Ventricular Septal Defects in the Infant Age Group

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An analysis was made of the 23 cases of isolated ventricular septal defects seen at autopsy over a 20-year period at a children's hospital. All of the deaths occurred during the first 15 months of life. In addition to the clinical and morphologic findings, cardiac catheterization data obtained on 6 of the infants are presented. A wide range of electrocardiographic variation with respect to ventricular hypertrophy was found. No simple relationship between the size of the defect and the clinical manifestations could be demonstrated. The surgical implications of the findings are discussed.

RECENTLY a few authors have recognized uncomplicated ventricular septal defects to be a major cause of heart failure and death in the infant age group, particularly in the early months of life. However, there are little data concerning the relative frequency of this malformation as compared with other serious cardiac anomalies and even less regarding the findings obtained at cardiac catheterization in cases of ventricular septal defect subsequently proved at autopsy. The purpose of this communication is to analyze a series of such cases from statistical, clinical, hemodynamic, and morphologic points of view and to discuss their surgical implications.

MATERIAL AND METHOD

A review of 2,586 consecutive autopsies performed at the Children's Hospital of Buffalo during the period of 1936 to 1956 revealed 288 cases of major congenital cardiac anomalies. Twenty-three of these proved to be uncomplicated ventricular septal defects. For the purposes of this study, stillborn infants and clinically unimportant cardiac abnormalities were excluded. In this series of ventricular septal defects the foramen ovale was closed in only one. However, in no instance did its valve appear incompetent. Two hearts with associated widely patent ducti were retained in this study. One of these occurred in a newborn baby who died at 3 days of age; the other in a baby who died on the eleventh day. In both, the patency of the ductus arteriosus was considered to be in the range of normal. In all other cases the ducti were either closed or very small.

The following additional cardiovascular anomalies were noted: left superior vena cava entering coronary sinus (cases 13 and 18); right aortic arch (case 5); bicuspid aortic and pulmonic valves (case 4). We did not think that these abnormalities had affected the clinical course of these patients.

Cardiac catheterization was performed on 6 of the infants in the usual fashion, with a Sanborn Poly-viso direct writing recorder and Statham strain gauges as pressure transducers. In each patient light general anesthesia with intravenous pentothal was employed in addition to local procaine anesthesia and premedication. The catheter was introduced into a vein in the groin (saphenous or femoral). Blood gases were analyzed by the Van Slyke method. Because of the size and condition of the patients we were unable to obtain respiratory gases. It is necessary to point out that all of the infants were ill and in varying degrees of heart failure at the time of the cardiac catheterization. In 2 of them it was found necessary to administer 100 per cent oxygen by mask during the procedure. No calculations of shunts or flows were made because we thought that, under the circumstances, the errors inherent in the assumptions that must be made were too great.

Special attention was given to the histologic findings in the lungs in an effort to determine vascular changes. The blood vessels were specifically studied in regard to changes in their walls. In addition to the usual hematoxylin-eosin stain, the orcein-Van Gieson method was used for elastic tissues and the smooth musculature in the blood vessels in all sections from the lungs.

As long as there is no standard measurement applied by all who are interested in the study of the relation between lumen and thickness of arterial walls containing elastic membranes, it seemed better to go by general comparisons of the individual cases, with assumed normal controls of the same age in this entire series. A variety of factors intrinsic in the lung tissue at the time of death (like atelectasis, pneumonitis, hemorrhage, edema, and, in particular, emphysema) and extrinsic factors of a technical nature, such as interval between time of death and postmortem examination, type of fixation, embedding material, and processing (in-
TABLE 1.—Relative Frequency of the Common Major Congenital Malformations of the Heart in Infants and Children as Seen at Autopsy

