The Diagnosis of Congenital Subaortic Stenosis

Application of Hemodynamic Principles

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Optically recorded subclavian pulse curves obtained with a simple cup-tambour system gave graphic evidence of aortic valve mobility. Differentiation between congenital subaortic stenosis and aortic stenosis was aided by the demonstration in the former of a sharp incisura accompanied by a good aortic second sound; these two features were lacking or rudimentary in the latter. Despite marked auscultatory findings, there is usually little hemodynamic impairment in subaortic stenosis.

SURGICAL treatment of stenotic valvular lesions in requires definitive diagnosis. The clinical differentiation of aortic stenosis from congenital subaortic stenosis has always presented certain difficulties. Such factors as age of onset, history of rheumatic fever, and the quality of the second aortic heart sound have aided in the differentiation.5-9

Review of the literature indicates that subaortic stenosis is not rare. According to Abbott4 subaortic stenosis occurred as a primary lesion in 1.2 per cent of 1000 autopsied cases of congenital heart disease (as compared with an incidence of 9.2 per cent for patent ductus arteriosus).

Since Chevers' original description of subaortic stenosis in 1842 numerous case reports of the lesion with and without associated anomalies have appeared.3, 4, 8-17 The relative rarity of the isolated lesion at autopsy as compared with its clinical incidence suggests the benignity of subaortic stenosis. It is also probable that minor degrees are missed at autopsy.

The congenital nature of the lesion described by Keith is generally accepted.18, 19 Subaortic stenosis is analogous to infundibular stenosis in the right ventricle in that it probably represents an arrest of involution of the bulbus cordis. Usually a fibrous ridge or bar just below the semilunar valves extending 1 mm. to 1 cm. into the ventricular cavity, it consists of much elastic and hyalinized connective tissue covered by intact flattened endothelium. The turbulent passage of blood through the narrowed region may produce some of the fibrous component of the lesion in adults. Figure 1 demonstrates the gross appearance of the lesion in uncomplicated congenital subaortic stenosis, while in figure 2 tetralogy of Fallot associated with subaortic stenosis is shown. Note that the aortic valve itself is not found to be involved.

Grishman, Steinberg, and Sussman3 collected 23 cases of aortic and subaortic stenosis. They stressed the importance of pulse tracings and phonocardiograms but did not attempt to differentiate aortic from subaortic stenosis. Young4 presented 10 cases of subaortic stenosis discovered clinically in routine examination of 1800 soldiers. He stated that in this condition the anacrotic pulse of aortic stenosis is not present. Brown5 has attempted to differentiate between congenital aortic and congenital subaortic stenosis on the basis of clinical signs. Kiloh21 after study of 15 patients with pure aortic stenosis, all under the age of 50, stated that the lesion is probably not rheumatic and cannot be differentiated from subaortic stenosis (except possibly by angiocardiography).

Levine and Harvey5 pointed out that in subaortic stenosis there is a loud systolic murmur of greatest intensity in the aortic area associated with a thrill transmitted over a broad area. As contrasted with aortic stenosis, the aortic sound is not decreased and may be

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accentuated because the aortic valve is not involved. Friedberg described the murmur as loud and harsh in the second right intercostal space and radiating to the cervical region. He published an excellent carotid pulse curve and phonocardiogram showing the characteristic features.

![Image 1](image1.png)

**Fig. 1.** Gross autopsy findings in uncomplicated subaortic stenosis. Note fibrous ridge (arrow). A 66 year old male who died suddenly following onset of severe precordial pain. (Courtesy Dr. C. K. Friedberg and the Department of Pathology of Mount Sinai Hospital, New York.)

**GENERAL PRINCIPLES**

The pulse curve obtained from the subclavian artery retains all of the characteristics shown in the aortic pulse. In the normal, the steep rise is interrupted by an anacrotic notch or peak due to the sudden increase in velocity of ejection (fig. 3). The curve rises to a rounded peak (maximum ejection). A fall in pressure denotes reduced ejection. A sharp incisura follows (ventricular relaxation), after which one or several small vibrations are observed due to closure of the semilunar valves. During diastole another wave of longer period may be seen.

