Saccular Aortic Aneurysm due to Aortic Valve Stenosis

By Theodore Ditchek, M.D., and Joseph J. Bookstein, M.D.

DISCRETE, non-mycotic aortic aneurysm associated with aortic stenosis is a rare entity. Only 11 cases have been reported in the literature, 10 associated with valvular lesions and one with subvalvular stenosis (table 1). That this association is not merely a coincidence has been demonstrated in several recent papers,5-9 and the pathogenesis of these aneurysms has been investigated.10-15

It is the purpose of this paper to present a unique demonstration of this rare combination of lesions and to emphasize the importance of the systolic jet as a causative factor in poststenotic aneurysm formation.

Case Report

An 18-year-old, asymptomatic, white boy was admitted to University of Michigan Hospital on December 13, 1961, for cardiac evaluation.

A heart murmur was first noted at the age of 3. At the age of 11 the patient had swelling and pain in the right foot with fever, and a diagnosis of rheumatic fever was made. He was treated with salicylates and penicillin, with disappearance of his symptoms in 3 days. The pa-

Figure 1

Posteroanterior (left) and lateral (right) chest films show cardiomegaly, configuration suggesting predominant left ventricular enlargement. The ascending aorta may be slightly prominent. On lateral view, note calcific crescent anterior to arch, subsequently found to lie within an aneurysm.
### Table 1

**Aneurysms Associated with Aortic Stenosis**

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>No. of cases</th>
<th>Age of pts.</th>
<th>Type of stenosis</th>
<th>Aneurysm location</th>
<th>Type of aneurysm</th>
<th>Cystic medial necrosis</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moore¹</td>
<td>1882</td>
<td>1</td>
<td>18</td>
<td>Congenital fibrous subaortic stenosis</td>
<td>Just below origin of innominate artery</td>
<td>Dissecting</td>
<td></td>
<td>Aneurysm dissected between layers of pericardium</td>
</tr>
<tr>
<td>Schnitker² &amp; Bayer</td>
<td>1944</td>
<td>2</td>
<td>Under 40</td>
<td>One rheumatic aortic valvular stenosis (slight), one combination of rheumatic aortic &amp; mitral valvular stenosis</td>
<td>Aorta</td>
<td>Dissecting</td>
<td></td>
<td>Cases obtained from literature</td>
</tr>
<tr>
<td>Lewes³</td>
<td>1951</td>
<td>1</td>
<td>65</td>
<td>Moderate aortic valvular stenosis</td>
<td>Ascending aorta</td>
<td>Dissecting</td>
<td>Present</td>
<td>Aneurysm rupture into pericardium</td>
</tr>
<tr>
<td>Petch⁴</td>
<td>1952</td>
<td>1</td>
<td>35</td>
<td>Probably congenital aortic valvular stenosis</td>
<td>Immediately above aortic valve</td>
<td>Dissecting</td>
<td>Present</td>
<td>Aneurysm dissected into pericardium. Sacular aneurysm, thought to be &quot;congenital.&quot; Patient did not display other features of the Marfan syndrome</td>
</tr>
<tr>
<td>McKusick⁵ et al.</td>
<td>1957</td>
<td>4</td>
<td>31</td>
<td>One (probably) congenital, others acquired aortic valvular stenosis</td>
<td>Ascending aorta starting within first few cm. of valves in all cases</td>
<td>Dissecting</td>
<td>Present</td>
<td>In one case, cystic medial necrosis present in arteries other than aorta</td>
</tr>
<tr>
<td>Heath⁶ et al.</td>
<td>1958</td>
<td>1</td>
<td>65</td>
<td>(Probably) acquired aortic valvular stenosis</td>
<td>5 cm. from base of ascending aorta</td>
<td>Dissecting</td>
<td>Present (limited to areas above valve)</td>
<td>Brother also had aortic valvular stenosis and arch showed medial necrosis, but no aneurysm present</td>
</tr>
<tr>
<td>Edwards⁷ (from unreported case of Hunt, J. C. et al.)</td>
<td>1960</td>
<td>1</td>
<td>46</td>
<td>Calcific aortic valvular stenosis</td>
<td>Ascending aorta approx. 2 cm. above aortic valve</td>
<td>Incomplete dissecting aneurysm (saccular)</td>
<td></td>
<td>Sudden interruption in aortic media characteristic of rupture seen in incomplete dissecting aneurysm</td>
</tr>
</tbody>
</table>
SACCULAR AORTIC ANEURYSM

Figure 2

Left. Retrograde aortogram. During systole, a radiolucent jet traverses the opaque column and strikes the anterior aortic wall at the mouth of a small saccular aneurysm. The catheter tip is also driven into the aneurysm with each systole. Right. Appearance during diastole. Note return of catheter tip to main aortic channel.

Patient continued to be very active, participating in competitive athletics without difficulty.

On physical examination the positive findings were limited to the cardiovascular system. The blood pressure was 94/80, pulse 80, and respiration 20. The patient was well developed and appeared well generally. The heart rate was regular, and the heart was enlarged to the anterior axillary line. A grade-VI systolic murmur was heard over the entire precordium, but loudest in the second right interspace, where a systolic thrill was easily palpable.

Chest x-ray showed an enlarged heart, with a configuration suggesting left ventricular enlargement. A crescentic rim of calcium was noted just anterior to the course of the aorta (fig. 1). The electrocardiogram showed unequivocal evidence of left ventricular hypertrophy.

