Griffith, J. P. C.

A CONTRIBUTION

TO

THE STUDY OF

FRIEDREICH'S ATAXIA.

WITH EXHIBITION OF CASES.

BY

J. P. CROZER GRIFFITH, M.D.,

INSTRUCTOR IN CLINICAL MEDICINE IN THE UNIVERSITY OF PENNSYLVANIA, AND ASSISTANT PHYSICIAN TO THE HOSPITAL; PHYSICIAN TO ST. AGNES' HOSPITAL; PATHOLOGIST TO THE PRESBYTERIAN HOSPITAL.

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[Read February 1, 1888.]

No better commencement of this subject can perhaps be made than by repeating the oft-quoted definition formulated by Friedreich himself, as follows: "There exists a chronic inflammatory degeneration of the spinal cord leading to atrophy, and which seems especially liable to develop at puberty under the influence of an hereditary predisposition. It is chiefly confined to the posterior columns, and, beginning in the lumbar part of the cord and spreading in both an upward and downward direction, it finds its limits in the medulla, after involving the centres and trunks of the hypoglossal nerves. The affection is characterized clinically by a disturbance of the coördination and harmony of the bodily movements, which develops very gradually, advances from the lower to the upper portion of the body, and at last invariably involves the organs of speech; though there is no disturbance of sensibility, and the special senses and cere-

bral functions remain intact. There are neither paralyses of the sphincters nor trophic affections. Less constantly there appear curvature of the vertebral column, vertigo, and nystagmus. From a clinical point of view the disease may be regarded as a chronic progressive paralysis of the power of combination of movements; and from the standpoint of pathological anatomy, as a chronic degeneration of the posterior columns of the spinal cord."

This definition still holds good in part, though in the light of the cases which have been reported since it was given, it must undergo certain modifications. The following, based upon it, will perhaps better accord with our present knowledge of the affection: Friedreich's ataxia is a chronic systemic inflammatory degeneration of the spinal cord, developing usually in infancy or childhood, and occurring in cases in which there has probably been an arrest of development of the cord during fœtal life, this being the result of some hereditary predisposition. It is situated chiefly in the posterior columns, the lateral pyramidal tracts, the direct cerebellar tracts, and the columns of Clarke, though other parts of the white and gray matter are very commonly somewhat affected, and the sclerosis extends slightly into the medulla. The affection is characterized clinically by a disturbance of the coördination of the bodily movements, developing gradually, advancing from below upward, and finally involving the organs of speech. Curvature of the spine, talipes, vertigo, and nystagmus are frequent. The patellar reflex is nearly always abolished. Paralysis and sensory disturbances are not uncommon in advanced cases. Trophic, vaso-motor, and visceral affections are unusual, and any involvement of the intellect is probably accidental.

In the course of some studies on this disease I found so many omissions in and discrepancies between the tabulated lists of cases made by various authors, and so many new cases in the literature, that I have investigated for myself all the reports of instances of the disease, so far as I have been able to learn of any. To render the results as accurate as possible, the original publications have been consulted in every instance but three; these being the article of Erlenmeyer, of which, however, a literal translation was found; that of Jakubowitsch, an abstract of which was seen in one of the German journals, and that of Palma, whose case is nevertheless detailed in full by Vizioli. Personal communication with Drs. J. J. Putnam, G. B. Shattuck, W. B. Gowers, W. Everett Smith, W. C. Warren, and Morton Prince has aided me greatly, and I take this opportunity to express my thanks to these gentlemen for their kind and valuable assistance.

I desire to offer to the College a new tabular arrangement of cases, with their most important symptoms. Table I. contains 143 cases,[1] comprising all those from

[1] Through an oversight, the case of C. L. Dana, and that of V. P. Gibney (New York Med. Record, Oct. 1, 1887, p. 465) were omitted. The important features of these cases are as follows:

CASE 144.—Dana's; male; aged nineteen. Father probably alcoholic; no other case in family. Ataxia of gait and speech, and constant thirst, appeared at seventeen years. Examination showed marked ataxia of gait; Romberg's symptom; some ataxia of arms; slight oscillation of head; slight paralysis; slight curvature of spine; slight talipes equinus; knee-jerk normal; no atrophy; hands cold and red; no pain; slight anæsthesia; slight diminution of muscle sense; legs sometimes numb; speech slow, thick, and scanning; almost imperceptible nystagmus; apparently beginning optic atrophy; sometimes vertigo; polyuria.

CASE 145.—Gibney's; male; aged seventeen to twenty years. Disease known to have existed at six to nine years; ataxia of gait very marked; speech ataxic; no pain; no cause of disease known.

the literature appearing to have any just claim to be considered probable instances of Friedreich's ataxia, and including those exhibited to-night by Dr. Sinkler, Dr. Osler, and myself. Objections might with propriety be urged against some of the number, but it is not the purpose of this paper to assume the right of discrimination beyond that which is absolutely necessary, but simply to collect and analyze the cases as an aid to future study. To Table II. (Additional List) are relegated: A. *Reputed cases*, containing all those reported and related by blood to those in Table I., but which have not themselves come under professional observation. B. *Possible or probable cases*, merely referred to and without published descriptions, so far as I have been able to ascertain. C. "*Too doubtful cases*," as they might be named, claimed by the observer or by critics to be instances of the disease, but which differ so widely from its complex of symptoms that it seemed almost certain that the general consensus of medical opinion would reject them.

I beg leave this evening to exhibit to the College three cases of Friedreich's ataxia, all of them inmates of the Home for Incurables and Cancer Hospital, of this city, where I have repeatedly examined them during my terms of service. For their presence here to-night I am indebted to the courtesy of my colleague, Dr. Horace G. Hill.

Case I.—Sadie T., aged twenty-three; single. Father and mother living and well; maternal grandmother died of apoplexy; no history of any nervous disease in family. There were 7 children, of whom 1 died of measles, 1 of some febrile affection; 1 brother and 2 sisters are well, and 1 brother (George) suffers from this same disease.

The history obtained from the mother and the patient herself is as follows:

She was well and active until about ten years of age, at which time the mother noticed that the child "was nervous." Her hands shook while eating, and her gait was somewhat unsteady and staggering, though she did not often fall. The mother, who is an intelligent woman, is positive in the statement that the disease did not resemble chorea, of which she had seen instances. On account of the nervous affection she was entered, a few months later, at the Children's Hospital of this city, and soon after passed through a severe attack of typhoid fever, followed very soon by measles. After recovery the nervous affection was found to be much worse, and the patient was unable to walk without crutches; and, even after strength returned to other parts, the legs remained weak. The affection of the upper extremities had not meantime grown worse, and was but slight. The patient complained at this time of a sense of burning in the feet and aching in the legs, but there is no history of true lightning pains. Sensibility in the legs to touch and to pain appears to have been undisturbed, for the mother used frequently to test it herself with the point of a pin, and could perceive no alteration. The patient walked with crutches until the age of sixteen, was able to go upstairs on the hands and knees, and to rise from the floor by grasping the furniture; yet the disease had been constantly progressing in the lower extremities.

In November, 1880, at the age of sixteen, she was examined at the Infirmary for Nervous Diseases, of Philadelphia, and the following note entered in the case-book: "She can feel perfectly. No tendon reflex. Adduction of thighs very weak, but the patient can make all movements of feet and legs, but is utterly unable to stand. Patient stout and of a good color. When on back, on attempting to lift legs, they fall outward. There is some choreic movement of the hands." She entered the Home for Incurables December 29, 1880, being still able to

walk with crutches, though with difficulty. The incoördination of the hands now grew worse. She rapidly lost the power to walk at all, and her speech became affected. It was impossible for her to sit upright, and a brace was made for her. Her mother says that her intellect has remained unaffected.

Present condition, August 16, 1887: The patient is of average size, and of a bright and intelligent expression. She sits during the day in a wheel-chair, which she is able to propel. She cannot sit erect, but slouches down in the chair, and holds her head far forward. There is some scoliosis to the right in the dorsal region. She is unable to walk or stand, or, indeed, barely to move the legs at all; nor can she raise herself from a recumbent to a sitting position while on the bed. Both feet are in a position of slight equino-valgus, but there are no other contractures. The muscles, both of the thighs and legs, are firm, and there is no more atrophy than would naturally follow lack of use. The patellar reflex is abolished. The grip of the hands is good; that of the left being somewhat better than that of the right hand. In grasping an object the fingers and hands are extended over it like a claw. The fingers are held rather flexed, and the patient is unable to extend those of the right hand very well, though there is no resistance to passive motion. There is no motion or tremor of the hands when in repose, but on voluntary effort a slow ataxic movement of the fingers develops, somewhat resembling athetosis. She picks up a pin with the greatest difficulty, and with marked incoördination, using the whole finger and not the tip, much as an infant would. She cannot cut her own food, but can feed herself. She can scarcely touch the tip of the nose with the finger, but this difficulty is not definitely increased when the eyes are shut. The muscles of the arms are perfectly well developed, and there is apparently no diminution in strength. The handwriting is almost illegible. During the effort to write, or under other excitement, the head develops irregular lateral oscillatory movements; and a slight jerking motion is almost constantly present when speaking, though there is no true tremor. The tongue is extended steadily. The speech is peculiar, somewhat

jerking, but not scanning; several words being spoken rapidly, then a brief pause ensuing, followed by another series of words, the pause sometimes occurring between syllables. The pupillary reflexes are normal. There is very slight and inconstant nystagmus on extreme lateral movement. The result of the ophthalmoscopic examination of the eye-ground, kindly made for me by Dr. Charles S. Turnbull, was entirely negative. The functions of the bladder and rectum are normal, though the urine is alkaline, albuminous, and contains numerous leucocytes. There is no affection of the thoracic and abdominal viscera, and menstruation is regular. There are no marked disturbances of sensation. The head and point of a pin are differentiated perfectly, both on the upper and lower extremities. The points of the æsthesiometer are recognized at six centimetres on the extensor surface of the legs, at four centimetres on the forearm, and at about one-half centimetre on the tip of the third finger. The sensibility to pain and to temperature is unaffected. There are no pains in the legs, but a girdle sensation is sometimes perceived. Owing to the slowness in speaking, it is uncertain whether any retardation of the conduction of sensation exists, though it seems probable. No electrical examination was made.

The history of the next case is most interesting, and, I believe, in certain respects, unique. It would seem as though some other nervous affection, perhaps neuritis or anterior poliomyelitis, had preceded the onset of Friedreich's ataxia by about eight years, and that the atrophy of the left leg is probably to be attributed to it.

Case II.—George T., aged twenty-one, brother of last patient. The mother relates that he was well as a child, but had a fall from a high step when two years old and hurt his left hip, though he soon recovered, and no traces of injury remained as far as could be detected. Soon after this he got his feet wet while wading, and on the same evening developed a

high fever, which disappeared the next day, but left him with feet and legs very much swollen as far as the hips, and entirely unable to stand. He was taken to see Dr. G. W. Norris, who pronounced it paralysis; and again, a few days later, to Dr. S. D. Gross, who gave the same opinion, but was not sure whether it was due to the fall or to taking cold. After one to two weeks the swelling began to disappear, and the child commenced to creep, and a few weeks later to walk. The right limb now appeared to be perfectly normal, but the left was weaker and decidedly wasted, and a brace was fitted to it. Slow improvement followed, and after two or three years the brace was replaced by a stiffened shoe. At the age of eight years he "threw" the left lower limb when he walked, and both leg and thigh exhibited considerable wasting; but the mother says that sensation to a pin-prick seemed entirely unaffected. She had further never noticed any affection of the hands, nor any other disturbance of the gait except that described. At nine years of age (in 1875) he was severely sick with measles, followed by pneumonia. On recovery it was found that he had lost the power of his lower limbs, and had difficulty in sitting erect. Curvature of the spine soon developed, but the upper extremities remained uninvolved; and after four months he was able to walk with the aid of some one or with crutches. He then learned to walk from one article of furniture to another rapidly, much as an infant would; but slow locomotion was accompanied by much swaying and frequent falls, from which he was able to rise again by putting his hands on his knees, or by grasping objects in the room. Improvement was very slow, if indeed there was any; and in November, 1884, at the age of fourteen, he was admitted to the Infirmary for Nervous Diseases, where the Achilles tendon of the left foot and some other tendon of the right foot were cut. After three months' treatment he was fitted with a brace for the left leg, reaching up to the waist, and with a short one for the right leg; walking tolerably well with the aid of these and of a crutch. He says the doctors at the Infirmary used to remark that it was strange he could not use his right leg better, as it was so well devel-

oped. He was admitted to the Home for Incurables in 1884, at the age of eighteen, and could still walk with crutches. Since admission his hands have become unsteady, and he is no longer able to sit erect without support, or to walk. The date at which the affection of speech began is uncertain. He has never had lightning pains or girdle sensation, though he sometimes experiences painless spasm of the extensors of the right leg at night.

Present condition, August, 1887: The patient, of average size, has a good color and general nutrition. He sits through the day in a wheel-chair, in the same unnatural position as his sister, though to a less degree. The lower extremities are rather cold and red. The muscles of the right leg and thigh are quite firm, and there is no marked wasting. He can flex the thigh on the trunk with some difficulty, and partially extend the leg, but the muscular power is decidedly diminished. The left lower extremity is very greatly wasted, the patient being barely able to move it at all. There is no knee-jerk on either side. The spinal column shows a marked scoliosis to the right in the dorsal region, and the patient cannot raise himself upright in his chair without the aid of his hands. The muscles of the arms are unusually large and well developed, and the grip is powerful. When in repose the hands lie quietly in the lap, but voluntary movements are executed with marked incoördination, which is not notably increased on closing the eyes. The patient buttons his clothes with difficulty, and is unable to bring the forefinger tips together or to touch the ear or nose with accuracy. He can feed and dress himself, and write, but the handwriting is very unsteady. There is occasionally a slight ataxic movement of the head, though the tongue is extended without tremor; the speech is a little jerking. There is slight ataxic nystagmus on lateral motion; the pupillary reflexes are normal. An ophthalmoscopic examination by Dr. Turnbull revealed no alteration of the eye-ground. The thoracic and abdominal organs appear to be normal. There is no pain; no girdle sensation; no affection of the bladder or rectum. There is retardation of the conduction of sensation

in the right leg, with slight diminution of tactile sensibility on the outer surface, but not over the anterior portion of the tibia. No positive sensory changes can be detected in the left leg. In the hands and arms there is evidently a diminution in the power to distinguish the point from the head of the pin, and the æsthesiometer points are recognized on the dorsum of the right hand at four centimetres and on that of the left hand at six centimetres. The muscle sense is decidedly involved in the lower extremities, as the patient cannot clearly define the position of his feet when his eyes are shut.

Case III.—Annie C., aged twenty-six years. The parents are living and well, and no history of any nervous disease in the ancestors can be elicited. Of 10 children, 2 died in infancy, 6 are in good health, and 1, a son, died of "weakness" at the age of nine years, having developed, after scarlet fever, the same unsteady gait as Annie at one time exhibited. She was well until five years of age, when she had scarlet fever, though not in a severe form, and without complications. She seemed, however, after recovery, to be very weak in the legs; had a swaying, somewhat staggering gait, with frequent falls, and could not run upstairs. When eight years old she wore, for a year, braces on both legs, extending to the hips. She went to school till twelve, but walking then grew so much worse that she was obliged to lean on her sister for support, and soon to leave school altogether, and she began the use of crutches. Weakness in the legs progressed, and at sixteen she could no longer walk at all. At twenty she was an inmate of the Jefferson College Hospital, where she received electrical treatment, but without benefit. When about twenty-two years old she first began to notice weakness in the arms, but she does not remember when the difficulty in sitting erect or in speaking began. She says that at times she cannot talk as fast as she wishes, and that she sometimes chokes when swallowing liquids; and I have myself once seen her in a severe attack of choking from swallowing saliva. She does not suffer from headache or

other pain, but complains often of cold feet. The left leg is sometimes drawn up spasmodically at night.

Present condition, August, 1887: The patient is of average size, for the most part well nourished, and of a slightly apathetic expression. Her position in sitting is like that of Case I. There is some scoliosis in the dorsal region. The feet are shorter than natural, with some talipes equino-valgus, and with some contraction of the flexors of all except the great toes. There is but very slight atrophy of the muscles of the lower extremities. She is not able to extend the legs at all, and can flex the thighs but very little. The knee-jerk is abolished on both sides. The hands are rather claw-shaped, with the proximal phalanges partially extended on the dorsum of the hand, and the remaining ones flexed; but the patient can straighten them at will, and there is no resistance to passive motion. The ataxia of the upper extremities is very great, and she is scarcely able to pick up a pin, or with the eyes closed to bring the forefingers together, or to touch the ear or nose. With the eyes opened this incoördination is very little, if any, less, but when in repose the hands remain quiet. The arms are somewhat wasted; the grip fairly good, but rather infantile, small objects being grasped with the radial side of the index finger and the thumb. The head exhibits slight irregular swaying and nodding movements, intensified when talking or under excitement. The tongue is extended with a slight trembling motion, and speech is slow and scanning. There is slight lateral nystagmus on extreme lateral movement of the eye; the pupillary reflexes are normal; and Dr. Turnbull reports ischæmia of the retina and venous pulsation in both eyes, but no other alteration of the eye-grounds. The intellect is entirely unaffected; the functions of the bladder and rectum are normal, and there is no disturbance of menstruation. Physical examination of the abdominal and thoracic organs reveals no change in them. There are no lightning or other pains, and no girdle sensation. The tactile sensibility of the legs is evidently diminished, although the head and point of a pin are differentiated quite well. She is unable to feel the two points of the æsthesiometer

at a distance of ten centimetres on the forearm, and on the dorsum of the hand their recognition is very uncertain, but she can nearly always distinguish the point and the head of a pin. The slowness of speech renders it difficult to determine whether the conduction of sensation is retarded.

The cases of Dr. W. C. Warren, of Waterford, Mississippi, referred to by Dr. Hammond in the *Journal of Nervous and Mental Diseases* for 1882, are so briefly reported there that I have written the observer for further particulars concerning them, which he has kindly sent me, with permission to publish the history of the cases up to date, including the account of a fourth patient in the same family.

Family history: The paternal grandmother is still living, but has been paraplegic for over thirty years. The father is healthy, but of a very excitable disposition. With these exceptions there is no nervous disease in the family, and no known history of tuberculosis, alcoholism, or syphilis. There are 9 children, 4 of them the patients to be described. The disease affected the 1st, 4th, 5th, and 8th children; the others being healthy and well developed.

Case I.—Nannie W.; aged twenty-nine; was healthy up to the age of eight years, when the parents noticed that she staggered when first rising in the morning, and could only stand steadily after balancing for some time with extended arms. After a little exercise the unsteadiness would disappear, but as time elapsed it grew constant, the gait being oscillating from side to side, and the feet striking the floor with considerable force. At about the age of twelve curvature of the spine was noticed and the arms became involved. At thirteen she had a severe attack of bilious remittent fever, to which affection she was subject, and has never been able to walk since. The motions of the arms also grew very incoördinate and of a choreiform nature, and the patient was unable to button her clothes

or to touch the tip of her nose with her index finger with the eyes shut. Talipes was noted at an early period. There had been only slight pain, between the foot and knee. At twenty-one the affection of speech began, and has advanced to an extreme degree. Up to the age of twenty-three the patient was able to extend her hand in salutation, though with difficulty, while the arm oscillated involuntarily up and down and from side to side several times before the hand of a friend could be grasped. For the last two years she has been entirely helpless. Menstruation began at twelve to thirteen, and has continued regular. The patient complains of vertigo, and suffers from some incontinence of urine, while at times there are involuntary painless spasms of the right leg, occurring chiefly at night.