<table>
<thead>
<tr>
<th>Malformation</th>
<th>Number of cases</th>
<th>Approximate per cent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete transposition of the great vessels</td>
<td>35</td>
<td>12</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>23</td>
<td>8</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>23</td>
<td>8</td>
</tr>
<tr>
<td>Primary endocardial fibroelastosis</td>
<td>21</td>
<td>7</td>
</tr>
<tr>
<td>Persistent ostium atrioventricular commune</td>
<td>18</td>
<td>6</td>
</tr>
<tr>
<td>All others</td>
<td>168</td>
<td>59</td>
</tr>
<tr>
<td>Total</td>
<td>288</td>
<td>100</td>
</tr>
</tbody>
</table>

Including the temperature used for stretching the paraffin sections) should be considered in any thorough evaluation. The most important facet, however, would be to agree on specified areas in the lung tissue for examining the blood vessels and, in particular, on the planes in which such blocks including the small arteries and their branches should be cut. Under such conditions, the direct measuring of numerous representative vessels appears preferable to the more general comparisons that we have used in this study.

RESULTS

Clinical Aspects. For purposes of comparison, an analysis was made of the 288 major cardiovascular anomalies revealed at autopsy. The relative frequency of the 5 most common in this series is presented in table 1. Ventricular septal defects proved to be a common cause of death in congenital heart disease. While it must be admitted that in 8 cases death followed unsuccessful surgery, such surgery was only performed out of desperation on infants otherwise expected to die in a short time.* Also, with the exception of primary endocardial fibroelastosis, a similar proportion of fatalities occurred following surgery in each of the other major malformations listed in table 1.

* The surgical procedures were as follows: Closure of the defect under direct vision with the aid of hypothermia (cases 12, 13 and 15); creation of pulmonic stenosis to reduce pulmonary hypertension and flow (cases 11 and 14); and ligation of ductus arteriosus (cases 1, 6, 9). In the last 3 it was mistakenly thought that a widely patent ductus might be present, causing part of the load on the infant's heart.

The ages of the patients at death varied from 3 days to 15 months. Twenty out of 23 deaths occurred during the first 6 months of life (fig. 1). The majority of the patients died during the first 3 months.

The clinical symptoms and physical findings were similar to those described by Engle in 9 infants who died with uncomplicated ventricular septal defects. Failure to gain and recurrent episodes of respiratory infections were the chief complaints in all 23 cases. In 7, ranging in age from 3 weeks to 5 months, the admission weight was the same or less than the birth weight. Respiratory distress associated with transient cyanosis was present in all. In 4 cases there was a history of cyanotic spells usually occurring during feedings. These episodes preceeded the development of respiratory distress or infection. A loud, harsh, prolonged systolic murmur, maximal along the lower left sternal border and accompanied by a systolic thrill, was present in 21 cases. There was no murmur in 1 instance (case 21), a newborn who died at 3 days. One patient, (case 16) was dead on arrival; hence, there were no data regarding the presence or absence of a murmur. Evidence of both right and left-sided congestive heart failure as manifested by dyspnea, rales, and hepatic enlargement was present on admission in 19 of the 23 cases. Peripheral edema was never manifest. In 7 cases the birthweight was less than 2500 Gm. A history of German measles in the first trimester of pregnancy was given in case 3.

Roentgenographic examination, which was performed in 17 cases, revealed cardiac enlargement of a moderate to marked degree in all. Increased hilar vascular markings and convexity of the pulmonary segment were invariably present. Radiologic evidence of left atrial enlargement was present in 6 of the 12 patients.
in whom fluoroscopic examination was done. In case 14 there was a complete collapse of the left lung, which re-expanded spontaneously in 6 days.

Electrocardiographic tracings, including precordial leads, were available in 15 patients (fig. 2 and table 2). None of the tracings were normal.* In 7 there was evidence of right ventricular hypertrophy manifested by an abnormally tall and usually delayed R wave in precordial positions V₁ and V₃₅, together with the progressive decrease in the R wave and increase in the S wave as the electrodes were moved to the left precordium ("Wilson phenomenon"). Miller⁴ has stressed the value and consistency of the latter finding in the diagnosis of right ventricular hypertrophy in small infants beyond the age of 1 week. Two electrocardiograms showed definite evidence of left ventricular hypertrophy manifested by abnormally tall R waves in V₅ and V₆ with depressed S-T segments and inverted T waves in the same leads. Six of the tracings revealed evidence of combined ventricular hypertrophy. This diagnosis was made whenever patterns of both right and left ventricular hypertrophy appeared in the same electrocardiogram. In 3 of these 6 electrocardiograms the evidence of left ventricular hypertrophy was more marked than that of right ventricular hypertrophy; in the other 3 the reverse was true.

