CLINICAL PROGRESS

Congenital Aortic Stenosis

I. Clinical and Hemodynamic Findings in 100 Patients

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II. Surgical Treatment and the Results of Operation

By Andrew G. Morrow, M.D., Allan Goldblatt, M.D., and Eugene Braunwald, M.D.

All of the various forms of congenital aortic stenosis may cause severe obstruction to left ventricular outflow without the clinical symptoms of diminished cardiac reserve that are so frequent in other forms of congenital heart disease. Conversely, in many patients with mild obstruction, the clinical findings may be striking. For these reasons, the management of patients with congenital aortic stenosis is often difficult and a detailed description was considered useful of a large group of patients with a firmly established diagnosis. In the first portion of this report the clinical features, phonocardiographic, roentgenologic, electrocardiographic, and hemodynamic findings, and the interrelationship among these data in 100 patients are presented. The second portion deals with the selection of patients for operation, and also describes the surgical techniques employed, and the clinical and hemodynamic results of operation.

Selection of Patients

The 100 patients with congenital valvular or subvalvular aortic stenosis who form the basis for this report were studied consecutively at the National Heart Institute between 1954 and 1962. Patients with supravalvular aortic stenosis and hypertrophic subaortic stenosis are not included, since detailed descriptions of our experience with these lesions have already been presented. Hemodynamically significant obstruction to left ventricular outflow was proved by left heart catheterization in each of the patients. Other patients with heart murmurs and clinical findings suggestive of aortic valve disease, but without gradients, or with only trivial peak left ventricular-arterial pressure gradients (less than 10 mm. Hg), have also been excluded. In all of the 100 patients, a heart murmur had been detected before the age of 4 years, and none had a history suggestive of active rheumatic fever. In 20 of them, the coexistence of other congenital cardiovascular anomalies helped to substantiate the congenital origin of the aortic lesion. Anatomic confirmation of the diagnosis was provided in 57 instances by the findings at operation and by autopsy in two other patients. In 23 of the 25 patients aged 20 years or more, an anatomic diagnosis was established. Selective angiocardiograms further delineated the lesion in 31 patients.

The 100 patients are subdivided into three groups. In 61 patients the obstruction was
proven to be valvular. It was localized at operation in 39 patients, at postmortem examination in two patients, by selective angiocardiography in 11 patients, and by a characteristic withdrawal pressure tracing in nine patients. In the 20 patients with congenital subvalvular stenosis of the discrete membranous, or combined (tunnel) types, the site of obstruction was localized at operation in 18 patients, and by selective angiocardiography in two patients. In the third group of 19 patients the precise site of obstruction has not yet been localized.

Clinical Descriptions

Age and Sex

The patients ranged in age from 2 to 51 years. Thirty-six were under the age of 10 years, 39 were between 11 and 20 years, 12 between 21 and 30 years, and 13 were over 30 years; since a number of the clinical findings in infants with aortic stenosis differ from those observed in older children and adults, it should be emphasized that the data presented are not necessarily applicable to children in the first 2 years of life. There were 80 males and 20 females, a sex ratio comparable to that noted by others.

Associated Malformations

Associated congenital cardiovascular malformations were noted in 20 of the 100 patients (table 1). Fifteen of the 20 patients with associated lesions had valvular aortic stenosis, while in the others the obstruction was subvalvular. All five of the patients with associated coarctation of the aorta had valvular stenosis; at operation a bicuspid aortic valve was noted in four of them and a tricuspid valve in the fifth.

The relatively frequent coincidence of patent ductus arteriosus and of coarctation of the aorta with congenital aortic stenosis has been noted by others. The triad of congenital aortic stenosis, patent ductus arteriosus, and coarctation of the aorta, was not observed in our series, but it has been noted by others. Combined stenosis of the aortic and pulmonic valves is apparently less common combination of anomalies, but its existence in our patients and in other patients emphasizes the need for right heart catheterization in patients with congenital aortic stenosis.

Symptoms

The symptoms most frequently noted by patients with congenital aortic stenosis are fatigability, exertional dyspnea, angina pectoris, and syncope. Since fatigability is such a common and nonspecific symptom, it was not correlated with any of the other findings, but it was noted frequently in this series. The presence of the other three symptoms was correlated with the severity of obstruction, as estimated from the peak systolic gradient between the left ventricle and brachial artery, in order to determine the reliability with which the severity of obstruction could be predicted from the history (table 2).

Exertional dyspnea occurred in 54 of the 100 patients; it was present in all of the 45 patients in whom the pressure gradient exceeded 70 mm. Hg and occurred in only nine of the 55 patients with gradients equal to or below 70 mm. Hg. Six of these nine patients had an associated congenital cardiovascular anomaly, and the other three patients who had exertional dyspnea, but a systolic gradient under 70 mm. Hg, were adults over the age of 30 with calcific valvular stenosis. From these observations it may be concluded that a definite history of exertional dyspnea in a patient with isolated congenital aortic stenosis usually signifies at least moderately severe obstruction.
Angina pectoris was recorded in 29 of the patients. Although this symptom occurred more frequently in patients with severe obstruction, the correlation between this symptom and the gradient was not precise (table 2). In view of the difficulty of obtaining an accurate history of chest pain in children and adolescents, the relationship between the presence of this symptom and the pressure gradient was analyzed separately in adults and children. In all 13 patients above the age of 20 years who complained of angina, the gradient was between 70 and 190 mm. Hg, and the average value was 110 mm. Hg. In contrast, each of the seven patients with histories suggestive of angina pectoris, and in whom the pressure gradients were below 50 mm. Hg, was under 20 years of age. Since, in these patients, the histories were usually obtained from the parents, true angina pectoris may not have been present. It is thus apparent that in adult patients who have a history of angina, the obstruction is usually severe. On the other hand, the absence of this symptom does not preclude severe obstruction. From the present experience it would appear that, in children, a history suggestive of angina is of little aid in estimating the severity of obstruction.

Syncope on exertion had been noted by 23 of the patients. This symptom occurred in only one of the 33 patients (3 per cent) with gradients under 50 mm. Hg, in 12 of the 41 patients (30 per cent) with gradients between 50 and 100 mm. Hg, and in 10 of the 26 patients (40 per cent) with gradients exceeding 100 mm. Hg (table 2). Thus, there was a tendency for syncope to occur with greater frequency among patients with more severe degrees of obstruction, but it was present in only one patient with a gradient of only 35 mm. Hg, and was absent in 16 of the 26 patients with gradients exceeding 100 mm. Hg.

Physical Findings

On physical examination, a prominent left ventricular lift was detected in 87 of the 100 patients. The 13 exceptions were all among the 17 patients whose systolic gradients were 25 mm. Hg or less (table 2). A precordial systolic thrill was palpable in 85 patients; it was felt best over the base of the heart and was generally transmitted to the jugular notch and along the carotid arteries. All 15 patients in whom a thrill could not be felt were among the 19 patients whose gradients were 30 mm. Hg or less. The absence of a left ventricular lift or of a systolic thrill thus ordinarily indicates that obstruction is mild. An early systolic sound (ejection sound) was audible at the base of the heart in 59 of the 100 patients. There was a tendency for this sound to be heard more frequently in patients with lower than in those with higher gradients; an ejection sound was heard in 30 of

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Table 2

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<th>Gradient</th>
<th>No. patients</th>
<th>Dyspnea</th>
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*Refers to the peak systolic left ventricular brachial artery gradient.

†Refers to the total number of patients in each group.
CONGENITAL AORTIC STENOSIS

Figure 1

Phonocardiogram, indirect carotid pressure pulse and apex cardiogram of a 41-year-old man with a peak systolic gradient of 90 mm. Hg due to a stenotic bicuspid aortic valve. At operation there was moderate dilatation of the aorta and the left leaflet of the aortic valve contained a large mass of calcific debris that rendered it completely immobile. Prominent ejection sound (E), third (S₃) and fourth (S₄) heart sounds were recorded at the apex. Indirect carotid pressure pulse tracing showed a slow anacrotic rise and plateau. The apex cardiogram revealed a presystolic impulse synchronous with a prominent fourth heart sound, a sustained systolic impulse, and an early diastolic filling wave synchronous with S₃.

the 33 (90 per cent) patients with gradients under 50 mm. Hg, 23 of the 41 (56 per cent) patients with gradients between 50 and 100 mm. Hg, and in only six of the 26 (23 per cent) patients with gradients exceeding 100 mm. Hg.

