Angiocardiography of Multiple Ventricular Septal Defects in Infancy

KENNETH E. FELLOWS, M.D., G. RICHARD WESTERMAN, M.D., AND JOHN F. KEANE, M.D.

SUMMARY Biplane cineventriculography in 364 infants 1 year of age or younger demonstrated multiple ventricular septal defects (VSDs) in 56 (15%). Among 111 infants with VSDs (with or without patent ductus arteriosus), 18 (16%) had multiple VSDs, whereas 14 of 39 infants (36%) with VSD and coarctation of the aorta had multiple VSDs. The incidence of multiple VSDs in infants with tetralogy of Fallot was 7% (eight of 117), in infants with d-transposition of the great arteries 19% (eight of 43), and in infants with common atrioventricular canal 15% (eight of 54). Perioperative axially angled cineangiography correctly predicted the presence of multiple VSDs in 13 of 15 infants (86%) who underwent operation.

THE MORTALITY RATE for surgery of congenital heart disease when associated with multiple ventricular septal defects (MVSDs) is 14–17%, 1,2 compared with 4% when a single VSD is present. 3 With the trend in many medical centers toward surgical correction of congenital heart defects during infancy, 1,4,5 precise preoperative diagnosis in babies is becoming increasingly important. Our previous experience, in which the presence of MVSDs was identified preoperatively in only 55% of patients operated upon, 1 indicates that preoperative diagnosis must be improved.

This study was undertaken to determine the incidence of MVSDs in infants with ventricular septal defect (VSD), VSD and coarctation (VSD/CoAo), tetralogy of Fallot (T/F), d-transposition (d-TGA) and common atrioventricular canal (CAVC) using cineangiography. A second objective was to evaluate whether the use of axially angled views improved preoperative diagnosis. Additionally, the incidence of spontaneous closure of one or more of these defects was examined among those with VSD and VSD/CoAo by clinical observation and subsequent cardiac catheterization, and among those with d-TGA by repeat cineangiography.

From the Departments of Radiology, Cardiac Surgery and Pediatric Cardiology, Harvard Medical School and Children’s Hospital Medical Center, Boston, Massachusetts.

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Address for correspondence: Kenneth E. Fellows, M.D., Department of Radiology, Children’s Hospital Medical Center, 300 Longwood Avenue, Boston, Massachusetts 02115.

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Materials and Methods

Between January 1, 1975, and December 31, 1980, 364 babies 1 year of age or younger with the diagnosis of VSD (111 infants), VSD/CoAo (39 infants), T/F (117 infants), d-TGA (43 infants) and CAVC (54 infants) underwent cardiac catheterization and biplane cineangiography at Children’s Hospital Medical Center, Boston. The catheterization data and cineangiograms from these studies were analyzed to determine the prevalence of MVSDs (i.e., more than one VSD in the same patient) (table 1).

Cineangiography was performed using either left ventricular or biventricular injections with biplane, image-intensified cine filming at 64 frames/sec. Injected volumes of contrast varied between 1 and 2 ml/kg body weight. Axially angled views in either the long axial oblique (20° cranial and 70° left oblique) or hepatocavicular (40° cranial/40° left oblique) projections were used in most cases between 1975 and 1978, and in all studies after 1978. Infants whose cineangiograms demonstrated more than one VSD constituted the study group;* questionable cases were excluded.

Between January 1, 1978, and April 30, 1981, 15 infants were found to have MVSDs at the time of

*A VSD is identified angiographically by a stream of contrast across the interventricular septum; all infants included in this study demonstrated streaming at more than one level in the septum. Although excluded when recognized, it is possible that a few patients in this study had a single defect, as viewed from the left ventricular side of the septum, which produced two or more streams because of right ventricular trabeculation and musculature. This was not thought to be a limitation of the study because the surgical approach, which usually is through the tricuspid valve and right ventricle, would cause these defects to appear to be ‘multiple’ also.
surgery: nine with VSD and VSD/CoAo, two with T/F, three with d-TGA and one with a double outlet left ventricle. The preoperative data of these 15 infants were viewed to determine in how many the MVSDs had been prospectively predicted by axially angled cineangiography.

Follow-up data on the infants with MVSD alone and MVSD/CoAo were obtained a mean of 11.5 months after an initial catheterization, and examined to determine the incidence of spontaneous closure of MVSD. Repeat catheterization was performed in nine of 18 and nine of 14 infants with MVSD alone and MVSD/CoAo, respectively. Repeat cineangiography was performed in six of 18 and seven of 14 babies in the respective groups. Follow-up cineangiography at a mean interval of 4 months was also evaluated in the six babies with MVSD/d-TGA.

