AORTIC SINUS ANEURYSMS

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The three slight dilatations of the aorta immediately above the corresponding valve cusps, known as the aortic sinuses of Valsalva, are almost entirely intracardiac and lie near important parts of the heart (Fig. 1). Aneurysms arising from these sinuses may extend into the cardiac chambers and encroach upon the pulmonary valve, the atrio-ventricular valves, the conducting bundle, or the pulmonary or coronary arteries, or may lead to aortic regurgitation by deforming the aortic ring. They may rupture into one of the cardiac chambers, the pulmonary artery, or the pericardium, and sometimes communicate from birth with a cardiac chamber or the pulmonary artery. To these various complications such aneurysms owe much of their interest.

Aneurysms confined to the aortic sinuses are uncommon; Smith (1914) noted that in 8138 necropsies only 7 were found and, in all, only 20 cases were confirmed at necropsy prior to 1914. These aneurysms may be due to any of the usual causes, but those of congenital origin are of particular interest. The first recorded case of this type appears to be that of Thurnam (1840) and when Abbott (1936) collected her 1000 cases of congenital heart disease she was able to find only 12 congenital aortic sinus aneurysms recorded (Bauer & Astbury, 1944).

Our objects in this paper are to describe four new cases, which present certain features of interest, and to review the available clinical and pathological material particularly so far as it illustrates the pathological differences between the acquired and congenital types of aneurysm or aids the clinical diagnosis of congenital aneurysms. Some preliminary discussion of the nomenclature of the aortic sinuses and of the topography of the aortic root is desirable to clarify the anatomical features of the cases to be described.

NOMENCLATURE OF THE AORTIC SINUSES

The many ways of naming the aortic cusps and their sinuses are so confusing that it is often difficult to identify the sinus described. In Table I we have enumerated five methods of nomenclature and in Fig. 2 have indicated the sinus to which each name refers. The standard nomenclatures [Old Terminology; Basle Anatomical Nomenclature, 1895 (B.N.A.); Birmingham Revision, 1933 (B.R.)] have assumed that one aortic sinus lies anterior and two posterior, and always placed in an anterior position the sinus from which the right coronary artery arises, but Walmsley (1929) has pointed out that sometimes two aortic sinuses lie anterior, and one posterior (Fig. 2); he suggested that sinuses related to the coronary arteries should be named right and left coronary sinuses, the third sinus being called the non-coronary sinus. This nomenclature is easy to understand and allows the sinuses to be named when the heart has been removed from the body and the sinuses exposed by opening the aorta; we have therefore adopted it in this paper.

TOPOGRAPHY OF THE AORTIC ROOT

The structures adjacent to the aortic sinuses are of considerable importance in discussing the clinical and pathological features of sinus aneurysms; these structures are illustrated in Fig. 1 and described in the caption.

The annulus fibrosus. Between the aorta proper and the main body of the left ventricle there is a tubular zone of fibrous tissue, the annulus fibrosus, which forms an important part of the wall of the aortic sinuses and extends downward to become incorporated in the aortic vestibule of the left ventricle. It is believed that congenital sinus aneurysms arise from defective development of the bulbar septum which divides the primitive exit

FIG. 1.—Position of the aortic sinuses. From a normal heart, fixed unopened, with vessels and chambers distended. The anterior and part of the right walls of the right ventricle and right auricle have been removed and the aorta opened. The anterior part of the ventricular septum (I.V.S.) has been sectioned. The right coronary sinus (R.C.S.) projects into the conus of the right ventricle (R.V.). The non-coronary sinus (N.C.S.) projects into the right auricle (R.A.) and the tricuspid valve lies below adjacent sectors of the non-coronary and right coronary sinuses. The left coronary sinus (L.C.S.) with its coronary artery can be seen postero-laterally. It is related behind to the mitral valve and the left auricle. S.V.C.—Superior vena cava. A.—Aorta. P.A.—Pulmonary artery.

tube of the heart, the bulbus cordis, into right and left halves. Since the annulus fibrosus arises from the bulbus cordis and forms part of the wall of the aortic sinuses, it is a particularly important structure to study when a sinus aneurysm is present.

CASE REPORTS

Case 1. A congenital aneurysm of the right coronary sinus associated with a bulbar ventricular septal defect, clinically simulating patency of the ductus arteriosus and complicated by subacute bacterial endocarditis.

In April, 1939, a schoolboy, age 12, was referred to Dr. Crighton Bramwell on account of a heart murmur first discovered during a respiratory infection in infancy. Apart from tonsillitis when 5, he

had enjoyed good health. At school he led a normal life and played games, including football, without cyanosis or unusual dyspnoea. At 10 years of age tonsillectomy was advised, but not performed owing to the cardiac murmur. He suffered from epistaxis on several occasions and in August and December, 1938, severe attacks occurred.

On examination his general development was normal; height 60 inches, weight 80 lb. His exercise tolerance was good and there was no cyanosis or finger clubbing. The pulse was regular, rate 80, the blood pressure 125/40. To the left of the sternum, at the base, systolic and diastolic murmurs were heard. Cardioscopy showed considerable enlargement of both ventricles, but a cardiogram was normal apart from R waves of

TABLE I
NOMENCLATURE OF THE AORTIC SINUSES

	Walmsley (1929)	Common position		Less common position	
	(a)	(b) B.R. 1933	(c) B.N.A. 1895	(d)	(e)
1	Right coronary	Anterior	Anterior	Right	Right anterior
2	Non-coronary	Right	Right posterior	Posterior	Posterior
3	Left coronary	Left	Left posterior	Left	Left anterior

exceptionally high voltage in the standard limb leads. The condition was diagnosed as patency of the ductus arteriosus.

In January, 1942, subacute bacterial endocarditis was suspected and he was admitted to hospital. Seven weeks prior to admission he had a pyrexial illness lasting a few days accompanied by vomiting and shivering. Subsequently his appetite failed to return and he complained of transient pains in the limbs, felt continually tired, lost weight, and was troubled by nocturnal sweating. For a week before admission he suffered from substernal pain which frequently awakened him at night.

