Pathology of Left Ventricular Outflow Tract Obstruction

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The anatomic sites at which obstruction to the egress of blood from the left ventricle may occur are three in number, as follows (fig. 1): the aorta, the aortic valve, and the subaortic region of the left ventricle.

Obstruction in the aorta may involve one of two general areas, namely, (1) the arch or just beyond, the lesions taking the forms of classical coarctation, tubular hypoplasia, or interruption of the arch, or, (2) it may involve the ascending aorta. As the clinical manifestations of obstructive lesions in the vicinity of the aortic arch are not usually confused with obstruction in the region of the aortic valve, the author has chosen not to consider these conditions further in this symposium. Rather, consideration to obstructive lesions of the aorta will be confined to those that involve the ascending aorta, lesions which, while varied in appearance, are usually congenital in nature and, collectively, may be termed supravalvular stenosis of the aorta.

Obstruction at the aortic valve itself, termed either aortic valvular stenosis or simply aortic stenosis, may be represented by a congenital malformation or may result from acquired disease affecting either an initially normal valve or one that is the site of a congenital malformation; the latter most commonly takes the form of a congenital bicuspid valve.

Obstruction occurring proximal to the aortic valve in the outflow tract of the left ventricle is commonly known as subaortic stenosis. This condition is represented by a wide variety of anatomic states, among which are intrinsic anomalies of the left ventricular outflow tract and conditions that primarily involve the mitral valve. Some of the latter conditions are congenital in nature, others are acquired, some being iatrogenic.

Supravalvular Aortic Stenosis

Among a small proportion of patients who show clinical signs of obstruction in the region of the aortic valve, detailed observations, such as those obtained through left-sided cardiac catheterization, aortography, operative exploration or necropsy, reveal the obstruction to be in the ascending aorta rather than in the aortic valve. Anatomic details vary among such cases which, collectively, may be said to represent supravalvular aortic stenosis. One may recognize three structural types of supravalvular aortic stenosis (fig. 2), although in individual cases the lesion may appear to represent a twilight stage between one designated type and another. The three commonly recognized types of supravalvular aortic stenosis1 in order of decreasing frequency

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Figure 1

Anatomic sites at which obstruction to the left ventricular outflow may occur. In the case of supravalvular aortic stenosis, the obstruction lies distal to the coronary arteries. In contrast, in aortic valvular stenosis and in subaortic stenosis, the coronary arteries arise distal to the obstruction.
are the hour-glass, membranous, and hypoplastic.

The *hour-glass type* of supravalvular aortic stenosis, as the name implies, is characterized by an hour-glass deformity of the external aspect of the ascending aorta (fig. 3, a and b). Corresponding with this deformity is a narrow state of the lumen of the aorta that lies at a level just distal to the ostia of the coronary arteries. Fibrous intimal thickening may be superimposed over the deformity at the level of greatest narrowing.

The *membranous type* of supravalvular aortic stenosis is characterized by the presence, in the aortic lumen, just distal to the ostia of the coronary arteries, of a fibrous diaphragm, containing a single perforation. The *hypoplastic type* involves the entire ascending aorta beginning distal to the ostia of the coronary arteries.

As a consequence of the severe degree of obstruction to the egress of blood from the left ventricle, the wall of this chamber usually shows pronounced concentric hypertrophy. The coronary arteries assume a peculiar role in supravalvular aortic stenosis, in contrast to aortic valvular stenosis and to subaortic stenosis.

**Figure 2**

Diagrammatic portrayal of the three major types of supravalvular aortic stenosis. a. The hour-glass type is the most common variety. b. The least common variety is the hypoplastic type, in which there is tubular narrowing of the entire ascending aorta. c. The membranous type, which is intermediate in incidence between the two foregoing types.

