Congenital Aneurysm of the Left Aortic Sinus
A Rare Lesion and a Rare Cause of Coronary Insufficiency

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An aneurysm restricted to the left aortic sinus is extremely rare. Aneurysms of an aortic sinus most often involve the right (related to the right coronary artery) or the posterior (noncoronary) sinuses. The left aortic sinus may participate with the other two in aneurysmal dilatation, such as occurs in the Marfan syndrome. Described in this communication is an aneurysm that involved the territory of the left aortic sinus in a patient with a congenitally bicuspid aortic valve.

The purposes of this report are to demonstrate that an aneurysm may occur in this region and to indicate that by displacing and compressing the left coronary artery, the aneurysm was responsible for coronary insufficiency and myocardial infarction.

Report of Case

A 54-year-old woman, known to be normotensive and nondiabetic, was admitted to The Charles T. Miller Hospital with acute pulmonary edema and shock. A grade-II (on the basis of IV) systolic apical murmur was heard over the entire precordium, but was maximum in intensity at the apex. Abnormalities of the second cardiac sound or of the period of diastole were not observed. An electrocardiogram failed to show signs of acute myocardial infarction. Right bundle-branch block, first-degree atrioventricular block, and sinus tachycardia were the only electrocardiographic abnormalities. Despite the administration of vasopressor agents, digitalis, and quinidine, the patient died within 6 hours after the onset of symptoms.

Although previous health had been considered good, the above-described systolic apical murmur was known to have been present for 10 years and symptoms of angina pectoris had been experienced by the patient for 5 years.

An electrocardiogram 7 months before death revealed marked left axis deviation as the principal abnormality (fig. 1).

Thoracic roentgenograms taken 4½ years and 7 months, respectively, prior to her death revealed cardiomegaly (fig. 2). In addition, a prominence was apparent along the left cardiac border in the usual position of the left atrial appendage. The ascending aorta was moderately prominent. A superior mediastinal density, considered to represent a substernal thyroid, was also apparent. Roentgenoscopy had not been performed.

At necropsy, the findings of major interest were in the heart, where a congenital bicuspid valve and an aneurysm, in the area of the left aortic sinus, were seen (fig. 3).

The two aortic cusps were of about equal size. One was oriented posteriorly, the other anteriorly. The posterior of the two aortic cusps was longer and thicker than normal. The anterior cusp was about the same size, and the right and left coronary arteries arose above it. The ostium of the left coronary artery was at a higher level than the right. A ridge of tissue, considered to be a congenital raphé, extended along the central part of the aortic aspect of the anterior cusp. In the depths of the left side of the anterior aortic sinus was the broad mouth of an aneurysm. The mouth of the aneurysm, the upper edge of which was about 1.0 cm. below the ostium of the left coronary artery, led into a roughly spherical aneurysm measuring about 3 cm. in diameter. Presenting into the epicardium, the aneurysm bulged beyond the left side of the aorta and above the left ventricle. Anterior to it, lay the pulmonary trunk; to its left, the left atrial appendage; and behind it, the main body of the left atrium.

More distally, as the body of the aneurysm bulged upward, it pressed against the left coronary artery and its two primary branches and narrowed them. No significant atherosclerosis of the coronary arteries was present.

A histologic section through the left side of the anterior of the two aortic cusps and the underlying left ventricular wall showed a normal relationship between the cusp and the wall (fig. 4). An important finding was the absence of aortic tissue inserting into the junctional zone known as the annulus fibrosus. The medial aspect of the wall of
the aneurysm abutted against the epicardial aspect of the left ventricle. The wall was composed of nonspecific fibrous tissue. As the aneurysm was traced superiorly, it joined aortic tissue. The aortic media in the region of junction of aortic wall with the aneurysm showed isolated elastic fibers en-
meshed in fibrous tissue. No histologic evidence of inflammation was encountered. Histologic examination of the aorta in areas removed from the aneurysm revealed no abnormality.

In addition to a small, healed, apical myocardial infarct, there were extensive areas of myocardial infarction involving the lateral wall of the left ventricle. The necrotic muscle was being removed (fig. 5).

Discussion

In this case, the pathologic evidence suggests that the basis for the aneurysm involving the territory of the left aortic sinus was avulsion of the aortic media from the annulus fibrosa. In this regard, the lesion is like that in so-called congenital aneurysms of the right or the posterior aortic sinuses.1

In our patient, localized prominence of the left cardiac border as shown in roentgenograms had been known for 7 years and angina pectoris had existed for 5 years. The aneurysm is classified as a congenital aneurysm but it is unlikely that it had existed throughout the life of the patient. Prevailing evidence suggests that so-called congenital aneurysms of the aortic sinuses become acquired. The congenital element is an assumed weakness at

Figure 1

Electrocardiogram made 7 months before patient’s death. Left axis deviation is present (QRS axis in frontal plane minus 45 degrees). The disparity between the initial and terminal forces is of minor degree. Minor abnormalities of T waves and slight depression of ST segments in left precordial leads are also present.