On examination he weighed only 84 lb. and was pale. His temperature was 100.2° F. Striking arterial pulsation was visible in the neck, the pulse was regular, rate 120; the blood pressure 120/30. A systolic thrill was palpable in the second and third left interspaces near the sternum accompanied by a harsh systolic murmur and a softer diastolic murmur, both maximal in the same position, but audible over the whole præcordium. A teleradiogram showed considerable cardiac enlargement (cardiothoracic

ratio 0.66) involving both ventricles. The cardiogram was similar to the previous record. The liver was palpable two inches below the costal margin; there was no œdema but a few moist sounds were heard at the lung bases. The spleen was palpable and there were scattered petechiæ on the abdomen, chest, neck, and arms. An Osler's node was found on the tip of the right little finger. Blood culture yielded a growth of *Streptococcus viridans*. His condition steadily deteriorated and three weeks after admission he collapsed suddenly and died the following day.

At necropsy a few petechiæ were found on the arms; there was subcutaneous œdema of the trunk and effusions were present in the pericardial, pleural, and abdominal cavities. There was generalized passive congestion and infarcts were present in the spleen and right lung; these appearances were confirmed histologically.

The heart was globular in shape and weighed 600 g. The right atrium was slightly hypertrophied but the left was normal. The right ventricle was slightly, and the left ventricle considerably hyper-

FIG. 2.—Nomenclature of the aortic sinuses. (1), (2), and (3) are the sinuses named in Table I. R.C.—right coronary artery. L.C.—left coronary artery. The percentages refer to Walmsley's (1929) estimate of the relative frequency of the two arrangements.

trophied. The foramen ovale and the ductus arteriosus were closed. In the anterior part of the ventricular septum, immediately below the right coronary cusp, there was a triangular opening, base 1.3 cm., height 0.8 cm., communicating between the conus of the right ventricle and the aortic vestibule of the left ventricle (Fig. 3). This opening lay anterior to the membranous septum and above the septal and parietal muscle bundles; it was therefore a defect of the true bulbar septum. Vegetations characteristic of subacute bacterial endocarditis were present around the margins of the septal defect.

The right coronary sinus was enlarged to form an

aneurysm with a mouth 2 by 2 cm. and a depth of 2.5 cm. From the upper limit of the mouth the wall of the aneurysm extended horizontally for a distance of 0.6 cm., and then curved downwards obliquely for 0.5 cm. to a well defined transverse ridge. Below this the outer wall of the aneurysm was related to the muscle of the right ventricle and then joined the right coronary cusp. The size of the right coronary cusp was about normal but, owing to the aneurysm, the intercommissural distance was small; consequently the cusp sagged downwards through the septal defect (Fig. 3A), its lower part forming the upper margin of the communication between the ventricles. This sagging

FIG. 3.—Case 1. From a drawing of the aneurysm. (A) Aortic and left ventricular aspect. (a)—The aneurysm. The sagging of the right coronary cusp can be seen. The arrows indicate the margins of the ventricular septal defect, which is surrounded by the vegetations of bacterial endocarditis. (B) Pulmonary and right ventricular aspect. (b) The apex of the aneurysm projecting into the right ventricle through the septal defect. The margins of the septal defect are indicated by arrows.

had made apposition of the aortic cusps impossible so that the valve was incompetent.

The non-coronary aortic cusp was enlarged, measuring 3 cm. between the commissures and the corresponding sinus was deeper than usual (1.9 cm.). The left coronary cusp measured 1.5 cm. between the commissures. The right coronary artery was much smaller than the left, its orifice lying in the part of the aneurysm adjacent to the non-coronary sinus. The left coronary orifice was puckered and lay near the commissure between its sinus and the non-coronary sinus.

On *histological examination* the aorta and aortic valve cusps were normal apart from the vegetations and the inflammatory changes of bacterial endocarditis. From the annulus fibrosus a tongue of fibrous tissue ran upwards to the transverse ridge on the aneurysm wall, lying between the muscle of the right ventricle and the endothelial lining of the aneurysm. The aortic media terminated at the transverse ridge.

In addition to the aneurysm, several congenital anomalies were present; a bulbar septal defect, an abnormal tongue of fibrous tissue arising from the annulus, and grossly unequal aortic cusps. The aortic wall was free from disease other than bacterial endocarditis, which might have arisen upon the developmental defects. In view of these findings, it seems justifiable to attribute the aneurysm to a developmental anomaly.

The presence of aortic incompetence accounted for the collapsing pulse, the large pulse pressure, and the diastolic component of the murmur. The systolic component and the thrill were explicable by the presence of the ventricular septal defect.

Case 2. A congenital aneurysm of the right coronary sinus, which ruptured into the conus of the right ventricle and led to progressive heart failure.

In January, 1944, a man of 41, a chauffeur, was admitted to hospital, complaining of breathlessness and swelling of the legs and abdomen. Heart disease was first found at the age of 24 when he had a severe attack of breathlessness and præcordial pain. From that time he suffered from dyspnoea on exertion and præcordial pain. Some weeks before admission he noticed swelling of the legs and abdomen and his dyspnoea became more severe. He had never had rheumatic fever nor chorea and cyanosis had not occurred in childhood.

On *examination* he was cyanosed, dyspnoic at rest, and the neck veins were engorged. The pulse was regular, rate 104 a minute; systolic blood pressure 140 mm., diastolic end-point uncertain, the sounds being audible to complete decompression. The heart was enlarged, and a coarse systolic thrill

was palpable over the whole præcordium, most intense in the third left interspace near the sternum. Very loud, coarse, systolic and diastolic murmurs of machinery type were audible over the whole præcordium, but best heard in the third left interspace near the sternum. The liver was enlarged and tender and free fluid was present in the abdomen. There was considerable œdema of the legs and sacral area.

A cardiogram showed left axis deviation with depression of the RS-T segments in leads I and II; these appearances were attributed to left ventricular enlargement. A teloradiogram revealed considerable cardiac enlargement (cardio-thoracic ratio 0.63) involving both ventricles, but principally the left. The pulmonary artery and the pulmonary vascular markings were prominent.

With treatment by rest, mercurial diuretics, and digitalis he gradually improved, but his œdema did not entirely disappear. During his fourth week in hospital his condition began to deteriorate, his œdema increased, he became very breathless, and was soon jaundiced and pyrexial; he died after four weeks in hospital.