**Figure 3**

Base of left ventricle and ascending aorta in a 23-year-old woman with the hour-glass type of supravalvular aortic stenosis. a. External view shows classical deformity (between arrows) of the hour-glass type of supravalvular aortic stenosis. The large vessels in the foreground are ramifications of the left coronary artery. b. Sagittal section through the ascending aorta and base of left ventricle. Obvious narrowing of ascending aorta immediately above the aortic valve is associated with the thickening of the aortic wall. Adhesion of the right aortic cusp to the site of supravalvular aortic stenosis (point of arrow) has excluded the right aortic sinus (R.S.) from the aortic lumen. Associated is a narrow and thin-walled right coronary artery (R.C.), while the left coronary artery (L.C.) is dilated and tortuous.
osis. Whereas, in the last two conditions the coronary arteries arise distal to the site of obstruction, in supravalvular aortic stenosis the coronary arteries arise proximal to the site of obstruction and, hence, are subjected to the same systolic pressure as is exerted by the left ventricle. Changes are recognizable in these vessels as a consequence. These are tortuous, with wide external dimensions (fig. 4a). The walls of the vessels display medial hypertrophy. Also, focal luminal narrowing may result from focal intimal changes in the form either of nonspecific fibrous thickening or of premature atherosclerosis (fig. 4, b and c).

According to a review of the literature by Peterson and associates, in about one quarter of cases in which anatomic observations have been made there are associated changes of the aortic valvular cusps. The most significant is characterized by adhesion of all or part of the free edge of one or several cusps to the intima of the ascending aorta at the site of the stenosis. In a case reported by Kreeel and associates, and in a case that we observed, the entire free edge of the right aortic cusp was so adherent, the adhesion being above the level of the ostium of the right coronary artery. The adhesion caused the right aortic sinus to be a blind sac from which the right

Figure 4

coronary artery arose. In each of these cases the right coronary artery was thin, while the left coronary artery was grossly changed in the manner usually seen in supravalvular aortic stenosis. It is assumed that in each of these cases, wherein the right coronary artery lacked a direct connection with the aorta, the ramifications of this vessel derived their blood from collateral channels joining the left coronary artery.

Supravalvular aortic stenosis has interesting but not universal associations with other conditions. Among these are peripheral pulmonary arterial stenosis (rarely hypoplasia of the pulmonary trunk), stenosis of the branches of the aortic arch (fig. 4 d), mental deficiency with a characteristic facies, and by hypercalcemia.

Beuren and associates described cases of supravalvular aortic stenosis in which each of the conditions that may be associated with supravalvular aortic stenosis occurred in the same patients. The full syndrome expressed in these patients consisted of supravalvular aortic stenosis, peripheral pulmonary arterial stenosis, mental retardation, a peculiar facial appearance, and complex dental malformations.

Bacterial infection originating in the intima of the stenotic segment of the aorta occurs as an uncommon complication. Such a complication was observed in one of the five pathologic specimens of supravalvular aortic stenosis that we have studied.

Another complication is dissecting aneurysm of the aorta beginning just distal to the zone of stenosis in the ascending aorta, a complication that also occurs occasionally in aortic valvular stenosis.

Aortic Valvular Stenosis

Among patients with obstruction to the flow of blood across the aortic valve, the anatomic basis for obstruction varies. In general, the type of valvular lesion to be found is related to the age at which signs of aortic valvular disease develop. Accordingly, patients with aortic stenosis may be placed into three categories: adults; older infants, children, and adolescents; and young infants.

Adults

In adult patients with obstruction at the aortic valve, the lesion is universally stenotic, in contrast to valvular atresia, which may be observed in some young symptomatic infants.

As in all forms of obstruction to outflow from the left ventricle, the high level of left ventricular systolic pressure resulting from aortic valvular stenosis is followed by concentric left ventricular hypertrophy. The thick mass of left ventricular myocardium offers more resistance to filling of the left ventricle than does a normally thick left ventricle, and there is a corresponding elevation of the left ventricular filling (end-diastolic) pressure, even when the left ventricle is compensated. Left ventricular end-diastolic pressure is elevated to even greater levels when left ventricular failure supervenes.

