The pulmonary gas exchange improved with mechanical ventilation, but the blood pressure decreased to 60/40 mm Hg, and vasopressor therapy with dopamine hydrochloride was initiated. A Swan-Ganz balloon flotation catheter was inserted, and showed the following values: pulmonary artery systolic pressure, 30 mm Hg; pulmonary artery diastolic pressure, 16 mm Hg; pulmonary capillary wedge pressure, 11 mm Hg; and central venous pressure, 10 mm Hg.

During the subsequent 24 hours, the respiratory status and the blood pressure stabilized. The vasopressor therapy was discontinued, and the ventilatory support reduced. Additional laboratory data demonstrated cardiac enzyme levels within normal limits, and a radionuclide myocardial scan showed no evidence of acute infarction. On the third hospital day, the endotracheal tube was removed, and arterial gas analysis disclosed normal gas exchange. Chest roentgenogram on the fourth hospital day showed clear lung fields (Fig 2). The patient was discharged on the ninth hospital day with a diagnosis of acute pulmonary edema—cause undetermined.

Approximately five weeks after the initial episode of pulmonary edema, the patient presented to the West Virginia University Hospital with acute shortness of breath. Once again, a history of hydrochlorothiazide ingestion earlier that day was obtained. The patient had taken no medication since her previous hospitalization. The physical examination and laboratory values were similar to the first episode of pulmonary edema, but the patient did not require ventilatory support. A Swan-Ganz pulmonary artery catheter yielded the following values: PA systolic pressure, 34 mm Hg; PA diastolic pressure, 18 mm Hg; pulmonary capillary wedge pressure, 14 mm Hg; and central venous pressure, 12 mm Hg. The patient improved rapidly and was discharged on the fourth hospital day with a diagnosis of pulmonary sensitivity to hydrochlorothiazide.

**DISCUSSION**

The patient described herein is, to our knowledge, the seventh reported case of pulmonary edema associated with hydrochlorothiazide therapy, and the first with direct measurement of cardiopulmonary hemodynamic pressures. A Swan-Ganz balloon flotation catheter demonstrated a pulmonary capillary wedge pressure consistent with normal left ventricular function. This finding confirms the noncardiogenic etiology of the pulmonary sensitivity to hydrochlorothiazide.

It has been postulated that the pulmonary reaction is specific to hydrochlorothiazide, and not common to other thiazide diuretics. Our patient gave a history of prior ingestion of chlorothiazide without adverse reaction, lending support to this contention.

The pathogenesis of the pulmonary sensitivity to hydrochlorothiazide is unknown. Although the reaction has many characteristics of an allergic phenomenon, recent attempts to define an immunologic etiology have been unrewarding.

Pulmonary edema associated with hydrochlorothiazide ingestion is a rare and life-threatening adverse reaction which requires prompt recognition and adequate supportive care. Since this medication is frequently prescribed for patients at risk for cardiac failure, we believe this clinically confusing entity should be considered in any patient with acute pulmonary edema and a history of recent hydrochlorothiazide ingestion.

**REFERENCES**

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**Supernumerary Mitral Valve Producing Subaortic Stenosis**

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A ten-year-old girl with severe subaortic stenosis was found to have relatively mature valvular endocardial cushion tissue (fibromyxomatous sheets with a chorda tendinea attached to a left ventricular papillary muscle) immediately beneath the aortic valve. This structure behaved like a valve mechanism, obstructing the left ventricular outflow tract during ventricular systole. This anomaly is an extreme on the spectrum of obstructive endocardial cushion malformations.

Obstruction to left ventricular outflow may be due to one of several malformations produced by abnormal differentiation of endocardial cushion tissue. Subaortic stenosis produced by sheet-like masses of fibrous tissue with chordae tendineae and papillary muscle arising partly from mitral and partly from tricuspid tissue has been described recently.1 We report a similar case in which the obstructive tissue was probably mitral in origin.