Present condition: She lies curled up in bed with the knees nearly touching the chin. There is marked curvature of the spine (lordosis). Intelligence at first appears deficient, but careful examination shows the diminution to be slight, as far as it is possible to judge in the presence of the great affection of speech. For a few months she has been almost constantly crying without any known cause. She is able to read ordinary print for hours, but sometimes vision fails, and the type seems blurred, though no ophthalmoscopic examination has ever been made. There is no nystagmus. Speech is nearly unintelligible, even to the parents, and there is slight trembling of the tongue. The arms are almost powerless, and are held flexed, as are the hands and fingers; and there is marked atrophy of the muscles, especially of the extensors. The legs are also very helpless; the muscles wasted, particularly the extensors; talipes equinus is present; the knee-jerk absent. There is no girdle sensation, and at present no pain. Cutaneous sensibility is about normal. The electrical reactions were not tested, but three years ago the faradic contractility was exaggerated. The surface of the body is cold; the pulse persistently rapid and small.

CASE II.—Thomas W.; aged seventeen; fourth child; healthy till eight years of age, when the disease began in exactly the

same way as in the case of the sister,—*i. e.*, with unsteadiness when arising from bed. The gait became more and more oscillating and uncertain, and the feet struck the floor with force. Finally, at fifteen, walking without assistance was no longer possible. At twelve the arms became involved, and at fifteen very great incoördination with choreiform movements had developed. The speech has quite recently become affected. There has been at times slight pain between the foot and knee, as in the sister's case. More recently there has been pain in the region of the dorsal spine.

Present condition: The intellect is unaffected; there is no vertigo; vision is good; no nystagmus; speech slightly slow and hoarse; no trembling of the tongue; head held steadily; no ataxic movements of the trunk; decided scoliosis in the dorsal region. The hands and arms exhibit marked incoördination, and the patient cannot button his clothes, and touches the tip of the nose with the index finger with difficulty. These and all motions become much more ataxic when the eyes are closed. Station is swaying, and the patient has a tendency to pitch forward. On closing the eyes the oscillation increases very greatly, and he falls unless holding to some support. The muscles in general are somewhat atrophied, and there is some loss of power. Knee-jerk is lost; talipes equinus of both feet is present; there is a slight tendency to spasmodic contractions of the legs at night. Cutaneous sensibility is normal; there is no girdle sensation, and no affection of the bladder or rectum. The surface of the body is cold, and the circulation feeble.

Case III.—Robert W.; aged fifteen; fifth child. The disease in this case also began when the patient was eight years of age, and with the same staggering when first rising in the morning. The oscillation in walking progressed; the arms became involved at twelve years of age; the speech is still unaffected. Some pain in the legs below the knees, and a sense of weakness in the back is complained of.

Present condition: Intelligence good; there is decided vertigo;

the vision is good; no nystagmus; no ophthalmoscopic examination has been made. The speech is unaffected, and there is no trembling of the tongue. The head does not oscillate; the movement of the arms is very ataxic and of a somewhat choreiform nature, and buttoning of the clothes is impossible; while incoördination is increased on closing the eyes. There are no ataxic movements of the trunk. Scoliosis in the dorsal region is present. The gait is oscillating, staggering, only possible when the patient supports himself by the furniture or uses a cane. The station is swaying, and becomes impossible when the eyes are shut. Talipes equinus is present. There is no patellar reflex. Decided atrophy of the muscles is observed in the upper and lower extremities, and there is a certain amount of paralysis. The cutaneous sensibility is unaffected. There is no girdle sensation, and no affection of the bladder or rectum. The surface of the body is cold and the circulation poor.

Case IV.—Nettie W.; aged eight; eighth child; is just beginning to show symptoms of the disease. She was always healthy, but since the age of six years it has been noticed that she staggers when rising in the morning, and only after some effort, and by repeated placing of the feet on the floor and balancing with the arms outstretched, can she succeed in preserving her equilibrium. After a little exercise the ataxia of station disappears. There is no affection of speech; no nystagmus; no curvature of the spine; no paralysis. The knee-jerk is diminished in both legs.

To study the characteristics of Friedreich's ataxia more in detail, the subject may conveniently be divided under the following headings:

History.

The first cases, 6 in number, were reported by Friedreich in 1861 before the Thirty-second Congress of Ger-

man Naturalists and Physicians, held at Spiel, but they were not published in full until 1863. In 1865[1] Carre described a case in his monograph on Tabes dorsalis. From 1872 to 1876, inclusive, 6 cases were reported in England by Bradbury, Carpenter, and Dreschfeld, and 2 in America by Kellogg; and in 1876 Friedreich published his last set, thus making 20 instances in the literature, including the 2 of Quincke quoted by Friedreich, but which, for the sake of convenience, are tabulated later, since Rütimeyer has described them more fully. By the end of 1880 the number had increased to 31; with the close of 1882 to 47; with the beginning of 1885 to 71; and now reaches 143. There have appeared several valuable critical reviews, with or without reports of cases, as, for example, those of Féré, Möbius, Cuche, Ormerod, Bury, Sepilli, Vizioli, and others. I would here especially acknowledge my indebtedness to those of Ormerod, Vizioli, and Bury, of which I have made free use.

Name.

Friedreich first described the disorder as "Degenerative atrophy of the posterior spinal column," but this title quite fails properly to distinguish it. The name "Friedreich's disease," favored by Brousse and probably by most writers, though at one time perhaps the best, has recently become very objectionable, since paramyoclonus multiplex is frequently spoken of under the same title. "Congenital ataxia," as employed by Mastin, may lead to misapprehension, since, though it is

[1] This is the date given in the reference in Friedreich's paper, and also that of the copy of Carre's work which I consulted, and which there is no reason to suppose was other than a first edition, though the usual date assigned by authors is 1862.

probable that the affection is congenital, it has not been proved so. "Hereditary ataxia" as it was called by Friedreich, is in few cases a proper appellation, for reasons which will be explained later. "Family ataxia," suggested by Féré, is a less objectionable name than the preceding, since all the 143 cases have occurred in but 77 families of brothers and sisters. 2 sets of Musso's cases were related as cousins, as were 4 sets of Rütimeyer's; while 2 of Vizioli's patients and 4 of Smith's were children of ataxics. The number is thus reduced by 6, making 71 entirely unrelated families. Yet it is to be noted that the disease does not always occur in several brothers and sisters, since there have been 39 instances in which but 1 ataxic child was *seen by a physician* in the family, and of 24 of these it is stated that the patient was the only child affected by the malady. It becomes evident, therefore, that this title is inapplicable, though in 4 of the 24 cases (99, 107, 111, 116) the patients were the oldest children, and others may consequently have developed the symptoms of the disease later; while in Cases 56 and 122 the patient was the only child, and in 30, 117, 118, and 127 the existence of other children is not mentioned. Smith proposes the name "Postero-lateral spinal sclerosis (generic origin)," or short, "Generic ataxia." The full title describes the pathological changes, but does not distinguish it from Ataxia paraplegia of Gowers—the Combined postero-lateral sclerosis which is so ably discussed by Westphal; while the term "generic" is open to the same objection which applies to "hereditary" and "family"; viz., that it is not always applicable, and describes as characteristic and even indispensable that which is by no means such. A still better title is, I

think, "Friedreich's Ataxia," which designates the most prominent characteristic, gives the disease a specific name, and does not impose any limitations to be violated. Though not preferring the use of a writer's name for this purpose, I cannot but think that in the present state of our knowledge this is the best title that can be chosen.

Etiology.

Heredity.—Friedreich's ataxia is essentially one of the hereditary diseases, but the adjective must be understood here in its broadest sense.

A. *Direct and similar inheritance—i. e.*, inheritance of the disease itself or of some form of ataxia—has occurred in only 33 cases in 16 families of brothers and sisters; some of which, however, are very doubtful examples of it. Some of the cases of Ormerod (66, 70), those of Musso, and the Blattner family of Rütimeyer had respectively the grandfather, great-uncle, and great-great-great-grandfather reputedly ataxic. Descended from the last mentioned, who went by the name of "Stulzi" (the stumbler), were 8 instances of Friedreich's ataxia (2 in Table II. A) in 4 families. The grandfather of 2 of Bury's patients (112, 113) had probably an ataxic gait. In Bradbury's, Brousse's, and Carre's cases the mothers are spoken of as ataxic, and in the last there were in addition 6 brothers and sisters of the patient, 8 of the mother, the grandmother, and a cousin, who were said to have the same disease. The father of Fowler's cases was reported to have a peculiar gait; 2 uncles of Mastin's patients were almost certainly subjects of Friedreich's ataxia; and the father as well as the brother were without doubt affected by the same symptoms as

Botkin's case exhibited. But in no instance were the reported ataxic parents actually under medical observation except in the case of Smith, and in 2 of those of Vizioli; and here a direct inheritance of Friedreich's ataxia appears to have been witnessed. The father (100) of Smith's patients (101, 104) became ataxic after the age of sixty-six, while the 2 cases of Vizioli referred to are especially interesting. Vincenzo Vitielli (81), namely, was the father of Antonio and Luigi (89, 90), and the table also contains a cousin (91) and 7 uncles and aunts of these boys who were subjects of Friedreich's ataxia. A sister of 91 is to be found in Table II. A. It is interesting to note that there are sometimes instances of crossed heredity—*e. g.*, among Musso's patients there were 3 females inheriting the disease through the male line; and 3 males (2 in Table I. A) through the female line; while in Mastin's cases, again, 3 males inherited it through the female line.

B. *Polymorphic inheritance*—as seen in the development of a nervous disease in the children of parents affected by some other neurosis—is of frequent occurrence, and even the simple constitutional degeneration of the antecedents from various causes may account for the production of Friedreich's ataxia or other disorders in the descendants. Under this broad definition of heredity in nervous affections as emphasized by Möbius there exist as possible additional ancestral predisposing causes of Friedreich's ataxia: 1. Other Neuropathies, or exceedingly nervous disposition in the parents, grandparents, uncles, or aunts. 2. Alcoholism. 3. Tuberculosis. 4. Syphilis. 5. Consanguinity. 6. Other unclassified causes.

1. *Neuropathies* of various kinds, including great

nervousness, have been reported present in the parents or the relatives mentioned in 58 cases. In the remainder there were either none discovered or no reference is made to the matter. In 25 of the 58 cases one or both of the parents (marked + in the table) alone suffered from nervous symptoms; in 23 (marked ±), grandparents, uncles or aunts were also nervously affected; and in 10 (marked —) only the latter (*i. e.*, excluding parents) were neuropathic.

2. *Intemperance* of the ancestors[1] is a feature to which considerable importance was attached by Friedreich, who found it present in 6 of his 9 cases. In the Süss family, indeed, the mother maintained that her children were conceived while the father was intoxicated. I find intemperance of the father—and in Mastin's cases, of the mother also—in 31 instances (marked + in the table) occurring in 10 families; while in 13 other cases (marked —) the parents were sober, but the grandparents or other parental relatives had been intemperate. In some of the remaining 98 cases it is stated that there was no intemperance in the family, but in most of them the subject is not referred to, and it is fair to presume that the habit was absent in a large number. In only 7 instances (1, 2, 20, 21, 74, 75, 134) could it be said to be the only hereditary predisposing cause detected, and intemperance is unfortunately so common that it may have been here a pure coincidence. Though it seems to be not without some influence, I think but comparatively little importance can be attached to it.

3. *Tuberculosis* of the parents only was noted in 9 cases (marked +). In Palma's case the mother was

[1] The term "ancestors" in this paper denotes parents, grandparents, uncles, and aunts.

consumptive and sprang from a very consumptive family, and in one of Bury's (114) both father and uncle died of the disease. In 10 instances (marked —), including the 3 cases of Freyer where tuberculosis had probably existed, grandparents, uncles, or aunts had died of the affection. In 21 cases, therefore, all told, tuberculosis was present in the ancestors, though in but 11 was it the only cause known. It is probable that its presence in the forefathers, by producing debility of the offspring and a liability to degeneration of all sorts, may predispose to the development of Friedreich's ataxia.

4. *Syphilis* is said to have been present in the grandfather of Vincenzo Luongo (91), and some of Freyer's patients had exhibited an eruption which was possibly syphilitic. Except in these instances there was no evidence of a deterioration of the nervous system through inherited syphilis.

5. *Consanguinity* of the parents (grandparents in Mastin's cases) is reported as existing in but 7 cases in 4 families, viz., those of Seeligmüller, Erlenmeyer, Mastin, and Mendel. In the last 3 it would appear to have been the principal predisposing influence, but in the first family there also existed a very neurotic history.

6. We read of no *other possible ancestral causes*, except that the mother of Freyer's cases is said to have been feeble, and that some of the uncles and aunts of Musso's patients suffered from general debility and other signs of poor constitution.

As regards *conditions discovered in the other children* of the same family which rendered probable the existence of a common inherited taint or tendency to degenerative processes, we find that in 20 families of brothers and sisters there are said to have been 31 individuals presenting

the symptoms of Friedreich's ataxia, but who did not come under professional observation; this does not include the relations of the cases reported by Fellows, where the number is not stated precisely. Other neuroses, including exceedingly nervous temperament, are reported present to a greater or less degree in 14 families —32 cases. In 13 families—27 cases—there existed other suspicious conditions in the brothers and sisters indicating an inherited feebleness of constitution. Thus in 5 families (13–15, 19, 56–58, 96–97, 132–133) there had been numerous early deaths, and in Musso's 2 families 7 stillbirths were observed. In 2 of Ormerod's families (69–75) one or more children had feeble reflexes, and in Freyer's there had possibly been syphilis in several children beside the patients themselves.

The brother of Smith's oldest case (100) lacked all moral sense, and the younger brother of Botkin's patient is described as having moral insanity. The sister of 2 of Seguin's cases (96, 97) died of phthisis. In Palma's report a curious occurrence is described. The phthisical mother suckled but 2 of her children, of whom one was the patient, Giuseppi; the other being a sister who died of phthisis. The others were nursed by another woman and remained healthy, and Palma believes that the milk from the phthisical mother was a factor in the development of the ataxic disease.

It must be noted in this connection that there are certain cases where no trace of any inherited or of inheritable taint whatever has been discovered, or at least reported either in the ancestors or other descendants. An inspection of the table will show this to be true in 11 instances (24, 44, 55, 76, 99, 107, 109, 116, 117, 122, 140), though the patient of Erlicki and Rybalkin was

a foundling, and nothing was known of her relatives; while the family history of McAlister's case is not published.

As regards heredity, then, statistics show that the existence of ataxia or of some other nervous disorder in the ancestors exerts a strongly predisposing influence on the development of Friedreich's ataxia in the descendants; that intemperance has a much less powerful and even questionable effect, and that the agency of other conditions is insignificant, except as they debilitate the parents' constitution.

AGE.—The influence of age in determining the onset of Friedreich's ataxia has been variously estimated. Friedreich made the claim, based upon his own cases, that the disease developed at about the time of puberty; but this is clearly too narrow a limit. Numerous instances have been reported in which it appeared much earlier than this, and a few are recorded in which it was not detected until after the age of 20. Looking through the table it will be seen that the first symptoms were observed as follows:

In 15 cases	by	2	years of age.

and, including these,

In 39 cases	before	6	" "

Further,

In 45 cases	at from	6 to 10	" "
" 20 "	"	11 " 15	" "
" 18 "	"	16 " 20	" "
" 5 "	"	20 " 25	" "

The age of 14 of the remaining 16 cases is not accurately enough stated to allow of placing it in any of the periods given above; but in all it was less than 16 years, and in all but 2 (4, 112) less than 12 years, while in

4 (76, 81, 82, 95) it was probably less than 5 years. The 2 remaining cases are those of Seeligmüller (23) and Smith (100). In the former, difficulty in walking was not observed by the patient until after the age of 27, though it is uncertain just when ataxia actually developed; and in the latter no symptoms were perceived until the age of 66. (See remarks on cases in Table I.) The statistics show, then, that in *over one-half* the recorded cases the first symptoms were perceived before the age of 11 years, and that in only a small number did they develop after 16.

Sex.—The statements as to the sex chiefly affected vary somewhat, depending simply on the number of cases of the disease which the various tabulators had collected. Of the 143 cases reported here 86 were males (60.14 per cent.), and 57 females (39.86 per cent.)

Acute Diseases.—It is a most interesting fact that the occurrence of certain acute, and usually infectious, diseases has sometimes precipitated the appearance of the ataxia. Most probably the necessary conditions for its development were already present and were only incited to activity. This precipitation is described in 20 instances as follows: Typhoid fever (16, 52); pertussis (47); variola (64); meningitis (56); acute spinal affection of a rather uncertain nature (116); periostitis (119); morbilli (142); scarlatina (49, 90, 105, 143); scarlatina and morbilli (114); rheumatism (100, 132); probably diphtheria (69, 70, 71); diseases not clearly defined (117, 137). As a rule, the febrile disease precedes the development of the ataxic symptoms either immediately or with only a short interval. In 3 of Musso's cases, however (64 and 2 in Table II. A.), 2 years had elapsed since the patients had suffered from variola, which the

author nevertheless considers the exciting cause; while in Erlenmeyer's, 1 of Dreschfeld's (16), and 1 of R timeyer's (47) cases the acute disease had occurred respectively 5 months, 2 years, and 1 year previously. A degree of doubt cannot but attach to the existence of any causal relation in some of these cases. Bury believes that chorea may precipitate Friedreich's ataxia, but it seems more probable that the choreiform movements witnessed in one of Rütimeyer's cases (in Table II. A.) and in certain other instances were but anomalous early symptoms of the ataxic disease.

Clinical History.

The disease usually begins early in life, and nearly always attacks the lower extremities first; the initial symptoms being unsteadiness and weakness, with an oscillating, staggering gait accompanied by frequent falls. Exceptionally other symptoms, perhaps accidental, appear before the affection of the legs. *Eclampsia* occurred repeatedly in a patient of Vizioli's (91); and in one of Rütimeyer's (53) it was seen at 6 weeks of age and left the patient feeble, so that he never learned to walk as well as the other children. Friedreich describes the early occurrence of *hysterical attacks* in one case, but considers them independent of the ataxia. In Botkin's case the symptoms were ushered in by a fit of uncontrollable laughing coming on in church,—possibly of an hysterical nature. *Pain* in the head or other parts has been noted in a few cases before, or with the earliest ataxic symptoms, as have been *vomiting*, *vertigo*, *curvature of the spine*, *dorsal flexion of the toes*, *palpitation of the heart*,

choreiform movements, etc. They will be discussed in the proper place.

All these early symptoms, however, are of very unusual occurrence, if indeed they are other than purely accidental; and disregarding them, it appears that the lower extremities were first attacked in 114 of the 143 cases. In 10 instances the involvement of the legs and arms was noted almost simultaneously, though in one of these (99), Sinkler's, the mother seemed to think that the awkwardness was first in the arms. In 8 instances the speech and the four extremities were affected together; in Descroizilles' case, and very probably in one of Vizioli's (91) also, the legs and the speech were simultaneously involved, no reference being made to the time when the arms became ataxic; and finally in 2 instances (106, 134) the upper extremities were first diseased. The part first attacked in the remaining 8 cases is not definitely stated.

