum; VS = ventricular septum; other abbreviations as in Figures 1 and 3.



Avoidance of term "membranous ventricular septal" defect. Some categories and descriptors in this study differ from those of others. No category of "membranous ventricular septal" defect is used, in large part because the phrase has had so many different meanings in the past. Also, Van Praagh and coworkers (5,6) have given the phrase the connotation that the defect is small, that all other cardiac structures are normal and that there is no identifiable hypoplasia or malalignment of the conal septum. However, in another morphologic study, the investigators encountered large defects that had no identifiable conal septal abnormality and that, except for size, could have been considered "membranous ventricular septal defects" (Kreutzer J, Jonas R, Kirklin JW, Van Praagh R, 1989, unpublished observations). In the present study, these would have been categorized as conoventricular defects without conal septal hypoplasia or malalignment.

"Perimembranous ventricular septal defects." Such a defect was originally conceptualized as being juxtatricuspid, juxtamitral and juxtaaortic (7). The phrase is useful and well understood as a descriptor of defects that are juxtaposed to the tricuspid annulus, the mitral annulus and the aortic valve and membranous septum, although others may prefer a term such as "paramembranous" (11). However, defects that are not perimembranous may abut the tricuspid valve in the region of the antero-septal commissure and in these the His bundle traverses the posteroinferior border of the defect just as in perimembranous defects.

"Subvalvular" defect. "Sub" is not used as a prefix in describing defects that are immediately adjacent to a structure such as the aortic valve because it continues to be used and to be useful in the rather different sense in which it was employed by Lev et al. (12) in their study of double outlet right ventricle. They used "sub" to mean that the defect was near or in the pathway to one or the other semilunar valve, not necessarily adjacent to it, and this meaning has been retained in this study.

The footnote in Table 1 emphasizes that the other morphologic details of hearts that have a septal defect must be included in diagnostic studies and surgical descriptions.

We express our sadness and deep regret that death prevented the genius of Ricardo Ceballos, MD from influencing on the final revisions of the manuscript. We express appreciation to our colleagues at the University of Alabama at Birmingham for their input into this work and to Robert Anderson, MD, Brian Barratt-Boyes, MD, Louise Calder, MD, Richard Jonas, MD and Richard Van Praagh, MD for valuable criticism of the earlier versions of the study (this is said without implying that they necessarily endorse the final version).

References

1. Kirklin JW, Harshbarger HG, Donald DE, Edwards JE. Surgical correction of ventricular septal defect: anatomic and technical considerations. *J Thorac Surg* 1957;33:45-57.
2. Kirklin JW, McGoon DC, DuShane JW. Surgical treatment of ventricular septal defect. *J Thorac Cardiovasc Surg* 1960;40:763-75.
3. Lauer RM, Ongley PA, DuShane JW, Kirklin JW. Heart block after repair of ventricular septal defect in children. *Circulation* 1960;22:526-34.
4. Becu LM, Fontana RS, DuShane JW, Kirklin JW, Burchell HB, Edwards JE. Anatomic and pathologic studies in ventricular septal defect. *Circulation* 1957;14:349-64.
5. Van Praagh R, Weinberg PM, Calder AL, Buckley LFP, Van Praagh S. The transposition complexes: how many are there? In Davila JC, ed. *Second Henry Ford Hospital International Symposium on Cardiac Surgery*. New York: Appleton-Century-Crofts, 1977:207-13.
6. Smolinsky A, Castaneda AR, Van Praagh R. Infundibular septal resection: surgical anatomy of the superior approach. *J Thorac Cardiovasc Surg* 198;95:486-94.
7. Soto B, Becker AE, Moulart AH, Lei JT, Anderson RH. Classification of ventricular septal defects. *Br Heart J* 1980;43:332-43.
8. Griffin ML, Sullivan ID, Anderson RH, Macartney FJ. Doubly committed subarterial ventricular septal defect: new morphological criteria with echocardiographic and angiographic correlation. *Br Heart J* 1988;59:474-9.
9. Studer M, Blackstone EH, Kirklin JW, et al. Determinants of early and late results of repair of atrioventricular septal (canal) defects. *J Thorac Cardiovasc Surg* 1982;84:523-42.
10. Baker EJ, Leung MP, Anderson RH, Fischer DR, Zuberhuhler JR. The cross sectional anatomy of ventricular septal defects: a reappraisal. *Br Heart J* 1988;59:339-51.
11. Huhta JC, Edwards WD, Danielson GK, Feldt RH. Abnormalities of the tricuspid valve in complete transposition of the great arteries with ventricular septal defect. *J Thorac Cardiovasc Surg* 1982;83:569-76.
12. Lev M, Bharati S, Meng CCL, Liberthson RR, Paul MH, Idriss FS. A concept of double outlet right ventricle. *J Thorac Cardiovasc Surg* 1972;64:271-81.