Stenosis of Left Ventricular Outflow Tract
Causes and Contrast Visualization

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Among the several pathologic entities capable of presenting the clinical syndrome of aortic stenosis are congenital and acquired aortic valvular stenosis, supravalvular and subaortic stenosis, and muscular hypertrophy of the left ventricular outflow tract. Differentiation of the responsible lesion has become a clinical necessity since surgical measures are often indicated. Knowledge of the site, character, and hemodynamic consequences of the obstructing lesion makes possible the competent selection of surgical candidates and permits advanced planning of an operative attack appropriate to individual circumstances. In the following survey of aortic stenosis and related obstructing lesions, particular attention is given to the critical diagnostic roles of selective left heart visualization and manometric studies.

Radiologic contrast visualization provides a means for inspecting the functioning left ventricle, aortic valves, and thoracic aorta. In certain respects it is superior to direct examination. Chief among these is the advantage of studying undisturbed structures under relatively normal functional circumstances.

During the past quarter century of experience with contrast cardiovascular visualization many technics have been proposed for left heart visualization. If the ineffective and unduly hazardous ones are disregarded, several different approaches remain. Regardless of the method employed, success in contrast visualization requires that an adequate concentration of contrast agent be so placed (in space and time) as to allow the desired radiographic study. Direct injection into the left ventricle is essential to the study of the left ventricular outflow tract. Conventional intravenous angiocardiography and selective catheter injections into the right heart or pulmonary arteries are not satisfactory for this purpose because of dilution and other factors. Occasional brilliant exceptions are unfortunately out of context. The amount and time of injection, route of administration, and the time, duration, and frequency of x-ray exposures are best determined on the basis of the patient's anatomy and physiology rather than by arbitrary standards.

Percutaneous retrograde catheterization of peripheral arteries permits the insertion of large-bore polyethylene tubing into the left ventricle and thereby makes possible both contrast visualization and manometric studies. Based upon the pioneer contributions of Seldinger and Prioton and associates, a technic employing a flexible coilspring guide has been used successfully in over 300 patients at the University of Oregon Medical School. The safety of the technic has allowed its use in outpatients, and its simplicity is evidenced by the usual achievement of left ventricular catheterization within 5 or 10 minutes after starting the arterial puncture. Unquestionably, aortic valvular stenosis increases both the need and the difficulty of transvalvular catheterization; fortunately it rarely prevents the accomplishment.

In brief, the technic begins with needle puncture of a femoral or other peripheral artery under local anesthesia. A 125-cm. long, flexible coilspring guide, resembling a guitar string and containing a piano-wire stiffner, is inserted into the arterial lumen through the

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Blood pressure findings in aortic stenosis. Redrawn "ideal" records. Upper line on each trace represents normal systolic level. Normally, left ventricular and aortic pressures are the same. In aortic valvular stenosis there are left ventricular hypertension and a significantly lower systolic pressure in the aorta. The drop in pressure is a guide to the severity of the lesion, whereas the location of the gradient indicates the anatomic site. Gradients are best considered in relation to the maximum systolic pressure rather than as absolute figures, since they are greatly influenced by stroke volume and heart rate.

Figure 1

needle. The latter is then withdrawn and removed by sliding it along and off the guide, which is kept in place. Suitable length of polyethylene tubing, usually PE 240, is next slipped sleeve-fashion over the guide and advanced until, with a rotary motion, its tip enters the arterial lumen. The tubing is shorter than the spring guide by a known amount, so that the relative positions of tubing and guide may be adjusted after their insertion.

Fluoroscopy is used during placement of the catheter, manipulation of which is generally best accomplished with 2 or 3 inches of guide protruding from the exploring end of the tubing. The major branches of the abdominal aorta, and brachiocephalic and coronary arteries can usually be entered as desired. Retrograde transvalvular passage is favored by the formation of a partial loop in the flexible protruding end of the guide following its impingement on an aortic valve cusp. Entry into the ventricle is effected when an appropriate combination of valve action and catheter manipulation allows the loop to snap straight. At this juncture ventricular extrastoles are usually noted on the monitoring cathode-ray oscilloscope. The polyethylene tubing is advanced along the guide into the desired location in the ventricle, and the guide is withdrawn to allow manometric study and the injection of contrast substance. Slight manipulation of the catheter will usually result in obtaining a position unassociated with ectopic beats.