An early, high-pitched, blowing diastolic murmur was heard along the left sternal margin in 22 of the 100 patients. It was present in eight of the 61 patients (13 per cent) with valvular stenosis, in 11 of the 20 (55 per cent) patients with subvalvular obstruction, and in three of the 19 (16 per cent) patients in whom the site of obstruction has not yet been localized. The severity of the aortic regurgitation, as judged by the direct arterial pressure pulse, was considered to be trivial in 20 of these 22 patients. The other two showed moderate widening of the arterial pulse pressure.
When a congenital malformation of the aortic valve is hemodynamically significant, the lesion is almost always pure stenosis. As noted above, in a minority of patients there is mild associated regurgitation, but with stenosis predominating in importance. Even less commonly, a congenital aortic valve deformity results in pure aortic regurgitation. A total of five such patients has been studied at the National Heart Institute, and the severity of regurgitation was mild or moderate in all. Probably the least common lesions in this spectrum are those that result in predominant regurgitation, but in which hemodynamically significant obstruction is also present. We have had the opportunity to study only one patient in this category, who is not included in the present series, which is limited to patients with pure or predominant stenosis. It must be recognized, however, that a purely stenotic congenital lesion may be modified by acquired pathologic changes. As calcification of the valve occurs it may be so deformed that it becomes incompetent. Bacterial endocarditis may also be responsible for such a combined lesion.

Phonocardiograms

Phonocardiograms were recorded in 61 of the 100 patients; 35 of them had valvular aortic stenosis, 14 had subvalvular stenosis, and in 12 the site of obstruction has not been localized. The sound recordings were obtained from the mitral, tricuspid, pulmonic, and aortic areas with a Sanborn two-channel photographic recording apparatus. The logarithmic amplifying system was employed at a paper speed of 75 mm. per second. An electrocardiographic lead, the external jugular venous pulse, the indirect carotid arterial pulse, and an apex cardiogram were recorded simultaneously with the phonocardiogram in most instances.

First Sound and Ejection Sound

The time interval from the onset of electrical depolarization of the ventricles to the mitral valve closure sound (Q-M) ranged from 40 to 70 msec. and averaged 52.8 msec., comparable to the values found in normal children. The first heart sound was followed by an ejection sound in 46 of the 61 patients; this sound was present in 31 of the 35 (88 per cent) patients with valvular aortic stenosis (fig. 1), in five of the 14 patients (36 per cent) with subvalvular stenosis (fig. 2), and in 10 of the 12 (83 per cent) patients in whom the site of obstruction is as yet undetermined. The aortic ejection sound was identified and differentiated from the sound produced by tricuspid valve closure and from the pulmonic ejection sound by its prominence at the apex, by its temporal relationships, and by its failure to show respiratory variations in intensity. The time interval between the first component of the first heart sound and the ejection sound in the 46 patients in this series in whom it was present ranged from 40 to 80 msec. and averaged 49 msec. The ejection sound tended to occur more frequently in patients with aortic dilatation; it was recorded in 11 of 19 (58 per cent) patients without detectable dilatation of the aorta on roentgenographic examination and in 35 of 42 (83 per cent) patients with poststenotic dilatation. There was no correlation between the presence of the ejection sound on the phonocardiogram and the peak systolic left ventricular-arterial pressure gradient, the left ventricular systolic pressure, or the effective aortic orifice size.

Hanceock concluded that the early systolic click which is frequently present at the cardiac apex of patients with aortic stenosis results from the opening of the aortic valve, as had been suggested previously by Wiferth and Margolies. The infrequent presence of this sound in patients with subvalvular aortic stenosis, whose valves are usually normal, and its absence in patients with calcific aortic stenosis, in whom the valves are immobile, was considered by Hanceock to be evidence supporting his view. On the other hand, Leatham and Reinhold et al. attributed the sound to sudden distention of the dilated aorta or pulmonary artery after the onset of ejection. Since the opening of the aortic valves and the onset of ejection are virtually simultaneous, it is difficult to determine the
CONGENITAL AORTIC STENOSIS

Figure 2

Top. Phonocardiogram of an 11-year-old girl with discrete subvalvular aortic stenosis and a systolic gradient of 59 mm. Hg. The mitral closure sound (S₁) is followed by a prominent ejection sound (E) that was best recorded at the apex. The murmur was best recorded at the left sternal border, but was also prominent at the apex. Bottom. The phonocardiogram and indirect carotid pressure pulse of an 8-year-old boy with a peak systolic gradient of 65 mm. Hg across the outflow tract of the left ventricle. At operation moderately severe subaortic obstruction and poststenotic dilatation were found. There was a high-frequency ejection sound (E) that followed the mitral closure sound (S₁) by an interval of 60 msec, and that was recorded best at the apex. The indirect carotid pulse demonstrated a prominent anacrotic notch and systolic vibrations.

mechanical event responsible for the production of the ejection sound in aortic stenosis.

In our observations the ejection sound followed the onset of the indirect carotid upstroke by an average of 33 msec.; the latter value had been corrected for the delay between the aortic and indirect carotid pressure pulses in each individual patient by subtracting the time interval between the aortic closure sound and the incisura on the carotid pulse tracing. The intervals were similar in patients with subvalvular and with valvular
The relationship between the $A_2 - P_2$ interval during expiration plotted along the abscissa and the increase of this interval during inspiration in normal subjects (closed circles), in patients with valvular aortic stenosis (open circles) and discrete subvalvular aortic stenosis (open squares). When $A_2$ (aortic closure) followed $P_2$ (pulmonic closure), the $A_2 - P_2$ interval was considered to be negative. All values to the left and below the broken line were outside the normal range.

Aortic stenosis. The time interval between the opening of the aortic valve, determined from the carotid pulse, and the ejection sound exceeded 20 msec. in 38 of the 46 patients in whom ejection sounds were recorded. It therefore seems unlikely that this sound originates as a result of aortic valve opening. However, the impingement of a high velocity jet on a dilated vessel may well be of importance in the genesis of the ejection sound. The lower incidence of this sound in patients with discrete subvalvular aortic stenosis than in those with valvular aortic stenosis may then be explained by the consideration that in patients with subaortic stenosis the high velocity jet does not enter a dilated vessel directly, but rather enters the subvalvular region, which is surrounded by ventricular muscle and which may not be capable of generating a high frequency sound. Similarly, the presence of a long subvalvular chamber in patients with idiopathic hypertrophic subaortic stenosis may explain the absence of ejection sounds in patients with this disease.

Systolic Murmurs

A rhomboid-shaped systolic murmur of varying intensity, duration, and configuration was recorded in each patient (figs. 1 and 2). The murmur was usually best heard between the aortic area and the apex, it radiated to the neck, and was commonly associated with a thrill. The correlation between the length of the murmur and the magnitude of the gradient was poor. The configuration of the murmur may prove to be of some help in predicting the severity of stenosis, since the peak of the murmur occurred during the last 60 per cent of ventricular systole in all of the patients in whom the gradients exceeded 75 mm. Hg; however, the converse was not always true. Neither the location, the intensity, the duration, nor the configuration of the systolic murmur was of help in differentiating
CONGENITAL AORTIC STENOSIS

Second Heart Sound

The characteristics of the second heart sound were analyzed in 60 of the patients with congenital aortic stenosis and the findings were compared with those obtained in 68 normal subjects (fig. 3). In the normal subjects the pulmonic component of the second heart sound followed the aortic component by 0 to 40 msec. during expiration. During inspiration this interval increased by 0 to 60 msec. In 29 of the 60 patients with aortic stenosis the second sound was single or closely split during expiration and during inspiration; increases in the interval were within the range found in normal subjects. The average gradient in this group of patients was 49 mm. Hg and it exceeded 70 mm. Hg in only six of them. Twenty-seven of the patients with aortic stenosis had a single second sound during expiration and no change, or a subnormal increase of the interval between its two components, during inspiration. The average gradient in this group of patients was 100 mm. Hg, and it exceeded 70 mm. Hg in 21 of them. In four patients the aortic component of the second heart sound preceded the pulmonic component during expiration, i.e., paradoxical splitting was present. All of these patients had gradients above 70 mm. Hg.

The Q-A2 interval, that is the interval between the onset of electrical depolarization and the aortic closure sound, was measured during expiration in 61 patients with congenital aortic stenosis, and these time intervals were compared with the values obtained from 67 normal subjects (fig. 4). As expected, there was an inverse relationship between the duration of this interval and the heart rate.

