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Subvalvular aortic stenosis associated with dynamic outflow tract obstruction

A case report

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Hemodynamic and angiographic findings are described in a 27-year-old patient, who had undergone surgical operation for subvalvular membranous aortic stenosis. Left ventriculography revealed persistence or regrowth of the subvalvular fibrous ring, and hemodynamic data revealed associated dynamic obstruction of the left ventricular outflow tract. The authors emphasize the importance of uncovering associated dynamic obstruction by provocative maneuvers and use of Beta blockers.

THE simultaneous occurrence of membranous and dynamic subvalvular left ventricular outflow obstruction has been previously reported.[1] Surgical removal of the fibromembranous obstruction usually abolishes both the fixed and dynamic subvalvular gradient. In some patients, however, the dynamic subvalvular gradient persists after the fixed gradient has been relieved and gradually diminishes over a period of time.[2] We report the hemodynamic, echocardiographic, and angiographic findings in a patient in whom the development of symptoms suggests the occurrence of dynamic left ventricular subvalvular obstruction ten years after surgical correction of fixed left ventricular outflow obstruction due to a fibrous band.

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Case report

A 27-year-old Caucasian male underwent a resection of a fibromuscular sobaortic ring at another institution at age 17. Although he had experienced exertional dyspnea since childhood, the preoperative evaluation was prompted by a syncopal episode.

At operation, a crescent-shaped fibromuscular band was found extending from the bulging hypertrophied ventricular septum to the anterior leaflet of the mitral valve. The band was removed without a septal myotomy or myectomy. The patient's symptoms improved postoperatively, but he was advised to avoid strenuous activities.

The current evaluation was prompted by the development of chest pains and dyspnea in July, 1975. Although the precordial chest pains usually occurred with exertion, they occasionally occurred at rest. The dyspnea which occurred with moderate exertion was usually mild. There were also episodes of "fast heart beats" but no paroxysmal nocturnal dyspnea, orthopnea, or syncope. There was no family history of sudden death or heart disease.

Physical examination revealed a blood pressure of 110/80, pulse 80/minute and regular, height 5'8" and weight 217 pounds. The carotid pulses were normal. The cardiac apical impulse was forceful and located in the fifth intercostal space, 3 cm to the left of the mid clavicular line. There were no thrills, gallops or ejection click. The first and second heart sounds were normal and physiologically split. A Grade III/VI ejection systolic murmur was heard at the base radiating into the carotid vessels. No diastolic murmur was audible.

Chest x-rays revealed some left ventricular enlargement and a slightly dilated ascending aorta. There was no calcification in the aortic valve. The

