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Reprinted from the Archives of Internal Medicine
July, 1915, Vol. xvi, pp. 38-53

CHICAGO
American Medical Association
Five Hundred and Thirty-Five North Dearborn Street
1915

A CASE OF OPEN DUCTUS ARTERIOSUS (BOTALLI), WITH NECROPSY *

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A case of open ductus Botalli in a girl of 17, with necropsy, occurring at the Peter Bent Brigham Hospital in the last year, was carefully observed on Dr. Christian's service in the hospital for fifteen days, and notes were obtained of a previous examination four years before entrance. Diagnosis of the lesion was made during life. The completeness of the record of this case gives it that value as an aid to the diagnosis of the lesion which is common to all carefully observed single cases. The purpose of this paper is to present and comment upon this case, to summarize the recent uncollected cases, and to consider such points as these data seem to throw light on, but to make no attempt to repeat the general discussion of the subject so fully presented by Hochsinger,[1] Wells,[2] Abbott,[3] and Goodman.[4]

In 1908 H. G. Wells collected all the uncomplicated cases of open ductus Botalli and found a total of forty-one, only twenty of which were in adults. Maude Abbott, in the same year, found thirty-seven, only nineteen of which had clinical histories, and three of these latter cases were unconfirmed by necropsy. E. H. Goodman, in 1910, determined the percentage occurrence of symptoms in thirty-four necropsied cases. It is not possible to tell from these papers how many cases confirmed by necropsy and carefully recorded clinically have occurred in adults, but the number is evidently extremely small.

Dr. Abbott's and Dr. Goodman's analyses of the cases reported are so complete that I have used them in my summary. I have carefully searched the literature for cases occurring since the publication of these papers, and have found twenty-two in all. In only five was the patient over 1 year old; one of these cases was complicated by slight aortic and mitral stenosis, and one by mitral stenosis with acute vegetations; none

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* From the Pathological Laboratory of the Peter Bent Brigham Hospital.

1. Hochsinger, K.: Zur Diagnose der Persistenz des Botallischen Ganges und der Erweiterung der Lungenarterie, Wiener Klinik, 1907, xxxiii, 311.
2. Wells, H. G.: Persistent Patency of the Ductus Arteriosus Botalli, Am. Jour. Med. Sc., 1908, cxxxvi, 381.
3. Abbott, M. E.: Congenital Cardiac Disease, Osler's Modern Medicine, 1908, iv, 323.
4. Goodman, E. H.: Report of a Case of Patent Ductus Arteriosus Botalli, with a Study of the Cases Heretofore Published, Univ. Pennsylvania Medical Bulletin, 1910, xxiii, 509.

had other important congenital lesions. Of the infantile cases eleven were complicated by other congenital abnormalities, leaving two uncomplicated infantile cases and two infantile cases in which an absence of other lesions was not noted. The symptomatology and pathology will be discussed after the presentation of the case I have studied.

REPORT OF CASE

Patient.—E. S., aged 17, female, unmarried, of German ancestry. At the age of 7 the patient had measles, and shortly afterwards mumps. Her general health had been good; she was not subject to tonsillitis or colds, and no other infectious diseases had been noted. There was no history of decompensation symptoms such as dyspnea, edema, or precordial pain. There were no symptoms referable to other organs, with the exception of headaches, occurring about once a week.

In March, 1910, at the age of 13, the patient entered the service of Dr. Christian at the Carney Hospital, Boston, and was discharged after thirteen days with the diagnosis of "Congenital heart. Pulmonic stenosis. (?) Open ductus arteriosus (?)."

First Examination.—On physical examination at entrance, the following heart condition was found: Dulness, the upper border at the third rib, the right border 1 cm. to the right of the midsternal line; the left border 3.75 cm. to the left of the midsternal line. A palpable thrill over the pulmonic area extended to the left and downward along the left sternal margin. Over the same area was heard a rough systolic rumble running into diastole. A systolic murmur was heard all over the precordia. The pulmonic second sound was accentuated. The action was regular.

March 16 the thrill was noted to be continuous through systole and diastole, but somewhat stronger in systole, with a late systolic accentuation. At the apex could be felt a short late diastolic and a systolic thrill.

June 19 the patient entered the Peter Bent Brigham Hospital on account of "spots on the legs." There had been no general malaise or other symptoms of illness. Four days before entrance a large black and blue spot had appeared spontaneously on the inner side of the right ankle, and soon after several small reddish spots on both shins. The spots had given no noticeable subjective sensations.

Second Examination.—A rather poorly developed and nourished girl was found, in no apparent discomfort. The only positive points on physical examination were with reference to the heart and extremities.

The heart examination showed: On palpation, an apex impulse in the fifth space, 8 cm. to the left of the midsternal line; over the pulmonic area a marked thrill running throughout the cardiac cycle with greatest intensity during systole; a sharp second sound, distinctly felt, over the pulmonic area. On percussion, dulness with the upper border at the third rib, the right border 2 cm. to the right of the midsternal line, and the left border 10 cm. to the left of the midsternal line in the fifth interspace. On auscultation, at the apex a first sound of good quality, followed by a systolic murmur somewhat rumbling in character, apparently transmitted from the pulmonic area; a normal second sound; a loud rough murmur over the pulmonic area, running throughout the cardiac cycle coincident with the thrill, with the greatest intensity during systole; a blowing diastolic murmur, distinct from the other murmurs, transmitted from the pulmonic area down along the left border of the sternum, becoming slightly musical in the third and fourth interspaces; aortic second not increased; pulmonic second markedly accentuated; the action regular and moderately rapid. The blood pressure was 118 systolic, 70 diastolic.

Extremities: The fingers were clubbed and the nails cyanotic. On the right leg, just above the internal malleolus, was a purplish-black echymosis, about 5 mm. in diameter, moderately tender. Scattered over the anterior surfaces of both shins were a number of small purpuric spots 1 to 3 mm. in diameter.