![Graph showing relationship between heart rate and Q-A2 interval in normal subjects and in patients with congenital aortic stenosis.](http://circ.ahajournals.org/)

**Figure 4**

Relationship between heart rate and the interval between the Q wave of the electrocardiogram and the aortic closure sound (Q-A2) during expiration in normal subjects (closed circles) and in patients with valvular aortic stenosis (open circles), and discrete subvalvular aortic stenosis (open squares). The broken line indicates the upper limits observed in the normal subjects.
in both groups. Although, at any given heart rate, there was considerable variation in the duration of the Q-A2 interval among the subjects within each group, 36 of the 61 patients with aortic stenosis had values above the highest values observed in normal subjects at comparable heart rates. The abnormal behavior of the second heart sound in many patients with severe aortic stenosis (fig. 3) may therefore be attributed to the prolongation of left ventricular systole, which accompanies resistance to left ventricular ejection.

**Third and Fourth Heart Sounds**

A third heart sound was recorded in 58 per cent of the patients in whom phonocardiograms were recorded (fig. 1). The presence or absence of this sound did not correlate with the magnitude of the systolic gradient, with the height of the left ventricular end-diastolic pressure, or with the mean left atrial pressure. The frequent occurrence of a third heart sound can be explained, in part, by the fact that the majority of patients were in the age group in which a physiologic, early diastolic filling sound is a common finding. A fourth (atrial) heart sound was recorded in 36 of 61 patients. Since a physiologic fourth heart sound is also frequently heard in normal children and in subjects with prolonged P-R intervals,22-24 the presence or absence of a fourth heart sound was related to the severity of stenosis only in patients above the age of 12 years who had normal atrioventricular conduction. The sound was present in 12 of 17 patients with peak systolic gradients above 70 mm. Hg, and in only one of eight patients with gradients below this level. Furthermore, in the patients above the age of 12 years, the atrial sound was generally associated with a left ventricular end-diastolic pressure above 11 mm. Hg and a left atrial a-wave peak above 13 mm. Hg.25 In many of the patients with loud fourth heart sounds a prominent presystolic apical expansion was palpable. The apex cardiograms revealed that the peak of the presystolic apical expansion was synchronous with the fourth heart sound (fig. 1). It is suggested that in the presence of marked left ventricular hypertrophy, the resistance offered by the ventricle to passive filling is increased and the contribution of atrial systole to ventricular filling is augmented. This phenomenon is manifested by an audible fourth heart sound and a prominent presystolic expansion of the apex. Thus, the relationship between the severity of obstruction and the presence of the fourth sound is probably an indirect one, reflecting hypertrophy of the left ventricle rather than obstruction per se.

**Indirect Pulse Tracings**

The external jugular venous pulse of patients with aortic stenosis frequently showed an increase amplitude of the atrial contraction (a) wave, even in the absence of pulmonary hypertension. It is likely that it is caused by reduction of the size of right ventricular cavity due to bulging of the ventricular septum, and the increased resistance to right ventricular filling that may result.26

The indirect carotid arterial tracings usually showed a prominent anacrotic shoulder or notch, a rounded or delayed peak, superimposed systolic vibrations ("carotid shudder") and a delayed incisura (figs. 1 and 2). The ejection period and the upstroke time were measured in 34 patients with valvular and in 13 patients with subvalvular aortic stenosis and they were compared with the values obtained from 25 normal subjects. Although both of these variables were markedly prolonged in patients with both types of stenosis, there was no correlation with the severity of outflow obstruction. Weissler et al.27 found close agreement in measurement of the ejection time, as determined from direct central aortic pressure pulses and from indirect carotid or subclavian pulse tracings, and demonstrated that the ejection time is prolonged in patients with aortic stenosis. A carotid shudder28 (fig. 2) was present in 70 per cent of the patients with valvular and in 92 per cent of the patients with subvalvular aortic stenosis. However, the presence of these coarse systolic vibrations did not correlate with the severity of the aortic stenosis. These

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findings are consonant with those of Smith,\textsuperscript{29} who could not find a direct relationship between the amplitude of the anacrotic vibrations and the severity of stenosis.

In summary, an ejection sound most easily audible in the aortic area, a rhomboid-shaped aortic systolic murmur, a vigorous systolic impulse in the apex cardiogram, a prominent anacrotic shoulder with superimposed systolic vibrations, and a delayed peak and incisura in the indirect carotid tracings are all characteristic findings in congenital aortic stenosis. None of these findings, however, is of significance in estimating the severity of obstruction. On the other hand, the characteristics of the second heart sound, the magnitude of the positive presystolic wave in the apex cardiogram, and the presence or absence of a fourth heart sound may be of value in such an assessment. Although the graphic technics described above permit the recording of these physical findings, it must be emphasized that it is possible to detect all of them at the bedside.

**Electrocardiographic Findings**

The opinion is widely held that the electrocardiogram is an extremely valuable technic in the clinical assessment of the severity of the obstruction in patients with congenital aortic stenosis.\textsuperscript{3} Taussig\textsuperscript{2} states that the electrocardiographic findings vary with the severity of the obstruction and that the electrocardiogram is frequently within normal limits when the stenosis is mild. Hancock and Fleming\textsuperscript{33} found a good correlation between the voltages in the precordial leads and the depth of T-wave inversion over the left precordium with the severity of the obstruction, as reflected in the systolic gradient or the left ventricular systolic pressure in 36 patients with acquired or congenital aortic stenosis. Spencer and associates stated that left heart catheterization is not necessary routinely, in patients with congenital aortic stenosis, because the electrocardiogram is a reliable guide to the severity of the stenosis.\textsuperscript{31} Marquis and Logan\textsuperscript{30} reported that electrocardiographic evidence of left ventricular hypertrophy had proved to be the most reliable single sign indicating severe congenital stenosis.

Detailed perusal of the literature, however, reveals some inconsistencies with the widespread point of view presented in detail above. Thus, two of the six children, reported by Braverman and Gibson,\textsuperscript{35} who died of severe congenital aortic stenosis, had normal electrocardiograms. One of the two patients, described by Kjellberg et al.,\textsuperscript{12} who died suddenly, also had a normal electrocardiogram. Reynolds and collaborators\textsuperscript{36} studied seven

\textbf{Figure 5}

Relationship between the peak systolic left ventricular-brachial artery (L.V.-B.A.) pressure gradient and the angle between the mean QRS and T-wave axes in the frontal plane. The panel on the left shows this relationship in patients under 10 years of age, while the panel on the right shows this relationship in patients over 10 years of age.
patients with hemodynamic evidence of severe aortic stenosis in whom definite electrocardiographic evidence of left ventricular hypertrophy and abnormalities of the ST segments and T waves were not evident. Baker and Somerville\(^{37}\) believed that assessment of the severity of obstruction, by left heart pressure measurements, is justified in children with congenital aortic stenosis even when the electrocardiogram is normal, presumably because severe stenosis is compatible with a normal tracing. Hugenholtz and associates\(^{38}\) reported that of their patients with severe congenital aortic stenosis, only three fourths had electrocardiographic evidence of left ventricular hypertrophy, and that one third did not show abnormalities of the ST segments and T waves. On the other hand, one fourth of their patients with mild stenosis also exhibited electrocardiographic voltages of left ventricular hypertrophy, and one eighth showed ST-segment and T-wave abnormalities. In view of these divergent opinions, a detailed analysis of the electrocardiograms in our 100 patients was carried out and the findings were correlated with the hemodynamic data.

The mean electrical axis of QRS in the frontal plane ranged between \(-20^\circ\) and \(+120^\circ\). There was no correlation between this angle and the peak systolic pressure gradient. Nine of the 100 patients had left axis deviation with a mean QRS angle between \(0^\circ\) and \(-20^\circ\), while two patients had right axis de-
CONGENITAL AORTIC STENOSIS

ECG—HEMODYNAMIC CORRELATION IN CONGENITAL AORTIC STENOSIS

Figure 7

Relationship between the depth of the S wave in precordial lead V1 and the systolic pressure gradient (top) and the left ventricular systolic pressure (bottom).

viation. The magnitude of the systolic pressure gradient did not correlate with the presence of left or right axis deviation. Correlation of the mean T-wave angle in the frontal plane with the systolic pressure gradient revealed that in all five children under the age of 10 years in whom the gradient exceeded 100 mm. Hg this angle was to the left of −40°. The T angle was between −40° and −160° in eight of the 36 patients under 10 years of age; the gradient exceeded 65 mm. Hg in seven of these eight patients. No correlation was observed between the direction of the T wave in the frontal plane and the systolic pressure gradient in the patients above the age of 10 years.

