

## ON HEREDITARY ATAXY, WITH A SERIES OF TWENTY-ONE CASES.

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SINCE Friedreich first described Hereditary Ataxy in 1861, many cases (in all less than a hundred) and series of cases have been reported, which have been supposed to belong to this disease; but in some instances the reports have been so meagre, and in others there has been such wide variations in the symptoms present, that it has so far been well nigh impossible, from the literature of the subject, to obtain a definite idea of the conditions which must be present in any given case, in order to entitle it to a place under this heading.

The establishment of a reliable and permanent criterial picture to be used for purposes of comparison will necessarily require many years for its completion, more especially in a disease so rare as this one. The establishment of such a standard, however, worked out from a critical scrutiny of all available cases, is of the utmost importance, even if it have to undergo considerable amendment from time to time, as the stock of definite facts accumulates.

The work of Dr. P. Ladame in this direction (BRAIN, vol. xii. p. 467) is highly commendable; and to it I would refer anyone who wishes to acquaint himself with a brief historical review of the subject, as well as with the views that many distinguished neurologists have held, and still hold, regarding the disease.

After a critical review of all available published cases, and an elaborate study of a case for some time under his

own observation, which he accepts as typical. Dr. Ladame proceeds to erect a criterial standard which he summarizes as follows :

"Slow and progressive ataxy of the four limbs, usually attacking several children of the same family, dating very often from very early age—commencing in the legs, extending gradually to the trunk and arms, the muscles of the larynx, those of the tongue and eyes; weakness of the legs, increasingly difficult gait; choreiform unsteadiness; static ataxy; difficulty of articulation; nystagmus; spinal curvature; paralytic club-foot; abolition of knee-reflexes; no sensory disturbances; absence of oculo-pupillary anomalies and of lightning pains; integrity of sphincters."

Further on Dr. Ladame states that there are no authentic cases known of onset after the twentieth year, that there is integrity of the special sense organs, and that horizontal nystagmus, static and dynamic, is rarely absent. He also speaks of scoliosis and scanned speech as common.

If the cases herein reported are accepted as instances of this disease, then the criteria as set forth in the above summary should be considerably modified. And the same may be said of the descriptions found in the text books or elsewhere, so far as my reading has extended.

CASE XVIII.—A business man, single, of temperate habits, with good family history, excepting that his mother became ataxic at about thirty-three, the disease progressing steadily, till she died of tubercular diarrhoea at forty-eight. Her mother and maternal grandmother were similarly affected, but I will not proceed farther with a verbal description of the heredity, as this may be seen by reference to the accompanying chart.

Patient was always active and vigorous in every way until attacked by his present disease, being rather among the foremost in all athletic sports and school work.

His attention was first attracted to this disease while he was working with a surveying party in Texas, and then it was noticed by others before he himself noticed it. He was then twenty years old, and the ataxia was so marked as manifested by a staggering gait, that his chief thought him intoxicated. But now he recollects that at least two years before this, or when he was eighteen years old, the draughtsmen in the office where he worked complained that he rendered their desks or drawing-boards so

unsteady when he leaned against them that they could not well go on with their work ; and he further distinctly remembers that on a certain occasion at about this time, when in company with other young men in the country, he was quite unable to read the large letters of an advertisement at a considerable distance, though each of his companions could read them with ease. He feels confident that this comparative visual defect had not always existed. I should say here that this patient has a good English education and that his intellectual capacity is distinctly above the average, so that, notwithstanding his affliction, he would at the present time be justly regarded as a well-informed man ; and in this same connection I wish to say further that I can hardly find words in which to properly express my admiration for the high humanitarian spirit and untiring energy which this gentleman has all along displayed, in assisting me to present to our profession all facts that might throw any light upon this obstinate disease.

He feels quite certain, too, and in this he is corroborated by his older relatives who have had an opportunity of observing him carefully, that at the age of fourteen, when his voice underwent the change incident to puberty, that there appeared a gradually increasing defect in utterance. His speech was slower and his syllables less definite and distinct than formerly. The ataxia has always been much more pronounced if he was fatigued, and he now remembers that at the age of eighteen he could not walk in a straight line when very much tired.

There has been some progressive loss of power in the legs from the first, but this has been insignificant throughout in comparison with the ataxia. The ataxia was first noticed in the legs and has progressed more rapidly in their muscles than in other situations, but it has been distinctly noticeable in the arms from a very early period, and has certainly progressed. An increase of the knee-jerk was probably an early symptom, because it has been so in all the other members of the series whom I have examined (in one, a younger brother of the patient, the disease is just now commencing to develop) ; and because the patient has shown me notes of his case made four years ago by such competent observers as Dr. Henry M. Lyman and Dr. Walter Hay. The former physician made, I believe, the diagnosis of ataxic paraplegia, which, at the time of his examination, and without the more complete family history since elaborated, was probably more nearly the proper nosological position of the disease than any other according to the standard text-books ; and even now I

think hereditary ataxia paraplegia is a designation that comes pretty near the mark in assigning to this series of cases a place in accordance with the usual classification of degenerative diseases of the nervous system.