The temperature was 103.4, pulse 129, respiration 31. The white count was 14,000, and the hemoglobin 65 per cent. A smear showed moderate achromia, polymorphonuclears 78 per cent., small mononuclears 7 per cent., large mononuclears 15 per cent., eosinophils none. The urine had a small trace of albumin. The Wassermann reaction on the blood serum was negative.

The clubbing of the fingers, with some cyanosis, the character and localization of the thrill and murmurs, and the lack of etiology for cardiac disease, suggested a congenital cardiac lesion, either a patent foramen ovale, or a patent ductus arteriosus. A severe active endocarditis superimposed on the congenital defect was indicated by the purpuric spots, fever, anemia, and heavy sweats; although this indicated infection was considered not necessarily localized in the heart.

Further Clinical Notes.—On June 22 examination gave similar heart findings, except that the systolic and diastolic murmurs were noted all over the precordia, and the diastolic murmur seemed of greatest intensity over the aortic area, suggesting a diagnosis of aortic insufficiency in addition to an open ductus arteriosus. The murmur over the pulmonic area is described as a "sawing to and fro murmur" through the whole cycle.

June 23: A high evening temperature, usually reaching 103, continued, but the patient felt comfortable and had no complaints. The purpuric spots had almost faded out.

June 25: Slight cyanosis was noted. Visible pulsations were found in the fifth space, nipple line, in the suprasternal notch, and in the second interspace to the left of the sternum. In the latter spot the impulse was wave-like. The time of maximum intensity was just after the first sound, as judged by apex and aortic pulsations. On percussion no increase of dulness was definite to the right of the sternum, while to the left in the second and third spaces increased dulness was found for a distance from 1 to 2 cm. from the sternum. On auscultation the first sound appeared blurred, and was followed by a low-pitched murmur transmitted into the axilla. The rhythm was a protodiastolic gallop. Otherwise the examination revealed no new facts. Another observer the same day noted a distinct capillary pulse.

June 26: The total diameter of the heart was made out by percussion to be 18.5 cm., the left border being 15 cm. to the left of the midsternal line at the level of the fifth rib, and the right border 3.5 cm. to the right of the midsternal line at the junction of the fourth rib. The apex was most distinct in the fifth space, but could be felt in the sixth. A pistol shot sound was heard in the femoral arteries.

June 27: A distinct systolic thrill at the apex was noted. The diastolic murmur seemed most intense in the third space and the systolic at the apex. A capillary pulse in the fingers and collapsing pulse in the small arteries were plain. The diagnosis at this time was aortic and mitral regurgitation, with possibly a patent ductus arteriosus, and with an acute process on the chronic valvular lesions.

Two blood cultures had showed no growth. Widal reactions were negative. The white count had risen to 34,000. The red count was 2,408,000. Occasional granular casts were found in the urine. The temperature and pulse continued the same course.

The patient's condition now began to grow worse; there was much vomiting, and no food intake. The temperature fell to 99 June 29 and remained about at that level until July 3, when there was an ante-mortem drop.

On July 2 dulness with many fine crackling râles were found at the right base, and the white count was 70,000.

July 3 the systolic murmur was heard in the vessels of the neck, and seemed most intense over the sternum at the level of the third costal cartilage; the diastolic was plainest over the pulmonic area and along the sternum; a capillary pulse, a Corrigan pulse, and pistol shot sounds in the femoral arteries were plainly observed. Cyanosis persisted, and there was dyspnea out of proportion to the temperature.

On July 4, after a comfortable night, the patient sat up in bed suddenly at about 9 o'clock in the morning, with a complaint of palpitation and dyspnea, and quickly became very cyanotic and almost pulseless. The right border of