The clinical diagnosis was aortic stenosis, probably congenital. Retrograde catheterization of the left ventricle from the right brachial artery was attempted, but it was not possible to pass the catheter beyond the aortic valve. The central aortic pulse showed a prolonged upstroke (0.23 second). A retrograde aortogram showed a systolic jet of nonopacified blood, 8 mm. in diameter, which appeared to strike a small saccular aneurysm of the proximal ascending aorta (fig. 2). In addition, the systolic jet appeared to thrust the catheter into the aneurysm with each systole.

The patient was operated upon on May 14, 1962, with use of extracorporeal circulation. The ascending aorta was dilated and a systolic thrill was easily felt. There was left ventricular hypertrophy. An aneurysm of approximately 3-cm. diameter was found on the anterior surface of the aorta above the right coronary artery. The aortic valve was tricuspid but stenotic with an opening of approximately 8 mm. in greatest diameter. The aneurysm was completely excised; the aortic valve was reconstructed by incising the rudimentary commissures toward the annulus.
Discussion

Although not described in earlier papers (table 1), cystic medial necrosis of the wall of the aorta has been recently recognized as a direct precursor of poststenotic dissecting aneurysm. In the “incomplete dissecting type,” described by Edwards, rupture may occur through the intima and a portion of the media, as in a typical dissecting aneurysm of the aorta. If no hematoma dissects into the aortic wall, only a zone of weakness at the site of the tear remains, and a saccular aneurysm develops. Aneurysms that develop in this manner may acquire secondary atherosclerosis and consequently may be confused with aneurysms of atherosclerotic origin. Edwards pointed out that in this type of aneurysm, in contrast to those resulting from atherosclerosis, an abrupt break in the continuity of the aortic media may be seen.

In the case presented here, only the roof of the aneurysm was available for pathologic study. On microscopic section, changes of cystic medial necrosis were not evident, but the media was very thin and atrophic, with hyaline material occupying much of the remaining thickness. The intima was thickened, and its margins were indistinct. Calcification was present within the intima, and probably extended into the very thin media. There was no evidence of an inflammatory or infectious process. While no distinct break in the aortic media was evident, the findings are similar to those illustrated by Edwards as incomplete dissecting aneurysm.

The cause of cystic medial necrosis and subsequent aneurysm formation in an area beyond a stenosis has been attributed by several authors to hemodynamic stress factors playing on the wall. Holman emphasized the effects of increasing lateral pressure in the poststenotic area, plus the repetitive effects of alternating high and low pressure eddy currents. Later studies by De Vries and van den Berg, and Robicsek have emphasized the singular importance of turbulence from eddy currents as the factor causing injury to the elastica of the media, with subsequent degeneration.

Little has been written concerning the direct effect of a repetitive jet of blood on an opposing vessel wall. It has been recognized as the cause of the intimal atheromatous “impingement plaques” of Hall and Ichikawa, and the intimal fibrotic “jet lesions” of Edwards. It has never been considered a prominent factor in poststenotic dilatation or aneurysm formation, and some investigators dismiss its effects entirely.

In the case described here, the saccular aneurysm arises abruptly from an aorta that showed little surrounding dilatation. The narrow jet of blood being forced through the stenotic valve is visualized striking the aortic wall at the very origin of the aneurysm. As if to emphasize the force and direction of this jet, the catheter tip was forced into the aneurysm with each systole. Because of the insignificant dilatation of the aorta anywhere but at the area where the jet strikes, it appears that the concentrated force of this stream of blood was the major factor in the formation of this aneurysm, perhaps through the production of an underlying medial necrosis.

It is in this “in vivo” demonstration of a factor long neglected in “in vitro” experiments that this case report assumes significance.

Summary

A case of congenital aortic stenosis with an associated discrete saccular aortic aneurysm is presented. The few previously reported cases of this combination of lesions are reviewed and the nature and mechanisms of the aneurysmal changes are discussed. The apparent etiologic importance of the systolic jet striking a localized area of vessel wall is emphasized.

References
4. Fetch, C. P.: Congenital aortic aneurysm with

Cases and Observations, Illustrative of Renal Disease Accompanied with the Secretion of Albuminous Urine
By Richard Bright—1827

I believe that our want of success in treatment, in what are considered the more recent attacks, is frequently owing to the fact, that the disease is far more advanced than we suspect, when it first becomes the object of our attention: and I am most anxious, in the present communication, to impress upon the members of our profession the insidious nature of this malady, that they may be led to watch its first approaches, with all the solicitude which they would feel on discovering the first suspicious symptoms of phthisis or of epilepsy. There is great reason to suppose that the seeds of this disease are often sown at an early period; and that intervals of apparent health produce a false security in the patient, his friends, and his medical attendants, even where apprehension has been early excited.

The first indication of the tendency to this disease is often haematuria, of a more or less decided character: this may originate from various causes, and yet may give evidence of the same tendency: scarlatina has apparently laid the foundation for the future mischief. . .

Intemperance seems its most usual source; and exposure to cold the most common cause of its development and aggravation.—Original Papers of Richard Bright on Renal Disease. Edited by A. Arnold Osman. London, Oxford University Press, 1937, pp. 93-94.
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