

A PATHOLOGICAL STUDY OF TWO CASES OF HEART-BLOCK WITH ADAMS-STOKES' SYNDROME *

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In a recent very interesting report of a case of terminal heart-block, Pardee¹ demonstrates that an extensive anatomical lesion of the auriculoventricular bundle may be present without disturbance of its function. He then gives three lists: (1) ten cases, in which complete destruction of a transverse section of the bundle was invariably accompanied by complete heart-block; (2) six cases,² with varying degree of block, in which no lesion of the bundle existed; (3) ten cases, (six of complete and four of partial heart-block) accompanied by very slight pathological changes. The intermediate degrees of bundle involvement he has not considered. That is, though it has been firmly established that lesions of the conductive system may cause partial and complete heart-block, it seems also true that such lesions may exist (even though of considerable extent) without block; and further that partial or complete block may exist without lesion of the bundle.

The extracardiac origin of heart-block has long been suspected. Holberton³ reported a case in which, after an injury to the neck, extreme bradycardia and "fainting fits" developed. At necropsy, adhesions pressing on the vagus were found with the heart normal. Again, Neuberger and Edinger⁴ found, in a man suffering from chronic constipation, a marked bradycardia and fainting spells during defecation. The necropsy, by Weigert, disclosed a normal heart, but an infarction at the decussation of the pyramids with a small varix pressing on the nuclei and roots of the vagus. Recently it has been demonstrated⁵ that stimulation of the left vagus can cause auriculoventricular dissociation.

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1. Pardee, H. E. B.: The Relation of Heart-Block to Lesions of the Auriculoventricular Bundle, with Report of a Case, *THE ARCHIVES INT. MED.*, 1913, xi, 641.

2. One of these reported by me, Adams-Stokes' Syndrome with Complete Heart-Block, without Destruction of the Bundle of His (*THE ARCHIVES INT. MED.*, 1910, v, 583), the first reported with complete block, Pardee incorrectly states to have followed typhoid. The exact cause was never ascertained, though it followed influenza, accompanied by a long period of physical and mental strain.

3. Holberton: *Med.-Chir. Tr.*, London, 1841, Series 2, vi, 76.

4. Neuberger and Edinger: *Berl. klin. Wchnschr.*, 1898, xxxv, 69.

5. Cohn, A. E.: *Jour. Exp. Med.*, 1912, xv, 49.

Furthermore, it is generally conceded that ganglion cells and a rich plexus of nerve-fibers accompany the muscular system.⁶ A complete pathological investigation should include, therefore, not only the sino-auricular node and the auriculoventricular system with its connections with the auricle and ventricle; but also the nerve-cells and fibers accompanying it, the vagi, and the central nervous system in neighborhood of the nuclei and roots of the vagi. This has not been done in the past for obvious reasons and will probably seldom be accomplished. It should also be remembered that even such an examination would fail to account for the functional type of block, caused by the action of toxins or deficient blood-supply.

For the purpose of determining the frequency of extensive lesions of the conductive system in the absence of heart-block, studies are now being conducted in this laboratory on appropriate post-mortem material.

In view of the increasing number of reports of heart-block, in which the clinical and pathological findings are contradictory, the following two cases are of interest, as in each case the findings at necropsy tally well with the clinical history. Unfortunately, in neither was it feasible to obtain polygraphic or electrocardiographic tracings during the period of block; but the clinical symptoms in each were of such a nature as to make it reasonably certain that at least transient block with Adams-Stokes' syndrome existed.

CASE 1.—Mrs. Kate H., aged 42, a housewife, was admitted to the Philadelphia General Hospital on the service of Dr. David Riesman Jan. 1, 1913, and died January 18.

Abstract of History.—For five weeks she had had cough with scanty and at times blood-tinged sputum. The cough had gradually been getting worse. There had been no definite loss of weight. On exertion there was dyspnea; but edema was not noted. She had to rise once at night to urinate. Her menses had ceased three months before. No noteworthy symptoms referable to the gastro-intestinal system were present.

She had no recollection of any previous illness, except an attack of rheumatism in 1903, which confined her to bed for seven weeks, and a "stroke" in May, 1911, which occurred with left-side paralysis during the night. The patient had been married ten years, had had no children, no miscarriages and no venereal history. She had used alcohol (principally beer) in moderation. A brother and a sister had died of nephritis.