In aortic stenosis (fig. 4) the subclavian pulse curve begins with a sharp, steep ascent which continues to a level of about one-third to one-half of the height of the upstroke. It ends with a vibration similar in character to that occurring in the normal pulse, differing only in that this primary peak occurs relatively low on the ascending limb instead of at the top. After this primary vibration, the curve rises more slowly, differing from the normal in that the summit is not reached in mid-systole but near the end of systole. The gradual ascent is broken by small vibrations, irregular in amplitude and period, which is the graphic representation of the systolic murmur and thrill of aortic stenosis. On the catacrotic side the fall resembles much the ascent in gradient. The sharp incisura is either lacking or rudimentary, and there are no sharp after-vibrations since the valve rigidity prevents proper closure.

The above description of the pulse in aortic stenosis may be applied to the pulse of sub-
aortic stenosis except that the diastolic fall is more rapid and there is a sharp incisura, followed by the normal after-vibrations (fig. 5). Thus, obstruction to ejection produces the anacrotic vibrations, but the normal aortic valve closure produces vibrations caused by the sudden impact of the blood column on the valve.

In aortic stenosis the leaflets are fused and rigid and no vibrations are seen. The evidence of a faint or absent second heart sound favors the diagnosis of aortic stenosis, but a second sound may be due to a transmitted pulmonary valve closure. The subclavian pulse curve then gives definite and graphic evidence of the presence or absence of aortic valve mobility.

The application of these principles has aided us in distinguishing between aortic stenosis and congenital subaortic stenosis, and has further served as a basis in evaluating certain aspects of the hemodynamics of subaortic stenosis.

**METHODS AND MATERIALS**

In a large series of patients with acquired or congenital heart disease, optically-recorded arterial pulse tracings were obtained in those with suspected aortic valve obstruction. Subclavian and radial artery pulse tracings were obtained by means of a cup-tambour system using a Frank segment capsule and optical registration as described by Wiggers. Heart sound recordings and electrocardiograms obtained with a Sanborn Stethocardiette were superimposed.

Using the method described by Katz and Feil, the duration of isometric contraction and total...
systole were determined in selected cases. Pulse transmission time from subclavian to radial artery was measured by noting the difference in time of the upstroke in the two pulse curves.

2. Normal physical and mental development.

RESULTS

In the past two years, 10 patients with suspected congenital subaortic stenosis have been observed. All pulse curves were considered characteristic of this lesion.

Their ages ranged from 3½ to 37 years. All 10 (four females and six males) had the following features in common:

1. Heart murmur present from birth. The murmur, accompanied by a thrill, was characteristically located over aortic area and transmitted toward the great vessels.

2. Normal physical and mental development.

3. No symptoms or disability referable to the lesion.

4. Minimal or no left ventricular enlargement on cardiac fluoroscopy.

5. Blood pressure within normal limits.

The pertinent data in these cases were as follows:

Case 1. A 15 year old boy (high school football player) had a blood pressure of 116/88 and an electrocardiogram showing left ventricular hypertrophy.
Case 2. A 12 year old boy with a blood pressure reading of 110/70 showed left ventricular hypertrophy in the electrocardiogram.

Case 3. A 10 year old boy, had had resection and repair of coarctation of the aorta. Electrocardiogram showed left ventricular hypertrophy before resection of coarctation and left axis deviation after.

Case 4. A 7 year old girl had a blood pressure of 110/70 and left axis deviation in the electrocardiogram.

Case 5. A 27 year old woman had blood pressure of 120/70 and a normal electrocardiogram. Patient had had a full-term pregnancy.

Case 6. A 26 year old man with blood pressure of 130/70 showed early left ventricular hypertrophy in the electrocardiogram.

Case 7. A 37 year old male truck driver had a blood pressure reading of 130/90 and a normal electrocardiogram.

Case 8. A 29 year old woman laboratory technician had a blood pressure of 110/70 and showed early left ventricular hypertrophy in the electrocardiogram.

Case 9. A 9 year old boy had a blood pressure of 80/60 and a normal electrocardiogram.

Case 10. A 15 year old girl with a blood pressure of 105/70 had a normal electrocardiogram.

The pulse tracings and heart sounds are shown in figure 5.

Comparison of the pulse curves with the normal reveals the characteristics to be found in subaortic stenosis. The accompanying systolic murmur and definite aortic second sound associated with the incisura are considered diagnostically. In one patient (case 3) the recorded aortic second sound is of poor quality although the dicrotic notch is present.

Duration of isometric contraction, total systole, and pulse wave transmission time were determined in cases 1, 2, 4, 5, 6 and 7 (table 1).