Elevated left ventricular diastolic pressure is associated with corresponding elevations of pressure in the entire pulmonary circuit and in the right ventricle. In aortic stenosis, right ventricular hypertrophy may, therefore, follow left ventricular hypertrophy.

In the usual adult with aortic valvular stenosis, no abnormal communications are present and no intracardiac shunt is present.

The aortic valvular stenosis seen in adult patients takes two main forms (fig. 5), the fibrous and the calcific. Among cases of each of these anatomic types there are two major backgrounds, namely, congenital deformity of the valve and acquired disease resulting from rheumatic endocarditis.

Among examples of the fibrous type of aortic valvular stenosis in adult patients, a rheumatic background is more common than a congenital one. Characteristically, fibrous aortic stenosis resulting from rheumatic endocarditis is represented by interadhesion between aortic cusps at two or at each of the three commissures. The binding effect of the interadhesion restricts the motion of the cusps and is the principal basis for stenosis. In instances wherein there is fusion of cusps at each of the three commissures, there is con-
Figure 5

Drawings of unopened aortic valves viewed from above, including the normally tricuspid aortic valve (a) and the congenitally bicuspid aortic valve (d). Congenital unicommissural aortic valvular stenosis illustrated in b is a form of fibrous aortic stenosis. Such valves may secondarily become calcified, as shown in c. The congenitally bicuspid aortic valve has a tendency to become calcified and to yield calcific aortic stenosis with the features shown in e. Rheumatic disease of the aortic valve may result in fusion of one or more of the aortic commissures. In f is shown the phenomenon of fusion at one commissure yielding an acquired bicuspid aortic valve. In g is illustrated fusion of two of the commissures yielding an acquired unicommissural aortic valve which is stenotic. Secondary calcification may occur. When fusion of each of the three commissures occurs, as in h, a severe form of fibrous type of aortic stenosis develops. Aortic valves altered as in g and h frequently are associated with rheumatic disease of other valves, particularly the mitral.

Considerable fixation of the cusps, to the extent that the only opening in the valve is a central perforation surrounded by the interadherent cusps. Such valves, while severely stenotic, are concomitantly incompetent. When fusion occurs at only two commissures there may be sufficient mobility of cuspid tissue to the degree that aortic valvular incompetence is not necessarily present.

The fibrous type of acquired aortic valvular stenosis appears to be a direct result of rheumatic endocarditis. Such valves frequently are associated with rheumatic disease of the mitral valve and not infrequently with rheumatic disease of the tricuspid valve, as well.

The congenital type of fibrous aortic valvular stenosis characteristically is represented by the unicommissural aortic valve. In this type of valve there is but one cusp and one commissure. Attaching to the aortic wall at one region, the single cusp runs toward the opposite wall of the aorta, but fails to join the aortic wall in that position. Instead, the cusp turns on itself and makes a second attachment with the aortic wall near the first attachment to create the single commissure characteristic of this valve. Being deformed as it is, the valve bears some resemblance to the pulmonary valve in so-called dome-shaped congenital stenosis. Usually two shallow equidistant raphes extend from the aortic wall onto the aortic face of the single aortic cusp.

The congenital unicommissural aortic valve is intrinsically stenotic and, in some instances, incompetence of the valve may be manifested additionally. In those instances wherein the single cusp is taller than average, incompetence is presented by buckling of one side of the single cusp. During ventricular diastole the buckled portion of the cusp presses upon the opposite wall of the cusp to create a flutter-valve action which prevents regurgitation.

The calcific type of aortic valvular stenosis is seen almost universally in valves that lack the usual three cusps. In the overwhelming number of instances the valve is bicuspid, either on an acquired or on a congenital basis. Uncommonly, calcification involves a congenitally unicommissural valve. In such instances the calcification itself does not increase the degree of stenosis, but the rigidity of the cusp imparted by its calcification may prevent a flutter-valve action during diastole and incompetence of the valve may appear additionally.