After observation and recording of intraventricular pressure, contrast agent (amounting to about half of the patient's estimated resting stroke volume) is delivered by means of a pressure injector. Rapid serial radiographic exposures are made for 4 to 6 seconds from the start of the injection. The left posterior oblique projection (approximately 35 to 40° rotation) provides a satisfactory view of the left ventricle as well as its mitral and aortic valves.

With the pressure-recording system operating, the catheter is withdrawn until its tip lies above the level of the coronary arteries within the proximal aorta. Pressure curves indicate when a supravalvular position has been achieved, but—in children at least—it may be wise further to confirm the catheter location by fluoroscopy, 1 or 2 ml. of contrast substance being injected into the otherwise radiolucent polyethylene catheter. The inadvertent injection of the aortic dose of contrast substance into a carotid, vertebral, or coronary artery is the major hazard of this technic. Fortunately, it is readily avoided by careful technic. Filming of the supravalvular contrast injection is best carried out with the patient in the right posterior oblique projection (60 to 80° rotation). As soon as the blood pressure has stabilized, the catheter is withdrawn during continuous pressure recording, and a sterile hemostatic pressure dressing is applied over the femoral puncture.

Observation of intraventricular and intracoronary blood pressures usually establishes the diagnosis of stenosis of left ventricular outflow
Normal aortic valve. During diastole (A and C), two of the aortic leaflets are profiled as thin, uniformly thick curved lines that continue the contour established by their respective sinuses of Valsalva and meet centrally (white arrows in C). The third cusp is usually visible (black arrow in C). During systolic ejection (B) the cusps are normally more difficult to identify but often one or two are shown, at least in their proximal portions, as vertically oriented shadows roughly parallel to the aortic walls (white arrow in B). Difficulty in visualizing the free margin is due to motion. Note also the varying appearance of the normal subvalvular infundibulum of the left ventricle.

tract early in the procedure (fig. 1). When ventricular irritability occurs as the catheter is withdrawn from the ventricle, a subvalvular chamber may not be represented faithfully in the pressure record. The differentiation of pressure changes due to valvular artifact, premature ventricular contractions, or a small chamber above or below the valve may be difficult or impossible. The interpretation of pressure records is also complicated by the use of side-holes near the catheter tip designed to prevent catheter recoil during injection. Nevertheless, pressure studies provide important information concerning the presence and severity of trans-stenotic pressure gradients and, at present, are regarded as indispensable in the selection of patients for surgical correction. Inasmuch as the actual site of the stenosis and other valuable anatomic information are demonstrable by angiography, it is fortunate that both types of information can be gained with a single procedure.

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Valvular Aortic Stenosis

Isolated aortic valvular stenosis in adults has been ascribed to degenerative sclerosis, rheumatic infection, and congenital aortic disease. Coexistent mitral disease offers strong, if circumstantial, evidence of rheumatic origin. In its absence and without a history of rheumatic fever, the presumptive diagnosis of congenital aortic stenosis is preferable, particularly in young patients. Since calcification may develop in congenitally stenotic valves (it has been observed in patients as young as 18), calcification per se offers no support for the diagnosis of rheumatic aortic stenosis. There remains uncertainty about the cause of the disease, particularly in older patients—a doubt shared by pathologist and clinician. In any event, if rheumatic carditis is excluded, the etiology probably does not influence prognosis or therapy.
Congenital valvular aortic stenosis: rigidity. The four sequential views here reveal a thickened dome that does not perceptibly change its position during various phases of the cardiac cycle. The left ventricular-aortic systolic gradient was 124 mm. Hg.