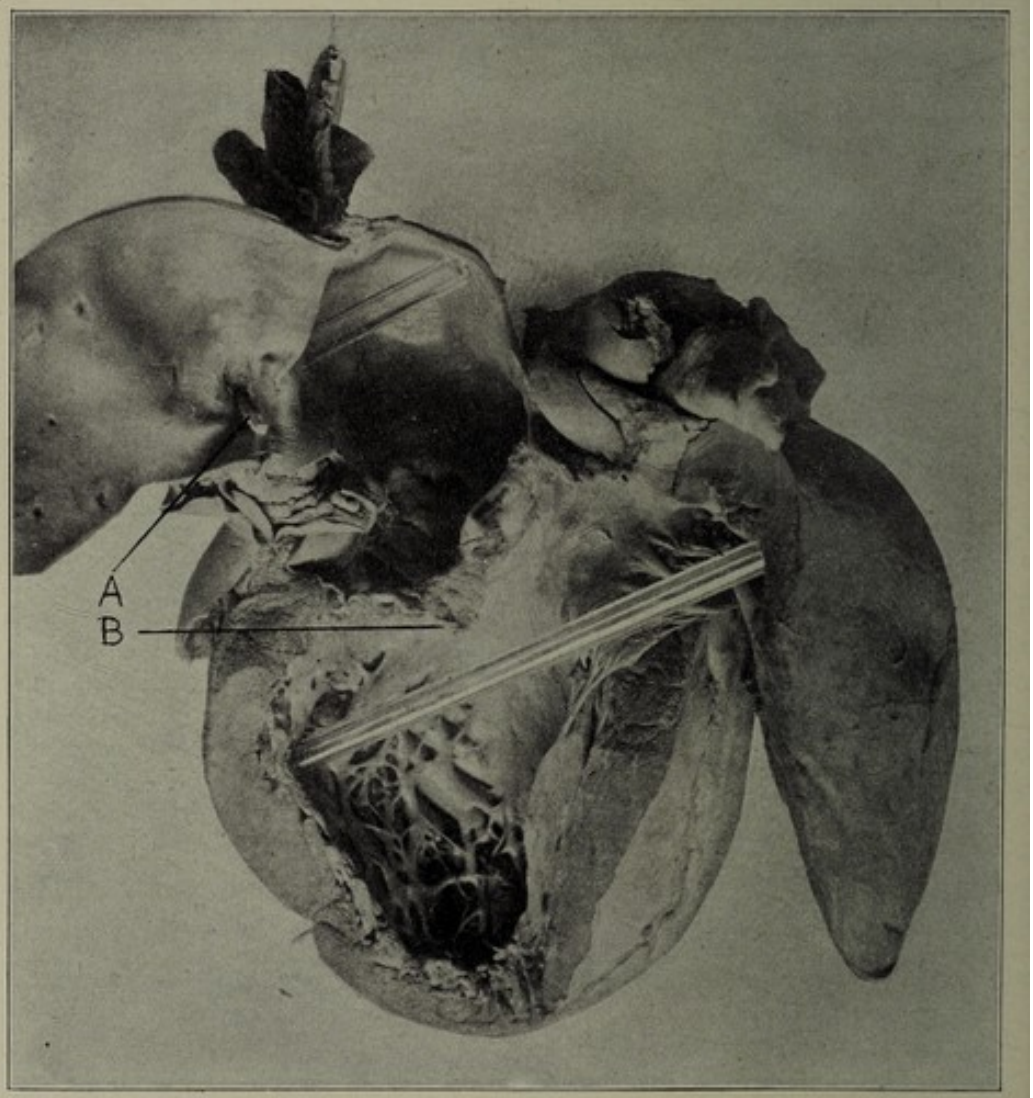

Fig. 1.—View of the heart from the left showing the vegetations on the aortic valve (*B*), and a probe in the open ductus (*A*). Beneath the probe can be seen the ridge on the aorta.

the heart was found to be 5 cm. (later 6 cm.) to the right of the midsternal line. The liver was felt 6 cm. below the costal margin and was pulsating. The previously felt thrills had almost disappeared, and the murmurs were of almost the same character, but of diminished intensity. A presystolic gallop rhythm was felt at the apex. The lungs appeared as before. The condition rapidly became worse, with greater cyanosis, deep quick respirations, and very rapid, scarcely audible heart sounds. The patient died at 9:35 a. m.

Blood cultures made July 4 showed a growth of an anaerobic hemolytic short-chained streptococcus, probably *Streptococcus viridans.*

Necropsy.—Two hours' post mortem.—Body: Emaciated, 165 cm. long. None but hypostatic discolorations of the skin.

Thoracic Cavity: When the thorax is opened the heart is found lying in a pericardial sac distended with fluid. The apex is in the fifth space, 9 cm. to the left of the midsternal line, and the right margin of the ventricle is 6 cm. to the right of the midsternal line. The plurae are smooth except for a small, easily broken adhesion at the right base posteriorly, and contain no free fluid.

Lungs: Left, weight 365 gm. At the apex is a dry, firm, red nodule 7 cm. in diameter. The remainder of the lung is crepitant and mottled with dark, brick-red areas on a yellowish ground. Right, weight 445 gm. The lung appears similar to the left one. Microscopically edema is found, with an infarct, and some areas of atelectasis.

Heart: Weight 252 gm. Right auricle: No dilatation or hypertrophy. The foramen ovale is closed. Some non-adherent, apparently post mortem, blood clot in the appendage. The coronary sinuses normal. Tricuspid valve: 10.5 cm., leaflets thin and covered with smooth endocardium; not retracted; attached to normal chordae tendinae. Right ventricle: Slight dilatation and hypertrophy; the walls measure 5 mm. Pulmonary valve: 7.5 cm. There are only two cusps, which are of equal length, and show no line of former fusion. No thickening or evidence of endocarditis can be seen. The valve apparently could function. A 5 mm. orifice, placed 2.5 cm. above the origin of the cusps, leads by a tube 8 mm. long into the aorta. This open ductus Botalli has just within its lumen a vegetation of fresh appearance, and a collection of similar pinkish vegetations from 1 to 3 mm. high form a streak leading from the orifice over the surface of the pulmonary artery in a posterior and slightly downward direction. The streak is from 1 to 3 cm. wide and aside from the vegetative roughenings the endocardium over it appears corrugated and darkened, but not finely irregular. The orifice of the ductus has no mound-like elevation about it as was described in Wells' case (Case 6 in the tabular summary). The duct is almost cylindrical in shape, but there is slight narrowing of the lumen near the middle. About the aortic mouth and throughout the lumen there is yellowish discoloration but no definite raised patches or sclerosis. On the aortic side the duct opens 5 cm. above the origin of the aortic valves, just beyond the left subclavian, by an orifice 5 mm. in diameter. The opening is smooth except for the presence, just below it, of a straight, ridge-like formation 5 mm. long and 3 mm. high, too stiff and thick to bend and act like a valve. Probably this represents the valve-like fold supposed by Strassman[5] to close the duct when blood pressures change at birth. A few vegetations are about this orifice of the duct. Left auricle: Dilated, but not definitely hypertrophied. The auricular appendage free from clot. Mitral valve: 9.5 cm.; there is slight but definite fibrous thickening of the edges of the cusps, but the endocardium is free from vegetations or ulcerations. Left ventricle: Slight dilatation. The walls are 1.2 mm. thick, and the muscle appears normal. Aortic valve: 8 cm. Two cusps only are visible; one is 4.7 cm. and the other 3.3 cm. The leaflets show evidence of severe acute vegetative endocarditis; there are along the line of closure of the largest cusp four soft nodular masses 1 to 3 mm. in diameter, with punctate elevated pink areas about them. The edge of the leaflet has attached to it two granular leaf-like appendages from 1 to 2 mm. thick and 1.1 cm. long, soft in consistency. Back of the vegetations are areas of fibrous thickening. The other cusp has similar but less well marked lesions. Near the posterior attach-