The speed with which the disease spreads varies very greatly. The average lapse of time in 35 cases between the first affection of the upper and lower extremities, where the date was accurately enough stated for calculation, and not including the cases of simultaneous involvement, was about 6 years; but the variation is so great that this average is of little value, and a better conception can be obtained by consulting the table itself.

In some of the patients the disease had not reached the upper extremities when reported, and in others it is not clearly stated whether or not they were affected. The longest interval mentioned, before the involvement of the arms was noticed, equals 20 years, and occurred in one of Dreschfeld's cases (16). Next in order comes one of my own (143) with an interval of 17 years.

These are, however, unusual, and the ataxia perhaps most frequently reaches the arms after the lapse of 1 to 3-5 years.

In Case 6 one leg was first attacked and then the arm of the same side, next the remaining leg and finally the other arm; or one leg may be involved and then that of the opposite side, as in Cases 2, 3, 35, 126. In certain instances one side of the body always remained more diseased than the other; as in 1, 6, 107, 122, and in Teissier's case (59) the upper limb was more incoordinate than the lower.

Ataxic movements may develop even in the head and trunk; either with those in the upper extremities or later. Bulbar symptoms, usually represented by nystagmus and affection of speech, are very constantly present. It is generally stated that they appear late in the disease, coming considerably after the affection of the arms; yet in 31 cases in which the date of the development of both classes is definitely given, the appearance of the bulbar symptoms (usually the affection of speech) averaged only about $1\frac{1}{2}$ years later than that of the incoördination of the arms; and the range of variation was not at all great. In 18 of these cases both developed at the same time, and in 6 the interval was but 1 year or less, while the longest intermission was but 9 years, viz., in one of Warren's cases (39). Yet these statements do not strictly express the truth; for doubtless bulbar symptoms would have developed later in many cases in which none were reported, and the average lapse of time would probably be increased.

As the disease advances, more or less paralysis with some muscular atrophy often makes its appearance, contractures of the muscles may develop, and talipes and

curvature of the spine commonly occur. Pain sometimes is present and exceptionally is severe. The patient becomes unable to walk, and loses more and more control over the bodily movements, until he is rendered almost completely helpless, while speech may be nearly or quite unintelligible. Finally death ensues from progressing asthenia; or oftener from some intercurrent disease.

Infectious or other febrile diseases have power to accelerate as well as to precipitate the ataxic disease. This aggravation has been either general, or limited to certain regions of the body, and is reported 14 times in 13 patients; one of Sinkler's cases (98) having once been made worse by scarlatina, and again by malaria. The acute diseases which have accelerated the progress of the nervous disorder are: rheumatism (13, 48); scarlatina (69, 89, 98, 133); malaria (98); typhoid fever (76, 83, 85); typhoid fever followed by measles (141); probably diphtheria (74); bilious fever (40); a tetanoid attack (128); some affection not clearly defined (102). The increase of the ataxia after recovery from the acute disease was so great in 5 cases (76, 83, 85, 98, 141) that the patients were found to have lost the power of walking entirely or except when assisted. Not included in the 13 cases is the second patient of Friedreich, in whom parturition at the age of 23 so augmented the severity of the symptoms that she was never again able to walk without assistance or unless holding to objects. A patient of Musso's (61) is also to be mentioned, in whom epileptiform seizures appeared to precipitate the involvement of speech.

Symptomatology.

Motor Symptoms.

Motor Ataxia.—*Motor ataxia of the lower extremities*, shown either in the gait or in intended movements, or in both, is reported present, or as having been present before loss of power supervened, in 128 cases. In the remaining 15 ataxia of the legs is not directly mentioned, though it is certainly implied in some of them. Freyer distinctly states that the gait was good in one of his cases (121), but the disease was only beginning. Hammond's effort in his article was to prove that the cases reported by Friedreich were not at all like locomotor ataxia. While, therefore, he will not attribute the motor affection in his own 6 patients to ataxia, he nevertheless admits that the feet "were moved exactly like those of a drunken man when he attempts to walk;" and in one case (36) distinctly speaks of lack of coördination, while in another (35) he implies that closing the eyes made an ataxia of station worse.

34 of the 128 cases exhibited increase of incoördination on closing the eyes, or in some instances had done this before the power of motion in the legs was lost; in 19 instances there was no such increase; in 1 (6) closure of the eyes had at one time produced it, but later did not, though the legs could still be moved. In the remaining cases no note is to be found in the original report.

The *gait* may be that characteristic of tabes dorsalis—*i. e.*, with the extreme anterior propulsion and elevation of the foot, and heavy fall of the heel; but I have found this stated in very few cases, as in 18, 25, 60, 80. Certain others—*e. g.*, 17, 66, 98—are described as of

"typically ataxic" gait; which may very probably indicate a gait like that seen in tabes. On the other hand, in over 45 instances I have noted it designated as "like that of a drunken man," or "oscillating," or as "not typically ataxic," etc. Musso described it as exhibiting a weakness, tremor, and a tendency to a *lateral projection of the feet;* and even as early as 1871 Bradbury said his case was not like tabes on account of the way in which the legs were thrown out at random. One of Musso's patients (61) said of herself that when walking she "occupied the whole road." In many other cases I have made no exact note of the character of the gait, or it is not mentioned by the authors; but there would seem to be no question that the oscillating, drunken, cerebellar gait is by far more characteristic of Friedreich's ataxia than is that usually seen in tabes.

The eyes are sometimes kept fixed on the floor while walking, as in both patients of Massalongo's, and in that of Carre's; sometimes the gaze is directed straight outward, as in Mendel's and some of Vizioli's cases.

Ataxic station is expressly mentioned or implied in 73 cases, and undoubtedly existed in many more. In 7 of the 73 it was certainly or probably absent when the eyes were open, but developed when they were closed. Of the 66 remaining it was increased by closing the eyes in 42 instances—*i. e.*, Romberg's symptom existed—and in 10 there was no such increase. In 13 it is not distinctly stated whether Romberg's symptom existed or not, and in 1 (6) it had been witnessed earlier in the case but afterward disappeared.

Ataxia on motion of the upper extremities is usually present and well marked. The movements are irregular and often jerky. Buttoning the clothes or threading a

needle becomes difficult or impossible; and even less delicate actions, such as touching the tip of the nose with the finger or carrying food to the mouth, are equally badly performed. In the attempt to make any voluntary motion the action is often overdone; thus in trying to grasp an object the hand may be advanced past it, as in 97. Prehension is often very peculiar, the fingers being spread over an object like a claw; as was well illustrated by one of my own cases (141). Bury also speaks of the claw-shaped grasp in Case 115, and Seguin describes the over-extension and spread of the fingers before grasping in 93 and 95. I have already referred to the peculiar infantile grasp in 2 of my own cases (141, 143). I find incoördinate movements of the hands or arms reported in 111 cases; increased by closing the eyes in 21 of these; not increased in 26; while in the remaining 64 the question of increase is not referred to by the authors. Many of the 32 cases in which ataxia of the upper extremities is not mentioned in the table undoubtedly possessed it, but the reporter has failed to state the fact clearly. In only 11 instances (17, 18, 23, 24, 28, 43, 58, 63, 75, 100, 108) is it distinctly said that there was none. Irregularities in distribution are sometimes witnessed; thus one of Ormerod's cases (71) exhibited ataxic movements in only one of the hands, viz., the left.

Muscle-sense.—Though coming somewhat out of its natural order, muscle-sense can best be considered here. Whether Romberg's symptom is due to a disturbance of muscle-sense, or to some entirely different cause, forms a most interesting question, but one too extended to be fully discussed in this paper. Friedreich claimed that his cases proved both locomotor and static ataxia

to be independent of any disturbance of sensibility. The case of Galassi's rendered that writer sceptical as to any relation between Romberg's phenomenon and muscle-sense. Musso, too, found Romberg's symptom in 4 of his cases, while in none of them was the muscle-sense otherwise affected, though carefully tested in various ways.

The muscle-sense is described in the table as normal in 32 instances. In some of these the authors merely state that it was unaffected and probably include the absence of Romberg's symptom, while in others the manner in which it was tested is detailed. It happens, now, that 15 of these 32 cases exhibited increased incoördination of the motion of the legs or of station on closing the eyes, yet the muscle-sense, strictly named, was undisturbed, as shown by the testing with weights, grasping with eyes closed after an object whose position was known, and in the knowledge of the position of the members in space. This would seem to confirm the view that Romberg's phenomenon depends on something more or other than the loss of this sense. An additional proof is found in the fact that in 11 cases the muscle-sense was more or less diminished, in 4 of which there was no Romberg's symptom, and in 3 others no mention is made of it, if it existed.

Static Ataxia.—By this title Friedreich designates the *ataxia of quiet action—i. e.*, of the muscular force required to keep any part of the body still while unsupported, as when holding the arm extended. In contradistinction to this, the term "locomotor ataxia," or, better, "motor ataxia," would indicate the incoördination which appears only when a voluntary movement is attempted. The ataxia of station is, of course, a variety

of static ataxia, and has already been discussed, but nothing has yet been said of its other forms. Friedreich states that static ataxia is almost the last to appear, and Charcot and Ormerod express the same opinion, while the former also claims that it is characteristic of the disease and does not appear in tabes. Though Pitt says that the history of the reported cases indicates that it is an earlier symptom than the locomotor ataxia, my own search through the literature would confirm quite decidedly the views of the writers first mentioned. I find it described in the extremities in 2, 6, 13, 15, 22, 46, 49, 68, 113, 114, 115, 117, 124, 127, 137, 138, 139, 141, and many others, usually of the severe forms. The exact number I have failed to record, though it is, I think, oftener absent or not described than present.

In the hands while lying passive in the lap, it may be seen as peculiar slow movements of the fingers, which sometimes resemble athetosis, as in Charcot's second case (127), one of Sinkler's (139), and one of my own patients (141). In the arms it appears as slow wavy oscillations when the member is held outstretched, or even as marked choreiform movements, as Erlenmeyer describes in his patient, which he, however, classifies as evidences of static ataxia, as does Charcot the constant non-rhythmic oscillations of the legs, seen in his first case (67). Botkin speaks of a similar condition in his own patient, and in one of Seeligmüller's cases also (22) there were observed the same constant trembling movements when the legs were in certain positions. Numerous other instances of static ataxia of the limbs are reported, but it is seen to best advantage in the oscillating movements of the head, which are described in 51 instances; and undoubtedly most of the cases exhibiting "tremor"

or "choreiform movements" of the head should be added to these. The oscillations may be slight or extensive, constant or seen only under excitement. Friedreich described in Case 6 an irregular nodding motion of the head, "like one going to sleep," and this has since been often witnessed, as in Cases 45, 127, and others.

Ataxia of the trunk is reported in 22 instances. In others its occurrence is implied, though with but one exception no reference is made to it.

Tremor.—Tremor and choreic movements may naturally be next described, since they are so closely allied to static ataxia. Tremor of some part of the body is referred to in 8 cases. In most of these it should probably be attributed to static ataxia—*e. g.*, 2 of Smith's cases (102, 103) had tremor of the head on voluntary motion, and one of Teissier's (59) exhibited little oscillations of the whole body when at rest. There is nothing found which at all recalls the intention-tremor of multiple sclerosis, except in Glynn's case, where there existed oscillations of the head and trunk, a forward and backward oscillation of the tongue, with a shaking of the arms which became violent when reaching for a cup. In Cases 22 and 133 the muscles of the face exhibited slight tremor.

Choreiform Movements.—Choreiform movements are referred to in 17 instances, and are, like tremor, probably in most cases evidences of static ataxia; being, in fact, recognized as such by some of the authors reporting them. They are usually seen in the extremities, but in 6 cases grimaces and twitchings of the face are alluded to (23, 56, 69, 71, 97, 127). Chorea has appeared as one of the earliest or even the initial symptom in certain cases, as, for example, in 2 cases of

Ormerod (69, 71) and in that of Gower's and Pitt's (27). Pitt states that 3 other cases in the family began with choreiform movements, which were, however, not referred to when Gowers reported them; and I do not think that statistics corroborate Pitt's view "that the first symptoms noticed are disorderly choreiform movements." Erlenmeyer's case is especially interesting in this connection, since Erb pronounced it to be an instance of chorea, while Friedreich deemed it some spinal affection, though not ready to admit it into the same class with his own cases. Erlenmeyer, however, insisted that it was an example of Friedreich's ataxia, and it has since been unquestionably accepted by Rutimeyer. There seems no reason to consider it other than a genuine case.

Spasm.—Involuntary spasmodic contraction of the muscles has been reported in 21 instances. These are sometimes painful, perhaps oftener not, and occur chiefly in the lower limbs when the patient is lying in bed or sitting. 4 of Friedreich's cases exhibited this symptom, apparently with cramp-like pains. It was seen also in a case each of Hammond's (33), Rütimeyer's (47), and Smith's (102); the legs of the last patient being at times drawn up to such an extent that she begged to have weights put on them to pull them down. In 2 of my own cases (142, 143) these spasms occurred at night, and the patients were unable unassisted to straighten the limbs immediately.

Fagge's patient suffered from a spasmodic action of the muscles of the face when an effort was made to talk, drawing the mouth into a meaningless smile; and in Botkin's patient there occurred spasm of the muscles

of the neck, hands, and feet, with constant grinding of the teeth, the case being unique in this last particular.

Paralysis.—More or less paralysis is a very common feature in advanced cases, and it may, indeed, appear early in the disease; but as Ormerod justly remarks, the initial weakness in the legs, and even the loss of the power of walking called by the friends "paralysis" are probably due simply to the incoördination in most instances. Many of the cases which have been subjected to careful medical examination in the early stages support this view. (See "Inability to Walk.") Muscular paralysis is reported present in 56 cases, absent in 53, and is not directly mentioned in 34, though in a number of these it almost certainly existed. Many of the cases in which paralysis is reported absent were not far advanced in the disease, and the percentage of cases affected would certainly be increased could the clinical histories be followed further. It may become extreme in advanced cases, as in one of Vizioli's (87), who had been affected in early infancy and who died at the age of 46, having reached a state of complete immobility apparently of all the muscles, excepting those of mastication and respiration. In Therese R. (123), diseased 28 years, the legs were totally paralyzed, and the arms and trunk largely involved; and Emma W., one of Smith's cases, was in a similarly helpless condition. The rapidity of the development of paralysis does not, however, always depend on the duration of the disease; since in Glynn's case, for example, which had lasted but 4 years, the muscular power of the limbs was decidedly diminished, and in Kahler's and Pick's patient, diseased 8 years, it was nearly gone; while in one of Freidreich's cases (6), on the other hand,

the patient died of typhoid fever with normal muscular power after suffering from ataxia for 24 years, and in one of Smith's (103) there was no paralysis after 18 years of the disease.

Inability to Walk.—In a number of cases standing or walking had become impossible as the disease advanced. This was due in some of them to the paralysis, but in others it clearly depended on the existence of excessive incoördination, since the actual strength did not seem to be at all diminished. Friedreich calls especial attention to this condition in Case 6. A review of the table shows that in 54 cases the patients were no longer able to walk, except with the aid of crutches or when assisted by some person. In 4 instances the authors do not make any statement concerning it, and in 86 the power of walking was still preserved, though often only when the patient steadied himself by the furniture or walls. The relation between the inability to walk and the paralysis may be seen by comparing these two columns in the table. In 10 of the 54 cases paralysis is stated to be absent; in 9 it is not mentioned, and in the remaining 35 there was more or less of it, though it is very certain that in many of them the term was wrongly applied, especially where the table shows that no atrophy of the muscles was to be found. In some of them paralysis is expressly stated to be very slight or even questionable, and was clearly not sufficient to account for the loss of the power of locomotion. The *age* at which this loss occurred is noted in the table whenever given in the original article, but the lapse of time from the onset of the disease to this event is so variable that to average it would be useless. In Cases 37, 38, 39, 87, and 88, the patients never walked; while

in 1, 32, 34, 44, 55, 139, and 141, walking unassisted was impossible within a year after the disease began, and in the last case within a month after the first onset. In Case 5 the patient was able to walk through 27 years of the ataxia, up to the time of death from an intercurrent disease, and the table shows other cases in which the ability persisted for years.

Contractures.—Contractures of various parts are referred to in a number of instances, and are usually among the later symptoms; and may be divided into Talipes, Curvature of the spine, and Other contractures.

Talipes is reported in 27 cases, in 14 of which it was of the variety equino-varus. The remaining 13 are named as follows: equinus (40, 41, 42, 58, 114, 115, 122); equino-valgus (141, 143); valgus (112); varus (134); cavus (126, 136). The last case, though not named directly "cavus" by the author, is best summed up by this word. Talipes is a late symptom, and, indeed, has only been seen in 7 cases which still preserved the power of walking. It is associated often (12 cases), though not always (9 cases), with some degree of muscular atrophy. In 2 of Smith's patients (101 and 102) the talipes was immovable; in one of Bury's (114) there was very little motion at the ankle-joint, and in one of Sinkler's (139) the tendo Achillis was very tense.

Curvature of the spine, considered by Rütimeyer a form of contracture, appears to be largely independent of the degree of paralysis. It is usually one of the later symptoms, though in Cases 4 and 10 there was curvature of the spine—perhaps accidental—2 years before other symptoms appeared, and in Cases 9, 71, and perhaps others it was among the early symptoms. It is reported present in 57 cases; being sometimes slight,

sometimes very marked; and is described in 28 of these as scoliosis, in 20 as cypho-scoliosis, in 8 as cyphosis; while in 1 the variety is not designated.

Contractures of other parts of the body are not infrequent as late symptoms, though in one of Bury's cases (113) they were present from the first. They are mentioned in 24 cases; all except 100, 101, 102, and 115 exhibiting dorsal flexion of the toes. There were also contractures of some of the fingers or of the hands in Cases 49, 50, 51, 101, 113, 115. In one of Smith's cases (101) the elbows were flexed; in another (102) the muscles of the thighs and calves were much contracted; in another (100) the calf-muscles were tense, and in one of Sinkler's (98) the plantar fascia was contracted.

Electrical Contractility.—The electrical contractility has been tested in only a few instances, and appears to reveal nothing characteristic. In 31 cases it is reported normal, sometimes the faradic current being specified, often the nature of the current not being named. In 2 instances (66 and 100) it was normal or increased. In 9 cases (2, 17, 19, 81, 89, 91, 103, 124, 136) it was slightly diminished; and in 2 (59, 60) the faradic contractility was increased. In 3 instances (113, 114, 115) the reaction of degeneration was detected; the faradic contractility being diminished in the first and normal in the second. One of Smith's cases (101) exhibited decreased faradic contractility but increased response to the galvanic current; while in Descroizilles' patient the faradic reactions were normal and the galvanic possibly diminished.

Examination of the electrical sensibility has been re-

ported in a few instances, but has taught nothing of importance.

