Supravalvular Aortic Stenosis

By J. J. Denie, M.D., and A. P. Verheugt, M.D.

A case of supravalvular aortic stenosis is described. The diagnosis was suspected from pressure curves obtained at retrograde catheterization of the left ventricle. At operation a circular narrowing of the aorta 2 cm. above the valvular ring was seen. No attempt at surgical correction was made; the patient died at the end of the operation.

Anomalies consisting of supravalvular partial occlusion of aortic flow seem to occur very seldom.1-12 Clinically they are very difficult to distinguish from aortic valvular or subvalvular stenosis. As the surgical resection of the aortic stenosis becomes more commonplace, however, these anomalies may well be encountered more frequently.

Case Report

A murmur was first heard in 1940 by a school physician when the patient was 8 years old. At the age of 19 the boy complained of easy fatigue. In 1951 a harsh systolic murmur, grade III to IV, was heard at the second right interspace near the sternum; it was accompanied by a thrill over the cervical vessels. A diastolic murmur was not heard. A rather loud second aortic sound was noted. The electrocardiogram displayed an R wave in V2 and V3 that was much smaller than in V1 (fig. 1). The roentgenogram showed a double aortic impression in the barium-filled esophagus. A venous angiocardiogram excluded gross anomalies of the aortic arch. A diagnosis was made of aortic stenosis, possibly subvalvular.

In January 1957 serious anginal discomfort and palpitation after very little exertion appeared. The patient was admitted to our hospital ward on August 29, 1957. On examination we found a debilitated young man (25 years), with some degenerative stigmata, such as long slender fingers and toes, prognathism, and thoracic kyphosis. He was 159 cm. tall and weighed 45.5 Kg. At the second right interspace we heard a grade V harsh systolic murmur, radiating to the carotid arteries, accompanied by a very evident carotid shudder. At the third left interspace a holodiastolic decrescendo murmur was heard. The phonocardiogram showed a diamond-shaped systolic murmur, ending just before a second sound of great amplitude. The electrocardiogram showed QS complexes from V2 to V3; in V1 and V5 the pattern of left ventricular hypertrophy and strain was evident (fig. 2). A roentgenogram of the thorax showed a rounded apex, but the heart seemed rather small (fig. 3). Catheterization of the right heart revealed no shunts and showed slightly raised systolic pressures. By retrograde catheterization of the left ventricle from the right brachial artery we obtained in duplicate a very interesting tracing (fig. 4). Superimposing the aortic on the ventricular tracing, we obtained a systolic gradient of about 90 mm. Hg. The aortic tracing seemed to rule out any serious valvular insufficiency (fig. 5).

The aortic valvular area was calculated to be 0.4 cm.2, by means of the modified formula of Gorlin and Gorlin, aortic regurgitation being neglected. A diagnosis was made of aortic stenosis, probably of congenital origin. Antistreptolysin titers were repeatedly less than 100 units. Serologic tests for syphilis were negative. Although some points seemed still unexplained, we decided on surgical intervention with the aid of hypothermia.

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At thoracotomy on September 10 the surgeon noted a distinct circular narrowing of the aorta immediately distal to the valvular ring. We made pressure tracings from the left ventricle, the aorta, and the area between the aortic cusps and the stenosis (fig. 6). The last tracing shows no great systolic gradient compared with the left ventricular tracing, but a significant lowering of the diastolic pressure. As this type of stenosis seemed inoperable, the thorax was again closed. As the last skin

Fig. 2 Top, left. Electrocardiogram before surgery. QS complexes evident.

Fig. 3 Bottom, left. Roentgenogram of chest before surgical intervention. Note rounded apex. No distinct poststenotic dilatation of the aorta is seen.

Fig. 4 Top, right. Pressure curve obtained on withdrawal of the catheter (no. 5, Cournand) from the left ventricle to the aortic arch.

Fig. 5 Bottom, right. Pressure curves superimposed. Aortic valve area was calculated from area between these 2 curves.
sutures were being placed, the electrocardiogram showed a very pronounced depression of the S-T segments and the arterial pressure decreased to 40 mm. Hg systolic. The thorax was opened again rapidly whereupon ventricular tachycardia began, which rapidly changed to ventricular fibrillation that could not be corrected.

