Multiple Muscular Ventricular Septal Defects

By William F. Friedman, M.D., Ali Mehrizi, M.D., and Allen L. Pusch, M.D.

Approximately 10 per cent of interventricular communications occur in the muscular portion of the ventricular septum. Of 259 cases of isolated ventricular septal defects, 4.0 per cent had multiple muscular defects. Scant mention of these multiple defects as a distinct entity can be found in the recent literature, although the incidence is significant.

We have observed characteristic findings in seven patients with multiple muscular ventricular septal defects. The purpose of this report is to describe the clinical picture, demonstrate the serious nature of this malformation, and emphasize the need for more thorough preoperative evaluation as well as careful inspection at the time of surgery. In addition, we wish to contrast this entity with the isolated muscular septal defect. A number of authors have commented that the latter anomaly has little functional significance.

Anatomic Classification

Defects in the ventricular septum are classified according to their anatomic relation to the crista supraventricularis and the tricuspid valve (fig. 1).

Type 1. The defect is supero-anterior to the crista supraventricularis.

Type 2. The defect is postero-inferior to the crista supraventricularis.

Type 3. The defect is located under the septal leaflet of the tricuspid valve.

Type 4. The defect is single and is located in the muscular portion of the septum.

Type 5. The defects are multiple and involve the muscular septum. The defects may be discrete and of variable size, and multiple small fenestrations may give the septum the resemblance of a sieve.

Material

Seven children with multiple muscular ventricular septal defects were seen at the Harriet Lane Home Cardiac Clinic between 1956 and 1962. The group was composed of four boys and three girls. All were Caucasian. They ranged in age from 4 months to 9 years; five were less than 2 years old.

Results

Clinical Findings

All seven patients had a history of poor feeding, slow weight gain, and frequent upper respiratory infections. A history of excessive sweating was obtained in four patients. Four children became intermittently cyanotic with crying before they reached their first birthday. One child became cyanotic with exertion at age 8 1/2 years. Cyanotic spells occurred in three patients, characterized by extreme irritability, excessive crying, and marked deepening of cyanosis. These spells were not accompanied by loss of consciousness.

Congestive heart failure occurred at an early age in six patients and at 8 1/2 years in one patient. Five children were digitalized before 14 months of age. Each child initially responded to conservative management. Gradual deterioration became evident, however, and refractoriness to all supportive measures subsequently occurred. Consequently, corrective surgery was attempted in each case.

Physical Findings

Each child was in less than the twenty-fifth percentile for weight for the respective age, although none had a premature birth weight. Five children became cyanotic with exercise or crying. The heart was enlarged in all. A systolic thrill was palpable in one patient. All had grade II to IV pansystolic murmurs (on the basis of I to VI grades), maximally heard in the third left intercostal space. Diastolic flow murmurs were described in four patients. The second sound in the
Convergent Locations of Ventricular Septal Defects

Crista supraventricularis
Pulm. valve
Tricuspid valve
Multiple vent, septal defects (sieve type)

Figure 1
Anatomic classification of defects of the interventricular septum (adapted from Kirklin and associates.8)

Pulmonic area was accentuated and split in each case.

Roentgenologic Findings
Progressive cardiac enlargement was noted in each instance. The pulmonary conus was prominent, and the pulmonary vascularity increased (fig. 2). Venous angiocardiography performed in three patients demonstrated reopacification of the right ventricle, pulmonary artery, or both. Although these findings were consistent with the diagnosis of ventricular septal defect, the precise anatomic site of the defect was not localized.

Electrocardiographic Findings
All tracings revealed an rsR' pattern in the right precordial leads and combined ventricular hypertrophy. Four patients had a prolonged P-R interval for age and rate. The QRS axis in five patients varied between $+95^\circ$ and $+175^\circ$. The QRS axis was $+65^\circ$ in one patient and $-165^\circ$ in another (fig. 3).

