Surgical Treatment of Idiopathic Hypertrophic Subaortic Stenosis: *

Technic and Hemodynamic Results of Subaortic Ventriculomyotomy

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IN THE MAJORITY of patients with obstruction to left ventricular outflow the responsible anatomic lesion is found to be stenosis of the aortic valve, a discrete fibrous subvalvular membrane or, more rarely, a constriction in the ascending aorta immediately above the valve. Recent reports from this and other clinics, however, have called attention to another and hitherto unrecognized form of obstruction termed idiopathic hypertrophic subaortic stenosis.[1-6, 9-16] In this malformation there is massive hypertrophy of the left ventricular myocardium, particularly in the area of the interventricular septum. During ventricular systole the outflow tract of the ventricle narrows to such an extent that resistance to left ventricular ejection occurs and a systolic pressure gradient develops within the ventricle. At the National Heart Institute the diagnosis of hypertrophic subaortic stenosis has been established in 15 patients and a detailed description of the clinical, hemodynamic and angiographic findings in them has been presented elsewhere.[3] Although the lesion has been encountered unexpectedly in several patients undergoing operations for what was considered to be one of the usual forms of aortic stenosis, it has been generally considered that resection of the hypertrophic muscle is not feasible and that this form of subaortic stenosis is, therefore, not amenable to surgical treatment.

A new operative method, designed to abolish the continuity of the sphincter-like contraction ring in the outflow tract, has been applied in two progressively symptomatic patients with severe hypertrophic subaortic stenosis. Brief descriptions of the clinical and operative findings in these two patients, the surgical technic utilized and the results of pre- and postoperative hemodynamic study will be presented.

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Case Reports

Case 1. C. F., a 10-year-old schoolboy with no family history of heart disease, had been found to have a heart murmur in early infancy. Although his growth and development were normal he developed fatigability which became progressive and at the age of eight years he was studied at another medical center where the clinical diagnosis of congenital subaortic stenosis was made. Because of the presence of cardiac enlargement, fatigability and dyspnea, the patient was admitted to the National Heart Institute.

The pertinent findings on physical examination were limited to the cardiovascular system. The peripheral pulses were brisk and regular and the blood pressure 85/55. A prominent left ventricular lift was palpable, and the heart was enlarged, with the apical impulse in the sixth intercostal space in the anterior axillary line. A systolic thrill was felt over the entire precordium. The second heart sound was paradoxically split at the base and there was a grade 4/6 ejection systolic murmur heard well over the entire precordium but not transmitted to the neck. The electrocardiogram (Fig. 1) showed first degree A-V block, left ventricular hypertrophy, and T-wave and ST-segment abnormalities. The chet x-ray (Fig. 2) revealed marked generalized cardiac enlargement and particular prominence of the left ventricle. There was no dilatation of the ascending aorta.

These clinical findings suggested the diagnosis of idiopathic hypertrophic subaortic stenosis and accordingly a retrograde left ventricular catheter-

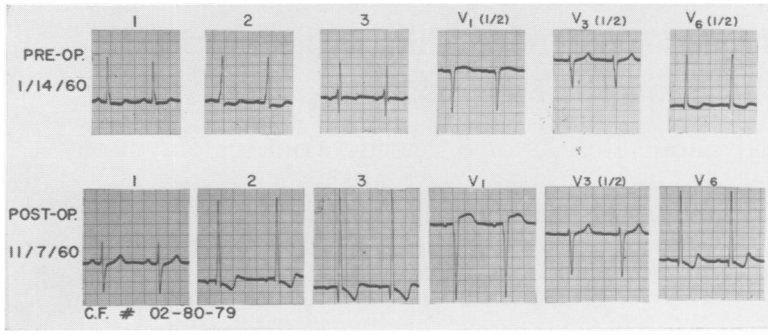

FIG. 1. Representative portions of the pre- and postoperative electrocardiograms in patient C. F.

ization was carried out. The aortic pressure pulse contour showed a brisk rise and there was no pressure gradient across the aortic valve. The pressure within the main left ventricular cavity, however, was 260/20 mm. Hg and there was a systolic pressure gradient of 185 mm. Hg within the left ventricular outflow tract (Fig. 3). A selective angiocardiogram with left ventricular injection demonstrated severe narrowing of the outflow tract during systole. The stenotic segment was not fixed, however, and was seen to open during diastole.