Figure 2

Thoracic roentgenograms in frontal plane. Left. Five years before death. Cardiomegaly. At the arrow is a prominence interpreted as representing the aneurysm described. Also present are prominence of ascending aorta and of aortic arch. Unexplained shadow in superior mediastinum. Right. Seven months before death. The deformity (arrow) caused by the aortic sinus aneurysm is prominent. Increase in cardiac size over earlier examination.
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Figure 3

Upper. Congenital bicuspid aortic valve from above. The two cusps are oriented from side to side, yielding anterior and posterior sinuses. At the depths of the left two-thirds of the anterior aortic sinuses is the mouth of an aneurysm. This is centered below the ostium of the left coronary artery (L.C.). Lower. Exterior of heart and great vessels viewed from the left and slightly above.

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the junction of the aortic media and the anulus fibrosus. Although the weakened element has not been specifically identified, it is probable that it sets the stage for avulsion and secondary aneurysm formation.1, 2

The basis for congenital aortic sinus aneurysm seems to be the same, regardless of which sinus is involved. The anatomic relationship of the aneurysm is dependent upon the anatomic relationship of the particular aortic sinus that is involved. Aneurysms of either the right or the posterior sinus present into a cardiac chamber, since the greater part of the aortic valve is related to intracardiac structures.2, 3 Only a small part of the aortic valve (that related to the origin of the left coronary artery) lies exposed to the epicardium. It is in this area that the aneurysm occurred in our case.

In this case, a bicuspid aortic valve was present. In a heart with a tricuspid aortic valve, the aneurysm would have involved the left aortic sinus.

Thurnam4 is credited with the earliest description of an aortic sinus aneurysm, in 1840. He also chose to locate and name the aortic sinuses by their relationship with the coronary arteries (right and left) and the posterior (noncoronary). Less than 150 aortic sinus aneurysms have been reported since Thurnam’s paper. Aneurysm of the left aortic sinus is extremely rare.5 Sakakibara and Konno5 were able to find only four cases in the literature as aneurysm in the territory of the left aortic sinus.

Wolcher6 in 1921 reported a case of aneurysm of the territory of the left aortic sinus. A semi-diagrammatic drawing is shown in his article that demonstrates a bicuspid aortic valve in which the cusps are oriented as right and left. Aneurysms of both aortic sinuses

The body of the aortic sinus aneurysm lies along the left side of the aortic root and above the left aspect of the ventricle. Arching over the superior aspect of the aneurysm is the left coronary artery (A) and its two primary branches (arrows). Anteromedial to the aneurysm lies the pulmonary trunk (PT), posteriorly the main body of the left atrium (LA), and laterally the left atrial appendage (AA).
were present. The right aortic sinus was larger than the left, and the aneurysm of this sinus had ruptured into the pericardium which, of itself, is a very unusual complication. Walcher’s case is similar to ours in that the valve was bicuspid, probably congenital, and involved an aneurysm in the territory of the left aortic sinus. His case differed from ours in that the valve was oriented in an anteroposterior plane and the aneurysm in the territory of the right aortic sinus ruptured into the pericardium. No occlusion of the coronary artery resulted from the left aortic sinus aneurysm.

Higgins, in 1934, reported a case of aneurysm of the “left posterior” sinus of Valsalva. In his report, the aneurysm was said to have ruptured into the right atrium. This particular complication of aortic sinus aneurysm is more commonly associated with aneurysms