Right Heart Catheterization
Right heart catheterization was performed in six instances; in case 2 the pressures were obtained during surgery with the patient under deep anesthesia (table 1). The data show a significant step-up in oxygen saturation at the ventricular level and marked right ventricular and pulmonary hypertension in each case. Moderate to marked left-to-right shunting was present. In case 5 a bidirectional shunt may have been present. The low arterial oxygen saturation in case 5 was probably due, however, to pulmonary hypoventilation and makes the calculation of blood flow subject to error. The pulmonary resistance was less than 33 per cent of systemic vascular resistance in four cases, as compared to a normal value of 10 to 20 per cent.13, 21

In case 1 the pulmonary artery was not
MUSCULAR VENTRICULAR SEPTAL DEFECT

entered. Right ventricular hypertension in the presence of markedly increased pulmonary flow radiologically led us to assume that high pressure existed in the pulmonary artery. There were insufficient data in cases 1 and 2 to calculate flows and resistances.

Operative and Pathologic Findings
Each patient underwent cardiac operation. Case 6 is doing moderately well 1½ years postoperatively. The remaining six patients died immediately after surgery and autopsies were performed in each instance. Case 2 was noted to have congenital absence of the middle lobe of the right lung. No other noncardiovascular anomalies were noted in the other patients. The origins of the aorta and pulmonary artery and the systemic and pulmonary venous returns were normal in all. There was no pulmonary infundibular or valvular stenosis noted in any case.

In each of the autopsied cases at least three blocks of lung tissue were available for microscopic study. Elastic-tissue and hema-toxyn and eosin stains were made from multiple sections of different lung areas. Structural alterations in the media and intima of the small pulmonary arterial vessels in each of the autopsied cases were graded 1 through 6, according to the classification of Heath and Edwards.14 Grade 1 changes were observed in cases 1, 3, and 5. These consisted of medial hypertrophy with distinct internal and external elastic laminae without evidence of intimal proliferation or fibrosis. Grade 3 changes were noted in cases 2, 4, and 7 and consisted of medial hypertrophy and cellular intimal proliferation, as well as development of cellular fibrous tissue under the endothelial lining of the smaller muscular pulmonary arteries and pulmonary arterioles.

Case Reports

Case 1 (JHH 90 78 34)
Surgery on this 5-month-old white girl was performed with hypothermia and arterial perfusion. A moderate narrowing of the aortic arch was noted. A ductus with an estimated internal

Figure 3
Representative electrocardiogram, case 7, showing incomplete right bundle-branch block and combined ventricular hypertrophy.
Right Heart Catheterization Data

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>SVC</th>
<th>RA</th>
<th>IVC</th>
<th>RV</th>
<th>PA</th>
<th>SA</th>
<th>Mean pressure (mm Hg)</th>
<th>Blood flow (L/min./M²)</th>
<th>Pulm./syst. flow ratio</th>
<th>PVR/SVR* (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4 mo.</td>
<td>66</td>
<td>71</td>
<td>63</td>
<td>82</td>
<td>—</td>
<td>—</td>
<td>t</td>
<td>60</td>
<td>45</td>
<td>—</td>
</tr>
<tr>
<td>21</td>
<td>14 mo.</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
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</tr>
<tr>
<td>3</td>
<td>21 mo.</td>
<td>64</td>
<td>67</td>
<td>62</td>
<td>86</td>
<td>90</td>
<td>97</td>
<td>45</td>
<td>54</td>
<td>5.8</td>
<td>1.2</td>
</tr>
<tr>
<td>4</td>
<td>7 mo.</td>
<td>72</td>
<td>71</td>
<td>82</td>
<td>86</td>
<td>91</td>
<td>83</td>
<td>110</td>
<td>6.8</td>
<td>14.1</td>
<td>2.1</td>
</tr>
<tr>
<td>5</td>
<td>26 mo.</td>
<td>64</td>
<td>67</td>
<td>67</td>
<td>78</td>
<td>78</td>
<td>85</td>
<td>71</td>
<td>86</td>
<td>5.8</td>
<td>1.2</td>
</tr>
<tr>
<td>6</td>
<td>9 yr.</td>
<td>66</td>
<td>67</td>
<td>84</td>
<td>81</td>
<td>95</td>
<td>72</td>
<td>92</td>
<td>6.9</td>
<td>2.2</td>
<td>31.8</td>
</tr>
<tr>
<td>7</td>
<td>39 mo.</td>
<td>68</td>
<td>74</td>
<td>—</td>
<td>89</td>
<td>90</td>
<td>100</td>
<td>58</td>
<td>73</td>
<td>4.0</td>
<td>23.2</td>
</tr>
</tbody>
</table>

\[
PVR/SVR* (%) = \frac{1}{PBF/SBF} \times \left( \frac{PA_m - 5}{SA} \right) \times 100.
\]

*RV pressure = 85/0 mm Hg.

