Critical Digests.

FRIEDREICH'S DISEASE.

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(Read before the Medical Society of Geneva. Translated by W. Dudley, M.B.)

DUCHENNE de Boulogne* had just published his classical study on progressive locomotor ataxy (1858 and 1859). This work aroused great interest, and immediately gave rise to numerous controversies, of which we still hear the last echoes. Duchenne has often been reproached, and with reason, with having caused an unfortunate confusion by this name, "locomotor ataxy," from his taking the symptom for the disease. Jaccoud† one of the first, in a historical critique too severe to be fair, accuses Duchenne of having fallen into an error, carefully avoided in recent times, when he named this disease from the functional trouble, from the symptom which it causes. "It is," says Jaccoud, "a nosological error of the same kind as if one called pulmonary phthisis, chronic cough; pneumonia, acute cough; or myelitis, paraplegia."

Nothing discloses this error better than the history of the disease described for the first time by Friedreich, September 18th, 1861, at the Congress of German naturalists and physicians at Spire. The eminent clinician of Heidelberg had indeed observed with the greatest care several cases of locomotor ataxy, the symptoms of which differed greatly from those which had been described two years before by Duchenne in the *Archives générales de Médecine*. Now Friedreich, who frequently had the opportunity, in

* DUCHENNE DE BOULOGNE, De l'ataxie locomotrice progressive. *Arch. gén. de méd.*, Dec. 1858, Jan., Feb., April, 1859.

† S. JACCOUD, Les Paraplégies et l'Ataxie du mouvement. Paris, 1864, p. 583

1858 and 1859, of showing his cases to several of his colleagues, among others to Eisenmann, Hasse, Kussmaul and Virchow, accepts only with reserve the clinical picture of Duchenne. He protests even against the pathological type created by the latter. "The complexus of symptoms," says he, "which constitutes progressive locomotor ataxy of Duchenne, cannot absolutely be considered as representing a distinct disease *sui generis*. It is a morbid entity, a group of symptoms which has its anatomical and physiological characteristics, but it is not a special disease, for progressive locomotor ataxy, the failure of co-ordination of voluntary movements, presents very great variations." Witness the cases observed and described by Friedreich, which offer very remarkable differences from those of Duchenne.

Friedreich, with his clinical insight, had therefore correctly guessed that his cases did not come under the description of locomotor ataxy of Duchenne. He did not think, however, that he ought to separate them entirely. He thought that the picture of Duchenne was incomplete, and as one of the fundamental differences between his cases and those of the great French pathologist was found to consist in the absence of sensory disturbances, he adopted the following hypothesis, which might, according to him, furnish a satisfactory explanation of it.

"If, therefore," says Friedreich (*), "in the different cases of locomotor ataxy which have been published, we find sometimes sensory disturbances, sometimes complete absence of them, it is because, in the first case, degeneration of the medullary fibres is not limited to the posterior columns, but has invaded other regions of the spinal cord."*

When he put forward this hypothesis, ingenious rather than well founded, Friedreich had already made three autopsies, the results of which would have permitted him to conclude that there might be combined sclerosis of the posterior and lateral columns, since this lesion manifestly existed in two of his cases.

Nevertheless, many years will pass before the ataxy of

* JACCOUD, *loc. cit.* p. 677, adopts this erroneous hypothesis.

Friedreich takes rank in nosography. For a long time, quite recently even, we have seen it, this morbid form will be considered by the authors, Topinard, Erb, Eulenburg, Jaccoud, Möbius, Grasset, Strümpell, &c., as a variety of tabes dorsalis (locomotor ataxy of Duchenne). To-day even, it is in the chapter on this affection that we must look for the description of *hereditary ataxy* in pathological works.

Hereditary ataxy! That is, indeed the name chosen by Friedreich as the title of his second memoir. The disease is not observed, he thought, in isolated individuals. Each time that he had seen a case of it he met with several others in the same family amongst the brothers and sisters of the patient. Eisenmann,† who speaks nowhere of Friedreich's cases, was the first to employ the words hereditary ataxy (*erbliche ataxie*), quoting as an example the case observed by Carre, and the opinion of Trousseau. This name of hereditary ataxy has been justly criticised. M. Féré ([33]), following Dr. Gowers ([25]), of London, and Brousse ([34]) of Montpellier, has shown that it was "manifestly wrong," since, on the one hand, classical locomotor ataxy recognised also very frequently for primary cause, either direct heredity, or more often neuropathic heredity, and, on the other hand, Friedreich's disease is absolutely distinct from locomotor ataxy, and consequently should not be confounded with it under a common name. Moreover, it is not heredity which is the essential and striking feature in this affection; it is rather its "familial" character, that is to say, the frequency of the development of the disease in several children of the same family. Dr. Everett Smith ([60]), of Boston, proposed for this reason to call it "postero-lateral spinal sclerosis of generic origin," or more simply "generic ataxy." This name, however, does not suit a disease which has nothing in common with progressive locomotor ataxy, as Pitt ([74]) says, except the abolition of the knee jerks.

After having been regarded as a particular form of locomotor ataxy of Duchenne, Friedreich's disease was also considered by some authors as a simple variety of dissemin-

† EISENMANN in Würzburg, Die Bewegungs-Ataxie. Wien, 1868, p. 205.

ated sclerosis, or at least as a combination of locomotor ataxy and multiple sclerosis. Charcot* and Vulpian† had in fact just described the symptoms of disseminated sclerosis. Everywhere interest was taken in the strange manifestations of this affection, of which a learned diagnosis was gladly made in medical clinics.

An attentive study of Friedreich's cases made M. Charcot at first think that perhaps he was dealing with disseminated sclerosis having invaded principally the posterior columns.‡ Such a supposition must indeed have presented itself naturally to the mind of observers of that period, who had never seen cases analogous to those of Friedreich. Hence, M. Bourneville examines in a special chapter§ Friedreich's cases under the title, "Cases in which there exist simultaneously the symptoms and the lesions of locomotor ataxy and of disseminated sclerosis."

After having given the translation of two of the principal cases of Friedreich, M. Bourneville expresses himself as follows:—

"The mode of onset indicated is not that which is ordinarily seen in ataxy, whilst it is much more conformable to that of disseminated sclerosis. Difficulty of speech, trembling of the head, nystagmus, are morbid phenomena foreign to the classical type of ataxy. Finally, contrary to what takes place so frequently in this disease, cutaneous sensibility and vision were not affected."

And with regard to the second case:—"The onset here has occurred in the same way as in the first case. . . . Moreover, we see that the patient has had spasmodic contractions in the peronei muscles, stammering, nystagmus, enfeeblement of motor power and of vision, and moreover that cutaneous sensibility was intact.

* CHARCOT, Leçons sur les maladies du système nerveux, t. I (Clinical lecture of 1868). *See* Bourneville's digest in *Mouvement médical*, 1868, Nos. 197, 235, 259 et 327.

† VULPIAN, Note sur la sclérose en plaques de la moelle épinière. *Union médicale*, 1866.

‡ See FETZER'S "Klinische Vorträge von Charcot," Stuttgart, 1874, p. 521 quoted by Friedreich.

§ BOURNEVILLE, Nouvelle étude sur quelques points de la sclérose en plaques disséminées in BOURNEVILLE et L. GUÉRARD, De la sclérose en plaques, p 191 Paris, 1869, chap. ii., p. 204.

"On the other hand, apart from lightning pains, there is nothing clearly belonging to ataxy, for the motor disturbances are so badly defined that one might ask if we are concerned with the trembling of disseminated sclerosis or with the inco-ordination of locomotor ataxy. . . ."

And M. Bourneville concludes:—"What is the outcome of this? It is that incontestably there is a mixture of two diseases . . . to wit, the co-existence in this case of progressive locomotor ataxy and of disseminated sclerosis."

Professor Charcot does not admit this combination of the elementary forms of these two diseases. "For myself," says he, in his Lectures on the Diseases of the Nervous System, "I have never met with, on the cadaver, the co-existence of multiple grey induration and posterior *fasciculated* sclerosis, and without denying that this association might exist, I believe it to be at least infinitely rare. It is pretty common, on the contrary, that the sclerous patches, which, as a rule, are principally seated in the antero-lateral columns, pass across the postero-lateral fissures, and encroach upon the posterior columns. Sometimes even I have seen them, having become continuous, occupy a large part of the thickness of these columns in the whole extent of one of the regions of the spinal cord—the lumbar, for example. Now, in all the cases of the latter kind, the ataxic symptoms had been, during life, manifested in different degrees."* And M. Bourneville adds in a note that the two cases of Friedreich, referred to above, belong to this category, what Charcot had not said expressly.

Friedreich ([11]) refutes this opinion by close reasoning, and demonstrates victoriously that his cases do not belong to disseminated sclerosis. "Why," says he, "has Bourneville made choice in my cases, reproducing some, and leaving others out? It is very surprising, for example, that he does not mention my first patient, Andreas Lotsch, since in this case the clinical and anatomical characters of degeneration of the posterior columns were certainly of the clearest." Friedreich then asks himself, by what signs one ought to

* CHARCOT, Leçons sur les maladies du Système nerveux, t. I, seconde edition. Paris, 1875, p. 248.

recognise "a true ataxic." Should it be, as Duchenne has laid down, by the absence of speech troubles? But one might upset this statement by arguing, on the other hand, that *true ataxics* are those in whom the want of co-ordination extends to speech and to ocular movements! "In my opinion," says Friedreich, "Charcot has attributed in my cases too great importance to nystagmus and to disorders of speech, whilst all the other more characteristic signs of disseminated sclerosis have been absolutely wanting during the many years that the patients have been under my observation. They have never presented diplopia, amblyopia, muscular contraction, spinal epilepsy, epileptic or apoplectic attacks, intellectual disturbance, bedsores, &c. The ataxic nature of the motor disturbances of my patients was so characteristic that it was impossible to doubt their significance, and because the ataxy invades the muscles of the tongue and those of the eyes, would that be a reason for not considering them true ataxics! Is it not, on the contrary, still more a reason for considering them as such?

"I find it incomprehensible," says Friedreich finally, "that anyone can still speak of disseminated sclerosis, when I have given the results of three carefully-made autopsies . . . and I hope that Charcot, in the vast field of observation which he commands, will sooner or later find a case analogous to those I have described."

This prediction was to be realised eight years later. I shall remark, moreover, that Charcot had never himself definitely pronounced on Friedreich's cases. He remained rather expectant, as we see by the quotations borrowed from his lectures. But in 1884, in one of his clinical lectures at the Salpêtrière, he comes boldly from his scepticism, and shows a young patient affected with hereditary ataxy, which he for the first time considers as a special affection, clearly differentiated from disseminated sclerosis and from locomotor ataxy. One may say that from that moment Friedreich's disease was clinically recognised in official teaching; henceforth it will occupy a distinct place in nosography among spinal affections.

"This disease," says Charcot, "approaches in symptoms

at the same time, locomotor ataxy properly called and disseminated sclerosis, whilst it distinguishes itself from them entirely in certain directions; it has been called hereditary ataxy, or Friedreich's disease" (45).

Two years before had appeared at Montpellier the important work of Dr. Auguste Brousse, who was the first to propose the name of "Friedreich's disease," to designate the particular morbid affection so well described by the professor of Heidelberg. This work gives a complete *résumé* of the subject at that time. It appears interesting to reproduce from it the principal conclusions:—

"The disease studied by Friedreich," says Brousse (84), "under the name of hereditary ataxy, is a special disease having a proper etiology, symptomatology, and morbid anatomy. It is characterised—

"1. From the etiological point of view, by its development in childhood or adolescence, due to hereditary influence, direct or indirect; the two sexes appear equally predisposed.

"2. From the symptomatic point of view, by ataxy of the four limbs, commencing in the lower extremities and developing so as to reduce the patient to almost absolute functional impotence; by difficulty of speech, by absence or late appearance of sensory disturbances; by absence of trophic troubles and preservation of tone of the sphincters; by slow and continually advancing progress; by very long duration; finally, by an always fatal termination, generally brought about by an intercurrent disease.

"3. From the pathological point of view, by fasciculated sclerosis of the posterior columns of the cord, extending upwards to the bulb, complicated by diffuse sclerosis of the lateral and anterior columns.

"4. From the diagnostic point of view it must be distinguished from progressive locomotor ataxy, from disseminated sclerosis, and, lastly, from cases described in Germany by Westphal and Schultze under the name of combined degeneration of the columns of the cord.

"5. From the nosographic point of view it renders necessary the creation of a new class of chronic spinal affections—the class of mixed myelitis."

In England hereditary ataxy was first described before the Clinical Society of London, October 8th, 1880. At this meeting Dr. Gowers showed three patients belonging to a family of nine children, five of whom were ataxics. He specially insisted on the characters which absolutely distinguish this disease from ordinary locomotor ataxy. Gowers recalled on this occasion that Dr. Carpenter (6) of Croydon, in 1871, had brought before the same Society two sisters affected with a curious disease hitherto unknown, and which was no other than Friedreich's ataxy. Since then, a brother of these two patients has also been attacked by the same disease.

At the meeting of November 13th, 1871, at which Dr. Alfred Carpenter exhibited his patients, the Clinical Society appointed a committee, composed of Drs. Richardson, Hughlings-Jackson, Lockhart Clarke, and Broadbent, to examine these remarkable cases, and to report at the next meeting. These two sisters presented the important symptom of lateral spinal curvature already mentioned by Friedreich, and which has been recognised as one of the characteristic signs of hereditary ataxy.

In recent years, the number of cases of this disease observed in France, Germany, England, Italy and America has so far increased that M. Soca (21) in his excellent thesis, in which are recapitulated almost all the known cases, reckons up 165. But that is a very exaggerated total; for if the published cases are more closely examined and seriously criticised, it is soon perceived that the list contains a very great number of cases which are incomplete, doubtful, or even absolutely foreign to Friedreich's disease. Before proceeding to this critical review, which will allow us to eliminate many cases which are inappropriate, and which obscure the clinical picture, it appears necessary for us to fix first of all the essential characters of this picture. To this end I shall give first of all a *résumé* of a case of my own which has been under my care for three years (April 1886), and which is thoroughly typical of Friedreich's disease

A. K., aged twenty-four (in 1886), a member of a liberal profession, has been affected from the age of eight, and has been

unable to walk for the last three years, since he had acute articular rheumatism, convalescence from which was prolonged for several months, at the age of twenty-one.

Family History.—Father, aged fifty-six, is very robust, it is said, and has never been ill; I have no information as to his health or his habits. Mother in good health also, but very nervous.

On the paternal side are several uncles and aunts, it appears, who are more or less affected with nervousness and peculiarities of character. There are some "hot-headed" members in the family. But no one is known to have presented symptoms analagous to those of our patient. A younger brother, aged sixteen, only walked when three-and-a-half years old; he is imperfectly developed for his age, very nervous, and of an irritable and troublesome temperament. A younger sister is in good health, but is also nervous.