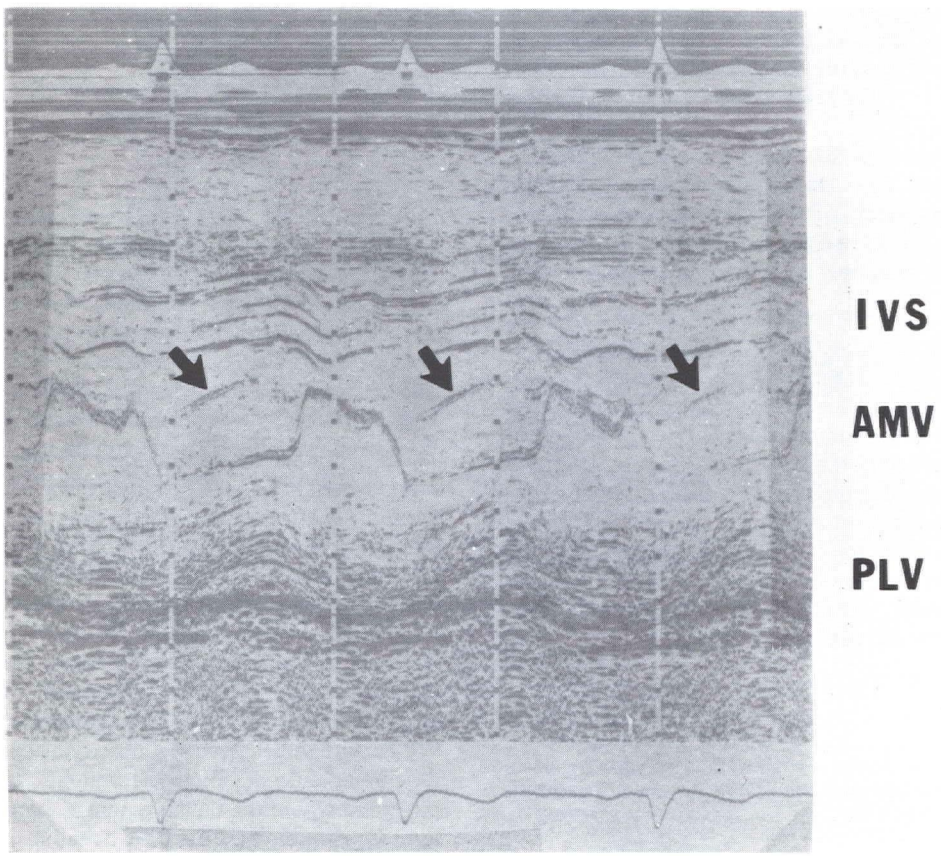

Figure 1
Echocardiogram: Note the thin echo (arrows) in the left ventricular outflow tract, suggestive of a systolic anterior motion of the mitral valve. There is no continuity in early and late systole.

electrocardiogram showed left ventricular hypertrophy with a strain pattern. Pulmonary function tests, hemogram and electrolytes were within normal limits.

The echocardiogram (Figure 1) revealed a thin echo in the left ventricular outflow tract, anterior to the mitral valve. Although the anterior mitral leaflet showed systolic anterior motion often observed in dynamic muscular left ventricular outflow obstruction, it was not typical because of the lack of contiguity in early and late systole. There was no septal hypertrophy and the septum to posterior left ventricular wall thickness ratio was within normal limits. Fluttering of the anterior leaflet of the mitral valve was consistent with aortic insufficiency.

Cardiac catheterization revealed normal right heart pressures, left ventricular end-diastolic pressure, O_2 consumption, cardiac output, and arteriovenous oxygen difference at rest. The left ventricular systolic pressure was 140 mm Hg at the apex and a 40 mm Hg systolic pressure gradient was documented as the catheter was pulled back to the subvalvular area from the apex with no further pressure drop across the aortic valve (Figure 2). The post ectopic beat showed an additional increase of 44 mm Hg in gradient with no change in the aortic pulse pressure. The Valsalva maneuver also increased the gradient. Amyl nitrate inhalation increased the gradient by an additional 40 mm Hg with a significant fall in systemic arterial pressure (Figure 3). Isoproterenol 0.002 mg IV caused a significant tachycardia, a rise in the systolic gradient to 120 mg Hg, and a fall in systemic arterial pressure (Figure 4). The patient complained of mid substernal pain during the isoproterenol infusion which was relieved as the systolic gradient was abolished with methoxamine. Subsequently, propranolol, 1.0 mg IV was given and the patient was rechallenged with isoproterenol 0.002 mg. At this time, despite a slight tachycardia, only a minimal systolic gradient developed and the patient reported no symptoms (Figure 5).

The left ventriculogram showed a mildly enlarged cavity with hypertrophied walls, good contractions, and no mitral regurgitation. A narrow subaortic chamber was present (Figure 6). The apical left ventricular cavity was not obliterated in systole. The motion of the anterior leaflet of the mitral valve appeared to be limited. There was no systolic apposition of the anterior leaflet against the septum. Aortic root angiogram showed moderate dilation of the ascending aorta, three cusps and 1+ to 2+/4+ regurgitation. Coronary arteriograms were normal.