Physiologic Aspects. The results of the data obtained at cardiac catheterization in 6 patients are summarized in table 3. In 4 of the infants the catheter was directed in the usual fashion through the right atrium and right ventricle, into the pulmonary artery. In the other 2 it passed from the right ventricle directly into the aorta (fig. 3). (It was not possible subsequently to direct the catheter into the pulmonary artery in either of these 2 patients.) In no instance was a significant pressure gradient present across the aortic or pulmonary valves.

The systolic pressure in the right ventricle was markedly elevated in all patients. In 5 of them it was equivalent to the systemic systolic pressure. In the sixth patient it was lower than the systemic pressure. No wedge pressures were obtained.

Evidence of a left-to-right shunt at the atrial level with a rise in oxygen content of the venous blood obtained from the right atrium occurred in 3 patients. The magnitude of these rises exceeded 2 volumes per cent. In all 6 patients there was a rise in the blood oxygen content at the ventricular level exceeding 1 volume per cent. The oxygen saturation of the arterial blood obtained from the aorta or femoral artery was below 90 per cent in 3

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* Normal values were determined according to Ziegler.⁵

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Fig. 2. Representative electrocardiograms showing variability of findings with respect to evidence of ventricular hypertrophy. Cardiac catheterization was performed on each of these infants. a. Case 12, age 3 weeks, right ventricular hypertrophy. b. Case 15, age 6 months, combined ventricular hypertrophy. c. Case 13, age 2 months, combined ventricular hypertrophy. d. Case 10, age 5 weeks, left ventricular hypertrophy.
TABLE 2.—Electrocardiographic and Postmortem Findings in Twenty-three Cases of Uncomplicated Ventricular Septal Defects