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**Figure 4**

*Left. A composite illustration of three photomicrographs. Along the left side of the illustration is shown the left aortic cusp (L.A.) and underlying left ventricular wall. L.V. = left ventricular cavity. A significant abnormality is absence of insertion of aortic tissue into the annulus fibrosus (situated at the junction of the valve cusp and the ventricle). The wall of the sinus aneurysm is against the epicardial aspect of the left ventricle. Dotted lines indicate position of aneurysm. In its upper lateral aspect is a low-power photomicrograph of the junction of the right wall of the aneurysm with the aortic wall. A portion of the ostium of the left coronary artery (L.C.) is shown as it arises from the aorta. Dotted lines from this ostium show course of coronary artery over outer aspect of aneurysm as was shown grossly in figure 3, Lower. Third photomicrograph (insert) taken from junction of aortic and aneurysmal tissue, which is outlined in bracket. In the upper part of this illustration several strands of aortic medial tissue are identifiable. In the lower part is shown the nonspecific fibrous wall of the aneurysm. Right. A composite illustration of two photomicrographs. In the lower part is shown the same area of the left ventricle and left aortic cusp as in Left. In the remainder is shown that part of the aorta above the bracket in Left. By joining the aortic wall to the annulus, the illustration portrays the authors' concept of the state of affairs before avulsion of the aortic media from the annulus had occurred and, in turn, served as the basis for development of the aneurysm. Ao. S. = aortic sinus.*
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that involve the territory of the right or the posterior aortic sinuses than those that involve the left aortic sinus. It is possible, therefore, that the aneurysm Higgins described was, in fact, of the posterior aortic sinus. Unfortunately, no photographs or diagrams accompany his report. In Higgins’ case serologic study for syphilis was positive; but from his description it seems likely that the aneurysm was of the so-called congenital type.

Another case of aneurysm of the left aortic sinus was that reported by Micks in 1940. He also reviewed three other cases. In his case, there was involvement of all three aortic sinuses. It was considered that the aneurysms were of congenital origin. The aneurysm of the left aortic sinus was the largest of the three. None of the aneurysms had perforated.

In 1949, Raman and Menon reported a case of congenital aneurysms of the left and the right aortic sinuses. The aneurysm of the left sinus was approximately the size of a marble and had ruptured into the epicardium, medial to the left atrial appendage. The type of aneurysm in the area of the left aortic sinus was similar to the one reported in this communication, except that there was no coronary arterial involvement. The aneurysm of the right aortic sinus bulged into the outflow tract of the right ventricle and ultimately ruptured into the left ventricle.

The phenomenon of coronary arterial narrowing, in association with aortic aneurysm, is usually indirect. Classically, it concerns patients who have a syphilitic aneurysm at a zone removed from the coronary arteries, while the ostia of these vessels are narrowed as an additional manifestation of the aortitis.

The case of Chipps bore some resemblance to our case. In that case, a syphilitic aneurysm involved the ascending aorta above the level of the aortic sinuses and the lower aspect of the aneurysm compressed significantly the left coronary artery. It will be recalled that in our case the aneurysm lay below the left coronary artery and pressed upward upon it.

In this respect, the case herein reported is similar to yet another case described by one of us (J.E.E.) and Burchell. In that case, an aneurysm was present at the base of the left ventricle, arising at the junction between the aortic and mitral valves and left ventricle (fig. 6). That aneurysm also presented into

Figure 5
Photomicrograph of lateral wall of left ventricle. Necrosis of muscle and the process of removal indicate acute myocardial infarction in a healing stage. (H & E; × 500).

Figure 6
Diagrammatic representation of two types of aneurysm causing left coronary arterial compression. Left. Illustration made from case described by Edwards and Burchell in which the aneurysm arose below the left side of the aortic valve, its cavity communicating with the left ventricle. L. C. = left coronary artery; L. V. = left ventricle. Right. In the case herein described the aneurysm arose above the left side of the aortic valve and is classified as an aortic sinus aneurysm in the territory of the left aortic sinus.

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the epicardium and, as in the current case, pressed upward against the left coronary artery. Acute myocardial infarction also resulted in that case.

Summary and Conclusions
This report concerns a 54-year-old woman in whom a bicuspid aortic valve and congenital aneurysm, in the area of the left aortic sinus, were present. This rare lesion compressed the left coronary artery and its primary branches. This resulted in coronary insufficiency manifesting as angina and myocardial infarction.

References

The Upright Posture
The simplest case in which the conditions common to most other animals have been drastically rearranged in man is the upright posture. The essential mechanics of the skeleton and of the circulation were established, and as it were tried out, in quadrupedal forms through an enormously long space of time. Upon this well-proved arrangement, and it would seem rather suddenly and without a fundamental mechanical reorganization, was imposed a posture which threw on the old scheme fresh strains that have tried it to within reach of the actual margin of safety. The results of this late and, so to say, hurried revision of the structural plan show themselves far and wide in the pathological field. It is not unreasonable to see these strains as important factors in the so-called static deformities, and in the excessive liability of the lower limbs to diseases of the bones and joints and to circulatory disorders on both the arterial and venous sides. Echoes of the same disturbing element are perhaps to be found in abdominal pathology, and even in the peculiarities of the cerebral circulation. In general, however, it is perhaps in relation to the special qualities of the circulation as a whole in man that the upright posture should arouse the most caution in applying the results of animal experiment.—The Collected Papers of Wilfred Trotter, F.R.S. London, Oxford University Press, 1946, p. 108.
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