Reflexes.—One of the most important studies connected with Friedreich's ataxia is that of the *patellar reflex*. In the 143 cases reported the knee-jerk was abolished in 91, while in 30 the condition is not stated. The absence would seem, then, to be a very constant symptom, as it is a very early one. In 2 of Ormerod's cases (72, 73), for example, it was observed before the diagnosis could be made. In Leubuscher's case the reflex was lost during the first half year; in Jakubowitsch's, which had only been diseased a month or so, it was very greatly diminished; and in 15, 21, 28, 56, 74, and others it disappeared within 1-3 years after the first appearance of the symptoms. The patellar reflex is nevertheless not always absent, though this claim has frequently been made. It is reported diminished in Fellows' patient and one of Warren's (43), and much diminished in 7 instances. But in 6 cases (63, 65, 85, 100, 137, 139) it is said to be normal; in Descroizilles' patient, normal or exaggerated; and in 6 cases (22, 23, 66, 68, 107, 138) exaggerated. Though all of these have a fair claim to be considered instances of Friedreich's ataxia, objection might be raised to some of them on the ground that in still other respects they were anomalous. (See remarks on Table I.) The patient of Prince's, however, though having exaggerated reflexes, appears to be an unquestionable instance of it, and Cases 63 and 85, with normal reflexes, are typical in other respects and belong respectively to the families reported by Musso and Vizioli, which contain some of the most striking cases known. Massalongo's 2 patients are beautiful examples of the disease; yet in one the knee-

jerk was normal, and in the other exaggerated; and I have personally examined the later patients of Sinkler—the MacDonald children—and cannot but agree with others who have seen them, that they are undoubted, well-advanced cases of Friedreich's ataxia, yet without the absence of knee-jerk.

As Westphal states, the knee-jerk will not be abolished unless the lumbar enlargement be involved; and it is easily conceivable that this involvement might fail to occur in undoubted cases of Friedreich's ataxia.

We cannot, therefore, but conclude that though abolition of the patellar reflex is a very important diagnostic sign of the disease, yet genuine cases may occur in which the reflex is slight, normal, or even exaggerated.

Very exceptionally *ankle clonus* has been observed. I do not recollect reading of it in any cases but those of Descroizilles' and Fellows'.

As regards the *cutaneous reflexes* the plantar reflex was normal in 40 instances, absent in 11, diminished in 4, and increased in 7. The other cutaneous reflexes were normal in 26 cases in which they were tested to some extent, diminished, at least in part, in 7, absent in 1, and increased in 1.

Trophic and Vaso-motor Symptoms.

These two sets of disturbances are so nearly allied that I have classified them together.

Atrophy of the Muscles is comparatively unusual even where paralysis is well marked. In 60 cases there was no muscular atrophy, and undoubtedly it was absent in many, if not most, of the instances in which no reference is made to it. The lower extremities are the parts usually attacked, though the arms are sometimes in-

volved. In 11 cases atrophy is said to be slight, and in 24 marked. This does not include my own case (142), in which the great atrophy of the left leg and thigh was evidently independent of the ataxic disease. Some of the patients exhibited extreme atrophy; thus Case 50 had both legs and arms much wasted, as had Cases 19, 40, and 64.

Other Trophic Changes—which are tabulated in the same column with vaso-motor affections—are very unusual. Bedsores are said never to develop in Friedreich's ataxia. One, however, was observed in Case 2, but was superficial and soon healed. The skin of the legs was in parts firmer, thickened, and adherent to the subjacent tissues in one of Sinkler's earlier cases (99); Vizioli reports an extreme development of adipose in the atrophied legs of his patient Vincenzo (81), and Smith reports enlargement of the feet of one of his cases (101), while in a patient of my own (143) they seemed smaller than normal. Two other patients, of Smith's and Vizioli's, respectively (102 and 85), and one of Seguin's (93), suffered from chilblains, which easily suppurated in the last case. These are so equally a vaso-motor affection that other disorders of this class may be expected to occur.

Vaso-motor Affections are represented in 19 cases by coldness of the surface of the body or especially of the feet, with more or less blueness and sometimes œdema (19, 40, 41, 42, 46, 47, 48, 51, 54, 57, 69, 74, 75, 93, 95, 101, 102, 142, 143). In 2 instances flushings are reported; of the head and face (63), and flashes of heat (2). [When both vaso-motor and trophic affections, not including muscular atrophy, are present, the sign in the table is +.]

Sensory Symptoms.

PAIN.—The absence of the severe initial lancinating pain of tabes is one of the most characteristic symptoms of Friedreich's ataxia; and their occurrence in Dreschfeld's and a few other cases contributed to throw doubt upon the diagnosis. Pain of some sort, before the other symptoms appeared, or early in the course of the disease, was present to some degree in only 22 cases, absent in 79, and probably wanting in most of the remaining. Botkin's and one of Musso's cases (61), as well as 2 of Friedreich's (13, 14), suffered from headache before other symptoms appeared, and Bradbury's case began with pain and weakness in the back. The patient of Carre's suffered from severe pain in the legs and feet; and that of Jakubowitsch's from pain in the knees, shoulders, and head at the very beginning of the ataxic affection, while Friedreich's Cases 3, 4, and 5 complained of considerable early pain of the lower extremities. With these exceptions pain at the beginning of the disease has been insignificant, and in many cases was probably accidental. Pain after the disease has become well established is comparatively more common. I find it recorded in 47 cases, in many of which it was certainly unconnected with the disease itself. In about half of this number it was well marked, and in the others slight; while in 63 cases it is stated to be absent and in the balance was presumably so. Among the cases in which it was severe may be noted 2 patients of Musso's (61 and 62), who suffered from intense headache at certain periods, accompanied or followed by a temporary increase of the ataxic symptoms. Severe lightning pains are rare, and are perhaps best illustrated by one of Fowler's patients (77).

Lissette Süss (5) suffered from gnawing and neuralgic pains and painful spasms in the limbs; and several other of Friedreich's cases exhibited similar sensory disturbance. Some of Seguin's (96, 97) and of Smith's (101, 102, 103) patients had marked pain in the lower extremities, though in 102 these seemed rather to be due to the intercurrent and fatal acute myelitis. Maria Vitielli (85) suffered from headache and pain in the articulations. Tenderness on pressure over the spine has been reported in Cases 63, 64, 85, and a few others.

Cutaneous Sensibility.—The cutaneous sensibility is generally not much involved. In 74 instances it was normal, in 16 no reference is made to the subject, and in 47 there was more or less diminution of tactile sensibility. In 2 cases (7 and 25) it was normal or increased, and in 3 cases (40, 60, 136) increased. In Descroizilles' patient it was at one time slightly diminished, but later normal.

The diminution of sensibility—usually tactile—has generally been slight and often questionable. Sometimes it could be detected by the æsthesiometer; sometimes the touch of the finger could not well be felt. Often the sensibility to pain was blunted also, and in certain cases the conduction of sensation was retarded. Among the most marked instances of anæsthesia may be noted the second case of Friedreich's, the first of Musso's, 1 of Dreschfeld's (16), and 2 of Smith's (102, (103); the last 2 patients being scarcely able to distinguish pinching and pricking of the skin from simple touching. In one of Freyer's cases (119) there was total anæsthesia in spots; but the most striking example is Therese R. (123), in whom the lower limbs had become totally anæsthetic after 7 years' illness from Friedreich's

ataxia. Evidences of a degree of anæsthesia are further seen in Cases 55, 103, and 100, in which there was respectively a sensation of cushions, of velvet, and of a sponge under the feet.

Muscle-sense has been already discussed in connection with Romberg's symptom.

Paræsthesia.—Some form of paræsthesia, including girdle sensation, is reported in only 18 cases. In 43 instances (0 in table) there was no mention of any form of paræsthesia, and it is stated that either girdle sensation, or paræsthesia of other kinds, or both classes were absent. It is fair to presume that the majority of the remaining cases were free from any such affection. In 8 instances a girdle sensation is reported, namely, in 22, 103, 132, 141, 100, 102, 114, 119. In the first 4 it was very slight, but in the others well marked; Smith's patient (102) having it perhaps most strongly developed. Seeligmüller's second patient felt as though the clothes everywhere were too tight. Paræsthesias of other kinds, such as formication, tingling, numbness, etc., were observed in 14 instances (6, 7, 9, 17, 22, 45, 46, 54, 69, 100, 102, 107, 119, 135). In few were they more than very slight, and in none severe.

Bulbar and Cerebral Symptoms.

Affection of Speech.—Affection of speech is one of the most prominent of the bulbar symptoms, as it is one of the most frequent characteristics of the disease, and one which would probably develop in all cases not cut short by some intercurrent affection. It is generally a later appearance, occurring as a rule about the same time or somewhat before nystagmus. As already shown

under "Clinical History," it does not always develop so long after the affection of the arms as has often been supposed, and may be affected from the first. It was present in 107 cases, absent in 21, and not referred to in the remainder. Speech may be scarcely perceptibly involved, or so greatly so that it is almost or quite unintelligible, as in one of Warren's (40) and one of Smith's (101) cases. It may be drawling and slow, as in one of Bury's (115) and in Joffroy's patients, or jerking and interrupted, or confluent, as in Jakubowitsch's case, or typically scanning. In Rudolf Blattner (49) there were frequent changes of pitch; in Hollis's case, speech was low and indistinct. Glynn's patient slurred his syllables, pronouncing "actually" as though written "axially." In Power's case it was sometimes slow, sometimes rapid and slurring, etc. Friedreich spoke of "ataxia of speech;" and the term applies especially well to the rapid pronunciation of words and syllables followed by a pause, and this again by the same jerking articulation as I have described it in one of my own cases (141). He considers it due to an incoördination of the articulating movements of the tongue. The peculiar rushing, tumbling speech which Seeligmüller observed in one of his cases (23) induced him to characterize the trouble in that instance as an "ataxia of thought;" since the patient seemed to begin a new sentence before he had finished the old one.

I have attempted to express by signs in the table the degree of the disturbance of speech in the different cases, but the character of this disturbance is too varied to be well shown. The scanning articulation is quite common, though it is not, I think, mentioned in the majority

of cases. As instances of it may be noted Cases 26, 59, 60, 61, 62, 64, 102, 112, 114, 122, 127, 143.

FACE, MOUTH, AND THROAT.—The *tongue* is not infrequently affected; usually by a fibrillar tremor, sometimes by an irregular jerking or a to-and-fro movement. It is reported involved in 24 cases, and not affected in 30, including Case 2, where a previously existing tremor had disappeared. In Quincke's and Rütimeyer's first case (46) the tongue could not be held still in the mouth, but this very active movement was afterward substituted by a simple tremor. In Erlenmeyer's case it would not remain quiet; in one of Teissier's (59) it exhibited little oscillations, and in Glynn's these were larger, while the tongue was seen "curling and uncurling in a most curious way." In Ferrier's it was unsteady when projected; in the first of Friedreich's it trembled and twitched, and in that of Mendel moved backward and forward but was without fibrillar tremor. Nearly all the other cases exhibited fibrillar tremor only. At one time in Case 2 the tongue would become motionless for some moments and the patient unable to speak. There was also some paresis in Joffroy's patient.

Mastication and Deglutition are usually spared, but may become involved in advanced cases. Thus deglutition is described as almost impossible in 2 of Smith's cases (101, 103), and a similar difficulty was present to some degree in Cases 33, 45, 49, 50, 94, 143. Choking attacks of a peculiar nature seemingly not dependent on deglutition were observed in a case of Ormerod's (133).

There is sometimes *inability to retain the saliva* in the mouth; apparently distinct from or in addition to ptyalism. This was the case in one of Seeligmüller's

patients (22), one of Smith's (101), one of Sinkler's (139), and others. True salivation will be referred to later.

The *face* has often a very expressionless appearance; as, for example, in 11, 64, 65, and 136; and this condition, when combined with great difficulty in articulation, sometimes gives an erroneous impression of an affection of the intellect. Slight twitching or tremor occasionally is witnessed, as stated elsewhere. In one of Dreschfeld's cases (17) there is said to have been slight atrophy of some of the muscles, and in Joffroy's some paresis; while in Ferrier's the imperfect movement of the upper lip did not properly disclose the teeth.

Eye.—*Strabismus* is mentioned in a few cases—*e. g.*, in 8, 23, 56, 71, 80, 109, 115, 131; but it is uncertain in how many of these it was accidental. *Diplopia* is referred to in a few instances, as 7, 68, 136. *Blepharospasm* I remember to have seen described in but 3 cases, those of Botkin, Descroizilles, and Erlenmeyer, in the first 2 of which the spastic element elsewhere was very well marked. Some degree of *ptosis* was observed in Joffroy's patient and in one of Ormerod's (71).

But the most important feature connected with the organ of vision is *nystagmus*, which Friedreich regarded as a very characteristic symptom, though it is one of the last to appear. It was present in 56 cases, absent in 46, and not mentioned in the remainder, though it certainly would be found to have appeared in many of them could the subsequent histories of the cases be followed up. It is a much less common symptom than is affection of speech, as is exemplified by Friedreich's cases; the latter symptom being observed in all of them, and the former in but 5. It very frequently develops at

the same time with the affection of speech or a little afterward, but it may appear much later; not being detected in one of Friedreich's cases (2), for example, until 10 years, and in another (6) until 19 years after the disturbance of articulation was first noticed. So in one of Warren's patients (40), speech had been affected for 5 years and had become almost unintelligible, yet no nystagmus could be discovered. On the other hand, in Erlenmeyer's patient nystagmus appeared at the outset of the disease, lasted 3 weeks and then disappeared permanently. A survey of the table shows that there are but 4 cases (22, 48, 124, 125) stated to have had nystagmus without difficulty of speech, while in 2 others (92, 103) the latter is not mentioned. The form is nearly always what Friedreich called "ataxic nystagmus;" namely, a more or less extended, transverse, bilateral oscillation of the bulbs, appearing only when the eyes are fixed upon an object, especially if held at the side. The nearer the object is approached, the more marked does the symptom become, but when the eyes are at rest it disappears completely. It is of less rapid motion than the ordinary or static nystagmus, and, indeed, differs widely from it. In Leo von K. (22) it appeared to cease only when the eyes were directed at certain angles, which varied at different times. Botkin's case is said to have had involuntary turning of the eyeball upward, a motion which appears to be allied to nystagmus. "Static nystagmus" also—*i. e.*, that seen when the eyes were at rest—was observed in Cases 46, 51, 54. The *pupillary movements* are in some instances said to be sluggish, as 64, 69, 111, and others; or to be dilated, as in 45, 65, 66, or con-

tracted; but in no case were they found not to react to light and on accommodation.

Atrophy of the optic nerve, so common in tabes, occurs in Friedreich's ataxia with the greatest rarity, if at all. I have found ophthalmoscopic examinations recorded in 38 cases, and in none of them was there any change of importance noted except in Power's case, in which the disks were rather white; and in one of Seguin's (96), in which there was partial atrophy of both optic nerves. The condition of the *color fields* might prove of interest in showing the possible relation of the disease to locomotor ataxia. Very little has been done in this direction, though Oliver made a careful examination of one of Sinkler's cases (99) and found narrowed fields, leading him to believe that there existed ocular changes allied to those of tabes.

Vision is reported impaired in a number of instances in which no ophthalmoscopic examination had been made, as for example in 7, 11, 17, 35, 40, 51, 96, 97, 123. In Botkin's case there was total blindness of the left eye, but probably not dependent on the principal disease.

Hearing was deficient in a few cases, as in 50, 51, and 123, but this was probably purely accidental.

Intellect.—One of the features which distinguish Friedreich's ataxia so sharply from disseminated sclerosis is that it rarely has any effect on the mental power. In 79 instances the intellect is reported normal, while in 21 it was possibly more or less affected, though the study of these cases shows that the deterioration was generally only apparent or trifling. Thus it is stated that the intelligence was low, but not out of proportion to the social position (19); that the patient often forgot words (35); that the intellect seemed at first to be decidedly affected,

but was in reality very slightly, if at all so (40); that the patient said he thought slowly (98), etc. In only a few instances does the intellect seem to have been actually impaired. In one of Smith's cases (102), for example, the memory had become very deficient, and the simplest mental acts were the result of laborious effort; while a sister (101) would frequently scream for hours for no discoverable reason, and had also had epileptic seizures during fifteen years, though it is not certain whether they had any connection with the ataxic disorder. A case of Seguin's (94) became practically insane during the last two months of life; and Power's case had but weak intelligence, and often laughed spasmodically. In Brousse's case the intelligence was low, and one of Hammond's patients (33) appeared to have suffered an arrest of mental development to some extent. It seems open to question whether Friedreich's ataxia had any direct bearing upon the deficient cerebration of any of these cases; and even Seeligmüller's patients, sometimes excluded on the ground of cerebral disturbance, exhibited no greater affection of this nature than did their father, who was in other respects a healthy man.

Vertigo.—Vertigo is a not infrequent early or late symptom, being present in 29 cases and absent or not recorded in the remainder. 4 of Friedreich's cases had it to some degree, and the patient of Ferrier's was early so annoyed by it that he was unable to play with his companions, though the symptom later disappeared entirely. In Botkin's patient it preceded other symptoms by about two years, but had probably nothing to do with the ataxic disease; and in one of Friedreich's (3), one of Rütimeyer's (46), and one of Bury's (112) it was

among the early symptoms. Vertigo later in the disease was seen, for example, in Power's patient, who suffered from attacks of giddiness, with loss of consciousness and fainting; and in Catharine L. (105), where it was well marked, and brought on by reading or in other ways, as also in one of Smith's patients (100), where it was a constant symptom, greatly increased by standing. Other cases have experienced it even when in the recumbent position, as in Friedreich's second case, in which it was for a time the chief complaint. In certain other instances it was severe, though it usually is a symptom of no great importance, and is often probably accidental.

Visceral and Secretory Symptoms.

These are usually slight and possibly accidental; are reported present in 42 cases in all, and are best considered in classes, as follows: [In the table marked + if more than one class is present; if only one, the sign is —.]

Affection of the Bladder is rarely met with, being referred to in 13 cases; in 8 of which (8, 23, 55, 59, 96, 111, 114, 133) it is described as incontinence which was slight or occasional, or as occurring when laughing (55), etc. In Power's case, and in one of Warren's (40), incontinence seems to have been more marked, though never excessive. Sinkler's first patient (98) had some difficulty at the beginning of urination; one of Musso's (61) was unable to urinate while lying in bed; and Friedreich's second patient could only pass water in drops, and after long straining.

Affection of the Rectum is very rare, excepting that several cases had troublesome constipation—cer-

tainly no unusual feature in many conditions. In one of Seguin's patients (97) this is said to have been extreme. In the first case of Seeligmüller (22), on the other hand, the feces were sometimes expelled before the clothes could be removed; and the brother (23) was obliged to hasten as soon as the desire to defecate or urinate was felt. Descroizilles' patient suffered from constipation alternating with involuntary passages of urine and feces.

Disorders of the Sexual Apparatus have been claimed by some writers to be quite common, being represented in the males by impotence and in the females by affections of menstruation. This claim was probably based on the fact that 2 of Friedreich's cases were impotent, while some of the women had disordered menstrual functions. Later experience shows that *impotence* has only been reported in 3 cases, all told (1, 6, 17), though the perverted sexual feelings of Seeligmuller's patient (22) rendered him virtually impotent, and he might be properly classified with the others. On the other hand, the males of Vizioli's first family, though severe cases of Friedreich's ataxia, exhibited normal or unusually great sexual power. *Disturbances of menstruation* were considered by Friedreich to be a complication rather than a result of the spinal affection. They are reported in 10 instances (2, 4, 19, 30, 54, 61, 62, 69, 116, 117), consisting of such disorders as "scanty menstruation, beginning in the sixteenth year" (19); "menstruation at twenty, regular but scanty" (62); "has not menstruated at nineteen" (69), etc. One of Friedreich's cases (2), who had at times decided menstrual irregularity, conceived and bore a child while suffering from ataxia. Probably the most serious disturbance reported is that of Salome Süss (4), who did not menstruate until the age of twenty-

two, and then very irregularly and with intervals of several years. Among any 57 females suffering from any chronic debilitating disease, it is probable that 10 instances of menstrual disorders would be found equally severe with those reported here.