Necropsy was performed by Dr. G. Stewart Smith. The skin and mucous membranes were jaundiced and there was some œdema of the legs. The whole upper lobe of the right lung was consolidated and typical of the stage of grey hepatization of lobar pneumonia. Both lungs were congested and œdematous. There was generalized chronic venous congestion.

The *heart*, which we examined, weighed 640 g. In the left half of the right coronary sinus there was a round opening 0.7 cm. in diameter with rounded, slightly irregular margins (Fig. 4B). This led to a globular thin-walled cavity (Fig. 4A), 2 cm. in diameter, which projected into the conus of the right ventricle between the septal (posterior) and right anterior cusps of the pulmonary valve (Fig. 4A). On the upper surface of the aneurysmal sac was an aperture approximately rectangular in shape, 0.8 cm. by 0.6 cm., with smooth, rounded and thickened edges. The irregular shape of this opening suggested that it was due to rupture of the aneurysm and, since the edges were smooth and rounded, healing had occurred. The aneurysm thus formed a communication between the aorta and the right ventricle. The right coronary artery arose near the commissure between its sinus and the non-coronary sinus (Fig. 4B).

In the outflow tract of the left ventricle, 0.5 cm. below the aortic valve, was a fibrous band, lying on the interventricular septum, and extending from the right upper margin to the left upper margin of the anterior curtain of the mitral valve (Fig. 4B).

FIG. 4.—Case 2. *From a drawing of the heart.* (A) *Pulmonary and right ventricular aspect.* The perforated, globular aneurysm lies in the conus of the right ventricle just below the septal (posterior) and right anterior cusps of the pulmonary valve. (B) *Aortic and left ventricular aspect.* (a) The opening of the aneurysm in the right coronary sinus. The right coronary artery arises near the commissure between its sinus and the non-coronary sinus.

This abnormality occurred at about the level at which sub-aortic stenosis develops. The wall of the left ventricle was slightly thickened (1.5 cm.) and its cavity was dilated; this had led to the displacement of the ventricular septum towards the right ventricle. The wall of the right ventricle was about twice the normal thickness (0.9 cm.) and the columnæ carnæ and chordæ tendineæ were hypertrophied. The conus of the right ventricle was considerably dilated but its body appeared only slightly dilated, though this was difficult to estimate owing to the displacement of the ventricular septum. The circumference of the pulmonary valve measured 8 cm.; the septal cusp was fenestrated. The circumference of the aortic valve was 6.8 cm., and the left coronary cusp was slightly fenestrated. The mitral valve measured 7.5 cm. in circumference and the tricuspid valve 13 cm.; both valves appeared normal.

The absence of acquired heart disease and the presence of a congenital sub-aortic band of fibrous tissue make it reasonable to suppose that this aortic sinus aneurysm was of congenital origin. While unruptured it is unlikely to have given rise to symptoms, and it seems probable that the onset of symptoms 17 years before death coincided with rupture into the right ventricle; the healing of the margins of the rupture is consistent with this opinion.

Case 3. A dissecting aneurysm of the right coronary sinus communicating with the right and left ventricles and leading to an intracardiac hæmatoma around the right coronary artery.

In March, 1941, a married woman of 54 was admitted to hospital, complaining of vomiting and palpitation. Her previous health had been good except for an attack of rheumatic fever at age 27, when she was confined to bed for a year; she was subsequently told that she had a "weak heart." At 50, she noticed breathlessness on exertion and began to suffer from attacks of palpitation of sudden onset and offset, lasting 20 to 30 minutes. In June, 1940, she noticed increasing thirst, polyuria, and progressive weakness; a diagnosis of diabetes mellitus was made. She remained fairly well until two weeks before admission when she lost her appetite and became easily tired; for 48 hours before admission she vomited repeatedly and became drowsy. On the evening before admission she had an attack of palpitation which lasted several hours; during the night she awoke owing to a recurrence of the palpitation which continued until admission.

On examination it was found that the heart was completely irregular at a rate of 172 a minute. Blood pressure 120/85. The cervical veins were

engorged and moist sounds were present at both lung bases. The temperature was 100° F. and the urine contained a considerable quantity of sugar and acetone; blood sugar 355 mg. per 100 ml. The condition was diagnosed as diabetes mellitus with hyperglycæmic pre-coma, and uncontrolled auricular fibrillation.

She was treated with insulin, glucose, and intravenous digoxin followed by digitalis by mouth. Her heart rate fell to 94 in 24 hours, when an apical diastolic murmur, typical of mitral stenosis, became audible. A cardiogram confirmed the presence of auricular fibrillation. A telerradiogram of the chest showed slight cardiac enlargement. The diabetic ketosis was rapidly controlled, but her temperature remained raised owing to a *B. coli* pyelitis which was treated by sulphapyridine. After three weeks she was allowed up for a short time; a week later she had an attack of paroxysmal nocturnal dyspnoea. She was confined to bed and the attacks ceased after four nights. Four days later she suddenly collapsed, complaining of tightness across the chest; she became cyanosed and died within a few minutes.

At necropsy effusions were present in both pleural cavities and there was generalized chronic venous congestion. The heart weighed 480 g.; the left ventricle was hypertrophied but the right appeared normal except where the aneurysm projected into it. Both atria appeared normal. The mitral valve was moderately stenosed; the aortic cusps were thickened but the pulmonary and tricuspid valves appeared healthy. There was slight atheroma of the coronary arteries, especially near the orifice of the right coronary.

The lower part of the right coronary sinus formed an aneurysm about 3 cm. deep extending towards, and projecting into, the conus of the right ventricle (Fig. 5). The upper part of the inner (aortic) wall of the aneurysm was formed by the right coronary cusp; the middle part of the wall was disorganized and interrupted by a perforation communicating with the left ventricle; the lowest part of the inner wall was formed by the upper part of the ventricular septum. The apex of the aneurysm projected into the right ventricle. The outer wall was formed by a large, grey, necrotic mass, 6 by 4 cm., which enclosed the first part of the right coronary artery. The upper part of this mass contained a recent hæmorrhage. The greater part of the aneurysm was lined by loose blood clot; when this was removed a smooth lining of adherent thrombus was seen.