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Electrocardiogram</th>
<th>Ventricular wall thickness (mm.)</th>
<th>Ventricular septal defect diameter (mm.)</th>
<th>Ratio of size septal defect to aortic orifice</th>
<th>Medial hypertrophy of the small pulmonary arteries</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. C. P.</td>
<td>3 mos.</td>
<td>CVH</td>
<td>3</td>
<td>4</td>
<td>0.42</td>
<td>Slight</td>
</tr>
<tr>
<td>2. F. C.</td>
<td>1 mo.</td>
<td>RVH</td>
<td>4</td>
<td>4</td>
<td>0.6</td>
<td>Distinct</td>
</tr>
<tr>
<td>3. K. B.</td>
<td>6 wks.</td>
<td>RVH</td>
<td>7</td>
<td>7</td>
<td>1.35</td>
<td>Not significant</td>
</tr>
<tr>
<td>4. J. R.</td>
<td>11 das.</td>
<td>RVH</td>
<td>8</td>
<td>12</td>
<td>2.2</td>
<td>Distinct</td>
</tr>
<tr>
<td>5. J. F.</td>
<td>3 mos.</td>
<td>RVH</td>
<td>8</td>
<td>9</td>
<td>1.2</td>
<td>Distinct</td>
</tr>
<tr>
<td>6. K. M.</td>
<td>3 mos.</td>
<td>RVH</td>
<td>7</td>
<td>10</td>
<td>1.2</td>
<td>Distinct</td>
</tr>
<tr>
<td>7. W. A.</td>
<td>2 mos.</td>
<td>RVH</td>
<td>8</td>
<td>3</td>
<td>0.27</td>
<td>Slight</td>
</tr>
<tr>
<td>8. M. M.</td>
<td>3½ mos.</td>
<td>CVH</td>
<td>5</td>
<td>10</td>
<td>1.1</td>
<td>Distinct</td>
</tr>
<tr>
<td>9. B. H.</td>
<td>2 mos.</td>
<td>LVH</td>
<td>8</td>
<td>10</td>
<td>1.25</td>
<td>Distinct</td>
</tr>
<tr>
<td>10. A. M.†</td>
<td>3 mos.</td>
<td>LVH*</td>
<td>7</td>
<td>10</td>
<td>1.25</td>
<td>Distinct</td>
</tr>
<tr>
<td>11. L. K.</td>
<td>5 mos.</td>
<td>CVH*</td>
<td>7</td>
<td>9</td>
<td>0.71</td>
<td>Distinct</td>
</tr>
<tr>
<td>12. G. C.</td>
<td>6 wks.</td>
<td>RVH*</td>
<td>10</td>
<td>7</td>
<td>1.16</td>
<td>Not significant</td>
</tr>
<tr>
<td>13. H. G.</td>
<td>2 mos.</td>
<td>RVH*</td>
<td>9</td>
<td>5</td>
<td>0.55</td>
<td>Not significant</td>
</tr>
<tr>
<td>14. K. S.</td>
<td>5 mos.</td>
<td>RVH*</td>
<td>3</td>
<td>7</td>
<td>0.75</td>
<td>Distinct</td>
</tr>
<tr>
<td>15. L. S.</td>
<td>6 mos.</td>
<td>RVH*</td>
<td>11</td>
<td>6</td>
<td>2.0</td>
<td>Distinct</td>
</tr>
<tr>
<td>16. T. C.</td>
<td>6 mos.</td>
<td>RVH*</td>
<td>8</td>
<td>19</td>
<td>0.42†</td>
<td>Not significant</td>
</tr>
<tr>
<td>17. S. S.</td>
<td>15 mos.</td>
<td>RVH*</td>
<td>4</td>
<td>4</td>
<td>1.0</td>
<td>Distinct</td>
</tr>
<tr>
<td>18. D. P.</td>
<td>6 wks.</td>
<td>RVH*</td>
<td>6</td>
<td>8</td>
<td>0.89†</td>
<td>Distinct</td>
</tr>
<tr>
<td>19. R. M.</td>
<td>5 mos.</td>
<td>RVH*</td>
<td>10</td>
<td>9</td>
<td>1.8</td>
<td>Not significant</td>
</tr>
<tr>
<td>20. D. B.</td>
<td>13 mos.</td>
<td>RVH*</td>
<td>10</td>
<td>8</td>
<td>0.86†</td>
<td>Not significant</td>
</tr>
<tr>
<td>21. D. F.</td>
<td>3 das.</td>
<td>RVH*</td>
<td>9</td>
<td>6</td>
<td>0.38†</td>
<td>Data unavailable</td>
</tr>
<tr>
<td>22. A. D.</td>
<td>2 mos.</td>
<td>RVH*</td>
<td>4</td>
<td>6</td>
<td>3</td>
<td>Data unavailable</td>
</tr>
<tr>
<td>23. J. M.</td>
<td>4 mos.</td>
<td>RVH*</td>
<td>6</td>
<td>5</td>
<td>Data unavailable</td>
<td></td>
</tr>
</tbody>
</table>

* Cardiac catheterization performed.
† Autopsy performed at outside hospital. No measurements given. Two defects present.
‡ Aortic orifice not measured. Described as normal. Average normal measurement for age used.