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5. Strassmann, P.: Ueber den Mechanismus des Verschlusses des Ductus Arteriosus (Botalli), Arch. f. Physiol., 1893, p. 566; Anatomische und physiologische Untersuchungen über den Blut Kreislauf beim Neugeborenen., Arch. f. Gynaek, 1893, xlv, 393.

ment of the anterior cusp is a small ulcerated area 3 mm. in diameter. The sinuses of Valsalva are shallower than normal, due to the retraction of the valves. The right coronary artery opens into the aorta by four small orifices. The arteries are otherwise normal. Aorta: There are several spots of yellowish discoloration but not of definite elevation or increased density. The aorta has four principal branches, the positions and sizes of which are shown in Figure 2. Circumference of aorta 1 cm. above the valve (in formaldehyd hardened specimen) is 6 cm. Circumference of pulmonary artery in corresponding position 6.8 cm. (There is considerable shrinkage in the formaldehyd specimen, but the comparative size of the arteries should be nearly the same, and these measurements show that the pulmonary artery is larger than normal in proportion to the aorta.)

Anatomical Diagnoses.—Open ductus arteriosus (Botalli). Anomalies of pulmonary and aortic valves, consisting in the presence of only two segments in each valve. Four primary aortic arch branches. Acute vegetative, and chronic endocarditis, with connective tissue proliferation, of the aortic valve. Vegetations of the ductus Botalli and pulmonary artery. Slight hypertrophy of the right ventricle. Infarcts of both lungs.

Microscopic examination shows an acute endocarditis of the aortic valves, and endarteritis of the aorta, ductus Botalli and pulmonary artery, with vegetations and masses of streptococci.

A section of the ductus stained with Verhoeff's elastic tissue stain demonstrates a large amount of elastic tissue, thus suggesting that deficiency in elastic tissue is not the only factor in producing patency.

COMMENT

This is one of the rare cases of patent ductus Botalli with "typical" signs, and one of the few cases diagnosed during life and confirmed by necropsy. In the series in the accompanying table only one other of the cases of adults had the lesion included in the antemortem diagnosis.

The endocarditis of the aortic valve above seems to indicate that the vegetations about the ductus Botalli and along the pulmonary artery, and also the lung emboli, owe their ultimate origin to this lesion, and that in fact the infection has made a vivid diagram by its lesions of a course of blood from the aorta through the ductus Botalli.[6] While the embolus which produced the infarction of the lung may have had its immediate origin in the vegetations on the pulmonary artery, it is also possible that it came from the lesions of the aortic valve through the open ductus. The possibility of this form of paradoxical embolism has not received the attention it deserves. It may be nearly as important as the usually cited passages through the foramen ovale, for in 412 cases of congenital cardiac disease patent ductus arteriosus occurred (counting cases with other congenital lesions) 106 times, as compared with 134 cases of open foramen ovale.

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6. A very similar case of acute aortic endocarditis with extension through the duct is reported by Schlagenhaufer: Ztschr. f. Heilk., 1901, xxii, 19.