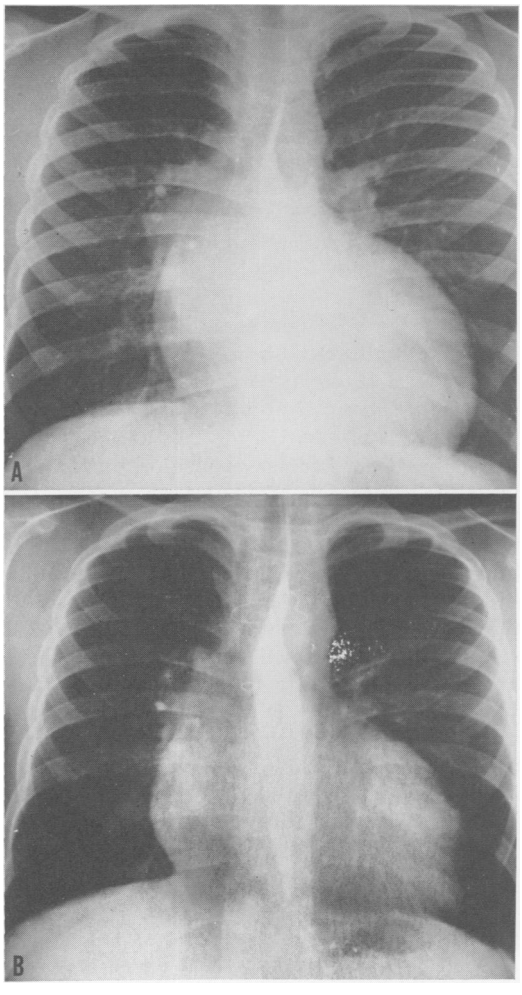

FIG. 2. Preoperative (A) and postoperative (B) chest x-rays of patient C. F.

On January 26, 1960, the patient was operated upon. Following the institution of cardiopulmonary bypass an aortotomy was made and the aortic valve was seen to be normal. When the leaflets were retracted a hypertrophic mass of muscle could be seen bulging into the outflow tract, primarily from the anterior or septal aspect of the ventricle, almost completely obliterating the lumen during systole. Following an unsuccessful attempt to resect some of the muscle mass a subaortic ventriculomyotomy was performed.

Postoperatively the child's convalescence was prolonged but his recovery was satisfactory. Five weeks after the operation the left ventricular and systemic arterial pressures were measured and the peak systolic gradient was found to have been reduced from 185 to 65 mm. Hg.

Eight months after operation the patient was readmitted for repeat study. His parents stated that he was improved and his activities were normal except that he still became somewhat dyspneic with strenuous exercise. The physical findings were unchanged, as was the size of the heart by x-ray (Fig. 2). The electrocardiogram (Fig. 1) showed right axis deviation, first degree heart block, a QRS prolongation to 0.12 secs. and left ventricular hypertrophy. A left heart catheterization was carried out by the transseptal route. The left ventricular pressure was 160/20 mm. Hg and the peak systolic gradient between the left ventricle and femoral artery varied between 20 and 30 mm. Hg (Fig. 3). The cardiac output was 4.63 L./min. and the calculated orifice area was 1.03 cm.^{2} Another left ventricular selective angiocardiogram was made and the previous abnormal configuration of the left ventricular outflow tract was still evident

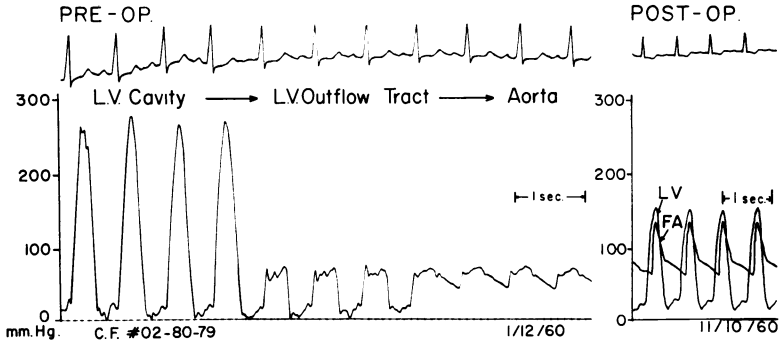

FIG. 3. Pre- and postoperative pressure tracings obtained by left heart catheterization in patient C. F. The preoperative withdrawal tracing (left) localizes the obstruction to a site within the left ventricular outflow tract. There is no systolic pressure gradient across the aortic valve. The simultaneous pressure tracings made postoperatively (right) show the systolic gradient between the left ventricle (LV) and femoral artery (FA) to be only 20 mm. Hg.