TABLE 3.—Data Obtained at Cardiac Catheterization: Six Proved Cases of Uncomplicated Ventricular Septal Defect

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Pressures (mm. Hg)</th>
<th>Per cent oxygen saturation</th>
<th>Capacity (O₂) ml./L.</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Aorta</td>
<td>FA</td>
<td>PA</td>
</tr>
<tr>
<td>10</td>
<td>3 mos.</td>
<td>89/52</td>
<td>92/0</td>
<td>9/0</td>
</tr>
<tr>
<td>11</td>
<td>5 mos.</td>
<td>104/73</td>
<td>108/50</td>
<td>108/0</td>
</tr>
<tr>
<td>12</td>
<td>1 mo.</td>
<td>99/42</td>
<td>99/0</td>
<td>8/0</td>
</tr>
<tr>
<td>13</td>
<td>2 mos.</td>
<td>104/60</td>
<td>104/0</td>
<td>104/0</td>
</tr>
<tr>
<td>14</td>
<td>5 mos.</td>
<td>75/50</td>
<td>70/37</td>
<td>82/0</td>
</tr>
<tr>
<td>15</td>
<td>6 mos.</td>
<td>102/57</td>
<td>75/23</td>
<td>75/0</td>
</tr>
</tbody>
</table>

FA = Femoral artery; PA = Pulmonary artery; RV = Right ventricle; RA = Right atrium;
SVC = Superior vena cava; IVC = Inferior vena cava.
* One hundred per cent oxygen administered throughout procedure.

patients with a lowest value of 83 per cent. Two patients showed normal peripheral arterial oxygen saturation. However, 100 per cent oxygen was administered throughout the procedure to both of them because of their poor clinical condition (the ear-oximetry readings were 66 and 68 per cent at the beginning of the procedure). It is clear that no conclusions re-
regarding the presence or absence of a right-to-left shunt can be drawn in these 6 infants. Both the existence of congestive heart failure and the use of a general anesthetic could easily explain the unsaturated state of the arterial bloods.

**Morphologic Aspects.** All of the 23 hearts examined were enlarged, showing both dilatation and hypertrophy. This enlargement, in all but 1 instance (case 10) involved chiefly the right heart. Admittedly, measurement of the thickness of the ventricular wall offers only a rough estimate of the degree of its hypertrophy, particularly in the presence of varying degrees of ventricular dilatation (table 2). Dilatation of the pulmonary artery was a constant finding. The aorta appeared normal in all except case 10 in which it was dilated. Pulmonary congestion, which was frequently marked and associated with patchy atelectasis, was an almost constant finding. The liver was usually hyperemic but of a lesser degree than the lungs.

The defects ranged in size from 3 to 19 mm. (table 2 and fig. 4). According to the recent classification outlined by Becu and associates their location on the septum was as follows: In 17 cases the defect was in the region of the outflow tracts posterior to the crista supraventricularis and below the right and posterior aortic cusps (usually described as the region of the membranous septum). In this group there were 2 examples (cases 5 and 10) of double openings. In both of these hearts there was a second defect in the region of the inflow tracts under the posterior and septal leaflets of the tricuspid valve. In cases 2 and 23 the defects were small (4 mm. and 3 mm., respectively) and were located in the region of the outflow tracts anterior to the crista supraventricularis just below the pulmonary valve. In cases 6 and 9 there were large (10 mm.) openings involving the apical portion of the ventricular septum. In order to determine the relative size of the defect the ratio of its diameter to that of the aortic orifice was calculated in the 21 cases where measurements were available (table 2 and fig. 5).

Noncardiovascular congenital malformations were present in 5 patients. They included right hydronephrosis and hydroureter, absence of the left kidney and ureter, hypospadias, mongolism, and oxycephalus. It was considered that in none of these cases had the associated lesion contributed to the death of the infant.

Examination of the small pulmonary arteries
in 21 cases revealed distinct abnormal medial hypertrophy in 12 and slight in 3 (table 3). In the remaining 6 cases no significant changes were seen as compared with the normal controls. Endarteritic intimal thickening was not observed in any one of the total series.

Discussion

Clinical Aspects. Our data regarding the frequency with which a ventricular septal defect may cause heart failure and death in the early months of life correspond roughly with those recently reported by others. Harned and associates found 5 cases of this anomaly among 55 congenital cardiac malformations taken from the protocols of 1,400 consecutive autopsies in a large general hospital. All 5 of the patients died before the age of 15 months. Edwards noted that of the 30 specimens of ventricular septal defects in the collection of the Mayo Clinic, 19 were from infants who died before the age of 6 months. Marquis found it to be the most common malformation in 110 consecutive cases of congenital heart disease seen at autopsy during the first 3 years of life.

