Membranous Subaortic Stenosis Complicated by Aneurysm of the Membranous Septum and Mitral Valve Prolapse

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SUMMARY The clinical, echocardiographic, and catheterization findings in a patient with discrete subaortic stenosis, aneurysm of the membranous interventricular septum, and mitral valve prolapse are presented. Echocardiography showed a subaortic membrane, abnormal aortic valve motion, accentuated systolic anterior motion of the membranous interventricular septum, and prolapsing mitral leaflets. Cardiac catheterization confirmed the diagnoses. The possible functional interrelationship of these lesions is discussed.

DISCRETE MEMBRANOUS SUBAORTIC STENOSIS is a rare cause of left ventricular outflow obstruction which may coexist with other congenital lesions such as membranous ventricular septal defect. 1, 2 A membranous ventricular septal defect may close spontaneously leaving a residual aneurysm of the membranous septum. 3 An association between discrete subaortic stenosis and aneurysm of the membranous septum therefore would not be surprising, and this association has been reported. 4 Further, an association between left ventricular outflow tract obstruction and anomalies of the mitral valve is recognized, resulting either from the bulging of accessory valvular tissue into the left ventricular effluent or from the abnormal insertion of a normal or cleft mitral leaflet to the interventricular septum causing narrowing of the left ventricular outflow tract. 5, 6 In these instances, however, a discrete fibrous membrane would be absent.

The echocardiographic features of discrete subaortic stenosis are narrowing of the left ventricular outflow tract with premature systolic closure and systolic fluttering of the aortic valve cusps; 7, 8 echocardiographic recording of the subaortic membrane has also been reported. 9, 10 Demonstration of an aneurysm of the membranous septum by echocardiogram has likewise been reported, showing accentuated systolic anterior motion of the membranous septum. 11 This report describes the clinical, echocardiographic, and

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catheterization features of a case with the combination of three lesions: discrete subaortic stenosis, aneurysm of the membranous septum, and mitral valve prolapse.

Case Report

A 27-year-old asymptomatic male was referred for cardiac evaluation because of a heart murmur first discovered during a routine physical examination nine years before. Since the patient had no cardiovascular complaints, the murmur had not been further investigated until its significance was questioned in regard to insurability. The only family history of congenital or acquired cardiac disease was the clinical and echocardiographic documentation of prolapse of the mitral valve in his mother several years previously.

Physical examination revealed a slender, normal appearing young male with a blood pressure of 116/78 mm Hg in both arms. Carotid upstroke was normal and symmetrical. A systolic thrill was palpable at the apex and left sternal border. An early systolic click was noted (fig. 1). A grade IV/VI systolic ejection murmur was audible, maximal at the left sternal border with radiation into the carotids bilaterally. The murmur decreased in intensity during the strain phase of the Valsalva maneuver. No diastolic murmur could be detected despite multiple examinations.

The electrocardiogram revealed prominent QRS voltage which may be within normal limits for a slender male under
30 years of age. There were no other criteria for left ventricular hypertrophy. The chest X-ray was normal.

**Echocardiographic Findings**

The echocardiogram was recorded from the fourth intercostal space at the left sternal border, using a Unirad echocardiograph/recorder and a 2.25 MHz transducer 13 mm in diameter with a 10 cm focal zone. Cardiac wall and cavity dimensions were within normal limits. There was no left ventricular outflow tract narrowing. However, an abnormal echo was recorded within the left ventricular outflow tract from a structure which moved sharply anteriorly during systole (fig. 2). High frequency systolic and diastolic vibrations of this structure were noted. The membranous septum showed accentuated systolic anterior motion (1.2 cm), coincident with the brisk anterior movement of the left ventricular outflow tract structure (fig. 3). The distal septum demonstrated a normal posterior systolic movement. The posterior aortic valve cusp partially closed early in systole and both cusps vibrated coarsely through the remainder of systole (fig. 4). There was no high frequency diastolic fluttering of the anterior mitral leaflet, but systolic posterior bowing was evident compatible with mitral valve prolapse.

**Cardiac Catheterization Data**

After informed consent was obtained from the patient, right and left heart catheterization was performed. On pullback from the left ventricular body to the ascending aorta, a 14 mm Hg peak systolic gradient was demonstrated across the left ventricular outflow tract in the subvalvular region (fig. 5). Left ventricular angiography revealed a subaortic membrane, an aneurysm of the membranous septum, and prolapse of the posterior mitral leaflet (fig. 6). Aortic root angiography showed grade II/VI aortic regurgitation. No left-to-right shunt was detected by hydrogen inhalation with the sensing electrode positioned in the main pulmonary artery.

**Discussion**

Discrete subaortic stenosis is frequently associated with other congenital cardiac defects including membranous ventricular septal defects1-2 or mitral valve anomalies. Membranous ventricular septal defects often close spontaneously during childhood. Formation of an aneurysm of the membranous septum may accompany diminution in the size or obliteration of membranous ventricular septal defects. Although the etiology of the aneurysm of the membranous septum in our patient is uncertain, it may have formed during the spontaneous closure of a membranous ventricular septal defect.

An early systolic click is not a reported feature of discrete subaortic stenosis, and its presence in a patient with clinical left ventricular outflow obstruction would usually suggest aortic valvular stenosis. The click in this patient, however, was a clue to the association of discrete subaortic stenosis with a second (or third) anomaly. Early systolic clicks have been reported to be associated with an aneurysm of the membranous septum and with mitral valve prolapse syn-
drome, either or both of which may have caused the click in our patient.

The echocardiographic features of discrete subaortic stenosis are left ventricular outflow tract narrowing and premature aortic valve systolic closure, with systolic fluttering of the aortic valve cusps. Direct recording of the subaortic membrane has been reported in two cases. In both, the membrane was situated between the anterior mitral leaflet and the interventricular septum throughout the cardiac cycle. In one case, there was slight systolic anterior motion of the subaortic membrane, and in the other, the structure labeled "fibrous membrane" moved posteriorly during systole. Since subvalvular membranes are typically attached to the membranous septum and to the anterior leaflet of the mitral valve, the prominent systolic anterior motion of the subvalvular membrane in this patient might possibly be secondary to the membrane's attachment to the bulging aneurysm of the membranous septum. Further, such an anatomical interrelationship may have resulted in traction on, and anterior displacement of, that portion of the anterior leaflet attached to the fibrous membrane. This in turn may have caused distortion of normal systolic apposition ("keystone support") of the two mitral leaflets, thereby permitting the posterior leaflet to prolapse during systole.

Since aortic insufficiency is commonly associated with discrete subaortic stenosis, high frequency diastolic fluttering of the subvalvular diaphragm would be an expected finding. In this case, the diastolic vibrations of the membrane strongly suggested the presence of aortic insufficiency despite the absence of a murmur, and this was confirmed angiographically. Significant aortic regurgitation is often associated with fluttering motion of the anterior mitral leaflet during diastole. This finding was absent in our patient. The membrane interposed between the aortic valve and the margins of the anterior leaflet of the mitral valve apparently shielded the anterior leaflet from the regurgitant jet and prevented its fluttering.

References

Membranous subaortic stenosis complicated by aneurysm of the membranous septum and mitral valve prolapse.
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