but the extent of systolic narrowing appeared less than in the preoperative study (Fig. 4, 5).

Case 2. I. R., a 33-year-old clerk with no family history of heart disease, was apparently well until the age of 27 years when he first developed epigastric and precordial pain with exercise. Shortly after this he began to have episodes of exertional dizziness and experienced several syncopal episodes. In 1955, he was studied at another medical center where the diagnosis of aortic stenosis was established by left heart catheterization. He was operated upon under general hypothermia and at this time direct pressure measurements showed an intraventricular pressure gradient of 40 mm. Hg and localized the obstruction to an area 3.0 cm. below the aortic valve. The surgeon believed the lesion was some unusual form of subaortic stenosis and that the relatively small gradient did not justify the risk of an attempt at surgical correction. Following the procedure the patient's

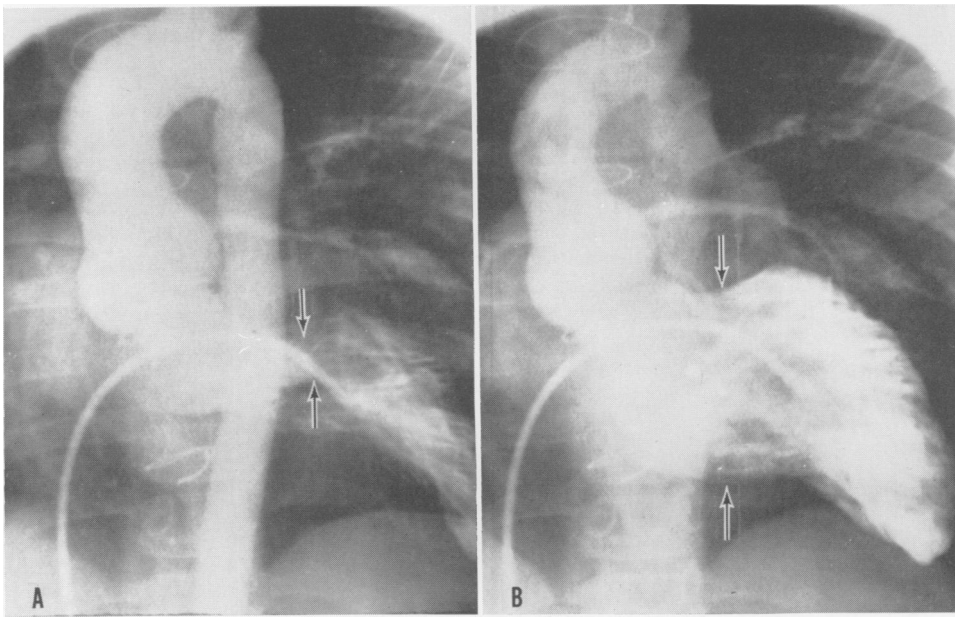

FIG. 4. AP projection of the left ventricular selective angiocardiogram during systole (A) and diastole (B) in patient C. F. The catheter has been introduced into the left ventricle by transseptal puncture of the interatrial septum.[8] The outflow tract (arrows) narrows markedly during systole but opens widely during diastole.

