

Rep  
ded

## MCGILL UNIVERSITY PUBLICATIONS

SERIES VIII. (MEDICINE).

No. 2.

Clinical and Developmental Study of a Case of  
Ruptured Aneurysm of the Right Anterior  
Aortic Sinus of Valsalva

by

MAUDE E. ABBOTT, B.A., M.D.

Reprinted from "Contributions to Medical and Biological Research,  
dedicated to Sir William Osler, in Honour of His Seventieth Birthday,  
July 12, 1919, by his Pupils and Co-workers."

i  
s  
f  
c  
i  
a  
a  
s  
a  
h  
a  
ac  
of  
lil  
re  
du  
sep  
is  
(2)

Coc

Sir  
Put

CLINICAL AND DEVELOPMENTAL STUDY OF A  
CASE OF RUPTURED ANEURYSM OF THE RIGHT  
ANTERIOR AORTIC SINUS OF VALSALVA

LEADING TO COMMUNICATION BETWEEN THE AORTA AND BASE  
OF THE RIGHT VENTRICLE, DIAGNOSED DURING LIFE. OPENING  
IN ANTERIOR INTERVENTRICULAR SEPTUM (PROBABLY  
BULBAR SEPTAL DEFECT). MALIGNANT ENDOCARDITIS<sup>1</sup>

By MAUDE E. ABBOTT, B.A., M.D.,

Curator of the Medical Museums, McGill University, Montreal  
(From the medical wards of the Royal Victoria Hospital)

**A**BNORMAL communications between the aorta and base of the right ventricle, or between aorta and pulmonary artery, are of not very infrequent occurrence. The event is accompanied in the great majority of cases by characteristic symptoms, and signs so striking as to be pathognomonic. In a few of the cases the communication is of congenital origin, and the clinical evidences of its presence have existed throughout life; but in far the greater number it is formed by the sudden bursting of an aneurysm of the base of the aorta into the pulmonic circulation, and symptoms then set in suddenly. Such aneurysms may be of the so-called "spontaneous" type, in which the wall of the ascending aorta is extensively diseased, from luetic or other causes. There is, however, an extremely interesting group of cases in which the aortic wall is perfectly healthy, and the right anterior aortic sinus of Valsalva is the seat of an opening leading into a finger or thimble-like process which projects into the conus of the right ventricle and represents an aneurysm of its wall and that of the aortic sinus, due not to disease, but apparently to a congenital thinning of the septum between the two great trunks. This was evidently so, and is the explanation put forward in the cases of Beck, (1) Hale-White, (2) Krzywicki, (3) and Kraus, (4) in which the walls of the aneurysm

<sup>1</sup> The anatomical portion of the study of this case was made under a grant from the Cooper Fund for Medical Research.

Reprinted from "Contributions to Medical and Biological Research, dedicated to Sir William Osler, in Honour of His Seventieth Birthday, July 12, 1919, by his Pupils and Co-workers."

were thin and membranous, without sign of inflammatory action of any kind, and rupture had taken place at the apex of the sac, evidently as a result of strain. In the case of Krzywicki the ventricular septum was entire, but in the other three above cited, and in several similar ones in the literature (Hart, (5) Thurnam, (6)) in which the conditions were obscured (as in our own case) by the existence of a malignant endocarditis, the aneurysm of the right aortic sinus was associated with a defect of the anterior interventricular septum immediately below the cusp, in a situation identical with that of the interventricular communication in the case which it is our privilege to report here, and both conditions were ascribed by those who recorded them to a defective development of the aortic (bulbar) septum at this point.

The clinical features of abnormal communications between the aorta and pulmonary circulation were clearly outlined by Thurnam (6) in 1840, who reported 1 case of ruptured right aortic sinus aneurysm (summarized below), and mentioned 5 others within his knowledge in which a thin-walled membranous sac projected into the right ventricle, but was not ruptured; by Peacock (7) in 1868, who included Thurnam's series in a review of 17 cases from the literature, and by Brocq (8) in 1886. In all the cases except 4 cited by these authors the communication was "accidental," due to rupture of a "spontaneous" aneurysm of a diseased aortic wall; of the other 4, which were all believed to be of congenital origin, 2, 1 by Thurnam, (6) and 1 by Rickards, (9) were cases respectively of ruptured aneurysm of the right aortic sinus, and congenital opening in this situation. In the other two by Wilks, (10) and Baginsky, (11) a smooth-walled aperture of communication between the aortic and pulmonary trunks lies above the valves but below the origin of the innominate artery (thus excluding patent ductus), and represents a defect at the upper part of the aortic septum. (See Plate IV, S. a. p.) Similar cases of congenital perforation at this point above the valves were reported by Fräntzel, (12) 1868, Girard, (13) 1895, and Hektoen, (14) 1900, who adds a valuable developmental study of aortico-pulmonary communications of congenital origin. He quotes a case by Charteris (15) of much interest in relation to the subject of this article, in which a hole behind the right aortic cusp leads directly into the base of the right ventricle without any sign of aneurysm or inflammatory action, evi-

dencing a true defect at this point and not merely a congenital thinning. Rickards (9) reported a similar opening combined with ventricular septal defect (summarized below).<sup>2</sup>

The characteristic clinical picture presented may be summarized as follows: Dyspnoea without cyanosis, precordial vibration and thrill of intense purring character, and loud sawing murmur, sometimes systolic, sometimes diastolic, but usually continuous, with systolic or diastolic accentuation, and synchronous with the thrill. These signs are differentiated from those of patent ductus by their extremely superficial character relatively to the chest wall, and by their point of maximum intensity being definitely at the base of the heart and over the middle of the precordium. In the very rare cases of congenital perforation of the trunks (Wilks, Baginsky, Fräntzel, Girard, Hektoen) or of the right aortic sinus (Charteris, Rickards), these signs were of course persistent throughout life. In the far commoner ones of ruptured aneurysm, a sudden onset in a person in apparently perfect health is a part of the symptom-complex. The patient may survive the rupture for years, the precordial thrill and murmur persisting unchanged.

