Aneurysm of the Membranous Ventricular Septum

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Aneurysm of the membranous portion of the interventricular septum is considered to be a rare congenital anomaly, and to date only a few publications have documented ante-mortem diagnoses. Taussig (1960) reviewed the published reports and was able to find 80 cases diagnosed at necropsy. Steinberg (1957), using angiocardiographic studies, reported the first living patient with this defect. Two recent reports brought the total number discovered by angiocardiography to 7 (Kasparian, Brest, and Novack, 1965; Edelstein and Charms, 1965). Using selective left ventricular angiocardiography we have found 10 patients at this centre during the past two years.

ANATOMY AND PATHOGENESIS

The membranous septum is the weakest portion of the interventricular septum. It not only separates the left ventricle from the right ventricle, but also the left ventricle from the right atrium behind the tricuspid valve (Larsen and Noer, 1960). Aneurysmal dilatation of this portion is usually of small size, varying from 1–3 cm. in diameter. Rarely these sacs expand to obliterate the right atrium or the out-flow tract of the right ventricle (Gould, 1960). When located in the muscular portion of the septum, these dilatations are usually considered to be acquired and many have been found in patients with a history of a myocardial infarction (Arons and O'Rourke, 1954; Valle-Cavero and Maquera, 1961). Most membranous aneurysms are congenital in origin, but cases due to trauma and infection have been described (Lev and Saphir, 1938). The most plausible theory for the development of an aneurysm in the membranous portion of the septum relates its formation to the natural spontaneous closure of a ventricular septal defect (Edelstein and Charms, 1965; Agustsson *et al.*, 1963; Arcilla *et al.*, 1963). Since this area is weak, the tissue responds to the pressure exerted by the left ventricle and bulges to the right.

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CLINICAL PICTURE

The clinical spectra of the 10 patients is summarized in the Table.

Age. The oldest patient was 20 and the youngest patient was 6 at the time of catheterization.

Sex. Male and female patients are equally distributed in this series.

Pre-catheterization Diagnosis. A ventricular septal defect was diagnosed in all but one patient, who was considered to have an atrial septal defect.

Signs and Symptoms. All the children were known to have had a heart murmur since birth or the murmur was discovered in early childhood. None of the patients was truly symptomatic. Cases 5 and 7 gave histories of repeated colds and easy fatigability. Case 9 had undergone surgical repair for her ventricular septal defect one year before admission and was restudied because a murmur persisted and she had developed the electrocardiographic pattern of complete right bundle-branch block.

Physical examination of the heart revealed a Grade 2–4/6 pansystolic murmur in some and ejection type murmur in others. The murmur was heard in the left præcordium with maximal intensity in the third and fourth left intercostal spaces along the left sternal border. In some cases the murmur was accompanied by a thrill. The second sound in the pulmonary area was either accentuated or normal with normal splitting with respiration. The intensity, location, character, and radiation of the murmur in no way served to distinguish these patients from those with ventricular septal defects.

Electrocardiogram. All tracings were within normal limits except for Case 6 which had the electrocardiographic pattern of a complete right bundle-branch block.

Radiographic Appearance of the Heart and Lungs. The heart and the lung parenchyma were within