Other Visceral and Secretory Affections, some of which are allied to vaso-motor disturbances, are recorded as follows: Salivation in 5 cases (2, 50, 56, 59, 68); palpitation of the heart in 13 cases (2, 61, 62, 63, 5, 4, 19, 22, 23, 101, 102, 103, 104), in the last 8 of these being one of the earliest symptoms; persistent acceleration of the pulse in 8 cases (2, 19, 40, 59, 60, 119, 123, 124); profuse sweating in 3 cases (2, 50, 63); gastric disturbance in 9 cases (8, 57, 132, 133, 45, 101, 102, 103, 104), being one of the initial symptoms in the last 5; dyspnœa in 5 cases (2, 4, 5, 19, 102). Besides these there are mentioned polyuria (2), cough (4), intermittent albuminuria (13), nervous crises (61), and præcordial anxiety (62, 63).

Pathological Anatomy.

Among the 12 autopsies which have been made I have included that of Erlicki and Rybalkin, whose case was an excellent example of Friedreich's ataxia, except that nystagmus and the affection of speech had not yet developed. The authors admit that these symptoms would not appear until late, and make their case a text for the discussion of Friedreich's ataxia, yet hesitate to call it an example of it on account of the absence of these symptoms. Since the symptoms in other respects and the post-mortem lesions are those of the affection in question, there is every reason to include the case here.

The autopsies, arranged in the order in which they were reported, are as follows:

I. Andreas Lotsch (1); Friedreich; reported 1863. Died at 35 of typhoid fever. Ataxic 18 years.

II. Justinia Süss (3); Friedreich; reported 1863. Died at 31 of typhoid fever. Ataxic 16 years. Specimens re-examined 16 years later by Schultze.

III. Salome Süss (4); Friedreich; reported 1863. Died at 28 of typhoid fever. Ataxic 12–13 years.

IV. Friedreich Suss (6); Friedreich; reported 1877. Died at 38 of typhoid fever. Ataxic 23 years. Microscopical examination made by Schultze.

V. Josefa Strasik (19); Kahler and Pick; reported 1878. Died at 23 of phthisis. Ataxic 8 years.

VI. Charlotte Lotsch (2); Friedreich; reported 1880 by Schultze. Died at 51 of nephritis. Ataxic 34 years.

VII. Marie R. (30); Brousse; reported 1882. Died at 32 of cerebral hemorrhage. Ataxic 8 years.

VIII. Clara W. (102); Smith; reported 1885. Died at 29 of acute myelitis. Ataxic 19–20 years. Microscopical examination by Putnam.

IX. J. W. (117); Erlicki and Rybalkin; reported 1886. Died at 18 of phthisis and typhoid fever. Ataxic 2 years.

X. Robert Shaw (27); Gowers; reported 1886–7 by Pitt. Died at 28–29 of cardiac dilatation. Ataxic 9–10 years.

XI. Heinrich Kern (46); Quincke and Rütimeyer; reported 1887 by Rutimeyer. Died at 20 of fatty heart. Ataxic 13–15 years.

XII. Bertha Kern (48); Rütimeyer; reported 1887. Died at 16; cause of death not clear. Ataxic 9–10 years.

The following analysis will perhaps best indicate the lesions found:

Brain.—The brain (not including the medulla) was certainly examined in 10 cases and nothing of importance discovered in 9 of them (I., II., III., IV., V., IX., X., XI., XII.). An examination was probably made in VI., but it is not positively stated; in VII. there were the lesions of cerebral hemorrhage, and the pons was small; and in VIII. the skull was not opened.

Cord and Medulla.—As regards the microscopical appearance—*i. e.*, the size and shape of the cord and medulla—the latter is stated to have been very atrophic in IV.; and the cord was "small" in IV., V., VI., X., and XI.; while in 10 instances, viz., I., II., III., IV., VI., VIII., X., XI., XII., the flatness of the posterior columns is particularly specified. In 11 instances, therefore, all told, the size of all or part of the spinal cord was below normal, no allusion being made to this point in VII. *Meningitis* was observed in 10 cases, viz., I., II., III., IV., V., VI., VIII., IX., XI., XII.; limited in 6 of them (I., II., III., IV., VIII., IX.) to the posterior portion of the cord. In XI. and XII. the pia was diffusely thickened in places; in V. and VI. the position of the inflammation is not clearly stated, and in the former could scarcely be called a meningitis; III. and IV. exhibited thickening of the ependyma of the fourth ventricle, while in VII. and X. there was no meningitis at all. Considerable fluid was found in the sac of the dura in a number of cases, and in Friedreich's first 3 autopsies the posterior fissure was obliterated.

Medulla.—The medulla has been examined microscopically in 8 instances; though whether lesions were found is not always clearly stated. In I., II., III., and

IV. there is said to have been slight extension of the posterior sclerosis to the medulla, with traces of change in the fourth ventricle. In IV. there was also sclerosis of the right anterior pyramidal tract extending a little above the decussation of the pyramids, and with slight increase of connecting tissue with numerous corpora amylacea in the restiform bodies. The funiculi graciles and funiculi cuneati are described as sclerosed in IX., X., and XII., but in the last up to the level of the upper decussation only, while in the first it is further stated that there was no change above the decussation. In IX. the funiculi rotundi were also involved to some extent. Case VII. exhibited a patch of sclerosis reaching to the level of the hypoglossal nucleus ; in VI. there was nothing found to note, and in V., VIII., and XI. the medulla was not examined.

Cord.—A microscopical examination has been made in all cases, though the improved means of investigation now at our command might show more extended changes in I. and III. than were reported.

The *Posterior Columns* were sclerosed from the lumbar to the cervical enlargement in all the cases, and even higher and into the medulla in those just described ; and the sectional area was usually sclerosed very nearly throughout. The postero-median columns were greatly degenerated in all the cases, but the postero-external columns less intensely and uniformly affected. In I., II., and III. the sectional area of degeneration was greatest in the lumbar region, though everywhere complete ; but in IV. the degeneration was most marked in the cervical and dorsal regions. The whole sectional area of V. was very uniformly sclerosed, except a healthy layer next to the gray matter. Case VI. is said to have been just like

IV. In VII. only the portion was affected adjacent to the central canal and to the cornua, and the sclerosis was most pronounced in, and reached the periphery only in the lumbar region, though almost doing so in the cervical cord. In VIII. the posterior columns were diseased nearly throughout, though more completely in the dorsal than in the lumbar region, and a narrow portion adjoining the posterior commissure was not involved. IX. exhibited a degeneration in section which was most intense above, but very general everywhere, except a small tip where the posterior commissure joined the posterior horns, and even this was diseased in the upper part of the cord. In X., also, the sclerosis was general, except a small portion contiguous to the neck of the horn in the lumbar and cervical regions. Finally, in XI. and XII. the degeneration of Burdach's columns increased somewhat in a downward direction as far as the lower dorsal region, below which it again diminished; in these cases, too, there remained a narrow strip of healthy tissue at the junction of the posterior horn with the posterior commissure, or along the posterior horn.

Gowers states that the cervical root zone never escapes in Friedreich's ataxia, as it sometimes does in tabes; but in the cervical cord of XI. the postero-external columns were scarcely at all involved.

The *Lateral Pyramidal Tracts* are reported sclerosed throughout nearly their whole length in 11 cases; and possibly the disease was overlooked in the remaining one for reasons already given. The degree of degeneration in sectional area in these cases varies considerably. In II. and III. only the portions adjacent to the posterior horns were affected throughout the cord, and in the latter the change could only be detected by the mi-

croscope. The degeneration in IV. occupied merely the posterior parts of the lateral tracts, but reached quite to the periphery, while nearly the whole length and sectional area were sclerosed in V. Case VI. is described as resembling IV. In VII. the degeneration was situated at the junction of the anterior and posterior horns, and reached the periphery in the lumbar cord only; VIII. exhibited more sclerosis in the dorsal than in the lumbar region; it was especially well marked in IX. in the cervical swelling, and in X. in the dorsal cord; while in XI. and XII. it seemed to decrease from above downward, and at any rate was less marked in the lumbar portion.

A very important feature is still to be remarked, namely, that in 7 cases (IV., V., VI., IX., X., XI., XII.) the diseased portion of the lateral tracts did not immediately join the posterior horns; but was separated by a band of healthy white tissue. No direct statement is made concerning the matter in II., III., and VII., but it would seem that no such band existed in these cases, and it was clearly absent in VIII.

The *Direct Cerebellar Tracts* would appear from descriptions and drawings to have been more or less involved in 7 cases (IV., V., VI., VIII, X., XI., XII.) throughout all or nearly all of their length, with a greater or smaller sectional area of degeneration.

A *Peripheral Degeneration* passing forward from the sclerosed cerebellar tracts has been described in 5 cases, viz., IV., VI., VIII., X., and XI. An excellent example is the case of Gowers' and Pitt's (X.), in which it was well marked, and extended even along the anterior median fissure from the cervical to the upper dorsal region, though below this point it was irregular and

slight. In IV. it was to be seen only in the cervical cord, and in VI. the position was the same, but the zone of degeneration was not so complete, and did not extend so far forward. Gowers figures slight peripheral sclerosis in the lumbar cord in a section which he examined from Smith's case (VIII.). Case XI. had but a partial development of this sclerosed zone, and only in the upper cervical region.

The *Anterior Pyramidal Tracts* were sclerosed to some extent in 6 cases (IV., V., VI., VII., VIII., X.). In IV. and V. the degeneration extended downward on the right side into the dorsal region, and in VI. both sides were irregularly sclerosed from the decussation downward. Case VII. showed irregular degeneration on both sides, chiefly in the neighborhood of the anterior cornua; in VIII. there was a bilateral sclerosis in the dorsal region; and in X. from the upper dorsal cord upward.

Changes in the *gray matter* are reported as follows:

The *Columns of Clarke* were found to be degenerated in 8 instances, viz., II., IV., V., VI., IX., X., XI., XII. Friedreich makes no reference to them in I., II., and III.; but Schultze found the degeneration years later in II.; and perhaps could have detected it in I. and III. Brousse (VII.) does not speak of the condition of gray matter. Smith (VIII.) says that the nerve cells of the posterior cornua were at all levels far less numerous than usual, and in this statement probably includes Clarke's columns, which would increase the number of cases to 9.

Other changes in the gray matter were sometimes seen. Thus in II. there was sclerosis of the middle and posterior portions from the cervical enlargement to the mid-dorsal region, and even the anterior horns were affected to

some extent. In IV. there was diminution of the volume of the gray matter, especially evident in the cervical enlargement. In VIII. the cells of the anterior horns as well as of the posterior horns (see above) were diminished in number. There was degeneration between the anterior and posterior horns in IX.; and in X. portions of the posterior horns were affected in some sections. Inflammation in and about the central canal was seen in Cases II., VII., and VIII., where the canal was filled with small round cells; and in XI., in which it was found obliterated in most sections. Supplementary canals have also been noticed in rare instances. In VIII. a small channel of this nature was situated a little posterior to the main tube, and extended throughout nearly the whole lumbar region. In X. there were 2 canals not more than 1 inch in length, lined by columnar epithelium, and situated one behind the other in the central portion of the cord at the decussation of the pyramids, thus taking the place of the usual single central canal. In III. there were 2 supplementary canals, about 1 line in diameter, beginning at the lumbar swelling and extending half way to the cervical enlargement. They were situated one on each side, chiefly in the gray matter, at the junction of the posterior and anterior horns, and were filled with a clear fluid. In the upper half of the cord they were represented by foci of gelatinous material occupying similar positions, and downward in the lumbar enlargement some traces of these were found.

NERVES.—*The posterior nerve roots* were sclerosed in all the cases, and in general extensively, as in VIII., where there was scarcely a fibre not degenerated. In a few cases the sclerosis was less severe, as in VII., where

the nerve roots were only affected in the lumbar region; and in X., where only some of the fibres were diseased. Some strands of the *anterior nerve roots* were sclerosed in VIII.; slight changes were found in the *sciatic*, *crural*, *brachial*, and *hypoglossal nerves* in I.; and the *median* and *sciatic* were somewhat degenerated in XII.

Histology.—The histological nature of the changes found in the spinal cord is described by Friedreich as simple sclerosis; an overgrowth of the neuroglia at the expense of the nerve fibres. A finely fibrillated or granular tissue develops, traces of the nerve fibres are scarcely to be found, corpora amylacea are frequently very numerous, and the large cells of the gray matter shrink and lose their processes. In Case III. the lateral columns underwent a simple softening instead of the usual sclerotic change.

Pathology.

The nature of Friedreich's ataxia and its position among other diseases have been matters of considerable discussion. Friedreich, Erb, Möbius, Carre, Ormerod, and Grasset classify it as a variety of tabes only. In fact, Eulenburg, Topinard, and Carre do not distinguish it at all from this affection, and the last two writers include Friedreich's cases in their works on locomotor ataxia. Friedreich, however, would seem to separate it from locomotor ataxia rather more sharply than do the others. Bourneville considered it a combination of tabes and disseminated sclerosis. There are, indeed, clinical reasons why the belief in its intimate connection with disseminated sclerosis could be entertained, for many of the symptoms, especially in some cases, are

strikingly similar, yet none of the autopsies which have been made reveal any instance of lesions *en plaques*. Friedreich's ataxia certainly belongs to quite another group of diseases, while both anatomical and clinical symptoms distinguish it also from tabes. Hammond was of the opinion that the ataxia was caused by cerebellar disease, but this is entirely unconfirmed by the results of post-mortem examinations; and though Vizioli, Galassi, and Musso agree with him regarding its cerebellar origin, they do not consider it due to disease of the cerebellum, but to interference with the innervation from this centre in its descent through the diseased cord. They entertain the view expressed by Seppilli, Brousse, and Charcot, and probably most generally held, viz., that Friedreich's ataxia is a distinct, independent affection. It is placed by some writers intermediate clinically between tabes and disseminated sclerosis, but more exactly by Gowers between tabes and ataxic paraplegia. The early age at which it appears led Friedreich to the conclusion that there existed a congenital disposition to it; that there was an hereditary arrest of development of the spinal cord *in toto;* and that the poorly developed organ was readily attacked by a sclerosing process. This same view is adopted by Schultze, Möbius, Vizioli, Musso, and others.

The question as to the primary lesion is an exceedingly interesting and difficult one. Friedreich says that the nature and position of the anatomical changes render it certain that the spinal disease cannot be secondary to an alteration in the medulla, or *vice versa;* and, basing his belief on the first 3 autopsies, he claimed that the meningitis was the primary cause and that other lesions were secondary to this. After the autopsy on

Friedreich Suss (IV.), however, he concluded that the degeneration of the posterior columns in all his cases wss primary, but that the disease of the lateral columns, and in IV. of the peripheral zone, was secondary to the meningitis which the posterior sclerosis had set up. Smith entertains the same opinion, though claiming that the secondary inflammation is not of a diffuse nature. Brousse maintains that the posterior sclerosis is primary and systemic, and that extension from it occurs in two ways: the one by means of the inflamed central canal, the other peripherally by a leptomeningitis; the disease of other parts of the cord being secondary and diffuse. Schultze considers the affection in Cases IV. and VI., which he examined, to be a degenerative atrophy of the entire posterior half of the cord, with certain slight exceptions, and claims that though the distribution of the sclerosis is too regular for disseminated sclerosis, it is too irregular for a genuine systemic disease. Kahler and Pick think that it is not an arrested growth of the cord *in toto*, but rather an imperfect development of the fibres of certain nervous systems which is the primitive cause of the disease. This development ceases at about the time of the sheath formation, and is induced by hereditary influences; and later in life, under the action of some agencies unknown to us, degeneration takes place in these imperfectly formed fibres. These authors were, I think, the first to assert that Friedreich's ataxia was a hereditary combined systemic disease of the cord—*i. e.*, a simultaneous and independent degeneration of two or more systems of nerve fibres. They regard the peripheral degeneration as a later and accessory change, perhaps due to a subsequently developing chronic meningitis. Rütimeyer adopts the same view regarding

both the nature of the disease and the origin of the peripheral sclerosed zone, and Erlicki and Rybalkin claim that the post-mortem lesions in their case can only be accounted for by admitting the presence of a combined systemic spinal disorder. Pitt proposes a very rational theory which well explains the arrested development of the cord *in toto*, and at the same time harmonizes with the views of Kahler and Pick. He believes that the general or partial smallness of the cord is congenital, being a consequence of an inherited tendency to early vascular degeneration. The posterior columns, being naturally the most vascular, of course suffer the most, and are the most diminished in size. The earliest histological changes consist less in an excessive growth of neuroglia than in an imperfect development of the medullary sheaths, followed later by a degeneration; the first cause being probably an endarteritis. These views necessitate the belief in a systemic degeneration of various parts of the cord, and are sustained by the fact that the tracts most sclerosed correspond very nearly to those which are the last to acquire medullary sheaths in fœtal life; and further that puberty or an acute illness, with the vascular changes which accompany both, often precipitates or accelerates the development of the symptoms of the ataxic disorder. Again, it is impossible in Cases X. and VII. that the affection could have spread from a meningitis, in spite of the peripherally sclerosed zone in the former, since no meningitis existed, and in IX. too, it did not pass beyond the surface of the posterior columns; and on the other hand, the 7 instances in which there was a band of healthy tissue in the lateral tract adjacent to the posterior horn confute the supposition that in these,

—the majority of cases,—there had been an extension of degeneration by contiguity from tract to tract, or through the posterior horn to the lateral tract. Therefore, although more autopsies are needed to settle the question positively, the evidence is strongly in favor of the view that Friedreich's ataxia is a combined systemic spinal disease, at least for certain parts of the cord. Whether there may also occur in some instances a diffuse secondary inflammation of other districts does not seem so clear, though it is possibly the case.

The cause of the bulbar symptoms cannot be satisfactorily explained by any of the pathological lesions yet discovered. The usual absence of pain and of diminution of cutaneous sensibility is likewise not easy of explanation. There is reason to believe that these disturbances are not connected with involvement of the exterior portion of Burdach's columns, as has been supposed, since in certain cases, as IX., the whole transverse section of the posterior columns was sclerosed, yet the cutaneous sensibility was unaffected. Rutimeyer advances the theory that they depend on disease of the boundary layer of the posterior horn (*Rand Zone* of Lissauer), since in his 2 autopsies this zone was unaffected, while in tabes its involvement is one of the most characteristic changes in the gray matter. Gowers states that Friedreich's ataxia differs anatomically from ataxic paraplegia in the greater degree of affection of the posterior columns in the lumbar region, and in the involvement of the posterior nerve roots. It is on this account that the exaggerated knee-jerk and ankle clonus are absent in the former and present in the latter disease.

Diagnosis.

The recognition of Friedreich's ataxia is not always easy; the principal and usual conditions on which a diagnosis may be based being, 1, the evidence of some hereditary influence; 2, the occurrence of several cases in a family; 3, the early age at which the symptoms appear; 4, the presence of motor ataxia spreading from below upward; 5, static ataxia in more advanced cases; 6, affection of speech; 7, nystagmus late in the disease; 8, talipes; 9, curvature of the spine; 10, frequently paralysis late in the disease. Negative symptoms are the absence, 1, of patellar reflex; 2, of marked sensory lesions, at least early in the disease; 3, of decided trophic lesions; 4, of affection of intellect; 5, of affection of the optic nerve; 6, of visceral or vaso-motor disturbances. But some of these symptoms may be absent in certain cases, and others may appear which suggest the presence of some other affection; so that in this, as in all diseases, the diagnosis must be based on the aggregation of symptoms rather than on individual ones. (For anomalous cases see Remarks on Table I.)