At autopsy the heart was removed in toto. The circular narrowing of the aorta was distinctly seen (fig. 7). By dividing the aorta transversely above the stenosis, the rather narrow aortic lumen came into view (fig. 8). The stenotic area measured about 0.5 cm.². The stenosis was accentuated by 2 features: a circular narrowing at the level of the insertion of the commissures and a hypertrophy of the plica which normally forms the margin of the sinus of Valsalva. The cusps themselves were of normal size. There was no fusion between the cusps, except at the very outer edge near the commissures. The right coronary artery originated normally in the right sinus of Valsalva. The left coronary artery originated in a blind pouch formed by the fusion of the free margin of the left aortic cusp with the aortic wall. This pouch communicated with the free aortic lumen via a narrow opening. The coronary arteries were otherwise normally permeable (fig. 9). The pulmonary artery and the
right ventricle were normal. The myocardium of
the left ventricle was greatly hypertrophied. Roent-
genograms of the specimen with barium contrast
medium showed very clearly the cusps and supra-
valvular stenosis. The left coronary artery was
very poorly filled (fig. 10).

On microscopic examination the aortic wall just
above the cusps showed a distinct hypertrophy of
the media, which was most pronounced at the
height of the stenosing ring. Except for this hy-
pertrophy of the media, the aortic wall in this area
showed no gross changes, in particular no leuko-
cytic infiltration or Asehoff bodies. There appeared
a slight grade of mucoid degeneration of the media
that resembled that seen in cystic medial necrosis
(fig. 11). This mucoid degeneration extended over
a short distance along the media of the coronary
arteries. At the fusion of the left cusp with the
aortic wall there were no distinct inflammatory re-
actions (fig. 12). A small atrophic muscular bundle
ran from the more ventral to the more dorsal com-
missure in the left aortic cusp a few millimeters
from the line of fusion (fig. 13). The aortic wall
3 cm. above the stenosis showed no abnormalities.
The myocardium of the left ventricle showed an
advanced stage of fibrous myocarditis.

Discussion

The origin of the lesion seems to be congeni-
tal. Clinically there are no reasons to suspect
a rheumatic etiology. There is probably a
relation between the mucoid degeneration and
the constitutional type of patient, which in
certain points resembles that of the Marfan
syndrome. The fusion of the left cusp with
the aortic wall explains the anginal com-
plaints, the electrocardiographic changes, and
the ventricular fibrillation at the end of the
surgical intervention. The aortic regurgita-
tion was also caused by this particular fea-
ture. We consider that this fusion developed
gradually and is correlated with the progres-
sive electrocardiographic changes and the
rather abrupt onset of very severe anginal
complaints. The fact that the very evident
auscultatory signs of aortic regurgitation
were not perceived at the first examination
of the patient in 1951 would also point to this
conclusion. The microscopic examination,
however, does not confirm this supposition.

The pressure curves show some interesting
peculiarities. Superimposing the aortic curve
and the left ventricular curve obtained at
operation, we see that the dicrotic notch of
the aortic curve falls distinctly by a time
interval of 0.02 second after the descending
branch of the left ventricular curve (fig. 14).
This is easy to understand when we realize
that the dicrotic notch of the aortic curve is
located on a higher pressure level than the
notch of the curve obtained between aortic
valve and stenosis. This last curve shows also
a lower diastolic level and a much steeper de-
scent than the aortic arch curve. It seems
that the stenosis minimized the aortic regurgi-
tation. When we observed these 2 features
more closely of a lower diastolic level just
after the cusps and a dicrotic notch in the
aortic curve that falls after the descending
branch of the left ventricular curve, we no-
**Table 1.—Survey of Reported Cases**

