Congenital Aneurysm of Aortic Root with Fistula to Left Ventricle

A Case Report with Autopsy Findings

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RECENTLY we observed an infant with severe aortic insufficiency and progressive left ventricular failure. Postmortem studies revealed aneurysmal dilatation of the base of the aorta and a large communication with the left ventricle. The aneurysm involved the right sinus of Valsalva but was not limited to that region. The rarity of congenital aortic insufficiency in general and of this lesion in particular and the increasing feasibility of surgical repair warrant a detailed description of the case.

Summary of Case

This Caucasian male infant was born on July 17, 1960, following a pregnancy that was complicated only by a fever during the first trimester. He remained apneic after birth for an interval of 5 minutes and acrocyanosis persisted after resuscitation. Physical examination revealed a weight of 6 pounds 1 ounce, respirations of 60 per minute, and a pulse of 128 per minute. Blood pressure by palpation was 110 mm. Hg systolic in the upper extremities. All peripheral pulses were bounding in quality and the head bobbed with each heart beat. The liver edge was felt 2 cm. below the right costal margin and was slightly rounded. The lungs were clear to percussion and auscultation. The heart was enlarged to the left axilla on percussion, and its apex impulse was displaced almost to the midaxillary line. A left ventricular thrust was diffusely felt over the lower left chest and the left axilla. A long thrill was felt over the midsternum and the mid-left sternal border. The first heart sound was normal in intensity at the apex. The second heart sound was difficult to hear at the base because of the loudness of the murmur present, but it was audible and did not appear to be accentuated. A prominent third heart sound was audible over the lower precordium. A systolic ejection sound was heard along the lower left sternal border. A grade-V, rough, continuous murmur was heard maximally at the mid-left sternal border but was also well heard over the entire precordium and was transmitted to the back (fig. 1).

The electrocardiogram (fig. 2) showed a mean QRS axis of pulse 75°, deep S waves in the right and mid precordial leads, and moderately tall R waves in leads V5 and V6.

At fluoroscopy the heart was markedly enlarged. A smooth rounded mass was apparent in the superior mediastinum. Marked pulsation of this mass was apparent and a rocking motion could be seen when the right margin of the mass was compared with the left. The barium-filled esophagus was displaced posteriorly (fig. 3).

The patient was digitalized, and his condition improved sufficiently to warrant an aortogram on August 2, 1960. Four injections of 90 per cent diatrizoate sodium (Hypaque) were made, with a hand syringe, while rapid x-ray exposures were taken. The angiocardiogram (fig. 4) disclosed marked dilatation of the ascending aorta, particularly its anterior aspect. A large volume of blood leaked immediately into the left ventricle. Slight regurgitation through the mitral valve was also evident. The major branches of the arch of the aorta were normal. An anomalous coronary artery communication was not observed, and there was no evidence of a left-to-right shunt.

During the night following the procedure, the patient's condition suddenly worsened, he became cyanotic even in oxygen, and he died.

At autopsy a slightly eccentric, massive dilatation of the base of the aorta accompanied by left ventricular hypertrophy was apparent (fig. 5). At the inferior extremity of this aneurysm were two openings into the left ventricle separated by a
relatively dense septum (figs. 6, 7, and 8). Three normally formed cusps were present within the right and posterior opening, and this opening was of approximately the same diameter as that of the pulmonary artery. The right coronary artery arose from the anterior wall of the dilated region of the aorta just superior to the fistulous left ventricular opening, which had no cusps. The pulmonary artery was separated from the right ventricle by three normally formed cusps. There were no interventricular or interatrial septal defects. Microscopic examination of the common wall between the pulmonary artery (with cusp) and the aortic-ventricular fistula (fig. 9) revealed a defect in the formation of the fibrous skeleton of the heart with apparent separation of the aortic annulus from the remaining portions of the common wall. This resulted in a wide-open communication from the aorta to the left ventricle.

Discussion

Isolated congenital aortic insufficiency is a rare lesion which has recently been studied from a clinical and hemodynamic approach by Frahm et al.\textsuperscript{1} Increased interest in sinus of Valsalva aneurysms, both ruptured and intact, has developed in recent years with the advent of retrograde aortography and the increasing feasibility of surgical repair.\textsuperscript{2-11} The majority of ruptured sinus of Valsalva aneurysms rupture in adult life and establish fistulas to the right atrium or right ventricle. The lesion in our patient is unusual because it ruptured before birth and because the resulting fistula from the aortic base communicated with the left ventricle. Two cases of right sinus of Valsalva aneurysm with rupture into the left ventricle have been reported.\textsuperscript{12} Harvey reported that of 600 cases of aortic insufficiency evaluated by him only two were considered congenital.\textsuperscript{13} One of these was found at postmortem examination to have a "separation of the aortic annulus from the wall of the aorta" and therefore may have had a lesion very similar to the one in our patient.
Most authors believe that congenital aneurysms of the sinuses of Valsalva result from a developmental weakness in the aortic wall just cephalad to the annulus fibrosus of the aortic valve. Mall demonstrated that while the aortic septum is growing to divide the primitive truncus arteriosus into aorta and pulmonary arteries, the aorta is shifting from the right side of the heart to its posterior position opening into the left ventricle. At the same time the inferior septum of the ventricles, which is to form the superior part of the ventricular septum, is shifting from left to right. The right margin of the aortic septum then fuses with the inferior septum to form the membranous septum. If this fusion is incomplete, a high interventricular septal defect will result. If fusion occurs in such a manner as to result in an attenuated area in the region of the sinuses of Valsalva, then the repeated impact of the pulse wave in the aorta will dilate the attenuated region and result in an aneurysm of the sinus of Valsalva.