Examination.—The skin was yellowish and transparent, mucous membranes very pale; nutrition fair. The arteries were hard and tortuous; pulse irregular, ranging from 70 to 80 at first, although occasionally it was 25 or 40. There was a tic of the facial muscles and tongue, more marked when talking. Severe pyorrhea alveolaris and follicular deposits in the tonsils were present. At the apex of the right lung were slight dulness and harsh breath-sounds. Posteriorly both lungs were resonant, but generalized bubbling râles were heard. The cardiac apex was palpable in the sixth interspace just inside the anterior axillary line. Dulness began above at the second rib, extended on the left to the apex and on the

6. Wilson: Proc. Royal Soc., 1909, lxxxi, Series B, 141.

right 1 cm. to the right of the right border of the sternum. On auscultation a loud, rough systolic murmur was heard at the pulmonary area, with reduplication and accentuation of the second sound; also a soft systolic murmur at the apex, not heard in the axilla or back. The abdomen was tympanitic. Tenderness could not be elicited and no organs or masses could be felt. The urine was reported negative, specific gravity 1.012.

Course.—January 8 the patient had a fainting spell. The patient next to her did not notice any convulsive movements. The pulse at 4:30 and 7 p. m. was 34. There were no visible pulsations in the neck.

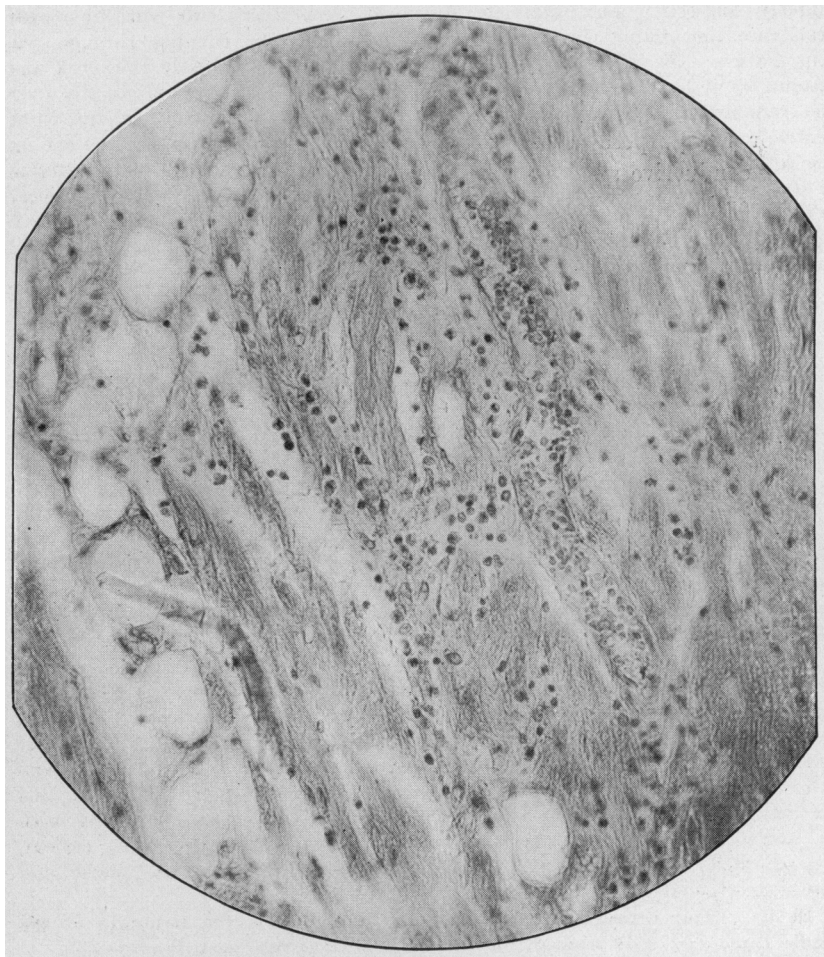


Fig. 1—Case 1, heart 31; cellular infiltration of main bundle (more marked around small blood-vessel). Zeiss DD, eye-piece 4.

January 16 the resident physician observed the patient in an epileptiform attack. The breathing was stertorous, the head was drawn to the left and the left arm twitched momentarily. The pulse dropped from 60 to 24, the patient was mentally confused and poorly oriented. The following day several attacks occurred, one of which was observed by Dr. Farr. Both arms moved slightly and the eyes turned upward. There was a brief period of unconsciousness and after-

ward the patient complained that everything looked black. Dr. John H. Musser, Jr., took a polygraphic tracing on this date after an attack, but found no evidence of heart-block. The pulse was then 60. The following day the patient died.