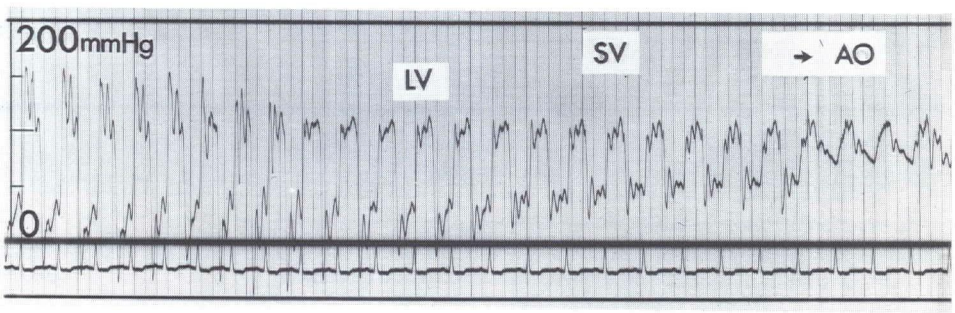

Figure 2
Pullback recording from left ventricle (LV) to aorta (AO). Note the systolic gradient as the subvalvular chamber (SV) is entered. There is no further pressure drop across the aortic valve.

Discussion

Fixed subaortic stenosis is a congenital anomaly characterized by the presence, at the subvalvular level, of a membranous or fibromuscular structure obstructing the outflow tract of the left ventricle. A rather uncommon anomaly, it occurs in four to ten percent of all cases of congenital left ventricular outflow obstruction.[3]

Reis et al classified congenital subvalvular aortic stenosis into discrete and tunnel forms.[2] The latter form was seen only in 9 of 33 patients studied. Based on angiocar-

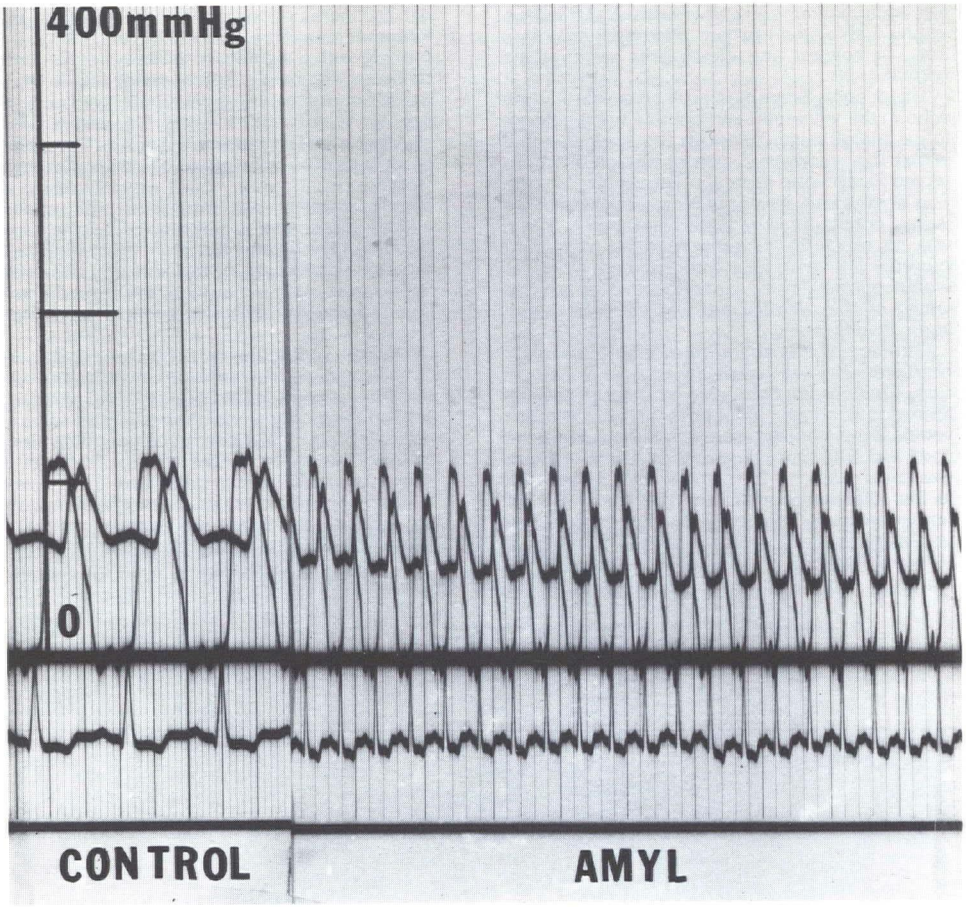

Figure 3
Simultaneous pressure obtained from the left ventricle and femoral artery. A gradient of 44 mm Hg developed after amyl nitrate inhalation.