At no time has there been any pain or any other disturbance of sensation. There has been no atrophy, spasm, or trophic manifestations. The sphincters have not been affected, and the patient thinks sexual power has not been more impaired than can be attributed to the general decline in bodily weight and strength, which has slowly supervened within the past six years, the weight having fallen from 136 to 112 lbs. in that period.

I should have stated that a tendency to choking while eating has all along been a very troublesome symptom, and is in some way (whether mainly from inco-ordination or paralysis, I can't say) due to the tendency of food to come into contact with the larynx; and this has been a troublesome symptom in several members of the series.

For several years past vision has progressively failed, so that he could read best in a dim light.

*Present Condition.*—Patient is considerably emaciated, though he eats fairly and sleeps well. He is of medium size and well formed. The sensibility is normal, the knee-jerk is greatly exaggerated and equal on both sides; there is a slight ankle-clonus, the skin reflexes appear in the main normal, but the cremasteric and abdominal are not strongly pronounced. There is marked ataxia in nearly all voluntary movements; so that the patient can only walk when supported by an attendant, and can only stand when leaning against some solid support. Closure of the eyes does not materially increase his difficulty. The gait is such as would usually be described as cerebellar, the patient leans rather backwards against his attendant and sways from side to side, and he has an uncomfortable sense of insecurity all the time, as if his head must fall violently to the ground. He experiences a distinct loss of power in the legs, which he thinks is even greater than could be accounted for by his general decline in bodily vigour, but the muscles are firm and well developed. All the voluntary muscular movements are slowly performed, and of this the patient is quite conscious. He cannot reach out his hand suddenly to seize any given object. The hand moves slowly and deviates several inches in various directions from the direct line that would normally be taken in such an effort. This tardiness of movement is plainly noticed when the patient raises the eyes; on being addressed or in winking, the eyelids will rise so slowly

as to be suggestive of temporary ptosis, but in the end they are raised too high, so that the sclerotic is often so much exposed as to display an expression, usually associated with some intense emotion, when in fact the patient is suffering from no emotional disturbance whatever. Though ataxia in the muscles that move the tongue would be difficult of conclusive demonstration, because the normal movements could hardly be definitely described (and the same might be said of the muscles that move the lips), yet a careful observer would see at a glance that the movement of both tongue and lips, especially the former in this case (because a full beard is worn), were far wider in range than usual. There is no difficulty in swallowing, excepting the tendency to choking already noticed, which might be explained by ataxia and tardiness of the muscles concerned, because no extraordinary effort has to be made, and there is no tendency for liquids to pass out through the nose.

There is marked inco-ordination of the various muscles of facial expression, which is easily observed when the changes are going on incident to the discussion of an absorbing topic, the action being more marked, now in this group of muscles and now in that, so that people casually meeting the patient frequently get the impression that there is some mental defect.

There is no spontaneous movement during sleep or waking repose, but a comparatively slight voluntary movement gives rise to very extensive and peculiar movements in muscles far removed from those required for the execution of the required act. For instance, where the patient puts out his hand to take a book from a table beside which he may be sitting, the whole upper part of the body goes through a series of irregular movements highly suggestive of chorea. This is particularly the case with the head, which is somewhat inclined forward, moved from side to side, and the chin is protruded; and there is often an associated movement of the other hand. In none of these movements is there ever anything approaching a jerk.

This case and five others of the series was thoroughly examined by Dr. W. F. Montgomery, Professor of Ophthalmology in the Women's Medical College, whose reports I herewith append. And two of the less advanced cases (19 and 20) were minutely examined by Dr. Casey Wood, Professor of Ophthalmology in the Postgraduate Medical School in this city.

Patient states that for ten years past he has suffered from some gradual impairment of vision, and has had double vision at times, of not more than a few days' duration, but not for several

years past. Examination shows marked ptosis when the patient is at rest, but by an effort he can raise the lids, showing sclerotic above the cornea when the eyes are directed in a horizontal plane. There is perfect co-ordination of the ocular muscles except to extreme right, where there is slight lagging of external rectus—not enough, however, for the production of diplopia; the lids and conjunctivæ are normal.

The pupils respond to light but very slowly; and the same is true of accommodation. Dilatation also occurs slowly on stimulation of the skin of the neck.