We do not wish to infer that heart failure due to this malformation necessarily carries a fatal outlook in early infancy. In the past 5 years we have observed 4 patients with similar signs, symptoms, and electrocardiographic and roentgenographic findings who survived congestive heart failure in early infancy and appear clinically much improved at ages varying between 2 and 4 years. Cardiac catheterization performed on these patients during the first year of life revealed essentially the same findings as in the 6 patients of this study: that is, marked right ventricular and pulmonary arterial hypertension approximating the systemic pressure, and a large left-to-right shunt at the ventricular level. A similar experience with 3 infants catheterized before the age of 8 months was reported by Harned’s group. This is, of course, in line with the concepts of Dammann and Ferenez and confirms the statement of Edwards: “It appears that in those patients who survive the critical six postnatal months, adaptation to excess pulmonary flow is reasonably well-tolerated.”

The symptomatology, physical findings, roentgenographic data, and clinical course of these patients were essentially the same as recently described by other authors in smaller groups of infants. Ferenez and co-workers have pointed out that these clinical findings are nonspecific and for the most part apply to other cardiac malformations that produce large hearts and increased blood flow to the lungs in infants. We have nothing to add to the discussions of the differential diagnosis presented by these 2 groups of authors. In our experience, even with cardiac catheterization, it is occasionally impossible to distinguish persistent ostium atrioventriculare commune, truncus arteriosus, patent ductus, and a single ventricle from an isolated ventricular septal defect.

We are unable to explain the variations obtained in the electrocardiograms regarding ventricular hypertrophy or to relate them to age, clinical, roentgenographic, or pathologic findings in this series of cases (table 2). Nor are the physiologic data obtained at catheterization helpful in this respect. Despite similar values as to pressures and shunts, a complete range of electrocardiographic variability with respect to right ventricular hypertrophy, combined ventricular hypertrophy, and left ventricular hypertrophy was found. Data regarding the pulmonary artery wedge pressures and the resistances in the pulmonary and systemic circuits might be helpful in this respect. The finding of Blount and co-workers that a left ventricular hypertrophy pattern is associated with large left-to-right shunts in the absence of severe pulmonary hypertension does not apply to case 10 in our series. In this infant the equivalent of the systemic arterial pressure was present in the right ventricle at cardiac catheterization. In case 9, in which clear electrocardiographic evidence of left ventricular hypertrophy had been obtained, the defect was large (10 mm.) and it seems probable that a marked degree of pulmonary hypertension had been present. An additional point is that in our patients with left ventricular hypertrophy, in contrast to the older children and adults analyzed by Blount’s group, the T waves over the left precordium were inverted (which,
according to Cabrera and Monroy, indicates systolic rather than diastolic stress).

Physiologic Aspects. It is now clear that there is an extreme range of variability insofar as pressures, volume, and direction of shunts are concerned in patients with ventricular septal defects. The findings summarized in table 3 are from 6 infants who had manifested obvious clinical evidence of heart failure prior to catheterization. In 5 of the infants the degree of right ventricular hypertension together with the evidence of a large left-to-right shunt indicated the presence of a defect of sufficient size to equalize the pressure in the 2 ventricles and a pulmonary resistance that was less than the systemic. In cases 12 and 13 an identical systolic pressure was demonstrated by the lack of a pressure gradient between the aorta and the right ventricle when the catheter was withdrawn from this vessel into the ventricle. In the sixth catheterized case (case 15) the systolic pressure in the femoral artery was 36 per cent greater than that in the pulmonary artery. Kroeker and Wood have demonstrated that the femoral artery systolic pressure may be 4 to 19 per cent greater than the central aortic pressure. If this applies to infants, it is unlikely that in this patient the aortic and pulmonary arterial systolic pressures were equivalent. In the 2 cases (11 and 14) in which data were available, the diastolic pressure in the pulmonary artery was significantly lower than that in the peripheral artery. This indicates, as Dammann and Ferencz point out, that a lower peripheral vascular resistance is present in the pulmonary circuit as compared to the systemic when the systolic pressures in the 2 great vessels are the same.

