Congenital Aneurysm of the Right Sinus of Valsalva, Diagnosed by Aortography

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This paper contains a brief review of the cases, published earlier, of aneurysms of the sinuses of Valsalva, and a case is reported in which the diagnosis was made before rupture of the aneurysm by means of thoracic aortography. No reports are to be found in the literature of the diagnosis of unruptured aortic sinus aneurysms and of the associated clinical findings. Roentgenograms, aortograms and catheterization findings are included.

The three sinuses of Valsalva, or aortic sinuses, form the proximal, partly intracardiac portion of the aorta. Congenital aneurysms, developing from the sinuses, are held by Abbott¹ to be caused by abnormal fusion of the aortopulmonary septum with the ventricular septum. This theory can only be applied to the aneurysms arising from the right and posterior sinuses. Micks² and Raman and Menon,³ however, have described congenital aneurysms of the left sinus of Valsalva. The theory of Venning⁴ that the aneurysms are due to defects in the elastic tissue of the sinuses, therefore, seems more plausible.

The rarity of aneurysms of the sinuses of Valsalva has been demonstrated by Schuster,⁵ who in 3000 autopsies found two cases. Snyder and Hunter⁶ among 5896 autopsies likewise found only two cases, and they stated that 10 aneurysms of the sinuses of Valsalva were found among 287 aortic aneurysms in 12,000 autopsies. The etiologic factors were maintained to be syphilis, atherosclerosis, and endocarditis. A group of cases was considered of unknown origin.

Abbott¹ in her review was able to find only 12 cases of congenital aneurysms in the literature. Morgan Jones and Langley⁷ in their recent review collected 23 cases and added two cases of congenital aneurysm. Since then Venning has published three additional cases. Micks⁸ reported a case of congenital aneurysm of all three sinuses of Valsalva and referred to three similar cases, published earlier.

Of the 33 cases published to date, 23 have developed from the right aortic sinus, five from the noncoronary sinus, one from the left aortic sinus, and in four cases aneurysms of all three sinuses were found. Depending upon the origin of the aneurysm, it extends into the interventricular septum or the wall of the ventricles or auricles. Other congenital abnormalities of the heart are often associated with the aneurysms, such as ventricular septal defect and bicuspid aortic valve.

Rupture of the aneurysm can occur at any time of life and is usually provoked by physical strain. While acquired aortic sinus aneurysms most frequently rupture into the pericardial sac,⁹ the congenital aneurysms usually rupture into the right ventricle. Eight cases have been reported which ruptured into the right auricle, and single cases have been reported which ruptured into the pulmonary artery and left ventricle, respectively.

Before rupture of the aneurysm, no symptoms or uncharacteristic symptoms are present. In the case published by Herson and Symons⁸ the patient was examined at the age of 12 years, several years before rupture occurred. A loud systolic murmur was heard over the whole precordium. X-ray examination showed no cardiac enlargement and the electrocardiogram was normal. Autopsy after rupture of the aneurysm 20 years later revealed that, besides an aortic noncoronary sinus aneurysm, a ventricular septal defect was present. The murmur was possibly attributable to the septal defect. In the case reported by Micks, a sys-

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tolic apical murmur was audible and roentgenograms showed enlargement of the heart.

Cardiac enlargement, prolongation of the auriculoventricular conduction time, and diastolic murmurs can probably be attributed to associated congenital abnormalities of the heart or complications due to progression of the aneurysm and encroachment on the conduction bundle, the pulmonary artery, the aorta, the auriculoventricular valves or the coronary arteries. Ostrum and coworkers by x-ray screening have observed intracardiac bulging caused by acquired aortic sinus aneurysms and have discussed similar bulgings in other conditions.

Rupture of the aneurysm produces an acutely critical situation with precordial or substernal pain, shortness of breath and feeling of weakness, often followed by collapse. After rupture into the right side of the heart, a systolic-diastolic murmur will develop, in some cases accompanied by a systolic, rarely a diastolic thrill, with its maximum in the third and fourth left intercostal space. When, after an acute onset, a collapsing pulse is combined with evidence of pulmonary hypertension, shown by x-ray examination, a fistula between the aorta and the lesser circulation is suggested, as pointed out by Venning.

The time of survival after rupture of the aneurysm into the right side of the heart has been described as varying from a few seconds to 17 years. The oldest patient in whom rupture of a congenital aortic sinus aneurysm has been reported was 64 years of age. Death after rupture is caused by heart failure.

Bacterial endocarditis is maintained to have been the cause of death in six cases of congenital aneurysm. Jones and Langley suggest that when bacterial endocarditis develops in a heart apparently previously healthy, an aneurysm of the sinus of Valsalva must be suspected. In the case reported by Micks, heartblock was the cause of death.