Vision is 20/200 in either eye. Snellen No. 5 *can* be read, though with difficulty, at 8 inches in an ordinary light, much more easily in a dim light.

The ophthalmoscope shows decided blanching of the optic discs and lessening of the calibre of the arteries, with slight but distinct atrophic changes in the retina.

There are only slight peripheral limitations of the field, and almost complete colour blindness, red only being distinguished with any degree of certainty.

The above description fairly represents the patient's condition, May 2nd, 1891, when I first examined him, and there was no material change until about October 15th, except that under tonic treatment with galvanism—20 to 60 m.a.—to the spine for ten minutes two or three times a week, there was a considerable improvement in the general health with a gain of about 10 lbs. in weight. At the last-named date the patient was attacked with a severe cold, and in a day or two became unable to walk even with assistance, on account of loss of power in the legs, without any alteration of sensation or the reflexes. There was some cough, but the appetite remained good. To-day (November 13th) there is considerable improvement in the legs, but the power is much less than before this attack.

CASE XIX.—Sister of the preceding, first consulted me May 5th. She is twenty-six and has been subject to headaches with nausea and vomiting since childhood; she first menstruated at eighteen, but this function has been too infrequent since, often occurring at intervals of from two to four months. Ataxia first showed itself in the leg when she was twenty, but had not invaded the arm, the organs of speech or the ocular muscles to any considerable degree till she was twenty-three. She then began to notice that vision was indistinct in a bright light, and she would move back from the window when reading. The ataxia had advanced so far in the legs that she was obliged to give up dancing.

*Present Condition.*—Patient is well formed, and of medium size; she has intellectual faculties above the average and a happy and cheerful disposition. She is in a rather reduced state of general health, as manifested by numerous very painful boils about the hips and thighs, which have been appearing in successive crops for the last two or three months.

The cutaneous and muscular reflexes are all distinctly exaggerated, especially the knee-jerk.

There is not and has not been any disturbance of sensation or of the sphincters. She walks well without assistance of an even surface, but deviates considerably from a straight line, no matter how hard she attempts to prevent it. Closure of the eyes does not materially increase the tendency to fall. The same ataxy of the movements of the hands, eyelids and face are present, as in the preceding case, and the same tendency to extensive associated movements, alike in quality but less in degree. There has been no tendency to choking.

The oculists report slight static ptosis with inco-ordinate over-action of the levators. All the movements of the ocular muscles and all the pupillary reflexes are sluggish. All are present, however, and in no case can nystagmus be produced. Vision is 20/80 and Snellen No. 3 is read well enough at 10 inches. Patient reads much better in a dim light. The average expert would pronounce the fundus normal without knowing anything of the condition of vision.

The colour sense is not much affected and there is some peripheral limitation of the field of vision.

The patient states that she is conscious of some difficulty of articulation, but there is no markedly positive defect.

It should be stated that this patient, the only girl in a family of five children, was, before the onset of this disease, very active physically and accustomed to join in the sports and exercises practised by her brothers; and indeed, all the members of this family were very active, so that in athletic contests at school they very frequently carried off the prizes. For this reason it is easier to determine the date of onset of ataxic symptoms than it might be under other circumstances. With the application of the galvanic current twice a week, the occasional exhibition of fractional grain doses of calomel, six week courses of Fowler's solution carried up to six drop doses after meals, and several short courses of simple bitters with port wine, the boils, the headaches and the irregular and painful menstruation have entirely disappeared, and the special symptoms of the disease have certainly not increased.

CASE XX.—Brother of the two former, æt. twenty-four, well developed, cheerful with correct habits, good business man. The disease began, without exciting cause, at age of eighteen. The unsteadiness of gait and difficulty of articulation came on together, and not until two years later was a defect of vision realized. He has carried a cane for the last few months, but can walk fairly well without one, though he deviates considerably from a straight line in walking. The defect in speech is marked, the labials are often defective, yet he whistles without much difficulty. Occasionally the labials are exploded.

The ocular condition is essentially identical with that of the preceding case according to the oculist's report and my own observation, excepting that ptosis is considerably more marked in the left than the right eye.

There appears to be a general relaxation of the facial muscles in repose and even during commonplace conversation, giving the patient a heavy and stolid expression which is quite at variance with the prevailing state of his mind.

He has had galvanism to the spine, as No. 18, and thinks there has been distinct failure in the power of walking in the past three months.

CASE XXI.—There are two other children in the family, twins æt. twenty-one, in one of whom there is distinct exaggeration of the cutaneous reflexes and slowness of speech, but no other indication of the approach of the disease. The remaining twin is entirely free, up to the present time.

In the next family three cases have occurred.

