CONGENITAL valvular aortic stenosis and congenital localized subvalvular aortic stenosis are commonly encountered, easily recognized at operation, and relieved by standard surgical technics. Idiopathic diffuse subvalvular aortic stenosis is uncommon and can be overlooked at operation. Its relief by indirect methods has been proposed by some because of the problems inherent in its exposure in direct surgical relief.

In two cases an excellent result has been obtained by exposure and direct surgical excision of the lesion through a small left ventriculotomy. The technic appears generally applicable to this lesion.

Identification at Operation

Patients with congenital aortic stenosis should be known to have a systolic pressure gradient between the left ventricle and aorta prior to operation. When such a patient is found at operation to have a normal aortic valve and no localized stenosis immediately beneath the aortic cusps, the diagnosis of diffuse subvalvular aortic stenosis is to be strongly suspected. This type of obstruction usually cannot be visualized through the leaflets (figs. 1 and 2). The diagnosis can be made positively, however, by passing the left index finger into the aortic valve and through the aortic valve into the cavity of the left ventricle, with the thumb remaining outside the heart. The characteristic diffuse thickening situated 1 to 2 cm. upstream from the aortic valve can then be palpated between the thumb and index finger: It is most readily appreciated along the ventricular septum.

Technic of Resection

Extracorporeal circulation is established as usual by utilizing a single large venous cannula in the right atrium to divert caval blood to the extracorporeal apparatus. A catheter is inserted through the left atrial appendage into the left atrium and attached to the intra-cardiac sucker system. The aorta is cross-clamped and the ascending aorta is opened between clamp and aortic valve. After identification of the lesion, an incision about 2.5 cm. long is made in the free wall of the left ventricle. The optimal point for this incision is selected with the left index finger in the left ventricle to palpate the most accessible and thinnest portion of the anterior ventricular wall. Due regard is taken of the distribution of the coronary arteries so that the ventriculotomy incision does not compromise any large branches.

Small catspaw retractors are placed in the ventriculotomy incision. The chordae, papillary muscles, and anterior leaflet of the mitral valve can be visualized, although with some difficulty, through this small incision in a thick ventricle (fig. 3). The aortic leaflets cannot be visualized, but the left index finger can be kept in the aortotomy incision against the aortic leaflets to identify their location during the early stages of the resection.

The hypertrophied musculature is excised. The use of a Brock punch was helpful in one of our cases, but in most cases excision is done best by sharp dissection with a knife, or scissors, or both. Because of the dangers involved in resecting near the mitral valve structures, most of the resected material is taken from the region of the ventricular septum and the anterior portion of the free wall of the left ven-

From the Mayo Clinic and the Mayo Foundation, Rochester, Minnesota.
Anatomy of diffuse subvalvular aortic stenosis. The obstructing hypertrophied portion of muscle is about 1 cm. upstream from the base of the valve and is considerably more prominent during ventricular systole. For these reasons, at operation, with the heart quiet and the base of the aorta opened, no obstruction is visible in or beneath the aortic valve.

The outflow tract of the ventricle can be greatly enlarged by resection of this muscle mass, just as in cases of subvalvular pulmonary stenosis secondary to valvular pulmonary stenosis.

Direct perfusion of the coronary artery was not employed in either of the cases we are reporting, since resection could be accomplished within 20 to 25 minutes. The root of the aorta is closed and the aortic clamp is released, thus allowing coronary perfusion to begin. After the heart has begun to beat and with the ventriculotomy still open, one can palpate the outflow tract of the left ventricle in order to determine whether sufficient tissue has been removed to relieve the obstruction. If it has, the left ventriculotomy is closed with interrupted silk sutures, and air is evacuated from the ventricle. Perfusion is gradually discontinued in the usual fashion.

Report of Cases

Case 1

A 17-year-old white boy had noted fatigue, dyspnea, and substernal tightness from moderately severe exertion for about 2 years prior to his admission to the Mayo Clinic. His family history was noncontributory. Physical examination revealed a coarse systolic murmur in the aortic area. A roentgenogram of the thorax revealed evidence of a possible enlargement of the left atrium. The electrocardiogram gave evidence of left ventricular hypertrophy. The blood pressure was 90 mm. of mercury systolic and 60 mm. diastolic. By percutaneous punctures, systolic pressure was found to be 175 mm. of mercury in the cavity of the left ventricle while that in the aorta was 100 mm.

Operation was performed on April 9, 1959. At operation pressure was found to be 159 mm. of mercury systolic in the left ventricle while it was 89 mm. systolic in the aorta. The aortic leaflets were normal. Diffuse muscular hypertrophy of the outflow tract of the left ventricle was noted about 1 cm. upstream from the aortic leaflets without any evidence of a localized fibrous or muscular ring. A 2-inch incision was made in the free wall of the left ventricle. Through this incision the obstructing muscular mass was excised. In the operating room after repair, pressure in the cavity of the left ventricle was 82 mm. systolic while that in the root of the aorta was 78 mm. systolic. Postoperative convalescence was uneventful.