Histology. Normally the annulus fibrosus is of triangular shape and covered by a thin layer of elastic tissue on its aortic and left ventricular surfaces (Fig. 6A); the aortic layer is continuous with

FIG. 5.—Case 3. From a drawing of the heart. (a) Aorta. (b) Aneurysm. (c) Hæmatoma.

the internal elastic tissue of the aorta and the ventricular layer joins the subendothelial elastic tissue on the left side of the ventricular septum and is continuous with the subendothelial elastic tissue of the right ventricle. In the specimen the annulus fibrosus had become separated from the aortic media. The left ventricular layer of elastic tissue of the annulus could be traced through to the elastic tissue on the left side of the membranous septum. At the apex of the aneurysm the distribution of elastic tissue strongly suggests that this was the reflected right side of the septum (Fig. 6C).

These observations are explicable if the aneurysm had been formed as suggested in Fig. 6. First, blood penetrated between the termination of the aortic media and the annulus fibrosus (Fig.

6B); then it spread into the subpericardial fat to form the large hæmatoma. It next separated the muscle of the right ventricle from the annulus and then extended in three directions, (1) to rupture through the endothelium of the right ventricle (2) to split the membranous septum and reflect it with its elastic layer towards the right ventricle, and (3) to sever the apex of the annulus from the septum and to rupture through the endothelium of the left ventricle (Fig. 6C). Thus the dissection lay between the annulus and the muscle of the right ventricle and had communicated with both ventricles.

Although there is little doubt that this was a dissecting aneurysm arising in the right coronary sinus at the junction of the aortic media with the annulus fibrosus, it is more difficult to decide the nature of the lesion that led to the dissection. Examination of the aorta showed none of the degenerative changes such as cystic medial necrosis often found in dissecting aneurysms. There were, however, inflammatory changes with a round-cell reaction, fibroblasts, and new capillaries. These changes were sometimes perivascular but, unlike the changes of syphilis, they were confined to the adventitia and did not invade the media. Although the inflammatory changes were most severe and acute at the point of rupture, they were also present in the whole of the intrapericardiac aorta. There were also striking histological changes in the aortic and mitral valve cusps, which were scarred and vascularized by thin-walled blood vessels, and in the myocardium small areas of perivascular fibrosis were found. In the pericardium there were recent inflammatory changes with some cellular foci which resembled Aschoff bodies, though giant cells were not found. The association of these changes in the valves and myocardium with mitral stenosis confirms their rheumatic origin and suggests that the recent inflammatory changes in the pericardium and aorta were due to a recrudescence of cardiovascular rheumatism. The close resemblance of the aortic lesions to those described as rheumatic aortitis by Pappenheimer and von Glahn (1924) strengthens this hypothesis. It is therefore probable that the dissection arose owing to the development of active aortitis, possibly rheumatic in origin.

Case 4. An aneurysm of the non-coronary aortic sinus projecting into both atria, probably due to bacterial endocarditis.

A married woman, aged 52, was first seen in June, 1942, on account of hæmatemesis due to a gastric ulcer. At this time a soft apical systolic murmur was noticed, but no signs of organic heart disease; blood pressure 135/85. Hæmatemesis recurred in

FIG. 6.—Case 3. *The formation of the aneurysm.* (A) Normal structure of aortic root in section through right coronary sinus. The distribution of elastic tissue is shown by interrupted lines. The structures are named in (B). (B) Mode of dissection. The annulus fibrosus has been separated from the termination of the aortic media; the subsequent directions of dissection are indicated by arrows. (C) Aneurysm. By comparison with (B) the way in which the aneurysm has formed can be seen.

June, 1943, and again in September, 1945, when she was re-admitted to hospital.

Examination. Blood pressure 140/90 mm. Apical systolic murmur still present. Hæmoglobin 74, falling to 64 per cent in a few days. Radioscopy showed the heart at the upper normal limit of size. Slight fever was present and persisted. White cell count 5800–7200 per c.mm. Gallstones were demonstrated by cholecystogram. Eleven septic teeth were extracted without any improvement in her condition. On January 4, 1946, she had a sudden gripping mid-sternal pain lasting twelve hours and became breathless. A systolic thrill and murmur then appeared in the 2nd left interspace near the sternum, and a high-pitched diastolic murmur along the left sternal border. The pulse became collapsing; blood pressure 145/55 mm. A diagnosis of ruptured aortic cusp due to infective endocarditis was made, and blood culture showed a non-hæmolytic, penicillin-sensitive streptococcus. In spite of apparent control of the infection with penicillin, she died from heart failure three weeks later.

At *necropsy* bilateral pleural effusion, general passive congestion, splenic and renal infarcts, and chronic gastritis were found. The heart weighed 470 g. From the non-coronary sinus an aneurysm, 2.5 cm. in diameter, projected backwards into the atria (Fig. 7 and 8). The interatrial septum ran across the aneurysm dividing it into two unequal parts, one-third lying in the right atrium and two-

thirds in the left. The non-coronary cusp was thickened and calcified and, in the portion lying above the mitral valve, there was a perforation 1.7 cm. in diameter, surrounded by small crumbling vegetations. The other two aortic cusps were thickened and the commissure between them was calcified. Many small vegetations were present on the margins of all cusps. No abnormality was found in any other valve. The cavity of the left ventricle was moderately dilated and the ventricular wall slightly thickened. The other chambers appeared normal. Atheromatous changes were present in the coronary arteries but were not severe.

Histology. The wall of the aneurysm consisted of three layers; an outer layer of fibro-elastic tissue (the atrial endocardium), a middle layer of muscle, and a thick inner layer of fibrous tissue. The inner and middle layers were separated by small islands of elastic tissue which could be traced from the wall of the aneurysm into the elastic layer of the aortic media. Embedded in the inner layer were small areas of calcification. The aneurysm was partly lined by organizing thrombus which was sometimes covered by a thin layer of fibrous tissue. In the aortic adventitia adjacent to the aneurysm, there was a widespread, mainly polymorphonuclear, inflammatory reaction which had, at one point, spread into the aneurysm wall. Newly formed fibrous tissue extended from this area to the inner wall of the aneurysm and into the vegetations on the

FIG. 7.—Case 4. *From a drawing of the heart.* (A) *Aortic and left ventricular aspect.* The arrow indicates the aneurysm; across its mouth a strip of tissue represents the remains of the non-coronary aortic cusp, below this lies the large perforation of the cusp. (B) The remaining part of the anterior aortic cusp has been removed to reveal the extent of the aneurysm. The vegetations of bacterial endocarditis are visible in (A) and (B).