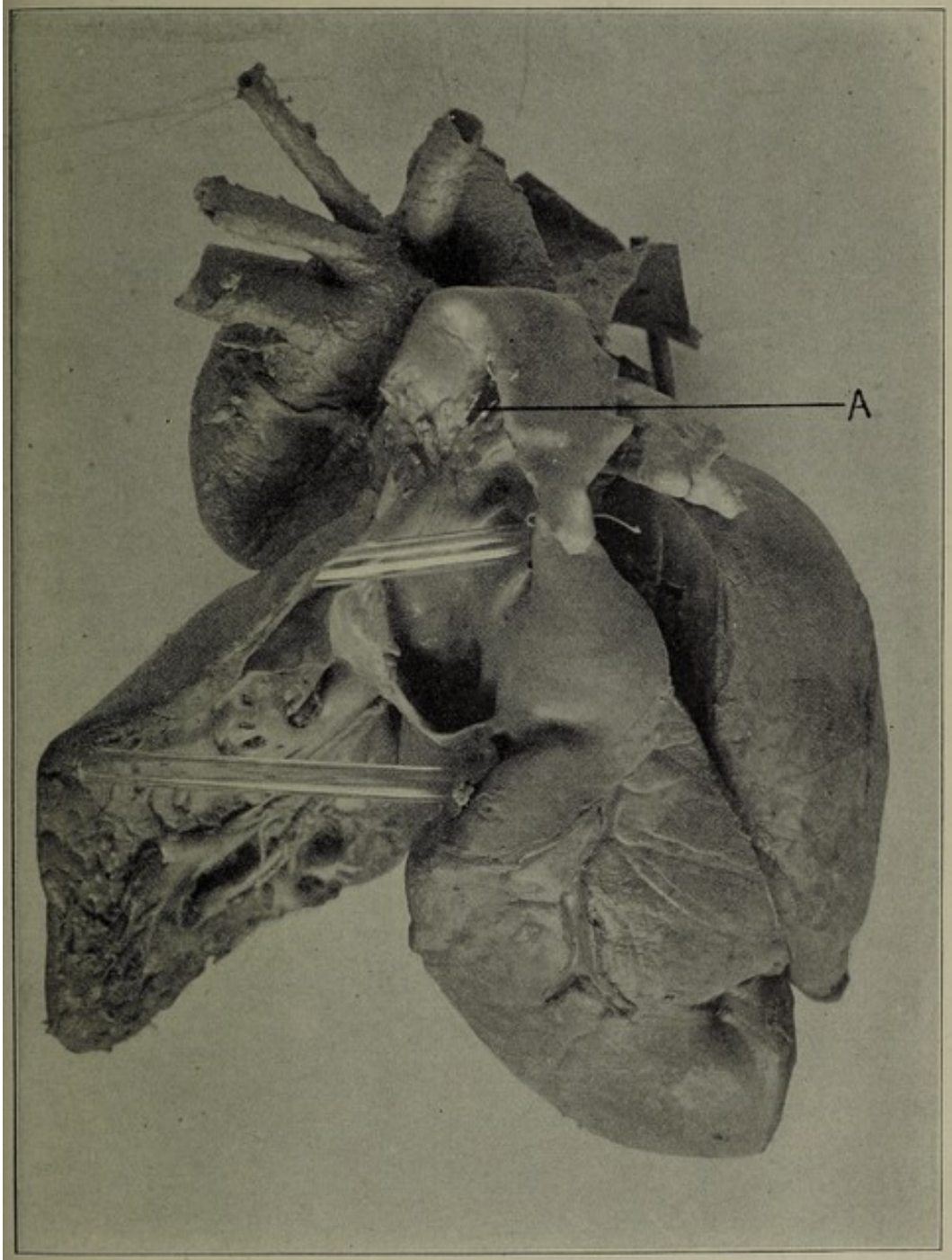

Fig. 2.—View of the heart from the right and above, showing the probe in in the open ductus (*A*), the vegetations on the pulmonary artery to the left of the probe, the four aortic arch branches, and the two cusps of the pulmonary valve.

Table of Reported Cases

Case	Age	Sex	Cyanosis	Clubbing	Dyspnea	Heart Examination			
						Right Border	Left Border	Thrill	Murmurs
I									
1	26	♀	0	0	0?	Incr.	Normal	Felt only once.	"Machinery" murmur through whole cycle, loudest in 3d lt. space.
2	26	♀	0	.....	In crises with palpitation.	.....	.....	.....	Presystolic roll at apex.
3	32	♂	Antemortem.	0	Antemortem	Normal	Normal	.....	Short presystolic over precordia.
4	35	♂	.....	.....	Slight	.....	.....	.....	.....
5	55	♀	Marked	.....	.....	.....	Normal	.....	Faint systolic at apex.
6	42	♂	.....	.....	.....	Incr.	Incr.	Suggested at apex.	Loud blowing systolic in 2d rt. space
7	17	♀	Slight	Present	0	Normal	Normal	Through cycle over pulmonic area.	Rough systolic running into diastole over pulmonary area.
II									
8	6½*	♂	Present	.....	.....	.....	.....	.....	0
9	6*	♀	Attacks	0	.....	Normal	Normal	.....	0
10	11½*	?	Moderate	Present	On exertion.	Incr.	Incr.	0	Systolic at base antemortem.
11	11½*	♀	0	.....	.....	Incr.	Normal	.....	Systolic over pulmonic area.
12	9†	♀	Slight attacks.	.....	.....	Incr.	Normal	0	Systolic loudest in 2d left space.
13	5†	♀	Marked	.....	.....	Incr.	Incr.	0	Systolic loudest 5th space 3 cm. out.
14	3†	♂	Slight	0	.....	Normal	Normal	0	Rough systolic loudest near apex.
15	7†	♂	Marked	.....	Present	Normal	Normal	.....	.....
16	6½*	♂	General	.....	.....	Normal	Normal	0	Systolic loudest opposite nipple.
17	17‡	..	Present	.....	.....	.....	.....	.....	0
18	2†	..	Present	.....	0	Incr.	Normal	.....	.....
19	2‡	♂	.....	.....	.....	.....	.....	.....	0
20	5‡	♀	Present	Present	.....	Normal	Normal	.....	Sawing systolic, loud at base.
III									
19	....	..	6	.....	6	Usually Incr	.....	7 — systolic or continuous.	Almost always loud systolic or continuous.
IV									
34	....	..	29%	2.9%	47%	.....	.....	Systolic over pulm. area —29% Continuous —5.9%	Systolic in pulm. area—38%. Continuous in pulm. area—5.9%

* Weeks. † Months. ‡ Days. ♀ Female. ♂ Male.

1. Mead, K. C.: Jour. Am. Med. Assn., 1910, lv., 2205.
2. Doufour, H., and Huber: Bull. et mém. Soc. méd. d. hôp. de Paris, 1911, xxxi, 510.
3, 4 and 5. Motzfeldt, K.: Deutsch, med. Wchnschr., 1913, xxxix, 2037.
6. Wells, H. G.: Am. Jour. Med. Sc., 1908, cxxxvi, 381.
7. This paper.
8. Weber, F. P.: Proc. Roy. Soc., Sect. Study Dis. Child., 1908, p. 193.
9. Cautley, E.: Proc. Roy Soc., Sect. Study Dis. Child., 1908, p. 34.
10. D'Espine, M. H.: Rev. de méd., 1908, xxviii, 941.