Clinically, Friedreich's ataxia is most closely allied to tabes dorsalis, disseminated sclerosis, and ataxia paraplegia.

Uncomplicated *tabes dorsalis* rarely appears before the age of 25–30, and the cases occurring during childhood are of the utmost rarity, if, indeed, genuine. It exhibits neither incoördination of speech nor nystagmus; though lesions of the optic nerve, affection of the pupillary reflex, and other disturbances connected with the eye are frequent and early symptoms. Severe pain is one of the first and most common signs of the disease;

other affections of sensation are usual and appear early; and visceral and atrophic affections are generally present. The gait of tabes is peculiar; the feet being projected too far forward, while in Friedreich's ataxia the projection is often more lateral, and the gait is oscillating, as has been described. Charcot says that "static ataxia" is rarely seen in tabes.

Disseminated sclerosis, whose symptoms still more nearly resemble those of Friedreich's ataxia, usually develops after the age of 20, though it may appear in infancy. The influence of heredity is exceptional, and the progress of the disease is not infrequently marked by remissions which seem almost like recovery. Nystagmus and affection of speech are common to both disorders, but in disseminated sclerosis the former is usually more extreme than in the other disease, and is not simply "ataxic nystagmus;" while the speech is typically scanning—a feature which may or may not be present in Friedreich's ataxia. Some of the more distinguishing symptoms of disseminated sclerosis are the exaggerated knee-jerk, ankle clonus, rhythmic oscillations, violent jerking movements, intention tremor, rigidity and spastic paraplegia, disturbance of intellect often with epileptiform and apoplectiform attacks, and the absence of Romberg's phenomenon. These symptoms either do not occur at all in Friedreich's ataxia, or are seen only exceptionally.

Ataxia paraplegia develops, according to Gowers, between 30 and 40 years of age, and does not occur in several members of a family. The knee-jerk is exaggerated, and there is no nystagmus. Marked affection of speech is wanting, though there is sometimes slight defect in articulation.

Of the other diseases with ataxic symptoms, *cerebellar tumor* can hardly be confounded with the disease in question. The gait is "drunken," and resembles many cases of the spinal disease; but there is no involvement of the arms, and the tendon reflexes are present; while tumor also produces headaches, optic neuritis, vomiting, epileptiform attacks, and marked vertigo. Certain instances of Friedreich's ataxia exhibiting excessive static ataxia or choreiform movements, as in Sinkler's last 3 cases (137, 138, 139), might raise the question as to the diagnosis of the affection from *hereditary chorea.* This disorder, however, is distinctly hereditary (direct similar inheritance) for generations, and without a break; nearly always appears after the age of 35–40 years; has no truly ataxic symptoms, and is almost invariably sooner or later associated with severe disturbances of intellectual power with a tendency to suicidal mania. There is, further, no diminution of the patellar reflex.

Duration, Prognosis, Treatment.

Duration.—The course of Friedreich's ataxia is steadily onward, and though there frequently occur periods during which no decided progress of the disease can be detected, yet improvement in the symptoms rarely takes place. This is the opinion generally accepted, and I recollect but 3 cases—one of Smith's (100), one of Teissier's (60), and that of Leubuscher—in which notable improvement was observed. In Teissier's case the symptoms largely disappeared during 2 years, and then returned; though not to so great a degree up to the time when the case was last examined. The dura-

tion of the disease is very extended, unless it be terminated by some intercurrent affection. Many patients are able to follow their avocations for years, with very little increase in the severity of the symptoms. 2 of Friedreich's cases (2 and 5) were ataxic 34 and 27 years respectively, and died finally of typhoid fever. One of Smith's cases (101) has had the disease 31 years; one of Stintzing's (123) 28 years; and one of Vizioli's (81) 41 years. But the longest duration reported is in another patient of Vizioli's (87) where the ataxia lasted from birth till he died at the age of 46. On the other hand, some cases have grown rapidly worse, and have died with advanced symptoms in but a few years from the onset. One of Hammond's cases (33), for example, suffered but 8 years; 2 of Coleman's patients (38, 39) never walked, and died at 3 years of age; and the other (37), as well as a case of Vizioli's (88), died at 6 years, and like the others had never walked. As far as I have been able to discover, 25 of the cases in the table are reported by the observers to have died. 5 of these (1, 3, 4, 6, 14, 38) are said to have died of typhoid fever; 1 (117) of typhoid fever and phthisis; 2 (19, 107) of phthisis; 1 (38) of some pulmonary affection; 1 (2) of nephritis; 1 (30) of apoplexy; 4 (5, 33, 34, 94) apparently of advancing weakness; 1 (46) of fatty heart; 1 (27) of fibroid heart; 1 (102) of acute myelitis; 1 (86) of "taking cold;" and 1 (88) of diarrhœa. In 1 (48) the cause of death is not clear, and in the remaining 4 (37, 39, 87, 128) there is no intimation given as to its nature.

Prognosis.—It will be seen from what precedes, that the prognosis is usually favorable as regards the duration of life, but most serious as regards recovery from

the disease. The patient will die of the progressive weakness in the course of Friedreich's ataxia, if not carried off by some intercurrent affection. How soon the habits of life will be seriously interfered with, and when the lethal *exitus* will probably take place, must be determined for each individual patient, by considering the former rapidity of progress in that particular case.

TREATMENT.—Treatment has proved unavailing in almost all cases. One of Teissier's patients (60) improved greatly, apparently through better hygienic surroundings. One of Smith's cases (100) improved temporarily under the use of nitrate of silver. This drug naturally suggests itself, and has, indeed, been used in a number of other cases, but without any decided benefit. Arsenic, phosphorus, iodine, zinc, may also be tried, and cauterization over the vertebral column has been proposed. The constant current applied to the spine seemed to be of benefit in Leubuscher's case. One of the chief indications is to sustain the general nutrition by tonics, iron, cod-liver oil, change of air, salt and other bathing, etc., and to stimulate by electricity and massage muscles which are apparently beginning to lose power. When the spinal curvature and the lack of control of, or lack of power in, the dorsal muscles render the patient unable to sit upright, a plaster jacket may be applied with great advantage. I have found this give great relief to one of my own patients (142). Case 102 (Smith's) also improved surprisingly under electrical treatment and the employment of a plaster jacket.

Remarks on some of the Cases in Table I.

I have already stated that I do not by any means consider that all the cases in the table are undoubted instances of Friedreich's ataxia, but that all of them have at least some claim to be considered such. The cases about which doubt has been or might perhaps be entertained are 7, 8, 11–12, 16–18, 19, 22–23, 31–36, 37–39, 55–56, 60, 68, 100, 108, 109, 111, 114–115, 117. Most writers have no question concerning Case 7 (Carre), but a few—*e. g.*, as Seeligmüller—doubt its genuineness chiefly on account of the very marked degree of initial and later pain. 8 (Bradbury) is not admitted by Bury, but has, I think, a good right to a place with other cases. Kellogg's cases (11–12) are not as well reported as could be desired, and are on this account excluded by many tabulators, but included by the Italian writers and by Bury. There seem to be good reasons to consider them genuine. Dreschfeld's patients (16–18) are certainly questionable, and some of them resemble tabes very greatly. They are tabulated by Ormerod, but rejected by Smith. The fact that there were 5 attacked in one family is their greatest claim for membership among this class of cases. Most authorities do not hesitate to call 19 an instance of Friedreich's ataxia, though Rütimeyer questions it, and Gowers assigns it to ataxic paraplegia. 22–23 (Seeligmüller's) are excluded by many writers, but considered genuine by Smith, Vizioli, Prince, and others, with whom I am disposed to agree. Hammond's cases (31–36) are so briefly and imperfectly described that Smith rejects them altogether, and they are very generally called in ques-

TABLE I.—OBSERVED AND REPORTED CASES. ARRANGED CHRONOLOGICALLY.

Number.	Name.	Sex.	Observer. (Brackets indicate those of one family of brothers and sisters.)	Ataxia in parents, grand-parents, uncles or aunts.	Other neuroses, including great nervousness in parents, etc.	Alcoholism in parents, etc.	Tuberculosis of parents, etc.	Syphilis, etc.	Consanguinity of parents or grandparents.	Other causes in parents, grandparents, etc.	Number of cases observed in children of one family.	Number of other children reputed ataxic.	Other neuroses in children of family.	Other suspicious conditions in children of family.	Where the ataxic symptoms began.	Age when first attacked.	Age when upper extremities affected.	Age when bulbar symptoms appeared.	Age when last examined, or when died.	Ataxia precipitated by acute febrile disease.	Ataxia accelerated by acute disease.
1	Andreas Lotsch,	M.	Friedreich,	0	...	+	...	...	...	...	2	0	0	0	l	17	25	26	35*	...	...
2	Charlotte Lotsch,	F.	"	0	...	+	...	...	...	...	2	0	0	0	l	17	25	25	51*	...	P
3	Justinia Süss,	F.	"	0	+	+	+	...	...	...	4	0	0	0	l	15	19	20	31*	...	...
4	Salome Süss,	F.	"	0	+	+	+	...	...	...	4	0	0	0	l	15–16	19	25	28*	...	...
5	Lisette Süss,	F.	"	0	+	+	+	...	...	...	4	0	0	0	l	14	<28	...	41*	...	...
6	Friedreich Süss,	M.	"	0	+	+	+	...	...	...	4	0	0	0	l	14	16	17	38*	...	...
7	Sophie C.,	F.	Carre,	±	...	...	...	...	...	...	1	6	...	...	l	22	...	..	28	...	...
8	M. C.	M.	Bradbury,	+	...	...	...	...	...	...	1	1	...	...	l	17	...	...	18	...	...
9	M. H.	F.	Carpenter,	0	0	...	...	...	...	...	2	1	...	...	l?	12?	...	...	28	...	...
10	E. H.	F.	"	0	0	...	...	...	...	...	2	1	...	...	l	14	...	...	26	...	...
11	....	M.	Kellogg,	0	...	...	...	...	...	0	2	1	0	0	l	6	...	...	10	...	...
12	....	M.	"	0	...	...	...	...	...	0	2	1	0	0	l	5	...	...	7	...	...
13	Louise Schulz,	F.	Friedreich,	0	0	0	...	...	...	0	3	0	0	+	l	12	12	13	30	...	R
14	Katharina Schulz,	F.	"	0	0	0	...	...	...	0	3	0	0	+	l	12	<13	13	19*	...	...
15	Marie Schulz,	F.	"	0	0	0	...	...	...	0	3	0	0	+	l a	12	12	<13	18	...	...
16	James B.	M.	Dreschfeld,	0	...	...	...	...	...	...	3	2	—	...	l	20	40	...	42	T	...
17	John B.	M.	"	0	...	...	...	...	...	...	3	2	—	...	l	23	...	...	32	...	...
18	Mrs. Martha C. (née B).	F.	"	0	...	...	...	...	...	...	3	2	—	...	l	20	...	...	34	...	...
19	Josefa Strasik,	F.	Kahler & Pick,	0	...	...	+	...	...	...	1	0	...	+	l	15	22	22	23*	...	...
20	George B.	M.	Schmidt,	0	0	+	...	...	...	0	2	0	0	0	l	11	...	...	21	...	...
21	Linus B.	M.	"	0	0	+	...	...	...	0	2	0	0	0	l	10	...	...	12	...	...
22	Leo v. K.	M.	Seeligmüller,	...	±	...	...	...	+	...	2	1	...	...	l	9–12	...	<22	26	...	...
23	Alexander v. K.	M.	"	...	±	...	...	...	+	...	2	1	...	...	1	27?	...	...	28	...	...
24	Harry G.	M.	Hollis,	0	0	...	...	...	...	...	1	0	0	0	l	9	...	...	13	0	...
25	...... Shaw,	M.	Gowers,	...	±	...	...	0	...	...	5	0	0	0	l	21	...	...	39	...	...
26	...... Shaw,	F.	"	...	±	...	...	0	...	...	5	0	0	0	l	18	...	...	29	...	...
27	Robert Shaw,	M.	"	...	±	...	...	0	...	...	5	0	0	0	l	19	...	...	28–29*	...	...
28	...... Shaw,	M.	"	...	±	...	...	0	...	...	5	0	0	0	l	19–20	...	...	22	...	...
29	...... Shaw,	M.	"	...	±	...	...	0	...	...	5	0	0	0	l	18–19	...	...	19	...	...
30	Marie R.	F.	Brousse,	+	...	...	—	...	...	...	1	...	...	...	l	24	...	...	32*	...	...
31	J. T.	M.	Hammond,	0	...	...	...	...	...	...	2	0	...	...	l	4	6	6	19	...	...
32	H. T.	M.	"	0	...	...	...	...	...	...	2	0	...	...	l	10	>11	>11	21	...	...
33	M. B.	M.	"	0	0	...	...	...	...	...	2	0	0	0	l	6	<8	8	14*	...	...
34	C. B.	M.	"	0	0	...	...	...	...	...	2	0	0	0	l	8	9	...	15*	...	...
35	E. M.	M.	"	0	0	...	...	...	...	...	2	0	...	...	l	3	6–7	6–7	30	...	...
36	H M.	M.	"	0	0	...	...	...	...	...	2	0	...	...	l	7	...	...	26	...	...
37	Green R.	M.	Coleman,	0	0	...	...	...	...	...	3	0	0	0	l	1	...	...	6*	...	...
38	Frank R.	M.	"	0	0	...	...	...	...	...	3	0	0	0	l	½	...	...	3*	...	...
39	Willie R.	M.	"	0	0	...	...	...	...	...	3	0	0	0	l	in	...	...	3*	...	...
40	Nannie W.	F.	Warren,	0	±	0	0	0	0	...	4	0	0	0	l	8	12	21	26	0	B
41	Thomas W.	M.	"	0	±	0	0	0	0	...	4	0	0	0	l	8	12	17	17	0	0
42	Robert W.	M.	"	0	±	0	0	0	0	...	4	0	0	0	l	8	12	...	15	0	0
43	Nettie W.	F.	"	0	±	0	0	0	0	...	4	0	0	0	l	6	...	...	8	...	...
44	....	M.	Leubuscher,	0	0	...	...	...	...	...	1	0	0	0	l	3	...	...	3½	0	...
45	....	M.	Power,	0	0	...	—	...	...	...	1	2	...	...	l	15	...	...	22	...	...
46	Heinrich Kern,	M.	Quincke and	0	+	...	—	...	...	0	3	0	0	0	l	5	<13	<13	20*	...	...
47	Hedwig Kern,	F.	Rütimeyer,	0	+	...	—	...	...	0	3	0	0	0	l	6–7	...	...	16	Ps	...
48	Bertha Kern,	F.	Rütimeyer,	0	+	...	—	...	...	0	3	0	0	0	l	6	...	...	16*	...	R
49	Rudolf Blattner,	M.	"	—	0	...	...	..	...	0	3	0	0	0	l	6	<13	<13	17	Sc	...
50	Gottlieb Blattner,	M	"	—	0	...	...	...	...	0	3	0	0	0	l	6–7	<15	<15	34	...	...
51	Marie Blattner,	F.	"	—	0	...	...	...	...	0	3	0	0	0	l	5	<14	<14	29	...	...
52	Rudolf Blattner, 2d,	M.	"	—	0	...	...	...	...	0	1	0	0	0	l	7	8	8	15	T	...
53	Karl Blatner,	M.	"	—	±	...	...	...	...	0	1	0	+	0	l a s	<5	<5	<5	10–12	...	...
54	Marie Magd. Blattner,	F.	"	—	0	...	...	...	...	0	1	2	0	0	l a	4–5	4–5	12–15	20	...	...
55	....	M.	Jakubowitsch,	0	0	...	0	0	...	0	1	0	0	0	l	10	...	...	10	...	...
56	Wm. Krämer,	M.	Erlenmeyer,	0	0	...	...	...	+	0	1	0	0	0	l a	5	5	...	8	Mn	...
57	Andreas Bösch,	M.	Wälle,	0	—	...	...	...	...	...	2	0	...	+	l	6	<11	<11	20	...	...
58	Heinrich Bösch,	M.	"	0	—	...	...	...	...	...	2	0	...	+	l	4	...	...	12	...	...
59	Louis D.	M.	Teissier,	0	—	...	...	...	...	...	2	0	...	...	l a	9–10	...	...	19	...	...
60	Marie D.	F.	"	0	—	...	...	...	...	...	2	0	...	...	a	10	...	...	18	...	...
61	Antonia Marchis,	F.	Musso,	—	±	...	—	...	...	—	3	0	—	+	l	15	21	26	28	...	...
62	Anna Marchis,	F.	"	—	±	...	—	...	...	—	3	0	—	+	l	12	22	...	27	...	...
63	Maria Marchis,	F.	"	—	±	...	—	...	...	—	3	0	—	+	l	13	...	...	20	...	...
64	Antonio Dardino,	M.	"	—	±	...	—	...	...	—	1	2	...	+	l	25	35	37	43	V	...
65	Luigia Rizzatti,	F.	Massalongo,	0	+	—	...	...	...	...	2	0	...	...	l	1½	...	5	10	...	...
66	Albino Rizzatti,	M.	"	0	+	—	...	...	...	...	2	0	...	...	l	1½	...	...	6	...	...
67	....	M.	Charcot,	0	0	...	...	...	...	...	1	0	—	0	l a s	in	in	in	18	...	...
68	Frederich Malluch,	M.	Botkin,	+	+	...	...	...	...	...	1	1	...	+	l a s	18	18	18	22	...	...
69	Kate Woodcock,	F.	Ormerod,	—	±	+	...	...	...	...	5	0	0	+	l a	11	11	...	18	D	Sc
70	John Woodcock,	M.	"	—	±	+	...	...	...	...	5	0	0	+	l	11	...	...	14	D	...
71	Hannah Woodcock,	F.	"	—	±	+	...	...	...	...	5	0	0	+	l?	16–19	20	...	20	D	...
72	.... Woodcock,	F.	"	—	±	+	...	...	...	...	5	0	0	+	...	9–10	...	...	9–10	...	...
73	.... Woodcock.	M.	"	—	±	+	...	...	...	...	5	0	0	+	...	7–8	...	...	7–8	...	...
74	Ruth Harmer,	F.	"	0	...	+?	...	...	...	...	2	1	—	+	l	13	...	...	20	...	D
75	Alice Harmer,	F.	"	0	...	+?	...	...	...	...	2	1	—	+	l	13	...	...	16	...	...
76	....	F.	Buzzard,	0	0	...	...	...	...	0	1	0	0	...	l	5–6	...	...	26	...	T
77	....	M.	Fowler,	+	...	+	...	...	...	...	3	0	...	...	...	...	...	...	14	0	...
78	....	F.	"	+	...	+	...	...	...	...	3	0	...	...	...	...	...	...	9	0	...
79	....	M.	"	+	...	+	...	...	...	...	3	0	...	...	...	...	...	...	5	0	...
80	Sebastiano Conti,	M	Fazio,	0	0	...	...	...	...	...	1	2	...	...	l	10–11	...	...	29	0	0
81	Vincenzo Vitielli,	M.	Vizioli,	0	+	+	...	...	...	...	8	0	+	..	l	<14	14	14	44	...	...
82	Andrea Vitielli,	M.	"	0	+	+	...	...	...	...	8	0	+	...	l	<15–16	15–16	...	33	...	...
83	Filomena Vitielli,	F.	"	0	+	+	...	...	...	...	8	0	+	...	l	3	13	...	32	...	T
84	Margherita Vitielli,	F.	"	0	+	+	...	...	...	...	8	0	+	...	l	3	>15	...	30	...	...
85	Maria Vitielli,	F.	"	0	+	+	...	...	...	...	8	0	+	...	l	3	...	...	28	...	T
86	Rosa Vitielli,	F.	"	0	+	+	...	...	...	...	8	0	+	...	l	3	12–13	12–13	34*	...	...
87	Giuseppi Vitielli,	M.	"	0	+	+	...	...	...	...	8	0	+	...	l	<2	...	...	46*	...	...
88	Francesca Vitielli.	F.	"	0	+	+	...	...	...	...	8	0	+	..	...	<2	...	...	6*	0	0
89	Antonio Vitielli,	M.	"	±	—	—	...	...	...	...	2	0	0	...	l	1¾	4	...	6	...	Sc
90	Luigi Vitielli,	M.	"	±	—	—	...	...	...	...	2	0	0	...	l	1½	...	...	3	Sc	...
91	Vincenzo Luongo,	M.	"	0	+	...	...	—	...	...	1	1	0	...	l	<4	...	<5	11	...	...
92	Giuseppe Perino,	M.	Palma,	0	+	...	±	...	...	...	1	0	0	—	l	<2	...	...	7	...	...
93	Lizzie K.	F.	Seguin,	0	+	...	...	...	...	0	3	0	0	0	l	4	15	>18	26	...	...
94	Lindley K.	M.	"	0	+	...	...	...	...	0	3	0	0	0	l	10	>15	>15	21*	...	...
95	Nettie K.	F.	"	0	+	...	...	...	...	0	3	0	0	0	l	<7	...	...	15	...	...
96	J. H.	M.	"	...	...	...	+	...	...	...	2	1	0	—	l	7	...	...	18	...	...
97	M. H.	M.	"	..	...	...	+	...	...	...	2	1	0	—	l	15	28	28	32	...	...
98	C. G.	M.	Sinkler,	0	...	...	—	...	...	...	1	0	0	0	l	<11	...	...	18	...	Sc–Ml
99	Mary G.	F.	"	0	0	...	...	...	...	...	1	0	0	0	l a	8	8	...	11	...	...
100	J. W. W.	M.	Smith,	0	+	+	...	0	0	...	1	0	0	+	l	66	...	...	69	R	...
101	Emma W.	F.	"	+	—	—	...	0	0	0	4	1	0	0	l	6	...	...	37	0	0
102	Clara W.	F.	"	+	—	—	...	0	0	0	4	1	0	0	l	9–10	16	...	29*	0	My
103	Jessie W.	F.	"	+	—	—	...	0	0	0	4	1	0	0	l	8	...	...	26	0	0
104	Myra W.	F.	"	+	—	—	...	0	0	0	4	1	0	0	l	10	...	...	18	0	0
105	Catherine L	F.	Putnam,	0	0	...	...	...	...	...	2	0	0	0	l	<5	...	..	20	...	...
106	Jerry L.	M.	"	0	0	...	...	...	...	...	2	0	0	0	a	7	7	...	16	Sc	...
107	Mary D.	F.	Prince,	0	0	...	...	0	...	...	1	0	0	0	l	15	...	...	19*	...	...
108	....	M.	Fellows,	...	...	...	...	...	...	0	1	+?	...	...	l	18	...	...	21	...	...
109	....	...	McAlister,	...	...	...	...	...	...	...	1	...	...	...	l?	19?	28–29	...	29	...	...
110	....	M.	Fagge,	...	...	...	...	...	...	...	1	0	+	...	l	16	25	25	30	...	...
111	Alfred D.	M.	Descroizilles,	0	0	0	...	0	...	...	1	0	+	...	l s	2	...	2	8	...	...
112	Edward G.	M.	Bury,	—	0	—	0	0	...	...	2	0	+	0	l	15–16	21	21	22	0	0
113	William G.	M.	"	—	0	—	0	0	...	...	2	0	+	0	l	12–13	14–15	...	19	0	...
114	John W.	M.	"	0	0	0	±	...	...	...	1	0	0	0	l	7	...	12–13	16	M&Sc	...
115	Jane R.	F.	"	0	0	...	...	...	...	...	1	0	+	0	l a	7	7	11	12	0	0
116	....	F.	Galassi,	0	0	0	0	0	0	0	1	0	0	0	l a s	8	8	8	16	+	0
117	J. N.	F.	Erlicki & Rybalkin,	...	...	...	...	...	...	...	1	...	...	...	l	16	18	...	18*	+	...
118	Frederick B.	M.	Glynn,	0	+	0	0	0	0	...	1	0	...	...	l	17	...	...	21	0	0
119	Otto N.	M.	Freyer,	0	0	...	—?	+?	...	+	3	0	—	—	l	10	...	...	19	Ps	...
120	Hans N.	M.	"	0	0	...	—?	+?	...	+	3	0	—	—	l	<5	...	...	16	...	...
121	...... N.	M.	"	0	0	...	—?	+?	...	+	3	0	—	—	l	...	...	...	9	...	...
122	Paul P.	M.	Blocq,	0	0	...	...	...	...	...	1	0	0	0	l	10	12	>12	20	0	0
123	Therese R.	F.	Stintzing,	0	0	0	...	...	...	...	3	1	—	0	l	4	...	...	32	...	...
124	Rosalie R.	F.	"	0	0	0	...	...	...	...	3	1	—	0	l a	8	8	...	15	...	...
125	Joseph R.	M.	"	0	0	0	...	...	...	...	3	1	—	0	...	8	...	...	13	...	...
126	A. F.	M.	Ferrier,	0	...	...	+	...	...	...	1	0	0	0	l	7	...	14	18	...	...
127	Paul X.	M.	Charcot,	...	...	...	...	...	...	...	1	...	...	...	...	10	...	...	20	...	...
128	Willis F.	M.	Mastin,	—	...	+	...	...	+	...	3	0	0	0	l	10–12	25	>25	32*	0	+
129	Ervin F.	M.	"	—	...	+	...	...	+	...	3	0	0	0	l	10	..	...	28	0	0
130	Abraham F.	M.	"	—	...	+	...	...	+	...	3	0	0	0	l	8	12	...	17	0	0
131	Emma F.	F.	Mendel,	0	0	...	...	0	+	...	1	1	...	...	l?	1	...	...	4	...	...
132	John Brand,	M.	Ormerod,	0	±	...	...	...	...	...	2	0	+	+	l	18	...	...	25–26	R	...
133	Linda Brand,	F.	"	0	±	...	...	...	...	...	2	0	+	+	l	5	17	17	29	...	Sc
134	Bridget Sullivan,	F.	"	0	0	+	...	...	...	...	1	0	—	0	a	16	16	...	21	...	...
135	H. O.	M.	Shattuck,	0	0	0	0	0	0	0	1	2	0	0	l	8	...	12	14	0	...
136	....	M.	Joffroy,	0	0	...	+	...	...	...	1	1	0	0	l	10	15	15	19	0	0
137	Annie MacDonald,	F.	Sinkler,	0	0	0	0	0	0	0	3	0	0	0	l a s	3	3	3	15	+	...
138	Michael MacDonald,	M.	"	0	0	0	0	0	0	0	3	0	0	0	l a s	2	2	2	12	...	...
139	Bernard MacDonald,	M.	"	0	0	0	0	0	0	0	3	0	0	0	l a s	2	2	2	11	...	...
140	Blanche Roberts,	F.	Osler,	0	0	...	0	...	...	...	1	0	0	0	l a s	3	3	3	11	...	...
141	Sadie T.	F.	Griffith,	0	0	...	...	...	...	...	2	0	0	0	l a	10	10	>16	23	0	T & M
142	George T.	M.	"	0	0	...	...	...	...	...	2	0	0	0	l	9	>18	...	21	M	0
143	Annie C.	F.	"	0	0	...	...	...	...	...	1	1	0	0	l	5	22	...	26	Sc	0