The angle between the mean QRS and T forces in the frontal plane showed some correlation with the systolic pressure gradient only in patients below the age of 10 years (fig. 5). In all five patients with gradients of 100 mm. Hg or greater the angle exceeded 100°, while the 14 patients in this age group whose gradients were under 40 mm. Hg all had QRS-T angles of less than 70°. However, as with the direction of the T wave, in patients above the age of 10 years there was no correlation between the QRS-T angle in the frontal plane and the gradient.

When the peak systolic gradient was plotted against the height of the R wave in V5 in patients below the age of 10 years, a moderately good correlation between these two variables was evident (fig. 6). Six of the
36 patients under the age of 10 years had R waves less than 20 mm., and in all of them the obstruction was relatively mild, the peak gradient being less than 50 mm. Hg. However, there was a significant number of patients with equally mild obstruction in whom the R wave was markedly elevated, between 30 and 40 mm. There was no correlation whatsoever between the R wave in V₅ and the peak systolic gradient in the 64 patients who were 10 years or over (fig. 6). It was considered that the hemodynamic burden on the left ventricle might be reflected more precisely by the peak left ventricular systolic pressure than by the systolic pressure gradient and, therefore, the peak systolic pressure was related to the height of the R wave in V₅. The results were similar to those observed when RV₅ and the gradient were related (fig. 6).

A moderately good correlation was noted between the depth of the S wave in V₅ and the systolic gradient in the patients below the age of 10 years (fig. 7). In this group there were nine patients with S waves less than 16 mm., and mild obstruction (gradient under 50 mm. Hg) was present in all of them. Although all of the patients with more severe obstruction had S waves that were deeper than 16 mm., a number of patients with mild obstruction also had such deep S waves. In the patients under 10 years the relationship between SV₁ and the peak left ventricular systolic pressure (fig. 7) was similar to that observed between SV₁ and the systolic gradient. However, no correlation at all between SV₁ and the gradient or the left ventricular systolic pressure was noted in patients 10 years of age or over (fig. 7).

An attempt was then made to determine whether the R/S ratio, in either leads V₁ or V₅, would be more helpful in predicting the severity of obstruction than the R or S waves in these leads taken individually. The relationship between R/S in V₁ and the peak systolic gradient is shown in figure 8. Al-
though an R/S under 0.40 was noted in patients with all degrees of obstruction, all 11 patients in whom this ratio equaled 0.40 or more had a gradient less than 75 mm. Hg. There was no apparent difference between the relationship of this ratio with the gradient in the patients of various age groups. No correlation whatsoever was noted in any age group between either the gradient or the left ventricular systolic pressure and the R/S ratio in V5 or the sum of RV5 and SV1.

Many observers consider that abnormalities of the T waves in the precordial leads are helpful in assessing patients with aortic stenosis. The amplitude and polarity of the T waves were correlated with the peak systolic gradient. Since associated lesions or the administration of digitalis, or both, might be expected to modify this relationship, patients in these special categories are distinguished by different symbols in figure 9. There was only gross correlation between the T wave in the left precordial leads and the gradient; although upright T waves were seen in patients with gradients up to 110 mm. Hg, no patient with a gradient in excess of this value had an upright T wave. On the other hand, inverted T waves occurred in patients with gradients ranging between 20 and 200 mm. Hg. However, no patient with mild aortic stenosis (gradient below 40 mm. Hg), who did not have an associated lesion and was not receiving digitalis, had an inverted T wave lower than -2 mm. (fig. 9).

In summary, no single electrocardiographic finding is a reliable index of the severity of obstruction in congenital aortic stenosis. However, in patients under the age of 10 years the electrocardiogram is distinctly more helpful than in older patients. In this younger age group severe obstruction was associated with T-wave angles to the left of $-40^\circ$, and with QRS-T angles exceeding $100^\circ$. The height of the R wave in V5 and the depth of the S wave in V1 both correlated to some extent with the systolic gradient and the peak systolic pressure. Analysis of the electrocardiographic voltages in the precordial leads is potentially helpful in identifying some of the patients under 10 years of age with relatively
mild degrees of obstruction. An RV<sub>5</sub> < 20 mm., an SV<sub>1</sub> < 16 mm., and an R/S V<sub>1</sub> < 0.40 all suggest that the obstruction is relatively mild. On the other hand, a significant number of patients with low gradients did not exhibit these particular electrocardiographic findings. The potential hazards of relying primarily on the electrocardiogram in the management of patients with congenital aortic stenosis are illustrated in the tracings shown in figures 10 and 11.

Roentgenographic Findings

The over-all size of the heart in the patients with congenital valvular aortic stenosis was considered to be normal or the degree of enlargement to be minimal in the majority of patients (fig. 12). These findings are consonant with the impressions of Klatte et al. and of Campbell and Kauntze. However, it was observed that 75 per cent of the patients with subvalvular aortic stenosis showed mild or moderate cardiac enlargement, whereas this finding was observed in only 39 per cent of the patients with valvular obstruction. Marked cardiac enlargement was not noted in the series. There was no correlation between the general heart size and the left ventricular arteriolar pressure gradient.

The left ventricle showed some degree of enlargement in the majority of patients with either valvular or subvalvular obstruction (fig. 12). No correlation was present between the roentgenographic size of the left ventricle and the effective orifice size, the left ventricular systolic pressure or the peak systolic gradient (fig. 13). The difficulty in predicting the severity of obstruction from plain roentgenograms is illustrated by the films reproduced in figure 14, which were obtained from two patients with congenital valvular stenosis; the patient whose film is shown in figure 14A had a peak systolic gradient of 150 mm. Hg, while the patient whose film is shown in figure 14B had a gradient of only 40 mm. Hg. In contrast, the patient whose film is reproduced in figure 15A had a peak aortic systolic

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**Figure 10**

Electrocardiogram showing left ventricular hypertrophy by voltage criteria (SV<sub>2</sub> = 36 mm., RV<sub>5</sub> = 32 mm.) and simultaneously recorded left ventricular and brachial arterial pressure pulses in a child with minimal obstruction and a gradient of 10 mm. Hg.
gradient of 160 mm. Hg whereas the one whose film is shown in figure 15B had a gradient of 59 mm. Hg.

It is now well recognized that the ascending aorta is usually dilated in congenital valvular aortic stenosis.9, 40, 41 It has been the impression of some observers that the finding of poststenotic dilatation of the aorta strongly suggests that the obstruction is valvular.42

In this series, aortic dilatation was noted in the majority of patients with subvalvular, as well as in those with valvular obstruction (fig. 16). However, it was more marked in those with a valvular than in those with a subvalvular form of stenosis; 42 per cent of the former had moderate or marked dilatation, whereas only 6 per cent of the patients with subvalvular obstruction had this degree of dilatation (fig. 16). No correlation existed between either the presence or the degree of aortic tortuosity or dilatation and the peak systolic gradient or the effective orifice size.

Slight enlargement of the left atrium was noted in seven of 57 (12 per cent) patients with valvular stenosis and in one of 16 (6 per cent) patients with subvalvular obstruction. Among these eight patients with left atrial enlargement only one had a peak aortic systolic gradient less than 90 mm. Hg. Thus, radiologic evidence of enlargement of the left atrium occurs in only a small percentage of patients with congenital aortic valvular and subvalvular stenosis, but its presence suggests that obstruction is severe.

Calcification in the region of the aortic valve was noted on roentgenographic or fluoroscopic examination in 11 of 57 (19 per cent) patients with valvular stenosis, but was not observed in any of the patients with subvalvular obstruction. As shown in figure 17, calcification occurred predominantly in the older patients; it occurred in eight of 11 patients above the age of 30 years, and in only three of the 46 patients below this age. Thus, the occurrence of calcification in the patients with valvular stenosis appears to be a function of the duration of the disease, a finding in agreement with that of other observers.39, 40, 44

No relationship existed between the presence of calcification of the aortic valve and the severity of stenosis.
Fig. 12

Incidence of varying degrees of heart size (top) and left ventricular size (bottom) in patients with isolated congenital aortic stenosis.
CONGENITAL AORTIC STENOSIS

Hemodynamic Findings

Pulmonary Artery Pressure

At right heart catheterization, which was carried out in all patients, 16 were found to have an elevation of the pulmonary artery pressure, i.e., a systolic pressure exceeding 30 mm. Hg, and a mean pulmonary artery pressure exceeding 20 mm. Hg. Pulmonary hypertension was relatively mild in 12 of these patients, the systolic pressure ranging between 30 and 45 mm. Hg. Seven of the 16 patients had an associated lesion, generally one resulting in a left-to-right shunt. These seven patients were all under 16 years of age and the aortic obstruction was usually of moderate severity; the systolic gradients averaged 65 mm. Hg, and exceeded 100 mm. Hg in only one patient. On the other hand, the gradients tended to be higher and averaged 100 mm. Hg in the nine patients with pure aortic stenosis and pulmonary hypertension. It may therefore be concluded that pulmonary hypertension is found relatively infrequently in patients with congenital aortic stenosis, occurring in 16 per cent of them in this series and in only 11 per cent of the patients with pure stenosis. Its presence suggests that an associated left-to-right shunt may be present and in patients with pure aortic stenosis it indicates that the obstruction is severe.