†Data obtained during operation.

SVC, superior vena cava; RA, right atrium; IVC, inferior vena cava; RV, right ventricle; PA, pulmonary artery; SA, systemic artery; PVR, pulmonary vascular resistance; SVR, systemic vascular resistance; PBF, pulmonary blood flow; SBF, systemic blood flow; and PA_m, pulmonary artery mean pressure.

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This 21-month-old white boy had open-heart surgery for closure of the ventricular septal defect, and at the time of open-heart surgery the muscular septum was resected. The patient died in the postoperative period with congestive heart failure.

Case 2 (JHH 84 02 86)

This 14-month-old white girl was thought to have a ventricular septal defect and a patent ductus arteriosus. A catheter was passed into the pulmonary artery, and the ductus arteriosus was found to be patent. The ductus was closed and resection, with the exception of the muscular portion of the ventricular septum, was performed. The patient died in the postoperative period with congestive heart failure.

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from the apex; in addition, there were several probe-patient muscular fenestrations.

Case 6 (JHH 104 52 60)

This 9-year-old white boy underwent open-heart surgery with cardiopulmonary bypass. Two muscular ventricular septal defects were noted and closed with sutures and reinforced with two Teflon patches. There was no leak after the procedure. A tracheostomy was performed.

Postoperatively the patient required the Mörch respirator for 8 days. He subsequently did well and was discharged in satisfactory condition. He has continued to receive digitalis. He is clinically stable 1 1/2 years after surgery and able to exercise to a moderate degree. The second sound in the pulmonary area is still accentuated. A moderate decrease in heart size has occurred.

Case 7 (JHH 68 73 45)

This 5-year-old white girl was operated on with the use of extracorporeal circulation. There were two defects in the muscular portion of the ventricular septum. They measured 1 by 1 cm. and 2 by 1 cm., respectively, and were located near the apex. Other defects were searched for but not found. Both defects were closed with sutures. Postoperatively ventricular fibrillation occurred and could not be successfully reverted. The child died shortly thereafter.

At autopsy (no. 26799) the heart weighed 250 Gm. and was greatly enlarged. The thickness of the left ventricular myocardium was 0.7 cm. and the right was 0.5 cm. The atrial septum was intact and the foramen ovale was covered by a flap. In addition to the two closed defects in the muscular ventricular septum, there were three 3.5-mm. defects very low in the muscular septum and several probe-patient interventricular communications. The septum resembled a sieve.

Discussion

A number of authors have considered isolated defects in the lower portion of the muscular septum of no clinical importance because they thought little hemodynamic derangement occurred. Edwards\textsuperscript{12} observed the defects to be ovoid in infancy and considered that they lengthened and almost coapted their edges in adulthood. Veasy\textsuperscript{15} and Selzer\textsuperscript{2,10} reasoned that contraction of the septal muscle mass in systole closed or decreased the size of the defects. Applying the same reasons as the latter two authors, Dammann et al.\textsuperscript{11} considered muscular defects to be functionally small, although they recognized the ten-
dency for the defects to be multiple and anatomiclly large. Imperial et al.\textsuperscript{4} believed that a multiple fenestrated defect offered more resistance to shunt flow than the equivalent cross-sectional area of a single defect and hence had less hemodynamic effect.

Contrary views have been expressed by others. Taussig\textsuperscript{16} has stated that although small in size, multiple small defects may create a large left-to-right shunt and are often associated with marked right ventricular and pulmonary artery hypertension. Young et al.\textsuperscript{5} and Heath et al.\textsuperscript{9} cited several patients in whom muscular defects produced marked cardiomegaly and severe pulmonary hypertension. Young et al.\textsuperscript{5} regarded multiple large defects in the lower part of the septum as basically a single ventricle.