**CASE REPORT**

A heart murmur consistent with aortic stenosis was heard in a five-week old girl. She remained asymptomatic, and cardiac catheterization at the age of six years demonstrated subaortic stenosis with a peak systolic pressure difference between left ventricle and aorta of 65 mm Hg. When the patient was ten years old, M-mode echocardiogram demonstrated left ventricular outflow tract obstruction (Fig 1). Electrocardiogram demonstrated normal sinus rhythm and

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**CHEST, 79: 4, APRIL, 1981**
right bundle branch block with left axis deviation. At cardiac catheterization, the left ventricular peak systolic pressure was 295 mm Hg and aortic was 100 mm Hg. Left ventriculography revealed an obstructing subaortic mass that was unusual in appearance (Fig 2).

At operation a month later, the aortic bicuspid leaflets were found to be thickened; mild commissural fusion was incised. The anterior leaflet of the mitral valve appeared thickened. A sheet-like mass opposite the mitral valve lay immediately under the aortic valve (Fig 3A). Its superior attachment was sessile, and its inferior border attached to the septum with a few thick chord-like fibers. One of these fibrous strands extended deeply into the left ventricle. Its termination could not be clearly visualized, but traction of the fibrous chord toward the aortotomy demonstrated its attachment to a structure that appeared to be a papillary muscle, which was transected. Histologic examination of the transected end of the chordal structure demonstrated a portion of myocardium surrounded by thickened endocardium. The remainder of the specimen was fibromyxoid, consistent with valvular tissue. The postoperative course was uneventful.

The patient's physical examination and echocardiogram remained abnormal and consistent with residual subaortic obstruction. Two dimensional cross-sectional echocardiograms showed the ventricular septum to be 2 cm thick and the posterior left ventricular wall to be 1.5 cm thick, satisfying the criteria for asymmetric septal hypertrophy; a discrete subaortic mass could not be clearly demonstrated.

Figure 1. A, Echocardiogram M-mode scan from aorta (upper left) to left ventricle (lower right). Note area of narrowing with multiple echoes in left ventricular outflow tract showing systolic anterior motion and diastolic motion to level of anterior mitral leaflet. B, M-mode echocardiogram of aortic valve demonstrating early systolic closure and late systolic fluttering of valve leaflets. Ao indicates aorta; IVS, interventricular septum; LA, left atrium; MV, mitral valve; and SVM, subvalvular mass.

Figure 2. Preoperative left ventricular angiogram in left anterior oblique projection, frame from ventricular late systole. Mass is seen in left ventricular outflow tract that seems to be attached to both mitral valve and ventricular septum (arrows). Ao indicates aorta; LV, left ventricle.
FIGURE 3A. Anatomy and physiology of supernumerary mitral valve. Upper left, original lesion is depicted in ventricular diastole; view is in long axis, left anterior oblique projection. Aortic valve is closed. Chorda tendinea extends from septal component of supernumerary tissue to a papillary muscle. Upper right, during ventricular systole, bicuspid aortic valve is open, and supernumerary valve tissue obstructs left ventricular outflow tract, acting like an inverted outflow valve mechanism. Circle, operative view of subaortic region clearly demonstrated STVS and HVS. The STMV was not appreciated because it appeared to be merely thickening of the anterior mitral leaflet. Abbreviations: A indicates anterior; Ao, aorta; HVS, hypertrophied ventricular septum; LA, left atrium; LVFW, left ventricular free wall; MVAL, mitral valve anterior leaflet; MVPL, mitral valve posterior leaflet; P, posterior; STMV, supernumerary tissue on mitral valve; and STVS, supernumerary tissue on ventricular septum.