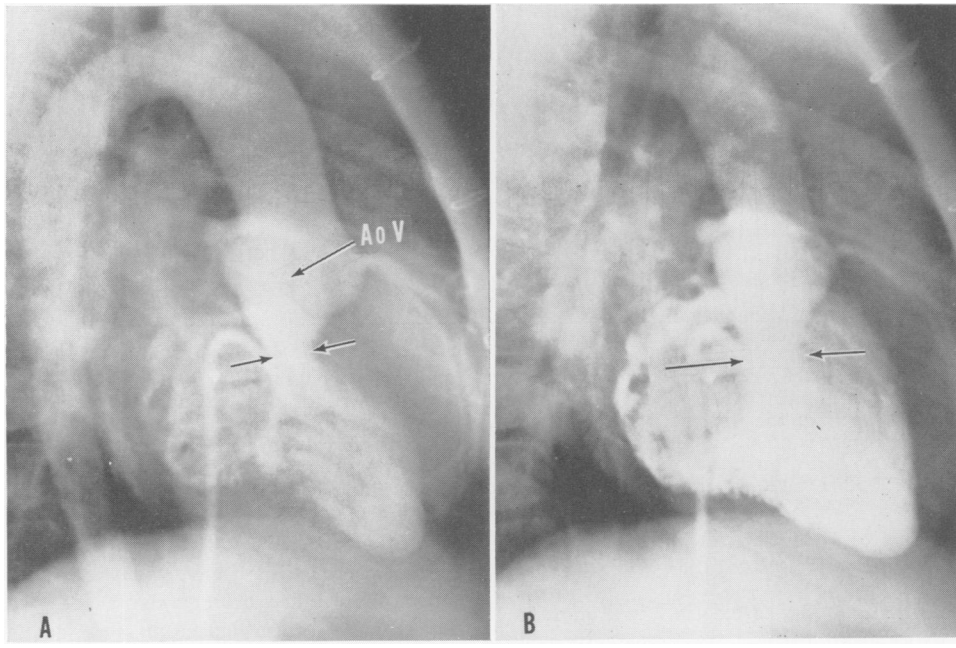

FIG. 5. Lateral projection of the left ventricular angiocardiogram in patient C. F. During systole (A) the outflow tract is seen to narrow resulting in the cone-shaped configuration characteristic of hypertrophic subaortic stenosis (arrows). The prominence of the mass anteriorly is well demonstrated. The normal aortic valve is indicated (AoV). During diastole (B) relaxation of the outflow tract is apparent.

symptoms gradually progressed and he developed signs of left ventricular failure.

In 1957, he was operated upon at a second institution. On this occasion it was noted by palpation that moderate mitral regurgitation was present; no pressure gradient could be demonstrated between the left ventricle and aorta. The surgeon concluded that obstruction was probably due to muscular hypertrophy and did not attempt any corrective procedure.

The patient was admitted to the National Heart Institute in January, 1960. He was orthopneic and in evident left ventricular failure. The blood pressure was 110/60 and the peripheral pulses were brisk. There was a prominent left ventricular lift in the sixth intercostal space at the anterior axillary line. A systolic thrill was present only at the apex. There was paradoxical splitting of the second heart sound, an atrial sound was audible, and a grade 4/6 harsh systolic murmur was audible at the apex. The electrocardiogram showed left ventricular hypertrophy with T-wave inversion and ST-segment depression in the standard leads and over the left precordium (Fig. 6). X-ray examinations revealed enlargement of the left ventricle but no evidence of calcification in the region of the aortic valve or of poststenotic dilatation of the ascending aorta (Fig. 7). At right heart catheterization the pulmonary artery pressure was 27/8 mm. Hg. There was no pressure gradient within the right ventricular outflow tract and no evidence of a circulatory shunt. At transseptal left heart

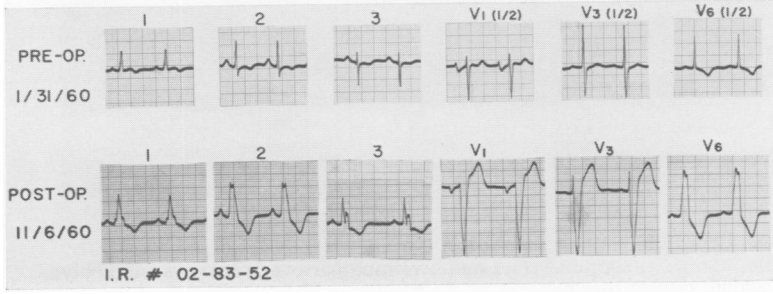

FIG. 6. Pre- and postoperative electrocardiograms of patient I. R.