Children with congenital lesions may experience angina, syncope, and heart failure. Sudden death as a complication of aortic stenosis is said to be frequent. In adults, the disease is reported to have been associated with sudden death in one out of every four autopsied cases. The classical symptoms of syncope, effort intolerance, dyspnea, and other manifestations of heart failure are well known. In adults, angina often indicates coexistent coronary arteriosclerosis. In this unfortunate and highly lethal combination, two distinct anatomic limitations additively oppose the increase in coronary flow demanded by the ventricular hypertrophy that results from the valvular obstruction. Life expectancy in such patients bears a direct relationship to the heart’s continuing ability to hypertrophy.

In severe aortic stenosis, regardless of the etiology, the peripheral pulse characteristically exhibits a low amplitude and prolonged rise-time, but this is not always so. Generally, the systolic blood pressure is below 140 mm. Hg but, again, exceptions have occurred. Usually aortic stenosis is associated with a systolic thrill at the base of the heart. A palpable increase in the left ventricular impulse is another commonly encountered physical sign in aortic stenosis and related obstructive lesions of the left ventricular outflow tract. The sine qua non of the clinical diagnosis is a loud systolic ejection murmur. Its point of maximum intensity is most often in the aortic area, but it can be at the apex. Appraisal of the relative intensities of the two valve-closure sounds and the modifications in their response to respiration are of value. A soft, or silent aortic closure is often but not always found in patients with aortic stenosis, whereas paradoxical splitting of the second sound is a common finding believed to be constant in severe stenosis.

Aortic diastolic murmurs are often heard in patients with aortic stenosis. It is clearly of clinical significance when due to aortic regurgitation of such a degree as to cause a diastolic thrill or the physical signs of abnormally high aortic run-off. Even in the absence of a diastolic murmur, associated aortic regurgitation of very slight degree may result in catastrophe during attempted open-heart surgical repair without coronary perfusion. Aortography not only can identify subclinical aortic regurgitation, it can exclude it.

The left ventricular enlargement that occurs in any type of aortic stenosis may be demonstrated by conventional radiologic and electrocardiographic techniques. Hypertrophy without dilatation of the left ventricle is common in the absence of heart failure or associated aortic or mitral insufficiency. Dilatation of the ascending aorta is a frequent manifestation of post-stenotic turbulence when obstruction antecedes the loss of aortic elasticity. It is also seen in aortic insufficiency without stenosis, syphilitic aortitis, hypertension, Marfan’s syndrome, coarctation of the aorta, aorticopulmonary defect, common
stenosis of left ventricular outflow tract

truncus, and other conditions including non-valvular congenital aortic stenosis. Like most of the findings already mentioned, aortic dilatation is at most confirmatory evidence of aortic valve narrowing. Considered in concert, the signs and symptoms of aortic valvular stenosis enable reasonably accurate diagnosis but it remains for left heart catheterization to provide essential knowledge of the individual anatomic changes and resultant trans-stenotic pressure gradient in blood pressure.

Pressures recorded from the left ventricle and proximal aorta provide quantitative data concerning the hemodynamic effects of aortic stenosis and guides to the adequacy of its surgical correction. In our opinion, surgery is indicated when nonvalvular aortic stenosis without significant aortic insufficiency has caused the left ventricular systolic pressure to exceed that in the aorta by more than 50 mm. Hg. It is far more difficult to assess the hemodynamic importance of aortic stenosis when mitral disease is also present. A resultant reduction in effective stroke volume may lower aortic valve gradients deceptively.

Contrast visualization of the left ventricle and aortic valve is indispensable to the competent preoperative work-up of patients with stenosis of left ventricular outflow tract. Adequate studies can be depended upon to reveal the structure and cyclic variations of the left ventricle, aortic valve, and proximal aorta. Directional and quantitative information about blood flow is also obtained and mitral insufficiency may be identified or excluded.