Patient's History.—His early childhood presented nothing particular. Suckled by his mother, the child developed normally, and walked at fifteen months. Very intelligent and precocious for his age, he early learned to read and write. At the age of eight the first difficulty in walking appeared, a certain weakness of the legs and of the movements, which was looked upon as "nervous." He frequently fell; he walked slowly, and he planted his feet one before the other. He was scolded, because it was looked upon as carelessness and idleness. Nevertheless, he was able to walk easily alone. He never complained of pain. About two years after, he became ungainly with his hands, and five years only after the onset (the first symptoms in the legs) his speech began gradually to become drawling and difficult. The boy was then aged thirteen. Never involuntary grimaces, but so-called nervous movements in the arms and legs during walking. One day when the child was suffering from headache and stomach ache, a medical man was called in, and his visit was taken advantage of to consult him on the singular movements observed in the little patient. The doctor had him undressed, made him walk before him, examined him carefully, and said: "Oh, it will be nothing, it is St. Vitus's dance"—ordinary chorea slightly developed. Now this supposed St. Vitus's dance has never disappeared, on the contrary, it has been accompanied little by little by other symptoms, so that the disease has insensibly become aggravated. For three consecutive years, however, the patient was treated for St. Vitus's dance.

At the age of thirteen, the boy, who was well-advanced at school, was apprenticed to a lithographer, with whom he remained

three years without making any progress. The employer said that he was too weak to follow this trade, and that he would never make anything at it. We must suppose that the ataxy of the arms, which had become more and more developed, was an invincible obstacle to the work of the young apprentice. He was then sent to college, and at the end of some months he had already outstripped his companions by the vigour of his memory and intellect. Henceforth he advanced rapidly and did not cease to devote himself to intellectual labour. However, his gait became more difficult, his handwriting more slow and trembling. At the same time his speech also became slower. At the age of seventeen the patient began to make use of a stick for walking, and henceforth did not give it up. But the progress of the disease had been so insensible and so slow for several years, that he attached scarcely any importance to these symptoms, and the less so as they were not at all painful. He did not perceive, so to speak, that his disease was progressive. Thus he reached the age of twenty-one. At this period he was attacked with acute articular rheumatism, which confined him to bed for three months, and at the end of several months' convalescence he perceived that he could no longer walk alone. In darkness especially he felt less firm on his legs, and for a long time, even before his rheumatic attack, he could not go out in the evening, whilst during the day he still walked pretty well with the assistance of a stick. His legs had become more and more weakened, and even while giving one arm to someone, and helping himself with a stick with the right arm, the patient continued to find more difficulty in walking.

Never has he experienced lightning pains, nor girdle pains. Never incontinence of urine, nor any other urinary trouble. No disturbance of vision. Habitual constipation. No seminal losses. Never syphilitic infection.

Present Condition.—(He was examined several times from April, 1886 to April, 1889). Body generally imperfectly developed. Muscular system rather feeble. Adipose tissue scanty. General pallor of integuments. Skeleton fairly formed. Head large, round, symmetrical, brachycephalic. Scoliosis, with convexity to the right very pronounced in the region of the dorsal vertebræ, the ribs bulging on the same side forming a well-marked prominence under the scapula. No other peculiarity to note as to the development of the skeleton except the deformity of the feet which we are about to describe, remarking that it is not of bony nature.

The feet are small, deformed, affected with talipes equinus,

and as if shortened. Their colour is of a darkish blue. They feel cold to the touch. The sole of the foot is much hollowed, and has undergone a certain twisting, which is seen by the transverse furrows of the skin, which have become oblique from within out, and from behind forward, as if the anterior part of the foot had been twisted on the posterior. There is no permanent contraction of the sole of the foot. The dorsum of the foot is prominent; there is a projection at the level of the tarsus. The toes, especially the great toe, are raised up clawlike, and the tendon of the extensor proprius pollicis appears prominently under the skin. These symptoms are especially marked when the foot is raised; they are largely effaced when the patient places his foot on the ground. The articulations are flexible. All passive movements are easily performed. The tibio-tarsal articulation is relaxed, and voluntary extension of the foot can be prevented by fixing it with the hand. During walking the toes are constantly affected with movements of athetosis.

Motor symptoms.—The patient cannot stand upright without support. His trunk, head, and limbs are always in motion. He says that he could never be photographed because he is too nervous, and his features are too mobile. In reality it is because he cannot hold still an instant; even when he is at rest, his head and his trunk are incessantly agitated. It is static ataxy, as Friedreich calls it; when seated in an armchair he is perpetually moving. They are a kind of badly marked choreiform movements. We can understand that at the commencement of the affection the disease was mistaken for chorea. We shall speak of it again in treating of the diagnosis. Let us add, however, that this "choreiform instability," as Charcot calls it, is not always alike. Sometimes very pronounced, it is much less so on certain days, especially if the patient is well rested.

All voluntary movements of the arms and legs can be executed, but are manifestly ataxic. It is impossible for the patient, when told, to touch with his index finger, at the first attempt, the tip of his nose or the lobule of his ear, even with the eyes open. He always places his finger at a certain distance from the point intended, then, without leaving the surface of the skin, he makes it glide slowly to the indicated spot. The patient, whose intelligence is highly developed, is well aware of how his actions proceed. At the moment of raising his index finger to execute the required movement, he knows very well where he is going, but he soon loses control over the direction of his arm, because he has no longer an exact notion of the position of the limb during move-

ment. We see it then perform oscillations which distantly recall the trembling of disseminated sclerosis, but do not sensibly increase in intensity at the final moment. If the patient is told to take up a small object from the table—a pencil, for example, a match, or better still, a pin—he advances his hand above the object on which his eyes are fixed, makes it hover an instant, hesitates, separates somewhat his fingers, then darts suddenly on the object to seize it as a bird of prey would do, with this difference, however, that the direction of the movement is sinuous and interrupted, instead of being direct as in the case of the bird who arrives by a straight line on the coveted object.*

The method of picking up the pin and the pencil from the table is also characteristic. The patient employs to this end all his fingers, which he keeps separate and then brings together, making the object glide into his hand till he holds it conveniently.

He buttons his clothes with difficulty, especially the small buttons. Nevertheless, even if his eyes are closed, he ends by reaching the small buttons, and when once they are held, he does not relax his hold. He cannot dress or undress without assistance, on account of the effort required to lift his clothes. At the moment of taking off his shirt, when it passes above his head and covers his eyes, he loses his equilibrium completely on the stool on which he is seated, falls backwards and remains with his legs in the air till someone comes to his assistance. However, he has made in the meantime great efforts to regain his equilibrium.

It is the incessant movements of the trunk and limbs which prevent the patient maintaining the upright position. If he is told to loose hold of his support, and he sees around him those who are ready to support him, he can remain standing an instant, his legs widely separated, and even if his eyes are closed he is not more unsteady at first; but his unstable equilibrium is soon disturbed, a more extensive movement of the body makes him lose his balance completely, and he would collapse if not supported immediately (Romberg's symptom).

In spite of this, however, it is easy to assure oneself of the integrity of the muscular sense. The patient distinguishes very readily, with his eyes closed, different coins placed in his hand, in the right hand equally with the left. He estimates also pretty correctly the difference of weights when he supports small objects in the hand.

* Carre had already observed this movement of the hand, which "hovers" over the object before seizing it, but it is to M. Charcot that we owe the comparison with the bird of prey, which appears to me appropriate and striking.

Dynamometer: right hand forty-one, left thirty.

The patient feeds himself but slowly, and with some difficulty.

The patient writes very well, although rather slowly. The letters are well formed, and not at all shaky. The writing is plain and easily read.

The muscular strength of the arms and legs is well preserved. His leg cannot be flexed with both hands when he maintains it in extension. There is marked enfeeblement of the movements of the feet. Since the patient has been unable to walk alone he thinks he has noticed certain changes in his gait. Formerly he walked more on the soles, the fore part of the foot; now more on the heels, after the manner of ataxics. At present he cannot make a single step without being supported on each side. When he wishes to walk he passes his left arm under the right arm of the person who strongly supports him, and he supports himself with a stick in the right hand. He advances then slowly and with difficulty, making each instant extensive movements of the head and trunk, whilst the inco-ordination of movement of the right arm, which holds the cane, causes the latter to go from side to side so that it often misses the ground, and makes the patient plunge forward at the moment that he felt secure of finding a support.

He drags his feet in walking and controls their movements by sight; at each step the point of the foot deviates inwards, and rubs on the ground, especially the left foot, whilst the heel scarcely touches it. The leg, moreover, is not thrown strongly forward as in ordinary ataxy, for the foot scarcely leaves the ground, and the gait strongly recalls that observed in spastic paralysis, except that the stiffness of the legs does not exist here. The steps are short, unequal and irregular, and when one foot has been put forward the other follows, its fore part being dragged along the ground, deviating strongly into equino-varus, whilst the trunk and the head are thrown back in a disorderly manner. When the foot is raised from the ground the inco-ordination of movement becomes very manifest; the leg is flung forward, and the heel falls heavily. To sum up, what characterises the gait is the fact that the trembling is more pronounced than the ataxy. With the least emotional disturbance, the patient can no longer walk. His legs give way under him. If he is tired his feet continually turn over, and he catches against every stone in the way. He can with difficulty mount stairs, but he descends them with yet greater difficulty.

When I took his foot-prints, after the method of M. Gilles de la Tourette, it was necessary to support him under the arms to

make him walk on the band of paper. This is what explains the absence of the characters which we have just described—the dragging of the front of the foot, and the heel hardly touching the ground. When supported the patient raised his feet, threw forward his legs and brought down the heel at every step. What the print especially verifies is the deep concavity of the sole of the foot, which is shortened, and the trembling, irregular walk of the patient, whose legs double up several times under him during the experiment; lastly, in several prints we find the double heel of ataxics.

The sensibility of the soles was, moreover, so exaggerated that the floor caused an insupportable sensation of cold, and the pressure caused pain; he cried out while taking the first steps with bare feet The toes were seen to be raised violently in a spasmodic way and the sole to become more hollow each time that the patient raised the foot. The feet very quickly became cold and more and more cyanosed. It was then very difficult to warm them. "When my feet become warm again," said the patient, "they readily perspire, and quickly lose their heat." He adds that the impression of cold to the feet was very painful to him and paralysed him. He felt plainly the ground and its inequalities, and did not experience any peculiar sensation in the sole of the feet as is the case in tabetics.

Disturbances of Speech.—It was towards the age of thirteen years that difficulty of speech was noticed. His parents remarked that the boy's words became drawling and thick. He did not himself notice it—at least he cannot fix the time when speech troubles first manifested themselves. He does not recall that he may have had an interval between the symptoms in the legs and arms and those of speech. He maintains that they have all been developed almost simultaneously.

His voice is monotonous, very low-pitched. Speech is slow, somewhat scanned, but without hesitancy. The mechanical articulation of the sounds and the duration of the words are alone affected. There is no stammering.

The patient pronounces long words with a single effort, although slowly, and somewhat scanning the syllables; but he has no hesitancy of speech, does not cut the words in two, does not stammer. He will say without stopping, but slowly, i-na-mo-vi-bi-li-té, all in the same drawling tone, no syllables being pronounced more rapidly than others. The small pauses are equal between each syllable. Whilst the general paralytic slurs his syllables, this patient articulates distinctly in a slow, monotonous, and drawling voice.

The tongue, when protruded, is of normal size. But it undergoes very marked ataxic movements; it contracts, it is suddenly drawn to one side, then is withdrawn, curving up behind the dental arch, to come out again with a sudden jerk. His reading is also slow and slurred. The soft palate reflex is absolutely normal. The knee reflexes are completely abolished.

Nystagmus.—Horizontal oscillation of the eyes when following a moving object is the last symptom which we should connect with ataxy. Nystagmus is not very pronounced. It must be looked for and brought out to be observed. It is not present when the eyes are at rest. It is a motor ataxy of the ocular muscles, a dynamic nystagmus, in opposition to a static nystagmus, a balancing of the ocular muscles in different directions, which is observed when the eyes are fixed on a point. The patient says that when he fixes an object his sight is soon fatigued. The oscillations of nystagmus are very rapid and of small amplitude; they do not exceed a millimetre in extent, and only take place at the moment when the eyeballs reach the corners of the eyes. However, they are still observed, though more rarely, when the eyes are directed in front; at the moment when the eyes are fixed on the finger placed a few centimetres before the forehead, opposite the root of the nose, some slight oscillations are seen to occur. It is what Friedreich had already clearly observed, remarking that in ordinary cases the existing nystagmus is thus made to cease, instead of being provoked.

Impulse to laugh.—The patient is often taken with an impulsive desire to laugh. He notices carefully these occurrences, which come on him unawares, and which he cannot resist. Lips habitually separate.

Sensibility absolutely normal in all its manifestations, to contact, pain, temperature and electricity. Nowhere in the legs are anæsthetic patches. He feels everywhere the lightest touch, and localises pin pricks perfectly. Never creeping sensations nor pain in the legs, particularly lightning pains. When the patient walked much, he experienced for several months a pain in the left knee. This articulation has never been swollen. Passive movements have always been free; the knees, left as well as right, supple and flexible. He then experienced great weakness in the legs, following fatigue; he could hardly drag himself along. He felt as if his legs were too long, "the nerves unstrung," especially during the north winds. The pain in the left knee then sensibly increased. He could no longer take long steps as usual. His feet dragged on the ground. This is why the patient

said: "The smaller the steps, the more awkwardly I walk." The pain, which he attributed to a rheumatic origin, has completely disappeared since the patient has been less on his legs, and has scarcely walked at all.

Pupillary reflexes perfectly normal to light, to convergence, and to accommodation. No diplopia nor strabismus; sight excellent.

Sphincters normal. The patient is not subject to constipation. He can retain his urine for long. No gastric troubles.

Genital functions.—No disturbance to note. No involuntary emissions. Erections normal. No impotence nor premature ejaculation.

Psychical functions normal. Intelligence remarkable, especially the poetic and imaginative faculties.

Electric reactions normal everywhere except in the muscles of the leg (antero-external group), where electro-muscular contractility is weakened.

During electric examination the patient, seated on a stool, cannot be left alone an instant; he loses his equilibrium at every movement, and as he is perpetually in motion he would fall if not watched. (Choreiform instability, static ataxy.) Moreover, he is weighed down, he is doubled up, and his head falls on his breast.

Galvanic Exploration of Nerves.—L. ulnar (K.C.C.) 1·5 ma.; R. 1·5. L. radial, 3; R. 3. L. peroneal, 2; R. 3.

Faradic Exploration of Muscles.—Tibialis anticus, R. 75 mm.; L. 80. Peroneal muscles, R. 85; L. 84. Gastrocnemius, R. 78; L. 82. Vastus ext., R. 83; L. 80. Interossei, R. 76; L. 80.

Galvanic Exploration of Muscles.—Tibialis ant., R. 8·5 ma.; L. 8. K.C.C.=A.C.C.; occasionally A.C.C.>K.C.C. Peroneal muscles, R. 7; L. 7, weak. Contractions normal, rapid. No R.D. Legs thin; atrophic on antero-external aspect.