Calcific aortic stenosis is the most common variety of aortic valvular stenosis seen in adults and, as indicated, usually involves a valve that is bicuspid. In the past, it was suggested that in the majority of instances the bicuspid nature was usually a result of rheumatic endocarditis; in only a small pro-
portion of cases of calcific aortic stenosis was a congenital background for the bicuspid nature suggested, until the reports of Bacon and Matthews.\textsuperscript{17} Recent study suggests that the reverse is true, namely, that a congenital background for the bicuspid aortic valve in cases of calcific aortic stenosis is more common than a rheumatic background.

Regardless of the specific background for a bicuspid aortic valve, its relationship to calcific aortic stenosis has been explained as follows. The dynamics of valvular motion are probably more stressful to the cusps of a bicuspid valve than to the three cusps of a normal aortic valve. The abnormal stresses on the bicuspid valve lead to alterations in the connective tissue of the cusps, alterations that culminate in calcification. Once calcified, the cusps become rigid and stenosis of the valve is a natural effect.

**Older Infants, Children, and Adolescents**

Of the three groupings by age under the subject of aortic valvular stenosis, the most uniform pathologic findings are in older infants, children, and adolescents.

Aortic valvular stenosis, which becomes apparent in this range of age, is probably always of congenital origin. The valve is fibrous and usually exhibits the unicommissural type of aortic stenosis (fig. 5b).

The secondary effects of aortic stenosis, which are observed in adults, are duplicated in this group. One deviation is that structural changes in the lungs characteristic of pulmonary venous obstruction tend to be more highly developed than in the adult. In this group, the foramen ovale is often sealed anatomically, and no basis for a central shunt is present.

Calcification is usually absent from the aortic valve but, when seen in this age group, occurs in the older subjects and usually is present only to a minimal degree.

**Young Infants**

In infants of all ages, obstruction at the aortic valve is congenital in origin, but in very young infants one finds the greatest variation in the basic process and in its secondary effects.

The variants may be placed into three main categories (fig. 6): (1) aortic valvular stenosis with a normally sized left ventricular chamber and either with or without significant endocardial fibroelastosis of the left ventricle; (2) aortic valvular stenosis associated with hypoplasia of the left ventricle and with endocardial fibroelastosis; and (3) aortic valvular atresia.

In young infants aortic valvular stenosis with a normally sized left ventricle (fig. 6a) may look like that seen in older groups. The basic problem revolves around the simple point of obstruction at the aortic valve. In those cases of aortic stenosis with a normally sized left ventricle and with endocardial fibroelastosis, the latter process influences the circulation. Mitral insufficiency commonly is present and the restricted excursion of the left ventricle peculiar to endocardial fibroelastosis is manifest.\textsuperscript{18} No shunt is present in this condition, however.

In cases of aortic valvular stenosis associated with left ventricular hypoplasia and endocardial fibroelastosis (fig. 6b), the aortic stenosis assumes a position of secondary importance relative to problems concerned with the left ventricle. The changes in the left ventricle are responsible for obstruction to the flow of blood into this chamber. Under these circumstances, the resultant elevated pressure in the left atrium is often responsible for herniation of the valve of the foramen ovale into the right atrium, thereby establishing an interatrial communication. Through this opening some of the left atrial blood is shunted into the right atrium. Since the interatrial opening is usually small and the left ventricle fails to accept a full complement of blood, pulmonary venous hypertension of considerable proportion may develop. This is associated with prominent right ventricular hypertrophy.

In such cases, as the most severe effects are concentrated in the pulmonary vascular bed, clinical attention is usually directed to the pulmonary venous system and the right
ventricle rather than to the aortic valve.\textsuperscript{19}

In the majority of subjects with aortic valvular atresia (fig. 6\textsuperscript{c}), the other valves usually are patent, although the mitral valve is hypoplastic. Less commonly, aortic valvular atresia is associated with mitral atresia. In either situation, there is major embarrassment of the circulation in the neonatal period, and only in exceptional cases does the patient live beyond a week of age.