Number.	Ataxia of lower extremities.	Ataxic station.	Ataxia of upper extremities.	Ataxia of head (neck)	Ataxia of trunk.	Tremor.	Choreiform movements.	Spasmodic contractions of muscles.	Paralysis.	Inability to walk; and age when it developed.	Curvature of spine; cyphosis; scoliosis.	Talipes.	Other contractures.	Electrical contractility.	Patellar reflex.	Plantar reflex.	Other cutaneous reflexes.	Atrophy of muscles.	Other trophic changes; and vaso-motor affections.	Pain, initial, or as early symptom.	Pain, later in disease.	Cutaneous sensibility.	Muscle sense.	Girdle sensation, or other paræsthesias.	Affection of speech.	Tremor of tongue.	Nystagmus.	Affection of optic nerve.	Affection of intellect.	Vertigo.	Visceral and secretory disturbances.
1	...	...	±	+	...	...	...	...	+	18	0	...	...	N	...	...	...	0	...	0	0	N	N	...	±	—	0	...	0	0	—
2	f +	...	± X	+	+	0	...	+	±	<31	CS	e r	0	—F	0	N	...	+	—	0	+	=	N	0	±	0f—	+	0	0	±	+
3	+	...	+	+	...	...	...	+	...	0	CS	...	...	N	...	...	...	0	...	+	+	—	...	...	±	...	+	...	0	+	...
4	...	...	±	...	...	...	...	—	...	+	CS	...	...	N	...	...	...	0	...	+	+	N	...	...	+	...	—	...	...	0	+
5	...	...	+	+	+	...	...	+	...	0	CS	...	...	...	...	...	...	—	...	+	+	N	...	...	±	+	0	...	0	0f+	+
6	±X f I	+X f I	± X	+	...	...	...	...	0	>24	S	...	...	N	0	N	N	0	...	...	...	N	N	—	±	...	+	...	0	+	—
7	+ X	...	+	—	...	...	...	...	0	0	...	...	...	...	...	...	...	0	...	±	...	N, +	...	+	+	...	0?	...	0	...	...
8	±	+ I	+ I	...	...	...	...	...	0	0	0	...	...	...	...	N	...	0	...	+	—	N	...	0	...	...	...	...	0	+	+
9	± I	± I	+ I	...	...	...	...	0	=	0	S	...	...	...	0?	0	...	...	...	0	0	—	...	0	—	...	...	...	0	...	...
10	+	...	+ I	...	...	...	...	...	...	0	...	...	...	...	...	...	...	...	...	0	0	...	...	...	—	...	...	...	0	...	...
11	±	...	+	+	...	...	...	...	0	+	...	...	...	N	...	...	...	...	...	...	...	—	...	...	+	...	+ ?	...	—?	...	...
12	+	...	...	...	...	...	...	...	0	0	...	...	...	...	...	...	...	0	...	...	...	...	...	...	0	...	0	...	...	...	...
13	± X	± X	± X	+	+	...	=	...	0	0	...	...	...	N	0	N	N	0	...	—	=	—?	N	0	±	0	...	...	0	...	—
14	± X	0 X	± X	...	...	...	...	...	0	0	...	...	...	...	...	...	...	0	...	—	...	N	...	0	+	...	0	...	...	...	...
15	+ X	+ X	+ X	...	...	...	...	...	0	0	...	...	...	...	0	N	N	0	...	0	0	N	N	...	+	0	+	...	0	...	...
16	+ I	+ I	— X	...	...	...	...	...	—	0	...	...	...	N	...	...	...	0	...	±	...	=	...	...	0	...	0	0	...	...	...
17	— I	±	0 X	...	...	...	...	...	...	0	...	...	...	=	...	...	...	+	...	±	+	N	...	—	0	...	0	0	...	...	—
18	+ I	...	0	...	...	...	...	...	0	0	...	...	...	...	...	...	...	0	...	+	...	—	...	...	0	...	...	...	...	...	...
19	f ±	...	+ X	—	—	...	...	...	±	22	S	...	0	=	0	...	...	+	—	0	0	N	N	0	+	...	0	0	—?	...	+
20	± X	f ± X	± X	+	+	0	...	0	0	>19	...	...	...	N	0	N	N	0	...	0	0	—	N	0	+	0	+	...	0	...	...
21	+	0	...	...	...	...	...	...	0	0	...	...	...	...	0	...	...	0	...	...	...	N	N	...	...	...	...	...	0	...	...
22	+ I	+ I	= I	—	...	—	...	...	0	0	S	...	...	N	+	0	...	—	...	...	+	N	...	+	0	+	±	...	—	—	+
23	+	0 X	0	...	...	0	—	...	0	0	C	...	...	...	+	0	...	0	...	...	—	N	...	=	=	...	—	...	—	...	+
24	+	+ I	0	...	...	...	...	...	0	0	...	...	...	N	0	N	...	...	...	...	—	N	...	0	+	...	0	...	...	+	...
25	+	...	+	...	...	...	...	...	—	0	...	...	...	...	0	N	N	...	...	0	0	N, +	...	...	+	...	0	...	0	...	...
26	+	+	+	0	...	...	...	...	—	0	...	...	...	...	0	...	...	...	...	0	0	N	...	...	+	...	0	...	0	...	...
27	+	+	+	+	...	...	+	...	0	0	...	...	...	...	0	N	—	...	...	0	0	N	...	...	+	...	0	...	0	...	...
28	—	— I	0	...	...	...	...	...	...	0	...	...	...	...	0	=	=	...	...	0	0	=	...	...	=	...	0	...	0	...	...
29	—	0 I	=	—	...	...	...	...	0	0	...	...	...	...	0	...	—	...	...	0	0	=	—	...	+	...	0	...	0	...	...
30	+	+ I	+ I	+	...	...	...	...	0	0	...	...	...	...	0	...	...	0	...	...	...	—	...	...	+	0	...	...	+	...	—
31	...	...	X	...	...	...	...	...	+	8	...	...	...	...	...	N	...	0	...	...	...	N	...	0	+	...	...	...	0	...	...
32	X	X	...	...	...	...	...	...	...	>11	...	...	...	...	...	...	...	...	...	...	...	N	...	0	+	...	...	...	...	...	...
33	X	...	...	...	...	...	...	+	+	0	...	...	...	...	...	...	...	...	...	0	+	N	N	0	±	...	...	...	+	+	...
34	...	...	...	...	...	...	...	...	...	9	...	...	...	...	...	...	...	...	...	0	—	N	...	0	+	...	...	...	...	...	...
35	...	I	...	...	...	...	...	+	+	0	...	...	...	N	0	...	...	0	...	0	=	N	...	0	+	...	+	...	=?	+	...
36	—	...	—	...	...	...	...	...	...	0	...	...	...	...	0	...	...	...	...	...	=	N	...	0	+	...	...	...	...	...	...
37	...	...	...	...	...	...	...	...	+	+	...	...	...	...	...	...	...	0	...	0	0	N	...	...	+	...	...	...	...	...	...
38	...	...	...	...	...	...	...	...	+	+	...	...	...	...	...	...	...	0	...	0	0	N	...	...	+	...	...	...	0	...	...
39	...	...	...	...	...	...	...	...	+	+	...	...	...	...	...	...	...	0	...	0	0	N	...	...	+	...	...	...	...	...	...
40	f +	f +	f ±	..	...	...	+	+	±	13	C	e	...	...	0	...	...	±	—	=	—	+	...	0	±	—	0	...	—	+	+
41	+ I	+ I	+ I	...	...	...	+	—	—	15	S	e	...	...	0	...	...	+	—	=	—	N	...	0	—	0	0	...	0	0	...
42	+ I	+ I	+ I	...	...	...	+	0	—	0	S	e	...	...	0	...	...	+	—	=	—	N	...	0	0	0	0	...	0	+	...
43	+	+	...	...	...	...	...	...	0	0	0	...	...	...	—	...	...	...	...	...	...	...	...	...	0	...	0	...	0	...	...
44	± I	...	—	...	...	...	...	...	0	3	...	...	...	...	0	0	N	—	...	0	0	N	...	...	+	0	0	...	0	...	...
45	...	...	+ I	+	...	...	...	+	+	17	S	...	0	N	0	N	...	+	0	—	—	—	...	+	+	...	...	—	+	+	+
46	f ± I	f + I	± X	+	+	...	...	...	±	14	CS	e r	+	...	0	N	N	+	...	0	0	=	N	=	+	+	+	...	0	+	...
47	± X	± X	+ X	+	±	...	...	+	0	0	S	...	+	...	0	N	N	0	—	...	—	—?	N	0	—	0	+	...	0	...	...
48	± X	± X	+	+	+	...	...	...	0	0	CS	...	+	...	0	N	N	—	—	0	0	—	N	0	0	...	+	...	0	...	...
49	± I	+ I	+ X	+	+	...	...	...	0	0	S	...	+	N	0	N	N	=	...	0	0	—?	N	0	+	+	+	0	0	0	...
50	f +	...	f +	...	...	...	...	...	±	16	CS	e r	+	...	0	N	N	±	...	0	0	=	...	0	±	+	+	...	0	...	+
51	f +	...	± X	...	...	...	...	...	±	>14	CS	e r	+	...	0	N	N	0	—	0	0	—	N	...	+	—	+	...	0	...	...
52	± I	± I	±	+	+	...	...	...	0	0	S	...	—	...	0	N	N	0	...	0	0	—?	N	0	+	0	—	...	0	0	...
53	— I	+ I	+ X	+	+	...	...	...	0	0	CS	...	0	...	0	N	N	...	...	0	0	—	N	0	+	0	+	0	0	0	...
54	±	+	+ X	...	...	...	...	...	+	>15	C	...	—	...	0	N	N	0	—	0	0	—	N	=	+	+	+	...	0	...	—
55	+	...	...	...	...	...	...	0	+	10	...	...	0	...	=	...	...	—	...	+	...	—	—	...	+	...	...	...	...	...	—
56	±	+ I	±	+ I	+ I	0	±	0	0	0	CS	...	...	...	0	...	...	0	...	0	0	N	...	0	+	+	0f+	...	0	...	—
57	± I	...	— I	...	...	...	...	...	...	18	CS	e r	+	...	0	...	...	+	—	—	...	—?	N	0	+	...	+	...	...	0	—
58	+ I	...	0	...	...	...	...	...	...	0	C	e	+	...	0	...	...	0	...	0	0	N	...	...	0	...	...	...	...	...	...
59	+ I	+ I	+ I	+	+	+	...	...	=	0	S	...	...	+ F	0	0	0	=	...	0	0	—	...	...	+	+	...	0	...	0	+
60	+ X	+ X	+	—	—	...	...	...	...	0	S	...	...	+ F	0	...	...	...	...	0	+	+	...	...	— f +	...	0	0	...	...	—
61	+ X	...	± X	...	...	...	...	...	=	21	0	0	...	...	0	0	...	0	...	±	±	—	N	0	+	+	0	...	0	+	+
62	+ X	...	± X	0	0	...	...	...	—	18	0	0	...	...	=	0	...	0	...	0	+	—	N	...	+	+	0	...	0	+	+
63	+ X	0	0	...	...	...	...	...	0	16	0	0	...	...	N	N	...	0	—	...	—	N	...	0	...	...	...	...	0	0	+
64	+ X	X	+	...	...	...	...	...	+	37	...	...	...	...	0	0	...	±	...	—	...	=	=	...	+	+	0	...	+	...	...
65	+ X	X	+	...	...	0	...	...	—	0	0	...	...	N	N	N	N	+	...	...	...	N	...	...	+	...	+	0	—	...	...
66	+ X	X	+	...	...	0	...	...	0	0	0	...	...	N, +	+	+	...	0	...	0	0	N	...	...	+	...	0	0	0	...	...
67	±	...	±	...	...	...	...	...	...	0	...	...	...	...	...	...	...	...	...	0	0	...	N	...	+	...	+	...	...	...	...
68	± X	+ X	+ X	±	±	...	...	+	+	0	CS	...	...	N	+	N	...	0	...	+	+	N	...	...	+	...	+	0	0	+	—
69	+	+ I	+	+	...	...	+	...	—	18	...	e r	+	N	0	N	...	—	—	=	...	—	N	=	+	...	0	0	...	...	—
70	+	+ I	...	—	...	...	...	...	...	0	...	...	—	...	0	—	...	...	...	...	...	...	=	...	+	...	0	...	...	+	...
71	— I	+ I	—	...	...	...	f+	...	0	0	+	...	...	N	0	...	...	...	...	...	—	N	...	...	...	...	0	...	...	...	...
72	...	...	...	...	...	...	...	...	...	0	...	...	...	...	0	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...
73	...	...	...	...	...	...	...	...	...	0	...	...	...	...	0	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...
74	+	+ I	= ?	—	...	...	...	...	—	0	0	...	...	N	0	...	...	0	—	...	+	N	N	...	+	...	0?	...	...	...	...
75	+	I	0	...	...	...	...	...	0	0	...	...	...	N	0	...	...	...	—	0	0	N	...	...	0	...	0	...	...	...	...
76	+	+	+	...	...	...	...	...	...	23	S	...	...	...	0	N	...	...	...	0	0	N	...	...	+	...	...	...	...	...	...
77	+	...	...	...	...	...	...	...	...	...	...	...	...	...	0	...	...	...	...	0	±	N	...	...	...	...	...	...	...	...	...
78	+	...	...	...	...	...	...	...	...	...	...	...	...	...	0	...	...	+	...	0	...	N	...	...	...	...	...	...	...	...	...
79	+	...	...	...	...	...	...	...	...	...	...	...	...	...	0	...	...	...	...	0	...	N	...	...	...	...	...	...	...	...	...
80	+ I	0 X	=	...	...	0	...	0	...	0	...	...	...	...	=	=	=	...	...	0	0	—	...	0	+	0	...	...	0	...	...
81	f +	...	±	...	...	...	...	...	±	<14	0	...	...	=	0	...	...	+	—	0	0	N	...	...	+	+	+	...	0	0	...
82	f + X	...	+	...	...	...	...	...	±	15–16	0	...	...	...	0	N	N	+	...	0	—	—	—	...	+	0	0	...	0	+	...
83	f +	...	+	...	...	...	...	...	±	13	0	...	...	...	0	...	...	0	...	0	0	N	...	...	=	0	—	...	0	0	...
84	+	0 X	+	...	...	...	...	...	0	0	0	...	...	...	0	...	...	...	...	0	—	N	...	...	=	0	0	...	0	0	...
85	f +	...	+	...	...	...	...	...	±	10	CS	...	0	...	N	...	...	+	—	...	+	—	N	...	±	...	+	...	0	+	...
86	+	...	+	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	+	...	...	...	+	...	...	...	0	...	...
87	f +	...	f +	...	...	...	...	...	±	<2	...	...	...	...	...	...	...	+	...	...	...	...	...	...	+	...	+	...	...	...	...
88	±	±	+	...	...	...	...	...	0	<2	0	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...	...
89	+	...	—	...	...	0	...	...	0	0	...	...	...	—	=	...	...	...	...	0	0	N	...	...	0	...	0	...	0	...	...
90	+	...	—	...	...	...	...	...	0	0	...	...	...	N	=	...	...	0	...	0	0	N	...	...	0	...	0	...	0	...	...
91	+ I	...	+	...	...	...	...	...	0	0	...	...	...	—	0	N	N	...	...	...	...	N	...	...	±	0	+	...	0	...	...
92	+	...	...	...	...	...	...	...	0	0	0	...	...	...	...	...	...	+	...	0	0	N	...	...	...	...	=	...	—?	...	...
93	+ I	f + I	+ I	+	...	0	0	+	=	23	CS	e r	+	N	0	±	...	0	—	0	—	—	...	0	+	0	0	0	0	...	...
94	+ I	+ I	± I	...	...	...	+	+	—	0	...	...	0	...	0	...	...	+	...	0	0	N	...	0	+	...	0	...	+	...	...
95	+ I	+	+ I	...	...	...	...	...	—	0	S	e r	...	...	0	...	...	...	—	0	0	...	...	...	0	...	...	...	0	...	...
96	+	+ I	+	...	...	0	...	...	—	0	...	...	...	...	...	...	...	...	...	0	+	...	...	0	0	+	...	+	...	...	—
97	+	...	±	...	...	...	+	...	—	18	...	...	...	...	...	...	...	...	...	...	+	—	=	...	+	+	...	...	—	...	—
98	+ I	...	+	...	...	...	...	...	...	17	...	...	+	...	0	...	0	0	...	0	0	N	...	0	+	...	0	...	=?	...	—
99	+	+ I	+ I	...	...	...	...	...	...	0	C	...	...	N F—G	0	N	N	0	—	0	0	N	...	...	0	...	...	0	0	...	...
100	+	+ I	0	...	...	...	...	...	...	0	...	...	+	N, +	N	...	...	...	...	...	+	...	...	±	...	...	...	...	...	±	...
101	f +	...	+	...	...	...	...	+	±	+	CS	e r	±	—F ± G	0	N	...	...	+	...	+	...	...	0	±	...	...	0	+	...	+
102	f +	...	+	...	...	+	...	+	±	>16	CS	e r	±	...	0	...	...	+	+	...	+	=	...	±	+	0	+	0	±	+	+
103	± I	+ I	+	...	...	+	...	0	0	0	S	...	0	—F —G	0	N	...	...	...	...	+	=	...	=	...	...	+	0	...	...	+
104	+	...	—	...	...	...	...	...	...	0	S	...	...	N	0	N	...	...	...	0	0	...	...	0	...	...	...	...	...	...	+
105	+ I	+ I	+ I	+	...	0	...	+	0	0	S	e r	...	...	...	+	...	...	...	0	0	—	...	0	+	...	...	...	—	+	...
106	+ I	+ I	+ I	+	...	0	...	+	0	+	S	e r	...	...	...	+	...	...	...	0	0	—	...	0	+	...	...	...	—	...	...
107	+	± I	+	...	...	0	0	+	=?	0	S	e l	+	...	0	...	...	0	+	0	0	N	N	f—	+	—	—	0	0	...	...
108	+ I	I	0	...	...	...	...	...	...	0	...	...	...	...	—	...	...	...	...	0	0	...	...	...	...	...	...	...	...	...	...
109	+	...	+	+	...	...	...	...	...	>26	S	...	...	...	0	N	N	—	...	...	—	N	...	...	+	...	...	0	...	+	...
110	+	...	+	...	...	...	...	—	...	30	...	...	...	...	...	...	...	...	...	...	...	—	...	...	+	...	+	...	...	...	...
111	+ I	+	+	+	...	+	...	0	...	0	...	...	0	...	N, +	...	...	0	...	0	0	N f —	...	...	+	...	+	...	—?	0	+
112	+ I	...	+ I	+	+	...	...	...	0	21	CS	v l	...	N	0	...	—	0	...	0	±	N	—	...	+	0	+	...	0	=	...
113	+ I	± I	+	+	+	...	...	...	—	0	CS	e r	+	r d	0	N	N	...	...	0	...	N	—	...	+	0	—	0	0	...	...
114	+	+ I	+	+	+	...	...	+	0	0	C	e	+	r d	0	N	—	0	...	...	+	N	N	+	+	—	—	0	0	...	—
115	+	±	+	+	+	...	...	...	+	8–9	S	e	±	r d	0	N	N	+	...	0	0	N	=	...	+	0	—	0	0	...	...
116	± I	± I	+ X	+	+	=	—	...	0	0	S	...	...	N	0	N	N	0	...	=	—	—	N	...	+	0	+	0	0	0	—
117	±	+ I	+	...	...	...	...	...	=	0	0	...	0	N	0	N	...	0	...	0	0	N	=	0	0	+	0	0	0	0	—
118	+ I	+ I	+ I	+	+	+	...	...	+	0	...	...	...	...	0	+	...	...	...	...	—	N	...	...	+	+	+	...	0	+	...
119	+	+ I	+	...	...	...	...	...	0	0	...	...	...	N F	0	N	N	0	...	—	...	=	N	+	+	...	+	0	0	+	—
120	+	+ I	+	...	...	...	...	...	...	0	...	...	...	...	0	...	...	...	...	0	0	—	...	...	+	...	0	...	0	...	...
121	0	0 I	...	...	...	...	—	...	0	0	...	...	...	...	0	...	...	0	...	...	...	N	...	...	0	...	0	...	0	...	...
122	± X	...	± X	+	...	...	...	...	0	18	...	e	...	N	0	...	...	0	...	0	0	N	...	...	+	...	+	...	...	...	...
123	f +	...	= f +	...	...	...	...	...	±	20	...	...	...	...	0	0	...	0	...	...	...	=	...	...	+	...	+	...	...	...	—
124	+	+ I	— X	...	...	...	...	...	0	0	...	...	...	—	0	+	+	0	...	...	...	N	N	...	0	...	+	...	0	...	—
125	+	+	+	...	...	...	...	...	0	0	...	...	...	...	0	...	...	0	...	...	...	N	...	...	0	...	+	...	0	...	...
126	+ I	...	+ I	+	...	0	0	0	...	14	S	c v	+	N	0	0	N	+	...	0	0	—	...	...	+	+	0	0	0	0f ±	...
127	+	+ I	+ I	+	...	...	+	...	0	0	...	...	...	...	0	...	...	0	...	0	0	N	...	...	+	...	+	...	...	...	...
128	+	...	+	...	...	...	...	...	+	+	...	...	0	...	0	...	...	+	...	0	0	N	...	...	+	0	0	...	—?	...	...
129	+	0 I	+	...	...	...	...	...	+	0	CS	...	—	...	0	...	...	—	...	0	0	N	...	...	0	...	0	0	0	...	...
130	+	0 I	+	...	...	...	...	...	...	0	C	...	0	...	=	...	...	0	...	0	0	N	...	...	0	0	0	0	0	...	...
131	± X	± X	—	...	...	...	...	...	=	0	...	...	...	...	0	...	...	0	...	0	0	N	...	...	+	+	+	0	0	...	...
132	+	+ I	f =	+	...	...	...	...	0	0	...	0	...	N F	0	N	...	0	...	...	+	—	...	—	—	...	+	0	...	...	—
133	f +	...	±	...	...	—	...	...	+	28–29	0	0	...	N F	0	0	...	+	...	...	+	N	N	...	+	0	+	...	...	...	+
134	±	± I	—	—	...	...	...	...	...	0	CS	v r	...	N F	0	...	...	0	...	...	=	N	N	...	=	...	=	0	0	...	...
135	+ I	+ I	+ I	+	+	...	...	...	0	0	S	0	...	...	0	N	...	...	...	0	0	—	...	+	+	—	0	...	0	...	...
136	+ I	+ I	+	0	...	...	...	...	—	0	S	c v	—	—	0	+	...	...	...	...	...	+	N	...	+	...	...	0	0	—	...
137	+	+ X	+ X	+	...	...	+	...	0	0	...	...	...	...	N	...	...	0	...	0	0	N	...	0	+	0	+	...	0	0	...
138	+	+ X	+ X	+	...	...	+	...	0	0	...	...	...	...	+	...	...	0	...	0	0	N	...	0	+	0	—	...	0	0	...
139	+	+	+ X	±	...	...	+	...	0	10	S	e r	...	...	N	...	...	—	...	0	0	N	...	0	+	0	—	...	0	0	...
140	+	+ I	+	+	...	...	0	0	0	0	0	0	...	...	=	...	...	0	...	0	0	N	...	0	+	...	0	...	0	...	...
141	f +	...	± X	+	...	0	0	...	±	10–11	S	el	0	...	0	...	...	0	...	0	+	N	...	—	+	0	—	0	0	...	...
142	+	...	+ X	—	...	0	0	+	+	>18	S	...	0	...	0	...	...	+	—	0	0	—	=	0	—	0	—	0	0	...	...
143	f +	...	± X	+	...	0	0	+	±	16	S	e l	+	...	0	...	...	0	+	0	0	—	...	0	+	—	—	0	0	...	...