Taking for our basis this case, and others which we have had the opportunity of examining in the wards of M. Charcot at the Salpêtrière, and of M. Dejerine at Bicêtre, and comparing them with the cases that have been hitherto published, we shall successively consider Friedreich's disease from the etiological, the symptomatic and the diagnostic point of view. Finally, we shall describe its pathological lesions, which class this affection among the combined scleroses of the spinal cord.

ETIOLOGY.

This case is distinguished from most of those which have been previously observed, by the absence of the *family* or *hereditary* (generic, as Smith calls it) character, which has for long been considered as the rule in this disease. Indeed, the brother and sister of the patient have not been attacked by the same affection, and if we credit M. Soca, who has deduced the law, from the observations of several authors, among others Gowers ([20]) and Rütimeyer ([41]), that the children of the same family become affected almost at the same age, we should be able to say that the patient's brother and sister no longer run any risk of having this affection, since they have both passed the age at which the disease commenced in their brother.

M. Soca formulates this law in the following way:— "Friedreich's disease commences at the same age, within two or three years, in all the members of the family which it attacks."

We shall, however, make some reservations on this point, and before adopting this law, the practical importance of which would be very great, we think that it is expedient to wait for a greater number of cases, and especially greater exactness among authors in fixing the onset of the first symptoms. Mr. Soca says so himself (p. 132):—

"The first phenomena which strike the patient and his friends vary somewhat in different cases, sometimes the onset of the disease dates even from birth. The little patients never learn to walk properly. It is only very late that they walk alone, and then their gait already presents all the characters of ataxy. Let me add that this mode of onset is pretty common."

In reality, the onset of Friedreich's disease is so often so insidious and insensible that the patient and his friends do not trouble about them till long after their appearance, when the weakness of the legs and the ataxy of movement have become sufficiently developed to interfere with walking. If the physician who is at length consulted then questions the parents they will tell him that they have noticed, for several years perhaps, certain signs which should have aroused their

suspicions, but without giving serious attention to them, because they attributed them to idleness and obstinacy on the child's part rather than to disease.

Must we then admit that external conditions, environment, occasional causes, are really without any influence on this disease which would thus fatally attack at the same age the members of a family? We must confess that our knowledge of the etiology of this disease is still so obscure that it is actually impossible to say anything on the subject. We think, however, that Friedreich's disease is no exception to the general laws of heredity, and we believe that one would commit a great error in wishing to withdraw it from external influences (always necessary for the development of hereditary morbid germs slumbering in the organism) by reason of the absolute ignorance in which we are of the occasional causes of this disease.

It has been observed in several attacks that the onset of the disease has declared itself after an acute disease—a fall, an injury, or after early habits of onanism. These are the common causes of which we must assuredly take account, but which can only be of influence in individuals hereditarily predisposed.

In the personal history of patients we sometimes find noted scrofula, tuberculosis, and infantile convulsions. But it is the eruptive fevers and typhoid fever which appear to have most often been the occasion of the onset of the weakness of the legs and the ataxy of the extremities. In one of Rütimeyer's patients the first symptoms of ataxy were manifested during a long convalescence following scarlatina of four weeks' duration Vizioli ([59]) has seen two cases of Friedreich's disease develop after scarlatina. Ormerod ([54]) also quotes a similar case. Whooping-cough and measles have also been observed to have preceded, by a longer or shorter period, the first manifestions of the disease. Rütimeyer and Vizioli have each seen a case in which the ataxy of Friedreich appeared after typhoid fever. Let us further note as antecedents the mention of febrile affections not attended by eruptive or cerebral manifestations.

Musso ([49]) insists at some length on small-pox as a

predisposing cause of the disease. He quotes numerous examples of affections of the nervous system observed after small-pox by Gubler, Westphal, Leyden, Bernhardt, Seppilli, &c., and thinks that in three of his patients it is the small-pox which has kindled the disease, which would otherwise perhaps have remained latent all through life. But in examining Musso's case more closely, it may justly be asked if the first symptoms of ataxy should really be attributed to small-pox. Indeed, in one case, the ataxy did not appear till two years after, in another two years and a half, and in the last not till three years after a mild small-pox.

It is to be remembered that our patient experienced a notable aggravation of his disease after an attack of acute articular rheumatism. At the end of convalescence from that disease he could no longer walk alone, as he could previously with the aid of a stick. In Friedreich's seventh case there is an analogous circumstance. Louise Schulz, whose disease commenced at the age of thirteen, was attacked at fourteen with acute articular rheumatism, after which the shaking of the limbs greatly increased, and speech became indistinct and stammering, which was not the case previously.

In other patients chorea has been mentioned as a pathological antecedent. Thus Rütimeyer ([41]) says that Jacob Blattner (his seventh case) had at the age of four years St. Vitus's dance for six months, and that after that affection he remained feeble in his legs and uncertain in his gait; since then, too, the disease invaded successively the upper extremities and speech, as in his eldest sister. We ask ourselves if this chorea should not rather be considered as a symptom of the onset of Friedreich's disease. We have seen that our patient was regarded for three years as suffering from chorea, and we know now that "choreiform movements" form part of the regular procession of symptoms of this disease. We think that chorea should henceforth be considered as belonging to the first period of Friedreich's disease, whenever the latter will appear to be the direct consequence of it. Other concomitant symptoms, such as the abolition of the knee reflexes, will serve in a doubtful case to settle the diagnosis.

Let us relate yet a negative etiological fact, which assumes a certain importance in the enumeration of the causes of so-called hereditary ataxy. We hardly ever find syphilis mentioned in the personal or hereditary antecedents of Friedreich's ataxics—contrary to what is observed so frequently in true tabetics.

On the other hand, alcoholic excess in the parents has been pretty often blamed. Friedreich was the first to insist on this point. The first six cases that he observed belonged to two families, Lotsch and Suss. Now, the fathers of these two families were drunkards, and the widow of Suss said that all her children were conceived during drunkenness. Other observers, notably Everett Smith, Quincke* and Rütimeyer have since related analogous examples. There are other cases, as a set-off, where alcoholic excess of the parents is absolulely excluded.

Direct heredity has, therefore, some share in the symtomatology of Friedreich's disease, as in that of myopathic atrophy and Thomsen's disease. There are, however, numerous exceptions to this rule, and isolated cases like the present one are to-day not rare in science. It would not be necessary to think on this account that these last cases escape the law of heredity. We establish most often, as in our patient, amongst ancestors and relatives nervous and psychical affections of diverse nature, so that these sporadic cases fall like the others under the general law of neuropathic heredity, although they do not present the "family" character, which is the index of an accumulated converging hereditary influence, that is to say, raised to higher power. (See subjoined the genealogical tables of Rütimeyer, Musso, Vizioli and Gowers.)

RÜTIMEYER (*Virchow's Archives*, 1883, vol. 91, p. 116 and 117). Blattner "Stulzi" (that is to say, "hobbling") married in 1710.

All the direct descendants of this Blattner are entered under this surname "Stülzi," in the official registers of the commune of Küttingen up till the "forties" of the present century, and the strange gait of this man must have strongly impressed his con-

* QUINCKE, in Friedreich's second paper, *Virchow's Archiv*, Bd. lxviii p. 165, 1876.

temporaries for it to have remained during so many generations in the recollection of the inhabitants of the country. However, the eight cases of Freidreich's disease have only quite recently shown themselves in this Blattner family, whilst previously neither nervous nor mental diseases were known in it.

These eight cases belong to four different branches, of which the great grandfathers were brothers and lived at the commencement of the present century, direct descendants of the famous Blattner-Stülzi. Here are the four branches (in the fourth generation probably, and not in the second, as Soca erroneously states).

BLATTNER-BLATTNER.
Ten children, of whom seven healthy.

Rudolf (1). Born 1805. ATAXIC (scarlatinal).
Gottlieb. Born 1848. ATAXIC.
Marie. Born 1853. ATAXIC.

BLATTNER-BASLER.
Seven children, six healthy.

Karl. ATAXIC. Mother affected with chorea in childhood. Maternal uncle paralysed in both legs suddenly (acute poliomyelitis).

BLATTNER-KYBURZ.
Seven children, six healthy.

RUDOLF (2). Born 1867. ATAXIC (following typhus).

BLATTNER-WEHRLI.
Ten children, seven healthy.

Marie Madeleine, age 20. ATAXIC.
Jacob. Died, aged 18. ATAXIC.
Fritz. Died of measles, aged 9. ATAXIC.

As for age and sex, we have but little to say of them. Friedreich thought that hereditary ataxy was a disease commencing at puberty, especially in girls. This observation is not always verified. Many cases date from infancy as well in boys as in girls. After the age of twenty it is extremely rare for Friedreich's disease to show itself.

Symptomatology.

The clinical history of our case furnishes a typical picture of the symptomatology of Friedreich's disease, which may thus be recapitulated: slow and progressive ataxy of the four limbs—usually attacking several children of the same family, dating 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 of the eyes; weakness of legs, increasingly difficult gait (tabeto-cerebellar, Charcot); choreiform unsteadiness; static ataxy (Friedreich); 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.

Sometimes other symptoms are added which are due to

complications, or which result from an error of diagnosis. It appears to us necessary from this point of view to eliminate a certain number of cases which are still classed under Friedreich's disease, although they have no right to be so considered. In reckoning these doubtful cases the symptomatology of this affection is confused, and elements of error are introduced which obscure the clinical picture of it.

Let us rapidly pass in review some of the cases which in our opinion ought no longer to figure in the category of Friedreich's ataxics.

There are first of all three patients of Hitzig,* referred to by authors and hawked about from one bibliography to another without any verification of the reference. As M. Soca remarks, this reference is inexact. It refers in fact to page 513 of the *Berliner Klinische Wochenschrift* of 1875, which is devoted to advertisements! In reality, it does not relate to a work of Professor Hitzig on Friedreich's disease, but simply to a discusion which took place at the Berlin Society of Medicine on January 20th, 1875, after a communication by Dr. Bernhardt on muscular atrophy. Hitzig said that he had had under observation for several months two brothers and a sister affected with muscular atrophy, in whom the disease had existed between five and ten years; seven other children of the same family perfectly healthy. The *mother* of these ten children was *tabetic* in a high degree. But there is not a word about difficulty of walking, which should have been observed in the little patients.

The error proceeds from Seeligmüller,† who thought that the atrophics of Hitzig ought to be classed under Friedreich's disease, perhaps on account of the tabes of the mother, and who speaks of difficulty of walking (*Gehstörunsgen*, loc. cit. p. 188), whereas there is no mention of it in the report of the discussion.

Seeligmüller, moreover, is not fortunate in the choice of observations on which he relies for the description of Fried-

* *Berliner klinische Wochenschrift*, 1875, *p*. 143.

† V. SEELIGMULLER in Halle, Krankheiten des Nervensystems II. Scleroses der Hinterstränge, p. 185.—In GERHARDT'S *Handbuch der Kinderkrankheiten*, vol. v., part ii., first edition, Tubingen, 1880.

reich's disease. Beyond the cases of Friedreich—the only really authentic ones at that period—he quotes *Du Castel*,* whose patient was not ataxic but paralysed; the two cases of Kellog (⁸), too incompletely described and presenting too many anomalies to be admitted as belonging to Friedreich's ataxy; the cases of *Bouchut*† (in which ataxy exists in name only), a boy eleven years of age, ill for fifteen days with tingling in the feet, sometimes diplopia with convergent strabismus and slight constipation, and a girl of fourteen years, an old rachitic subject, ill for two years, who walks with a firm but slow step, without trembling and without defect of muscular co-ordination; moreover, "one may tickle in any way the soles of her feet without making them move" (!!); lastly, the case of *Kahler* and *Pick* (¹⁵), combined sclerosis of the posterior and lateral columns, which is by no means a case of Friedreich's ataxy, the dominant symptom being paraplegia.

Let us add, that to crown his mistake, Seeligmüller (²⁵) has published two cases which certainly do not belong to Friedreich's disease; that Friedreich himself, consulted by the author, has refused to recognise them as analogous to his own, but which Seeligmüller persists in spite of Friedreich's authority, and in spite of evidence, in considering as authentic examples of hereditary ataxy with nystagmus. These are the cases:—

Leo von K——, age twenty-six, descended from an old noble family, in the genealogic tree of which neuro-psychopathies abound. Uncertainty of gait at twelve years, at fourteen progressive myopia, from fifteen to sixteen melancholia. Onanism. Sexual perversion. At twenty-one he enlists as a volunteer in the cavalry. He is a bad horseman. He has learned to swim, but he is a bad swimmer. He dances, but he is a bad dancer, for he easily becomes giddy if he turns round in dancing, and if he rides round. Girdle pains. Urinary troubles. Various psychical disorders. What is most striking in his physiognomy is his nystagmus, which the patient can arrest when he wishes,

* Du Castel, Observation de sclérose primitive des cordons de Goll. *Gazette médicale de Paris*, Jan. 17, 1874, No. 3, p. 33.

† Bouchut, Ataxie locomotrice et sclérose des cordons postérieurs de la moelle chez les enfants. Signes ophthalmoscopiques. *Gazette des Hôpitaux*, March 31. 1874, No. 38, p. 297.

so much control has he over himself! He is able to arrest the nystagmus when he fixes his look fifty centimetres in front of him. He then feels firm on his feet. The patient is never at rest, but he can always arrest his movements by an effort of will. No locomotor ataxy. With eyes closed he goes pretty straight to a fixed point in front, but his legs are too wide apart and he walks irresolutely. He mounts a chair, &c., without difficulty. Plantar reflex abolished; cremasteric absent on the right side. *Patellar reflexes very exaggerated.*

Alexander von R——, age twenty-eight, married, robust, very strong, but awkward. From childhood a grimacer; shakes his head; spasms; rabbit-like movements of the nose and lips. Syphilis. Gonorrhœa; vesical catarrh followed by violent pains in the right hip. For six months he has had difficulty in walking (October, 1878), yet he has walked for several hours—limping rather—to go to the chase. Asymmetry of face. Left strabismus. No nystagmus. Speaks very quickly, and gets flurried. Ataxy of thought but no trace of ataxy of speech. No spinal curvature. Walks with his legs apart, and with swaying of the body. His eldest daughter has the same gait. When upright, with eyes open, the patient does not stumble, nor does he if his eyes are closed. No trace of ataxy in the upper limbs. Plantar and cremasteric reflexes abolished. Knee reflexes very lively.

A sister, whom Seeligmüller was not allowed to examine, suffered, it appears, from symptoms in all respects like those of her elder brothers.

It is indeed sufficient to enumerate these symptoms to understand that Friedreich had good reason not to find in them an image of the disease which he had so conscientiously studied and described. Nor is Seeligmüller's indignant exclamation in any way justifiable, when he says (*loc. cit.*, p. 242): "To conclude, I must protest strongly against the assumption of Bourneville, who argues that the hereditary ataxy of Friedreich is a disseminated sclerosis, with predominant lesion of the posterior columns. The clinical picture offered by the patients of Friedreich, two of whose ataxics I have myself seen at Heidelberg in the autumn of 1878, as by my own is absolutely different from that of disseminated sclerosis."