CASE IX.—Is a farmer's wife, aged sixty-seven, of regular habits, who has borne three children. The disease first showed itself at the age of about thirty-five, by ataxy in the gait, and in the hands a year or two later. At the age of about fifty she could walk only with assistance, and at fifty-eight could not pour tea. For several years, at about this time, she went about the house on hands and knees, but for the past four years has been entirely helpless.

Failure in articulation and vision in right eye began to be manifested at about fifty-seven and advanced very rapidly. Vision with the eye first affected was nearly gone in three years, when it began to fail in the other, about the same course being run. No defect of sensation or the sphincters. Memory has failed much within the past four or five years.

*Present Condition.*—Permanent spastic contraction of the legs, the three being flexed at about a right angle, which can be partly



overcome by force slowly and steadily applied. When the patient attempts to talk the tongue appears to move in every conceivable position without being protruded; the face undergoes various inco-ordinate movements, the head is repeatedly bent forward and moved from side to side; there is protrusion of the chin, and the arms were flexed and moved forward and backward. The movements are highly suggestive of chorea but less rapid. In articulation there is a marked dwelling on the vowel sounds. When I briefly explained the nature of my errand, the patient partly succeeded in saying she was glad to see me, said she had no pain when I forcibly extended her legs, and correctly identified the parts touched when sensibility was being tested. But she said she had only been blind about two months, showing her great failure of memory.

*Oculist's Report.*—Husband states that patient used to be a great reader and was able to read good print till about nine years ago. At present she can only just distinguish a bright light.

There is almost complete ptosis. If the lids are held open, as is necessary in making an ophthalmoscopic examination, they remain open for a short time and then slowly close. There is scarcely any action of the external ocular muscles, the eyes staring directly forward when the lids are raised. Pupils of medium size and not responsive to light; media clear. The discs present a typical picture of complete atrophy of the optic nerve.

CASE X.—Son of No. 9; farmer, age thirty-eight, single, temperate. First noticed disease in legs at age of thirty-one, and in his hands about two years later. Defects in vision and articulation were first noticed at the age of thirty-five, at which time he began to use a cane. Has occasionally had shooting pains in the legs for the past two years, not very severe, however; for the past few months has not been able to retain his urine as usual, and has had to urinate much more frequently than before, having to make great haste to prevent an accident.

Excepting that he still walks with a cane, and that there is more exaggeration of the reflexes, there being well marked foot-clonus on both sides, the objective symptoms in this case are identical with those already described at length as belonging to Case No. 18. There is no alteration in sensations, as might be suggested by the pain and the bladder symptoms. The oculist's report is also practically identical with that of Case No. 18. The general health of this patient was good, though he said he had lost a few pounds in weight.

CASE XI.—Brother of No. 10; died of exhaustion of this disease at age of twenty-six. First symptoms appeared at age of sixteen, as simultaneous ataxy of the legs and eye muscles. He walked with a cane at nineteen, and was helpless at twenty-one. Voice and vision were both somewhat affected at seventeen, but not very markedly so at the time of his death. There was no defect of sensation nor of sphincter action.

Though I had already obtained a pretty full history of the next three cases from their near relatives, I happened to learn that so competent an observer as Dr. Norman Bridge of this city was temporarily living very near them (about 2,000 miles from here), and he examined them and kindly sent me the following notes :

CASE V.—“Patient is sixty-seven years of age. She has a shrunk, cadaveric body, her muscles having undergone extensive atrophic change; the head is in almost constant motion, much like that produced by chorea; this is especially the case when she is talking, moving her hands, or conscious of being observed; the only difference is that the head movements are less regular than in localized chorea. When lying or sitting she can move her legs and even kick, but these voluntary movements are performed with some ataxic irregularity. She is utterly unable to rise from a chair, and has been so for the past year. She moves her hands and arms about freely, but with the shaking, irregular movements peculiar to the ataxic. She cannot, with her eyes closed, accurately touch her nose nor any other desired point on her body. The voice has the same irregularity as the hand movements; it is not a tremor, but a shaking; she does not hesitate in her enunciation, but, so to say, shakes her words out with poor but not abolished labial and palatal action.

“There is no contraction anywhere. The knee-jerk is considerably exaggerated, but there is no foot-clonus. The reflexes are alike on each side. Nystagmus is absent, but the eyesight is so much impaired that she can only see the spots on playing cards (she plays solitaire much of her time): I tried to make an ophthalmoscopic examination, but under great difficulties, so that I do not feel able to say with any degree of positiveness what the condition of the fundus is. Vision is the same in each eye.

“There is no constipation nor vesical symptoms of any sort. Attempts to swallow fluids often provoke coughing and strangling, doubtless from their entrance into the trachea to some extent. She has learned to swallow with great care and deliberation. Her appetite is poor, tongue clean; she insists that her soft

palate is too low, and it is, in fact, relaxed; she insists, also, that she has intense heat in her back, but to my touch it is normal. Pulse 112. The pupils are only very slightly responsive to light."