The following case, which presents a picture pathognomonic in all details of an aneurysm of the base of the aorta rupturing into the pulmonary artery or base of the right ventricle nine years before death, occurred in the public service of Dr. W. F. Hamilton, who kindly accorded to me the fullest privileges of observation during the nine months of the patient's stay in the hospital, and gave me the opportunity of making the ante-mortem diagnosis. The clinical notes were made by Dr. C. R. Joyce, house-physician in charge, with my own collaboration.

CASE. G. R., Englishman, aged thirty-six, intelligence above the average, never had chorea, rheumatism, or venereal disease, and had never used tobacco or alcohol. Admitted to the Royal Victoria Hospital, Montreal, on October 27, 1914, immediately upon his arrival in Canada from England, being referred by the Immigration authorities on account of his cardiac condition. Complained of slight dyspnoea, weakness, and sense of pulsation in the temples. Gave a history of having been in apparently perfect health, and leading a strenuous life following his trade

<sup>2</sup> Good examples of so-called "spontaneous" aneurysms rupturing into the pulmonic circulation with life maintained for some time are the cases by Roberts (20) and Gairdner (21).

as a carpenter, and also working as a Methodist preacher, until the age of twenty-seven, when he suddenly became aware of a feeling of cardiac irregularity and of throbbing in the temples. He took to bed under medical advice, and a month later was admitted unimproved to the Grantham Hospital, Surrey, England, where he remained in bed for nine months. Here *the diagnosis of aneurysm was made and the intense precordial vibration, which is perfectly evident to his own senses, is stated by him to have been first noted.* After his discharge he remained in bed at home for fifteen months longer, when the signs of cardiac insufficiency passed off, and he was able from this time until his sailing from England to follow his work as a Methodist preacher (though not his trade as a carpenter), without subjective symptoms. *The precordial vibration, however, had persisted to the present time.* He was extremely seasick on shipboard.

Examination showed a tall, spare, poorly nourished man, of flabby musculature, dyspnoic on exertion, without cyanosis, clubbing, or œdema. Posterior cervical, sub-maxillary, and axillary glands slightly enlarged. Marked pyorrhœa alveolaris. Temperature 100°, remittent type. Wassermann negative, hæmoglobin 65 per cent, R.B.C. 3,230,000, W.B.C. 12,300. Pulse 84, waterhammer. Marked pulsation in temples and vessels of neck. Capillary pulse.

*Heart.* Slight precordial bulging and widely distributed heaving impulse over whole precordium. Apex beat visible, precordial vibration and *very strong diastolic, almost continuous, thrill*, felt over the precordium, of *maximum intensity in second and third left interspaces*, where it is so strong that a vibration may be felt when the fingers are held half an inch away from the chest. This thrill is transmitted to the right nipple line, below to the seventh rib, above to the clavicle, and to the left to the midaxillary line. Slight presystolic thrill at apex, and systolic thrill in vessels of neck. Heart dullness at second space, 3½ cm. to right and 15½ cm. to left of midsternal line. At apex systolic and diastolic murmurs, transmitted to posterior axillary line, and a rough presystolic murmur. At the base a *very loud rough diastolic murmur almost continuous, being interrupted for only a brief time in systole with maximum intensity in third left interspace. This murmur is extremely superficial and may be heard with the ear 2 inches from the chest wall.* Another rough systolic murmur is heard best over pulmonary cartilage. To-and-fro murmurs behind from apex of left lung to level of fifth dorsal spine.

During the succeeding months the temperature became high and of septic type (98° to 105°), with occasional severe chills. Blood cultures on November 10th and December 21st negative. Weakness and dyspnoea became progressively worse, vomiting set in, albuminuria, hæmaturia,

and slight œdema developed, but never any cyanosis. The patient died March 5, 1915.

*Clinical Diagnosis.* Aortic insufficiency. Congenital or acquired communication (probably ruptured aneurysmal) between aorta and pulmonary artery or base of right ventricle. Malignant endocarditis, or endarteritis, about the margins of the communication.

The autopsy was done by Professor Horst Oertel, from whose report the following abstract is quoted, with his kind permission.

Body, 180 cm. long, of indifferent physique and poor nutrition. Skin pale, petechial hæmorrhages on thighs, œdema of legs. Chest bulging.

On opening thorax precordial area occupied largest part, goodly amount of clear yellow fluid in both pleural and peritoneal cavities. Pericardium contains 200 c.c. clear fluid, parietal pericardium free, thin, no pleuro-pericardial adhesions. Aortic and pulmonary valves the seat of an extensive productive ulcerative endocarditis which leads to the formation of massive polypoid vegetations and loss of substance on the cusps of the valves and on the right side on the parietal endocardium adjoining the valve. Two perforations exist, leading to communications between left and right sides of heart. The first takes its origin from the sinus of Valsalva of the middle aortic cusp, and forming an aneurysmal pouch or trumpet which projects into the right ventricle between the septal and middle cusps of the pulmonary, has perforated into the right heart. The second perforation exists immediately below this, and extends through the septum of the ventricles by an irregular ulceration of the musculature, immediately below septal cusp of the aorta to immediately below septal cusp of the pulmonary artery. A few small verrucose vegetations on mitral. Tricuspid quite free, also auricles. Marked hypertrophy of both sides of heart.