Clinical Diagnosis.—Chronic myocarditis, mitral insufficiency, Adams-Stokes' syndrome, pulmonary edema, old hemiplegia (embolic).

Anatomical Diagnosis.—In addition to the preceding, mitral stenosis, sclerotic and infarcted kidneys.

Gross and Microscopic Examination.—Heart 31, hardened in formaldehyd. Weight 390 gm. Moderate amount of fat in epicardium. Coronary vessels slightly sclerosed. Left ventricle 10 to 14 mm. thick, cuts with increased resistance and numerous fibrous threads can be seen running through the musculature. On the septal surface the endocardium is diffusely thickened and opaque, as if by extension from the thickened mitral valve, but especially over the ramifications of the left branch of His' bundle. Similarly in the right ventricle there is fibrous thickening at the bases of the chordae tendinae in the septum. The surface of the septum shows a dense mass of fibrous tissue in its middle, and near the insertion of the mitral valve a cartilaginous, almost calcified mass. The aortic valves are distinctly thickened, irregular and calcified. Small patches of sclerosis are seen on the intima of the aorta. The mitral valve is in the form of a distinct "buttonhole," the opening being about 5 by 15 mm. The pulmonary and auriculoventricular valves are normal.

HISTOLOGICAL STUDY OF HIS' BUNDLE AND KEITH'S NODE

A section 3 cm. long and 3 cm. wide was cut from the septum to include Tawara's node, His' bundle and its bifurcation. The anterior and posterior borders of the piece are almost parallel with the long axis of the right ventricle, the inferior being slightly more anterior. Considering the heart as being upright in the body, oblique (almost sagittal) serial sections were cut from behind, forward and up. About one thousand were cut, of which five hundred were mounted and about one hundred and fifty stained. Every fourth slide was stained with hematoxylin and eosin, and the succeeding slide with Mallory's anilin-blue. In all the sections both auricle and ventricle appear, with the delicate auriculoventricular valve on the right side, and the thick stumps of the aortic and mitral valves on the left.

In the earlier sections (that is, posterior) the network of Tawara's node is well shown, lying close to the fibrous base of the mitral valve. Herein are considerable delicate fibrous connective-tissue and occasionally small collections of fat-cells, but this picture shows nothing definitely abnormal. The cellular infiltrate, however (small round cells and plasma cells), is distinctly more than is ever seen in normal tissue. As far as can be ascertained, after the fixation in formaldehyd, the muscle-fibers of the node, their connection with the auricular musculature and the auricular musculature itself are normal, with characteristic narrow fibers, faint cross striations, numerous nuclei and loose connective tissue in normal amounts.

In its course through the auriculoventricular groove, the structure of the bundle cannot be well studied, but the cellular infiltrate is still noticeable.

Lower down, where the main bundle sits on the ridge of the ventricular septum (owing to the plane of the section the bifurcation appears in the same sections), the fat and cellular infiltration is well shown. The right branch can be followed for a short distance imbedded in the ventricular musculature and then disappears, enveloped on three sides by a dense mass of scar tissue (Fig. 2). The sections above and below this show that it is a real obliteration. The left lies superficially beneath the endocardium and also shows the increased number of cells.

Histological Diagnosis.—Chronic interstitial myocarditis and valvulitis (aortic and mitral), cellular and fatty infiltration and fibrosis of His' bundle.

CASE 2.—Mrs. Hattie R., aged 41, a housewife, was admitted to the Presbyterian Hospital on the service of Dr. R. Pemberton, March 3, 1913, complaining of dyspnea.

History.—The usual symptoms of a chronic, severe heart-trouble were present; namely, dyspnea, edema of the legs and abdomen and occasional palpitation and