I have given Dr. Bridge's notes in detail because I thought it important to have an entirely independent statement from so high a source. I will add that the first symptoms in this case appeared, at thirty-five, in the legs, and has progressed quite steadily down to the present condition. There has been no pain or sensory disturbance, unless the intense heat in the back be so regarded.

CASE XV.—Son of. No. 5; aged forty-nine, also examined by Dr. Norman Bridge, whose notes I quote: "He walks with an ataxic gait, and his upper extremities show to all tests the same condition as the lower. Patient states that the disease first appeared in the lower extremities four years ago, and in the upper a year or two later; says that the uncertainty of gait is much greater sometimes than at others; walks with a cane, and sometimes has great difficulty in rising from his seat. The knee-jerk is enormously exaggerated, and there is a typical ankle-clonus, these conditions being alike on both sides. There is no nystagmus, no contractions, and no bowel nor bladder symptoms.

"The sight of the left eye is abolished, and has been so for the last four or five years. It has slowly declined since he was serving in the army at the age of about twenty. The sight of the right eye is much impaired, but he can read with a good lens, and says that the print then looks correct to him. Careless swallowing of fluids is apt to carry some into trachea and set up coughing. His voice is like that of his mother's, only in a less degree; he says that his voice was normal until a year ago. He has lost 22 lbs. in the last three or four years. Pulse (sitting) is 104. His face has a congenital twist, it is noticeably longer on the left side. He is apathetic. He denies ever having had any pains in the back or extremities."

I will add that there have been no sensory symptoms, and that acute observers among his relatives have assured me that positive signs of ataxia in the gait were present at forty.

CASE XVII.—Female, sister of the preceding, was first affected at fourteen and died from exhaustion, extremely emaciated, at twenty-eight. The disease developed rapidly in her case, and early in its course she had severe pains in the hips and legs, and marked paralytic constipation. The voice and vision were much involved.

CASE XVI.—Male, aged forty-two, son of No. 5, brother of 15 & 17, has shown some ataxy for the past two or three years; quite obtrusive when tired, but not otherwise. Ataxia of gait the only symptom.

CASE VI.—Female, aged sixty-three. First symptoms at forty-five, consisting of ataxy of legs. The course of the disease has been slow. She can still walk on an even surface without assistance, but has to go very slowly and sways about considerably, usually steadying herself by holding on to furniture. The hands are certainly involved, but not greatly, as she can still do needlework and pour tea. Vision is considerably affected, but she can read coarse print in a dim light. Cannot read in a bright light. Speech has become very slow, but is quite distinct. General health fairly good. Other symptoms negative. History given by a relative, himself affected, who has seen her almost daily for years.

CASE VII.—Female, died at forty-six of tubercular diarrhœa. First affected, at twenty-eight, with slight ataxy of hands; a few years later it was noticed in the legs, but it progressed very slowly till she was thirty-five when, after child-birth, it advanced more rapidly. With the exception of the last six months, when she became greatly emaciated from chronic diarrhœa, she could walk alone, but would deviate several feet to either side of her line of direction. Until the last two years of life was able to pour tea. Her voice gradually became very slow and somewhat indistinct, but this did not show itself till about the fortieth year and progressed very slowly.

Vision began to be impaired at thirty-six, and by the time she was forty she could only read very large print in dim light; after forty vision failed more rapidly. Other symptoms negative.

CASE VIII.—Female, died at thirty-two, of consumption, only slightly affected. Two of her children are living, aged respectively twenty-two and twenty-five, and are healthy.

CASE II.—Female, died at about sixty-five. The disease was well developed in all its usual features at fifty, and for the last fifteen years of life the gait showed cerebellar divergence, if I may use that term, to almost the same extent as obtained in No. 7. This statement is the result of a conference among her surviving children and other near relatives, and though I did not attempt to extract from them evidence of earlier symptoms, it is pretty safe to say, judging from the other cases, that they existed at forty.

CASE IV.—Male, died at about forty; I could not learn the

cause of death. He was only slightly affected for two or three years preceding death. No further particulars available.

CASE XII.—Female, died of pneumonia at thirty-one. Married at twenty-three and had four children. First symptoms appeared at about twenty, consisting of ataxy in the hands and arms, three or four years later the legs became affected. The disease progressed quite rapidly in her case. Was unable to walk without assistance at the age of twenty-eight. No pain at any time. Mind impaired during the last two or three years of life—hebetude mainly.

