CLINICAL PROGRESS

Clinical Manifestations of the Unperforated Aortic Sinus Aneurysm

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ANEURYSMS of the aortic sinuses (of Valsalva) are rare and are either congenital or acquired. The acquired types are chiefly due to syphilis or bacterial endocarditis. The congenital aneurysms are thought to be due to a developmental defect in either the aortico-pulmonary septum or the elastic tissue of the aortic sinuses.

Morgan Jones and Langley in 1949,1 in a classic paper, reviewed the literature; they recorded 43 autopsied cases of aortic sinus aneurysms and added 4 new ones. Venning in 19512 reported 3 additional cases, and Falholt and Thompson in 19533 described a case of unruptured congenital right aortic sinus aneurysm diagnosed by retrograde aortography and increased the number of congenital sinus of Valsalva aneurysms to 34. In 1954, Besabe and associates4 added a case of congenital aortic sinus aneurysm with rupture into the right atrium due to bacterial endocarditis, which also caused embolic gangrene of a foot.

Nineteen cases of syphilitic aneurysm of the aortic sinuses have been reported in the English literature since 1914; the antemortem diagnosis was made only once. However, the advent of contrast visualization of the aorta has made possible the recognition of aortic sinus aneurysms in 9 additional cases.4

In a 10-year period, during which over 2,500 patients were studied by intravenous angiocardiography, 19 cases of unruptured aortic sinus aneurysms have been diagnosed at this center. Seventeen cases have been reported previously and of these, 8 were congenital; 3, associated with coarctation of the aorta6; 1, with coarctation of the aorta and bacterial endocarditis7; 1 with pseudocoarctation of the aorta8; and 3 with arachnodactyly.9 The 9 acquired cases were due to syphilis: of these, 6 were associated with aneurysms of the ascending aorta; while 3 had aortitis with localization of the aortic sinuses.8

The 19 cases of unperforated aortic sinus aneurysms observed by us form the basis of this report. Experience gained from their study indicates that there are significant clinical clues that should suggest the diagnosis. Confirmation may then be had by angiocardiography.

NORMAL ANATOMY OF THE AORTIC SINUSES

The aortic sinuses are 3 small dilatations in the wall of the aorta immediately above the aortic valves. Each sinus lies just above the attachment of an aortic cusp. The right and left coronary arteries usually originate within 2 of the aortic sinuses; however, they may emerge immediately distal to the aortic sinuses. The aortic sinuses and their corresponding aortic valves are named according to the source of the coronary arteries. Thus, the right and left coronary arteries emerge from the right and left aortic sinuses, respectively. The remaining aortic sinus usually lies posteriorly, does not contain a coronary artery and, according to the newest nomenclature, is designated the non-coronary sinus.1

The aortic sinuses are intracardiac and cannot be identified on conventional roentgenography. During angiocardiography they appear as small dilatations at the root of the aorta immediately above the aortic valves, and are best visualized in the left anterior oblique projection.10 Inconstant filling of the coronary arteries during angiocardiography

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does not allow complete identification of the sinuses. However, in the left anterior oblique view, the right coronary sinus is anterior and just behind the sternum. The aortic sinuses of Valsalva are in close relation to all the cardiac chambers, particularly the right atrium and ventricle. The superior vena cava and the pulmonary artery, the left ventricle and left atrium are adjacent.

**CLINICAL FEATURES OF UNPERFORATED CONGENITAL AORTIC SINUS ANEURYSMS**

*Age, Sex, and Race.* The ages in 9 congenital unperforated aortic sinus aneurysm patients varied from 7 to 47 years. All but 2 were under 30 years; the older patients had associated arachnodactyly. Six were male and 3 were female. Only 1 was Negro.

*Associated Anomalies.* Four patients with aortic sinus aneurysms had the classical findings of coarctation of the aorta, i.e., hypertension of the upper extremities, rib notching, and absent or diminished blood pressures of the lower extremities. One of the patients with coarctation of the aorta had subacute bacterial endocarditis and aortic regurgitation. Three had the classical findings of arachnodactyly. One patient had pseudocoarctation of the aorta; and 1, the youngest, aged 7 years, also had congenital mitral and aortic stenosis with generalized hypertrophy and dilatation of the cardiac chambers.