Identification or exclusion of aortic insufficiency may similarly be accomplished by means of supravalvular aortic injections. Here also, careful attention to technical details such as the orientation of the catheter tip will permit reliable conclusions. Clinically appreciable aortic regurgitation always results in radiologically visible retrograde passage of contrast agent through the abnormal valve into the left ventricle. When there is regurgitation sufficient to produce clinical evidence of functional impairment, the entire ventricle fills. In the functionally unimportant regurgitation, which often accompanies nonrheumatic aortic stenosis, there is apt to be a more transitory subvalvular puff of contrast agent. Functionally important aortic insufficiency produces other gross angiographic findings.

With supravalvular contrast injections the normal aortic valve is seen as a trilobulate expansion of the root of the aorta. The three dilatations are the shadow of contrast agent within the sinuses of Valsalva and are roughly equal in size. Films exposed during ventricular ejection usually show an irregular interface between radiolucent blood and contrast agent in the aorta. Estimates of the volume...
of ejection by measurement of the radiolucent area have serious shortcomings. When contrast agent is injected into the left ventricle and radiographs are exposed during diastole, two of the three aortic cusps are outlined in profile while the third is outlined en face by a contrast agent in its respective sinus. The normal aortic valve leaflets are symmetrical, smoothly contoured, evenly thin in radiographic cross section, and flexible as judged from exposures made during various phases of the cardiac cycle. In systole, the cusps are not as readily outlined because of their motion (best results are obtained through the use of short exposures). Often one or two of the cusps may be recognized as a short radiolucent line extending upwards from the point of attachment for a distance of about half the length of the cusp; motion or superimposed contrast agent obscures the free edge. The characteristic configuration of the normal aortic valve is well shown in figure 2.

When valvular stenosis is present, selective contrast visualization regularly demonstrates gross and characteristic abnormalities of the valve leaflets (figs. 3 to 8). In the absence of degenerative changes, congenital aortic stenosis is most commonly seen as a curved radiolucent line with convexity directed upwards. This is the cross sectional shadow of the dome formed by the incompletely segmented cusps and is usually best shown during systolic ejection. When contrast agent is injected above the stenotic valve, the latter outlines a rounded radiolucent defect at the base of the aorta. Diastolic exposures often reveal a surprisingly normal appearance although there is likely to be flattening of the sinus pockets. Figure 3 shows the striking systolic-diastolic variation that occurs in the still flexible, but congenitally unsegmented, stenotic aortic valve. That the dome in congenital aortic stenosis may be rigid is illus-

**Figure 6**

Congenital valvular aortic stenosis: degenerative change and eccentricity. Supravalvular injection reveals marked distortion of the sinuses of Valsalva. One large, evidently quite flexible valve cusp is present. Blood reaches the aorta through the eccentric orifice at the site of calcific deposits seen fluoroscopically. Pressures at time of visualization were left ventricle 210/10, thoracic aorta 140/90. Following surgery these fell to 150/5 and 140/110 respectively.

**Figure 7**

Rheumatic aortic valvular disease, predominant stenosis. A. Left ventricular injection of contrast agent. B and C. Supravalvular injection. The aortic valves are thickened, straightened, and quite rigid in all phases of cardiac cycle. The appearance is more like that of a tent than the rounded dome in early congenital stenosis. Despite chronicity of the disease process, the abnormal valve is more or less symmetrical in appearance. Moderate aortic regurgitation is demonstrated. The shadow of a thickened mitral valve is seen in A.
trated in figure 4. Diastolic exposures during supravalvular injections are most likely to reveal relatively minor aortic regurgitation but it should be noted that impingement of the catheter tip or the jet issuing from it can simulate regurgitation. In congenital aortic stenosis, supravalvular injection often reveals radiolucent blood spurtling through the stenotic orifice and gives rise to what might best be described as a systolic jet sign. This filling defect is narrow at its base and frequently is eccentrically located. The extent to which its axis deviates from the central axis of the aorta probably correlates with the presence of superimposed sclerotic change. The distortion and rigidity that accompany sclerotic degeneration (evidenced by calcification, age, rheumatic origin) are less likely to be associated with the jet sign. Instead, the films disclose asymmetric thickening, rigidity, and irregularity of the valve cusps (figs. 4 to 6). Asymmetrical involvement is not rare and, as shown in figure 6, may be associated with both an eccentric orifice and an abnormally large single sinus of Valsalva. Despite what appears to be prolapse of the relatively normal single cusp, we have not observed gross insufficiency in this situation. The large, flexible, relatively unaffected cusp may be used to achieve a competent valve in conjunction with surgical relief of the stenosis.8 Figure 7 shows a rigid, stenotic, incompetent but symmetrical valve in rheumatic heart disease of long standing. It also shows rigidity and calcification of the mitral valve. In figure 8, the lesion of rheumatic aortic stenosis and insufficiency was also relatively symmetrical. On the basis of these studies the superimposition of sclerotic degenerative change on a congenitally stenotic aortic valve seems more likely than rheumatic disease to cause an eccentric leaflet deformity.