Who would have doubted that Seeligmüller had seen Friedreich's patients? Doubtless Bourneville was deceived,

but we must confess that nothing resembles more a patient affected with disseminated sclerosis than an ataxic of Friedreich, with his choreiform unsteadiness, his nystagmus and his slow and scanned speech. It seems quite natural that one may have supposed, in 1869, at the time when disseminated sclerosis had just been discovered, and was not yet known under all its clinical varieties, that the first cases of Friedreich might be classed under that disease. We have already seen how Friedreich has replied to it; we shall not return to it.

Hammond ([28]) has enriched the statistics of hereditary ataxy by twelve cases, of which the very incomplete, superficial and aphoristic observations do not permit us in the case of any of them to subscribe without reserve to the diagnosis. It is absolutely certain, on the other hand, that most of these cases do not belong to Friedreich's disease. What can the following cases, for example, have to do with this affection? In the family of R—— there are three brothers who died young, and a sister in good health, of whose heredity Hammond says nothing.

1. A little boy, G. R——, ill since dentition, general paralysis and articulatory trouble; no pains; sensibility always normal; died aged five years.

2. His brother, L. R——, paralysed at six months; paralysis gradually increasing; died aged three years.

3. A third brother, W. R——, died aged three years; paralysis and articulatory difficulty, without atrophy and anæsthesia.

And authors would regard these patients as cases of Friedreich's ataxy! The twelve cases of Hammond should henceforth be erased from the list of this disease.

As much may be said for the two cases of Drs. *Oxley* and *Pollard* observed at the Liverpool Children's Hospital, and reported by *Davidson* ([31]).

Case 1. (Oxley).—M. R——, girl, aged nine years. Six weeks ago she was attacked without known cause, in two or three days, with muscular inco-ordination of the four limbs, as if she had chorea. Mastication and deglutition difficult. Speech embarrassed. Urine in the bed. Patellar reflexes exaggerated. Sensibility normal. Rapid improvement by appropriate hygiene and good nourishment. After five to six weeks she could walk alone.

CASE 2. (Pollard).—J. G——, little boy, aged seven years. Ten weeks previously an attack of rheumatic fever; for the last eight weeks has entirely lost speech and the use of his legs. At rest in bed no involuntary movements As soon as he wishes to make a movement, the latter is exaggerated and violent as in chorea. For a reply he makes a motion with his head. Sphincters normal. No exaggeration of tendon reflexes. Sensibility normal. Six weeks after entering the hospital he could walk alone, and stand with his eyes closed. Speech unimpaired.

It must be said that Dr. Pollard does not consider this patient as an ataxic of Friedreich, but considers him to be affected with paresis and functional ataxy, recalling certain hysterical manifestations.

The custom which has been followed, since Seeligmüller, of considering exaggerated or simply normal knee reflexes as belonging to the symptomatology of Friedreich's tabes, has contributed largely to lead astray the authors who have considered under the heading of this disease a number of cases with exaggeration of the knee reflexes, which ought not to figure in it.

Seeligmüller recognises that the symptoms of his patients differ from those which had been given by Friedreich as characteristic of his disease. In his manual he mentions in passing hereditary ataxy in the middle of the description of the symptomatic forms of tabes, and says (85) : "The symptoms of the cases observed by other authors and by mysel offer a clinical picture more or less different " (*in mehrweniger ausgesprochener Weise !*) to those of Friedreich.

I shall yet quote the two patients of *Massalongo* (48), the one of Descroizilles (68), and the one of Dalché,* which certainly do not belong tc Friedreich's ataxy. The abolition of patellar reflexes is an absolute rule in this disease, and one will do right always to mistrust the diagnosis of this disease whenever the knee reflexes will not be lost, although all the other symptoms would seem to confirm it.

Another category of patients, on the other hand, in whom the tendon reflexes are abolished, have also been improperly reckoned among the cases of Friedreich's disease, and should

* P. DALCHÉ, Maladie de Friedreich. Pseudo-tabes. Discussion *Progrès médical*, June 30, 1888, p. 507.

no longer be classed under it. The anomalies which they present suffice to remove them from this affection, and to place them either under disseminated sclerosis, under the combined scleroses, or amongst other spinal or general nervous diseases.

Brousse's patient, for example (34), whose first symptoms, weakness and inco-ordination of the legs appeared only at the age of twenty-four years, had at the end of his life apoplectic attacks which do not form any part of Friedreich's disease. Moreover, the ataxy and the articulatory troubles have peculiar characters foreign to this affection. This is how M. Brousse expresses himself:

"Walking is not possible without assistance. When it is attempted it causes trembling of the whole body. . . .

"When the patient is at rest, if he is questioned rather suddenly his head is affected with trembling. . . . There is difficulty in his speech, it is hesitating, stammering."

The autopsy revealed a combined sclerosis, fasciculated of the posterior columns, and diffuse of the antero-lateral, with central myelitis. It was not therefore Friedreich's disease.

The cases of *Erlenmeyer* (40), *Hollis* (26), *D'Arcy Power* (32), *Fazio* (56), and many others besides, are doubtless liable to the same objections. The case of Carre (7), too, offers certain symptoms which render it suspicious. The disease began at twenty-two years by pains in the sole of the feet and in the thigh, then on the inner side of the hands, which were *dead* (sensibility blunted). Diplopia and weakness of sight. Speech impeded; a kind of stammering; one would say that the tongue, hindered by an obstacle, has difficulty in detaching itself from the palate, *then a series of words succeed one another rapidly*—all that is absolutely foreign to the symptomatology of Friedreich's disease.*

* I had written this critical review of cases of Friedreich's disease before reading the excellent article which Professor F. Raymond has devoted to tabes dorsalis in the Dictionary of Dechambre (See Bibl., No. 62). I see with pleasure that M. Raymond no longer admits the following cases as examples of the disease described by Friedreich—(1) the case of Kahler and Pick (35); (2) the two cases of Seeligmüller reported above; (3) the case of Brousse; (4) Hammond's case. The author remarks, moreover, that Carre's

After throwing out many cases considered by different authors as belonging to Friedreich's disease, we propose, as a set off, to add one which appears to us truly typical We found it in the work of Topinard (4), and as it has passed entirely unnoticed, we give of it the following *résumé:*

Case 215.—Edouard L——, aged twenty-one years, tiler. Father and mother in good health. Eight brothers and sisters, of whom five have died before the tenth year. Scrofula; onanism from the age of thirteen, afterwards venereal excess. Periodic migraine every fortnight. Towards the age of eleven years, at la Pitié, M. Michon proposed an operation to remedy a sinking of the left scapula. Towards the age of seventeen, vertigo and cramps. His legs supported him with difficulty; tendency to fall backwards. His speech became difficult. He became humpbacked. Blows with a stick on the back, followed by dull pains for a month. To sum up, during two or three years the above-described phenomena; failures of equilibration, difficulty of pronunciation and gradual deviation of the trunk constituted the whole disease and became more and more pronounced.

Intellectual power excellent; sight and hearing excellent. Neither strabismus nor diplopia. Since the invasion of his disease, no headache, loss of consciousness, nor convulsions. All the movements of the tongue are freely executed; no indication of paralysis of the soft palate. These facts are all the more necessary to note, as the speech is thick, nasal at times, as if the tongue was swollen.

Impressions of cold, of pain, of simple contact, of tickling, as perceived as usual in the face, the trunk, the arms, the legs, the soles of the feet. Muscular sense everywhere unimpaired. Neither numbness, nor tingling, nor pains of any sort. No trembling. Muscular strength very considerable everywhere.

For some months L. has been awkward in his hands, and has allowed objects to fall. When he has his eyes closed he is unable to carry his spoon straight to his mouth, or the point of his index finger to the tip of his nose. In the lower limbs analogous

case (3) and Teissier's (4) present features foreign to Friedreich's disease, and ends by saying that an attentive reading of most of the cases which it has been sought to connect with Friedreich's disease shows that authors like Bourneville may have wished to see in this disease a defaced form of disseminated sclerosis. Judson Bury (69, p. 20), thinks too that the cases of Seeligmüller belong rather to disseminated sclerosis, by reason of the trembling, of the exaggeration of the patellar reflexes, and of the nystagmus which they present.

symptoms. In bed, he can execute all movements. . . With eyes open and without any assistance, even a stick, he can go about all day and can ascend or descend stairs. His legs feel stiff, he says, but not weak. Exercise does not tire him. But from time to time he loses his equilibrium, and would fall if he did not find a support within reach.

When his eyes are closed the difficulty of maintaining equilibrium is more conspicuous. He separates his feet to enlarge his basis of support, and throws his arms about in order to balance himself. His movements are not disorderly. His foot alone, moderately raised, falls heavily on the ground a few inches beyond or within the proper spot. He walks more quickly than he wishes to preserve his centre of gravity. At long intervals, when he is strongly exerting himself, cramps and spasms in his legs and toes. Genital functions, micturition, defæcation, normal.

After having left the hospital some months he entered the Hotel Dieu with increased difficulty of walking, which resembles that of individuals affected with locomotor ataxy. *He would be supposed to be drunk.* He separates his legs widely. Persistence of sensibility in its different manifestations—muscular, cutaneous, and of the mucous surfaces. Left on account of *insubordination.*

Topinard adds (4): "MM. Duchenne and Vigla have decided as to the diagnosis in favour of progressive ataxy; we have not the courage to do so. The peculiarities of gait in this case differ too much from what is seen in this disease, and are too identical with those of Case 25 (4), (p. 20) (cerebellar tumour), the diagnosis of which is not doubtful, for us not to maintain reserve."

This reserve was justified. The case without doubt belongs to Friedreich's disease. The youth of the patient, the special characters of the tabeto-cerebellar gait, the spinal curvature, the absence of sensory disturbances, the integrity of the sphincters, the articulatory difficulty, finally the incoordination—everything concurs to corroborate this diagnosis.

Analysis of Certain Symptoms.

The symptoms of Friedreich's disease offer certain peculiarities which deserve to be examined more closely. I shall not make an exhaustive study of them; I shall limit myself to remark upon some among them which appear to me the most important from the clinical point of view.

The ataxy has a quite special feature in this disease, and it is the observations of the Professor of Heidelberg that the partisans of the motor theory of ataxy always quote to demonstrate that sensory disturbances are not at all necessary for the genesis of inco-ordination of movements.

One might be perhaps spared the long discussions which have taken place, and which do not appear to be near termination between advocates and opponents of ataxy by sensory disturbances, if one had at the commencement clearly distinguisned the cases of Friedreich's disease from those which belong to tabes properly so-called.

In the latter disease, the theory sketched out in 1862 by Charcot and Vulpian* and developed in the following year by Leyden,† appears to explain pretty clearly the ataxy of tabetics in whom the control of sight supplements at least in part the absence of knowledge of position of limbs, which sensory impressions from the periphery habitually give in the healthy condition. The ataxy is then the result of a lesion in the centrifugal conducting paths, the integrity of which is necessary for the co-ordination of movements. It is a *sensory ataxy*. I shall not pass in review the very serious objections which have been made against this theory, but I will say that it is absolutely insufficient to explain the disorders of movements in Friedreich's disease. There cannot be any question here, in fact, of sensory ataxy. The characters which distinguish the ataxy in this affection are precisely, on the contrary, the absence of disturbances of sensation and of the muscular sense, coinciding with a pronounced and generalised motor inco-ordination.

The following fact, which I have often verified in the patient whose case I was just now reporting, appears to me quite characteristic and capable of furnishing on this question a decisive argument against the sensory origin of ataxy in Friedreich's disease.

I have already related in this case that the patient,

* CHARCOT et VULPIAN, Sur un cas d'atrophie des cordons postérieurs de la moelle épinière, &c., *Gazette hebdomadaire de méd.*, No. xviii., May 2, 1862, p. 281.

† LEYDEN, Die graue Degeneration de hinteren Rückenmarksstränge, Berlin, 1862.

GENEALOGICAL TABLE OF A FAMILY AFFECTED WITH FRIEDREICH'S DISEASE.

Musso (*Clinical Review of Bologna*, 1884, p. 865).

MARCHIS–RIVOTTI.

ANTONIA.

Melancholia, terminated by dementia. A brother ataxic.

IGNATIO.

Healthy, but nervous, of feeble constitution; his wife healthy.

CATTERINA.

Wife of Dardino Guiseppe, both healthy.

- Antonia. Hereditary Ataxy.
- Anna. Hereditary Ataxy.
- Three still-born.
- Maria. Hereditary Ataxy.
- Giovanni. Healthy.

7 children.

- Maria.
- Bernardo.
- Antolo. Herditar Atay.
- Domenico. Hereditary Ataxy. Died, aged 40.
- Magdalena.
- Angela.
- Still born.
- Ignazio. Hereditary Ataxy. Died, aged 46.
- Antonio.
- Giovanni.
- Three still-born.

13 children.

First cosins.

NOTE.—The three ataxic boys are descended from the insane grandmother through the maternal line. The three ataxic girls are descended from the insane grandmother through the paternal line.

GENEALOGY OF THE FAMILY VITIELLI AFFECTED WITH FRIEDREICH'S DISEASE.

VIZIOLI (*Giornale di neuropatalogia* anno iii. fasc. 1°e 2°, 1885, p. 15).

FATHER.

ANIELLO VITIELLI.

Died, aged 75, of cerebral apoplexy. Gouty. Good health. Strong up to the evening of his death. A wine drinker, irascible temperament, bright intellect.

MOTHER.

LUIGIA LUISI.

Died, aged 72, of dry gangrene. Several attacks of convulsions in early life. Subject to migraine all her life. Character odd, original.

Sixteen children, of whom three only in good health. Eight ataxic, and two grandchildren ataxic.

- Marianne. Left facial hemiatrophy, congenital.
- Guiseppe. Died, aged 8
- Rosa. Died, aged 26. Paralysis of the four limbs, speech troubles. ATAXY. Was never able to walk.
- Raffaelo. Died, aged 10 months.
- Guiseppe. Died, aged 16, of cerebral hemiplegia. ATAXY. General strength well developed. Kypho-scoliosis.
- Antonio. Hypochondriacal, died aged 40.
- Vincenzo. Paralysis of legs. ATAXY. Speech troubles. Nystagmus. Marries and has five children, of whom two ATAXIC. Onset at two and three years, respectively. Characteristic gait. Two others with insomnia. One healthy, died, aged 6, of scarlatina.
- Raffaelo. Irascible.
- Aniello. Intemperate.
- Lucia. Irritable. Married, no children.
- Filomene. Married. No children. ATAXY of four limbs, with atrophy and paralysis of lower.
- Margharita. Married. Two children, healthy. Did not suckle. ATAXY of four limbs, speech troubles.
- Andrea. Married. Two sons in good health. ATAXY of four limbs, body and limbs imperfectly developed.
- Maria. Paralysis and atrophy of lower limbs. ATAXY.
- Luigi. Healthy, age 30. In no way abnormal.
- Francisco. Died, aged 6, of an intercurrent disease. ATAXY. Was never able to walk.

GOWERS (*Transactions of the Clinical Society*, t. xiv., 1880).

- MOTHER. Choreic.
- FATHER. Bright's Disease.
- UNCLE. Bright's Disease.
- UNCLE. Insane.