Left Heart Catheterization and Angiocardiography

The patients described in this report were studied during a 7-year period in which a number of different methods of left heart catheterization were employed at the National
Heart Institute. Transbronchial left heart catheterization was employed in 19 studies,\textsuperscript{45} percutaneous left ventricular puncture in 34,\textsuperscript{46} retrograde left ventricular catheterization in 15,\textsuperscript{47} and transseptal left heart catheterization in 42 studies.\textsuperscript{48} A total of 110 left heart catheterizations were performed in the 100 patients.

At the present time transseptal left heart catheterization is utilized almost exclusively in the initial study of patients who are considered, on clinical grounds, to have obstruction to left ventricular outflow. By means of percutaneous puncture of the femoral vein, right heart catheterization is first carried out in order to determine whether or not an associated lesion involving the right side of the heart is present, and to measure the pulmonary artery pressure. By the same approach, transseptal puncture is then performed and left atrial and ventricular pressures are measured simultaneously with the brachial artery pressure. Cardiac output is determined by the indicator-dilution technique, and the systemic arterial pressure response to ventricular premature contractions is recorded. Anal-
CONGENITAL AORTIC STENOSIS

AORTIC DILATATION IN CONGENITAL AORTIC STENOSIS

Figure 16

Presence and degree of aortic dilatation (as estimated roentgenographically) in patients with congenital aortic stenosis.

AGE DISTRIBUTION; CONGENITAL VALVULAR AND SUBVALVULAR AORTIC STENOSIS

Figure 17

Incidence of calcification in the region of the aortic valve, demonstrated radiologically, in patients with congenital aortic stenosis in various age groups.

ysis of this response has been found to be helpful in determining the presence or absence of hypertrophic subaortic stenosis, and the arrhythmia must often be induced purposely by moving the catheter within the left ventricular cavity. If the systolic pressure gradient between the left ventricular cavity and the systemic artery exceeds 50 mm. Hg, and there are no obvious clinical or hemodynamic contraindications to operation, contrast medium is injected through the transseptal catheter into the left ventricle and either a biplane angiocardiogram in the frontal and lateral projections, or a cine-angiocardiogram in the left lateral or left anterior oblique projection is then exposed. Such angiocardiographic studies permit the delineation of the precise site of obstruction.

While the advantages of the percutaneous transseptal technic have been described previously, the major drawback of the method in the study of patients with obstruction to
Transseptal selective angiograms with left ventricular injections in two patients with valvular stenosis. On the left, the systolic jet is indicated as “J”. The oblique arrows indicate the narrowed orifice. In the film on the right the thickened valve leaflets are indicated by the arrow and moderately severe poststenotic dilatation of the aorta (Ao) is evident. The left ventricular cavity (LV) is small. In both films there is no evidence of subvalvular narrowing.

Selective angiogram in a patient with discrete subvalvular stenosis (SVS) (denoted by arrows). The left ventricular cavity (LV) is small and there is reflux of dye into an enlarged left atrium (LA). The aortic valve (AoV) is normal, and the right coronary artery (RCA) is visualized.

left ventricular outflow is the inability to record a continuous tracing as the catheter is withdrawn across the site of stenosis. However, in our experience, left ventricular pressure can be measured more easily in patients with aortic stenosis by the transseptal technic than by any of the other methods. As already indicated, the method also provides the opportunity for performing selective left ventricular angiograms, which are helpful in delineating the site of stenosis (figs. 18 and 19). The angiogram will also distinguish discrete subvalvular from hypertrophic subaortic stenosis, outline the thickness of the left ventricular wall, determine the mobility of the valve cusps, and delineate the coronary arteries, the sinuses of Valsalva, and the ascending aorta.

Although the left side of the heart and aorta can sometimes be adequately visualized by means of intravenous angiocardiography, or following injections into the right side of the heart or the pulmonary artery, the results of such indirect studies have been dis-
appointing in this clinic. By the time the contrast material reaches the left side of the heart, it is so dilute that precise identification of the detailed anatomy of the left ventricular outflow tract and the aortic valve is impossible. Retrograde arterial catheterization of the left ventricle permits selective left ventricular angiography, but this technic is of limited value in patients with aortic stenosis, since it is frequently difficult in children to pass the catheter through the stenotic orifice. Selective left ventricular angiography performed through a needle introduced into the left ventricle by percutaneous puncture has been found to be a valuable technic in the study of patients with congenital aortic stenosis, but this approach has been associated with significant morbidity, making routine use of this procedure undesirable. The transseptal technic for left ventricular angiography appears to us to be the method of choice in the study of patients with congenital aortic stenosis at this time.

**Left Atrial Pressure**

The mean left atrial pressure was within normal limits (up to 12 mm. Hg) in 47 of the 54 patients in whom it was recorded. This pressure exceeded the upper limit of normal, and ranged between 13 and 20 mm. Hg in the other seven patients; in all seven of them the obstruction was severe and the gradients ranged from 100 to 188 mm. Hg. Conversely, however, there were 10 patients with gradients of equal magnitude and normal mean left atrial pressures. While the v wave is the tallest wave in the left atrial pressure pulse of subjects without heart disease, the a wave was more prominent than the v wave in a large percentage of the patients with congenital aortic stenosis (fig. 20). The frequency of an audible atrial sound in patients with congenital aortic stenosis has already been noted (fig. 1), and the close correlation between the presence of the fourth heart sound and a prominent a wave in the left atrial pressure pulse of the patients with aortic stenosis has been presented previously.

**Left Ventricular End-Diastolic Pressure**

It has been shown that atrial contraction is of hemodynamic importance to patients with left ventricular hypertrophy, since it elevates the left ventricular end-diastolic pressure but permits the mean left atrial pressure to remain at a lower level. It was therefore of interest to determine the frequency of elevation of the left ventricular end-diastolic pressure in these patients with aortic stenosis. Twenty-one of the 100 patients had diastolic pressures between 13 and 36 mm. Hg. Nine of the 11 patients with the highest values (exceeding 17 mm. Hg) had severe obstruction, with gradients of 90 mm. Hg or above. The other two patients with such marked elevation of the left ventricular end-diastolic pressure had relatively mild obstruction; moderate aortic regurgitation undoubtedly contributed to the elevation of the end-diastolic pressure in one of them, and myocardial hypertrophy associated with, but perhaps unrelated to, minimal aortic valvular
obstruction was apparently responsible for the markedly elevated end-diastolic pressure in the other patient. Twenty-one patients with severe obstruction and gradients exceeding 100 mm Hg had left ventricular end-diastolic pressures in the normal range (below 13 mm Hg). The normal values in these patients suggest that their left ventricular function was not impaired at rest and that their left ventricular hypertrophy had not sufficiently altered the compliance of this chamber to elevate the end-diastolic pressure to an abnormal level. The absence of severe symptoms in many patients with marked obstruction is also compatible with this hemodynamic finding.

**Cardiac Output**

Cardiac output was measured by the indicator-dilution technic at the time of left heart catheterization in 54 patients. It was observed that the cardiac output exceeded the upper limits of normal for this laboratory (3.80 L/min./M²) in 28 patients. The outputs were normal (between 2.50 and 3.80 L/min./M²) in 20 patients, and below normal in only six. There was no correlation between the peak systolic gradient and the cardiac output; patients with very high gradients had high, normal, or low outputs. However, all six patients with outputs below normal had gradients exceeding 60 mm Hg (fig. 21). These findings extend to congenital aortic stenosis, the observations of other investigators who have reported that the resting cardiac output is generally within normal limits or slightly elevated in patients with acquired aortic stenosis.