FIG. 8.—Case 4. *From a drawing of the atrial aspect of the aneurysm.* The atrial septum, which crosses the aneurysm, has been cut. The left atrium and the mitral valve (M) lie to the left of the cut edge; the right atrium and the tricuspid valve (T) to the right.

non-coronary cusp. The aortic media was entirely free from inflammatory changes. The normal architecture of the non-coronary cusp was greatly distorted and much calcification was present. The histology of the mitral valve was normal. Sections from the ventricular septum showed a diffuse, fairly acute, myocarditis.

The aneurysm might have developed owing to the bacterial endocarditis, or it could have been of congenital origin and the focus for the bacterial endocarditis. It is not difficult to understand how the bacterial endocarditis could have led to this aneurysm for the position of the inflammatory reaction in the aorta was such that it might have caused a separation of the annulus fibrosus from the aortic media, thus exposing the muscle of the atria, which would be stretched by the intra-aortic pressure to form the aneurysm. In another case of subacute bacterial endocarditis we have seen the first stage of this process, for the annulus fibrosus of the non-coronary aortic cusp had been separated from the aortic media by a gap of 2 mm.; through this gap blood had penetrated to form a hæmatoma between the annulus fibrosus and the atrial muscle. If this had occurred in the present case and the separation had extended to expose the atrial muscle, thrombosis would have occurred on the exposed

muscle surface and subsequent organization could have given rise to the fibrous lining of the aneurysm. The mode of formation thus resembles that in Case 3, but there the dissection started in the right coronary sinus and blood leaked into the loose subpericardiac tissues forming a hæmatoma. In the present case the leakage was limited by the close relationship of the non-coronary sinus to the atrial muscle, so a large hæmatoma could not form. We believe this to be the probable explanation of the histological structure of the aneurysm wall, so we do not suggest a congenital origin in this case. The absence of other congenital anomalies, which nearly always accompany congenital sinus aneurysms, supports this conclusion. There was no proof of an old lesion of the aortic valve upon which the bacterial endocarditis could have arisen, but the valve was so greatly deformed by the infection that old rheumatic disease cannot be excluded.

Cases 3 and 4 illustrate the difficulty that may be encountered in deciding the ætiology of sinus aneurysms, and emphasize the importance of adequate histological examination in these cases.

DISCUSSION

Aneurysms of the aortic sinuses may be divided into two groups; acquired (so-called "spontan-

eous") aneurysms, which arise in a diseased aorta, and aneurysms due to developmental defects. Since there appear to be important pathological, radiological, and clinical differences between the two types, which have not usually been described separately, we have considered it worthwhile to compare verified cases from both groups including our own cases and have collected 25 reported cases of congenital sinus aneurysms proved at necropsy, together with 22 acquired aneurysms confined to the aortic sinuses and confirmed pathologically. We have not included acquired aneurysms reported prior to 1914 since these were often inadequately described and the ætiological importance of syphilis was not appreciated. The sources of these cases are given below.

Congenital Aneurysms

*R.C.S.** Thurnam (1840); Beck (1842); Rickards (1881); Charteris (1883); Livingston (1883); Cayla (1885); Kryzwicki (1889); White (1892); Kraus (1902); Hart (1905) 3 cases; Eppinger (1916); Abbott (1919); Jacobi and Heinrich (1933); Hirschboeck (1942); King (1942); Macleod (1944); Our cases 1 and 2.

*N.C.S.** Goehring (1920); Laederich and Pomeau-Delville (1928); Duras (1944); Kawasaki and Benenson (1946); Herson and Symons (1946).

Acquired Aneurysms

*R.C.S.** Smith (1914) Cases 1 and 2; Noack (1919); Scott (1924) Case 1; Sheldon (1926); Abbott (1932); Benson, Hunter, and Manlove (1933) Case 1; Snyder and Hunter (1934) Case 2; Schuster (1937); Hamilton-Paterson and Castleden (1942) Case 3; Our case 3.

*N.C.S.** Marty and Froncin (1924); Norris (1932); Higgins (1934); Wright (1937); Our case 4.

*L.C.S.** Scott (1924) Case 2; Abbott (1932); Benson, Hunter, and Manlove (1933) Case 2; Snyder and Hunter (1934) Case 1; Ostrum, Robinson, Nichols, and Widman (1938) Case 5; Chipps (1941).

MORBID ANATOMY

The figures below refer to the number of cases in which the feature mentioned has been recorded; since the descriptions are often insufficient, the figures are necessarily incomplete.

Congenital Aneurysms (25)

Sinus involved: *R.C.S.* 20; *N.C.S.* 5; *L.C.S.* 0.

Origin from sinus: Sac with round or oval opening 0.5 to 1.5 cm. sinus not dilated, 12. Whole

sinus dilated, no separate aneurysmal sac, 2. Both dilated sinus and aneurysmal sac, 2.

Position of opening in sinus: *R.C.S.* right half, 3; central, 2; left half, 4. *N.C.S.* Adjacent to *R.C.S.*, 3; central, 2.

Size: None larger than 4 cm. diameter.

Shape: Globular, unless ruptured; then collapse, unless thickened, to form fistulous channel 1.5 to 2.5 cm. long.

Walls: Thin, often transparent, but occasionally thickened.

Cardio-aortic fistulæ: *R.C.S.* 17 led to fistulæ; 13 to conus of right ventricle, 2 to right atrium, 1 to left ventricle, 1 to pulmonary artery. Fistulæ due to rupture of aneurysm, 12; probably present from birth, 5. *N.C.S.* 4 ruptured into right atrium. No congenital aneurysm ruptured outside heart.