The additional finding in 3 patients of a large left-to-right shunt into the right atrium should, we believe, be ascribed to functional tricuspid insufficiency in view of the fact that no anomalies other than the ventricular septal defects were found. In no instance did it appear that the defect communicated directly with the right atrium. The existence of functional tricuspid insufficiency in the presence of a ventricular septal defect was recognized by Baldwin and associates in 1945. Obviously, this finding of left-to-right shunts at both the atrial and the ventricular levels complicates the differential diagnosis. Such patients may be subjected to surgery in an attempt to repair erroneously suspected atrial septal defects. Watkins and Gross report this experience with 3 patients (apparently not infants) with "adequate pre-operative catheterization studies."

Pathologic Aspects. Several authors have accepted Selzer's contention that the size of the defect determines its clinical importance. Insofar as our material is concerned, there was no apparent relationship between the absolute size of the defect and the ages at death (fig. 4). In accordance with the observation of Selzer and Laqueur that the relationship of the size of the opening to the size of the aortic orifice is important, we determined the ratio of the 2 in terms of their diameters. Again, there was no apparent correlation with the ages at death (fig. 5). However, 12, or more than half, of the hearts of our series had defects as large as or larger than the aortic orifice. Also, the ratios in the catheterized cases are compatible with Selzer's reasoned assumption that when the ventricular septal defect is larger than one half of the aortic orifice the pressure differential between the 2 ventricles may disappear. In all 5 catheterized patients with available autopsy measurements the ratio was greater than 0.5.

The 4 cases with small defects and ratios below 0.5 are difficult to explain. There were no apparent significant differences in the clinical, roentgenographic, electrocardiographic, or pathologic data in this group as compared with the infants who had large defects. Catheterization findings would have been most interesting in this group. Engle, in her series of 9 infants, reported 1 with a defect only 2 mm. in diameter. Her suggestion that the relationship of the location of the aorta to the defect is of importance would not apply to our case 23. In this heart there was only a 3-mm. opening located just below the pulmonic valve, anterior to the crista supraventricularis.
Our data do serve to confirm the findings of Selzer8 and of Becu et al.,7 that the location of the defect in the septum is not important in determining its effect on the cardiovascular system. The 4 patients (cases 2, 6, 9, and 23) with single openings in other than the usual site were in no respect different from the other 19 infants.

In septal defects with the same systolic pressure in the pulmonic and systemic circuits Dammann and Ferenez9 have described 3 distinct clinical syndromes or phases. Although we obviously cannot assume the same hemodynamic situation in all of our patients, it is apparent that clinically all of them fit into what these authors term “Phase I.” This phase is manifested by retarded growth, recurrent pulmonary infections, dyspnea, excessive blood flow to the lungs, cardiac enlargement, and high-output cardiac failure. Adaptation and survival are ascribed largely to an increase of the pulmonary vascular resistance produced by mediat hypoplasia in the small pulmonary arteries. Distinct changes of this type present in 12, or over half, of our cases suggest partial adaptation to the altered hemodynamics produced by the defect. However, analysis of the clinical data in these failed to reveal any significant differences as compared with the 6 patients in whom no significant changes in the small pulmonary arteries were found. The same holds true with respect to the age at death and onset of symptoms in our series. In all but 1 of the 9 infants reported by Engle,1 the fetal character of the small muscular pulmonary arteries was retained. In 3 of our hearts with small defects less than half of the diameter of the aortic orifice, the pulmonary vascular changes were described as slight or not significant. This suggests the possibility that the patients involved (cases 1, 7, 17) may not have had severe pulmonary hypertension. Several of the patients with large defects, on the other hand, had either slight or no significant medial hypertrophy of the small pulmonary arteries.