![Fig. 1. Aneurysm of the right coronary sinus and coarctation of the aorta. Left anterior oblique angiogram of a 27-year-old woman demonstrates an enlarged left ventricle, a dilated ascending aorta with an aneurysm of the right coronary sinus (arrow), coarctation of the aorta (arrow), and enlarged brachiocephalic vessels with prominent collateral arteries. (Previously published, reproduc with the permission of the New England Journal of Medicine.)](image-url)
Symptoms. All the patients with coarctation of the aorta were symptomatic; however, the symptoms were not due to the aortic sinus aneurysm but to the associated aortic disease. Headache, dizziness, epistaxis, and Jacksonian epilepsy led to physical examination and the diagnosis of coarctation of the aorta in 3; fever and bouts of dyspnea resulted in the discovery of a complicated subacute bacterial endocarditis in the fourth patient.

Two of the patients with arachnodactyly were free of cardiovascular complaints; the third had sudden onset of chest pain at the age of 47 years, apparently due to the onset of aortic regurgitation and mild heart failure. The patient with pseudocoarctation complained of nervousness and backache, probably due to a neurotic disposition; while the ninth case, a 7-year-old child, was chronically ill with dyspnea and easy fatigability.

Murmurs. All patients with congenital aortic sinus aneurysms had heart murmurs. Common to all was a loud, rough (grade 2 to 4) systolic murmur at the apex of the heart, heard all over the precordium and especially to the left of the sternum. In 3 instances, aortic valvular lesions were also present and produced diastolic and systolic murmurs at the base of the heart.

In the 4 patients with coarctation of the aorta, there were in addition, interscapular bruits due to arterial collaterals. Resection of the coarcted segment and re-anastomosis of the aorta completely relieved the hypertension and
did away with the interscapular murmurs, but the cardiac murmurs were unaltered.

**Blood Pressure.** Systolic and diastolic hypertension was present in 3 of the patients with coarctation of the aorta; the fourth with aortic regurgitation had a systolic pressure of 190 mm. Hg but the diastolic blood pressure was 0. Two patients in this group had absent blood pressures in the legs and in 2, the lower extremity pressures were present but diminished. The patient with arachnodactyly and aortic regurgitation had an elevated pulse pressure, (140/60 mm. Hg); in the other 2 cases the blood pressure was normal.9

**Electrocardiography.** Left ventricular hypertrophy on electrocardiography was present in 2 of the 4 patients with coarctation of the aorta. Two others and the patient with pseudocoarctation had normal electrocardiograms. In the group with Marfan’s syndrome, I had a normal electrocardiographic tracing, although conventional and angiocardiographic roentgenograms showed left ventricular hypertrophy; another had partial heart block with a normal-sized left ventricle; the third had left axis deviation and left ventricular hypertrophy.9 The child with generalized cardiac chamber enlargements showed peak P waves indicative of atrial enlargement and inverted S-T and T waves suggestive of myocardial disease.

**Roentgenography of Congenital Unperforated Aortic Sinus Aneurysms**

**Conventional Roentgenography.** On fluoroscopy, unusual pulsation of the root and ascending aorta was frequent. However, the hypertension and dilated ascending aorta due to coarctation of the aorta and the aortic valve involvement in the majority of the cases limited the value of this observation.

An abnormal cardiac silhouette was present in all roentgenograms of patients with congenital aortic sinus aneurysms except in 1 instance; a patient with arachnodactyly had a normal heart. All cases with coarctation of the aorta, in addition, exhibited rib notchings and

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**Fig. 3.** Syphilitic aortitis with localized aneurysm of the aortic sinuses. Frontal roentgenogram of a 64-year-old man showing enlargement of the heart, pulmonary artery segment, and a massive calcified mass occupying most of the right upper cardiac silhouette (arrows).
an enlarged left ventricle. Evidence of poststenotic enlargement of the descending aorta was seen and verified by indentation of the esophagus in 3 instances. The patient with pseudocoarctation had a prominent shadow in the region of the aortic knob and a dilated descending aorta; however, no rib notching was recognized. Two of the patients with arachnodactyly had enlarged left ventricles. In the patient with aortic and mitral valve stenosis there was also enlargement of cardiac chambers.