Lungs compressed, œdematous, and contain multiple hæmorrhagic infarcts. Congestion of ileum and œdema and congestion of large intestine. Cyanosis of liver. Spleen weighed 375 grams, enlarged, firm, dark red. Hæmorrhagic and productive nephritis.

A detailed description of the heart, which was handed over to the writer for further study, follows:

A heart greatly dilated and hypertrophied in its left ventricle and auricle, and also, though to a less extent, in its right chambers. Depth of left ventricle from base of anterior aortic cusps to apex 12 cm., thickness of wall 1.6 cm., thickness of right ventricle 6 cm. Pulmonary artery and orifice dilated, circumference at valves 8.5 cm. Circumference of aorta at same level 6.5 cm. Aortic and pulmonary cusps thickened and insufficient, and covered with recent vegetations which extend upon the



upper surfaces of the aortic cusps and upon the mural endocardium of the base of both ventricles, and are especially redundant in the conus of the right ventricle on its anterior and septal walls.

The right anterior<sup>3</sup> aortic sinus of Valsalva presents in its floor, below the orifice of the right coronary artery, a large orifice admitting the finger, which leads into a tubular trumpet-shaped process, 2 cm. long, which projects into the right ventricle between the septal and median pulmonary cusps, and represents a ruptured aneurysm of the sinus. The walls of this tubular canal are thick and tendinous, and are covered externally in the conus of the right ventricle by polypoid vegetations, and the free borders of its ruptured orifice are fringed with vegetations which have impinged upon the opposite (anterior) conus wall and have evidently led by secondary infection to the development of a great patch of vegetations 5 cm. square, which occupies this position. (See Plate II.)

Directly below the right anterior cusp in the left ventricle is seen a second opening leading into the conus of the right ventricle. It is of ovoid shape, about 1 cm. long, and has as its upper border the base and ventricular surface of the right aortic cusp, which on its aortic surface is continuous with and helps to form the floor of the trumpet-shaped aneurysm of the sinus. The lower and anterior borders of this inter-ventricular opening on the side of the left ventricle are rounded and muscular, but this is partly obscured by tendinous thickening and by a superimposed flattened film of vegetations. (See Plate I.) The posterior border of the defect lies 1.5 cm. anterior to the pars membranacea, that is, it is not in the situation usually occupied by interventricular septal defects, but lies in the extreme anterior or bulbar part of the inter-ventricular septum in the usual situation occupied by bulbar septal defects. Seen from the right ventricle this interventricular communication opens into the conus immediately below and to the left of the ruptured sinus aneurysm. Its edges are ulcerated and destroyed, and it is surrounded by masses of vegetations.

The left anterior aortic cusp is displaced downward 5 cm. below the left coronary, and an interval of 1 cm. exists between it and the right anterior cusp. This interval is filled with a mass of vegetations, and these have burrowed freely into the myocardium adjacent.

REMARKS UPON THIS CASE. *Clinical.* 1. As has been said, the diagnosis of a communication between the base of the aorta and

<sup>3</sup>A note upon the nomenclature of the aortic cusps is necessary. Gray, Morris, mention an anterior (right coronary) and two right and left posterior (left coronary) cusps. But that used by the older writers and in the German articles is right and left anterior (right and left coronary) and posterior (free) cusp, and it is this terminology that is followed here.

the pulmonary circulation, which was so obvious here, was based on the localization of maximum intensity of the continuous murmur and thrill over the middle of the precordium, and the intensity and extraordinary proximity of these signs to the chest wall, which differentiated it from patent ductus. The question as to the time the communication occurred, that is, whether it had been present throughout life (indicating a congenital perforation) or had existed for some years (pointing to a ruptured aneurysm), or was synchronous only with the present attack of malignant endocarditis, and due possibly to its ravages, was of equally great interest. In view of the intelligence of the patient, his emphatic assurance that the precordial vibration, which was perfectly evident to his sight and touch, had appeared at the time of his first cardiac breakdown, nine years ago, and had persisted since, was important. Since, however, the diagnosis of an aneurysm rupturing at this time depended on the accuracy of this statement, it was important to confirm it. Through the kindness of Professor Arthur Keith, London, the following facts were obtained from the Grantham Hospital, England. The patient was sent in by Dr. G. M. Shipman, and the entry relating to him in the hospital case-book read: "G. R., aged twenty-seven, admitted August 14, 1905. Discharged May 3, 1906. Disease, aneurysm of the aorta. Result, in statu quo." Dr. Shipman further wrote Dr. Keith "Re G. R.," that he "remembered the case of aneurysm of the aorta perfectly, and would look up some notes which he had about him." Owing to the war exigencies these notes have not yet been received, but the above information is sufficiently confirmatory of the patient's statements to make the diagnosis of aneurysm of the base of the aorta rupturing into the pulmonary circulation nine years before death a practical certainty.

2. The marked *diastolic* character of the almost continuous murmur was of interest. This was present also in several of the cases, but is not a constant feature, and may have had to do with the rough vegetations at the orifice of the trumpet-shaped tube through which the blood passed in diastole. The complexity of the other murmurs present is readily explained by the aortic and pulmonary insufficiency that existed, and the masses of vegetations that blocked the pulmonary conus.

3. The marked dyspnoea, without cyanosis, was confirmatory

evidence that the course of the blood through the ruptured aneurysm and ventricular communication was from the left to the right heart, owing to the fact that the pressure is physiologically highest in the left (systemic) ventricle. In such cases the volume of blood in the pulmonary circulation is necessarily increased. An effort was made to obtain confirmation of this by determining the basal carrying tension of the alveolar air for oxygen according to the method suggested by Plesch, (16) but no results were obtained owing to inadequacy of apparatus.