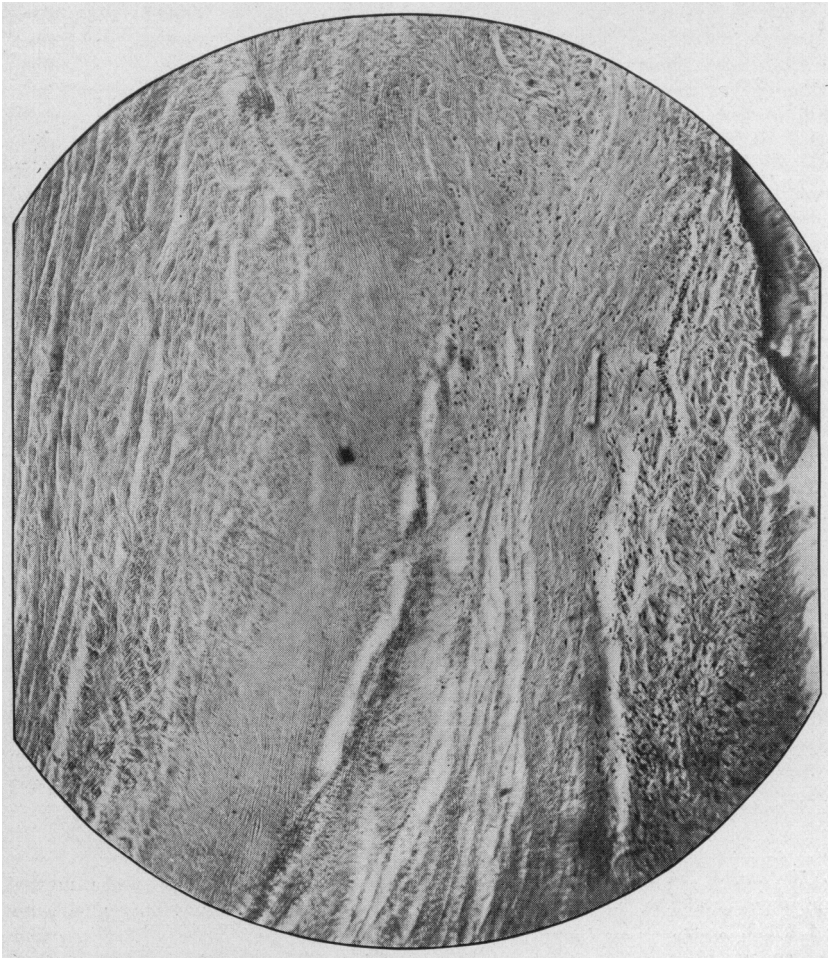


Fig. 2.—Case 1, heart 31; right branch, obliterated by fibrous tissue. Zeiss A, eye-piece 4. Two vertical bands of fibrous tissue (more dense and homogeneous than the surrounding myocardium) enclose the right branch of His' bundle. Near the center of the figure some cellular infiltrate appears in the muscle bundle, and just above this the two bands of fibrous tissue meet to obliterate it.

precordial pain, so severe that she could hardly walk. For three months she had had a severe cough. Four years before admission she had a febrile attack of rheumatism, sufficiently severe to confine her to bed for some days. Except for the usual diseases of childhood, her history and that of her family were negative.

Examination.—On admission her temperature was 99, pulse 108, respiration 38. Cardiac dullness extended from 1 cm. beyond the right border of the sternum to 2 cm. to the left of the midclavicular line. A presystolic murmur was noted with an accentuated second pulmonic sound. There was movable dullness in the abdomen and edema of the legs. Examination of eye fundi showed hemorrhagic retinitis. The urine contained a cloud of albumin and numerous hyalin casts. The Wassermann reaction was negative.

Course.—After three weeks of aggravation, and then of amelioration of the cardiac symptoms, fever set in with a leukocytosis of 26,000, and pelvic inflammation was diagnosed. On the day of her death, four typical Adams-Stokes' attacks were observed by Dr. Pemberton. With the pulse at 48, a violent epileptiform convulsion occurred, with loss of consciousness, jerking of limbs, bulging eyes, protruding tongue and stertorous breathing, and lasted perhaps three minutes. The pulse dropped to 14, but returned to the former rate after the attack. After recovering sufficiently to ask questions, she suffered another similar attack in which the pulse dropped to 24. In the third attack it was noted that the heart stopped entirely for some seconds; in the fourth and fatal attack, respirations stopped two minutes after the cessation of the heart-beat. No opportunity was offered for graphic registration of heart-beat and pulse. Necropsy was performed by Dr. C. B. Farr four hours after death.

Anatomical Diagnosis.—Double pyosalpinx, acute seropurulent peritonitis, acute fibrinous pericarditis and pleuritis with right-sided effusion; hypertrophy and dilatation of heart; mitral stenosis; atheroma of aorta; chronic diffuse nephritis; chronic passive congestion of liver and spleen; parenchymatous degeneration of liver.