Subaortic Stenosis

Though less common than the valvular variety, subaortic stenosis will be encountered in any thoroughly studied group of young patients with congenital "aortic stenosis" (8 of 21 proved cases in one reported series.9).

Figure 8

Rheumatic aortic valvular disease, predominant insufficiency. It was impossible to reach the left ventricle and supravalvular injection only was done. The cusps appear normal, but there is marked regurgitation into the left ventricle due to rheumatic aortic insufficiency.

In this probably congenital anomaly there occurs a fibrous ring or ridge, which partially or completely encircles the left ventricular infundibulum at a point 1 to 3 cm. below the normal aortic valve. Important transinfundibular pressure gradients have been repeatedly demonstrated. The symptoms, prognosis, and indications for surgery are related to the degree of obstruction, which varies considerably from case to case. In general, the clinical manifestations resemble those of congenital valvular stenosis.

The differentiation of subaortic and valvular stenosis has been attempted on the basis of the physical findings and the arterial pulse curve. The murmur is similar in both conditions. The presence of a diastolic notch in the arterial pulse curve, in combination with evidence of aortic stenosis has been put forth as indicative of subaortic stenosis. That this and
Figure 9
Congenital subaortic stenosis: long-segment infundibular type. A. Supravalvular injection. B-D. Left ventricular sequence. As shown in A, the cusps are normal, save for the presence of a small volume regurgitation, which could represent catheter artifact or subclinical insufficiency. Clearly shown on B-D (arrow) is an indentation of the left ventricular outflow tract border a short distance below the aortic valve. Pressures found at catheterization: Lower left ventricle, 260/20; aorta, 100/60. Pressures were not obtained in the infravalvular chamber owing to its small size. Following surgical correction of a fibrous ring encircling and deforming the outflow tract, left ventricular and femoral arterial systolic pressures of 125 mm. Hg were recorded. Hypertrophic muscular infundibular outflow tract played no surgically recognized role in this case.

Figure 10
Congenital subaortic stenosis: diaphragm type. A and B. Two different exposures of a subvalvular diaphragm (white arrows) located 3/4 inch below normal aortic valve cusps. As is illustrated here, the finding may be subtle in comparison to the pressure gradients produced. Lesion corrected at surgery. C and D. Subvalvular fibrous ring or diaphragm producing the pressure record illustrated below. Here, outflow chamber pressure was 175/0; the left ventricular subvalvular, 110/0; aortic 110/80. D also reveals the presence of minor aortic insufficiency. Open heart surgery confirmed the anticipated presence of a fibrous subvalvular ring.

dependable. Conventional fluoroscopy and radiographic examination of the heart is of little value, since the changes are common to both conditions. Calcification in the aortic valves supports valvular stenosis but even here exceptions have been noted.