**Differentiation between Valvular and Subvalvular Stenosis**

Considerable effort has been directed toward developing reliable criteria for the preoperative differentiation of congenital valvular and discrete subvalvular obstruction. Spencer and associates have indicated that subvalvular obstruction rarely causes severe symptoms in infancy, and that, when either congestive heart failure or angina occurs in

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*Figure 21*

Relationship between cardiac index and the peak systolic gradient. The two broken horizontal lines indicate the limits of normal for the cardiac index in this laboratory.
an infant, the obstruction is generally at the valvular level.\textsuperscript{5} On the other hand, these authors have also commented upon the rare occurrence of subvalvular stenosis in adults and, in fact, did not observe this lesion in patients over 18 years of age. Brock\textsuperscript{50} has also suggested that subvalvular aortic stenosis is generally a severe lesion, death usually occurring before adult life in untreated patients; the oldest patient whom he reported was 27 years of age. We noted no difference in the age distributions between the patients with valvular and subvalvular stenosis. The oldest patient with proved discrete subvalvular stenosis was aged 44 years, and the ages of four of the 20 patients with subvalvular stenosis exceeded 18 years.

The sex ratios in patients with these two forms of obstructions have not been commented upon previously; there were 52 males and nine females (M/F = 5.8/1.0) among the patients with valvular stenosis, whereas there were 13 males and 7 females (M/F = 1.9/1.0) among the patients with subvalvular obstruction. This difference in the sex ratios was found to be statistically significant ($p < .05$).

A number of observers have expressed the opinion that the clinical findings are of little, if any, value in the differentiation of these two lesions.\textsuperscript{4, 9, 61} Taussig\textsuperscript{3} and Lees and associates,\textsuperscript{8} have commented upon the relatively greater frequency of diastolic murmurs in patients with subvalvular than in those with valvular obstruction. Lees et al.\textsuperscript{8} regard the absence of a diastolic murmur as strong evidence against the diagnosis of subaortic stenosis. The higher incidence of diastolic murmurs in patients with subvalvular than in those with valvular aortic stenosis was confirmed in our patients, but it should be noted that this murmur was absent in nine of the 20 (45 per cent) patients with subvalvular obstruction.

The presence or absence of an ejection sound has also been considered to be a helpful sign in the differentiation between the two sites of obstruction. Hancock\textsuperscript{19} commented upon the rarity of the sound in subaortic stenosis. Our experience is in general agreement with this point of view, since the ejection sound was recorded in the phonocardiograms of 88 per cent of the patients with valvular stenosis and in only 36 per cent of the patients with subvalvular obstruction.

The electrocardiogram was not found to be helpful in predicting the site of obstruction, but the chest roentgenogram showed that a moderate or marked degree of poststenotic dilatation occurred in only 7 per cent of the patients with subvalvular obstruction. As already indicated, since roentgen evidence of valvular calcification occurs only in patients with valvular stenosis and was found in eight of the 11 patients with valvular stenosis above the age of 30 years, the presence or absence of this roentgenographic sign is particularly useful in adult patients with obstruction to left ventricular outflow.

A comparison of the left ventricular-brachial artery systolic pressure gradients showed that in the patients with valvular stenosis the gradients averaged 80 mm. Hg (1 standard deviation equaled 43 mm. Hg). The gradients averaged $88 \pm 28$ mm. Hg in the patients with subvalvular stenosis. This finding would not appear to substantiate the view that the obstruction in patients with congenital subvalvular stenosis is generally more severe than in those with the valvular type.\textsuperscript{60} However, the interpretation of these values is limited by the consideration that the site of obstruction was usually delineated only in those patients in whom the pressure gradient was elevated, since the majority of patients with mild degrees of obstruction were among those in whom the site of stenosis has not been localized.

In summary, it must be concluded that the various clinical findings discussed, with the exception of evidences of valvular calcification, provide only suggestive evidence as to whether the stenosis is valvular or subvalvular. This differentiation can be made with certainty only by the analysis of pressure tracings recorded as a catheter is slowly withdrawn across the outflow tract and valve or
The operative exposure and cannulations employed in the open correction of congenital aortic stenosis during cardiopulmonary bypass. The left ventricular drainage cannula is passed percutaneously into the pericardial space from the left precordial or subcostal region. It is introduced into the ventricle through an apical stab wound and anchored with a mattress suture of heavy silk. Throughout the period that the aorta is open the operative field is flooded with 100 per cent CO₂ (15 L./min.).

by the application of selective angiography.

Congenital Aortic Stenosis II
Surgical Treatment and the Results of Operation

In 57 of the 100 patients described in the first section of this report, operative treatment was carried out. These patients were selected for operation on the basis of the severity of outflow obstruction as determined by hemodynamic study. With one exception, all had peak systolic gradients between the left ventricle and brachial artery of 50 mm. Hg or more. Selection was based on hemodynamic assessment rather than symptoms and clinical findings because of the known fact that sudden decompensation or even death may occur in a previously asymptomatic or only mildly symptomatic patient. Furthermore, the correlation presented above between symptoms, physical signs, and the severity of obstruction demonstrates that the interpretations of clinical findings are not sufficiently reliable to permit them to be utilized in the important decision as to operation. This point is illustrated by the fact that seven of the 57 patients who were operated upon were entirely asymptomatic. Of the remainder, 24 evidenced only mild fatigability or slight exertional dyspnea, while 28 had experienced the more serious symptoms of severe dyspnea, angina, syncope, or congestive heart failure. A detailed summary of the findings and operative results in the first 15 patients of the series, in whom a closed operative technic or general hypothermia was employed, has been presented previously along with a summary of the experiences in other clinics as of that date (1958). The results in these first patients indicated that safe and effective relief of congenital aortic stenosis could not be achieved in all patients during the brief period of aortotomy permitted by hypothermia. Extracorporeal circulation greatly extends the time available to the surgeon, and the advantages of an open operation during cardiopulmonary bypass have led to general adoption of this method. The discussion which follows, therefore, is limited to the 44 patients who have been operated upon at the National Heart Institute with the aid of cardiopulmonary bypass between 1958 and 1962. Two of them had had previous inadequate operations under hypothermia.

The 44 patients ranged in age from 3 to 51 years; 13 were less than 10 years of age, 21 were aged 11 to 20, and 10 were over 21. There were 30 males and 14 females. In seven of the patients preoperative study revealed the presence of another congenital cardiovascular anomaly in addition to aortic stenosis. Patent ductus arteriosus was present in three patients, ventricular septal defect in two, and in one patient coarctation of the aorta and in one partial persistent atrioventricular canal were demonstrated. Except in the patient...
CONGENITAL AORTIC STENOSIS

The operative appearance of a stenotic bicuspid valve, the most frequent anatomic lesion encountered. There are only two well-formed and properly suspended leaflets and a single fused commissure, most often anterior. The raphe in the left leaflet is seldom in continuity with a true third commissure, and an incision in this leaflet will often result in valvular incompetence. The inset shows the method by which oxygenated blood for coronary artery perfusion is diverted from the arterial return line. At a mean perfusion pressure of 60 mm. Hg each cannula delivers approximately 300 ml./min. when there is no outflow resistance. The coronary cannula accommodates itself to the direction of the artery by means of a swivel tip.

Figure 24
The four types of valvular congenital aortic stenosis encountered. The bicuspid and tricuspid types are amenable to incision of the fused commissures as indicated. In the combined ("tunnel") stenosis, obstruction results not only from thickened leaflets but also from hypoplasia of the valve ring and fibrous narrowing of the ventricular outflow tract. It is considered that stenosis due to a unicommissural valve can only be relieved by valve replacement.