The fundamental problem is that there is no normal route for the flow of blood through the left ventricle. As a consequence, blood from the left atrium is delivered into the right atrium through a narrow opening in the atrial septum. The ventricular septum is intact and the right ventricle functions as a common ventricle, supplying the lungs through the pulmonary arteries, and the aorta through a patent ductus arteriosus.

The combination of systemic levels of the pulmonary arterial pressure and pulmonary venous obstruction (as a consequence of the narrow opening in the atrial septum) appears to be the factor most responsible for the characteristic early death, through the development of pulmonary edema.

In aortic atresia blood flows in a retrograde direction through the arch and ascending portions. As the coronary arterial ostia lie distal to the atretic aortic valve, blood to these vessels is delivered through the ascending aorta. The latter structure has no function but to carry blood to the coronary arteries. The ascending aorta is so hypoplastic that it is at times overlooked in the pathologic dissection. Under such circumstances, a faulty patholog-

\textit{Figure 6}

The range of aortic valvular obstruction that may be observed in young infants. a. The aortic valve shows the unicommissural type of stenosis as that in figure 5 b. The left ventricle is of normal size and the wall is hypertrophied. Endocardial fibroelastosis of the left ventricle may be associated and when present may represent a secondary phenomenon. b. Hypoplastic left ventricle with endocardial fibroelastosis and aortic stenosis. In this situation, the aortic valve is like that shown in a, but the left ventricle is hypoplastic and its endocardium grossly thickened with fibrous and elastic tissues. The latter process may contribute more toward dysfunction than does the aortic stenosis. The process restricts filling of the left ventricle. Under these circumstances, a secondary interatrial communication may develop leading to a left-to-right interatrial shunt. Not shown in this illustration is a patent ductus arteriosus, although some infants with this type of malformed heart may exhibit a patent ductus arteriosus, as well. c. Aortic atresia. In this condition the normal route of outlet of blood from the left side of the heart is absent. An interatrial communication allows flow from the left atrium into the right. Arterial supply is derived from the pulmonary trunk by way of the patent ductus arteriosus.
ic diagnosis of persistent truncus arteriosus is made, in which the enlarged pulmonary trunk is considered to be a persistent truncus arteriosus. An awareness of the fact that the coronary arteries do not arise from this vessel should stimulate further search that would lead to identification of the hypoplastic ascending aorta and of the coronary arteries arising from it.

Subaortic Stenosis

Reference to the classification of subaortic stenosis (table 1) reveals that this is a complex subject. This is derived from the peculiarities of the anatomic features of the left ventricular outflow tract. It is therefore proper to review the highlights of this region before considering the various entities which may be responsible for subaortic stenosis.

Anatomy of the Left Ventricular Outflow Tract

The anterior, right, and posterior walls of the left ventricular outflow tract are formed by the anterior, septal, and posterior walls of the left ventricle (fig. 7). The medial wall is formed by the anterior leaflet of the mitral valve, which accounts for a changing caliber of the tract during the cardiac cycle. During ventricular diastole, the subaortic area is narrowed as the anterior leaflet of the open mitral valve is directed toward the ventricular septum. During systole, this leaflet moves away from the ventricular septum. In so doing, the leaflet participates in closure of the mitral valve and at the same time allows the subaortic tract to enlarge. This is ideal, since at this stage blood flows through the subaortic tract on its way to the aorta.

It is evident from the foregoing that the mitral valve may play a significant role when abnormal processes involve the subaortic tract. Primary conditions of the subaortic tract may affect the mitral valve secondarily and, contrariwise, certain conditions that are primary in the mitral valve may cause subaortic stenosis, secondarily.