<table>
<thead>
<tr>
<th>Author and reference no.</th>
<th>Age/Sex</th>
<th>Description</th>
<th>Place of obstruction</th>
<th>Clinical findings</th>
<th>Cause of death Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Archer (1)</td>
<td>M 39</td>
<td>Band, 1.3 cm. long with free edges</td>
<td>Just above insertion of aortic cusps</td>
<td>Autopsy finding</td>
<td></td>
</tr>
<tr>
<td>Brody (2)</td>
<td>M 24</td>
<td>Band with free edges</td>
<td>Just above insertion of cusps</td>
<td>Autopsy finding</td>
<td>Patent foramen ovale, Ductus arteriosus persistent</td>
</tr>
<tr>
<td>Burry (3)</td>
<td>W 37</td>
<td>(1) Circular narrowing of aorta with (2) Two ridges</td>
<td>(1) Just above the cusps at the upper margin of sinuses of Valsalva, (2) Between commissures above left and anterior cusp</td>
<td>No murmurs recorded, Physical signs of Marfan syndrome</td>
<td>Died of congenital failure, Small apical myocardial infarction with endocardial thrombosis, Deepening of sinuses of Valsalva, Narrowing of aperture of left sinus of Valsalva</td>
</tr>
<tr>
<td>Kretz cited by Hektoen (5)</td>
<td>—</td>
<td>Two fibrous bands across lumen of aorta</td>
<td>Just above insertion of aortic cusps</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lev (6)</td>
<td></td>
<td>Circular narrowing of aorta</td>
<td>At the level of the entry of the sinuses of Valsalva</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lukasch (7)</td>
<td>M 76</td>
<td>Tendinous chord across lumen of aorta</td>
<td>2-3 cm. above aortic cusps</td>
<td>Autopsy finding</td>
<td></td>
</tr>
<tr>
<td>Mönckeberg (8)</td>
<td>M 26</td>
<td>Circular narrowing of aorta</td>
<td>1-2 cm. above valvular ring</td>
<td>Those of aortic coarctation</td>
<td>Short aortic arch, aortic coarctation, Died of rupture of aneurysma below coarctation</td>
</tr>
<tr>
<td>Nikiforoff cited by Rohrie (9)</td>
<td>Infant</td>
<td>Band with free edges in the lumen of aorta</td>
<td>Between commissure of left and noncoronary and commissure of left and right cusp</td>
<td>Autopsy finding</td>
<td></td>
</tr>
<tr>
<td>Rohrie (9)</td>
<td>2 weeks</td>
<td>Band with free edges in the lumen of aorta</td>
<td>Between commissure of left and noncoronary and commissure of left and right cusp</td>
<td>Autopsy finding</td>
<td>Died of colitis</td>
</tr>
<tr>
<td>Rosenberg cited by Rohrie (9)</td>
<td>M adult</td>
<td>Band with free edges in the lumen of aorta</td>
<td>Between commissure of left and noncoronary and commissure of left and right cusp</td>
<td>Musical systolic murmur aortic region</td>
<td></td>
</tr>
<tr>
<td>Stephen (10)</td>
<td>M 30</td>
<td>Stenosing membrane in lumen of aorta</td>
<td>Inserting at the height of the commissures, fused with lunulae of non-coronary and right cusp</td>
<td>Those of aortic stenosis and aortic regurgitation. The regurgitation was caused by fusion of the left and noncoronary cusps with the membrane</td>
<td>Abdominal blind pouch in which originated a thrombus, Died of congestive failure</td>
</tr>
<tr>
<td>Torres and Calvacanti (11)</td>
<td>M 32</td>
<td>Band with free edges across aortic lumen</td>
<td>Between commissure of left and noncoronary and commissure of left and right cusps</td>
<td>Systolic and diastolic aortic murmur</td>
<td></td>
</tr>
<tr>
<td>Torres and Calvacanti (11)</td>
<td>Stillborn</td>
<td>Band with free edges across aortic lumen</td>
<td>Between commissure of left and noncoronary and commissure of left and right cusps</td>
<td></td>
<td>Atrophy of left aortic cusp, Died of congestive failure</td>
</tr>
<tr>
<td>Weinstein (12)</td>
<td>M 50</td>
<td>Anomalous band across lumen of aorta</td>
<td>2 cm. above the cusps</td>
<td></td>
<td>Atrophy of left aortic cusp, Died of intestinal obstruction</td>
</tr>
</tbody>
</table>
SUPRAVALVULAR AORTIC STENOSIS