	Autopsy					
Heart, ft.,Gm.	Right Ventricle	Left Ventricle	Pulmonary Artery	Valves	Other Organs	Other Defects
395	Dil. and hyper.	Hyper.	dil.	Slight mitral and aortic sten.	.....	None.
...	Hyper.	Hyper.	.....	Mitral sten. and veg.	.....	None.
590	Hyper.	Hyper.	dil.	Normal	.....	None.
480	Hyper.	Hyper.	sl., dil.	Normal	.....	None.
700	Hyper.	Hyper.	dil.	Normal	.....	None.
300	Slight hyper.	.....	.....	Normal	Liver cirrhosis	None.
252	Slight hyper.	0	sl., dil.	Aortic veget.	Lung infarcts	Two leaflets on aortic and pulmonary valves. Four aortic branches.
...	Slight hyper.	Slight hyper.	.....	Normal	Atelectatic patches	None.
...	Slight hyp. and dil.	Slight	.....	.....	.....	Slight narrowing of aorta, patent foramen ovale and ventricular septum.
95	Hyper. and dil.	.....	.....	.....	.....	Aorta rides ventricles, obliterated pulmonary artery, patent ventricular septum.
...	.....	Hyper.	Lumen ¼ in.	.....	Slight emphysema	Aorta rides ventricles, patent ventricular septum, atresia pulmonary artery.
...	.....	.....	sl., dil.	.....	Pneumonia	Absent ventricular septum, slight narrowing aorta, four aortic branches.
...	.....	.....	.....	.....	.....	Absent interauricular septum, fused mitral and tricuspid valves, four aortic branches.
...	Dil. and hyper.	.....	.....	.....	Broncho-pneumonia	Large ventricular septum defect, small open foramen ovale.
...	Hyper.	.....	Atresia	Aortic sten.	Emphysema	Atresia pulmonary artery, open foramen ovale.
90	Hyper.	.....	Atresia dil. above	Sl. aortic sten.	Broncho-pneumonia	Atresia pulmonary artery, open foramen ovale.
...	.....	.....	Atresia	.....	.....	Atresia pulmonary artery.
...	Hyper.	.....	Oblit.	Normal	Atelectasis	Obliterated pulmonary artery.
...	.....	.....	.....	.....	.....	Aorta and pulmonary artery arise in common.
...	.....	.....	.....	.....	Broncho-pneumonia	
...	Usually hyper.	Often hyper.	Often dil.			
...	Hyper. in 32.1%	Hyper. in 32.1%				

11, 12 and 13. Carpenter, G.: Brit. Jour. Child. Dis., 1908, v, 396.
14. Sawyer, J. E. H.: Birmingham Med. Rev., 1909, lxvi, 152.
15. Bach, S.: Arch. f. Kinderh., 1909, l, 31.
16. Bradley, W. N.: New York Med. Jour., 1909, lxxxix, 1302.
17. Vallois, M.: Bull. Soc. Obst. de Paris, 1910, xiii, 107.
18. Edwards, E. P.: Cleveland Med. Jour., 1911, x, 748.
19. Gierke, E.: Charité Ann., 1907, xxxii, 299.
20. Kingsley, C. R.: Johns Hopkins Hosp. Bull., 1911, xxii, 56.

Table 1.—In the first division of the table are the cases since 1908, of patients over 1 year of age—the youngest happens to be 17; in the second division are the cases since 1908 of under 1 year of age; in the third division are the cases collected by Dr. Abbott, and in the fourth those of E. H. Goodman. The case in this paper is tabulated as Case 7. The table summarizes practically all recorded uncomplicated cases of open ductus Botalli, and a few (occurring since 1908) complicated cases. Notes 1 to 20 are the references to the cases in the order given in the table.

That the lesion had no great disturbance of function as its sequence is plainly seen by the absence of cardiac symptoms in the case history. Clubbed fingers with slight cyanosis of the nails constitute the only recognized effect. In the heart itself there is slight right ventricular hypertrophy, and on consideration it seems probable that there was a distinct disturbance in the pressure in the pulmonary circuit, the cause for which is of course to be sought in the aortic-pulmonary short circuit, and the result to be seen in the dilated pulmonary artery, and hypertrophied right ventricle, and proved clinically by the palpable and loud pulmonic second sound, which was a striking feature of the heart examination. Very probably the clubbing of the fingers is to be traced to this changed pressure condition.

Were the thrill and murmurs due to the patent ductus Botalli or to the aortic lesion? No fact is conclusive proof one way or the other, but there is an accumulation of probabilities, all of which point in one direction, and which give a basis for a theory very reasonable in appearance. In the first place, why should there be a murmur from the passage of blood through this tube any more than through other branches of the aorta? Of course there is a possibility for the production of a murmur in the interference of the ductus current with the current in the pulmonary artery. Examination of the specimen, however, gives a plainer answer; the ductus comes away from the aorta at a peculiar angle, and at the proximal side of the opening is the sharp ridge described above, an arrangement admirably suited to break up the flow and produce the continuous murmur. Assuming that murmurs are transmitted with the blood flow, this murmur would be heard as in the pulmonary artery. The pathological examination indicated that the aortic lesion was a rather recent one, for there was none of the left ventricular hypertrophy and dilatation of a four years' aortic insufficiency. Clinically, also, the absence of signs of aortic insufficiency at the examination four years before death, and the absence at that time of the cardiac history to be expected in the production of the aortic lesions, point in the same direction. The murmur itself does not correspond to the usual character of an aortic insufficiency murmur, and the continuous humming top thrill would be difficult to explain on that basis. Most probably, then, the continuous thrill and the accompanying murmurs were unconnected with the aortic lesion, but were due to the sharp ridge on the aorta near the

orifice of the ductus Botalli. The diastolic murmur, greatest over the aortic area, the systolic thrill at the apex (and perhaps part of the other murmurs) coming at the time other signs of aortic disease were recognized, and at the time when the pathological examination and clinical history indicate aortic disease began, suggests that they were probably due to the aortic lesion.

An important point in the physical examination is the mention on June 25 of visible pulsation and dulness in the second left interspace, just after the first sound. This corresponds with the strip of dulness, and the Roentgen-ray shadow so much emphasized by several observers and supposed to be made by the dilated pulmonary artery. Pulsation in the suprasternal notch was noted at the same time.

To compare this with previous cases I shall make use of Dr. Abbott's and Dr. Goodman's summaries, and my own of the cases published since.