Dammann and Ferenez9 found that of a group of 27* infants who had manifested in life the clinical findings of “Phase I” and who had been shown to have equal systemic and pulmonary artery pressures, the majority had pulmonary arteries with thicker walls and smaller lumens than the average normal of the same age. (Most of them, however, fell into what was considered to be the range of normal.) Our failure to correlate the changes noted in our series with the ages at death and the clinical findings probably indicates that other factors are involved and the observed medial hypertrophy was not sufficiently marked to benefit these patients.

**Surgical Implication.** The data as presented have a direct bearing on the surgical approach to this problem: (1) The frequency of early death necessitates surgery in the first few months of life if a significant reduction in the fatality rate is to be achieved; (2) even in infants who survive the early months of life, pulmonary vascular changes are commencing that may well preclude surgery at a later date; and (3) the data accumulated in our series and that of Becu and co-workers7 are indicative of the range of anatomic variations that the surgeon must be familiar with and be expected to cope with—the wide range in the size of the lesions, multiple defects, and unusual locations of the defects in the septum.

**Summary**

The clinical features and postmortem findings have been analyzed in 23 infants who had a ventricular septal defect and died in heart failure. This defect comprised 23 (8 per cent) of 288 major cardiac malformations observed at autopsy over a 20-year period at a children's hospital. The majority of the deaths occurred during the first 3 months of life.

Hemodynamic data obtained on 6 of these infants revealed a large left-to-right shunt and marked right ventricular and pulmonary artery hypertension in all. In 3 of the patients the shunt appeared at the atrial as well as the ventricular level. In 5 the systolic pressure in the right ventricle was equivalent to the pressure in the systemic artery.

There was a wide variability in the electrocardiographic findings with respect to evidence of ventricular hypertrophy. Patterns of right,
left, and combined ventricular hypertrophy were found without apparent correlation with the clinical, hemodynamic, or postmortem findings.

No apparent relationship was found between the size of the defect or its location on the septum and the clinical manifestations or age at death. Four of the hearts had small defects less than half the diameter of the aortic orifice.

Distinct medial hypertrophy of the small pulmonary arteries was present in 12 cases; in 6 cases, no significant changes were seen.

The surgical implications of these findings are discussed.

SUMMARIO IN INTERLINGUA

Es discutite le signification de iste constatazioni ab le puncto de vista chirurgic.

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Medical Eponyms

By Robert W. Buck, M.D.


“The examination of a patient who had been referred to me was the beginning of a series of determinations of the borders of the lung apices as well as the lung margins which I will briefly report here. After I had determined the anterior supraventricular margin of the lung in the usual fashion, that is by gentle percussion, I proceeded, still percutting very lightly to the posterior aspect, and thereby obtained the following results: On the right side, as on the left, there appeared a line which extended medially in a wide arch with its convexity directed inward and which approached to within a centimeter of the midline, on the left at the level of a line between the second and third thoracic spines, on the right at the level of the fourth thoracic spine. In this case the right apex was diseased . . . while the left showed a normal condition . . . Inasmuch as I had obviously been successful in determining not only the height of the lung apices but also their breadth . . . I tried to determine similarly the lateral margin . . . The determination of the posterior lateral border is easy in many cases, especially in thin individuals, but is frequently difficult in powerfully built, extremely muscular, or fat persons. The lateral border which I have outlined on the anterior surface is extremely trustworthy. It runs from about the middle of the anterior margin of the trapezius muscle, curves down sharply, cuts the clavicle at about the line between its middle and outer third, and then courses outward diagonally to the axilla. From the configuration of these normal clinical margins, it will now be possible, without great difficulty, to hypothesize the necessary shift which will occur when there are pathological changes in the lung apices. Diseases which reduce the air content will shift the medial border outward and the lateral border inwards.”
Ventricular Septal Defects in the Infant Age Group

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