Fig. 13. In left aortic cusp muscular elements were found running a few millimeters from the aortic wall.

ticed that both were already present on our pressure curves obtained at catheterization (figs. 4 and 5). In analogy with subvalvular stenosis and in accord with other authors, we classify this case as supravalvular stenosis. That a supravalvular stenosis indeed may occur, is not generally known. We can roughly divide the recorded cases in 2 groups (table 1). The first comprises membranous chords (with muscular elements), or membranes that protrude in the free aortic lumen, a second group local narrowing of the aorta in the ascending part.

The cases of Nikiforoff, Rorhle, and Rosenberg (cited by Rorhle), the 2 cases of Torres and Calvacanti show the same localization of the supravalvular aortic band. This band runs from the commissure between the left and noncoronary aortic cusp and the commissure between the left and right aortic cusp. It is most remarkable that in our case upon microscopic examination a muscular band was found in the left deformed cusp which showed approximately the same localization. It seems then that in our case both of the previously described mechanisms are involved. The recorded physical signs are those of aortic valvular stenosis, mostly accompanied by those of aortic insufficiency. Sometimes the peculiar musical quality of the aortic systolic murmur is noted. Except for a loud second aortic sound, there seem to be no physical signs that permit a distinction between a supravalvular and a valvular stenosis. Curves as obtained by retrograde left ventricular catheterization should, when examined closely, contain important clues for the diagnosis of either subvalvular or supravalvular stenosis. With the aid of aortography it would probably have been possible in our case to locate the site of the stenosis more accurately.

SUMMARY

A case is described of supravalvular aortic stenosis that was suspected on clinical examination and was confirmed by surgery. The Marfan habitus of the patient correlated with the mucoid degeneration of the media found at microscopic examination. Certain peculiarities in the curves obtained at catheterization and at surgery are discussed. A review is presented of 14 cases confirmed by autopsy.

ACKNOWLEDGMENT

We wish to express our appreciation to Dr. A. Schaepkensvyn Riemst and Dr. A. Gründemann who performed the operation and allowed us to use their data. May we further thank Dr. H. van der Linden for his cooperation in providing the autopsy material.

SUMMARIO IN INTERLINGUA

Es describite un caso de stenosis aortica supravalvular. Le presentia del condition
esseva suspicite al examine clinic e confirmate al operation. Le habitus Marfan del patiente esseva in correlation con le degeneration mucoid del media, trovate al examine microscopic. Es discutite certe peculiaritates in le curvas obtenite al catheterisation e al operation. Es presentate un revista de 14 casos, omnes necropticamente confirmate.

REFERENCES


Atrial fibrillation, usually associated with some form of heart disease, may occur in individuals without evidence of cardiac or other systemic disorders. Two brothers with atrial fibrillation of 17 years' and 5 years' duration were studied. No evidence of heart disease could be detected; in addition, the son of 1 of the brothers was found to have the same condition. Despite the long duration of arrhythmia in these 2 patients, large doses of quinidine were effective in restoring the heart to normal sinus rhythm. Increasing doses of the drug were employed until single doses of 1.5 Gm. and 2.0 Gm. respectively were reached. The drug was withdrawn gradually after reversion to normal rhythm, which had persisted for 5 years and 3 years. The plan used for treatment of atrial fibrillation is that of digitalization for correction of rapid ventricular rates; heparinization to prevent embolization; and careful increasing of the quinidine dosage until revision to sinus mechanism occurs. The drug is stopped immediately if the blood pressure decreases, pulse rate increases, QRS complex widens by 50 per cent or more, or if respiratory difficulty, syncope, or blurred vision develops. A high percentage of fibrillators with normal hearts respond well to quinidine in contrast to the results obtained with such treatment in patients with mitral stenosis.

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