CASE XIII.—Female, aged twenty. First symptoms noticed consisted of ataxy in legs, beginning at fourteen. The arms were involved in a few months, and vision and voice failed rapidly. The voice is described as slow, harsh, and indistinct. There have been darting pains in the legs, but she still walks with a cane. This and the preceding patient live 1000 miles away, and the above incomplete history was obtained by correspondence carried on by a near relative. I hope later to get a more exact report.

The same correspondent informs me that he has learned from a reliable source that No. 12 has a younger child distinctly affected, but has no further particulars.

CASE III.—Male, died at fifty-five of pneumonia. First affected at forty, after a fall; was able to walk with a cane at time of death. Arms, voice and vision distinctly, but only slightly, affected.

CASE XIV.—Male; died of phthisis at sixteen; affected at eleven; the symptoms began in legs after a fall; he was never able to walk afterwards. Later they appeared in the arms. I have no information as to voice and vision.

CASE I.—Female, died at seventy-two. The first symptoms appeared at forty-five, after the patient had sustained a fracture of the hip by a fall. They consisted in marked ataxia in the hands; after injury she was never able to walk without assistance. No more definite or satisfactory account of the symptoms can be obtained. Various relatives who remember her well, and have since seen numerous and well marked instances of this disease, are certain that she was affected with it for many years prior to her death, in the same way as the other advanced cases.

CASE XXII.—A girl of eight has ataxia of the legs with spastic symptoms so that she walks on tip-toe. Her symptoms have developed gradually during the last two or three years. I have not seen the case, but my information is reliable, though meagre.

CASE XXIII.—A boy of six, with paralysis of both legs, from anterior polio-myelitis (?), at the age of two.

CASE XXIV.—A boy of thirteen has never learned to walk, though the legs are not stiff or atrophied. He is reported as all right in every other way.

This completes the list so far as my present information extends.

Cases XXIII. and XXIV. are only cited as having a collateral bearing on the subject; and though there is some probability that XXII. and XXIV. are affected with this disease, I could not reasonably insist upon it from my present information.

In Cases I. and IV. the history is meagre, and though the period at which patient No. 1 lived is remote, yet the fact that this series of cases has been confined to families who have always maintained a good position in society, and whose family record has been well kept and can be readily traced, and also the very obtrusive nature of the symptoms, together with the fact that they are in most cases progressive through a variable number of years, warrant a diagnosis of a comparatively meagre history. For my own part I have become intimately acquainted with several branches of the family, and particularly with that branch from which I have had most of my information; and I can vouch for their superior integrity and intelligence.

Four cases—Nos. 18, 19, 20 and 21—have been under my observation steadily for seven months, and Nos. 9 and 10 for three months; and two cases, Nos. 5 and 15, have been well examined by Dr. Norman Bridge.

Taking these cases alone for a text and assuming them to be cases of hereditary ataxy, the following diagnostic criteria might fairly be deduced:

Hereditary ataxy is a disease which may be traced through several—at least four—generations, increasing in extent and intensity as it descends, tending to occur earlier in life and advance more rapidly. It usually attacks several members of the same family. It occurs most frequently between the ages of sixteen and thirty-five, but it may begin as early as eleven and as late as forty-five. It shows no

marked preference for sex, but it descends through females four times as frequently as through males. Atavism rarely occurs. The influence of an exciting cause can rarely be demonstrated, but in some instances a fall or injury has appeared to determine the onset; and any cause like child-bearing or lactation, which very much depresses the vital forces, may produce a rapid advance of all the symptoms. There is always considerable inco-ordination of all the voluntary muscles, and a sluggishness of the movements which they produce when the disease is well established. This is usually noticed first in the muscles of the legs, but in a few months or years extends to the arms, face, eyes, head, and organs of speech. Sometimes it occurs first in the upper extremities, and sometimes in the organs of speech.

The ataxy is often extreme, and the gait devious, the patient deviating several feet on either side of the intended line of progression, before he loses the power of walking. The ataxy is not markedly increased by closure of the eyes. The sense of posture is perfect.

Some weakness of the muscles of the legs, without atrophy, is frequently an advanced symptom, and occasionally there is permanent spastic contractions of the legs. In developed cases there are usually extensive choreiform movements of the head and often of the arms accompanying all voluntary movements. These irregular movements occur in the hands, legs or head whenever it is attempted to maintain either of these parts in a fixed position by a voluntary muscular effort. Movement ceases during sleep. The pulse rate may be increased to 112 in advanced cases or may be normal.<sup>1</sup>

There is usually some degree of static ptosis, with overaction of the levator on looking upward. In rare cases there may be temporary diplopia, in the early stages, due to weakness of the external rectus. There is no nystagmus of any kind.