Encroachment on intracardiac structures: *R.C.S.* Pulmonary valve, 6; tricuspid valve, 5; ventricular septum, 2; right ventricle, 1; both ventricles, 1. *N.C.S.* Right atrium, 5; tricuspid valve, 3; conducting bundle, 1.

Aortic incompetence: *R.C.S.* 4.

Associated lesions: Congenital: 23. Anomalies of aortic cusps, 15 (bicuspid, 6; much enlarged cusp, 4; rudimentary cusp, 1; thickened cusp, 4; calcified cusp, 2; fenestrated cusp, 2). Subaortic stenosis, 2. Abnormal extension of annulus fibrosus, 2. Ventricular septal defects, 10 (bulbar, 3; probably bulbar, 6; ruptured ventricular septal aneurysm, 1). Coarctation of the aorta, 2. Pulmonary conus stenosis, 1. Single coronary artery, 1. Patent foramen ovale, 1.

Acquired: Subacute bacterial endocarditis, 6. Rheumatic heart disease, 1.

Cardiac enlargement: Myocardial hypertrophy, 20; enlargement both ventricles, 14; mainly left ventricle, 4; mainly right ventricle, 4.

Heart weight: Less than 300 g., 3; 350 to 450 g., 2; 500 to 650 g., 5.

Summary. Congenital sinus aneurysms have been confined to the right coronary sinus and the adjacent two-thirds of the non-coronary sinus. They are always small but, owing to their thin walls, commonly rupture to form cardio-aortic fistulæ (21 of 25) usually communicating between the right coronary sinus and the right ventricle, or between the non-coronary sinus and the right atrium. They remain entirely intracardiac and do not affect extracardiac structures nor rupture outside the heart, but frequently cause disturbance of intracardiac structures, especially the pulmonary

* *R.C.S.* Right coronary sinus. *N.C.S.* Non-coronary sinus. *L.C.S.* Left coronary sinus.

FIG. 9.—Formation of the bulbar septum. Fusion of the aortico-pulmonary and the proximal and distal bulbar septa occurs last at the areas represented as gaps. The suggested defects which may arise at these sites are indicated on the right.

FIG. 10.—Formation of the aortic cusps. Growth of the cusps indicated by interrupted lines. Only two cusps can be related to the bulbar septum.

valve, which is often interfered with by right coronary sinus aneurysms, and the tricuspid valve which may be encroached upon by aneurysms arising in either sinus. Congenital sinus aneurysms are nearly always associated with other developmental faults, usually anomalies of the aortic cusps or bulbar ventricular septal defects. Apart from bacterial endocarditis, acquired heart disease has occurred in only one case.

Acquired Aneurysms (22)

Ætiology: Associated with syphilis, 17; with bacterial endocarditis, 4; dissecting, 1; atheromatous, 1.

Sinus involved: R.C.S. 11. N.C.S. 5. L.C.S. 7.

Size: Often very large, e.g. admitting two fists.

Rupture: R.C.S. 7 (right ventricle, 1; pulmonary artery, 2; pericardium, 1; left pleural cavity, 1; externally, 2). N.C.S. 3 (right atrium, 2; pericardium, 1). L.C.S. 1 (pulmonary artery).

Encroachment of intracardiac structures: R.C.S. Pulmonary artery, 1; right coronary artery, 4; tricuspid valve, 1; septum and conducting bundle, 1. N.C.S. 0. L.C.S. Pulmonary artery, 2; left atrium, 3; left coronary artery, 2; left ventricle, 1; mitral valve, 2.

Associated lesions: Aortic incompetence, 8 (syphilitic, 6; bacterial endocarditis, 2). Bicuspid aortic valve, 2.

Summary. Acquired sinus aneurysms may arise from any of the aortic sinuses. Owing to their large size they tend to extend upwards, often becoming extracardiac and rupturing outside the heart. Cardio-aortic fistulæ were present in only 6 cases. They encroach upon intracardiac structures less often than congenital aneurysms. Congenital cardiac defects were present in only two cases, but acquired heart disease was invariably present, usually syphilis or bacterial endocarditis.

EMBRYOLOGY

Congenital sinus aneurysms are believed to arise from defective development of the distal bulbar septum. This structure separates the systemic and pulmonary halves of the bulbus cordis, the primitive exit tube of the heart. It arises by endothelial outgrowths from each side of the bulbus cordis, and is completed by fusion distally with the aorto-pulmonary septum and proximally with the proximal bulbar septum, which represents the bulbar part of the ventricular septum (Fig. 9).

Congenital weaknesses are likely to develop at points of fusion, that is, distally, where the distal bulbar septum fuses with the aorto-pulmonary septum, between the two halves of the distal bulbar septum, and proximally where the distal meets the proximal bulbar septum (Fig. 10). It is reasonable to suppose that congenital aorto-pulmonary fistulæ

develop at the distal site, aortic sinus aneurysms and cardio-aortic fistulæ at the intermediate site, and bulbar septal defects at the proximal site of fusion.

Although all the aortic sinus aneurysms collected by Abbott (1919) arose from the right coronary sinus, aneurysms undoubtedly arising from the non-coronary sinus have been subsequently reported, but there is no reported instance of a congenital aneurysm arising from the left coronary sinus. From Fig. 10 it is clear that only two sinuses can be related to the distal bulbar septum, so it is not surprising that these aneurysms are confined to the right coronary and non-coronary sinuses.

Micks (1940) collected three cases of aneurysmal dilatation of all three aortic sinuses and added a new case; Brown (1939) also mentions a similar case. Micks tentatively suggested that such cases might be of developmental origin, but since neither syphilis nor cystic medial necrosis was excluded by adequate histological examination in any of these cases, we do not regard their developmental origin as proven.

CLINICAL FEATURES OF CONGENITAL SINUS ANEURYSMS

Patients with congenital sinus aneurysms are usually male (19 of 24). Signs and symptoms might arise from (a) a cardio-aortic fistula, (b) interference with intracardiac structures, (c) other congenital defects, or (d) bacterial endocarditis.

Cardio-aortic fistulæ. A defect in the wall of an aortic sinus gives rise either to a congenital cardio-aortic fistula or to aneurysmal dilatation of the sinus that leads to a cardio-aortic fistula by rupture later in life; signs and symptoms due to the fistula may therefore either be present from birth or appear suddenly in later life.