Nine children.

- Son. Age 30. ATAXY.
- Daughter. Died. Aged 10.
- Son. Age 35. Healthy.
- Son. Age 33. Healthy.
- Daughter. Age 29. ATAX.
- Son. Age 26. ATAXY.
- Son. Age 23. Healthy.
- Son. Age 22. ATAXY.
- Son. Age 19. ATAXY.

whether his eyes were closed or not, could not touch at the first attempt the tip of his nose or the lobule of his ear with the index finger of one hand when told to do so. Now what appears to me of fundamental importance in order to judge of the nature of the ataxy in this case is that when once his finger touches his face at a certain distance from the point sought, the patient slowly glided his index to the point indicated, *without leaving the surface of the skin.* In thus drawing his finger over the epidermis he evidently guided himself by sensation to remedy his ataxy. The motor incoordination, far from being provoked by sensory disturbances, was on the contrary corrected, thanks to the integrity of the sensory tract.

This character, as well as that of the generalisation of the ataxy, of its propagation from the lower limbs to the muscles of the larynx, of the tongue and of the eyes, prove that the seat of ataxy in Friedreich's disease must be central, placed in the very focus of the centrifugal motor paths of co-ordination of muscular movements—that is to say, in the medullary extension of the cord. It is a *bulbar ataxy.*

Hammond has gone farther. He supposes that the seat of the lesion is in the cerebellum. Most physiologists since Flourens consider the cerebellum as presiding over the functions of co-ordination of movements. Hammond relies on this physiological doctrine to advance the theory that Friedreich's disease has its origin in the cerebellum and the medulla, to be propagated thence gradually to the spinal cord, which is consequently, according to this supposition, secondarily diseased. We shall see, in treating of the pathological anatomy, that the autopsies made hitherto do not allow this view to be adopted. Let us remark, however, from now, that it is contrary to clinical observation, since the symptoms of ataxy are propagated from below upwards, and not from above downwards, as they would be if Hammond's hypothesis had some probability. We have seen, moreover, that the cases of the American Professor do not belong to Friedreich's ataxy, so that the unusual symptoms observed in his patients—vertigo, pains in the back of the

head, &c.—should not be reckoned in the symptomatology of this disease. We shall speak of the choreiform unsteadiness, of the movements of athetosis of the toes, of the articulatory troubles and of the spinal curvature when we shall treat of the diagnosis.

As for *nystagmus*, Friedreich had already made a very profound study of it. He had remarked that the patients had no suspicion at all of their nystagmus, although the oscillations of the ocular globe were very pronounced. Moreover, contrary to what is often observed, the nystagmus in Freidreich's cases was produced when an object was fixed, whilst in the ordinary cases of nystagmus of ophthalmologists, it was then made to disappear, as Professor Seeligmüller has expressly observed in one of his patients. In the case we have described, as we have seen, the nystagmus behaved exactly as Friedreich has described it. We know that the latter writer distinguishes two kinds of ataxy, according as the inco-ordination is observed during movements, or at rest during the continuous contraction of the muscles, which maintains the equilibrium of the body and limbs—locomotor ataxy and static ataxy. The ocular movements also present these two kinds of ataxy.

Nystagmus is a late symptom in Friedreich's disease—one which appears generally several years after the weakness and ataxy of the legs, sometimes even eighteen or twenty years after the onset of the disease. Its importance is great, especially as a diagnostic sign.

The other ocular troubles, so frequent in the tabes of Duchenne, are almost unknown in Friedreich's disease. However, Joffroy ([80]) mentions diplopia and drooping of the upper eylid, and Mendel ([81]) converging strabismus (double paralysis of the sixth pair). In these two cases the pupillary reactions were normal to light, as well as with convergence and accommodation.

Deformity of the foot in Friedreich's disease is a symptom which may exist very early. Rütimeyer relates that they had observed in the families Blattner and Kern the early prominence of the extensor proprius pollicis, which was considered as a sign of very bad augury; the father also

of the children Kern wrote to the doctor that "each time that this traction of the great toe by a sinew" was observed, the parents lost all hope of seeing their child escape the disease.

Friedreich had already observed deformity of the feet in one of his patients, Charlotte Lotsch (his second case): "The group of dorsal flexors of the foot is weakened," says he (the feet are permanently extended by the action of the calf muscles). "*The toes also are permanently flexed,*" and he adds in his second memoir, when speaking of the same case, fourteen years later, "her feet are in a position of permanent equino-varus."

If, therefore, Friedreich has not given a complete description of all the kinds of club-foot which are observed in this disease, we cannot, however, say with Soca that he did not recognise this sign, and we ought to add, in opposition to the statement of Rütimeyer, that he has also proved the existence of dorsal flexion of the toes.

When deformity of the foot is complete, it is accompanied by certain characters which it is important to recognise to differentiate it from other deformities of the feet, especially the tabetic club-foot, as we shall do in the chapter on diagnosis, where we shall more particularly study this deformity.

What distinguishes the club-foot in Friedreich's disease is, first of all, the condition of equinus which results from the predominance of the muscles of the calf over the antero-lateral group of leg muscles, weakened, and even somewhat atrophied. There is indeed a certain degree of foot-drop in these patients, as we have verified in the case we have related. Some authors think that the condition of equinus must be attributed to the contraction of the sural triceps, but we do not experience any resistance in the tibio-tarsal articulation when we raise the foot, and the tendo achillis is not stretched unduly, so that we can readily admit retraction of the calf muscles, but surely not their contraction in the ordinary sense of the word. We must recollect, however, that the contracted muscles behave in a quite peculiar way in tabetics whose reflexes are abolished.

Westphal demonstrated in 1879 that spasmodic phe-

nomena disappear in the combined scleroses, when to the lesion of the lateral columns is added that of the posterior columns, as is the case in Friedreich's disease.

The articulations remain flaccid. Debove has recently quoted an example of it in an ataxic affected with cerebral hæmorrhage with secondary degeneration of the pyramidal tract, and manifest contracture of the paralysed limbs. Now in this case the reflexes were abolished and the joints remained lax on the hemiplegic contractured side. Might there be analogous contractures in Friedreich's ataxics? That is possible, and then we should have the explanation, not only of the equinus, but also of the dorsal flexion of the toes and of the prominence of the tendon of the extensor proprius pollicis, which also is shortened. This is the opinion of Rütimeyer, who explains also by contracture the deviation of the spinal column.

Be it as it may, one could not interpret in a satisfactory manner the club-foot in Friedreich's disease simply by the contractures of certain muscles. The paralytic weakening of the antagonists of the shortened muscles also enters into it largely in part, if indeed it is not the essential cause of it. It may well be that the claw-like character of the toes may be the consequence of paralysis of the interossei, and the excavation of the soles of the feet that of the relaxation of the plantar muscles. There are still, as may be seen, many obscure points in the genesis of this club-foot. The phenomena which are observed in these feet give to this disease a particular stamp, which separates it clearly from ordinary tabes, in which muscular atrophies and arthropathies, which are complications relatively pretty rare, give rise to deformities of the foot of quite another form, and quite another nature.

In Friedreich's disease the club-foot is a usual phenomenon which is, perhaps, in intimate and direct relation with the central lesion of the columns of the spinal cord. In every case the particular form of the foot is absolutely different from that which M. Dejerine* has recently so well

* J. Dejerine, Sur l'atrophie musculaire des ataxiques (névrite motrice périphérique des ataxiques), Paris, 1869, and *Revue de médecine*, Feb., March and April, 1889.

studied in his important work on the peripheral motor neuroses of ataxics, in whom muscular atrophy is presented under the form of equinus with plantar flexion of the toes, and particularly of the great toe—that is to say, in a position precisely the opposite of that which is observed in Friedreich's disease.

The *psychical condition* of Friedreich's ataxics deserves special mention. Some authors think that they always suffer from an arrest of intelligence. The case which we relate proves the contrary. I think it is rather in the temperament that anomalies must be looked for. To several patients great irritability of temper is ascribed. Some are apathetic and indolent, others have a caustic turn of mind. We have already spoken of the impulsive laughter which gives such a singular characteristic to the physiognomy.

Diagnosis.

The clinical study which we have just made of the etiology and the symptomatology of Friedreich's disease allows us to simplify very much the discussion of its diagnosis.

In the present state of our knowledge we cannot admit indeed, as belonging to this disease, either the cases with exaggeration of the tendon reflexes (Seeligmüller, &c.), or the forms with paralysis and contractures (Soca). M. Soca says that Friedreich's disease can appear under three different forms, according to the nature of the motor disturbances, which may be of three kinds: 1st, ataxy; 2nd, paralysis; 3rd, contracture. Clinical observation does not, however, authorise us to distinguish these hypothetical classes. M. Soca himself admits doubts on the form with contractures, and in order to prove the existence of the paralytic form he is reduced to quote some cases of Hammond and the case of Kahler and Pick. We have already given the reasons on account of which we could not consider these cases as belonging to Friedreich's disease. We know that this disease presents itself clinically as a well-defined type of ataxy dating from childhood, and before placing under its

title other forms foreign to this type, we must evidently wait till the study of the combined scleroses of the spinal cord is more advanced, which will, perhaps, permit us to connect with Friedreich's disease certain unusual cases which it would be at least premature to actually class in it.

In keeping, therefore, to the clinical type which we have learned to recognise in the preceding chapter, we shall say that the diagnosis of Friedreich's disease is based upon the following characters:

Ataxy and weakness of the legs developing gradually in childhood, almost always without pains, but often simulating choreiform movements. Very slow progressive course, from below upwards, of the disease, which attacks successively the trunk, the arms, the muscles of the larynx, of the tongue, and of the eyes. Ataxic and trembling gait, becoming gradually aggravated. No remissions.

Speech slow, drawling, scanned, as in disseminated sclerosis. Static and dynamic nystagmus. Abolition of knee reflexes. Scoliosis. Peculiar deformity of the foot.

To which must be added negative symptoms which are of very great importance:

Absence of lightning pains. Integrity of cutaneous sensation in all its manifestations. Normal reaction of pupils. Integrity of sight. Absence of genito-urinary troubles.

No syphilitic antecedents. *Family* character of the disease. These are the two principal etiological conditions.

The differential diagnosis will bear essentially on the following affections:—

1. Chorea.
2. Tabes.
3. Disseminated sclerosis.
4. The other combined scleroses of the spinal cord.

I.

Friedreich's disease can be confounded with *ordinary chorea* only at its onset, before the ataxic movements have become sufficiently developed in the lower limbs to interfere with walking. This error of diagnosis, we have already

said, has been made in several cases. We must not, therefore, lose sight of the possibility of a similar mistake, and we shall take care in the cases of chorea which appear suspicious by their manner of walking and their duration, to examine the nature of the ataxic choreiform movements, which are never unilateral in Friedreich's disease, and generally do not invade the facial muscles. We shall assure ourselves also of the presence or the absence of the tendon reflexes, and we shall not neglect to examine the feet, for one of the first signs of Friedreich's disease may be, as we have seen, the dorsal flexion of the great toe, with prominence under the skin of the tendon of its proper extensor.

II.

Locomotor ataxy and Friedreich's disease are distinguished one from another by the most decided characters. Tabes is an affection of adult age; Friedreich's ataxy a disease of childhood. It would not, however, be necessary to go so far as to say with M. Soca: "A child or a youth, who is ataxic is a case of Friedreich's disease. This is not absolutely true, but it is sufficiently exact for practical purposes."

Although tabes of adolescents may be a rare disease, there exists, however, now a sufficient number of authentic cases to invalidate this proposition. On the contrary, it is very necessary to guard against making a like diagnosis; and before admitting that an ataxic child may be affected with Friedreich's disease, it is necessary to search with much care for the presence of the symptoms which characterise this disease. We know that all kinds of cases of infantile ataxy have been too readily attributed to Friedreich's disease, without selection and without criticism, at the risk of spoiling the clinical picture of it.

The tabes of children, if we are to judge of it by the cases which have been published, does not differ from tabes in the adult. The ocular symptoms, the sensory disturbances, the lightning pains, the paræsthesiæ, the gastric crises, and the

urinary troubles, furnish sufficient differential characters to prevent any hesitation in the diagnosis. The importance of syphilis as an etiological antecedent has quite recently been emphasised by a case of Professor Strümpell at Erlangen It relates to a little girl with hereditary syphilis, who was affected with tabes and general paralysis at the age of thirteen years.* The same holds good in the case of the little patients of E. Remak,† who suffered from hereditary syphilis, and all three presented atrophy of the optic nerve without ataxy. Dr. Berbez,‡ who relates fourteen cases, concludes that in early tabetics, heredity is strong, and that intensity of the disease and multiplicity of symptoms are the necessary accompaniments of early tabes. He remarks also after the teaching of M. Charcot, that, although essentially hereditary, early ataxy has nothing in common with the hereditary ataxy of Friedreich.

MM. Gombault and Mallet§ have quite recently published a case of tabes occurring in childhood, with an autopsy, the report of which appears to us particularly instructive for the discussion of the differential diagnosis of Friedreich's disease and locomotor ataxy. Here is a *résumé* of this report.

Man, aged fifty-seven, infirm from childhood, died a few months after entering the hospital. The patient has, therefore, been only a very short time under observation at the close of his life. Inquiries as to etiology are almost entirely wanting. His father died when the child was seven years old and was already infirm. There is no information as to him. His mother died of acute mania. The patient was placed in a hospital because, it was said, he was permanently insane, and refused to take food. All that we can learn of his history is that the onset of his disease dated from childhood, that he was not seven years old when he fell ill, and that at ten years of age he was definitely admitted as

* A. STRÜMPELL. Progressive Paralyse mit Tabes bei einem 13 jährigen Mädchen. *Neurologisches Centralblatt*, March 1, 1888, no. v., p. 122.

† E. REMAK, Drei Falle von Tabes im Kindesalter. *Berliner klinische Wochenschrift*, 1885, no. vii., p. 105.

‡ P. BERBEZ, Tabes précoce et hérédité nerveuse. *Progrès médical*, no. xxx., July 23, 1887, p. 59, *cf.* W. JACUBOWITSCH, Tabes dorsalis im Kindesalter. *Archiv für Kinderheilkunde*, V Band, 1884, p. 187.

§ Albert GOMBAULT et MALLET. Un cas de tabes ayant débuté dans l'enfance. Autopsie. *Archives de médecine experimentale*, first year. no. iii., May, 1889, p. 385.

a patient in an asylum. From that period continuous and progressive course of the disease, which consists principally in disturbance of the voluntary movements, involving the four limbs. The patient could not maintain the erect position unless he held on to the furniture, and kept looking at his feet. He fell if his eyes were closed. At the time of admission to the infirmary flaccid paralysis of the lower limbs; absence of tendon reflexes; motor inco-ordination of the upper extremities; inability to appreciate the position of the limbs; muscular atrophy; muscles of the thigh and of the leg altogether atrophied; ape-like hands; pronounced scoliosis, the date of development of which cannot be stated, because the patient does not remember it. Did it exist in childhood? Diplopia; slight nystagmus; profound disturbance of sensibility in all the limbs; manifest retardation of tactile and painful sensations; anæsthesia; analgesia; thermo-anæsthesia; sensibility is intact only in the face; no trembling; no brain disturbance; no incontinence of urine or of fæces; pains from time to time, not definitely of the lightning character.