Angiocardiography. Angiocardiography provided the definitive diagnosis of aortic sinus aneurysm. Generalized aneurysmal dilatation of the aortic sinuses was present in all but 1 instance. In that case (fig. 1), only the right aortic sinus was involved. In the cases of coarctation of the aorta, the point of coarctation, the dilated brachiocephalic and collateral arterial branches as well as the enlarged left ventricle were clearly outlined (fig. 1). In contrast, in the case of pseudocoarctation (fig. 2), although there was an unusually dilated left subclavian artery, an apparent point of coarctation, and a dilated descending aorta, there was no collateral arterial circulation.

In summary, the only clue to the presence of an unperforated congenital aortic sinus aneurysm is a grade 3 to 4 systolic murmur over the heart, especially to the left of the sternum. Such a murmur if associated with coarctation of the aorta, anomalies of the aortic valves or arch, or arachnodactyly should arouse suspicion of an aortic sinus aneurysm. Angiocardiography will establish the diagnosis.

Syphilitic Unperforated Aortic Sinus Aneurysms

Pathologic Aspects. Syphilitic involvement of the cardiovascular system is commonest at the

Fig. 4. Left anterior oblique angiocardiogram reveals the localized aortic sinus aneurysm (arrow) surrounded by calcified thrombus. The left ventricle is enlarged. (Previously published, reproduced with the permission of the American Journal of Medicine.)
The aortic valve and ascending aorta. It begins with weakening and destruction of the intimal surface of the aorta and elastic fibers of the media. The aortitis becomes manifest as either a diffuse aneurysmal dilatation or a localized saccular aneurysm of the ascending aorta. Calcium plaques are frequently deposited in the damaged intima; their recognition in the root and ascending aorta becomes an important clue of syphilis.\(^\text{11, 12}\) If the aneurysm is large, stasis of blood occurs and a laminated thrombus partially fills the lumen. The aortic sinuses may similarly be involved and aneurysmal dilatation or sacculation of the sinuses results. Dilatation of the aortic ring and the production of aortic insufficiency are usual.\(^\text{5}\)

Unlike congenital and mycotic aneurysms of the aortic sinuses, which are paper thin and prone to rupture,\(^\text{1}\) syphilitic aneurysms usually become very large and laminated before they perforate.\(^\text{1, 12}\) Any of the aortic sinuses may be the site of origin of a syphilitic aneurysm. This is in contrast to the congenital type, which is believed to involve chiefly the right and non-coronary sinuses.\(^\text{1}\) Contrary to reports, there is no predictable relation between the sinus of origin and the structure into which an aneurysm perforates.\(^\text{13}\) Eight of the 11 perforated cases reported in the literature (73 per cent) ruptured into the lesser circulation (pulmonary artery 4, right atrium 2, and right ventricle 2), and produced a left to right shunt.\(^\text{5}\) Syphilitic aortic sinus aneurysms, unlike the congenital, may also rupture outside the heart. This is probably

**Fig. 5.** Syphilitic saccular aneurysm of the ascending aorta with aneurysmal involvement of the aortic sinuses. Conventional lateral roentgenogram of a 53-year-old man shows enlargement of the left ventricle and a large saccular aneurysm of the ascending aorta, lined by calcium (arrow). Note that the linear calcification extends into the intracardiac portion of the aorta and clearly outlines the dilated aortic sinuses.
because the syphilitic aneurysms attain such a large size.

In the literature, the most common cause of death in patients with syphilitic aortic sinus aneurysm, even with perforation, was heart failure. Rupture in 8 cases with the establishment of aortocardiac or aorticopulmonary shunts hastened failure; none survived more than 2 months. On the other hand, rupture outside of the heart caused sudden death. In 3 instances, a coronary artery occlusion due to pressure of the adjacent aortic sinus aneurysm was present. In 1 case, myocardial infarction was found distal to the occluded left coronary artery and caused sudden death. In another, the aneurysm was intact but bulged into the interventricular septum and pulmonary conus causing pulmonary stenosis and delayed atrioventricular conduction, presumably due to pressure on the bundle of His.5

Clinical Aspects of Syphilitic Unperforated Aortic Sinus Aneurysms

Age, Sex, and Race. In our series of 10 cases of syphilitic aortic sinus aneurysms, all the aneurysms were unperforated. The ages ranged from 44 to 60 years and averaged 54 years. Men predominated; only 1 patient was a woman. Five were white and 5 Negro.