Besides these may be mentioned the case of Dr. C. H. Dunn,[7] in which an infant with a murmur and slight cyanosis, but no thrill or heart enlargement, was found at necropsy to have an open ductus; and that of Dr. A. Hayashi,[8] in which a baby of 10½ months, with normal heart boundaries, a visible pulsation in the fourth, fifth and sixth spaces, a palpable systolic thrill, and a loud systolic and soft diastolic murmur over the precordia, was proved post mortem to have excessive hypertrophy of the left ventricle, active dilatation of the right ventricle, dilated auricles, and a ductus Botalli of a 5 mm. diameter.

The most interesting single case was reported by Mead. It is Case 1 in the table. Observations covering a period of three years were made. At first slight lateral enlargement in both directions, with a noisy systolic murmur at the base and apex, transmitted to the right of the sternum and almost to the axilla were found. Two years later a strong thrill in the third and fourth right spaces was noted. An examination by Dr. Thayer of Johns Hopkins showed a long machinery murmur over the right ventricle, strikingly loud at the base and the first left interspace, with a late accentuation—almost diastolic—high up. The right border was 5.5 cm. to the right, the left 8.5 cm. No thrill was felt. An impulse was felt in the second and third spaces. A soft systolic and a soft diastolic murmur were heard at the apex. The pulmonic second sound was loud. The diagnosis was made of septum defect or open ductus arteriosus.

In 1909 paralysis of the right vocal chord was diagnosed.

Roentgen-rays showed a bulge on the left of the heart close to the descending aortic arch, considered to be a hypertrophied and dilated

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7. Dunn, C. H.: Trans. Am. Pediat. Soc., 1913, xxv, 237.
8. Hayashi, A.: Monatschr. f. Kinderh., 1912, xi, 224.

right auricle. The patient then had irregularities and a loud aortic murmur, and died after several attacks of faintness.

At necropsy open ductus arteriosus was found, with a thickened ring and a fold of membrane about it. The heart had three ruptures of the right ventricle. The right ventricle was dilated and hypertrophied, the myocardium degenerated, the pulmonary artery twice the size of the aorta; there was slight aortic and mitral stenosis.

In the analysis of cases in the accompanying table it seems evident that characteristic symptoms or physical signs are rare. Cyanosis occurred in 70 per cent. of the cases as against the 31 per cent. of Dr. Abbott, and was about as frequent in the uncomplicated cases as in the whole. Clubbing of the fingers was noted in only 15 per cent.; in 25 per cent. of the cases there was dyspnea, as against Dr. Abbott's 31.5 per cent. A constant definite thrill was noted only once; Dr. Abbott finds it in 37 per cent., and many writers (De la Camp[9] and others) speak of it as one of the most useful diagnostic signs. A systolic murmur over the base, extending into diastole, was noted only twice (both of them cases of adults). A systolic murmur, loudest over the base, was present in five other cases (only one of them a case of an adult). Dr. Abbott, however, finds that a peculiar loud murmur is nearly always produced, almost invariably beginning in systole, and localized near the base of the heart. A murmur of some kind was present in 65 per cent. of the cases above.

Goodmann, in his collection of 34 cases with necropsy, found twenty females and eleven males. Cyanosis was found in 29 per cent., dyspnea in 47 per cent., palpitation in 37 per cent., clubbed fingers in 2.9 per cent., pulsation in the second left interspace in 5.9 per cent., a systolic thrill over the pulmonary area in 29.4 per cent., a systolic and diastolic thrill over the same area in 5.9 per cent., a systolic murmur over the area in 38 per cent., and continuous murmur in 5.9 per cent. The pulmonic second sound was accentuated in 17.2 per cent. The left ventricle was hypertrophied as often as the right—32.1 per cent.

In this series, whether analyzed by classifying the cases according to the division into simple and complicated cases, or into infantile and adult, the sexes are as evenly divided as possible. Dr. Wells, in his forty-one cases, found a remarkable preponderance of females (63 per cent.), which in the light of this series now seems probably was a matter of chance.

Evidently none of the symptoms or signs just discussed are to be depended upon for a constant diagnosis.

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9. De La Camp, Familiäres Vorkommen Angeborener Herzfehler — Zugleich ein Beitrag zur Diagnose der Persistenz des Ductus Arteriosus Botalli, Berl. klin. Wchnschr., 1903, xl, 48.

The pathological finding of hypertrophy with or without dilatation of the right ventricle was noted in all of the adult cases and in seven of the infantile. Dr. Abbott states that it is the usual pathological finding. Dilatation of the pulmonary artery was found in five of the adult cases and one of the infantile, and clinically the dulness in the second interspace near the sternum emphasized as its result was actually observed four times in the adult cases. (Of course in these statistics it must be remembered that in the infantile cases the examination was evidently often far from thorough.) The pulmonic second sound was noted to be exaggerated in three of the adult cases. It is evident, then, that clinical signs depending on an increased pressure in the pulmonary circuit are the most constant signs in this series. They are common to so many other conditions, however, dilatation of the pulmonary artery often occurring in congenital lesions such as defects of the lower part of the interauricular septum, widely patent foramen ovale, defects of the base of the interventricular septum, transposition of the arterial trunks, stenosis of the aorta, and without other defects, that they do not seem to be of great diagnostic importance.