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TABLE

Case No.	Age (yr.)	Sex	Pre-catheterization diagnosis	Signs and symptoms	Electrocardiogram	Chest radiograph	Cardiac catheterization data	Left ventricular cine-angiograms	Congenital or acquired
1	20	F	ASD	Heart murmur at 5 mth.; soft systolic grade 2 murmur best heard at pulmonary area; P2 increased and split; asymptomatic	Normal	Prominent pulmonary artery	Gradient of 10 mm. Hg between RV and PA; no oxygen step-up on right side; PA 20/5; RV 30/0	Aneurysm of VS; small VSD	Congenital
2	11	M	VSD	Asymptomatic; pansystolic murmur and thrill since birth; murmur and thrill disappeared in past five years; P2 normally split	Normal	Normal	Normal pressures and no oxygen step-up	Aneurysm of VS and small septal defect	Congenital
3	13	M	VSD	Murmur since infancy; asymptomatic; systolic thrill and grade 3 pansystolic murmur left sternal border; P2 split	Normal	Pulmonary vasculature slightly increased	Normal pressures and no oxygen step-up on right side of heart	Aneurysm of VS and small septal defect	Congenital
4	9	M	VSD	Murmur since infancy; repeated upper respiratory catarrh; systolic thrill and grade 3-4 ejection systolic murmur at left sternal border (3rd and 4th intercostal space)	Normal	Pulmonary vasculature and heart size slightly increased	Gradient of 15 mm. Hg between RV and PA; no oxygen step-up on right side of heart; PA 20/5; RV 35/0	Aneurysm of VS; small VSD	Congenital
5	9	F	VSD	Murmur since birth; repeated colds; grade 3-4 ejection systolic murmur over left praecordium; P2 normally split	Normal	Pulmonary vasculature increased	Normal	Aneurysm of VS; small VSD	Congenital
6	14	F	Questionable VSD	Heart murmur since birth; asymptomatic; grade 3-4 pansystolic murmur over left praecordium; P2 split	Normal	Minimal fullness of pulmonary artery	Normal pressures; step-up of oxygen; PA 85%; aorta 97%; RA 70%	Aneurysm of VS with small VSD	Congenital
7	11	M	VSD	Heart murmur since birth; some fatigability for one year; grade 3 ejection systolic murmur left third intercostal space; P2 normal	Normal	Normal	Normal	Small VSD and ventricular septal aneurysm	Congenital
8	15	F	VSD	Heart murmur since birth; grade 3 or 4 pansystolic murmur third and fourth left intercostal space; P2 accentuated; asymptomatic	Normal	Prominent pulmonary artery	Normal	Persistent left superior vena cava; small VSD and ventricular septal aneurysm	Congenital
9	6	F	VSD (post-op.)	Murmur since birth; cardiac failure at 5 years of age; repair of VSD; now thrill and grade 3-4 ejection systolic murmur persisting in third and fourth left sternal border; asymptomatic	Normal	Slightly increased lung markings	Before repair RV and PA pressures equal to systemic pressure; VSD but no aneurysm before operation	Small ventricular aneurysm in membranous portion	Acquired
10	8	M	VSD; question of pulmonary stenosis	Heart murmur since age 3 years; thrill and systolic murmur in left sternal border; P2 normal	Normal	Increase in pulmonary vasculature	Normal pressures; step-up of oxygen in pulmonary artery; oxygen saturation: PA 87%; RA 73%; aorta 98%	Aneurysm of membranous septum	Congenital

ASD atrial septal defect; VSD ventricular septal defect; PA pulmonary artery; RA right atrium; RV right ventricle.

normal limits in all parameters in three cases. The remainder had either prominent pulmonary artery segments or hypervascular lung fields or both. Heart size in one case was considered to show minimal enlargement in the transverse diameter.

Hæmodynamic Studies. Five cases had absolutely normal pressures and oxygen saturation on both

sides of the heart. Cases 6 and 10 showed oxygen step-ups in the pulmonary artery of 15 and 14 per cent, respectively, while the pressures were within normal limits. Cases 1 and 4 showed normal oxygen saturations but had gradients of 10 mm. and 15 mm. Hg across the pulmonary valve, respectively. Case 9 had pulmonary artery and right ventricular systolic pressures equal to systemic pressures before the surgical repair of the ventricular septum.

FIG.—Cine-angiogram of injection into the left ventricle. Oblique view of the heart. The frame to the left is taken during ventricular diastole and the frame to the right during systole. The arrow indicates the aneurysm.

One year after repair, repeat hæmodynamic studies were within normal limits.

Selective cine-angiographic studies of the left ventricle in postero-anterior and oblique views showed 1–3 cm. size aneurysmal dilatations of the membranous septum (Fig.). Most studies also demonstrated a small ventricular septal defect. Case 8, in addition to a ventricular septal defect, showed a persistent left superior vena cava.

The following two narratives are of interest.