Atrophy of the optic nerve is a constant and early symptom, and usually progresses slowly with the other

<sup>1</sup> I am inclined to attribute the pulse rate noted by Dr. Bridge to emotional disturbance, because I have made repeated examinations in the six cases under my care and found it only subject to normal variations.

symptoms. Rarely it begins earlier in one eye than the other.

The response of the iris to light and accommodation is sluggish and diminishes with the advance of optic nerve atrophy; when this latter is complete, as may happen in advanced cases, there may be complete internal and external ophthalmoplegia.

There is always marked disturbance of the articulation, probably due to inco-ordination of the muscles concerned, for weakness cannot be demonstrated. In some cases there is a troublesome tendency to strangulation in swallowing liquids, due to their getting into the larynx, but otherwise swallowing is in no way difficult.

Occasionally the sphincters are slightly, but positively, affected, this symptom only appearing in those cases where spontaneous pains in the legs co-existed, having some of the characteristics of those occasioned in locomotor ataxy. Excepting the spontaneous pains already mentioned, there is no disturbance of sensibility. There are no vaso-motor or trophic symptoms, but there is a marked tendency to emaciation; there is no hypertrophy or valvular lesions of the heart.

The knee-jerk is always exaggerated and there is frequently ankle-clonus, and the cutaneous reflexes are also always exaggerated, but to a less degree. The exaggeration of the reflexes is an early symptom, and they often decline considerably when the disease is far advanced.

There is never paralytic club-foot, nor any other deformity excepting rarely permanent spastic contractions of the legs in advanced cases. In none of these cases have the patients ever suffered from rheumatism so far as I can learn.

I wish to repeat that the above summary of the symptomatology of hereditary ataxy is only intended to apply to this particular series of cases; and I have only presented it in this way so that it might be the more easily compared with other series.

I regret that I am unable to show any sections illustrative of the pathological anatomy of the disease. Of the pathology it seems pretty evident that the prominent fea-



tures consist in an extensive degenerative process affecting mainly in the upper motor segment, that is the cortical cells and the fibres extending from them to the cells in the cord and medulla; and that the tissues concerned have derived a deficient vital endowment from the parent; this deficiency manifesting itself as frequently after, as during, the age of development.

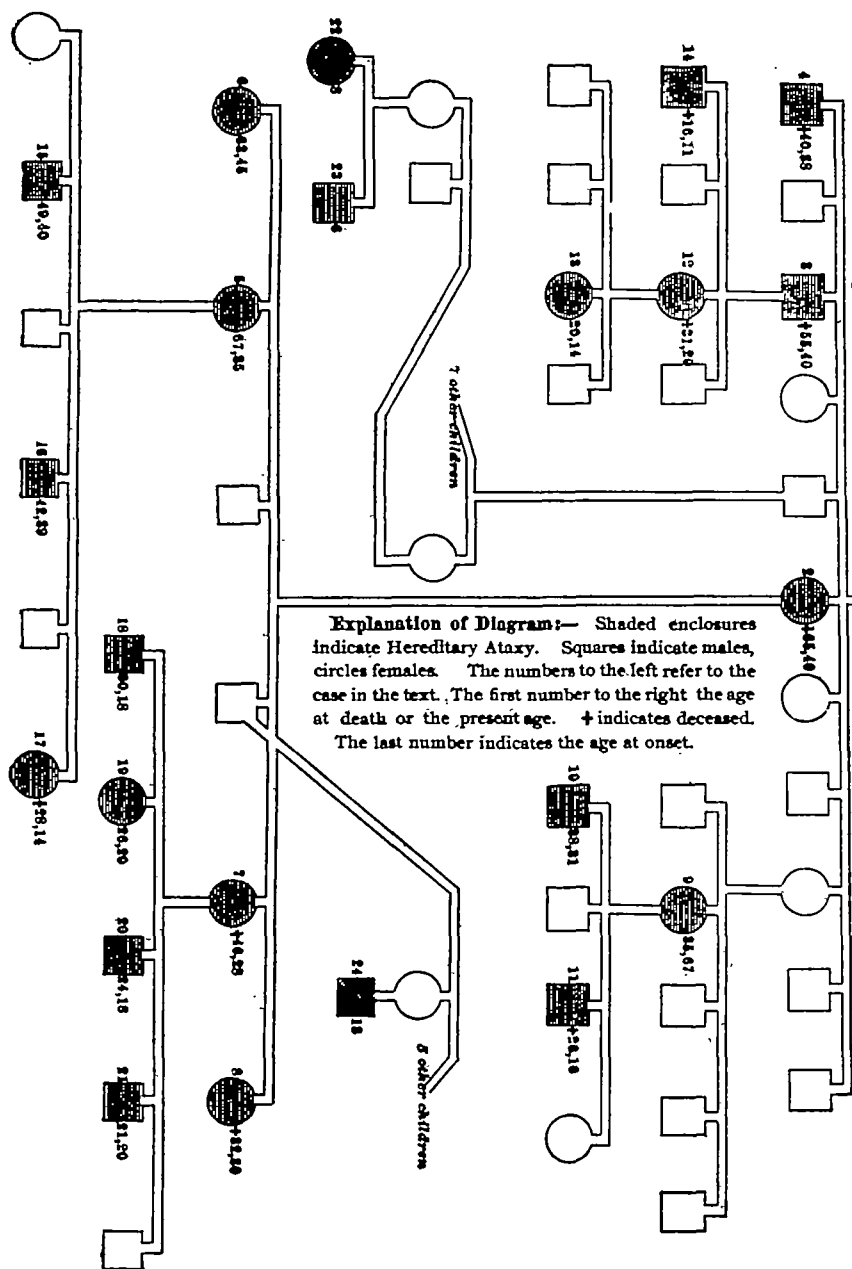
The optic nerve atrophy, the occasional spontaneous pains, associated with muscular weakness of the bladder and rectum, together with a tendency to emaciation, suggest that tissues outside of the upper motor segment may be either primarily or secondarily involved.