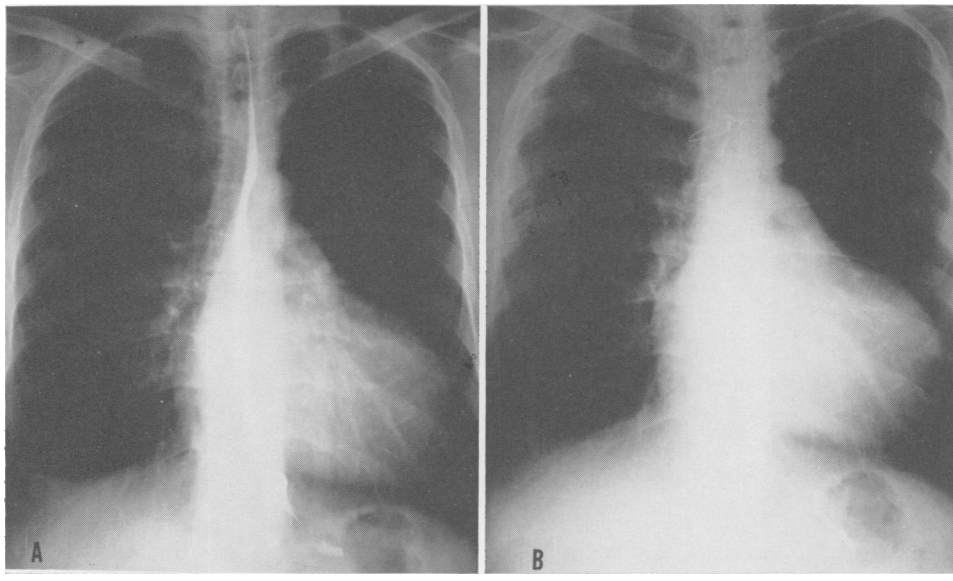

FIG. 7. Preoperative (A) and postoperative (B) chest x-rays of patient I. R.

catheterization the catheter passed from the left ventricle, where the pressure was 190/15 mm. Hg, into the aorta. The withdrawal tracings showed no gradient across the aortic valve but as the catheter tip traversed the outflow tract a systolic pressure gradient of 70 mm. Hg was demonstrated within the left ventricle (Fig. 8). The cardiac output was 3.94 L./min. and the calculated effective orifice size was 0.50 cm.^{2} An angiocardiogram was not performed since the diagnosis of hypertrophic subaortic stenosis seemed clear and it was thought that the patient's precarious cardiac status precluded any unnecessary study.

Operation was carried out on February 16, 1960. The aorta was of normal size and no thrill was felt within it. When the aortic valve was exposed it was found to be normal. Inspection of the outflow tract of the ventricle, however, revealed the myocardium to be diffusely thickened and there was particularly prominent hypertrophy in the region of the interventricular septum. A subaortic ventriculomyotomy was performed. Postoperatively the patient's course was uneventful. All postoperative electrocardiograms showed left bundle branch block. The left ventricular pressure three weeks after the operation was 140/15 mm.

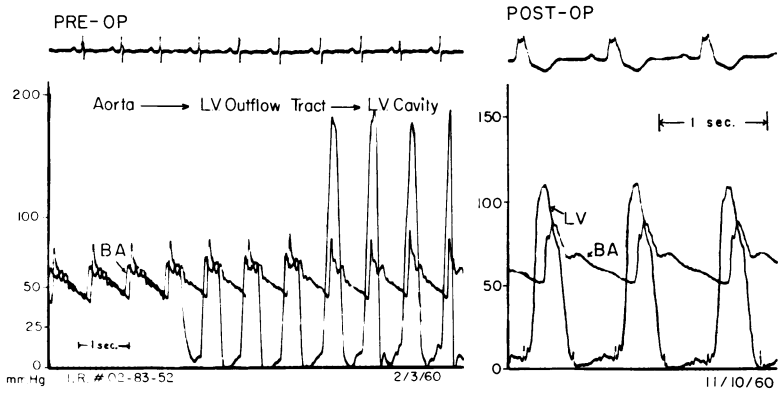

FIG. 8. Pre-and postoperative pressure tracings obtained by left heart catheterization in patient I. R. The site of obstruction is again localized by the withdrawal tracing to the outflow tract of the left ventricle. Postoperatively the gradient between the left ventricle (LV) and brachial artery has been reduced to 20 mm. Hg.