Gross and Microscopic Examination.—Heart 15, received in formaldehyd, weight 650 gm. Parietal and visceral pericardium much thickened with heavy deposits of fibrin, obscuring the various relations of heart. Pneumogastric terminations could not be found. Heart is homogeneous, yellowish-brown, firm, and cuts with increased resistance; fibrous strands are visible here and there, and several grayish, translucent nodules are seen. The left ventricle is from 12 to 14 mm. thick; the right ventricle from 4 to 6 mm. The trabeculae are well developed. The aortic valves are distinctly thickened with numerous atheromatous patches on the ascending arch of the aorta. The mitral valve is much stenosed and thickened but without distinct nodules. The left ventricular branch of His' bundle is visible as a fan-shaped, spreading bundle. One so-called abnormal tendon fiber with very slight endocardial thickening runs to the posterior papillary muscle. No other abnormality is visible. The coronary arteries are diffusely sclerosed.

HISTOLOGICAL DESCRIPTION

The whole heart septum from the lower border of the fossa ovalis in the auricle to the apex of the ventricle was removed and cut into blocks suitable for serial section. The anterior and posterior borders were cut almost parallel to the long axis of the right ventricle with the lower portion slightly forward and the superior and inferior at right angles. One piece of tissue was cut to include Tawara's node and as much as possible of His' bundle, and sections were taken on the frontal plane. In spite of fixation in formaldehyd excellent histological detail was as a rule shown.

In the earlier sections, the muscular network of Tawara's node was found normal. Adjacent to it, beneath the endocardium of the right ventricle, were numerous swollen, vacuolated, faintly striated cells resembling Purkinje fibers. Some of these contained hemosiderin pigment granules. A small collection of plasma and small round cells were present on the edge of Tawara's node.

In later sections these small collections were found to contain many polymorphonuclear leukocytes (10 to 40 per cent). They occur both subendocardial and buried deep in fibrous or muscular tissue, usually around small vessels.

In later sections, in which the bundle was found traversing the fibrous band between the auricle and ventricle, it was approximately normal with little or no increase of fibrous tissue. The same deposits of small round cells, plasma-cells and leukocytes were found in it.

Still later the division of the bundle was shown, but the right branch could not be followed well owing to poor fixation. The left appeared in cross-section or oblique section immediately beneath the endocardium. The musculature in its upper portion, was about normal, though it also was poorly fixed. Further down, the chains of Purkinje-like cells, well separated by loose fibrous tissue from each other and from the ventricular musculature, were well shown. The cellular foci before described were also present and were much more abundant than through the ventricular muscle proper. These foci could be followed the length of the septum, with occasional extensions into the septa between the ventricular muscle bundles. The actual junction of the Purkinje fibers with the muscle-fibers was not seen. The base of the auriculoventricular valve appeared edematous and contained similar cellular foci. The valve showed no acute endocarditis. A diffuse fibrosis of considerable extent existed throughout the ventricular musculature. In places, dense fibrous masses without nuclei were found at times with a core of more fibrous tissue, and one of these occurred subendocardially directly in the path of the left branch (gumma).

Sections from Keith's node (parallel to the crista terminalis, 8 by 20 mm.) failed to reveal any abnormality, other than the inclusion of a small number of cells similar to those described above.

Histological Diagnosis.—Acute inflammation of Tawara's node and bundle of His; diffuse fibrosis and multiple gummata of ventricle.

SUMMARY

In Case 1, in which there were transient attacks of heart-block, complicated with Adams-Stokes' syndrome during the last eight days of life, there was found a marked cellular infiltration of the auriculoventricular conductive system. Furthermore, the right branch was found completely obliterated, showing that total destruction of one branch is compatible with an at times normal pulse.

In Case 2, chronic salpingitis, in a woman who sought the hospital for relief from a chronic rheumatic endocarditis with failing compensation, the hitherto localized septic process caused, three weeks after admission, a general peritonitis, pleuritis, pericarditis, and localized areas of acute myocarditis, especially in His' bundle and Tawara's node. On the day of her death, due presumably to the heart-block thus caused, there occurred four typical attacks of Adams-Stokes' syndrome, the last of which was fatal. The only other reported case that I know of in which an acute septic condition of the conducting system was associated with heart-block is that reported by Gerhardt,⁷ in which a patient who had

7. Gerhardt: *Deutsch. Arch. f. klin. Med.*, 1908, xcii, 485.

just recovered from an attack of acute articular rheumatism with temporary heart-block succumbed to typhoid fever, and at necropsy a perivascular cellular infiltration was found in His' bundle, the residuum of an acute inflammation.

I am indebted to Drs. Riesman, Farr and Pemberton for the clinical reports and necropsy material.