Multiple Muscular Ventricular Septal Defects

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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.

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 8½ 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 8½ 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
Concise locations of ventricular septal defects

\[ \text{Crista supraventricularis} \]
\[ \text{Pulm. valve} \]
\[ \text{Tricuspid valve} \]

Multiple vent. septal defects (sievel type)

**Figure 1**

Anatomical classification of defects of the interventricular septum (adapted from Kirklin and associates.6)

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 angiocardiology performed in three patients demonstrated recocapsulation 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° and +175°. The QRS axis was +65° in one patient and −165° 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

**Figure 2**

Representative roentgenograms, cases 2 (left) and 4 (right). Cardiomegaly, a prominent pulmonary conus, and increased pulmonary vascularity are demonstrated.
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 hematoxylin 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. 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

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*Figure 3*

Representative electrocardiogram, case 7, showing incomplete right bundle-branch block and combined ventricular hypertrophy.
lumen of 2 mm. was ligated. A small opening in the muscular septum was ligated and thought completely closed. No other defects were noted. The patient died of intractable congestive failure several hours after surgery.

At autopsy (no. 25686) the foramen ovale was closed. The right atrium and right ventricle were dilated and hypertrophied. The left ventricle was hypertrophied. There was a sutured 5-mm. defect low in the muscular septum and another defect in the muscular septum lying posterior behind a papillary muscle of the tricuspid valve. It measured 1.3 by 0.8 cm. from the left ventricular side and 0.6 cm. from the right ventricular side. There was a mild coarctation of the aorta. The ascending aorta was 2.3 cm. in diameter, the arch 1.1 cm. and the descending aorta 1.7 cm.

Case 2 (JHH B-45893)

This 14-month-old white girl was thought to have a ventricular septal defect and a patent ductus arteriosus. At exploratory thoracotomy the ductus was found to be obliterated. The chest was closed and reoperation with the aid of cardiopulmonary bypass was planned. On the third postoperative day poor air exchange was noted and the child was bronchoscopy. Cardiac arrest occurred after the insertion of the bronchoscope; unfortunately, resuscitative measures failed.

At autopsy (no. 27740) the heart weighed 120 Gm. and showed enlargement of the right atrium and both ventricles. The thickness of the left ventricular myocardium was 0.8 cm. and the right 0.7 cm. The aorta was normal. The ductus was obliterated. The atrial septum was intact, and the foramen ovale was sealed. There were many separate defects in the muscular ventricular septum (fig. 4).

Case 3 (JHH 82 02 86)

This 21-month-old white boy had open-heart surgery for closure of his ventricular septal defect. At operation multiple defects were found in the muscular portion of the ventricular septum. The septum was described as "sieve-like." The defects were unsuccessfully sutured, and the child died shortly after leaving the operating room.

At autopsy (no. 28438) the heart weighed 170 Gm. All four chambers were enlarged. The left ventricular myocardium was 1.0 cm. thick, and the right was 0.6 cm. The pulmonary artery was dilated. The ductus arteriosus was obliterated. The foramen ovale was closed and the atrial septum was intact. Despite sutures along the course of the muscular septum, it was still sieve-like. Channels coursed back and forth between hypertrophied trabeculae. In addition, there was a defect 1.0 cm. in diameter located in the lowest portion of the septum.
MUSCULAR VENTRICULAR SEPTAL DEFECT

Case 4 (JHH B-33876)

This 8-month-old white boy underwent surgery with the use of cardiopulmonary bypass. The ductus arteriosus did not appear patent and no thrill was felt. Several holes were seen in the muscular septum but blood continuously welled up in the right ventricle and the operation was stopped before complete closure. The patient died shortly after completion of surgery.

At autopsy (no. 26826) the heart was markedly enlarged, particularly the right ventricle. The ductus was probe-patent and had an internal lumen of 1 to 2 mm. The foramen ovale was open 3 mm. and covered by a flap. There were multiple honeycomb-like fenestrations in the muscular ventricular septum. In addition, a 4 by 5 mm. and a 3 by 7 mm. defect were located in the muscular septum.

Case 5 (JHH 86 59 98)

This 27-month-old white boy was operated on with cardiopulmonary bypass. The muscular ventricular septum was noted to contain two longitudinal slits between the trabeculae. Their size was difficult to estimate because of their irregular shape. The defects were sutured but leakage from the left ventricle continued and a Teflon patch was incorporated into the repair. There remained a minimal leak with each beat. The ventriculotomy was closed and bypass discontinued. Ventricular fibrillation and ultimately asystole occurred shortly after his departure from the operating room.

At autopsy (no. 30605) the ductus arteriosus was obliterated. The atrial septum was intact, and the foramen ovale was sealed. The ventricular septal defects described above were noted 2 cm. from the apex; in addition, there were several probe-patent 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-patent 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. Edwards12 observed the defects to be ovoid in infancy and considered that they lengthened and almost coapted their edges in adulthood. Veasy15 and Selzer2,10 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.11 considered muscular defects to be functionally small, although they recognized the ten-
dency for the defects to be multiple and anatomi-
cally 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 hyper-
tension. 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
views. Zacharioudakas et al.\textsuperscript{7} reported the
defaths 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 pulmo-
nary hypertension in both. The multiple de-
fects 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 carnea.

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 angiocardiog-
raphy 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 ra-
diographically 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 mul-
tiple 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 ventri-
cles. The catheterization data and the histo-
logic survey of the pulmonary vasculature
did not describe especially significant pul-
monary 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 vas-
cular 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 pre-
cluded 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 re-
duce the size of the defects due to septal
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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