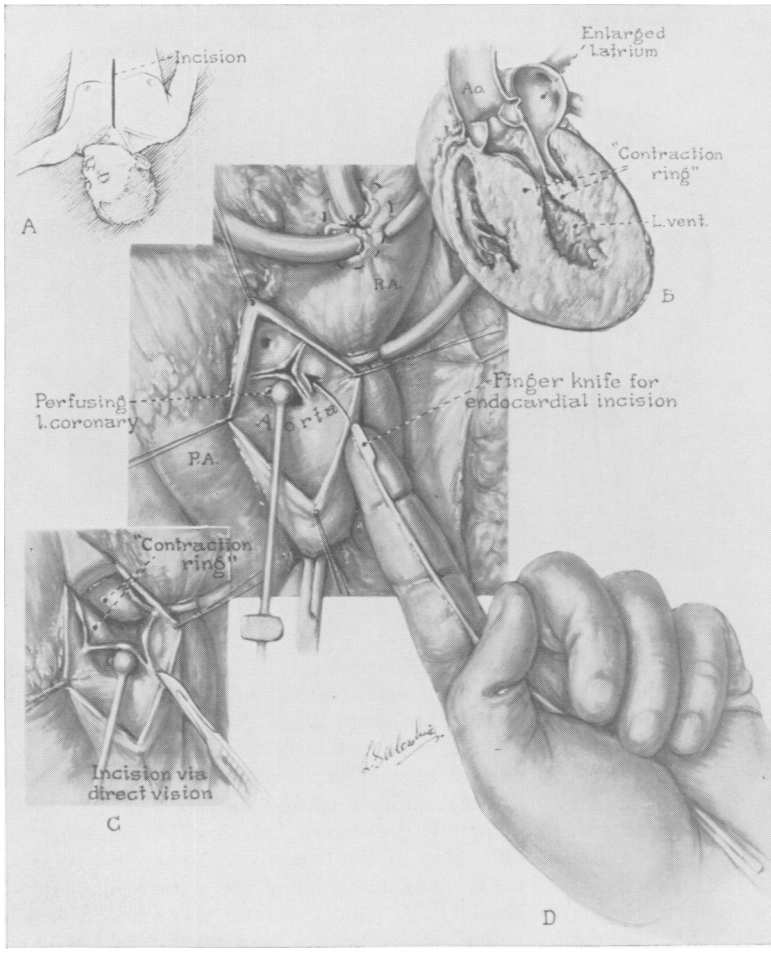

FIG. 9. Operative method for subaortic ventriculomyotomy. Myocardial contractility is maintained by perfusion of the left coronary artery. The contraction ring (B) is initially incised under direct vision (C) or with a finger knife (D).

Hg, the peak systolic pressure gradient between the left ventricle and brachial artery was 30 mm. Hg and the effective orifice size was 1.61 cm.^{2}

The patient returned for detailed postoperative study eight months after operation. He stated he had been markedly improved. He had experienced no precordial pain, dizziness or syncope and had returned to full-time work as a clerk. His only symptom was moderate fatigability. On examination the physical findings were unchanged and the heart remained enlarged by x-ray (Fig. 5). Left bundle branch block was still evident electrocardiographically (Fig. 6).

Left heart catheterization by the transseptal route showed the left ventricular pressure to be 110/10 mm. Hg and the peak systolic gradient between the left ventricle and brachial artery was only 20 mm. Hg (Fig. 8). The calculated effective orifice size was 1.48 cm.^{2} A selective angiocardiogram with left ventricular injection showed typical systolic narrowing of the outflow tract and persistent evidence of left ventricular hypertrophy.

Surgical Technic

Subaortic ventriculomyotomy was performed in each patient in a similar manner. A complete median strenotomy was employed and the pericardium widely incised. The aorta was freed from the pulmonary artery and stripped of its adventitia from the origin of the right coronary artery to the level of the innominate artery. Following cannulation of the venae cavae and the femoral artery, complete cardiopulmonary bypass was instituted. A vertical aortotomy was made and the aortic valve exposed (Fig. 9). The left coronary artery was perfused throughout the period of aortic occlusion with oxygenated blood supplied through a cannula connected to the arterial line from the heart-lung machine. The valve leaflets were retracted to permit visualiza-

tion of the obstructing muscle mass in the outflow tract. A vertical incision was then made through the endocardium and into the superficial muscle layer from the apex of the heart to the aortic annulus. The incision was placed over the most prominent part of the protruding muscle and in each patient was made anteriorly into the interventricular septum.