No reports are to be found in the literature of the diagnosis of unruptured congenital aortic sinus aneurysms during life. Based upon the history of an acute onset, a continuous murmur, evidence of pulmonary hypertension, and a collapsing pulse, Venning made the diagnosis of a ruptured aneurysm of the sinus of Valsalva during life. The differential diagnosis of the condition being discussed and aortic septal defect and even patent ductus, however, can be of great difficulty. If rupture of the aneurysm has taken place into the outflow tract of the right ventricle or into the pulmonary artery, cardiac catheterization will be of no great help, unless the ductus or aortic septal defect are directly catheterized, since the physiologic conditions will be the same in any of these instances.

The performance of aortography after introduction of a cardiac catheter into the ascending aorta has greatly widened the possibilities of studying the anatomy of the aorta in the living subject. In the case to be reported, aortography was made by the technic developed by Broden and associates.

**CASE REPORT**

A 17 year old girl from Iceland was referred to us by Dr. S. Samuelsson, of Reykjavik, because of cardiac complaints. She gave no family history of congenital disease; there was no history of syphilis and clinical examination revealed no symptoms of congenital syphilis. At 1 year of age she had had a severe attack of measles but had suffered from no other infectious diseases. Her physical abilities had always been diminished, compared with that of her contemporaries, because of dyspnea and palpitations.

At school she was not able to take part in gymnastics. Cyanosis was never observed, and there had been no attacks of precordial pain or severe dyspnea.

The diagnosis of congenital heart disease was made at the age of 13 years.

Half a year before admission to the hospital she had been employed drying codfish which involved lifting heavy boxes of fish. Because of dyspnea and palpitations she had to give up this work. She suffered no discomfort from walking or climbing stairs.

Her height was 163 cm. and weight, 54.4 Kg. The temperature remained normal during her hospitalization. The blood pressure in an arm was 130/90. Her intelligence was normal. There was no cyanosis of the skin and mucous membranes, and no dyspnea at rest. The lungs were free of abnormality on examination.

The apex beat was palpable in the fifth intercostal space 1 cm. lateral to the mideavicular line. A systolic blowing murmur, grade 2, and a short proto-diastolic murmur were audible at the apex. The intensity of the murmurs increased toward the sternum, and in the left third and fourth intercostal space there was a continuous murmur similar to
that of a patent ductus, though of higher frequency. In the right fourth intercostal space the diastolic murmur was dominating, harsh in character and peculiarly superficial. The murmurs were weak, though audible in the second intercostal space to the right of the sternum. There was a weak systolic thrill with its maximum in the fourth space to the left of the sternum. No transmission of the murmurs to the arteries in the neck was observed. The pulse was considered normal. A faint pistol shot sound was audible over the femoral arteries. There was found no evidence of capillary pulsation nor clubbing of the fingers. The liver was not palpable. No edema was present.

The sedimentation rate was 8 mm. in 1 hour, the hemoglobin 95 per cent and the red blood cell count 4.18 million. The hematocrit was 36 per cent. The antistreptolysin titer was found to be 160. The Wassermann and Kahn reactions were negative. The basal metabolic rate was 102 per cent. No abnormal constituents were found in the urine.

Electrocardiograms showed regular rhythm and a rate of 70 per minute. The P-Q interval was 0.16 second and the QRS interval was 0.08 second. In all standard leads the R wave was of highest voltage; in lead I, 18 mm., and in lead III, 7 mm. in height. The unipolar limb leads showed a semivertical heart. The precordial leads gave normal tracings.

Roentgenograms showed the heart to be normally shaped; the cardiac index was 12/24. The left ventricle was slightly enlarged. The vascular markings in the lungs were normal (fig. 1).

Oximetry demonstrated no abnormal fall in arterial saturation during three minutes of exercise (405 kilogram-meters per minute).

Cardiac catheterization* showed the following findings. Pressure in the pulmonary artery was 22/9 mm. Hg, in the right ventricle 22/0, and the mean pressure in the right auricle was zero. The oxygen saturation in the superior vena cava and the right auricle was 74 per cent, and in the pulmonary artery 75 per cent. The arterial saturation, determined by the van Slyke method, was 94 per cent. No evidence of shunts was found. The catheterization revealed no explanation of the cardiac symptoms.

Aortography: Under nitrous oxide oxygen-ether anesthesia, 60 cc. of 70 per cent Diodon were injected through a number 9 cardiac catheter, which was introduced into the ascending aorta via the right radial artery. Twenty aortograms in the right and left position were taken within 10 seconds.

Fig. 1. Roentgenogram in the anterior-posterior view.

Fig. 2. Aortogram in the right oblique position.