Congenital cardio-aortic fistulæ. Five of the twenty-one fistulæ are believed to be congenital. In one case death occurred from pulmonary atelectasis ten days after birth and in another enterocolitis led to death at four months old; the aneurysms were incidental necropsy findings. In two other cases cardiac symptoms were minimal until two months and three years prior to death from heart failure at ages of 30 and 31; both patients had multiple congenital cardiac lesions including septal defects. The remaining patient never had cardiac symptoms, and death at age of 53 followed two weeks illness of unknown nature associated with severe vomiting and abdominal pain. Thus, death was due to intercurrent disease in three cases, and in the other two cases cardiac symptoms did not appear until the end of the third decade.

Cardio-aortic fistulæ due to sinus aneurysm rupture. Eppinger (1916) has described the event of rupture. A man of 43, previously fit and active, was climbing a mountain and bent down to lift a heavy stone; when about to cast this aside he was seized by a sudden pain in the chest and immediately noticed a "whirring" sensation in the mid-sternal area "like a half-filled bottle being shaken" in the chest. These sensations gradually diminished in severity and he was soon able to lead the way down the mountain. Eight days later he noticed difficulty in breathing and palpitation and soon became unable to walk more than a few steps; he was confined to bed and treated with digitalis without improvement; he developed œdema and died nine months later.

Of 25 congenital aneurysms, 15 are believed to have ruptured during life; in 6 cases death followed within five weeks and 6 died between two and thirteen months. Of the remaining 3 cases, one died four years after rupture due to further tearing of the aneurysm, a second died nine years later from bacterial endocarditis, and the third (our Case 2) survived seventeen years with intermittent failure, eventually dying with lobar pneumonia.

It therefore appears that the heart tolerates a congenital cardio-aortic fistula much better than its sudden development in later life, which usually leads to progressive heart failure, death occurring within a few weeks in over one-third of cases and within about a year in four-fifths of cases; in exceptional cases symptomatic recovery takes place, death being due to a second rupture or to intercurrent disease.

The physical signs of a cardio-aortic fistula are an important feature of congenital sinus aneurysms; for in only 4 of 25 cases was the aneurysm wall intact at death; in 13 cases the fistula led to the right ventricle, in 6 cases to the right atrium, in 1 case to the left ventricle, and there was an aortopulmonary fistula in the remaining case.

Both patency of the ductus arteriosus and aortopulmonary communications (Shepherd, Park, and Kitchell, 1944) give rise to a systolic-diastolic murmur at the base of the heart; and a collapsing pulse. But a communication between the greater and lesser circulations lying below the valve cusps, as in ventricular septal defect, causes only a systolic murmur. In cardio-aortic fistulæ one end of the fistula lies above the cusps, and one below, but there is no doubt that the associated murmur usually extends into diastole. This was so when there was no other congenital lesion (our Case 2) or only congenital lesions associated with systolic murmurs (10 cases). The character of this systolic-diastolic murmur is striking, for it is often described as superficial, simulating pericardial friction, and

nearly always coarse, harsh or rasping in character, accompanied by a thrill, usually systolic, but in Abbott's (1919) case there was a diastolic thrill so intense that it could be felt through the bedclothes. The systolic and diastolic components may merge into each other to form a continuous murmur so that the cardio-aortic fistula murmur is more likely to be confused with the murmurs of patency of the ductus or of a congenital aorto-pulmonary communication than with the to-and-fro murmurs of acquired aortic valve disease. The cardio-aortic fistula murmur tends to be lower in position than other continuous systolic-diastolic murmurs, and it was definitely louder in the third or fourth spaces than in either the second space in 6 of 9 cases in which this is recorded. The physical signs are essentially similar whether the fistula leads to the right ventricle or the right atrium. In the case reported by Herson and Symons (1946) the physical signs are of exceptional interest for, at the age of 12, before rupture, there was only a loud musical systolic murmur, maximal at the inner ends of the fourth and fifth interspaces, presumably due to the associated ventricular septal defect; whereas after rupture at the age of 31 the typical systolic-diastolic murmur became audible.

(b) *Interference with intracardiac structures:* The aortic valve was often affected and was incompetent in 7 cases, probably due to distortion by the aneurysm in 4 cases. There do not appear to have been striking symptoms until the aneurysm ruptured or bacterial endocarditis developed. The tricuspid valve was involved in 8 cases. In only 2 was a clinical diagnosis of tricuspid disease made; in these cases a systolic murmur was heard at the lower end of the sternum and hepatic pulsation was present. In 2 others, the necropsy findings suggested significant interference with the tricuspid valve, but in all 4 the patients had apparently been well and active until their aneurysms ruptured thirteen months, one month, four and a half weeks, and ten days before death. The pulmonary valve cusps were pushed aside in 5 cases; in none of these was pulmonary valve disease suspected during life, although pulmonary incompetence was diagnosed in another case in which the valve was not affected.

Pressure on the conducting bundle was believed to have led to the complete heart block in one case and pressure on the A-V node was regarded as the cause of A-V nodal rhythm in the other case. Encroachment upon the cardiac chambers has often occurred, but there is no evidence that it has impaired cardiac efficiency.

(c) *Associated congenital cardiac defects.* If a cardio-aortic fistula is present, its signs and symptoms overshadow those of other less striking con-

genital lesions, which can be studied only in cases with unruptured aneurysms and by reviewing the history prior to rupture in other cases. In two cases with unruptured aneurysms no clinical details are available, and in the other two there were no symptoms until bacterial endocarditis developed in the one case and Stokes-Adams attacks in the other. Of 15 patients whose aneurysms ruptured there were no cardiac symptoms prior to rupture in 14, and in the remaining one the incapacity was due to bacterial endocarditis. Ventricular septal defects were present in 6 cases, aortic incompetence in 3 cases, and slight coarctation of the aorta in 2 cases. The congenital defects associated with aortic sinus aneurysms appear to be of an almost asymptomatic type.