Primary Anomalies of Left Ventricular Outflow Tract

Anomalies of the left ventricular outflow tract that cause subaortic stenosis classically are of two principal types, the membranous and the muscular (so-called hypertrophic). The membranous type is characterized by an encirclement of the outflow tract by a membrane of fibrous tissue (fig. 8). As the mitral valve is involved by this process, mitral insufficiency may be associated with this form

Table 1

Classification of Subaortic Stenosis

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<tr>
<th>A. Anomalies of the left ventricular outflow tract</th>
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<tr>
<td>a. Membranous subaortic stenosis</td>
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<td>b. Muscular (hypertrophic) subaortic stenosis</td>
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<td>B. Diffuse or multifocal involvement of ventricular myocardium</td>
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<td>a. Muscular subaortic and subpulmonary stenosis</td>
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<td>b. Idiopathic myocardial hypertrophy with secondary subaortic stenosis</td>
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<tr>
<td>c. Glycogen-storage disease of striated muscle with secondary subaortic stenosis</td>
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<tr>
<td>C. Lesions of the mitral valve</td>
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<td>a. Malinsertion of prosthetic mitral valve</td>
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<td>b. Accessory tissue</td>
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<td>c. Anomalous basal attachment of anterior leaflet</td>
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<td>d. Anomalous insertion of chordae</td>
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<td>D. Complexes of anomalies</td>
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<tr>
<td>a. Persistent common atrioventricular canal (so-called endocardial cushion defect)</td>
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<td>b. Parachute mitral valve, supravalvular ring of left atrium, coarctation, and subaortic stenosis</td>
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<tr>
<td>c. Corrected transposition and anomalous muscle bundle</td>
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<td>d. Ventricular septal defect and subaortic stenosis</td>
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<tr>
<td>1. Defect distal to obstruction</td>
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<td>2. Defect proximal to obstruction</td>
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of subaortic stenosis. The muscular form of subaortic stenosis is characterized by localized hypertrophy of the ventricular septum in the subaortic area (fig. 9). In this condition, the primary cause of obstruction appears to be protrusion of an abnormal mass of myocardium into the channel of the left ventricular outflow tract. Secondary endocardial thickening may occur over the protruding muscle and on that portion of the mitral valve which lies opposite the mass. This new tissue may compound the obstruction caused by the abnormality of the muscle, and it may also cause sufficient immobilization of the anterior mitral leaflet to be responsible for mitral insufficiency.

**Diffuse or Multifocal Involvement of the Ventricular Myocardium**

Diffuse or multifocal involvement of the ventricular myocardium may be responsible for complex abnormalities, of which subaortic stenosis may be a part.

In *idiopathic myocardial hypertrophy*, a condition that has a familial tendency, the myocardium of both ventricles is grossly hypertrophied. Subaortic stenosis alone or infundibular stenosis of the right ventricle, as well, may be associated. In this condition, subaortic stenosis, when present, is probably an incidental feature in a more fundamental process involving the myocardium, generally, or idiopathic hypertrophy and localized muscular subaortic stenosis may be variants of the same basic process.

Another process that involves the myocardium diffusely and that may be associated with secondary subaortic stenosis is *glycogen-storage disease of striated muscle*. In this condition, both cardiac and skeletal muscles are involved. Subaortic stenosis does not occur universally in this condition.

A condition that may be a variant of muscular subaortic stenosis is that in which there is localized hypertrophy of the base of the ventricular septum that causes both subaortic and right ventricular infundibular stenosis. In order to avoid oversimplification, this combination is placed under this category, in the classification employed, rather than as a vari-
ant of primary muscular subaortic stenosis. As greater experience is gained with this condition, it may, however, be so considered in the future.

**Lesions of the Mitral Valve**

The possibility that lesions of the mitral valve may cause subaortic stenosis was emphasized by Ferencz in 1957. This subject was the basis of a recent report by Sellers and associates from our institutions. Several conditions are involved.

*Malinsertion of a prosthetic mitral valve* may cause subaortic stenosis. This circumstance involves certain patients in whom a diseased mitral valve is replaced by a *Starr-Edwards valve* and in whom the anterior mitral leaflet is incompletely removed. If the artificial valve is sewn to this leaflet some distance from its basal attachment, the seat of the valve and the attached tissue form a bulky mass that protrudes into and obstructs the left ventricular outflow tract (fig. 10).