Symptoms and Signs. As a rule, prior to rupture, aortic sinus aneurysms do not by themselves produce symptoms. Rather, syphilitic involvement of the aortic valvular ring causes aortic insufficiency and heart failure. Most of our 10 patients were moderately to severely ill; all had aortic incompetence. Aortic diastolic and systolic murmurs, pounding pulses, and wide pulse pressures were usual. In 3, heart failure was present in addition. Pre-

Fig. 6. Left lateral angiocardioiagram demonstrates the aneurysm of the aortic sinuses, the dilated ascending aorta (aortitis) and the sacculated retrosternal aneurysm of the aorta. (Previously published,4 reproduced with the permission of the American Journal of Medicine.)
cordial pain probably due to coronary ostial stenosis occurred in several.

Blood Pressure. Prominent features in the reported cases and in our cases of syphilitic aortic sinus aneurysm were systolic hypertension, high pulse pressures, and low diastolic pressures, which were related to the aortic insufficiency. Two of our patients, however, had normal blood pressures.

Electrocardiography. Electrocardiographic examination in 9 living cases showed either left axis deviation or left ventricular hypertrophy. These findings reflect the enlargement of the left ventricle due to aortic regurgitation and hypertension. In 1 patient (fig. 3 and 4), there was right axis deviation and first degree heart block. In this instance compression of the pulmonary conus, pulmonary artery, and its right branch by the aneurysm was demonstrated, and is believed to have caused chronic cor pulmonale.\(^4\) In the literature, 2 cases also exhibited right axis deviation of the electrocardiogram, but this was associated with perforation of the aortic sinus aneurysm into the lesser circulation. The right axis deviation in those instances was probably due to acute cor pulmonale from the overloading and hypertension of the right heart.\(^5\)

Roentgenography of Syphilitic Unperforated Aortic Sinus Aneurysms

Conventional Roentgenography. The diagnosis of syphilitic aortic sinus aneurysm can often be made after conventional roentgenography. A large boot-shaped heart due to the large left ventricle caused by the isolated aortic insufficiency was present in all but 1 case. Pulsatile dilatation of the ascending aorta and fusiform or saccular aneurysms of the ascending aorta are other frequent findings. Linear calcification of the aorta is common and has long been recognized as an important sign of syphilitic aortitis.\(^1,\)\(^2\) When the calcification extends into the intracardiac origin of the aorta and outlines the aortic sinuses (fig. 5 and 6), or becomes massive in this region (fig. 3 and 4), the diagnosis of syphilitic aortic sinus aneurysm becomes almost a certainty.

Rarely, a patient with right heart enlargement, right axis deviation of the electrocardiogram, and calcification near the left atrium may be mistaken for rheumatic heart disease with mitral and aortic valvular involvement and left atrial calcification.\(^5,\)\(^6\) However, the presence of linear calcification in the ascending aorta should suggest the syphilitic etiology.

Angiocardiography. Aortic sinus aneurysms are readily demonstrated by angiocardiography. In the syphilitic, 2 types are recognized: in 7 instances, the aneurysms were continuations of huge fusiform or saccular aneurysms of the ascending aorta (fig. 6); in 3, the aneurysms were localized at the aortic sinuses and associated with aortitis (fig. 4). In all instances, the syphilitic aneurysms were considerably larger than the congenital types and, therefore, created pressure effects in the surrounding cardiac chambers and great vessels. In 1 case (fig. 3 and 4), the deformity of the pulmonary conus and the pulmonary artery and its left branch was sufficient to cause right ventricular hypertrophy and chronic cor pulmonale.\(^5\)

In summary, the presence of a syphilitic aortic sinus aneurysm should be suspected whenever a fusiform or saccular aneurysm of the ascending aorta seems to extend into the heart. If, in addition, there is linear calcification of the ascending aorta reaching into the intracardiac origin of the aorta and, especially, if there is linear or massive calcification in the region of the aortic sinuses, the diagnosis of a syphilitic sinus of Valsalva aneurysm becomes almost a certainty. Angiocardiography by outlining the root of the aorta will make the definitive diagnosis and will establish whether the aortic sinus aneurysm is continuous with an ascending aorta aneurysm or localized, producing pressure on the neighboring cardiac chambers and great vessels.