4. The youth of the patient at the first onset of symptoms, together with the complete absence of luetic history, suggested a traumatic or congenital origin of the ruptured aneurysm. He gave a history of striking his chest violently over the precordium against a pointed instrument (semaphore) at the age of four, and this trauma was insisted upon as a cause by himself and by his father, who wrote us several letters. The idea was discarded by me as improbable, in favour of a probably congenital origin with rupture as a result of strain.

5. The septic temperature and chills, which grew progressively worse, pointed definitely to an acute infective process which was believed by me to be secondary to the ruptured aneurysm.

*Pathological Anatomy.* The pathological interest of this specimen lies in: (1) the ruptured aneurysm of the right aortic sinus of Val-salva; (2) the interventricular communication directly below the same aortic cusp; (3) the marked thickening of aortic and pulmonary valves and evidences in the great size and thickness of the left ventricle of an aortic insufficiency of long standing; (4) the extravagant vegetations of malignant endocarditis, which cover the aortic and pulmonary cusps, surround the margins of the inter-ventricular communication, especially in the right ventricle, fringe the borders of the ruptured aneurysm, and cover the opposite wall of the right ventricle.

The presence of such an extensive acute infective endocarditis, together with the evidences of an old inflammatory process, obscures the nature of the two openings into the right ventricle somewhat, and led to a difference of opinion at the autopsy as to their nature—their irregular ulcerated outline, especially on the side of the right ventricle, and their encrustment with vegetations suggesting a

C  
E  
L  
  
V.  
O.  
be  
tri  
tu  
th

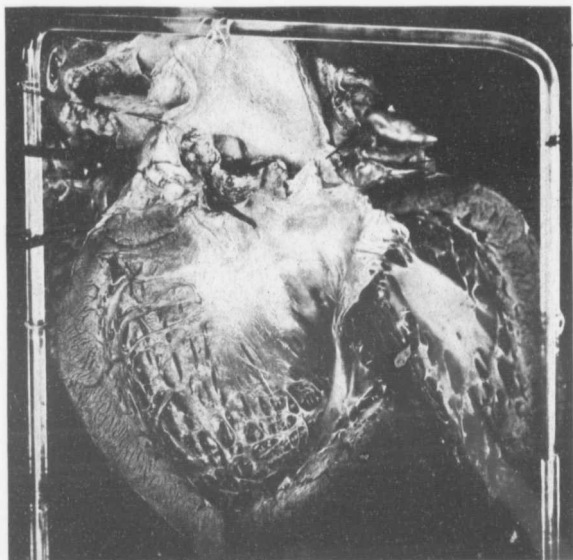


FIG. 1. ANEURYSM OF RIGHT AORTIC SINUS OF VALSALVA, RUPTURING INTO RIGHT VENTRICLE AT LEVEL OF PULMONARY VALVES.

Associated with Opening in Anterior Part of Interventricular Septum Leading into Conus Arteriosus of Right Ventricle (probably Bulbar Septal Defect). Malignant Endocarditis of Aortic and Pulmonary Valves and Adjacent Endocardium. View from Left Ventricle.

The Aortic Cusps are seen Thickened and Deformed by Old Endocarditis and Surmounted by Recent Vegetations with Ulceration of Subjacent Myocardium. Two Probes are Passed, the Upper through the Opening in the Right Aortic Sinus, the Lower through the Interventricular Communication Immediately below the Cusp, and are both Seen Emerging Close Together in the Conus Arteriosus of the Right Ventricle. The Interventricular Communication is Seen to Lie in the Anterior Part of the Septum Some Distance in Front of the Pars Membranacea, and Directly below the Anterior Half of the Right Aortic Cusp in the Extreme Anterior (Bulbar) Part of the Interventricular Septum. From the case here reported.



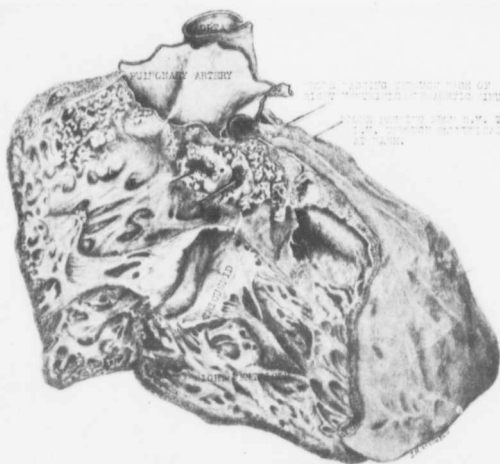


FIG. 2. VIEW FROM RIGHT VENTRICLE OF THE CASE SEEN IN FIG. 1.

Showing Probes passing through (a) Trumpet-Shaped Tube,\* in the picture, Fringed with Vegetations (the Ruptured Aneurysm of the Sinus Valsalva) Projecting into the Conus Arteriosus Directly below the Junction of the Posterior and Left Anterior Pulmonary Cusps; and (b) the Communication lying also in the Conus directly subjacent of the Septal Defect with the Left Ventricle; (c) Vegetative and Ulcerative Endocarditis of the Pulmonary Valves and of the Mural Endocardium Adjacent and Opposite to the Ruptured Aneurysm; (d) Dilatation of the Pulmonary Artery.