*Accessory tissue of the mitral valve* occurs rarely and may be represented by masses that present in the subaortic area and obstruct the pathway to the aortic valve. Successful removal of such tissue from one patient with subaortic stenosis was reported by MacLean and associates. This anomaly has its counterpart in corrected transposition. In the latter condition the accessory tissue involves the venous (right-sided) atrioventricular valve and causes subpulmonary stenosis.

In the experience of Björk and associates, subaortic stenosis may be observed as a result of *anomalous basal attachment* of the anterior leaflet of the mitral valve.

In the material that we have studied personally, there are examples of subaortic stenosis caused by *anomalous insertion of mitral chordae*. This involves the situation in which, in the absence of septal defects, there

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**Figure 8**

Interior of base of left ventricle and of ascending aorta in a 12-year-old boy in whom coarctation of the aorta had been repaired at 4 years of age. The illustration shows the membranous type of subaortic stenosis in which the left ventricular outflow tract is narrowed by the deposition of a collar of fibrous tissue along its walls, including that part of the wall formed by the anterior leaflet of the mitral valve.

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**Figure 9**

Muscular subaortic stenosis. a. The base of the left ventricle and aortic valve opened in the conventional manner. Beneath the aortic valve the ventricular septum (V.S.) bulges prominently into the left ventricular outflow tract. Over the prominence of muscle the left ventricular endocardium (E.) is thickened. b. The aortic valve and subaortic zone of the left ventricle viewed from above. The leaflets of the valve have been retracted to reveal, beneath the valve, the prominence (E.) caused by the ventricular septum.
Subaortic stenosis induced by placement of an artificial mitral valve. Sagittal section through the heart of an adult in whom both artificial aortic (A.) and mitral (M.) valves were inserted. The patient’s anterior mitral leaflet (A.M.) had not been excised and the seat of the artificial valve had been sewn into the anterior leaflet at a level well below the annulus. The bulk of the seat of the valve and the underlying natural mitral valvular tissue form a mass which obstructs the subaortic area (Sub. A).

is a cleft in the anterior leaflet of the mitral valve, and chordae that arise at the edges of the cleft mitral leaflet insert into the ventricular septum. These chordae prevent the normal movement of the anterior mitral leaflet away from the ventricular septum during ventricular systole. Subaortic stenosis is caused by the abnormally restrained mitral leaflet. In this type of subaortic stenosis the restrained and cleft anterior mitral leaflet is also responsible for mitral insufficiency.

**Complexes of Anomalies**

**Persistent Common Atrioventricular Canal**

(So-called Endocardial Cushion Defect)

In persistent common atrioventricular canal (so-called endocardial cushion defect), the characteristics of the atrial septal defect and of cleft atrioventricular valve or valves are well known. The matter of deficiency of the ventricular septum requires emphasis, particularly as it is pertinent to the subject of sub-

aortic stenosis. The deficiency of the ventricular septum lies anterior to the mitral valve in that part of the septum which contributes to the wall of the subaortic tract of the left ventricle.

In some cases, the ventricular aspect of the anterior mitral leaflet is adherent to the posterior edge of the ventricular septum. In this circumstance the subaortic area is intrinsically stenotic.

In other cases, the union of the anterior mitral leaflet with the ventricular septum is indirect, through chordae (fig. 11). In such cases, interventricular communications occur between the chordae and, in the natural state, no subaortic stenosis is present. If, however, during attempted surgical correction the anterior leaflet is sewn directly to the ventricular septum, subaortic stenosis is induced.

**Complex of Supravalvular Stenosis of the Left Atrium, Parachute Mitral Valve, Subaortic Stenosis, and Coarctation**

A developmental complex described by Shone and associates is characterized by four obstructive anomalies in the left side of the circulation, one of which is subaortic stenosis (fig. 12). In the region of the mitral valve, the obstructive anomalies are (1) a constricting ring in the lower aspect of the left atrium at the base of the mitral valve and (2) a deformity of the mitral valve known as a parachute mitral valve, in which there is but one papillary muscle into which all of the chordae converge. Coarctation of the aorta of the classic variety, usually associated with a bicuspid aortic valve, is a third component of the complex. The subaortic stenosis is of the muscular variety but may be compounded by proliferation of endocardial fibrous tissue over the obstructive anomaly. While the coarctation and obstruction of the mitral valve are readily identifiable clinically, the subaortic stenosis may be occult.