In one patient (C. F.) a conventional knife was employed while in the other (I. R.) the incision was more conveniently made with a malleable finger-tip mitral valvulotome (Fig. 9). With the finger alone the muscle fibers were then split to a depth of approximately 2.0 cm. along the length of the initial incision. In each instance the resulting cleft in the myocardial "contraction ring" opened widely and by palpation the degree of systolic narrowing was considered to be decreased.

Discussion

Three patients with hypertrophic subaortic stenosis were operated upon at the National Heart Institute before the true nature of the malformation was appreciated. Preoperatively each was considered to have the usual or discrete form of subaortic stenosis. In the first two patients the heart was arrested with potassium citrate and under these conditions the mechanism of obstruction was not apparent. In the third, myocardial contraction was maintained by coronary perfusion and the sphincter-like contraction ring in the outflow tract was easily palpable. Following this observation the hypertrophic form of subaortic stenosis was clearly recognized as a distinct disease entity which could be distinguished preoperatively on the basis of characteristic clinical, hemodynamic and angiographic features.

Among the 15 patients whom we have had the opportunity to study,[3] 12 were males and although the majority were young adults, four were less than six years of age. Five of these patients were members of two familites and in them the disease certainly seemed to be familial.[3, 4, 10, 14] The symp-

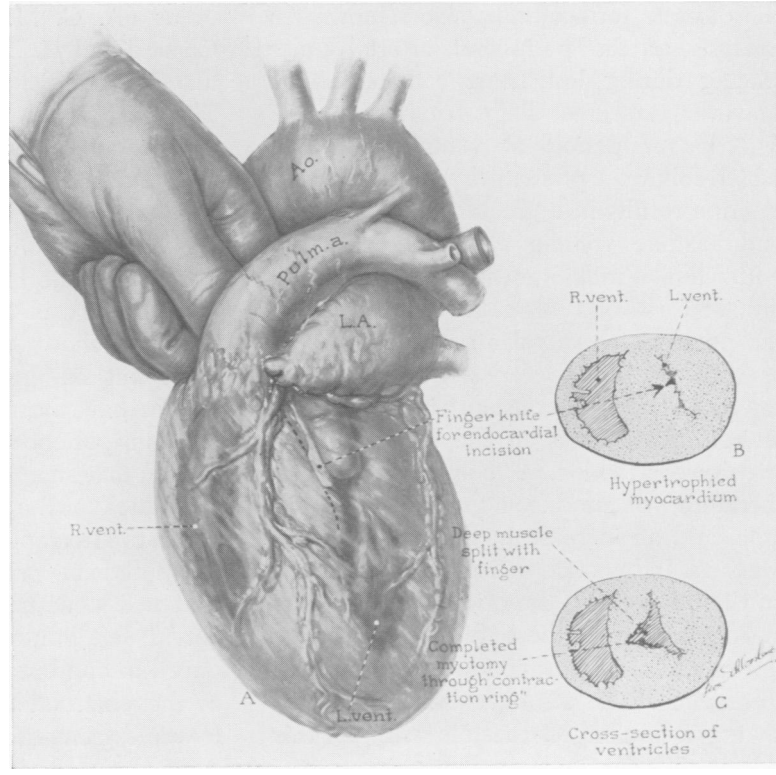

FIG. 10. After the initial vertical incision into the obstructing muscle mass has been made (A, B) the muscle fibers are deeply split with the finger to achieve the cleft shown in C.

toms of hypertrophic subaortic stenosis, when present, are not specific and usually consist of dyspnea and fatigability although angina, syncope or left ventricular failure may occur. Most of our patients were initially thought to have either ventricular septal defect or mitral regurgitation. On examination the physical findings associated with the usual form of aortic stenosis are not present. The peripheral pulse is usually quite brisk. A thrill is usually felt at the apex or along the left sternal border and a left ventricular lift is always present. The systolic murmur is most prominent at the apex and left sternal border and does not radiate to the carotid vessels. By x-ray (Fig. 2, 5) there is both left atrial and left ventricular enlargement but valvular calcification and dilatation of the ascending aorta are never seen. The electrocardiogram usually reveals only left ventricular hypertrophy and is not diagnostic.