The integrity of the muscular sense and the peculiar nature, wide extent and extreme degree of the ataxy, together with the state of the reflexes, suggest that the difficulty lies rather in the efferent than in the afferent paths.

The degeneration is uniformly progressive, though it presents considerable variation in the rate at which it advances in different individuals and at different times in the same individual. I have applied galvanism along the spine in three cases about three times a week for the last five months, using a current varying from 25 to 60 m.a. for ten minutes. The more advanced patient has an impression that the strong current did harm, and he is inclined to attribute the recent considerable loss of power in the legs to that cause; and while such an influence cannot be certainly demonstrated, I should in future cases hesitate to apply a current of more than from 20 to 30 m.a. I changed the poles usually several times during each sitting.

I have paid careful attention to the general health, using wine with a bitter tonic and arsenic from time to time; and all the patients have gained several pounds in body weight. In the case least affected, I cannot observe any advance of the symptoms, but in one there has been an advance of the ataxy noticeable in the gait, and in the third a somewhat rapid onset of weakness in the legs coincident with a bad cold, already mentioned. Now, after about ten weeks, muscular power in the legs has returned to a considerable degree, but not so as to permit the patient to walk.

Dr. Sanger Brown.

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The main points of difference between the symptoms observed in my series, and the criteria established by Dr. Ladame, I will not occupy space to further indicate, as I have already gone far beyond my intended limits.

With reference to the etiology and symptomatology set down by Dr. Gowers in his text-book, if the age at the time of onset were considerably advanced, and if it were stated that the knee-jerk and skin reflexes might be greatly exaggerated, that in some cases there might be ankle-clonus, and that there might be optic atrophy, the cases herein reported would be fairly well provided for. Indeed, it would seem that there may be much more variation in the age at which the disease first makes its appearance, the distribution and intensity of the pathological process and the consequent symptoms, than has hitherto been suspected.

## REMARKS ON DR. SANGER BROWN'S CASES.

### I.—BY DR. ORMEROD (LONDON).

Dr. Sanger Brown brings forward a long series of interesting cases, broadly characterised by the title of hereditary ataxy. Ataxia, indeed, was the leading symptom, while the hereditary nature of the disease in the families of this stock is sufficiently obvious. As to the date of onset<sup>1</sup> Dr.

<sup>1</sup> The dates of onset, given in his diagram, fall as follows:—

#### 1st Generation—

Case 1, F, at 45; died at 72

#### 2nd Generation—

Case 4, M, „ 38; „ 40 }  
 „ 3, M, „ 40; „ 55 }  
 „ 2, F, „ 40; „ 65 }

#### 3rd Generation—

Case 14, M, „ 11; „ 16 }  
 „ 12, F, „ 20; „ 31 }  
 „ 6, F, „ 45; „ }  
 „ 5, F, „ 35; „ }  
 „ 7, F, „ 28; died at 46 }  
 „ 8, F, „ 30; „ 32 }  
 „ 9, F, „ 35; „ }

#### 4th Generation—

Case 18, F, at 14; }  
 „ 22, F, doubtful }  
 „ 23, M, „ „ }  
 „ 15, M, at 40; „ }  
 „ 16, M, „ 39; „ }  
 „ 17, F, „ 14; died at 28 }  
 „ 18, M, „ 18 }  
 „ 19, F, „ 20 }  
 „ 20, M, „ 18 }  
 „ 21, M, „ 20 }  
 „ 10, M, „ 31; }  
 „ 11, M, „ 16; died at 26 }