Talipes equino-varus of the bedridden; white and soft œdema of the malleoli; speech distinct and easy; no hallucination; penis and testicles fully developed; scar on the sacrum; death.

Autopsy.—Calcareous plates in the arachnoid, all along the cord at its posterior part, especially in the dorso-lumbar region. Issuing from the lumbar enlargement, the posterior roots are bulky, translucent, gelatinous, nodulated, covered with an abundant meshwork of vessels. The anterior roots also are very much hypertrophied. Spinal cord altogether bulky; grey condition and gelatinous appearance of the posterior columns. Peripheral nerves bulky without nodosities, vascular, rose-coloured, translucid.

The grey matter is much reduced in size, atrophied, and its shape has altered. It is thin and delicate in the lumbar region, which one would take at first sight for a section of the cervical region. Antero-lateral columns relatively large. They are furrowed by connected perivascular tracts larger than in the normal condition. Posterior columns small, and stained of a rose colour by carmine, especially at the inner part of the posterior zones. Sclerosis of the posterior columns. In the posterior cornua diminution, almost disappearance, of the bundles of fibres belonging to the posterior roots. In the anterior cornua diminution in number and atrophy of the large motor cells.

Vesicular column of Clarke normal in the dorsal and dorso-lumbar region. The pia mater is everywhere notably thickened.

In the cervical region, sclerosis of the columns of Goll; the posterior root zones are sclerosed in this region only here and there.

Hypertrophic sclerosis of the peripheral nerves is much more developed than in the spinal roots. An almost complete disappearance of the myeline has been observed in the peripheral nerve-tubes which have been examined.

In the discussion of this case, the authors say that it is hardly necessary to point out the characters which differentiate it from Friedreich's disease. However, when we read the description of symptoms which they give, we cannot help thinking that the patient of MM. Gombault and Mallet presented in the course of his disease several symptoms absolutely analogous to those of Friedreich's disease:—motor inco-ordination commencing in childhood; abolition of tendon reflexes; course of the affection slow, unobtrusive, continuous and progressive, attacking the four limbs, without pains at the commencement; talipes equino-varus. Have we really in this case the club-foot of the bed-ridden, as some authors think? Deformity of the vertebral column:—MM. Gombault and Mallet remark in reference to this deformity, that it frequently accompanies various generalised affections which have commenced in childhood. But we have now to find out the differential diagnostic signs which allow us to distinguish this particular case from Friedreich's disease, and scoliosis is precisely a character common to the two cases, the importance of which must not be minimised. Nystagmus:—it is true that the authors quote the absence of nystagmus (*loc. cit.*, p. 403) as contributing to exclude Friedreich's disease in their case. They have forgotten that some pages before (*loc. cit.*, p. 387) they themselves have expressly mentioned the existence in their patient of a "slight nystagmus." Finally, the integrity of the sphincters: That is quite as many symptoms as one could refer to Friedreich's disease. And when the authors affirm that the complete absence of cerebral symptoms (mental symptoms being necessarily put on one side) bears witness in their case against Friedreich's disease, they forget again that they have mentioned diplopia (*loc.*

cit., p. 387) in the antecedents of their patient. What, then, are these cerebral symptoms which characterise "the now classical description of Friedreich's disease"? It is precisely, on the contrary, in the absence of these symptoms that we must find a character of the first order to distinguish this disease from ordinary tabes. The authors add, it is true, "and in particular, absence of articulatory troubles," showing thus that they include these troubles among the cerebral symptoms. But those with nystagmus are the only really classical bulbar symptoms which belong incontestably to Friedreich's disease!

We must seek elsewhere for the fundamental reasons which do not allow us to admit Friedreich's disease in the case of MM. Gombault and Mallet. They are especially the generalised muscular atrophies, and the intensity and the extent of the sensory disturbances. These are the essential symptoms which absolutely exclude, in this case, Friedreich's disease.

The authors, who found at the autopsy the peripheral nerves more diseased than the spinal roots, put forward the fairly probable hypothesis of the possibility of a polyneuritis followed by an affection of the cord. Perhaps, say they, we have to deal in this case with multiple peripheral primary neuritis, with consecutive involvement of the spinal cord in the course of development. They arrive, however, at the provisional conclusion, "that we must consider the case as an example of locomotor ataxy of spinal origin, developed in early age."

There is a hypothesis which pleases us more, and we have been struck by not seeing it expressed in the interesting work of MM. Gombault and Mallet. We shall willingly admit in this case a syphilitic affection, perhaps congenital. We know how frequent syphilis is in the hereditary antecedents of tabetic children—we have just given several examples of it. It would therefore be quite possible that syphilitic infection may have also played its part in the patient of MM. Gombault and Mallet. We must not forget either that there existed in this case the remains of a very pronounced anterior poliomyelitis, which also complicated the symptomatology of it.

Let us return to the differential diagnosis of locomotor ataxy and Friedreich's disease, and let us prove first that the symptoms which they offer in common, when examined more closely, are not at all alike.

The ataxy itself presents considerable differences in the two diseases, and the footprints show that the gait is far from being identical in tabes and in Friedreich's disease. In the latter affection the patient progresses in a very irregular zig-zag manner, instead of keeping in a straight line as tabetics do. Nothing displays better than the method of taking footprints, the fundamental difference which exists between the clubfoot of Friedreich's disease, and the deformities of the foot in tabes. The tracing of tabetics affected with arthropathies of the feet shows the print of flat feet, whilst that of the clubfoot of Friedreich is characterised by the complete absence of the print of the sole of the foot corresponding to the excavation which we have described. M. Joffroy (86 and 103) judiciously remarks, moreover, that the deformity of the foot is never developed in tabetics, except when the patient has been for some time confined to bed, whilst in Friedreich's disease the deformity of the foot exists in the patient who is able to walk, long before he is confined to bed.

Romberg's sign discloses also, as we have observed, a fundamental difference in the two diseases. Whilst in tabes closure of a patient's eyes makes him lose his equilibrium, because he has no longer knowledge of the position of his limbs, in Friedreich's disease it is in consequence of choreiform instability. The patient sways whether his eyes are opened or closed, but in the latter case having lost the control of vision, the swaying movements easily pass the limits of equilibrium, and he falls, although he may have preserved the notion of the position of his limbs.

Must we consider, with M. Joffroy, diplopia as a symptom that may belong to the picture of Friedreich's disease (103). We should not dare to speak affirmatively on this point. Diplopia, indeed, in M. Joffroy's cases has been transitory. It is noted in the history of the patient, who belonged to a tuberculous family, but it was not directly observed, we

think, in the medical examination. In every case it does not appear to have presented the lasting characters which it sometimes manifests in tabes. Transitory diplopia is met with in a great number of nervous affections. It is not rare in certain neurasthenics. It has been several times seen in cases of poisoning by tainted meat. Prof. Bouchard* and many other authors have pointed it out in dilatation of the stomach, in which we have many times proved it. It would not, therefore, be right to conclude from its presence in the history of a patient suffering from Friedreich's ataxy that it necessarily forms a part of the assemblage of symptoms in this affection. We must wait for further observations before admitting diplopia as a definite symptom of Friedreich's disease, of the same importance as in locomotor ataxy. And even, if it ever is admitted as such, it will not remain less exceptional in the first, whilst it is usual in tabes.

We shall not insist any further on the differential diagnosis of these two diseases, which is now well known, and does not offer any difficulty in ordinary cases.

A word only on nystagmus, very frequent as is known in Friedreich's patients, and which was thought never to exist in tabetics. It is observed, however, very clearly in some cases of tabes, but it is true that these cases are very rare. M. Gilles de la Tourette has published three cases of it. I have seen it myself in two cases, one of which is briefly reported in the note on my observations on suspension in tabes.†

In the following comparative table is a résumé of the differences which exist between the two diseases, the symptoms being put parallel :—

Friedreich's Disease.	Tabes.
"Family" character. Several children are usually attacked in the same family.	Rarely more than one tabetic in the same family.
Scarcely ever syphilis in the antecedents.	Syphilis is usually found in the history, either acquired or hereditary.

* Ch. Bouchard, Leçons sur les auto-intoxications dans les maladies. Paris, 1887, p. 174.

† Ladame, De la suspension dans le tabes. *Rev. méd. de la Suisse rom.*, June 20, 1889, p. 347.

The disease begins in childhood. There are no authentic cases known of onset after the 20th year.	Onset almost always after the 20th year. Infantile tabes is most often of syphilitic origin.
No lightning pains either at the onset or during the course of the disease. They have been only exceptionally observed, and in these cases are they really lightning pains?	Lightning pains perhaps preceding ataxy for several years. Scarcely ever absent in tabes.
No preataxic stage. The commencement of the disease may simulate a slight chronic chorea for several years.	Long preataxic stage, presenting the most diverse symptoms: ocular, visceral, &c.
Ataxy during rest and during movement. Choreiform instability. Movements incoordinate, feeble and not sudden. Ataxy is general. It progresses slowly from below upwards. Gait, tabeto-cerebellar, ataxic and festinating. Closure of eyes causes loss of equilibrium only by reason of the choreiform instability, due to static ataxy.	Ataxy during movement. At rest the patient is quiet. Ataxic movements violent, sudden, as of a spring which is let go. The ataxy may be localised in the legs or in the arms (cervical tabes). Gait plainly tabetic. The patient follows a straight line. Closure of the eyes causes loss of equilibrium, because it takes away the knowledge of the position of the limbs (Romberg's sign).
Integrity of sensation in all its manifestations. Towards the end of the disease only, in some cases, slight affections of the cutaneous sensation.	Anæsthesia and hyperæsthesiain patches. Paræsthesia. Retardation of transmission of painful sensations and other perversions of sensation.
Absence of knee reflexes. Cutaneous reflexes normal. Plantar reflexes rather exaggerated on account of the great sensibility of the soles of the feet.	Absence of knee reflexes. Cutaneous reflexes often weakened or abolished on account of affections of sensation; the plantar reflexes especially are frequently absent.
Integrity of the special sense organs. Horizontal nystagmus, static and dynamic, is rarely absent. Very rarely affections of the muscular innervation of the eyes.	Grey induration of the optic nerve. Amaurosis. Sclerosis of the auditory nerve. Buzzings in the ears. Menières' vertigo. Myosis and Argyll-Robertson sign. Pareses of muscles of the eye; diplopia, ptosis. Scarcely ever nystagmus.
Absence of visceral affections.	Laryngeal crises (spasms of the glottis). Gastric, intestinal, anal, vesical, nephritic, crises.

No lesions of the skin, the joints or the bones. Club-foot of special kind, habitual, in consequence of pareses and muscular contractures. Athetosis movement of the toes in walking. No peripheral neuritis.	Cutaneous lesions (zona). Trophic affections of the nails. Shedding of the teeth. Spontaneous fractures. Arthropathies. Tabetic foot. Muscular atrophies. Peripheral neuritis, sensory and motor. Other trophic troubles. Perforating ulcer of the foot.
Integrity of the genito-urinary functions and of the sphincters.	Genito-urinary troubles. Impotence. Paresis and hyperæsthesia of the bladder. Retention and incontinence of urine. Constipation.
Scoliosis.	No spinal curvature.
Speech slow, scanned.	No speech troubles.
Progressive, uniform, very slow development of the disease, without crises or prominent episodes.	Pretty often relatively rapid development of the disease by successive crises, followed by obvious aggravation, with numerous fresh symptoms from time to time.

III.

In the memoir of Marie,‡ and in the more recent one of Unger,* will be found very complete bibliographic notices on *disseminated sclerosis* in children, to which we shall add the work of Professor D'Espine, of Geneva.† It is seen from these works that disseminated sclerosis most often presents in children the cerebro-spinal type which commences by convulsions. The only positive symptoms which may be common to this affection and to Friedreich's disease are the nystagmus and the slow and scanned speech. But these troubles are late in Friedreich's disease, and only appear long after the ataxy, the loss of knee reflexes and the other symptoms which cannot leave any doubt as to the diagnosis,

‡ PIERRE MARIE, De la sclérose en plaques chez les enfants. *Revue de Médecine*, 1883, p. 536. *See also* MONCORVO, Contribution à l'étude de la sclérose multiloculaire chez les enfants, Paris, 1884.

* L. UNGER in Wien, Ueber multiple inselförmige Sclerose des Centralnervensystems im Kindesalter, Leipzig und Wien, 1887.

† A. D'ESPINE, Deux formes de paralysies chez les enfants. *Revue médicale de la Suisse romande*, March 20, 1889, No. 3, p. 129. *See also* D'ESPINE et PICOT, Manuel des mal. de l'enf., fourth edition, p. 441.

whilst the exaggeration of the reflexes, the rigidity of the legs, the contractures, the trembling on voluntary movement, the disturbances of the motor innervation of the eyes, diplopia, nystagmus, strabismus, &c., are the first signs of disseminated sclerosis. Moreover, all the other symptoms so characteristic of disseminated sclerosis, apoplectiform and epileptiform attacks, vesical paralysis, trophic and mental disturbances, frequent remissions in the course of the disease, all these are invariably wanting in Friedreich's disease, as this author himself had already plainly stated.

Friedreich's Disease.	Disseminated Sclerosis.
Family character of the disease. Insidious onset.	Case isolated in a family. Sudden onset, often with convulsions.
Very slowly progressive course of the affection. No remission of symptoms.	Frequent remission of symptoms. Irregular course of the disease.
Choreiform instability. Athetosis movements of the toes. Paresis of legs, with loss of tendon reflexes. Progressive ataxy of the four limbs.	Trembling on voluntary movement. Spastic paresis. Exaggeration of tendon reflexes. Ankle-clonus. Contractures.
Tabeto-cerebellar gait. Integrity of special sense organs and of the ocular muscles. Late nystagmus.	Spastic gait. Paralysis of ocular muscles. Diplopia. Amblyopia. Early nystagmus.
Scoliosis. Special club-foot. integrity of genito-urinary organs and of the sphincters. No visceral tabetic troubles.	Congestive, apoplectiform and epileptiform attacks. Vesical paresis and other tabetic symtoms, generally slightly marked. Mental disturbances.

Characters Common to the two Diseases.

Absence of sensory disturbances, slowness of speech, scanning of words. Nystagmus.

Vertigo has also been noted as a symptom common to the two diseases, but we do not yet know if vertigo really belongs to this affection, although it may often have been mentioned in reports, since the first case described by Friedreich. Our patient has never experienced vertigo.

IV.