**Corrected Transposition and Anomalous Muscle Bundle**

The anatomic features of corrected transposition are generally understood. The atrioventricular valves and ventricles are inverted. Certain malformations that are peculiar to

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particular structures of the normally oriented heart occur on the opposite side of the heart if corrected transposition is present. Such malformations may be termed "inverted malformations."\textsuperscript{31}

Pertinent to a discussion of subaortic stenosis is the fact that the homologue of the obstructive anomalous muscle bundle of the right ventricle of the normally oriented heart is represented by an obstructive anomalous muscle bundle in the subaortic area of the arterial ventricle in corrected transposition.

**Ventricular Septal Defect and Subaortic Stenosis**

When ventricular septal defect and subaortic stenosis coexist, the features of the ventricular septal defect usually are dominant clinically, while the subaortic stenosis may go unrecognized.

Among cases with both anomalies there are two major groups, depending upon which of the two anomalies is proximal to the other.\textsuperscript{32}

When the ventricular septal defect is distal to the subaortic stenosis, the basis for the subaortic stenosis varies. In some cases it is of the muscular type, in others of the membranous type, and, in still others, is related to malinsertion of mitral chordae. In this combination, the systolic pressure in the left ventricle exceeds that in the right ventricle and in the great arteries. The systolic pressure is essentially equal in the latter three compartments.

The phenomenon of the ventricular septal defect being proximal to the subaortic stenosis is usually part of a developmental complex with remarkable regularity in anatomic structure among cases so involved.\textsuperscript{33} In this complex, a muscular spur protrudes from the

![Figure 11](https://circ.ahajournals.org/)

**Figure 11**

*Persistent common atrioventricular canal and its relation to subaortic stenosis. a. The complete variety of persistent common atrioventricular canal as viewed from the interior of the left ventricle (L.V.) and of the aorta (A.). Characteristic for this malformation is deficiency of the ventricular septum (V.S.) in the subaortic area. In this particular case, the anterior mitral leaflet (A.M.) is adherent to the deficient ventricular septum through the chordae. Intervertricular spaces allow interventricular communication. b. A second case in which the malformation was repaired surgically. Suturing of the anterior mitral leaflet (A.M.) directly to the posterior edge of the deficient ventricular septum (V.S.) results in considerable crowding of the subaortic area. In some instances with persistent common atrioventricular canal the mitral valve is adherent naturally to the ventricular septum in the manner obtained surgically in this instance. Under those circumstances, subaortic stenosis may be an intrinsic part of the malformation while, in the instance shown here, the subaortic stenosis was induced by the surgical procedure.*
Lesions causing obstruction to the outflow of blood from the left ventricle may reside in the ascending aorta, the aortic valve, or the outflow tract of the left ventricle.

Conditions involving the ascending aorta are usually congenital, and designated as supravalvular aortic stenosis. A tendency is present for this condition to be associated with mental retardation, stenosis of peripheral pulmonary arteries, stenosis of branches of the aortic arch, and a peculiar facies. Secondary narrowing of coronary arteries may occur.

Obstruction at the aortic valve is usually represented by a stenotic lesion. In some instances, the basis for stenosis is congenital; in others, it is acquired either through the direct effects of rheumatic endocarditis or through calcification of an acquired or congenital bicuspid aortic valve.

Atresia of the aortic valve causes death in infancy.

Subaortic stenosis is a complicated subject. It may be represented by a primary anomaly of the left ventricular outflow tract or may be secondary either to conditions primary in the mitral valve or to generalized diseases of the myocardium.

In some instances of subaortic stenosis, this process is one part of a complex cardiac malformation.

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LEFT VENTRICULAR OUTFLOW OBSTRUCTION

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