The recording of blood pressure during the withdrawal of a catheter from the left ventricle to the aorta is a productive approach to the problem. In subaortic stenosis, there is an intermediate zone where ventricular complexes exhibit systolic peaks lower than those of the main ventricular cavity but equal to
those in the aorta. Here, too, artificial problems abound. In order to minimize catheter recoil and potentially hazardous displacement during forceful contrast injections, the catheters used for this purpose have a number of side orifices near the tip. If these are spread out over too long a segment, the curve may reflect the simultaneous effects of pressures on both sides of a stenotic lesion! Catheter withdrawal must be accomplished at a steady rate and in such a manner as to minimize the curve-corrupting effects of ventricular irritation. Side-holes in the catheter should be closely grouped near the tip. Pure chance appears to play a major role in any event.

Radiologic demonstration of subaortic stenosis requires the injection of contrast agent into the left ventricle. When present in its classical form (figs. 9 and 10), the subvalvular ring is represented by narrow, triangular, radiolucent indentations in the opposing margins of the otherwise tubular left ventricular infundibulum. Because of anatomic variations or projectional factors, this defect is likely to be visualized incompletely. The suprastenotic portion of the left ventricle has not appeared dilated in any of our patients. Although the lesion of subaortic stenosis may create seemingly minor changes on left ventriculography, pressure gradients may be major (fig. 10). The collective data obtained from both radiographic and pressure studies have thus far proved diagnostically sufficient.

**Supravalvular Aortic Stenosis**

Though a rarity, recent reports and our own experience suggest that congenital supravalvular stenosis of the aorta occurs more frequently than has been supposed. In this condition, the aortic lumen is narrowed by an inwardly protruding ring or band-like obstruction situated just above the aortic valve or 1 or 2 cm. distally, closely related to the point of origin of the coronary arteries. The ring is composed of dense, hyalinized collagen. Anomalies of the aortic valve are commonly encountered in association with the lesion. Bands from the free edges of the cusp to the aortic wall occur, as well as insufficient and bicuspid aortic valves.

That the anomaly is hemodynamically significant is evident from its appearance as well as the fact that young patients predominate in reported autopsy-proved instances. The effects of left ventricular obstruction are reflected in cardiac enlargement and congestive heart failure. Significant trans-stenotic gradients exist. Angina has been a symptom, but syncope has not thus far.

Though one reported patient is said to have had “no significant murmur,” a systolic murmur that decreases in intensity with quiet inspiration is characteristic in our experience. It is difficult or impossible to distinguish this lesion from congenital aortic valvular stenosis by conventional diagnostic means. The aortic valve closure sound should be normally audible and normal or reversed splitting of the second sound is possible. An associated
Figure 12
Supravalvular aortic stenosis, congenital: no valvular involvement. A and B. A supravalvular constriction is associated with marked enlargement of the left ventricle and hypertrophy of the coronary system. There is hypoplasia of the aorta distal to the site narrowing. Despite a mechanically satisfactory operative procedure, the patient failed to survive surgery, presumably because of left ventricular insufficiency. Had the lesion been picked up sooner, the result might have been different. C and D. Case reported in detail elsewhere.10 Frontal and lateral films with injection into the left ventricle disclose a pronounced constriction of the ascending aorta at the level of the coronary artery origin. Catheter side-holes made impossible the manometric identification of the supravalvular chamber. Pressures recorded at time of surgery were left ventricular 165/10; aortic 100/50. The surgical procedure was planned on the basis of the films and consisted of an incision carried through the constriction down into the noncoronary sinus. The vertical incision was closed with an oval patch of Ivalon restoring approximately normal aortic caliber. Postoperative result has been considered excellent.

murmur of aortic insufficiency may denote that one or more aortic cusps are involved in the deformity. Conventional roentgenologic examination has revealed left ventricular enlargement and the changes of congestive heart failure, but usually does not show aortic dilatation. Though the lesion can be corrected by surgery in certain patients, open-heart procedures are required. In addition, the problem of attachment of aortic valve cusps to the ring-stenosis and the relation of the latter to the coronary orifices provide added technical problems. The foregoing make mandatory the use of trans-stenotic catheter studies in the preoperative evaluation of patients with congenital aortic stenosis. Such studies are fortunately definitive in supravalvular aortic stenosis.