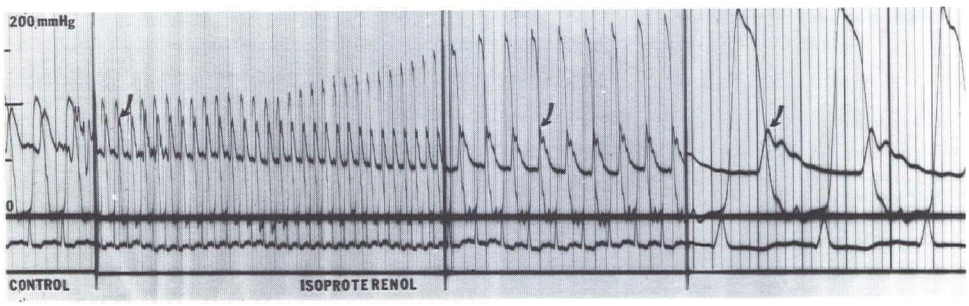

Figure 4
Simultaneous pressure tracings obtained from LV and FA. A gradient of 120 mm Hg developed after isoproterenol 0.002 mg IV.

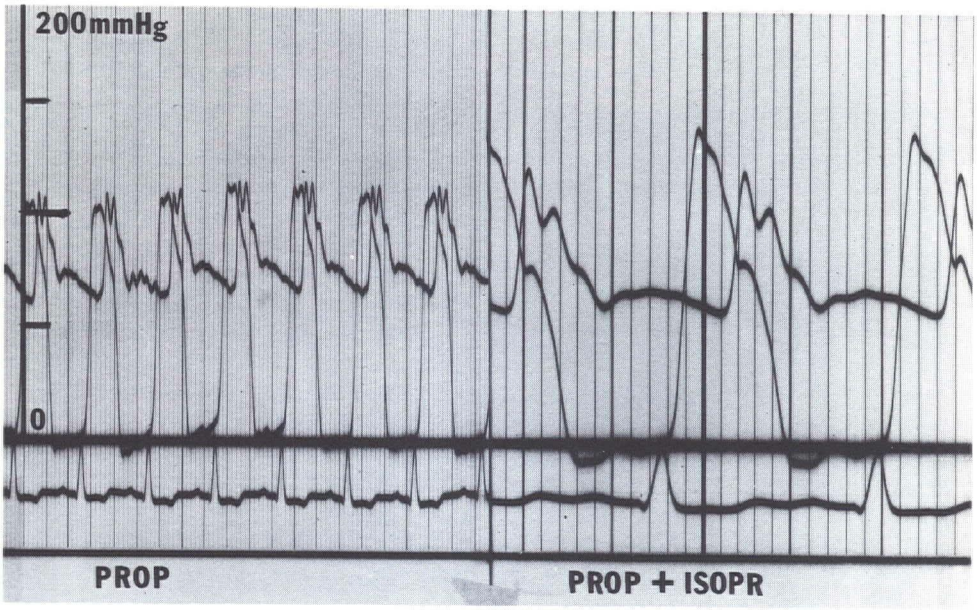

Figure 5
Rechallenge with isoproterenol 0.002 mg after pretreatment with inderal, 1 mg IV. Only a minimal gradient of 14 mm Hg developed.

diographic features, Deutsch et al[4] further classified fixed subaortic stenosis into four types:

Type I, a thin membranous diaphragmatic stenosis.

Type II, a fibrotic ring stenosis.

Type III, a fibromuscular additional tissue stenosis.