EXPLANATION OF SIGNS USED IN TABLE I.

- \+ increased, or marked.
- ± much increased, or very marked.
- — diminished, or slight.
- = much diminished, or very slight.
- 0 none, or absent.
- ? not clearly stated, or the author himself is in doubt.
- , or.
- \> [as >12] after or more than the age of.
- < [as <12] before or less than the age of.
- * died at the age mentioned.
- a upper extremities.
- B bilious fever.
- C cyphosis [or lordosis].
- cv cavus.
- D diphtheria.
- e equinus.
- el equino-valgus.
- er equino-varus.
- F faradic.
- f that the condition indicated by the following sign was formerly present.
- G galvanic.
- I increased on closing the eyes.
- in infancy.
- l lower extremities.
- M measles.
- Ml malaria.
- Mn meningitis.
- My myelitis.
- N normal.
- P parturition.
- Ps pertussis.
- R rheumatism.
- r d reaction of degeneration.
- S scoliosis.
- s speech.
- Sc scarlatina.
- T typhoid fever.
- V variola.
- vl valgus.
- vr varus.
- X not increased on closing the eyes.

tion by European writers, though tabulated by Ormerod and Vizioli. Careful reading "between the lines" of the author's report renders it probable that they are genuine, as he claims. 37–39 are also most incompletely described, but information received from Dr. Coleman through Dr. Smith renders it at least possible that they are instances of this disease, and I feel therefore obliged to place them in the table. There certainly appears to be no other affection to which they can be so well assigned. Jakubowitsch's case (55) is by no means typical, and is excluded by Bury; yet it seems to accord with the symptom-complex of Friedreich's ataxia better than with any other disease. Erlenmeyer's case (56) most probably belongs here, and Rütimeyer is of the same opinion, though Friedreich and Erb were opposed (see "choreiform movements"). The girl (60) described by Teissier might be excluded from the list on account of the improvement in the symptoms, had not her brother (59) been a well-marked case of the disease. 68 is a very anomalous case and might possibly be disseminated sclerosis; but the distinct hereditary and family history strengthen Botkin's claim that it was Friedreich's ataxia. He believes that accidental focal lesions may also have existed. The age of the patient and some of the symptoms lead Gowers to relegate 100 to ataxic paraplegia. Considerable correspondence with Dr. Smith as to the course of the case since it was reported, leads me to place it in the table. 108 is a very doubtful case, but not sufficiently so to nullify Fellows' claim that it is an instance of Friedreich's ataxic. The report is most incomplete. 109 (McAlister's) is a case included in this class by Bury, but of which an exceedingly imperfect account is pub-

lished. Descroizilles' patient (111) had some symptoms much like disseminated sclerosis, though it very probably belongs to Friedreich's ataxia. Bury hesitates to call 2 of his own cases (114–115) instances of this affection, on account of the absence of a history of heredity. They seem in other respects to be good examples of it. Erlicki's and Rybalkin's patient (117) has already been referred to under "Pathological Anatomy."

TABLE II.—ADDITIONAL LIST, ARRANGED CHRONOLOGICALLY.

A. *Reputed cases, related by blood to those in Table I., but not under professional observation.*

Reporter.	No.	Relationship to cases in Table I.
Carre,	17	Grandmother, mother, 8 uncles and aunts, 6 brothers and sisters and a cousin said to have similar symptoms.
Bradbury,	2	Mother and brother similarly affected.
Carpenter,	1	A third member of the family later became ataxic.
Kellogg,	3+	A brother, aged 5–6, and several cousins ataxic.
Dreschfeld,	2	A brother attacked at 19, and a sister at 21 years of age.
Seeligmüller,	1	A sister, aged 21, possibly affected.
Brousse,	1	The mother had the same disease.
Power,	2	A brother attacked at 15, and a sister at 17.
Rütimeyer,	3	Great-great-great-grandfather said to have been ataxic; 2 brothers of Case *54* showed the first symptoms in their 4th year.
Musso,	3	Possibly the great-uncle had the same disease; 2 brothers of Case *64* were certainly affected.
Botkin,	2	Father and a brother said to have had similar symptoms.
Ormerod,	2	Grandfather of Cases *69–73* reputed ataxic. Brother of Cases *74–75* attacked at age of 13
Fowler,	1	Father said to have had a peculiar gait; possibly the same disease.
Fazio,	2	2 brothers with the same disease.
Vizioli,	1	A sister of Case *91* died at 5 years of age with all the symptoms of Friedreich's ataxia.
Seguin,	1	A sister attacked at 15 years of age.
Smith,	1	A sister attacked at 6–7 years of age.
Fellows,	2+	Several members of the family are said to be similarly affected.
Bury,	1	Grandfather of Cases *112–113* probably had an ataxic gait, and may have had the same disease.
Stintzing,	1	A brother died at 31 with the same disease from which the others of the family suffer.
Mastin,	3	2 uncles attacked at 15 and 12–14; and a cousin at 9 years of age.
Mendel,	1	A brother died at 4½ with symptoms of Friedreich's ataxia.
Shattuck,	2	A brother attacked at 7, and a sister at 5–6 years of age.
Joffroy,	1	A sister affected at 12.
Griffith,	1	A brother of Case *143* died at 9 with symptoms of the disease.
	57+	

B. *Possible and probable cases; under observation and reported, but without description.*

Reporter.	No.	
Eulenburg,	3	Mentions a case of "locomotor ataxia" under 10 years of age, and 2 under 20; both illustrating heredity. Are very possibly instances of Friedreich's ataxia, as the author does not distinguish this affection from tabes.
Duchenne,	3	Says he saw with Trousseau 3 brothers attacked by ataxia.
Vulpian,	?	Says he has seen cases analogous to those mentioned by Duchenne.
Schultze,	1	Reported a case briefly before the Wanderversammlung der Südwestdeutschen Neurolog. u. Irrenärzte; but I can find no description of it.
Putnam,	2	Saw 2 separate cases which he believes to be of this type, though no others of the family were affected.
Goodhardt,	2	Saw an ataxic child with an ataxic father. No detailed description. They may be instances of Friedreich's ataxia.
Ormerod,	4	2 cases of this disease, brothers, were shown him by Dr. Hughlings Jackson. Also saw a sporadic case under the care of Dr. Jackson, and another under that of Dr. Gee.

C. *Too doubtful cases; claimed as cases by those reporting them, or by others, but probably without good reason.*

Reporter.	No.	
Bouchut,	2	A boy of 11 and a girl of 14 are reported by the author as examples of locomotor ataxia. They are occasionally supposed to be Friedreich's ataxia, but are very properly rejected by nearly every writer on the disease.
Hitzig,	3	The author rather intimates that these cases belong to progressive muscular atrophy. Kahler and Pick suggest that the disease may have been Friedreich's ataxia, but it does not seem clear on what grounds, except that the mother was tabetic.
Fellows,	1	A case described as Friedreich's ataxia, but without, I think, any claim to be considered an instance of it.

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