Combined Sclerosis of the Spinal Cord.‡ We shall not undertake here the study of the subject of the combined sclerosis, which is, perhaps, really the most obscure of spinal pathology. From the anatomical point of view, Friedreich's disease is a combined system sclerosis of the posterior and lateral columns, which could not, in many cases, be distinguished, as regards its spinal localisation, from other combined sclerosis, having a like anatomical seat, with very different clinical symptoms. It is evident that the special evolution of the lesions, their variable extent and the successive separate invasion of different systems of fibres, have a decisive importance for the symptomatalogy. Friedreich's disease which evolves on an incompletely developed cord, forms a very distinct group, and henceforth is classed among the combined system scleroses.*

In spite of the anatomical lesions which it presents, we yet do not think, with Professor Grasset, that Friedreich's disease can be put in the category of "combined tabes." We think it would be preferable to reserve this appellation for the cases of tabes which are complicated by a diffuse sclerosis of the lateral columns, most often following a posterior spinal meningitis, meningo-myelitis by propagation, as Friedreich and M. Dejerine† have proved it. As for the combined scleroses properly so-called of the postero-lateral columns, with primary system lesions of the posterior columns and pyramidal tracts, we actually possess a number of cases of them sufficient to sketch out the

‡ *See also* J. L. PREVOST, Sclérose des cordons postérieurs, compliqué d'une sclérose systématique des cordons latéraux. *Archives de Physiologie*, 1877, t. iv., p. 764. V. BABESIU, *Virchow's Archiv.*, 1879, vol. lxxvi., p. 74. EDES, *Boston Med. et Surg. Journal*, September 21, 1882. RAYMOND, *Arch. de Physiol.*, 1882, x., p. 456. DAMASCHINO, *Gazette des Hôpitaux*, No. 1, 1883. HOPKINS, BRAIN, October, 1883. ORMEROD, id. April, 1885. BABINSKI et CHARRIN, *Revue de Méd.*, 1885, November, p. 962. MASSALONGO, *Medic. Contemp.*, October and November, 1886. BORGHERINI, *Riv. sper. d. Freniat*, XIII., p. 137, 1887, &c., &c.

* Among recent treatises that of Professor L. HIRT of Breslau (*Pathologie und Therapie der Nervenkrankheiten*, Wien, 1890, p. 364), is to our knowledge the only one in which Friedreich's disease is separated from tabes and placed among primary combined scleroses.

† DEJERINE, *Archives de Physiologie*, 1884, iv., p. 456; *Semaine Médicale* 1886, No. 18, p. 181.

symptomatology of them. There exist, doubtless, very considerable differences in the symptoms described by observers in the different cases that we know. Nevertheless, we can provisionally distinguish from the clinical point of view, four or five categories of patients affected with combined scleroses, which belong to the following types (without mentioning syphilitic myelitis and others, which give rise to secondary scleroses more or less diffused, and which may simulate system scleroses) :—

I. *Ataxic paraplegia* (Gowers‡). In this we may have all the usual symptoms of tabes with predominance of paralytic weakness of the legs belonging to sclerosis of the lateral columns. If the lesion of the posterior columns stops in the dorsal region, or even if it descends in the lumbar region without attacking the posterior root zones, the knee reflex will be intact, or even exaggerated, in consequence of the sclerosis of the pyramidal tract (Westphal*).

II. *Spastic tabes dorsalis* (Charcot, Erb, Strümpell). On several occasions combined sclerosis of the postero-lateral columns has been discovered at the autopsy of patients having presented the clinical picture of spastic spinal paralysis, whenever sclerosis of the pyramidal tracts involved all the lumbar region.

III. *Ataxo-spastic tabes* (Grasset†). A mixture of the symptoms of locomotor ataxy of Duchenne with those of spastic tabes.

IV. *General paralysis.* Westphal has shown that primary sclerosis of the lateral columns combined with system sclerosis of the posterior columns is especially frequent in general paralytics. According to this observer primary

‡ R. GOWERS, Clinical Lecture on Ataxic Paraplegia. *The Lancet*, 1886, vol. ii., p. 1, 61 and 130.—DANA, Progressive spastic ataxia (combined fascicular scleroeis) and the combined scleroses of the spinal cord. *The Medical Record of New York*, 1887, July 2, vol. xxxii., No. 1, p. 1. DANA, Ataxic Paraplegia, BRAIN, January, 1889, p. 490.

* WESTPHAL, *Virchow's Archiv*, Band XXXIX. and XL.; *Archiv. für Psychiatrie.* Band V., p. 803, Bd. VIII., p. 469, Bd. IX., p. 413 and 691, Bd. XV., p. 224. *See also* STRÜMPELL, Ibidem, Band XL. p. 27, Band XVII. (1886), p. 217. DEBOVE, De l'hémiplégie des ataxiques, *Progrès Médical*, 1881. No. 52, p. 1021, No. 53, p. 1042.

† GRASSET, *Archives de Neurologie*, 1886, t. xi., p. 156 and 380, t. xii., p. 27. Cf. BALLET and MINOR, Ibidem, 1884. t. vii., p. 44.

isolated sclerosis of the lateral columns has hitherto been certainly observed only in general paralysis.‡

We have no need to go into the arguments which allow us to make the differential diagnosis between Friedreich's disease and the combined scleroses of the spinal cord which belong to the four preceding groups. This diagnosis can offer no difficulty.

It is not the same with a final group in which may be included all the cases which do not enter into the above categories. But it is here precisely that investigations and authentic cases are wanting. Some facts might raise a suspicion that there exists perhaps a form of Friedreich's disease peculiar to adult age; but we are still reduced on this point to a simple hypothesis which requires confirmation. Under these circumstances there can be but one course to follow. We must keep rigorously to the symptomatology of Friedreich's disease such as we know it, with its hereditary characteristics, and eliminate from it as we have done, all the cases which do not exactly correspond with it. The analogies, still very obscure, of pathological anatomy, ought not to serve us as a guide in these questions, but much rather clinical observation, till a deeper study of the combined scleroses, system or diffused, will better enable us to enlighten ourselves in these delicate and complicated researches of spinal pathology.

The report presented by Dr. Oppenheim at the meeting of the Society of Psychiatry of Berlin, Nov. 12, 1888, is particularly instructive on this point.* It is a case of combined sclerosis of the spinal cord in a child aged 15 years, who suffered from a progressive affection of sight from the age of 10 years, after measles, but whose other spinal symptoms only appeared at the age of 12 years, after a fall in the water. At first choreiform movements in the left half of

‡ ZACHER, Beiträge zur Pathologie und pathologischen Anatomie der progressiven Paralyse. *Archiv. für Psychiatrie*, Bd. XIV. 1883, p. 463, Bd. XV., 1884, p. 359. SIOLI, Ein Fall von combinirter Erkrankung der Rückenmarkstränge mit Erkrankung der grauen Substanz. *Archiv. für Psych.*, vol. xi., 1881, p. 638.

* OPPENHEIM, Ueber einen Fall von combinirter Erkrankung der Rückenmarkstränge im Kindesalter. *Neurologisches Centralblatt*, December 1, 1888, No. 23, p. 647.

the body, especially in the face and the left arm, then weakness and slight contracture in the joints of this arm. A second group of symptoms is allied to the clinical picture of tabes. Double optic atrophy. Pupils fixed. Ptosis on the right side. Absence of knee reflexes. Weakness and slight ataxy of the limbs. Romberg's sign. No sensory nor sphincter troubles. Death from bedsore. Sclerous atrophy of pyramidal tracts and of the columns of Goll and Burdach. The lesion of the posterior columns was less marked than that of the lateral columns.

We see from the above description of symptoms how easy it was to exclude Friedreich's disease in this case. Oppenheim's case shows that there exists a combined sclerosis of the postero-lateral columns in children with symptoms in great part tabetic, and essentially different from those of Friedreich's disease. This case would be sufficient to distinguish clearly the latter disease from the combined scleroses, and to give it a place apart in the primary system lesions of the spinal cord, in spite of the anatomo-pathological analogies which bring it near to other combined scleroses.

Pathological Anatomy.

The number of autopsies which have allowed us to fix the anatomical lesions of Friedreich's disease is still very restricted, and if one can say henceforth that these lesions belong to the group of primary combined system scleroses, there yet remains no less obscurity on their nature and their precise localisation.

We have altogether nine autopsies of authentic cases, of which five belong to Friedreich's patients, one to Everett Smith, one to Newton Pitt, and two to Rütimeyer.

Friedreich ([11]), who had himself made the autopsy on four of his patients, arrives at the following conclusions :—

The "hereditary" character of the disease is explained, says he, by an arrest of development of the spinal cord. The volume of this organ was very much reduced in all the cases in which an autopsy was made. The spinal cord was

slender, flattened, and as if atrophied in its entirety. It was not difficult to convince oneself that the bulb, and also the cord, had not reached their complete development, so that the sclerosis degeneration of the posterior and lateral columns had invaded an organ still incompletely developed. This is confirmed by the discovery of Dr. Schultze, who found, on microscopical examination, the nervous elements of the healthy portions of the cord very small and very slender.

At the first autopsy Friedreich saw only sclerosis of the posterior columns. The anterior and lateral columns are noted as healthy. In the three others the invasion of the lateral columns by the sclerosis has been clearly determined. But what especially struck Friedreich is the posterior spinal meningitis, which is the true cause, according to this author, of the propagation of the chronic inflammation from the posterior columns to the lateral columns. That is why, says he, in the lower portions of the cord, where the meningitis is more recent, the margin of the lateral columns is less degenerated than in the cervical portion, where all the periphery is degenerated like a ring. We know now that this explanation of Friedreich has no longer much value. In fact, the difference which he had proved between the extent of the cortical degeneration in the lumbar and cervical regions proceeds from this, that in the latter the direct cerebellar tracts are degenerated; there we have a primary system sclerosis which forms part of the combined lesions of Friedreich's disease, the significance of which was completely ignored at the time of the first autopsies made by this eminent observer.

It is true that in Friedreich's cases, and especially in the one, the autopsy of which was made by Schultze (see later), the cortical degeneration went appreciably beyond the region of the direct cerebellar tracts. But the lesions of the cord were examined in these cases only after the disease had lasted twenty-three and thirty years, whilst in Rütimeyer's cases, the duration of which was only thirteen and nine years, the lesions were confined, as we shall see, to these tracts. We must therefore suppose that in Friedreich's

cases the sclerosis has extended itself, and has gone beyond its first limits owing to the long duration of the disease, so that the diffuse cortical degenerations may be regarded as accessory, having slowly developed themselves, perhaps under the influence of the slight spinal meningitis, and at length complicating the primary system changes.

The cells of Clarke's columns are in intimate relation with the direct cerebellar tracts, of which they represent the trophic centres. Now Friedreich had already remarked that there existed in one of his cases a double canal in the dorsal region of the cord. This double canal, says he, is probably nothing else than the degeneration of Clarke's columns, a degeneration verified in all the other cases, in which the number of Clarke's cells was very much diminished, sometimes even reduced to a solitary one in the lower dorsal portion, that is to say in the region in which they are usually most developed.

A fifth patient of Friedreich was examined post-mortem by Dr. F. Schultze (20), who found lesions similar to the preceding ones: grey degeneration of the posterior columns and of the posterior portions of the lateral columns; flattening of the cord from before backwards; slight thickening of the pia mater. Small size of the healthy portions of the cord. As a difference no circular complete cortical degeneration in the upper cervical region, but degeneration of the inner parts of the anterior columns along the longitudinal fissure extending up to the pyramids. Degeneration of the columns of Clarke. Posterior roots atrophied; integrity of the ganglia of the medulla oblongata.

Everitt Smith gives drawings of sections of the spinal cord of his patient, and describes the anatomical lesions as follows:—

Spinal meninges injected and adherent to the bony walls of the canal. The cord presents an asymmetric atrophy, it is pale and softened in places. The dorsal and lumbar regions, carefully examined, show marked sclerosis of the posterior columns in their whole extent, with the exception of a small part, relatively healthy immediately behind the posterior commissure. Sclerosis of the direct and crossed

pyramidal tracts less intense than in the posterior columns. Thickening of the pia mater; atrophy of the posterior roots; the anterior normal. Ganglion cells of the anterior and posterior cornua less numerous than normally, and showing in a high degree degenerative change.

Numerous figures demonstrate the seat and extent of the combined sclerosis in Dr. Pitt's case (74), the autopsy of which is described with the most minute details. The author gives a series of comparative measurements of a healthy cord and of the diseased cord, which prove how much smaller the sections of the latter were than those of the other throughout the length of the cord. Posterior roots very slender also, especially in the cervical and lumbar enlargements. Nowhere opacity or thickening of the meninges, which were found completely normal. Grey degeneration of the posterior and lateral columns. The hardened cord was examined by Weigert's method. We shall not reproduce the interesting details which Dr. Pitt gives of each region examined, we shall limit ourselves to presenting a *résumé* of his conclusions (*loc. cit.*, p. 389).

I. The spinal cord is extremely slender.

II. Extreme sclerosis of the columns of Goll in their whole length from the lumbar enlargement to their termination in the floor of the fourth ventricle.

III. Intense sclerosis of the posterior part of the columns of Burdach, in which, however, some healthy fibres are scattered, in greater number at the upper part of the cord.

IV. The zone of the columns of Burdach, which bounds the cornua and the posterior root, is intact. (This region bears now the name of marginal zone of Lissauer).

V. Sclerosis not distinctly limited (much less intense than in the posterior columns) of the crossed pyramidal tracts.

VI. Sclerosis of the ascending cerebellar tracts, visible up to the decussation of the pyramids. Very slight and irregular sclerosis of a few scattered fibres in the antero-lateral columns, especially at the periphery, and in some sections along the anterior fissure.

VII. Degeneration of the columns of Clarke in some sections.

VIII. Degeneration of some fibres of the posterior roots, and of the posterior cornua.

IX. Friability and shrivelling of the affected regions.

What was most striking to the naked eye was the great diminution of the transverse section of the cord, as well marked in the cervical enlargement as in the dorsal region and in the lumbar enlargement, the posterior portions being proportionally more diminished than the anterior.

Columns of Goll extremely sclerosed. Numerous healthy tubules scattered in the columns of Burdach and in the anterior third of the posterior columns. A fasciculus almost intact along the posterior cornua. Sclerosis of the crossed pyramidal tracts, and of the direct cerebellar, less pronounced. A slight annular degeneration extends from the periphery to the anterior part of each side of the anterior fissure.

The most marked sclerosis is always in the posterior columns. Posterior nerve roots in part degenerated. In the lateral columns the sclerosis is limited to the crossed pyramidal tracts, and to the direct cerebellar tracts. No degeneration around the posterior commissure. Clarke's columns are sclerosed.

Posterior columns almost completely sclerosed. Anterior quite normal. The triangular sclerosed patch of the lateral column corresponds exactly with the crossed pyramidal tract in this region. It is separated from the posterior root by a band of healthy tissue (marginal band of Lissauer).

Dr. Pitt remarks finally on the frequency among these patients of cardiac troubles, which have not yet been noticed by authors, who give for the most part little attention to abnormal conditions of the circulation and of the blood-vessels, which are frequently degenerated, according to Pitt, in Friedreich's ataxics.