Operative Methods and Findings

The operative technics described are those presently utilized at the National Heart Institute and have evolved in the course of this experience. In each of the 44 patients the aortic valve and subvalvular area were exposed during cardiopulmonary bypass. The operative approach and cannulations utilized are shown in figure 22. After complete median sternotomy, a single venous drainage cannula is passed into the right atrium and the femoral or iliac artery is cannulated for arterial return. The aorta and main pulmonary artery are freed from one another so that each may be occluded. Cardiopulmonary bypass,
at a flow rate of 2.0 L./min./M.² is then instituted, the main pulmonary artery is occluded, and the patient's esophageal temperature is reduced to 30 C. by a heat exchanger in the circuit. We have not employed deep hypothermia and circulatory arrest, as described by Gordon,⁴³ because of the additional perfusion time required and the other potential hazards of the method. While the patient is being cooled a drainage cannula is passed through the chest wall and into the left ventricle through a stab wound in its apex; the cannula is connected to the intracardiac aspirating system. The ascending aorta is then occluded, just below the origin of the innominate artery, and a vertical incision is then made in it and extended obliquely downward to the aortic annulus in the noncoronary sinus of Valsalva; this incision has been found preferable to transverse aortotomy (fig. 22). The left coronary artery, and usually the right as well, is perfused with oxygenated blood supplied through cannulae connected to side arms of the arterial return line (fig. 23). This technic, and the cannulae employed, are those described by Littlefield, Lowicki, and Muller.⁶² By this means, constant oxygenation of the myocardium is maintained and the impairment of left ventricular function which follows a period of ischemia is obviated. After completion of the specific operative procedure the left ventricle is care-

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Figure 25

A tricuspid valve, converted to a functionally bicuspid one by calcification and scarring. The left coronary leaflet is rendered mobile by debridement and the irreparable right and noncoronary leaflets are replaced with a single prosthetic one of Teflon.

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Figure 26

A bicuspid valve in which there is no fusion of the commissures but in which both leaflets are rigid and extensively calcified. They are resected with bone scissors, and calcium in the valve ring is removed with a rongeur. A series of mattress sutures is placed to anchor the prosthetic leaflets.

In 30 patients the site of stenosis was found to be the aortic valve. The most common anatomic lesion encountered by others has been a stenotic bicuspid valve⁵⁷,⁷⁴ and this malformation was found in 18 of the present patients; it is illustrated in figure 23. Most often a single well-defined fused commissure and an eccentrically placed orifice were identified. In addition, a third incomplete or rudimentary commissure, usually in the left leaflet, was frequently apparent. Eleven of the bicuspid valves, encountered in patients...
between the ages of 4 and 20 years, were not calcified, and only division of the fused commissure was necessary (fig. 24). In the other 7 patients, aged 18 to 51 years, calcification of variable degree was present and debridement as well as commissurotomy (4 patients) or partial or total valve replacement (3 patients) was required (figs. 25-27). Individual leaflets were replaced with the Teflon prostheses devised by Bahnson, it and when total replacement was required the tricuspid Teflon valve described by Muller was employed.

In 11 patients the stenotic aortic valve had three leaflets (fig. 24). In six of them (ages 9 to 20 years) no calcification was present, and only commissurotomy was carried out. In five patients (ages 18 to 43 years) calcification necessitated debridement of the valve as well. In one patient valvular aortic stenosis resulted from an unicommissural valve (fig. 24). This malformation, and its surgical significance, have been described elsewhere in detail. It is of interest that in the total series of 100 patients, two died in the hospital while awaiting operation and, at autopsy, both proved to have this unusual form of valvular stenosis. The lesion has also been encountered by Ellis and Kirklin.

The discrete form of subaortic stenosis was present in 11 patients. The membrane usually consists of thick fibrous tissue that forms a crescent-shaped ridge across the anterior two thirds of the ventricular outflow tract. It is in continuity with the aortic anulus, and the bases of the right and noncoronary aortic leaflets and the insertions of the membrane are into the aortic leaflet of the mitral valve and its annulus. While the valve leaflets are retracted with forceps or a nasal speculum, the free edge of the membrane is grasped with forceps and excised, ordinarily with a knife. Care must be taken to avoid damage to the mitral annulus and the leaflets of the aortic valve. The operative appearance of this lesion is shown in figure 28 and the method by which the obstructing membrane is excised is illustrated in figure 29. In one adult patient with discrete subaortic stenosis there was also a perforation in the left coronary valve leaflet near its base. After excision of the membrane, the perforation, which had been responsible for moderate aortic regurgitation, was closed by direct suture.

In the remaining four patients obstruction resulted from a combination of valvular and subvalvular stenosis. This lesion, included in figure 24, is characterized by thickened valve leaflets and commissures that extend into a hypoplastic valve ring and a narrowed and fibrotic outflow tract. The obstructive process is quite distinct from either the discrete membranous or hypertrophic muscular forms of subaortic stenosis, and the continuity of the areas of stenosis suggested to Spencer the term "tunnel stenosis." In three patients with tunnel stenosis, commissurotomy and multiple incisions of the subvalvular obstruction were performed and, in the fourth, total valve replacement was carried out in conjunction with the subvalvular incisions. In the first section of this report, and in subsequent analyses here, these four patients have, for the sake of convenience, been grouped with those having discrete subaortic stenosis.

**Results of Operative Treatment**

Thirty-nine of the 44 patients operated

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*Figure 27*

The first prosthetic leaflet has been placed and retracted as fixation of the second is begun (A). The final appearance of the reconstructed bicuspid valve is shown in B.
upon are living. Four of the deaths occurred among the first five patients operated upon and 38 of the 39 patients subsequently operated upon are living. Three patients died in the immediate postoperative period. One patient developed jaundice and renal failure; her death is probably attributable to an inadequate perfusion. A second patient died after prolonged postoperative bleeding and multiple transfusions. The third operative death was in the patient with a unicommissural valve. The lesion was not recognized as such at the time of operation, 1958, and an incision through the single cusp resulted in overwhelming aortic regurgitation. It would appear to us that total replacement is the only means by which stenosis due to a unicommissural valve can be relieved without the creation of serious aortic regurgitation.

There have been two deaths in the late postoperative period. In one child of 8, the first with subaortic stenosis in the series, mitral regurgitation was produced in the course of the operation. She developed heart failure, her parents refused to permit another operative procedure designed to correct her traumatic mitral lesion, and she died 7 months after operation. The other late death occurred in an adult who had had total replacement of a bicuspid valve. She was well and evidenced both symptomatic and hemodynamic improvement at study 8 months postoperatively. One month later she died suddenly, and autopsy disclosed partial detachment of one of the two prosthetic valve leaflets.

Significant complications of operation in the surviving patients were postoperative bleeding, necessitating evacuation of the hemothorax (three patients), and the occurrence of the postpericardiotomy syndrome in one patient. In two children with discrete membranous subaortic stenosis the aortic leaflet of the mitral valve was damaged in the course of resection of the membrane. In each, serious mitral regurgitation resulted but both children have done well following second
operations at which the mitral valve was repaired. In one it was only necessary to reattach the mitral leaflet where it had been torn from the annulus, but in the other child a specific traumatic lesion could not be identified and a mitral annuloplasty was carried out.

In four other patients, all with valvular stenosis, significant aortic regurgitation has followed aortic valvulotomy. In each of their valves three true commissures were thought to be present and were incised. It is likely that all these valves were truly bicuspid ones and that regurgitation resulted from incision of the false or rudimentary commissure shown in figure 23. This supposition was substantiated in one of the patients who has been operated upon again. Following the initial procedure his diastolic arterial pressure fell to 40 mm. Hg from a preoperative level of 80 mm. Hg and his heart size increased. Two months after this procedure chest x-rays showed an anterior mediastinal mass, which subsequently enlarged. An exploratory thoracotomy revealed the mass to be a benign thymoma, and following its excision the aortic valve was again exposed. At this time it was apparent that the valve was a bicuspid one and that one of the two leaflets had been inadvertently incised at the location of a false commissure. This leaflet was reapproximated by direct suture, and postoperatively the child’s arterial diastolic pressure is 70 mm. Hg. In two of the other three patients the creation of valvular incompetence as a result of incision of a false commissure was appreciated at operation and the offending incision was partially closed. In these two patients the diastolic arterial pressures are 60 and 48 mm. Hg respectively and each evidences slight cardiac enlargement. The fourth patient is asymptomatic with a diastolic arterial pressure of 45 mm. Hg but has also shown cardiac enlargement.

Of the total group of surviving patients all are asymptomatic save three. Two of these are among the patients with aortic regurgitation described above and they experience mild dyspnea and fatigability and are being maintained on digitalis. The other symptomatic patient has the tunnel form of stenosis; no reduction of his left ventricular pressure was achieved at operation, and his preoperative symptoms of dyspnea, fatigue, and angina are unchanged.

In the majority of the patients a systolic murmur is still audible at the base of the heart. In most instances, however, its intensity and duration are greatly reduced in comparison to the preoperative findings. In eight patients, all of whom had valvular stenosis, diastolic murmurs, of grades I or II/VI were audible for the first time after operation. The clinical and hemodynamic findings in these patients indicate that only trivial regurgitation is present, in contrast to the four patients with significant regurgitation described above.