Results

Fifty-six of 364 infants (15%) had MVSDs. Eighteen among the 111 with VSD alone (16%) had MSVDs (three of whom had a patent ductus arteriosus [PDA]), and 14 of 39 (36%) with VSD/CoAo, eight of 117 (7%) with T/F, eight of 43 (19%) with d-TGA and eight of 54 (15%) with CAVC had MVSDs (fig. 1). Of 18 infants with MVSD alone, 12 of 18 had a perimembranous defect plus mid- and apical muscular defects (fig. 2); six of 18 had multiple muscular defects (fig. 3). Among the 14 infants with MVSD/CoAo, nine of 14 had multiple anterior, mid- and apical muscular defects (fig. 4) and five of 14 had perimembranous plus muscular defects. Among the eight infants with MVSDs and d-TGA, five had combined perimembranous and muscular defects and three had only mid- and apical muscular septal deficiencies (fig. 5). T/F anatomy was complicated by the additional presence of both posterior (atrioventricular canal) and anterior muscular defects, while infants with CAVC (fig. 6) had additional anterior and apical muscular defects only.

Preoperative diagnosis in 15 infants found to have MVSD at surgery was correct in 13 (86%). In two

![Figure 1](http://circ.ahajournals.org/)

**Figure 1.** Frequency of multiple ventricular septal defects (MVSD) in infants, 1975–1980. VSD = ventricular septal defect; CoAo = coarctation of the aorta; TOF = tetralogy of Fallot; AVC = atrioventricular canal; d-TGA = d-transposition of the great arteries.

![Figure 2](http://circ.ahajournals.org/)

**Figure 2.** Left ventriculogram, long axial oblique projection. Perimembranous ventricular septal defect (double arrows) and additional muscular defects (single arrows). LV = left ventricle; Ao = aortic valve.
FIGURE 4. Left ventriculogram. Right oblique view (A) demonstrates multiple anterior muscular defects (surgically verified). Left long axial oblique view demonstrates anterior and midmuscular defects (s) and severe coarctation (arrow heads) on a later frame (C). LV = left ventricle.

By clinical examination and repeat catheterization, spontaneous closure of MVSDs occurred in one of 18 infants with MVSDs alone and in two of 14 with MVSD/CoAo (table 2). In two other babies with MVSD/CoAo, the shunt volume markedly diminished, suggesting decrease in the size or number of MVSDs. Of these five infants, closure of MVSDs was documented angiographically in only one.

Discussion

MVSDs were more common (15%) in these infants studied largely by axial angiography than in older children studied routinely, in whom the incidence has been reported to be 2–10%.7–9 The shunts in most of these infants were large, as documented by catheterization (table 1) and angiography, so that in most, closure during corrective surgery in infancy would have been necessary. Because the presence of MVSDs may have variable implications, depending on the other associat-

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<th>TABLE 2. Clinical and Catheterization Follow-up of Infants with Multiple Ventricular Septal Defects (Mean 11.5 Months)</th>
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<td>MVSDs alone (n = 18)</td>
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<tr>
<td>1/18 — spontaneous closure</td>
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<tr>
<td>4/18 — no change (after PAB)</td>
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<td>13/18 — surgical closure</td>
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<td>3/13 — CoAo repair + PAB</td>
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Abbreviations: MVSDs = multiple ventricular septal defects; PAB = pulmonary artery banding; CoAo = coarctation of the aorta; Qp/Qs = pulmonary-to-systemic flow ratio.
patients at Children’s Hospital (fig. 7), the incidence of MVSDs alone or with CoAo is three times more common in infancy. Because VSDs are thought to close spontaneously,10,11 the higher incidence in infancy is expected. Also, MVSDs are often associated with large left-to-right shunts leading to symptoms and clinical investigation early in life. The shunts calculated in these babies were generally greater with 2:1 (Qp/Qs) (table 1), although many had shunts at atrial and ductal levels so that the Qp/Qs ratio does not reflect ventricular shunting alone.

The high incidence (36%) of MVSDs in the group of infants with VSD/CoAo is surprising. However, in fewer infants, a similarly high incidence was also shown.5 Since MVSDs are more common in the first year of life, they are perhaps more likely to be visualized angiocardiographically when associated with coarctation because of higher left ventricular pressures. Infants with VSD/CoAo also present clinically (and undergo cardiac catheterization) relatively early, often within the first few weeks of life.4 The mean age of infants with MVSD/CoAo in our study was 26 days (range 2–150 days); the mean age for MVSDs alone was 3.3 months (range 0.5–9 months). Additionally, the high incidence of MVSDs in these infants may be associated with a reported increasing incidence of VSD in the population in general.12

VSDs have been reported to close or to decrease in 44% of (nonbanded) infants after repair of CoAo4 and a similar closure rate was evident in our study. Of nine infants who had CoAo resection and PDA ligation (with and without pulmonary artery banding), four of nine (44%) showed evidence of complete or partial VSD closure (table 2). Because most infants with a MVSD alone underwent surgical closure soon after

FIGURE 5. Left ventriculogram, long axial oblique projections in an infant with d-transposition of the great arteries, demonstrates mid- and apical muscular defects (arrows). LV = left ventricle; PA = pulmonary artery.

ed cardiac lesions, the infant groups are discussed separately.