*From a drawing by Dr. J. H. Atkinson.*

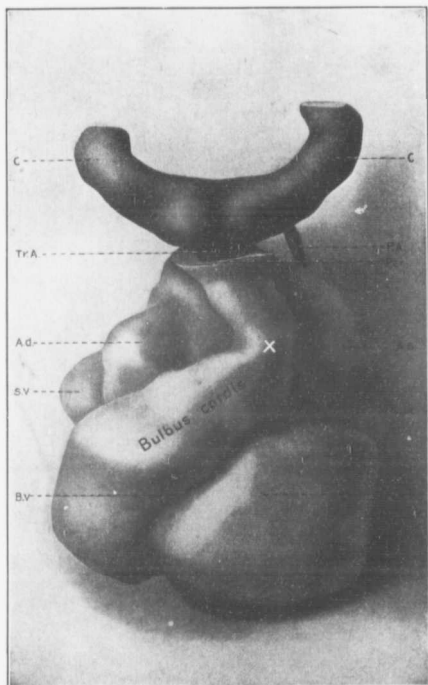


FIG. 3. MODEL OF THE HEART OF A HUMAN EMBRYO 4.6 MM. LONG X 108, TO SHOW THE RELATION OF EMBRYONIC BULBUS CORDIS TO VENTRICLE AND AORTIC ARCHES.

(The Division of the Bulbus Cordis into Aorta and Pulmonary Artery begins a little above the level of the Bulbo-Ventricular Cleft (B.V.), and from this point to the level of the Aortic Semilunar Cusps the "Bulbar" Septum Forms the Anterior Part of the Interventricular Septum of the Heart.) C., Carotid Arch; P. A., Pulmonary Artery; Per., Pericardium; Tr. A., Truncus Arteriosus; A. d., Right Auricle; A. s., Left Auricle; A. v., Common Auriculoventricular Orifice; B. v., Bulboventricular Cleft; V., Common Ventricle. Model by F. T. Lewis and M. E. Abbott, (Dr. Begg's Embryo).

(From the Anatomical Laboratory of the Harvard Medical School.) Republished from Osler and McCrae's "System of Medicine," 2nd Edition, 1915, p. 324.

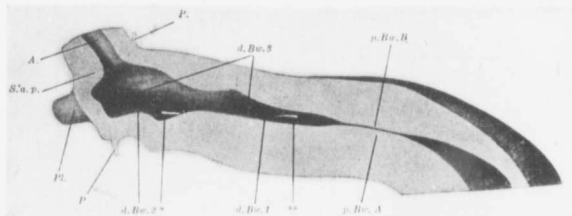


FIG. 4. LEFT HALF OF MODEL BY JULIUS TANDLER OF THE BULBUS CORDIS OF THE EMBRYO II, DIVIDED LONGITUDINALLY.

Showing Stage of Development in which the Distal Bulbar Swellings 1 and 3 (from which the Aortic and Pulmonary Cusps Originate) have Fused to Form the Distal Bulbar System, the Proximal Bulbar Swellings (*p. Bw. A, B.*) have Fused to Form the Proximal Bulbar Septum, and the Septum Aortico-pulmonale (*S.a.p.*) has Grown Downward for a Short Distance, Leaving two Points of Communication between the Aortic and Pulmonary Trunks, Immediately above and below the Distal Bulbar Septum (Aortic and Pulmonary Cusps). The Sound in the Picture has Disappeared in the Lumen of the Pulmonary Artery and Reappears in the Common Lumen above and below the Distal Septum at these Two Points of Communication. These Points thus Correspond in Location to the Openings above and below the Right Aortic Cusp Seen in Case Reported.

"A., Aorta (4th Aortic Arch); D. Bw. 1-3, Distal Bulbar Swellings; P, Attachment of Pericardium; p. Bw. A, B, Proximal Bulbar Swellings A, B; Pl., Pulmonary Artery (Sixth Aortic Arch); S.a.p., Septum Aortico-pulmonale;\*, point at which the sound of the Lumen of the Pulmonary Artery disappears, being covered by the Fusion of the Distal Bulbar Swellings 1 and 3, forming the Distal Bulbar Septum;\*\*, point at which the sound again appears in the Common Lumen. The subdivision of the Common Efferent Tube is produced distally by the Septum Aortico-pulmonale, in the middle region by the Distal Bulbar Septum, and Proximally by the Proximal Bulbar Septum. Between these three portions of the partition there are two points of communication, in which the ends of the sounds are visible."

From Keibel and Mall's "Embryology," Vol. II, Fig. 384, p. 552.



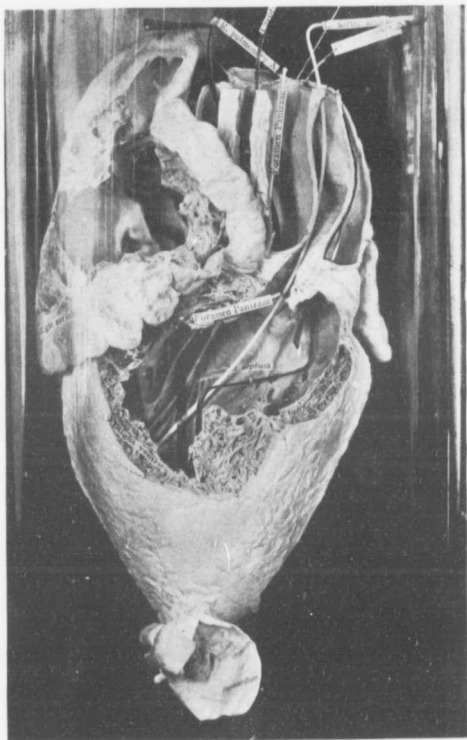


FIG. 5. HEART OF ALLIGATOR MISSISSIPPIENSIS.

View Showing Right Chambers, Pulmonary Artery and Left Aortic Arch (Arising Anteriorly from Right Ventricle), also Right Aortic Arch (Arising Posteriorly from Left Ventricle).

The Foramen Panizzæ lies behind the Posterior Cusp of the Left Aorta (in right ventricle), and Communicates with the Right Aorta just behind its Anterior or Right Coronary Cusp (in left ventricle) (which is the Location of the "Aneurysm of the Right Aortic Sinuses of Valsalva"). It is marked by a white rod. The Right Aortic Arch is marked by a dark rod. It slants obliquely upward from left to right and gives off in its course the Carotid Arch.