This analysis indicates that in a large number of cases there is nothing in the way of signs or symptoms to make us sure of an open ductus arteriosus, or even to make us suspect it. Hochsinger emphasizes the difficulty of diagnosis in infants, but hardly in adults. Before a conclusion based on such a small number of cases is accepted, however, it should be determined whether it is supported by reason. What should we expect as the clinical result of the lesion? It does not seem to me that a murmur or thrill is by any means a necessary or even likely result from the slight intermixture of currents arising from the presence of an additional aortic branch at an acute angle, emptying by a small opening into the pulmonary artery. (Hochsinger makes this mixing of currents his main explanation of the origin of the murmur, but there are no facts to support this view except the lack of a murmur in certain cases in which such mixing would not be expected. He has too few cases, however, and murmurs are lacking too often, to allow his arguments to convince.) It is interesting to observe that in the three adult cases in which a thrill or murmur at the base of the heart was noted, there was a distinct fold of endocardium about the aortic orifice of the duct (see description of the pathological findings in this and in Mead's case), and in one case a mound-like elevation about the pulmonary orifice (Wells' case). It seems much more reasonable to consider that the murmur or thrill is not the result of the patency alone, but depends on the presence in addition of some endocardial projection, or other roughening or vegetation, such as is recognized in other situations to give rise to

murmurs. This especially, when it is considered that in the cases with necropsy the ductus was open to almost a constant diameter, thus giving each time about the same anatomical reason for a murmur. The clinical findings of absent murmurs in many cases is in perfect accord with this reasoning. The conditions due to increased pressure in the pulmonary circuit could hardly be expected to be very marked when the small size of the ductus is considered (barely over 5 mm.), and there is of course nothing pathognomonic about them when discovered. Combinations of the signs discussed can be effected by combinations of lesions, which are often found in congenital heart disease. No signs are necessarily present when the ductus is open, then, which are sufficiently specific to afford us means of diagnosis. Even the peculiar murmur occasionally produced accessorily may be closely simulated in pulmonary stenosis, or in defects of the ventricular septum. In occasional cases the peculiar humming systolic murmur, loudest over the pulmonary area, with or without a thrill, combined with signs of increased pressure in the pulmonary artery, such as loud and palpable pulmonic second sound, increased dulness in the second left interspace, increased in the middle Roentgen-ray shadow, and suprasternal pulsation, with or without cyanosis, clubbed fingers, and dyspnea, justify the inclusion of "open ductus" in the differential diagnosis. Of course a continuous murmur over the pulmonary area makes a more certain diagnosis possible. The indefinite murmurs, etc., often present, may give rise to confusion with acquired valvular lesions in a way which can easily be seen from the tabulated cases. The present series of cases brings out the rarity of recognizable symptoms, especially of the thrill and characteristic murmur, rather than new symptoms or signs.

Many articles have been published (Arnheim,[10] De La Camp,[9] Miller,[11] Wessler and Bass[12]) in which it is assumed or claimed that the diagnosis can be made frequently, or almost always, by using the Roentgen-ray and careful percussion to determine the dilatation of the pulmonary artery. It is noteworthy, however, that most of these articles are based on cases without necropsy.

In the cases since 1908 there are only two with roentgenoscopy showing a dilated pulmonary artery confirmed by necropsy; no necropsy was made in the three Roentgen-rayed cases of Dr. Abbott. In view of these facts and of the non-characteristic nature of the findings, any emphasis on this method for diagnosis seems unwarranted.

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10. Arnheim, G.: Persistenz des Ductus Botalli, Berl. klin. Wchnschr., 1903, xl, 616.

11. Miller, R., and Orton, G. H.: A Case of Open Ductus Botalli with X-Ray Examination, Brit. Jour. Child. Dis., 1913, x, 109.

12. Wessler and Bass: Persistent Ductus Botalli and Its Diagnosis by the Orthodiagraph, Am. Jour. Med. Sc., 1913, clxv, 543.

In regard to the prognosis, the table brings out strongly the apparent fact that the patients that die in infancy are those with other congenital heart lesions, while those that live over one year, have an indefinite term of life. It will be noted that none of the adult patients had another important congenital heart lesion, while of the patients that died under 1 year of age only two failed to have one. The pure cases of open ductus, in other words, have little interference with function.

In spite of all that has been written on the subject, conclusions must be tentative until the extraordinarily small number of cases carefully and thoroughly observed is much increased.

SUMMARY AND TENTATIVE CONCLUSIONS

1. The case presented here is one of the rare cases with those signs of open ductus arteriosus usually regarded as typical.

2. It is one of the rare cases diagnosed during life and confirmed by necropsy.

3. The case illustrates the actual course of blood during life through the ductus.

4. The possibility of a practically unrecognized form of paradoxical embolism is shown.

5. A summary and discussion of cases seems to show that:

(*a*) The physical signs formerly regarded as characteristic are more often absent than present, and the possibility of diagnosis must be rare.

(*b*) Most of the signs discussed are really not absolutely characteristic.

(*c*) Combinations of the signs can occur in combinations of other lesions.

(*d*) When there is the rare combination of signs formerly regarded as diagnostic, the presence of the lesion is probable. but not certain.

(*e*) Far too few cases have had roentgenoscopy and necropsy to determine the value of the Roentgen-rays for diagnosis.

(*f*) The Roentgen rays determine only a dilatation of the pulmonary artery, which is present in several other lesions than open ductus, so that the Roentgen ray findings will not make a certain diagnosis possible.

(*g*) The characteristic murmur is probably not the result of the patency of the duct alone, but requires in addition the presence of endocardial folds, vegetations or other roughenings, about the ductus.

(*h*) The only result of the patency is increased pressure in the pulmonary artery (with its secondary effects).

(*i*) The former view of preponderance of cases in females was merely the result of chance.

(*j*) The open ductus alone does not lead to early death, but when complicated with other congenital heart lesions death is usual within one year.

(*k*) Very few cases have been carefully observed and recorded. More cases are needed before conclusions can be definite.

I wish to express my thanks to Dr. Henry A. Christian, chief of the medical service, for his permission to report the case; and to Dr. W. T. Councilman, for valuable suggestions in the treatment.

In addition to the references mentioned in the text, the following may be consulted:

Zinn, W.: Zur Diagnose der Persistenz des Ductus Arteriosis Botalli, Berl. klin. Wchnschr., 1898, xxxv, 433.