Left heart catheterization will localize the site of obstruction to an area within the left ventricle, but this finding alone does not, of course, distinguish the lesion from the discrete form of subaortic stenosis. An analysis of the peripheral arterial pulse tracing during left heart catheterization, however, is of great diagnostic importance.[7] The longer period of ventricular filling which follows a premature ventricular contraction results in a greater left ventricular end-diastolic volume and, in accordance with the Frank-Starling mechanism, a greater force of ventricular contraction. In the presence of valvular aortic stenosis or of discrete subvalvular stenosis the narrowed orifice is of constant size and is not altered by changes in the force of ventricular contraction. A more forceful beat, therefore, will produce a greater stroke volume, and an increase in the arterial pulse pressure. In hypertrophic subaortic stenosis, on the other hand, the orifice narrows during contraction of the outflow tract and its size is, therefore, a function of the force of contraction. The stronger contraction following a prolonged diastole will, in this condition, narrow the outflow tract yet further, reduce the stroke volume and, therefore, the arterial pulse pressure. This paradoxical narrowing of the arterial pulse pressure is thus a reflection of the "contractile stenosis" which is the hallmark of the lesion. Such a mechanism may also be shown by selective left ventricular angiocardiography and this study is certainly indicated if the discrete and hypertrophic forms of subaortic stenosis are to be differentiated with confidence preoperatively. The angiographic appearance of hypertrophic subaortic stenosis is typified by the films reproduced in Figure 4.

The operation described for the relief of outflow obstruction due to hypertrophic subaortic stenosis may be considered analogous to those procedures employed in the treatment of other obstructive lesions characterized by muscular spasm or hypertrophy. The Fredet-Ramstedt operation for pyloric stenosis and the Heller operation for cardiospasm immediately come to mind. The concept of subaortic ventriculomyotomy was suggested during a visit by one of us (A. G. M.) to Messrs. William P. Cleland and H. H. Bentall at the Postgraduate Medical School and Hospital, London. These surgeons encountered hypertrophic subaortic stenosis unexpectedly at operation and attempted to resect a portion of the muscle mass in the outflow tract. Only a small amount of muscle could be removed but left bundle branch block was noted in postoperative electrocardiograms. This patient has shown striking clinical improvement but no postoperative hemodynamic studies have been reported.[9] On the basis of this experience Cleland and Bentall postulated that an anterior vertical incision into the muscle of the outflow tract, of sufficient depth to interrupt the left bundle, might be attempted in patients with hypertrophic subaortic stenosis. In the patients described herein, left bundle branch block was produced in one but not the other while relief of the obstruction was achieved in each. It would seem, therefore, that the operation

relieves obstruction by direct mechanical interference with the sphincter-like contraction ring and that disturbance of the conduction mechanism is, *per se*, unnecessary.

It must be emphasized that the operation described must be considered a palliative one since the underlying pathologic process, idiopathic myocardial hypertrophy, continues to be evident in each patient. In both, however, the left ventricular pressure was reduced immediately by the operation and has remained low for nine months. On the basis of this experience it would seem that subaortic ventriculomyotomy should receive further clinical trial in symptomatic patients with hypertrophic subaortic stenosis in whom a large intraventricular pressure gradient is demonstrated.

Summary

Hypertrophic subaortic stenosis, a form of obstruction to left ventricular outflow caused by massive, idiopathic myocardial hypertrophy, is being recognized in increasing numbers of patients and the lesion has been encountered unexpectedly at operation by many surgeons. Since attempts at resection of the muscle mass have generally been unsuccessful, this form of subaortic stenosis has not been considered amenable to surgical treatment.

A new operative method, subaortic ventriculomyotomy, has been utilized in two symptomatic patients with hypertrophic subaortic stenosis. The distinguishing features of the disease, the surgical technic utilized, and the results of pre- and postoperative hemodynamic studies in these patients are described.

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