Since the lesion exists at a supraventricular level, manometry is not likely to be complicated by the multiple ectopic beats so often associated with the pull-out pressure records in valvular or subvalvular disease. The shorter the supravalvular pocket, the more important it is that the catheter side-holes be confined to its distal portion. In our three cases of supravalvular aortic stenosis, the constriction was sufficiently far above the level of the valve to permit the recording of the pathognomonic pressure curves. In this condition, three distinct pressure zones exist.

High ventricular systolic pressure is accompanied by similarly high but otherwise unremarkable proximal aortic pressures although aortic insufficiency may result in abnormally low aortic diastolic pressures. Distal to the stenosis, the systolic level falls abruptly, diastolic pressure being relatively unchanged. It would be difficult to distinguish between the pressure tracings of coarctation of the aorta and the ideal pull-out record of supravalvular stenosis were the catheter positions not known. The important clinical, physiologic, and therapeutic differences between these two conditions are primarily a matter of their location.

Supravalvular aortic stenosis, in the three cases we have observed, has appeared as an unmistakable, bizarre, symmetrical constriction of the aortic lumen at about the level of origin of the coronary arteries, i.e., approximately 1 inch above the aortic valve ring. Although in one of our cases the lesion was shown by conventional angiocardiography, the...
superiority of selective injection was unequivocally established when this technic was carried out in the same patient. In one case (fig. 11) a band-like connection was shown between the site of stenosis and the aortic valve. Surgery in this patient has been deferred in the absence of evident left heart embarrassment. That it will be necessary in a few years' time seems likely. Angiographic visualization is of aid, not only in establishing the anatomic diagnosis, but in planning the operative repair. After considerable study of the films reproduced in figures 12C and D, a vertical incision through the aortic wall into the noncoronary sinus and transecting the stenotic zone was made and closed with an oval Ivalon patch with excellent results.10 A similar surgical procedure was accomplished in another patient (figs. 12A and B), but he did not survive.

Hypertrophic Muscular Obstruction of the Left Ventricular Infundibulum

This curious syndrome, also known as familial muscular subaortic stenosis, has been recognized only recently. In rare patients, the left ventricle hypertrophies so as to create an intrinsic obstruction to its own outflow. Important, and occasionally very large, pressure gradients have been recorded, the transition between high and low pressure regions being found in the subaortic region. Brock11 has described such an occurrence resulting from systemic hypertension and regards it as an inoperable form of stenosis. Subsequently, others demonstrated a familial incidence of the lesion but cast no light on the cause of the marked cardiac hypertrophy that was found (since systemic hypertension was not present). In the familial form, deaths have occurred in young adulthood. Evidently in some patients with aortic valvular stenosis, resultant subaortic muscular hypertrophy may progress to the point at which an actual impediment to left ventricular ejection is created. Such a condition has its analog in right ventricular infundibular hypertrophy. Muscular subvalvular stenosis associated with valvular aortic stenosis regressed following aortic valvulotomy in a patient reported by Morrow and Braunwald.12

Diagnosis is simplified when the disease exists in its familial form. An ejection murmur may be heard along the left sternal border and at the apex better than in the aortic area, a finding that does not exclude aortic valvular stenosis. To our knowledge, poststenotic dilatation has not occurred in re-