In virtually all of the patients the electrocardiograms after operation have not differed strikingly from preoperative ones. A detailed analysis was made of the electrocardiograms obtained at the time of postoperative catheterization in 14 patients in whom this study was carried out 6 or more months postoperatively. In these patients the left ventricular-systemic arterial systolic pressure gradient was found to have been reduced by an average of 75 mm. Hg. In 12 of them, however, there was a change in the amplitude of the R wave in lead V5 of less than 5 mm. (fig. 30). In one patient whose gradient fell from 130 to 48 mm. Hg, R V5 decreased from 50 to 29 mm. In the remaining patient a fall in gradient from 110 to 37 mm. Hg was accompanied by an increase of R V5 from 28 to 42 mm. Similarly, comparison of preoperative and postoperative x-rays has revealed no correlation between the hemodynamic result and the size of the heart.

**Postoperative Hemodynamic Assessments**

As noted above, the severity of aortic stenosis was assessed preoperatively in every patient by means of left heart catheterization. At the conclusion of the operation, before the thoracotomy was closed, left ventricular pressure was measured by direct puncture of this
chamber and recorded simultaneously with the radial artery pressure on a multichannel oscillograph. Such intraoperative records were obtained in 41 patients, and a comparison of the preoperative and operative systolic pressure gradients is furnished in figure 31. A substantial immediate reduction in the gradient was achieved in every patient and in 25 of the 41 the gradient fell to less than 20 mm. Hg.

In the final analysis the results of any operative procedure designed to correct aortic stenosis must be evaluated by a measurement of the extent to which left ventricular outflow obstruction has been relieved. The peak systolic gradient, determined at postoperative catheterization, is here utilized as this standard. Thirty-three of the patients have returned for such studies at intervals ranging from 2½ to 17 months after operation. The average interval between operation and postoperative study was 9 months. The reductions in the gradient noted at operation were confirmed at late postoperative study in all but one patient (fig. 31). This patient, noted above to have continuing symptoms, was the one in whom tunnel stenosis was ineffectively relieved. The gradients recorded at postoperative catheterization tended to be slightly higher than those measured in the operating room, presumably because of variations in cardiac output between the two studies. In

Figure 30

Electrocardiograms and records of left ventricular (LV) and brachial arterial (BA) pressure obtained before operation and 7 months postoperatively in a patient with valvular stenosis. Although the pressure gradient was completely abolished, the electrocardiographic evidences of left ventricular hypertrophy are unchanged. Although the arterial pulse pressure is wider postoperatively, there was no evidence of aortic regurgitation. Leads V₃ and V₄ are one half standard.
Measurements of the peak systolic pressure gradients between the left ventricle and brachial artery recorded preoperatively, at the conclusion of the operative procedure (operative), and at the time of postoperative cardiac catheterization. The solid circles designate the patients with valvular stenosis and the open circles those with subaortic and combined lesions.

only seven of the 33 patients, however, did the two postoperative gradients differ by more than 20 mm. Hg.

The postoperative studies in the 33 patients show that the systolic pressure gradient was less than 20 mm. Hg in 18 of them. A less satisfactory, but acceptable result was achieved in 10 additional patients in whom residual gradients of 25 to 50 mm. Hg were found to be present. The postoperative gradient exceeded 50 mm. Hg, the value employed in selecting operative candidates, in only five patients. Among them pure tricuspid valvular stenosis was present in one, discrete subaortic stenosis in two, and tunnel or combined stenosis in two patients. The difficulty in effectively relieving the obstruction caused by the latter lesion has already been referred to and is underscored by the result in the adult patient in this group. In her, total valve replacement and extensive incision of the subvalvular area were carried out but the gradient was only reduced from 144 mm. to 60 mm. Hg. In another patient with a poor hemodynamic result, a child with discrete subaortic stenosis who was operated upon early in the series, the obstructing membrane was only dilated, and because of her residual gradient (66 mm. Hg) another operation has been planned.

A more detailed analysis of the data reveals a definite correlation between preoperative and postoperative gradients; the hemodynamic results in the patients with the highest preoperative gradients were not so satisfactory as in those with less severe stenosis preoperatively. Thus, among the 17 patients with gradients of more than 100 mm. Hg before
operation the average postoperative gradient was 43 mm. Hg. In the 16 patients with preoperative gradients below 100 mm. Hg, the average gradient after operation was only 13 mm. Hg. Similarly, the 18 patients with postoperative gradients less than 25 mm. Hg had an average preoperative gradient of 73 mm. Hg and in only one of them did the preoperative gradient exceed 100 mm. Hg. Among the patients with postoperative gradients in excess of 25 mm. Hg the average preoperative level was 128 mm. Hg and in 13 of these 15 patients the preoperative gradient was greater than 100 mm. Hg.

All the data required for calculations of the effective orifice size (valve area) by the Gorlin formula\textsuperscript{18} were available in 10 patients both before and after operation. These areas are compared graphically in figure 32. An excellent result, indicated by an orifice size greater than 1.4 cm.\textsuperscript{2}, was achieved in five patients while a less satisfactory result was shown by the others. The left ventricular end-diastolic pressures recorded before and after operation in the 29 patients without significant aortic regurgitation are plotted in figure 33. Preoperatively the average value was 12 mm. Hg, and the pressure exceeded the upper limit of normal (12 mm. Hg) in eight patients. Postoperatively, the end-diastolic pressure was lower in virtually all patients. The average postoperative level was 8 mm. Hg, and the value was abnormal in only one patient. In all four patients in whom significant aortic regurgitation was induced at operation the ventricular end-diastolic pressure rose.

The diastolic arterial pressures recorded directly from the brachial or radial artery in all patients preoperatively, in the operating room, and at the time of postoperative study are plotted in figure 34. No consistent differences were noted in the preoperative and postoperative values. In only three patients...
Diastolic pressures recorded directly from the brachial or radial artery preoperatively, at the conclusion of operation (operative), and at the time of postoperative cardiac catheterization. The value designated by the arrow was obtained at the first postoperative catheterization in the child in whom aortic regurgitation was produced at operation and later corrected.

Some Unresolved Questions

The information provided by the present patients, and those described by others, clearly indicates that congenital aortic stenosis, in its various forms, is a malformation susceptible to early clinical recognition and precise hemodynamic definition. Intelligent selection of patients for surgical treatment is therefore possible and, in the vast majority of patients operated upon, effective relief of obstruction to left ventricular outflow can be accomplished. Several fundamental questions concerning patients with congenital aortic stenosis remain unanswered, however. After a satisfactory valvulotomy, will the residual abnormality of the valve cause it to calcify and again become stenotic with the passage of time? Will the child with mild valvular obstruction, in whom operation seems unnecessary on hemodynamic grounds, develop severe calcific aortic stenosis in later life? If so, would an early prophylactic operation forestall this pathologic progression? Finally, following the relief of severe obstruction, what is the fate of the left ventricular myocardium that has been subjected to this type of abnormal hemodynamic burden for many years?
Will hypertrophy regress, and will ventricular function, if impaired, return to normal? Only careful and continuing studies of patients with congenital aortic stenosis will provide the answers to these questions and perhaps alter our present plan of management for them.

References


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Conversing with the Patient and his Friends

To patient or to patient’s friend simple and unmistakable language should be used; veiled statements are too apt to be misinterpreted. The words must be chosen thoughtfully. The word “disease” should never be used; disease of the heart conveys at once to most patients the idea of something incurable and threatening. The language should be as untechnical as it can be made. A patient has a right to be told what ails him, if he so desires, in terms that convey an idea of the magnitude and significance of the trouble; he has no right to technical diagnostic terms, and it is very rarely wise to parade these before him. Accurate information cannot be conveyed by means of strange words, which to unacclimated ears bring unintended meanings; and for many the word once caught up becomes a matter requiring search in medical books, or a topic of debate with other patients. Thus the word “angina” should never come first from a doctor, if it is understood at all it will convey in almost all instances an ominous meaning; yet the prognosis varies up to fifteen years. The word “dropsy” is to be avoided, and the adjective “malignant” eliminated completely from the medical man’s vocabulary. The valves should not be named, nor murmurs mentioned. These names and details should be of no concern to patients. Methods of examination likewise require little or no explanation; a clear example of the unhappy effects of unnecessarily disclosing technical detail is the well-known instance of high blood-pressure readings.—Sir Thomas Lewis (Diseases of the Heart Macmillan Co. Ltd.). The Quiet Art: A Doctor’s Anthology. Compiled by Dr. Robert Coop, Edinburgh & London, E. & S. Livingstone Ltd., 1952, p. 237.
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