FIGURE 3B (left) Anatomy after the first operation. Septal component of supernumerary tissue has been removed. Unresected tissue on mitral anterior leaflet is still obstructive, but less so than it was preoperatively. FIGURE 3C (right) Anatomy after second operation. Trough has been cut from hypertrophied septal muscle. Removal of all supernumerary tissue has resulted in equalization of systolic pressure in left ventricle and aorta.
A year after operation, recatheterization demonstrated left ventricular systolic pressure of 200 mm Hg and aortic pressure of 90 mm Hg. Selective left ventricular angiography revealed a mass in the subaortic area that appeared attached to the mitral valve and adjacent septum.

At reoperation, the subaortic region was inspected through the aortic valve; no discrete obstructive mass could be seen. There was hypertrophy of the upper septum. The echocardiographic findings of asymmetric septal hypertrophy suggested that her obstruction might be due in part to the subaortic septal muscle. A segment of septum, 5 mm wide and 15 mm deep, was therefore resected for a distance of 6 cm from the aortic annulus toward the apex. This seemed to widen the outflow tract further. An abnormal mass, attached to the mitral valve, could not be visualized. Because of the angiographic suggestion of its presence, however, the left atrium was opened and the anterior leaflet of the mitral valve was found to be thin and pliable. Bidirectional manipulation of the mitral apparatus through the left atrium and the aortic root revealed the apparent thickening of the anterior leaflet on the left ventricular side to be separate from the leaflet (Fig 3B). The structure was sheet-like, attached to the mitral leaflet near its base, and continued for a distance of a few millimeters onto the septum immediately beneath the aortic valve. No chordae tendineae arose from this valve-like sheet of tissue. After excising it through the aortic root, the anterior leaflet of the mitral valve came into full view, and the left ventricular outflow tract was widely patent (Fig 3C). After cessation of bypass, pressure recorded from a catheter withdrawn from the left ventricular apex to the aorta revealed no systolic pressure difference.

Light microscopic appearance of the excised mitral membrane was similar to that of the mass excised at the previous operation. Light and electron microscopy of the excised left ventricular septal muscle was consistent with severe myocardial hypertrophy, but showed neither frank fiber disarray nor other ultrastructural changes associated with asymmetric septal hypertrophy.8

Discussion

The known types of subaortic left ventricular outflow obstruction are discrete subaortic membrane,4 fibrous tunnel,8 hypertrophic muscle,6,7 and atriointerventricular valve anomalies. The malformation presented by this case belongs in the last category.

Accessory tissue of the tricuspid valve has been known to prolapse through a ventricular septal defect, producing left ventricular outflow obstruction.8 Several abnormalities of the anterior leaflet of the mitral valve may produce subaortic obstruction. These include accessory valve tissue attached to the anterior leaflet,8 sometimes with chordae tendineae,10 restriction of anterior leaflet motion by chordae tendineae inserting anomalously into a cleft mural valve or fusion of the leaflet to the ventricular septal wall,9 and displaced annular insertion of the anterior leaflet.11

In a recently reported case1 similar to ours, the patient died after operation because the extent of the obstruction was not recognized and was not completely resected. Detailed anatomic study revealed a ventricular septal defect and two "sacs" in opposing subaortic sites. The septal sac had chordal attachments to the tricuspid valve through the defect, and the opposing sac had chordal attachments to the mitral apparatus. In our case, there was no septal defect and the septal sheet had chordal connection to left ventricular papillary muscle, while the opposing sheet was attached to the mitral anterior leaflet. Thus, the entire obstructing apparatus in our case seems mitral in origin. Because its morphology was consistent with relatively mature valve tissue, it may properly be named "supernumerary mitral valve" to distinguish it from obstructive amorphous masses that have been called accessory mitral valve.7

The existence of this entity is not surprising in view of the known propensities of the primitive endocardial cushions both to produce malformations of their derivatives and to elaborate accessory tissue in the presence of otherwise normally formed derivatives.6,8 Awareness of this unusual malformation by cardiologists and surgeons is important because failure to recognize the full extent of obstruction may result in the patient's death or need for a second operation.

References