Instances of multiple muscular ventricular defects can be found in a number of other reviews. Zacharioudakas et al.\textsuperscript{7} reported the deaths of two 3-month-old infants with right ventricular and pulmonary hypertension. In the series of Warden et al.,\textsuperscript{8} a 6-year-old and a seriously ill 5-month-old patient died from their cardiac condition. In an autopsy analysis Mason and Hunter\textsuperscript{17} cited a 7-year-old girl with marked cardiomegaly. The two cases in Ash's study\textsuperscript{18} were in severe congestive heart failure in infancy. Cardiac catheterization demonstrated right ventricular and pulmonary hypertension in both. The multiple defects were uneventfully closed at age 4 in one patient. In the second patient it was not technically possible to close all of the multiple defects.\textsuperscript{19}

Difficulty in surgically identifying multiple muscular ventricular septal defects has been experienced by Cooley et al.\textsuperscript{20} and Bernhard, Innis, and Gross.\textsuperscript{3} The latter group described two patients in whom surgical exploration detected but one of the defects. Reoperation on their one survivor was especially difficult. As a result of their experience, Bernhard et al.\textsuperscript{8} developed an instrument for intracardiac transillumination of the ventricular septum. Similarly, cases 1, 5, and 7 described herein had incomplete surgical repairs. The defects were hidden within the spaces between the hypertrophied trabeculae carneae.

The consequences of incomplete surgical repair resulting from a failure to recognize the multiple nature of the defects has recently led to our inclusion of left heart angiocardiography in the routine evaluation of ventricular septal defects. The authors have not yet diagnosed the lesions prior to surgery and are aware of only one case in which multiple muscular ventricular septal defects were radiographically demonstrated during life.\textsuperscript{22}

What accounts for the severity of the clinical picture in our young patients? Certainly the many factors that influence the type of patient seen at a particular institution must be considered. Our group of patients may not be representative of all those with multiple muscular ventricular septal defects. The consistent picture of severe heart disease in each of these patients does suggest, however, that this malformation creates findings more compatible with a high membranous defect than an isolated muscular defect. Let us examine the possible reasons for this.

In no case did the aorta override the defect and directly communicate with both ventricles. The catheterization data and the histologic survey of the pulmonary vasculature did not describe especially significant pulmonary vascular obstruction. The pulmonary hypertension was most likely of the high-flow, low-resistance type. Although the pulmonary resistance was elevated in case 5, this may have been the result of a falsely low arterial oxygen saturation. Grade 1 pulmonary vascular changes were noted in case 5 at autopsy.

We cannot precisely relate the anatomic size of the defects to the pathophysiologic findings. It is likely that in many of our cases the septal fenestrations behaved much like a fish net and allowed greater functional stress than would otherwise have been supposed. In these cases the thinness of the septum precluded the notion that septal contraction would be capable of significantly altering the left-to-right shunt in systole. In the remaining cases the additive cross-sectional areas of the multiple defects approximated the size of a single large defect. Although systole may reduce the size of the defects due to septal

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contraction, in these cases it may be that a significant left-to-right shunt occurred in diastole.

Finally, it is necessary to recall that several of our patients were subjected to open repair at a time when technical methods were less optimal than at present. Had our current attitudes been applied, a pulmonary artery banding procedure would have been recommended in the majority of these cases.

Summary

Defects in the muscular portion of the interventricular septum are frequently multiple. Although their incidence is significant, scant attention has been paid them as a distinct entity. This report concerns seven patients with multiple muscular ventricular septal defects seen between 1956 and 1962. The patients ranged in age from 4 months to 9 years; five were less than 2 years old. Symptoms and signs of growth retardation, failure to thrive, and marked cardiac decompensation were present in each case. Electrocardiographic evidence of combined ventricular hypertrophy and incomplete right bundle-branch block was noted in each instance. Right heart catheterization demonstrated marked pulmonary hypertension of the high-flow, low-resistance type. These routine studies could not distinguish multiple muscular ventricular septal defects from the more usual membranous septal defect. All patients had cardiac surgery. There was one survivor. In three instances difficulty in identifying all of the multiple fenestrations prevented complete surgical repair. Postmortem examination was done in six patients. The sievelike appearance of the muscular ventricular septum was the most striking gross finding in the majority of the cases. Histologic analysis of the pulmonary vascular structural alterations did not reveal marked changes in any of the cases.

In contrast to the widespread belief that most isolated muscular septal defects are innocuous, these seven examples of multiple muscular defects were associated with a picture of severe heart disease. Every effort should be made to diagnose the lesion prior to surgery. Recognition of the multiple nature of the defect is a necessary preliminary to successful therapy. It is hoped that preoperative left heart angiocardiography will successfully demonstrate the multiple lesions.

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