The aortic valves are rather sharply outlined. A narrow band of contrast substance is seen, extending from the aortic valve into the outflow tract of the left ventricle where the contrast disappears. This must be an expression of insufficiency of the valve.

A rounded shadow of contrast about the size of a walnut appears below and a little in front of the aorta. A thin connection is seen, extending from the right aortic sinus to the shadow previously described. After its disappearance from the aorta, dye is still seen to be retained in the area of this shadow. No evidence of emptying of the dye into the lesser circulation is found.

Conclusion: A chamber below and a little in front

*Pressures were recorded by means of the electrical condenser manometer of Hansen. The oxygen saturation of the blood samples was determined by the Brinkman and Zylstra17 hemorefractometer. The oxygen capacity was determined by the van Slyke apparatus.
of the aorta, but situated deeply in the heart shadow, has been demonstrated. This chamber is only in communication with the aorta. It is undoubtedly an unruptured aneurysm of the right sinus of Valsalva. Furthermore, evidence of insufficiency of the aortic valve has been found (figs. 2, 3 and 4).

Fig. 3. Aortogram in the left oblique position

Fig. 4. Aortogram in the left oblique position, taken four seconds later than those in figures 2 and 3. Dye is still seen to be retained in the aneurysm.

DISCUSSION

No definite diagnosis was made on the clinical findings in the case reported. Based upon the location and character of the murmurs, the diagnosis of an aortopulmonary fistula, possibly an aortic septal defect, was entertained. The roentgenograms and catheterization findings did not support this diagnosis, as there was no evidence of pulmonary hypertension nor of an arteriovenous communication within the heart or involving the pulmonary artery. Aortography proved the presence of an unruptured aneurysm of the right sinus of Valsalva and an insufficiency of the aortic valve.

Aneurysms developing in the area under discussion could be either of the right aortic sinus or the right coronary artery. The congenital saccular aneurysms of the right coronary artery develop from the points of partition of the artery, usually 0.5 to 2 cm. distal to the sinus of Valsalva.8 These aneurysms are located on the surface of the heart. In the case reported, the aneurysm, like an aortic sinus, is deeply buried in the musculature of the heart, and a fistula from the sinus of Valsalva to the aneurysm can be followed on aortograms.

It seems reasonable to explain the murmurs found in this case as resulting from the intracardiac location of the aneurysm. During contraction of the heart muscle, the aneurysm will be partially compressed and, since no rupture of the aneurysm was found, blood will pass from the aneurysm into the aorta during systole. After relaxation of the muscle, blood will pass from the aorta into the aneurysm during diastole. In this way a systolic-diastolic murmur is produced. The murmur caused by the associated insufficiency of the aortic valve would be of another location and character than the one described in the previous section.

Murmurs, similar to the ones found in our case, have been reported in earlier published descriptions of aortic sinus aneurysms. The peculiar impression of the superficial location of the murmur has been described by Abbott and by Jones and Langley. In their cases, the aneurysm, in contrast to the present one, had ruptured. Consequently the aneurysm would have emptied into the right side of the heart during systole, and have been refilled from the aorta during diastole.

Insufficiency of the aortic valves, according to the literature, is a frequent accompaniment of congenital aneurysms of the sinuses of Valsalva. It can be due to congenital abnormality of the aortic valves, bicuspid valves, or encroachment by the aneurysm on the valve or outflow tract of the left ventricle.
The opinion that our case is of congenital origin is based upon exclusion of the known causes for the development of acquired aneurysms of the sinuses of Valsalva. There was no evidence of either congenital or acquired syphilis; the patient had had no serious infectious diseases; examination of the arteries revealed no signs of premature arteriosclerosis; and kymograms showed normal pulsations of the aorta.

**SUMMARY**

The clinical, prognostic and pathologic experiences from cases of congenital aortic sinus aneurysms, published earlier, are briefly reviewed and the embryology discussed.

In a 17 year old girl a case of congenital aneurysm of the right sinus of Valsalva was diagnosed by aortography. A report is given on the history and the clinical findings. Roentgenograms and cardiac catheterization rendered no help in establishing the diagnosis.

The diagnostic problems and origin of the murmurs in the case reported are discussed.

**SUMARIO ESPAÑOL**

Este trabajo contiene un breve repaso de los casos de aneurismas de los senos de Valsalva y se informa un caso en el cual el diagnóstico se estableció antes de la rotura del aneurisma por medio de aortografía torácica. En la literatura no se encuentran informes de casos diagnosticados antes de la rotura del aneurisma en aneurismas del seno de Valsalva ni de los hallazgos clínicos relacionados. Radiografías, aortogramas y hallazgos de cateterismo se incluyen.

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