Figure 13

Hypertrophic muscular infundibular stenosis of the left ventricle. Two views made during catheter injection into the left ventricle disclose a curved defect and posterior displacement of the outflow tract of the left ventricle. The catheter recorded a pronounced gradient across the infundibulum. There is in addition to the foregoing, a bimarginal indentation in the outflow tract approximately 2.5 cm. below the level of normal aortic valve. Surgery was not attempted and this patient died within 6 months of progressive left heart failure. Autopsy disclosed an unimpressive subvalvular connective tissue ring and considerable hypertrophic enlargement of the intraventricular septum. The latter is believed to have been responsible for the pronounced pressure gradient that was encountered at left heart catheterization. It is possible that the infundibular hypertrophy occurred as a result of the subaortic ring. Catheterization pressure record (below) reveals a systolic gradient in the outflow tract.
Infundibular obstruction of left ventricle: congenital subaortic ring and associated or secondary hypertrophic muscular obstruction. A-D. Two views showing a discrete conical subvalvular outflow tract chamber. While the appearance of the outflow tract suggests muscular hypertrophy of the interventricular septum, the well-defined subvalvular chamber in which the catheter lodged repeatedly probably is due to a fibrous subvalvular ring, i.e., subaortic stenosis. Since the patient has not yet been operated upon, it cannot be determined whether hypertrophy is significant and, if so, whether it is primary or secondary in etiology. Pressures obtained at catheterization (illustrated) were left ventricle, 145/5; subvalvular chamber, 100/5; aorta 100/72.

Figure 14

In response to muscular obstruction of left ventricular outflow tract. Insofar as the direct approach is concerned, this lesion is not generally considered to be surgically correctible.

In two cases we have studied (figs. 13 and 14), there was left infundibular stenosis and radiologic evidence of enlargement of the interventricular septum. Unlike the more familiar process in the right ventricle, the filling defect seemed to reflect enlargement of the upper half or two thirds of the interventricular septum. The diagnosis of intrinsic muscular infundibular obstruction of the left ventricle by means of left ventriculography should not be accepted unless the "abnormal" radiographic findings are supported by the demonstration of a pressure gradient. Systolic contraction of the normal or ordinarily hypertrophied left ventricle can simulate the finding.

Conclusions

Competent differential diagnosis of the lesion responsible for occlusion of the outflow tract of the left ventricle requires that maximum advantage be made of the specialized technics afforded by left heart catheterization. If, in the course of study of patients with evident aortic stenosis, left ventricular and aortic pressure determination and contrast visualization are routinely carried out, it will be possible to select patients for surgery wisely. By providing precise anatomic information about the location, cause, and abnormal anatomy of the individual obstruction, selective left heart contrast visualization has assumed a critical diagnostive role. Fortunately, it may be accomplished by a safe, reasonably simple percutaneous procedure, which also permits the study of important transstenotic pressure gradients. It is hoped that through further information gained from such technics, the over-all spectrum of obstruction of the left heart outflow tract will be resolved and clarified well beyond the extent achieved in this report.

References


Sir Thomas Browne
1605–1682

Sir Thomas Browne, one of the most illustrious medical philosophers, attended Oxford, studied medicine and practiced for 45 years in Norwich where he led a quiet, uneventful life. He had traveled widely on the continent and is said to have received his M.D. degree at the University of Leyden. His eldest son, Edward, became president of the Royal College of Physicians. The fame of Sir Thomas Browne rests almost entirely on four works: Religio Medici, Pseudodoxia Epidemica, a discourse on many errors; Urne-Buriall, a discourse of the sepulchral urns lately found in Norfolk; The Garden of Cyprus, the network plantations of the ancients.

In 1642 a copy of his Religio Medici, which he described as “a private exerise directed to myself” was printed from one of his manuscripts and became popular with remarkable rapidity. His erudition, his speculations on the problems of existence, and the quaintness of his style have sustained the appeal of his writings over the intervening centuries. A total of over 55 editions has been published in English, French, Dutch, and German. “I love,” he says, “to lose myself in a mystery, to pursue my reason to an O, Altitudo.” Interested in natural history, archeology, literature, and theology and blessed by a wide circle of scientific friends, his reflections reached into innumerable areas of inquiry.

In 1664 he was made an honorary Fellow of the Royal College of Physicians and in 1671 he was knighted by Charles II. As Osler remarks, “A life placid, uneventful and easy, without stress or strain, happy in his friends, his family and his work, he expressed in it that harmony of the inner and of the outer man which it is the aim of all true philosophy to attain, and which he inculcated so nobly and in such noble words in the Religio Medici and in the Christian Morals.”

The end came unexpectedly in his 77th year on his birthday, October 19, 1682.
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