In the six cases studied, the period of isometric contraction was well within normal limits. Total systole and time of pulse wave transmission from subclavian to radial artery were also within normal limits.

Cardiac catheterization was performed in case 1 to rule out any associated anomaly. Pressures in the pulmonaray artery, right ventricle, and right auricle were found to be normal. Cardiac output was 6.5 liters per minute. Brachial artery mean systolic pressure, directly recorded, was 89 mm. Hg. All values were within normal limits.

Hemodynamics

Certain interesting aspects of the hemodynamics of subaortic stenosis and of aortic stenosis may be inferred by liberal and approximate application of the formula derived by Gorlin.

Table 1.—Duration of Isometric Contraction, Total Systole, and Pulse-Wave Transmission Time in Six Patients with Suspected Congenital Subaortic Stenosis

<table>
<thead>
<tr>
<th>Case</th>
<th>Isometric Contraction (sec.)</th>
<th>Total Systole</th>
<th>Cycle Length</th>
<th>Transmission Time (Subclavian to Radial)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0.04</td>
<td>0.32</td>
<td>0.80</td>
<td>0.10</td>
</tr>
<tr>
<td>2</td>
<td>0.025</td>
<td>0.255</td>
<td>0.80</td>
<td>0.08</td>
</tr>
<tr>
<td>4</td>
<td>0.04</td>
<td>0.30</td>
<td>0.65</td>
<td>0.00</td>
</tr>
<tr>
<td>5</td>
<td>0.04</td>
<td>0.31</td>
<td>0.82</td>
<td>0.09</td>
</tr>
<tr>
<td>6</td>
<td>0.025</td>
<td>0.305</td>
<td>0.79</td>
<td>0.11</td>
</tr>
<tr>
<td>7</td>
<td>0.025</td>
<td>0.28</td>
<td>0.70</td>
<td>0.08</td>
</tr>
</tbody>
</table>

and Gorlin for calculation of the cross-sectional area of the aortic valve:

\[
AVA = \frac{AVF}{C \times 44.5 \sqrt{LV_{sm}} - BA_{sm}}
\]

\[
AVA = \text{aortic valve area in sq. cm.}
\]

\[
AVF = \frac{\text{aortic valve flow in cc./second}}{\text{systolic ejection period in seconds/minute}}
\]

\[
LV_{sm} = \text{left ventricular mean pressure in mm. Hg during systolic ejection}
\]

\[
BA_{sm} = \text{brachial systolic mean pressure in mm. Hg}
\]

\[
C = \text{empiric constant (although this has not been derived it is assumed to be 1 for purpose of this paper)}.
\]
The following values obtained during cardiac catheterization of case 1 may be substituted:

Cardiac output = 6.5 liters/minute
Systolic ejection = 25.2 seconds/minute

Therefore

\[ AVF = 260 \text{ cc./second} \]

By integration from directly recorded brachial pressure curve:

\[ BA_{sm} = 89 \text{ mm. Hg} \]

Substituting these values in the formula we obtain

\[ AVA = \frac{260}{44.5 \sqrt{LV_{sm}} - 89} \]

or

\[ LV_{sm} = \frac{34}{AVA^2 + 89} \]

We can now solve for \( LV_{sm} \) by substituting assumed values for \( AVA \):

Assumed \( AVA \)  \( LV_{sm} \)
4 sq. cm.  91 mm./Hg
2 sq. cm.  98 mm./Hg
1 sq. cm.  123 mm./Hg
0.5 sq. cm.  226 mm./Hg
0.25 sq. cm.  639 mm./Hg

These may be plotted:

Thus subaortic (or aortic) narrowing would be expected to produce little significant increase in the left ventricular systolic ejection mean pressure until the valve opening was less than 1 sq. cm.

**DISCUSSION**

It has been shown previously\(^{27,28}\) that the aortic orifice must be reduced to less than one-fourth of its natural size in experimental animals before systolic discharge, the blood pressure, or the pulse pressure are affected. Compensation in aortic stenosis is obtained by increased diastolic volume and intraventricular pressure which causes a more forceful systolic contraction and restoration of normal stroke output in accordance with Starling's Law.\(^{24}\) In aortic stenosis the isometric contraction period is slightly prolonged, the ejection phase and total systole are considerably prolonged, thus compensating for the reduced rate of ejection.\(^{29}\)

In this series of cases of subaortic stenosis, isometric contraction is well within normal limits, if not somewhat shortened.\(^{23}\) Total systole and pulse wave transmission time do not appear to be prolonged.