Type IV, a tunnel-like stricture of the left ventricular outflow tract.

Subvalvular left ventricular outflow obstruction can also be dynamic in nature related to inotropic state, left ventricular volume, or after-load alterations, causing changes in size of the outflow tract with variable degrees of obstruction such as in hypertrophic subaortic stenosis.

The association of both dynamic and fixed obstruction of the left ventricular outflow tract occurs more infrequently although it has been observed in some patients.[1,2] The dynamic obstruction is attributed to resultant ventricular hypertrophy secondary to a prolonged fixed anatomical obstruction. In some patients this could lead to persistence of a subvalvular gradient even after surgical repair. Recently, Bloom et al[1] have shown that this phenomenon is related to abnormal motion of the anterior leaflet of the mitral valve, secondary to severe concentric hypertrophy of the left ventricle and the septum. Occasionally, idiopathic hypertrophic subaortic stenosis has been found in association with fixed subvalvular obstruction.[5] Although anatomical, as well as hemodynamic echocardiographic, and angiographic criteria are helpful in differentiating these two entities, they may overlap at times[6] and the dynamic obstruction only expresses itself after the fixed obstruction is relieved.

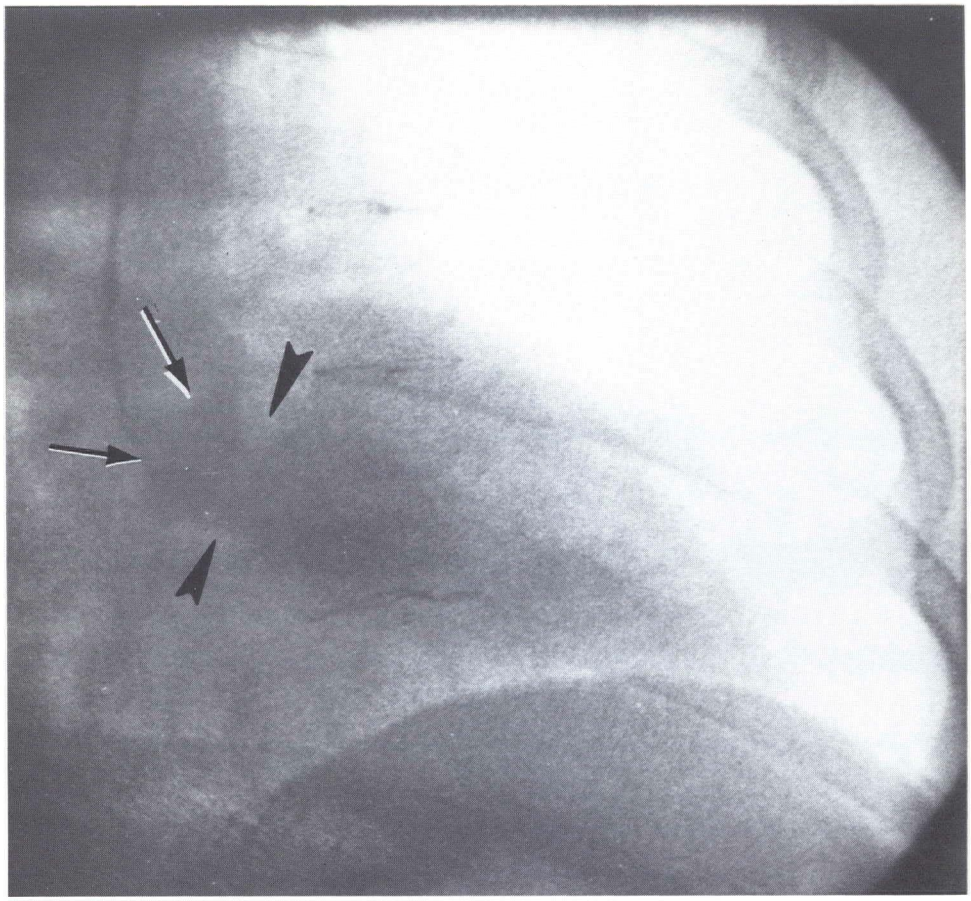

Figure 6
Left ventricular angiogram in RAO projection. Large arrows indicate subvalvular narrowing (lucency). The small arrows outline the aortic cusps.