From a specimen in Professor Huntington's Anatomical Museum, College of Physicians and Surgeons, Columbia University, New York.

possible inflammatory origin. The clinical history of the case, however, pointing to a ruptured aneurysm of nine years' standing; the finger-like character and position of this aneurysm, identical in all respects with the cases reported, in which an exactly similar tube with thin membranous walls and without any sign of inflammation projected into the right ventricle in an exactly similar situation; the location of the ventricular communication directly below the same aortic cusp and having this cusp as its upper border; its situation in the anterior upper part of the ventricular septum and opening into the conus of the right ventricle in the typical situation for bulbar septal defects (cases reported by Tate, (17) Coupland, (18) Rolleston (19)); its shape and character as seen from the left ventricle, where the ravages of the malignant endocarditis are less serious; above all, the combination of these two conditions, namely, aneurysm immediately above and ventricular communication immediately below the right aortic valve, as occurred in the other cases recorded in which there was no sign of inflammation and in which a congenital origin was concluded, led the writer to conclude that both openings are here of congenital origin, due to a thinning above and a defect below of the bulbar septum between the two great trunks which, in the submergence of the embryonic bulbus cordis, forms the extreme upper and anterior part of the interventricular septum, the congenitally thin wall of the right aortic sinus yielding under the pressure of the circulation and forming the trumpet-shaped pouch which projected into the right ventricle and ruptured as a result of strain. The great redundancy of the vegetations in the pulmonary conus supports the view of their secondary origin about the defects, for it is not usual for endocarditis to develop in this situation without determining cause.

*Development.* A study of the development of the aortic and anterior part of the interventricular septum gives striking confirmation to the view that both sinus aneurysm and septal communication are due to a defective development of the embryonic bulbar septum. As is well known, in the earlier stages of the embryonic heart, the arterial trunk or efferent tube consists of the muscular bulbus cordis of the ventricle, which gives off the embryonic aortic arches from its upper border (see Plate III), and is lined by endocardial swellings spirally arranged (which persist in some of the

fishes as rows of valves). As division of the heart proceeds by development of its septa the division of the primitive arterial trunk takes place in the part derived from the bulbus cordis by fusion of the "proximal" and "distal" pairs of bulbar swellings, and at the extreme distal end by a septum growing down from above between the aorta and the pulmonary artery. The distal bulbar swellings correspond to the site of the future aortic cusps. There thus exists a stage in the development of the aortic septum in which the septum aortico-pulmonale is growing downward from above, the distal bulbar swellings have united in the middle and the proximal bulbar swellings are united below, leaving two apertures where the arterial trunk is still common immediately below and above the future aortic and pulmonary cusps. This stage of development has been reconstructed by Julius Tandler and is figured in Keibel and Mall's "Embryology," 2d edition, Fig. 384, and the plate is reproduced in this article. The probe passes from the lumen of the pulmonary artery through two apertures which occupy the exact location of the defects seen in the cases of aneurysm of the right aortic sinus of Valsalva and the anterior interventricular septal defects in the cases described by Hale-White, Thurnam, Kraus, Hart, and in the specimen which forms the subject of this paper.

*Comparative Anatomy.* A study of the hearts of reptiles and amphibians, made for confirmatory purposes in Professor Huntington's Collection at Columbia University, New York, elicited the extremely interesting information that this aperture above the anterior aortic cusp of the right aorta in the crocodile remains permanently open and allows the aerated blood from the right aortic arch (which arises from the left ventricle and supplies the systemic circulation), to pass into the left aortic arch (which arises from the right ventricle and receives unaerated blood) through an aperture above its posterior cusp. This is the so-called foramen Panizzæ of the crocodile (see Plate V), and the aneurysm of the right aortic sinus would seem to represent a persistence of the same opening. It was seen in the human subject not as an aneurysm, but as a complete defect in the cases reported by Rickards and Charteris. This view is confirmed by a perusal of the comprehensive studies by Langer (22) and Greil (23) upon the developmental changes in

the reptilian bulbus cordis in successive stages, which gives interesting confirmation, from the field of comparative embryology, of Tandler's reconstruction.

*Summary of Literature.* For purposes of comparison the cases in the literature of congenital communications, or aneurysm, of the right aortic sinus of Valsalva into the right ventricle, are herewith briefly summarized.

- I. Thurnam (7) (1840). "Ruptured Aneurysm of Right Aortic Sinus of Valsalva. Malignant Endocarditis." Aneurysm projecting into and communicating by two rounded openings with base of right ventricle; recent endocarditis; pulmonary, aortic, and mitral insufficiency; hypertrophy and dilatation of heart.

Male, aged thirty-three. Rheumatism at twenty. Sudden onset while in perfect health, with sense of "cracking" in heart region, of faintness, palpitation, dyspnoea, hæmoptysis, anasarca, extraordinarily superficial continuous sawing murmur with tremor intense in left second interspace. Death eleven weeks later.

- II. Beck (2) (1842). "Ruptured Aneurysm with Interventricular Communication." Right aortic valve calcified, sinus enlarged, presenting round opening leading into collapsed sac-like glove-finger three-quarters inch long, projecting into right ventricle between healthy pulmonary valves. Three rounded openings in tip and sides. Below it, interventricular communication admitting goose-quill, believed by author to be congenital.

Male, aged thirty-one. Palpitation on exertion always. Signs of cardiac insufficiency and marked dyspnoea three years, anasarca developed. Very superficial sawing murmur with tremor, loudest in diastole but continuous, with maximum intensity at base of heart.