The superimposition of irregular vibrations on the ascending limb of the central pulse curve corresponds to the systolic murmur and may be present even though the valve area is adequate. It should be emphasized that a loud murmur and definite alteration of the pulse wave contour are not necessarily evidence of dynamic impairment of cardiac ejection. Presumably the outflow tract of the left ventricle would have to be narrowed to less than 1 sq. cm. before any significant load was thrown upon the left ventricle. Rarely is such a degree of subaortic stenosis found.

It has long been obvious that a ventricle with a congenital stenosis of its outflow tract is able to sustain a higher pressure than that in acquired valvular stenosis. In this laboratory we have observed systolic pressures as high as 200 mm. Hg in the right ventricle in patients with isolated congenital pulmonic stenosis with no evidence of failure. The hypertrophy associated with congenital stenosis may be considered developmental as well as compensatory so that the cardiac reserve presumably is relatively normal.

In general it may be said that in isolated congenital subaortic stenosis there may be little or no left ventricular enlargement. The electrocardiogram may be within normal limits or show left ventricular preponderance.\(^{20}\) Two of our cases (cases 1 and 2) showed a very tall R wave and terminally inverted T wave in the lateral precordial leads.
The presence of a definite incisura followed by after-vibrations gives good evidence of aortic valve closure. When a good second sound is associated with a definite incisura and when the history of a murmur since birth is obtained the diagnosis of subaortic stenosis is readily established.

The need for clear differentiation of congenital subaortic stenosis from aortic stenosis is obvious. The surgical considerations are such that aortic valvular stenosis, congenital or acquired, appears to be amenable to surgery, whereas subaortic stenosis is probably not. The prognosis of uncomplicated subaortic stenosis is much better than that of acquired valvular disease and the lesion may in fact produce little or no disability. Although in some cases of subaortic stenosis the turbulent passage of blood through the narrowed region may produce some of the fibrous component, the stenosis is not progressive as is the acquired valvular lesion. Death from congestive failure in uncomplicated subaortic stenosis occurs late in life.

Reports of subaortic stenosis combined with mitral valve disease have appeared in the French literature. It appears probable that these are cases of acquired endocarditis extending to the subaortic area. Chevers originally stated,

"The part of the orifice immediately below the valves is probably less frequently the seat of marked organic lesions than any other portion of the aortic outlet: still, it is liable to become generally rigid and contracted from inflammatory change; and one portion of its compass—the upper part of the larger part of the mitral curtain which is attached to the bases of the two aortic valves—is essentially found coated with masses of fibrinous or other deposit: this layer of fibrous structure is also apt to become hardened and rather contracted, after attacks of endocarditis, and in cases of disease affecting the left auriculoventricular orifice; in this way forming a cause of narrowing of the lower part of the aortic ostium, which I believe to be frequently overlooked."

This involvement of the aortic cusp of the mitral valve, producing a type of subaortic stenosis, may account for the anacrotic vibrations of the subclavian pulse and the basal systolic murmur occasionally present in "pure" mitral stenosis.

Summary and Conclusions

The clinical diagnosis of congenital subaortic stenosis can be made with greater certainty with the aid of optically recorded arterial pulse tracings. Subaortic stenosis is distinguished from aortic stenosis by the presence in the former of a sharp incisura accompanied by an aortic second sound of good quality. Pulse tracings obtained in 10 patients fulfilling the clinical criteria for congenital subaortic stenosis demonstrated the characteristic features. The phonocardiograms demonstrated the typical systolic murmur. In one case the aortic second sound was only fairly well heard. The 10 patients, ranging in age from 3½ to 37 years, had no symptoms referable to the congenital lesion.

The clinical frequency of this lesion as compared with the autopsy incidence suggests its benignity in the uncomplicated form. The congenital lesion probably represents an arrest of involution of the bulbus cordis, leaving a fibrous ridge below the semilunar valves. The hemodynamics appear to be such that ordinarily the left ventricle is not greatly burdened by congenital subaortic stenosis. In the absence of subacute bacterial endocarditis or associated anomalies, there is little disability and the prognosis is relatively good. Congestive failure is a late complication.

Subaortic stenosis should be distinguished from aortic stenosis in that the latter may be amenable to surgical treatment.

Acknowledgment

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References

8-24

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