Our patient represents a case of discrete subvalvular stenosis associated with dynamic outflow tract obstruction present ten years after surgical repair of the fixed obstruction. It is possible that the fibrous ring was not completely excised at the time of surgery or that it has recurred as described previously.[7] The cause of the dynamic obstruction is not entirely clear. Extreme care was taken in avoiding apical catheter entrapment during fluoroscopy and angiography and we do not believe this represents a spurious gradient. It would be expected that any dynamic obstruction secondary to the fibrous band present at the time of operation would have regressed or at least remained unchanged. The improvement in symptoms after surgery suggests that there was at least partial relief of the left ventricle outflow obstruction. Echocardiography demonstrated a structure in the left ventricular outflow tract which could be a remnant of a subvalvular fibrous ring. Such echoes have been described in patients with discrete subaortic stenosis.[8] Since provocative maneuvers were not used during hemodynamic studies prior to surgery ten years ago, the presence of an associated dynamic obstruction at that time cannot be excluded. However, the present studies clearly demonstrate the dynamic nature of the outflow tract gradient.

Although most cases of combined discrete and dynamic subaortic stenosis are found in older children, provocative testing of all cases of discrete subaortic stenosis should be considered. The echocardiogram can be used in selecting the patients for pharmacologic testing. The danger of overlooking a concomitant dynamic outflow tract obstruction during surgery for subvalvular aortic stenosis has been stressed recently.[5,7] Early deaths have been reported in the postoperative period due to low cardiac output resulting from the associated functional obstruction.[9,10] As a precautionary measure to prevent this from occurring, some surgeons advocate myotomies at the time of repair of discrete subaortic stenosis.[7] Beta adrenergic blockade was effective in preventing this demonstrable dynamic component to the obstruction of the left ventricular outflow tract, and relieving symptoms in our patient. Their use should be emphasized in patients with discrete subaortic stenosis associated with dynamic obstruction.

References

1. Bloom KR, Meyer RA, Bove KE, Kaplan S: The association of fixed and dynamic left ventricular outflow obstruction. *Am Heart J* **89**:586, 1975

2. Reis RL, Peterson LM, Mason DT, Simon AL, Morrow AG: Congenital discrete subvalvular aortic stenosis. *Circulation* **43** (Supplement I), I-II, 1971

3. Keith JD, Rowe RD and Vlad P: *Heart Disease in Infancy and Childhood*. Second Edition, MacMillan, New York, 1967, p 250

4. Deutsch V, Shem-Toy A, Yahini JR, Neufeld HN: Subaortic stenosis (discrete form). *Radiology* **101**:275, 1971

5. Chung KJ, Manning JA, Gramiak R: Echocardiography in coexisting hypertrophic subaortic stenosis and fixed left ventricular outflow obstruction. *Circulation* **49**:673, 1974

6. Roberts WC: Valvular, subvalvular, and supravalvular aortic stenosis: Morphologic features. *Cardiovas Clin* **5**:104, 1973

7. Champsaur G, Trusler GA, Mustard WB: Congenital discrete subvalvular aortic stenosis. Surgical experience and long-term follow-up in 20 pediatric patients. *Br Heart J* **35**:443, 1973

8. Popp RL, Silverman JF, Franch JW, Stinson EB, Harrison DC: Echocardiographic findings in discrete subvalvular aortic stenosis. *Circulation* **49**:226, 1974

9. Block PC, Powell WJ Jr, Dinsmore RE, Gladblatt A: Coexistent fixed congenital and idiopathic hypertrophic subaortic stenosis. *Am J Cardiol* **31**:523, 1973

10. Parker DP, Kaplan MA, Connolly JE: Coexistent aortic valvular and functional hypertrophic subaortic stenosis. *Am J Cardiol* **24**:307, 1969