STOKES-ADAMS DISEASE

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Definition.—A condition of slow pulse with syncopal, apoplectiform, or epileptiform attacks associated either with (a) derangement of the junctional system of the heart, or (b) disease of the nerve-centres of the vagi or of the nerves themselves.

History.—Morgagni gave the first description of the disease. "I will just skim over . . . those many things which I have observed for a long time in my fellow-citizen, Anastasio Poggi, a grave and worthy priest. He was in his sixty-eighth year, of a habit moderately fat, and of a florid complexion, when he was first seized with the epilepsy, which left behind it the greatest slowness of pulse, and in like manner a coldness of the body. But this coldness of the body was overcome within seven hours, nor did it return any more, though the disorder often returned; but the slowness of the pulse still remained." Our modern knowledge dates from the work of two Irish physicians, Adams and Stokes, whose original descriptions may be summarised as follows:

Adams' patient, aged sixty-eight, was of full habit, and subject to oppression of breathing and cough. He was first seen when recovering from the effects of an apoplectic attack, which had come on suddenly three days before, but he was sufficiently well to be about the house and even to go out. What attracted Mr. Adams' attention was the character of the breathing and the remarkable slowness of the pulse, 30 to the minute. His regular attendant informed Mr. Adams that during seven years this patient had had not less than twenty apoplectic attacks; after a day or two of heaviness and lethargy he would fall down completely insensible, and on several occasions had hurt himself. The pulse would become slower than usual, and the breathing loudly stertorous. He never had any paralysis after the attacks. Death followed an attack, and the heart was found to be very fatty, the valves being sound. There was no statement about the coronary arteries.

In his much more important contribution, entitled *Observations on Some Cases of Permanently Slow Pulse*, Stokes describes the case of a man, aged sixty-eight, who had recurring fainting fits which, however, did not leave any unpleasant effects behind. In the course of three years he had had at least fifty seizures, which were induced by any circumstance tending to impede or oppress the heart's action, such as sudden exertion or a distended stomach. He was never convulsed and never had any paralysis. The duration of the attack was seldom more than four or five minutes, and during this time he was perfectly insensible. On admission his general health seemed very good. There was an apical systolic murmur,

and a pulse of 28 in the minute. The arteries appeared to be in a state of permanent distension, "the temporal arteries ramifying under the scalp just as they are seen in a well-injected subject." An interesting feature in this case was that the patient could ward off attacks by a peculiar manœuvre; "as soon as he perceives symptoms of the approaching attack he directly turns on his hands and knees, keeping his head low, and by this means, he says, he often averts what otherwise would end in an attack." It was noticed that, while his heart-beats were reduced to 28, there were occasional semi-beats between the regular contractions, 8 of them in the minute. On readmission an entirely new sign was observed, namely, a remarkable pulsation in the right jugular vein, the rate of which was more than double that of the manifest ventricular contractions. Subsequently cases were reported elsewhere from time to time; in France Charcot called attention to it in 1872, and in his *Traité des Maladies du Cœur* (1889), Huchard gave an admirable description of the condition, which he named Adams-Stokes disease. In this country very little attention was paid to it until the appearance of Dr. Webster's paper, in which the cases were carefully studied by the graphic methods. In 1903 I drew attention to the comparative frequency of the condition and to the various groups of cases. In Germany the studies of W. His, junior (1899), Jacquet (1902), and Luce (1902) stimulated a physiological interest in the subject, and brought the syndrome into relation with the phenomena of heart-block described twenty years before by Dr. Gaskell. In the United States the clinical features of slow pulse were very fully considered by Prentiss and by Edes (1901). Recently the brilliant anatomical demonstration by Tawara of the intracardiac junctional system of fibres, the physiological studies of Erlanger on heart-block, and the pathological investigation of the cases by Dr. Keith and others have combined to arouse the keenest interest in the subject. More than sixty separate papers have appeared within the four years 1905-8, references to which may be found in the *Index Medicus*, and in Pletnew's critical digest in Vol. I. of the *Klinische Ergebnisse* (1908). Bachmann has collected 177 cases of the Stokes-Adams syndrome.

Nomenclature.—Bradycardia, a very common condition, is due either (i.) to influences acting on the nerve-centres (or the vagi) or (ii.) to changes in the heart itself; and the cerebral features which characterize the Stokes-Adams syndrome may be associated with both these, neurogenous and cardiac, groups. The disturbance of rhythm which we know as heart-block is not an invariable accompaniment of bradycardia. I do not know that its existence has been determined in the cases due to organic disease of the medulla, the cases in which the heart simply slows down, without change in its rhythm. The syncope and epileptiform features are epiphenomena of a bradycardia however induced. Whilst in some ways it is a pity that the syndrome was ever labelled a disease, yet, as in the case of angina pectoris, the clinical picture is very definite however varied its pathology. Since it is too

late to attach Morgagni's name to it, we may continue to call the syndrome or disease by the name Stokes-Adams; and, remembering the dictum of Socrates, that "it matters little what names you give to things so long as you tell us just what the things are," I may repeat that under this designation a description will be given of the cases with slow pulse and syncopal and epileptiform attacks, and in two groups—cardiac and neurogenous.

Incidence, Age, and Sex.—That the disease is not uncommon is shewn by the number of cases reported in recent years. In 1903 I reported 13 cases, and since then I have seen 7 additional examples. Men are more often affected than women—in my series all were men. The age-incidence varies with the cause; in a small group, due to acute infections, young persons may be attacked; Schuster indeed reported it in a girl of four years of age. In the group due to syphilis, men between the ages of twenty and forty are affected; but the great majority of the victims are men who have reached the period of arterial degeneration. Of my 20 cases, 2 were between thirty and forty, 6 between fifty and sixty, 8 between sixty and seventy, and 4 above seventy. My oldest patient was a man of seventy-four.

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