Congenital Aneurysm of the Membranous Portion of the Ventricular Septum

A Cause for Holosystolic Murmurs

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Congenital aneurysms of the membranous ventricular septum are rare. In 1936 Lev and Saphir1,2 found 70 cases in the world's literature. They added two cases of their own. We have been able to find 17 cases reported subsequently,3-9 of which three were diagnosed prior to death or surgery by means of cineangiography.10-12 We wish to present a fourth case diagnosed by cineangiography that was manifested clinically by a loud holosystolic murmur in spite of absence of an intracardiac shunt or valvular lesion.

Case Report

A 29-year-old woman was referred to the Indiana University Medical Center for evaluation of a heart murmur that was first detected when she was 13 years old. At that time she had a febrile illness associated with arthralgia and jaundice. Subsequently, the patient had three un-

Figure 1

Phonocardiogram showing a holosystolic murmur which is louder at the left sternal border in the fourth intercostal space than at the base.
complicated pregnancies and remained relatively well until May 1963, when during hospitalization for a severe respiratory illness, he had sudden, transient blurred vision, light-headedness, dyspnea, tachycardia, hypotension, and cyanosis. The etiology of this episode is unclear. No cardiac arrhythmia was recorded at that time.

Physical examination revealed a healthy, well-developed woman. There was no cyanosis or clubbing. The blood pressure was 140/88 mm Hg. The pulse was 90 beats per minute and regular. There was a grade-III/VI blowing, holosystolic murmur, heard best at the fourth interspace along the left sternal border. There was no thrill or diastolic murmur. The second heart sound split normally (fig. 1).

Electrocardiogram was normal. Cardiac fluoroscopy revealed normal heart size, but was unusual in that the pulmonary outflow segment of the left heart border was concave and the aorta was mildly elongated.

Cardiac catheterization and indocyanine dye curves revealed no evidence of peripheral arterial desaturation and no evidence of a shunt. The pressures in all four cardiac chambers were unremarkable, except that a small systolic gradient (7 mm Hg) was present between the right ventricle and the main pulmonary artery. Cineangiograms were remarkable in that they showed a “cauliflower” aneurysm of the high portion of the ventricular septum (fig. 2). The apex of the aneurysm projected into the right ventricle (fig. 3). No shunts were demonstrated during cineangiography.

**Discussion**

Aneurysms of the membranous part of the ventricular septum are thought to be congenital. In 1912, Mall proposed that this defect resulted from insufficient shifting of the ventricular septum to the right and the aorta to the left. Lev and Saphir's embryologic review supported Mall's theory. They concluded that the defect was, in fact, a mild form of transposition.

Congenital aneurysms of the membranous

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

**Figure 2**

Left. Left ventricular injection, left anterior oblique view. The aneurysm appears lobulated and arises high on the interventricular septum. Right. A retouched frame of the cine sequence.
ventricular septum are often of no clinical significance; however, they tend to be associated with other anomalies such as Mongolism and subaortic stenosis.\textsuperscript{14}

Ventricular septal aneurysms are sometimes associated with ventricular septal defects. It is not clear whether the aneurysm perforates, or whether the defect is part of the same developmental error.

Guccione\textsuperscript{15} reported a case of septal aneurysm associated with the formation of a thrombus in the aneurysm and endocarditis of the aortic valve. He concluded that the endocarditis resulted in the aneurysm; however, most authors favor the theory of congenital origin of Mall.

Peräsaolo\textsuperscript{8} and colleagues reported a case with an extremely interesting complication. Cardiac catheterization data suggested infundibular pulmonary stenosis with a systolic gradient of 50 mm. Hg between the infundibulum and body of the right ventricle. At operation, a large aneurysm of the membranous septum was found to be causing obstruction to right ventricular outflow. There was no pulmonic stenosis. It is possible, but highly conjectural, that our patient's symptoms in May 1963 were caused by temporary obstruction of the right ventricular outflow tract by the aneurysm. Another possible cause for our patient's symptoms might have been the occurrence of arrhythmia, for congenital aneurysms of the ventricular septum have been associated with arrhythmias.\textsuperscript{4, 5}

Holosystolic murmurs heard best at the fourth left interspace or apex are usually due to ventricular septal defects or incompetent atrioventricular valves. In our patient, the only lesion found was an aneurysm of the

\textbf{Figure 3}

\textit{Left. Right atrial injection. Contrast material fills the right atrium and right ventricle. The aneurysm presents as a negative filling defect in the right ventricular cavity. Right. A retouched frame of the cine sequence.}
ventricular septum well visualized on the cineangiograms. No shunt or valvular lesion was present; consequently, it is reasonable to conclude that the septal aneurysm alone was responsible for the murmur. Only two other cases of isolated aneurysms of the ventricular septum with systolic murmurs have been described in the literature. Lekisch 11 presented a case with a “grade III late systolic murmur. High pitched and of musical type, it was heard best over the apex and along the left sternal border.” He also attributed the murmur to the aneurysm. In one of Larsen and Noer’s 7 cases, a “thin sibilant murmur in late systole” was present. At autopsy the aneurysm protruded into the tricuspid ostium and appeared to elevate the septal valve into the ostium. The authors suggested that the valve deformity gave rise to the murmur. Tricuspid insufficiency probably did not give rise to the murmur in our patient. The right atrial pressure tracings do not suggest tricuspid insufficiency and the cineangiograms do not suggest that the aneurysm is deforming the valve.

Summary

A case of an isolated aneurysm of the ventricular septum with a loud holosystolic murmur at the fourth left interspace is presented. The origin of the murmur is ascribed to the aneurysm of the ventricular septum.

References

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