(d) *Subacute bacterial endocarditis.* Of 6 unruptured aneurysms, 4 became infected; 2 of these subsequently ruptured. Of 13 aneurysms that ruptured during life only one subsequently became infected, and that nine years later. The short duration of life after rupture may account for the rarity of subsequent infection. None of the five congenital communications became infected. Age does not appear to be a factor in determining the preponderance of infection in unruptured aneurysms, for the average age at death was similar in all groups and bacterial endocarditis developed at ages from one and a half to 56 years.

CAUSES OF DEATH

Heart failure was the cause of death in 15 of 25 cases, and in 12 could be attributed to rupture, in 2 to congenital cardio-aortic fistulae, and in 1 to rheumatic heart disease associated with an unruptured aneurysm. *Bacterial endocarditis* led to death in 6 cases, and *intercurrent disease* was the immediate cause of death in 4 cases. Thus, nearly two-thirds of these patients died from heart failure, usually due to the cardio-aortic fistula, and one quarter from subacute bacterial endocarditis.

In this summary of the clinical features of congenital sinus aneurysms the signs and symptoms associated with a cardio-aortic fistula have been emphasized, for this complication is present in the majority of cases, dominates the clinical picture by its often dramatic onset and striking physical signs, and usually leads to heart failure which is the commonest cause of death. By comparison, the effects of local extension of the aneurysm and the signs of other congenital defects are unimportant.

RADIOGRAPHY

Congenital sinus aneurysms are usually small and almost always project into a cardiac cavity rather than externally. It is therefore not surprising

to find that, in six published cases in which radiological findings are given, and in our two cases, no evidence of localized aneurysmal swelling was observed. Roesler (1943) and Ostrum *et al.* (1938) have described small projections from the vascular pedicle, often evident only in the oblique views, in cases of sinus aneurysm, but we conclude that these observations relate to the acquired variety. In eight cases with radiological findings, the heart was enlarged in all. In seven of them a cardio-aortic fistula was present and probably responsible for the enlargement; in the remaining case a ventricular septal defect and aortic incompetence were present. The available evidence indicates that the enlargement involves both ventricles. The aortic shadow was not abnormal in four cases where this point is recorded.

ELECTROCARDIOGRAPHY

Only two electrocardiograms have been published, and the findings are described in three other cases. Including our two cases, we have information about the standard limb leads in only seven cases. In two cases right axis deviation was associated with signs suggestive of aortic incompetence, though actually due to the cardio-aortic fistula; some diagnostic significance was assigned to this association by Hirschboeck (1942) and Herson and Symons (1946) but it has not occurred in the other cases, left axis deviation being present in three cases. Complete heart block and auricular fibrillation have each occurred in one case. The electrocardiographic findings are so inconstant that they give little assistance in diagnosis.

DIAGNOSIS OF CONGENITAL SINUS ANEURYSMS

An unruptured congenital aortic sinus aneurysm is almost asymptomatic and without physical or radiographic signs, though the presence of congenital heart disease may be recognized by the signs of an associated lesion such as a bulbar septal defect. Unruptured sinus aneurysms are, however, very prone to become infected, as we have shown, and when bacterial endocarditis develops in a heart apparently previously healthy, this is one of the silent underlying lesions that may be suspected.

The situation is quite different when a cardio-aortic fistula is present, either congenital or due to the rupture of a sinus aneurysm, for this is associated with striking physical signs and, if due to rupture in later life, with a dramatic clinical history. Eppinger (1916) was able to make a correct clinical diagnosis in his case by comparing the event of rupture during effort and the physical signs that followed with the similar case described by Kraus (1902). Rupture of

a congenital sinus aneurysm is perhaps most likely to be confused with rupture of an aortic cusp, though this is usually associated with either syphilis or bacterial endocarditis. If these two diseases can be excluded rupture of a sinus aneurysm may justifiably be suspected, unless it is to be admitted that a healthy aortic cusp can rupture owing to exceptional trauma. In this differential diagnosis the physical signs will be helpful for the characteristic harsh, superficial, systolic-diastolic murmur, often accompanied by a thrill, maximal in the third left interspace near the sternum should differentiate a cardio-aortic fistula from aortic incompetence but the accompanying Corrigan pulse gives little help for it is present in both conditions.

It is more doubtful whether a precise diagnosis can be made when the cardio-aortic fistula is present from birth for the advantage of the characteristic history of rupture is lost. The nature of the murmurs and their presence from early life will suggest a congenital heart lesion but it will almost certainly be impossible to distinguish between an aortopulmonary fistula and a cardio-aortic fistula and the differential diagnosis from patency of the ductus arteriosus may be difficult. The more superficial and harsher murmurs, situated unusually low for that of a patent ductus, and the more striking Corrigan pulse, may lead to the suspicion of a cardio-aortic fistula, but these qualities are merely a matter of degree and it must be admitted that no reliable differential sign is available. Electrocardiography and radiography will give little help unless an enlarged pulmonary artery suggests patency of the ductus, for it has not occurred in aortic sinus aneurysms. Since surgical ligation of the ductus has become commonplace, it is fortunate that congenital cardio-aortic fistulae are rare.

SUMMARY

The nomenclature of the aortic sinuses is discussed and the topography of the aortic root illustrated.

Four cases with aortic sinus aneurysms, two congenital and two due to aortic disease, are described.

Including these cases, a verified series of 25 congenital and 22 acquired sinus aneurysms has been collected.

The pathological features of the condition are illustrated by an analysis of this series of cases.

The embryology of congenital sinus aneurysms is briefly discussed.

The clinical features of congenital sinus aneurysms are described and diagnosis is discussed. Attention is directed to the striking clinical features associated with a cardio-aortic fistula.

When a congenital sinus aneurysm ruptures in later life we may, from the combination of history and physical signs, expect to recognize the presence of cardio-aortic fistula; if a congenital cardio-aortic fistula is present the existence of a congenital heart lesion will be manifest and, from the physical signs we may suspect the presence of a congenital cardio-aortic fistula or an aorto-pulmonary communication. Finally, although we cannot hope to diagnose an unruptured aortic sinus aneurysm, it is, like a bicuspid aortic valve, one of the silent congenital lesions to be suspected when bacterial

endocarditis develops in a heart apparently previously healthy.

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