The aneurysm in our case was not strictly limited to the region of the sinus of Valsalva. It seems likely, however, that the basic defect

Figure 3

Posteroanterior and lateral views of chest showing posterior displacement of esophagus.

Figure 4

Lateral view of angiogram showing marked dilation of the ascending aorta, particularly its anterior aspect, and leakage of dye from aorta into the left ventricle. An, aneurysm; LV, left ventricular outflow tract.
Figure 5
Anterior view of organs of chest showing marked dilatation of the aortic base and cardiac enlargement. An, aneurysm; AA, aortic arch; PA, pulmonary artery.

Figure 6
Schematic drawing of the base of the great vessels. The left drawing illustrates normal relations and the right illustrates the defect found at autopsy in our case. “P” indicates the plane through which the section illustrated in fig. 9 was made and passes from the fistulous opening, through the common wall between the aneurysm and the pulmonary artery, and into the right posterior cusp of the pulmonary valve.

Figure 7
View of base of aorta seen from above after aneurysm has been opened and the wall reflected. The pulmonary artery is collapsed and is not seen in this view. F, opening of fistula to left ventricle; AV, relatively normal aortic valve; RCA, right coronary artery.

Figure 8
View of heart from left showing probes passing through the two openings at the base of the aorta and into the left ventricle. An, aneurysm; ALV, anterior left ventricle; PLV, posterior left ventricle.
CONGENITAL ANEURYSM AORTIC ROOT

Figure 9
Photomicrograph of spiral section through wall of aneurysm, common wall of aneurysm and pulmonary artery, fibrous skeleton of heart, and superior portion of interventricular septum. Notice the distortion of the fibrous skeleton of the heart in the area between the site of separation of the aortic annulus and the site of attachment of the pulmonic valve cusp. PA, pulmonary artery; A, aorta; A\(\text{a}W\), aneurysm wall; F, fistula; PV, pulmonary valve; IVS, interventricular septum; RV, right ventricle; LV, left ventricle.

was similar. In our case the attenuated area was at the site of fusion with the annulus and most likely resulted in early separation of the annulus from the aortic wall. When separation from the aortic wall occurred, a direct communication between the aorta and left ventricle resulted. Following this there was marked turbulence of the stream at the base of the aorta and increased pulse pressure. This turbulence and high pulse pressure then resulted in a more diffuse dilatation than is found in most sinus of Valsalva aneurysms. The microscopic examination revealed no medial degeneration or other striking defects in the wall of the aneurysm. This finding lends support to the thesis that a major portion of the dilatation was due to the hemodynamic effects of separation of the annulus and not due to Marfan's disease or other extensive intrinsic aortic wall defects.

This case illustrates that aortic regurgitation may lead to cardiac failure and death early in life. Methods for preventing congenital lesions similar to this are not known at present. It is hoped, however, that increased surgical skill and diagnostic acuity will lead to the salvage of children with these lesions.

Summary
Clinical and autopsy findings in a case of a congenital aneurysm of the base of the aorta involving the right sinus of Valsalva and communicating with the left ventricle are presented. The mechanism of development and possible etiologic factors are discussed.

Addendum
Since this article was submitted for publication Levy et al. have reported three very similar cases with surgical cure in one of them.

References
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The Hippocratic Glossary

The eminent medical historian Karl Sudhoff wrote a very sound essay entitled "What is History of Medicine?" in which he decries the habit of dating everything from Hippocrates "although (as Sudhoff writes) we now know that not only in those parts of Asia Minor where Greek was spoken and in the outlying islands, many an able medical thinker flourished before Hippocrates, and that to the Greeks the name of Hippocrates did not represent the beginning of medical science and medical art."

Sudhoff is very right in this, but nevertheless in the field of medical terminology it is to the Hippocratic writings we must turn for the first appearance of much of our medical vocabulary. Of course, Hippocrates merely recorded the current terminology including such modern sounding words as arthritis and pneumonitis, but we do not know their earlier history. Egyptian contributed words including the root of hyalos, meaning glass, which we use in hyaline, and from Crete came a group of terms ending in nth or nx, such as, for example, salpinx and meningitis from which we get salpingitis and meningitis. But the bulk of his vocabulary was of earlier Greek origin. A mere listing of the Hippocratic glossary would give a reasonably good picture of the medicine of that era.—O. H. Perry Peiper, M.D. Opuscula Medica. (Reprinted from Transactions & Studies of the College of Physicians of Philadelphia, 4 Ser., 18: 29, April, 1950).
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