Case 2. An 11-year-old Caucasian boy was the full-term product of unrelated parents. His mother had no ailments during the antenatal period. A loud grade 4/6, pansystolic murmur and a thrill were detected in infancy and still noted at 5 years of age. Over the years the patient remained under strict medical surveillance. The thrill disappeared and the murmur was hardly audible at the time of catheterization at 11 years of age. The electrocardiogram, cardiac series, and other routine laboratory studies remained normal. Hæmodynamic studies showed normal pressures and oxygen saturations. Cine-angiographic studies showed a 2×1 cm. aneurysm of the membranous portion of the interventricular septum and a small ventricular septal defect. No surgical intervention was contemplated and the patient remains under active medical follow-up.

Case 9. A 6-year-old Caucasian girl was the full-term product of unrelated parents. No ailments were encountered during the pregnancy. A harsh ejection systolic murmur of grade 3–4/6 and a thrill were detected in the left præcordium in infancy. At 5 years of age, she developed mild congestive heart failure. Hæmodynamic studies at this time revealed a left-to-right shunt at the ventricular level. The right ventricular and pulmonary

artery systolic pressures were equal to systemic pressure. The chest radiograph showed a slight increase in heart size, and the electrocardiogram was within normal limits. Cine-angiograms of the heart showed a ventricular septal defect in the upper portion of the septum. Surgical repair of this defect was carried out and the child had an uneventful post-operative course. A repeat cardiac catheterization was carried out due to the persistence of a thrill and a murmur. The data showed normal pressures and oxygen saturation. Cine-angiograms revealed a 1.5×1 cm. aneurysm of the membranous septum and a residual ventricular septal defect. No further operations were carried out as the child became completely asymptomatic.

FOLLOW-UP

All 10 cases have remained asymptomatic. None of them warrant surgical intervention. As little is understood about the natural history of these aneurysms, a strict medical surveillance is considered necessary.

DISCUSSION

Selective angiocardiology is the best means to diagnose membranous ventricular septal aneurysms, and they are consistently seen as an outpouching immediately below the root of the aorta (Edelstein and Charms, 1965).

The abnormalities most commonly associated with these aneurysms are aortic insufficiency (Leckert and Sternberg, 1950), membranous sub-aortic stenosis (Rae, 1936), complete atrio-ventricular block (Clark and White, 1952) and other arrhythmias (Rogers, Evans, and Domeier, 1952), mongolism (Lev and Saphir, 1938), and coarctation

of the aorta (Kolesov, 1963). None of our cases showed the above associated features. Obstruction of the right ventricular outflow tract by an aneurysm was described by Das, Jahnke, and Walker (1964). Two of our cases showed mild pulmonary valve gradients. However, the gradient may represent mild valvular pulmonary stenosis rather than mild right ventricular outflow tract obstruction due to aneurysm. Rupture, infection, and thromboembolism have been documented (Peräsalo *et al.*, 1961) as complications of septal aneurysms. All the cases in our series presented as ventricular septal defects; the clinical picture, and electrocardiographic and radiographic findings, did not help to differentiate them from classical uncomplicated ventricular septal defects.

It is intriguing to apply the hypothesis that these aneurysms of the membranous septum occur during the spontaneous closure of an interventricular septal defect. Especially in Case 2, we were convinced that the diminution in the intensity of the murmur, the disappearance of a thrill, and the normal radiograph and electrocardiographic findings together with the over-all well-being of this boy, represented closure of a septal defect. Since no studies were performed early in life, we do not know whether an aneurysm is a late manifestation or an early accompaniment of a ventricular septal defect. As yet, membranous septal aneurysms have not been documented in infancy.

With so little known as to the course of patients with this defect, we plan no immediate surgical intervention on patients who are asymptomatic. These patients are being followed closely and plans have been made to repeat the angiographic studies at appropriate intervals.

SUMMARY

Ten cases of membranous ventricular septum aneurysm (nine congenital and one acquired) have been described. Their clinical features, hæmodynamic and cine-angiographic studies have been outlined. The only sure way of diagnosing this anomaly is by selective angiocardiography.

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