MVSDs and MVSDs with CoAo

The combined incidence of MVSDs alone (a PDA was present in three infants) and MVSD/CoAo (PDA in 10) was 21% (32 of 150). Compared with older

FIGURE 6. Left ventriculogram, hepatoclavicular view in common atroioventricular canal. Early in the injection (A), the abnormal cleft mitral valve, mild mitral regurgitation and shunt at the canal level are shown. Additional defects in the muscular septum (arrows) begin to opacify and are best seen on a later frame (B).
cardiac catheterization, the incidence of spontaneous closure could not be determined in that group.

T/F and MVSDs

Eight of 117 infants (7%) with T/F had MVSDs. Thirty-six (all younger than 7 months of age) were previously reported as having a 14% incidence of MVSDs. The discrepancy is attributed to differences in the groups. The present study was larger (117 vs 36 infants) and included patients with pulmonary atresia. Because the babies in the present study were older (less than 1 year vs less than 7 months of age), it is also possible that some muscular defects became smaller or closed. Nevertheless, the 7% incidence of MVSD in T/F was higher than the 2% previously reported.

Among the infants with T/F and MVSDs in this study, seven of eight (88%) had a typical malalignment VSD plus additional muscular defects. In the remaining infant, the defects consisted of the malalignment defect and a defect in the atrioventricular canal portion of the septum. Although the two defects in this infant were confluent rather than separate, they represented angiographically recognizable deficiencies in different parts of the septum and presented special problems for surgical closure.

**d-TGA and MVSDs**

Of 43 infants with d-TGA (mean age 16 days), eight (19%) had MVSD. Six of eight (75%) had perimembranous and muscular defects and two (25%) had only multiple muscular defects. Previous reports have indicated an incidence of MVSD in d-TGA of 2–4%; repeat angiography at a mean age of 4 months in 86 infants demonstrated no change in the size or number of MVSD in 76, but fewer MVSDs in the other three (whether due to VSD closure or decreased visualization because of lower left ventricular pressures was not evident).

**CAVC and MVSDs**

Eight of 54 infants (15%) with partial, intermediate or complete forms of atrioventricular canal defect had, in addition, anterior, mid- and apical muscular septal defects. Because the posterior interventricular septum is deficient in the presence of an atrioventricular canal anomaly, and because the abnormal attachments of the mitral valve tissue to the septum often create multiple streams across the canal defect, axially angled angiography in the hepatoclavicular view is generally necessary to identify these additional anterior muscular defects. Although the hepatoclavicular view best separates the posterior and anterior segments of the septum, in cases where the posterior or canal defect is small, the long axial oblique projection may also help to identify additional muscular defects.

**Preoperative Angiography**

The optimal view for evaluating the membranous and muscular portions of the interventricular septum is
the long axial oblique projection\textsuperscript{16-18} (figs. 2-5). Not only does it allow analysis of most of the septum from the aortic valve to the left ventricular apex, but its biplane reciprocal view (a right oblique projection) localizes muscular defects in the extreme anterior part of the septum (i.e., between the septal band and the right ventricular free wall) (figs. 4A and 8A).

The four-chamber or hepatoclavicular projection\textsuperscript{16-18} has a special advantage in patients suspected of having an atrioventricular canal defect. This projection displays the posterior (basal) interventricular septum, demonstrates the relationship of the common atrioventricular valve to the interventricular septum, and allows identification of additional defects in the muscular part of the septum (fig. 6).

Improved preoperative diagnosis of MVSDs in infants was demonstrated by the use of these cranially angled views. MVSDs were recognized preoperatively in 13 of 15 infants (86%) in whom axial projections were used exclusively, compared with 55% in infants and children in whom they were only occasionally used.\textsuperscript{1}

We cannot explain our failure to recognize MVSDs in two patients even on retrospective viewing. The muscular defects may have been superimposed on and obscured by the large malalignment defects present. It is also possible that in the presence of a large malalignment or perimembranous VSD, additional smaller defects with a longer course through the muscular septum do not opacify because they shunt little or no blood. This explanation is supported by postoperative angiography; after patching of large perimembranous defects in some infants, previously unidentifiable, small muscular defects are occasionally easily visualized by repeat ventriculography.

Early reoperation has been recommended for residual MVSDs in children.\textsuperscript{1} Improved accuracy in preoperative diagnosis should diminish the need for surgical reexploration in search of additional defects.

References

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