- III. Rickards (1) (1881). "Congenital Communication in Right Aortic Sinus with Right Ventricle. Septal defect." Right and left anterior aortic valves congenitally fused, behind right half large round orifice with smooth membranous funnel-shaped walls opening into right ventricle between healthy pulmonary cusps. Immediately below same cusp circular aperture in septum with smooth membranous walls passing into conus of right ventricle. Both openings considered by author congenital.

Male, aged thirty. Precordial discomfort and dyspnoea always. Loud, rough, double murmur practically continuous, systolic element loudest, maximum between third cartilages, intense purring double precordial vibration. Epistaxis, hæmoptysis.

IV. Charteris (15) (1883). "Congenital Communication in Right Aortic Sinus with Right Ventricle." Immediately behind right aortic valve rounded opening with firm margins leading into right ventricle. Patch of endocardial thickening on opposite wall of right ventricle.

Male, aged fifty-three. Died from cardiac insufficiency. Systolic murmur most distinct at apex.

V. Krzywicki (4) (1889). "Ruptured Aneurysm of Right Aortic Sinus." In sinus oval opening with tendinous borders leading into thimble-shaped cavity 2.5 cm. deep, extending into right ventricle with thin, almost transparent, membranous covering formed of very delicate connective tissue with endocardial covering derived from right ventricle. Bean-shaped perforation at apex.

Female, aged twenty. Nine months before death pleurisy, precordial pain, palpitation, anasarca. Systolic murmur over base.

VI. Hale-White (3) (1891). "Ruptured Aneurysm of Right Aortic Sinus with Septal Defect." Septum deficient just below right aortic valve, ovoid opening margins thickened, septum for three-quarters inch around thin, semitransparent. Sinus above valve, thin-walled pouch which bulged three-quarters inch into right ventricle with aperture at bottom. Two patches of endocardial thickening on wall of right ventricle opposite defects.

Male, aged fifteen. Symptoms four months, dyspnoea, oedema, precordial vibration, rasping to-and-fro murmurs simulating pericardial friction, maximum at third interspace.

VII. Kraus (5) (1902). "Ruptured Aneurysm of Right Aortic Sinus. Interventricular Communication. Old Endocarditis." Wall of right aortic sinus thinned and membranous, pushed into right ventricle just below pulmonary valves as sac 2.5 cm. long, carrying on anterior surface diverticulum with delicate walls, perforated at apex. Walls of sac smooth and glistening. Semilunar opening in septum just below right aortic cusp. Endocardium surrounding this extensively scarred.

Male, aged twenty-seven. Symptoms four years, onset after severe muscular strain. Dyspnoea, palpitation, cyanosis, anasarca, hæmoptysis. Marked systolic thrill and coarse, long systolic murmur almost continuous, with short, hissing, roaring diastolic element, maximum at second and third interspaces.

VIII. Hart (6) (1905) (Case III). "Ruptured Aneurysm of Right Aortic Sinus. Septal Defect. Malignant Endocarditis." Semilunar opening in septum just below right aortic cusp, opening in conus of right ventricle. Just above it, sacculation, size of walnut, of right aortic sinus with extremely transparent membranous wall reaching forward

into pulmonary conus in semicircular form just between right pulmonary cusps. Fibrous ring below aortic valves. Malignant endocarditis of margins of septal defect, left pulmonary cusp, and fibrous ring, considered by author secondary to septal defect and sinus aneurysm of congenital origin.

- IX. Author's Case (1919). "Ruptured Aneurysm of Right Aortic Sinus. Interventricular Communication (believed to be Bulbar Septal Defect). Malignant Endocarditis."

Male, aged thirty-six. Onset of symptoms nine years before death with dyspnoea, precordial vibration and sawing, continuous murmur with diastolic accentuation. Cardiac efficiency re-established after two years' illness, but physical signs persisted. Second cardiac breakdown with malignant endocarditis nine months before death.

### Conclusions

1. A case is reported of aneurysm of the right aortic sinus of Valsalva rupturing into the right ventricle associated with interventricular communication and malignant endocarditis.

2. The presence of the characteristic physical signs of communication between the base of the aorta and the pulmonary circulation, namely, coarse precordial vibration and continuous sawing murmur with diastolic accentuation superficially placed, and with maximum intensity over the second and third interspaces, justified this diagnosis, which was made during life.

3. The clinical evidence accumulated was sufficient, in the judgment of the writer, to warrant the conclusion that the aneurysm ruptured into the right ventricle nine years before death. During this time the patient maintained a moderate degree of cardiac efficiency, showing that communication between the two circulations is compatible with life for a long time.

4. The aneurysm of the right aortic sinus and the interventricular communication are believed by the writer—on the ground of their location immediately above and below the right aortic cusp, the shape and appearance of the septal opening seen from the left ventricle, the evidence of identical cases in the literature unassociated with malignant endocarditis, and the facts of development and comparative anatomy,—to be of congenital origin, and due to defective development of the bulbar septum between the aortic and pulmo-

nary trunks which forms the upper anterior part of the ventricular septum in the submergence of the bulbus cordis.

5. There is a stage in the development of the aortic septum at which two such apertures exist above and below the "distal bulbar septum" which is the site of the aortic and pulmonary valves. The upper of these apertures remains permanently open in the crocodile as the foramen Panizzæ, which is thus the homologue of the aneurysmal thinning or congenital opening which occurs in the right aortic sinus of Valsalva.

6. The malignant endocarditis, which is so extensive in this case as to obscure the two openings in the side of the right ventricle, is believed by us to be secondary to the two defects. The extravagance of the vegetations in the conus of the right ventricle supports this assumption, for such processes rarely develop in this situation without a determining cause.