Rupture of Aortic Sinus Aneurysms

Sudden onset of severe dyspnea, right heart overloading and failure, enlargement of the pulmonary artery, and evolution of machinery-like murmurs may mean rupture of an aortic root aneurysm into the pulmonary artery or right heart chambers.\(^4\) Confirmation may be had by cardiac catheterization.\(^5,\)\(^6\) A recent attempt to repair a ruptured aortic sinus was not successful.\(^7\)
TREATMENT AND PROGNOSIS OF AORTIC SINUS ANEURYSMS

Surgical repair of syphilitic sinus of Valsalva aneurysms is formidable because of their large size and serious pressure effects on the neighboring cardiovascular structures. However, in the very rare congenital type (fig. 1) when the aneurysm is localized to a single aortic sinus, approach through the right atrium appears feasible. In our case, surgery of the aneurysm appeared especially dangerous because of the associated aortic regurgitation, coarctation of the aorta, and the recent bacterial endocarditis. However, the patient withstood surgical correction of the aortic coarctation very well. Even though congenital sinus of Valsalva aneurysms are paper thin and tend to rupture, surgical repair of the diffuse aneurysmal types of unruptured congenital sinus of Valsalva aneurysms does not seem warranted.

Early diagnosis and treatment of syphilis will prevent aortitis and aneurysms. Evidence is being accumulated that penicillin treatment of aortitis, either simple or complicated by aneurysm, is also beneficial. Of our 10 cases of syphilitic aortic sinus aneurysm, 3 are fatal, 3 were lost to observation, and 4 are alive and well and have been under medical care for periods up to 5 years.

The 9 patients with congenital types of aortic sinus aneurysms are all alive. The 4 with coarctation of the aorta had resection of the coarcted segments with complete relief of hypertension after end-to-end anastomosis of the aorta; all but the patient with recent subacute bacterial endocarditis are well and working. One of the 3 patients with arachnodactyly is still troubled by mild congestive heart failure; the other 2 continue to be free of cardiovascular complaints. The patient with pseudo-outflow of blood from the subclavian aorta is also free of cardiovascular symptoms and is working; while the child with the multiple valve disease and heart enlargement remains disabled.

The long-term prognosis of patients with unperforated aortic sinus aneurysms is unknown. However, even though there have been only a few years of observation, the general well being of the majority of our patients suggests that the prognosis is not immediately fatal. Once the aneurysm ruptures, the situation is usually hopeless. Therefore, even though the surgical approach is difficult and hazardous and the circulatory hemodynamics are desperate, an attempt to repair the perforation should be made.

SUMMARY

There are no diagnostic features for the recognition of unperforated aortic sinus aneurysms except for a grade 3 to 4 cardiac murmur. Linear or massive calcifications in the intracardiac portion of the root of the aorta continuous with the linear calcifications of the ascending aorta sometimes occurs in syphilitic aortic sinus aneurysms. Accordingly, a high index of suspicion and angiocardiography are advocated in order to establish the diagnosis during life.

Surgical repair of a localized congenital aortic sinus aneurysm through the right atrial approach seems feasible if there are no serious complicating aortic or cardiac lesions. The diffuse unruptured aneurysmal congenital and syphilitic aortic sinus aneurysms are better left alone. If rupture occurs, surgical repair, even though heroic, should be attempted.

ADDENDUM

Since this review was submitted for publication 5 more congenital aneurysmal dilatations of the aortic sinuses have been discovered. Four were associated with Marfan's syndrome (2 in identical twins 2 years old), the fifth with coarctation of the aorta. Two recent reports from England also describe operations in 2 patients for closure of perforated congenital aortic sinus aneurysms.

SUMMARIO IN INTERLINGUA

II non ha characteristicas diagnostic pro le recognizion de non-perforate aneurispermas del sino aortie, excepte un mormure cardiae de grado III a IV. Calcificationes linear o massive in le portion intracardiae del radice aortie, continuante in le calcificationes linear del aorta ascendente, occure a vices in syphilitic aneurispermas del sino aortie. Ergo, un alte grado de suspicion e le uso de methodos angiocardiographie es recommendabile si on vole establir le diagnose durante le vita del patiente.
In le absentia de serie aortic o cardiac lesions complicatori, il pare practicable reparar un localisate e congenite aneurysma del sino aortic via le atrio dextere. In casos de diffuse non-rupturate aneurysmas del sino aortic, tanto congenite como etiam syphilitic, il vale melio abstener se de omne intervention. Si un ruptura se effectua, le tentativa de un reparo chirurgic debe esser interprendite, ben que un tal es alora heroic.

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