The patient returned to the clinic for reexamination on March 11, 1960. He had been entirely well since his operation and had resumed full activity including strenuous farm work without symptoms. No longer did he experience angina,
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dyspnea, or undue fatigue. On physical examination no murmur was heard in the aortic area but a faint systolic murmur, grade I, was heard in the fourth left interspace and at the apex. He had gained 30 pounds. A roentgenogram of the thorax showed a slight decrease in the size of the cardiac silhouette. No change was apparent on the electrocardiogram. The patient’s postoperative result was considered excellent.

Case 2

A 20-year-old white woman presented with a history of exertional dyspnea and fatigue with occasional episodes of paroxysmal nocturnal dyspnea since the age of 11. Symptoms had been more marked in the year prior to admission. On physical examination the blood pressure was 124 mm. of mercury systolic and 76 mm. diastolic. A grade-I systolic murmur, which became grade III with exercise, was heard in the aortic region. The murmur was heard only faintly in the neck. A roentgenogram of the thorax revealed evidence of cardiac enlargement and the electrocardiogram revealed advanced left ventricular hypertrophy. Pressures were recorded at 216 mm. systolic and 8 mm. diastolic in the cavity of the left ventricle and 81 mm. systolic and 51 mm. diastolic in the radial artery by the percutaneous puncture technic.

Operation was accomplished on September 13, 1960. The appearance of the left ventricle was striking. Massive hypertrophy prevailed, the ventricle being so large and firm that it seemed almost to be a solid nonbeating tumor. Leaflets of the aortic valve were normal. No localized obstruction was found immediately beneath the valve, but about 1 cm. upstream from the base of the valve was a greatly hypertrophied mass of muscle, which was particularly large along the septal side of the aortic outflow tract. Most of this mass, and particularly that along the septal side, was excised through a 3-em. incision in the left ventricle. After closure of the aortotomy the aortic clamp was removed and the heart became hard in ventricular fibrillation. Palpation of the region of resection through the left ventriculotomy now revealed no obstruction. The ventriculotomy was closed. After the heart had again taken over the circulation, pressure in the cavity of the left ventricle was 108 mm. systolic and 12 mm. diastolic, while that in the left arm by the cuff method was 105 mm. systolic.

Convalescence was uneventful. On the eleventh postoperative day a needle was passed percutaneously into the cavity of the left ventricle where the pressure was recorded at 100 mm. systolic. Unfortunately a needle could not be passed into the brachial artery, but the pressure in the right arm was 115 mm. systolic by the cuff method.

Figure 3

Surgical exposure through left ventriculotomy incision. The chordae and papillary muscle of the mitral valve must be visualized and protected during resection of the hypertrophied muscle forming the obstruction. In actual practice the surgeon’s index finger is passed through the aortotomy (not shown) to protect by palpation the aortic valve leaflets during resection.

Discussion

Brock, in 1957, first referred to the diffuse, acquired subvalvular aortic obstruction observed in patients with systemic artery hypertension. Brachfeld and Gorlin found that such diffuse muscular subvalvular aortic stenosis could be idiopathic, rather than secondary, in some patients. Morrow and Braunwald reported two such cases, and stated that in neither was the lesion amenable to surgical correction. They properly directed attention to the fact that this entity should be suspected when operating for congenital aortic stenosis in a patient who, at operation, is shown to have neither valvular stenosis nor a localized subvalvular obstruction.
Although the technic described herein has been used in only two cases as yet, it would seem applicable generally to patients with this uncommon type of subaortic obstruction. Data available in these two cases seem sufficient to indicate that effective relief of the subaortic obstruction has been obtained.

Concern must be had for the placement of an incision in the wall of the left ventricle. In neither of these patients, however, was there evidence of undue reduction of cardiac output after operation, nor have there been any late ill effects that could be attributed to the left ventriculotomy.

Summary
Direct surgical relief of diffuse subvalvular aortic stenosis has been accomplished in two cases by open operation through a left ventriculotomy.

References

When Harvey's discovery, like an earthquake, had broken up galenism and other outworn sophistries, his masterly work stood forth against a more lurid background of folk superstitions—of vampires, witch-burning, magic, cabbalism, astrology, alchemy, chiromancy, and water-casting. In physics, terrestrial and celestial, Galileo, persecuted as he was, had some current with him and before him; Copernicus had preceded him, Kepler was beside him: but in physiology the waters had closed upon the path of Galen as upon the wake of a great ship; the anatomists, themselves galenists, had given Harvey little help; and the share of Servetus, Colombo, and Fabricius was but small in the discovery of the central fact of the science, and of the method which opened the way to Pecquet and Aselli, to Glisson, to Steno, to Wharton and Willis, to Haller and Bernard. Harvey's discovery was the first step to a transfiguration of medicine; and though after Harvey there arose much false physiology and therewith again great floods of medical sophistry, yet from his time medicine has had to reckon with physiology, the only source of scientific nosology and therapeutics.—Thomas Clifford, Allbutt, M.A., M.D.

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