In conclusion, the writer's sincere thanks are due to Professor W. F. Hamilton for the privilege of studying this remarkable case during life, to Professor Horst Oertel for placing the heart at her disposal, to Dr. Arthur Keith for the valuable information obtained through his kindness from the Grantham Hospital, to Professor G. S. Huntington for the privilege of studying his admirable embryological and anatomical Museum Collections, and for his kindness in giving this case his consideration and confirming the explanation here given of the developmental origin of the two defects, and to Sir William Osler for inspiration and encouragement in the study of congenital hearts.

#### BIBLIOGRAPHY

1. Beck, "Aneurysm of Ascending Aorta Bursting into Right Ventricle with a Communication between the Two Ventricles," *Medico-Chir. Tr.*, 1842, XXV, 15.
2. Hale-White, "A Case of Patent Ventricular Septum, together with an Aneurysm of the Base of the Aorta Opening into the Right Ventricle," *Tr. Patb. Soc.*, Lond., 1891-92, XLIII, 34.
3. Krzywicki, "Aneurysms of the Right Aortic Sinus of Valsalva," *Zieg. Bertr.*, 1889, VI, 473.
4. Kraus, "True Aneurysm of the Right Aortic Sinus of Valsalva," *Berl. klin. Wechnschr.*, 1902, XXIX, 1161.

5. Hart, "Aneurysm of the Right Sinus of Valsalva of the Aorta and its Relation to the Upper Ventricular Septum," *Virch. Arch. f. patb. Anat.*, 1905, CLXXXII, 167.
6. Thurnam, "Aneurysms, and Especially Spontaneous Varicose Aneurysms of the Ascending Aorta, and Sinuses of Valsalva." Case VII. "Spontaneous Varicose Aneurysm of the Right Aortic Sinus and Summit of the Right Ventricle of the Heart," *Medico-Chir. Trans.*, 1840, XXIII, 337.
7. Peacock, "Aneurysm of the Ascending Aorta, Pressing upon the Base of the Right Ventricle and Opening into the Origin of the Pulmonary Artery; with Remarks on the Communication of the Sacs of Aneurysms with the Cardiac Cavities and Adjacent Vessels," *Tr. Patb. Soc. Lond.*, 1868, XIX, 111.
8. Brocq, "Étude sur les Communications entre l'Aorte et l'Artère Pulmonaire autres que celles qui résultent de la persistance du Canal Artériel," *Rev. de méd.*, 1885, V, 1046, and 1886, VI, p. 786.
9. Rickards, "Six Cardiac and Vascular Cases. Case II. Communications between the Aorta and Pulmonary and between the Right and Left Ventricle; Two Aortic Segments," *Brit. M. J.*, 1881, II, 71.
10. Wilkes, "Communication between the Pulmonary Artery and Aorta," *Trans. Patb. Soc., Lond.*, 1860, XI, 57.
11. Baginsky, "Communication between Aorta and Pulmonary Artery," *Berl. klin. Wchnschr.*, 1879, XLIII, 420.
12. Fräntzel, "A Case of Abnormal Communication of Aorta with Pulmonary Artery," *Virch. Arch.*, 1868, XLIII, 420.
13. Girard, "Case of Congenital Communication between Aorta and Pulmonary Artery," Zurich Thesis, 1895.
14. Hektoen, "Rare Congenital Anomalies. Case I. Large Defect in Septum between Pulmonary Artery and Aorta, the Heart normally Developed. General Infection with Bacillus Mucosus Capsulatus," *Tr. Patb. Soc. Chicago*, November 12, 1900.
15. Charteris, "Notes on a Case of Congenital Malformation of the Heart, Opening between Aortic Valves and Right Ventricle, *Med. Press. & Cir.*, 1883, XXXV, 354.
16. Plesch, "Zur Diagnose der kongenitalen Vitien," *Berl. klin. Wchnschr.*, 1909, XLVI, 390.
17. Tate, "Case of Malformation of the Heart with Perforation of Ventricular Septum," *Tr. Patb. Soc. Lond.*, 1892, XIII, 36.
18. Coupland, "Defect in the Ventricular Septum of the Heart, Probably Congenital; unusual Site of Aperture," *Tr. Patb. Soc. Lond.*, 1879, XXX, 266.



19. Rolleston, "Communication between the Ventricles of the Heart," *Tr. Path. Soc. Lond.*, 1891, XLII, 65.
20. Roberts, "Aneurysm of Root of Aorta Communicating with Pulmonary Artery," *Brit. M. J.*, May 2, 1868, No. 383.
21. Gairdner, "Aneurysm of First Part of Artery Opening into Pulmonary Artery and Conus Arteriosus of Right Ventricle," *Glasg. Hosp. Rep.*, 1899, 1.
22. Langer, "On the Development of the Bulbus Cordis in Amphibia and Reptiles," *Morph. Jabrb.*, XXI, 1894.
23. Greil, "Contributions to the Anatomy and Development of the Heart and Truncus Arteriosus in Vertebrates," *Morph. Jabrb.*, XXXI, 1903.

r.  
l-  
o-  
p.  
nd  
rt  
l,

1  
2  
3  
4  
5  
6  
7  
8  
9  
10  
11  
12  
13  
14  
15  
16  
17  
18  
19  
20  
21  
22  
23  
24  
25  
26  
27  
28  
29  
30  
31  
32  
33  
34  
35  
36  
37  
38  
39  
40  
41  
42  
43  
44  
45  
46  
47  
48  
49  
50  
51  
52  
53  
54  
55  
56  
57  
58  
59  
60  
61  
62  
63  
64  
65  
66  
67  
68  
69  
70  
71  
72  
73  
74  
75  
76  
77  
78  
79  
80  
81  
82  
83  
84  
85  
86  
87  
88  
89  
90  
91  
92  
93  
94  
95  
96  
97  
98  
99  
100