In favour of an association of Friedreich's disease with a tendency to vascular degeneration and to the imperfect development of the spinal cord, Dr. Pitt draws attention to the following points:—

1. That the posterior columns which are the most vascular, have always been found smaller and more atrophied than the rest of the cord;

2. That vascular lesions have been demonstrated in several cases, and that the disease has always been much aggravated after acute intercurrent affections;

3. The development of the cord begins in the most vascular regions, so that the degenerations attack by preference the parts which are developed in the last place. These parts have also the least important functions, and, according to the general law of Hughlings Jackson, these are the first which degenerate;

4. The sclerosed regions correspond to the tracts, the filaments of which are covered with myeline after the 5th month of fœtal life;

5. The symptoms become more marked at the time of puberty, which is accompanied, as we know, by great vascular modifications in the organism.

The two most recent autopsies are those of Dr. Rütimeyer, of Bale (7). By the courtesy of this distinguished writer I have been able to examine with him the interesting series of sections which he has made in his two cases. I can, therefore, speak with authority, and I embrace this opportunity to address again my thanks to Dr. Rütimeyer. The following is a résumé of these two autopsies:—

1. H., age twenty years, died rather suddenly of general weakness after 13 years' illness. Medulla oblongata and spinal cord very much atrophied throughout. Thickening of the meninges and multiple adhesions, especially in the lumbar region. Posterior roots very much thinned. Firm consistence. With the naked eye grey degeneration of the column of Goll is very well seen in the upper cervical region, and of the columns of Burdach, and of the posterior portions of the lateral columns in the cervical enlargement and the dorsal region. This degeneration corresponds exactly in the lateral columns with the direct cerebellar tracts and the crossed pyramidal tracts. It is clearly separated from the posterior cornu on either side by a narrow zone of healthy white matter. It is in the lower dorsal region that the scleroses attains its maximum intensity, as well in the lateral as in the posterior columns. In the upper lumbar

region the middle fibres of the radicular zone are much degenerated, which involves the abolition of the knee reflexes during life, as the researches of Westphal have shewn. Absolute integrity of the marginal zone of Lissauer, and of the grey substance generally; whilst in the dorsal region the columns of Clarke, in the grey matter, are degenerated and their cells atrophied. This atrophy is at its maximum in the lowest dorsal region, where ganglion cells are no longer to be found.

Symmetrical degeneration of the lateral columns, strictly limited to the direct cerebellar and crossed pyramidal tracts. Maximum scleroses in the medium portions of Goll's columns.

Almost complete degeneration of the posterior root zones. The degeneration of the lateral column corresponds exactly with the section of the crossed pyramidal tract in this region.

2. Girl, age 14, ill for nine years, died of general debility following typhoid symptoms. Spinal cord very slender, especially in the dorsal region. The same symmetrical lesions of degeneration in the posterior and lateral columns (crossed pyramidal and direct cerebellar tracts) as in the first case. The medulla of extremely small dimensions. Microscopic examination, made with the greatest care, as in the foregoing case, reveals the same sclerous degenerations in the different regions of the spinal cord. In the grey matter of the dorsal region the columns of Clarke are much degenerated; also the posterior roots. Moreover, the posterior cornua are inact. The same integrity of the zone of Lissauer as in the first case.

Intense degeneration of the columns of Goll. Degeneration of the columns of Burdach reaches to the neighbourhood of the posterior columns from which it is separated by an intact zone. In the lateral columns the sclerosis involves exactly the section of the direct cerebellar and crossed pyramidal tracts.

The maximum sclerosis is always in the columns of Goll, next in those of Burdach. The degeneration of the lateral columns is less pronounced. A slender band of normal

tissue borders the posterior cornua. The antero-external zone of the posterior columns is normal. The degeneration of the lateral columns extends in front beyond the limit of the cerebellar tracts. Is this an individual anomaly or a marginal cortical sclerosis? Accessory zone of Lissauer absolutely intact.

In these two cases the histological nature of the lesions was identical with that which we meet with in tabes. The atrophied nerve tubes are scattered in a very close meshwork of extremely fine connective fibrils crossing in all directions. Nowhere do we find ectasis of vessels, nor thickenings of the walls of vessels which penetrate from the pia mater into the cord. The diffuse general thickening of the pia mater is but slightly pronounced, and is not at all more marked at the periphery of the cord at the places affected by sclerosis. Contrary to what Friedreich thought, there cannot then be any question here of a propagation of the inflammatory process of posterior lepto-meningitis to the lateral columns.

Rütimeyer has also found some degenerated nervous fibres in the median and in the sciatic, but the degeneration of these nerves was, to sum up, very slight. On the other hand the posterior roots were everywhere degenerated, least so in the cervical, most in the lower dorsal and lumbar regions.

Primary, combined, system sclerosis was very distinct in the columns of Goll, of Burdach, in the crossed pyramidal and the direct cerebellar tracts. The lesion of these diverse systems could be followed from above downwards in the spinal cord from the highest cervical to the sacral region. We refer to the important work of Rütimeyer for the detailed description of these lesions. We shall, however, remark that in the two cases the posterior radicular regions of Westphal, the integrity of which is necessary for the existence of the tendon reflexes, were completely degenerated in the whole lumbar cord. Moreover, the columns of Clarke, and their ganglion cells in the grey substance, were affected with increasing intensity from the upper dorsal to the lumbar region.

Among the portions which have remained intact we must

quote the direct pyramidal tracts, and the "antero-external zones" of the posterior columns, and especially the marginal zone of Lissauer,† which has always been found hitherto much degenerated in ordinary tabes.‡ If this degeneration is constant in Duchenne's ataxy and is regularly wanting, as it seems to be up till now in Friedreich's ataxy, this will be a new anatomical character important to differentiate between these two diseases. (See Lissauer's diagram of the lumbar posterior cornu in BRAIN, October, 1888, p. 329.)

To sum up, after the results of the different autopsies to which we have just referred, we may conclude that from the anatomical point of view, Friedreich's disease is clearly distinguished from tabes and from the other combined scleroses.

1st. It is distinguished from tabes by the primary combined system, degeneration of the direct cerebellar, and of the crossed pyramidal tracts, by the pronounced atrophy of the cells and of the fine fibres of the columns of Clarke, whilst the marginal zone of Lissauer always remains intact. In tabes, this zone is, on the contrary, constantly degenerated, whilst the cells of the column of Clarke, the direct cerebellar, and the crossed pyramidal tracts are intact. In tabes frequent multiple degeneration of the peripheral nerves (motor and sensory nerve roots, Déjerine); in Friedreich's disease, exceptionally a few degenerated fibres in the nerve trunks (Rütimeyer). We do not, therefore, share on this point the opinion of Rütimeyer, who admits, as a symptom common to the two diseases, degenerations in different degrees of the peripheral nerves. There is certainly a considerable and characteristic difference in this respect between the two diseases. The only anatomical characters that they have in common would thus be the integrity of the grey matter of the anterior cornua, the sclerosis of the posterior

† H. LISSAUER, Beitrag zum Faserverlauf im Hinterhorn des menschlichen Rückenmarks und zum Verhalten desselben bei Tabes dorsalis. *Archiv für Psychiatrie*, Bd. XVII. p. 877.

‡ OPPENHEIM und SIEMERLING, Beiträge zur Pathologie der Tabes dorsalis und der peripherischen Nerven-Erkrankung, *Archiv f. Psych.*, Bd. XVIII., Heft. 1 and 2. W. B. HADDEN and C. S. SHERRINGTON. The Pathological Anatomy of a case of Locomotor Ataxy. BRAIN, Part XLIII., October, 1888, p. 329. "Throughout the lumbar region, Lissauer's tract is completely sclerosed."

columns (of Goll and of Burdach), especially in their middle part, with relative integrity of the antero-external bands and atrophy of the posterior roots.

2nd. Friedreich's disease is distinguished from the other combined scleroses by the regularity of the seat of its system localisation, whilst in the other combined scleroses we meet with the most varied lesions; and especially because the disease invades the structure of which is congenitally incomplete. In all the cases of Friedreich's disease which have been examined *post mortem*, the authors have been surprised by the smallness of the cord and of the bulbs which have always been found slender and atrophied, especially in their posterior part, as well as by the smallness of the nervous elements of these organs, cells and fibres, as seen by the microscope.

Friedreich's disease, which forms an absolutely distinct clinical group, presents, therefore, also anatomo-pathological characters, which allow us to class it apart in spinal diseases. It is a combined primary system sclerosis of the spinal cord, in which several systems of fibres have been affected from birth or during infancy, with an arrest of development (posterior columns, pyramidal, and direct cerebellar tracts), and degenerate before having attained their full growth.

Prognosis and Treatment.

Two things must be distinguished in prognosis; as regards the cause of the disease, and the gravity of the affection, *quoad vitam*. If Friedreich's disease does not directly threaten life, if the patients do not die from it, since those who are affected by it may reach a comparatively advanced age, thirty, forty years, and even more (when they are not cut off by an intercurrent affection, which is most usually the case), hereditary ataxy does not the less bear a grave prognosis, for it is progressive, in an extremely slow way, it is true, but without permitting us to hope for remissions in its fatal course. We know of no case of cure of this disease. It is true that hitherto it has only been diagnosed when it was incurable. Shall we be more

fortunate in the future, when we shall be able to recognise it from its first symptoms?

We have unfortunately but little to say about the treatment in face of such a prognosis. As prophylactic measures, in the case of predisposed children, we ought to advise good hygiene, hydrotherapy, rational gymnastics, careful education, and intelligent supervision, to prevent the child contracting fatal habits of masturbation, which would, doubtless, favour the evolution of the disease by exhausting an already feeble spinal cord. We have observed on several occasions excesses of onanism in the antecedents of those who suffered from this disease, as we have also pointed out alcoholism of the parents, of the father especially, in the family history.

As for treatment properly called, we may have recourse, without hoping too much from it to two different systems. First of all to electrotherapy. Application of continuous currents, stabile and labile, to the vertebral column, by means of large electrodes (ten to fourteen milliamperes, three times a week). The patient generally feels well after the applications of electricity. He feels stronger, more active, less weak, and less uncertain on his legs. By continuing this treatment for several years, with regular intervals, we may, perhaps, obtain a decided delay in the progress of the disease. This is at least the impression which I have derived from the treatment, to which I have submitted the patient whose case has been related above. He has come to have the current applied for three years, omitting the applications two or three times each year, and he has told me that he found benefit from them. "I should not come so regularly," said he to me, "if I did not experience any benefit."

The second method is that of suspension, recommended by M. Charcot. They have obtained at the Salpêtrière some good results in this disease. I have suspended my patient more than a hundred times with a certain success. Speech has improved; the general condition has derived good from it. The patient has often found benefit from it, but the local symptoms of ataxy of the extremities and of weakness

of the legs have not been at all modified. The club-foot, the scoliosis, the nystagmus, the abolition of knee reflexes, the choreiform instability, all these exist after as before the treatment by suspension. Would more have been gained by employing Sayre's jacket, as M. Gilles de la Tourette proposed? We do not think so. But it is evident that we ought to try everything to combat a disease against which we do not really possess any efficacious means of treatment.

Appendix.

We must add to the cases which do not belong to Friedreich's disease that of J. Botkin.(63) By the courtesy of Prof. Dragomanoff, who wished me to translate it, I am able to give here a résumé of this case. Here is, first of all, the exact title of the article.

Jac. Botkin. Sloutchai Friedreich-oviboliezni. (A case of Friedreich's disease, *Medical Review*, Moscow, 1885, No. 1, p. 32-38).

Theodore Malych, age twenty-two, single, admitted November 7th, 1884, to the hospital of Yaroslaw. Mother hysterical; insane for two years after the birth of this son. Father died ten years ago, after having suffered for fifteen years from a convulsive affection, which probably had an analogy with the disease of his son, as all those who knew him affirm. The same mimicry, the same gestures, the same bearing, the same grinding of the teeth, the same movements of the eyes and of the head. The eldest brother of the patient had also in childhood convulsive attacks. His youngest brother has symptoms of "moral insanity." The patient began to walk only after his second year. He has often been ill, and in his illnesses frequently had convulsions. From eight to fifteen years of age pleurisy, scarlet fever, and typhoid fever. Each of these diseases accompanied by convulsions. Theodore Malych is timid, irascible, indolent, and obstinate. Expelled from the fifth class of the gymnasium on account of bad conduct. At sixteen years of age he enters at the special school as military surgeon's assistant. But headaches and diplopia compelled him to interrupt his

studies after seven months. After that swelling in the joints (rheumatism) for fourteen months. At the age of eighteen years (April-June, 1880), bad headaches, insomnia and melancholia. The latter lasted till December 6th. On this day on finding himself at the church of the convent of the Trinity he was taken with violent, irresistible laughter. The police took him to the hospital, where he remained four months. It is then that several symptoms of his real disease manifested themselves in the space of a few days, according to his own statement, with violent vomiting.

Present condition. Height, 167 cent.; weight, 137 Russian pounds. General pallor. Skull symmetrical. Sight normal on the right side. Left eye amaurotic. Normal reaction of pupils to light. Nystagmus. Sphincters normal. Copious salivation.

The bearing, the gestures, and the facial expression of the patient attract the attention especially. He stands erect, with the legs apart, the body thrown backwards, and leaning to the left. Head inclined backwards towards the right shoulder. Chin turned to the left and upwards, as in contracture of the sterno-mastoid. Hands crossed over the stomach. Hair thin. Apathetic expression. Jaws closed, and lips partially opened.

The voluntary movements are accompanied by a series of involuntary movements. The head is agitated by zig-zag movements, the teeth are ground. The eyes go from left to right. The arms are elevated and depressed, are flexed and extended. The hands execute movements of prehension. The body undergoes convulsive movements, forward, backward, to the left, to the right, is bent and again straightened. The feet are separated widely during walking. Trembling of the lower extremities in consequence of involuntary muscular contractions. In walking the patient sways; he is thrown from side to side. The direction of locomotion is always in a zig-zag. The legs give way sometimes and the patient falls.

The ataxy is so marked in the upper extremities that the patient has difficulty in dressing himself, and buttoning his clothes. He writes badly, and he cannot thread a needle.

When he closes his eyes, there is no increase of the ataxy, either static or dynamic. Romberg's symptom not present.

The more complicated the movement is, the more attention it demands, and the more agitated the patient is, the more do the involuntary contractions increase. The ataxy is especially pronounced when the patient walks or when he endeavours to narrate his history in a clear and distinct manner, for he never succeeds in it. It is impossible to describe the complication of the involuntary movements which are then observed. One must see them to understand to what extent they are developed.

At first sight, these movements appear disorderly, but after an attentive examination it is noticed: 1st. To each voluntary movement a definite group of involuntary movements corresponds; 2nd however numerous and varied they may be, these movements cease rapidly immediately the patient lies down horizontally and ceases efforts. The grinding of the teeth and the ocular movements form the only exception and cease only during sleep, when all the other involuntary movements also cease. Clonic contraction of the lower extremities as soon as the toes rest on the ground or on the mattress. Spasms of the muscles of the neck and of the arms. These spasms increase with the voluntary movements, and render the latter difficult, as also passive movements.

Speech is varied in tone. Far from being monotonous the sound of the voice is changed during conversation or reading; the patient passes involuntarily from one tone to another. He pronounces certain words indistinctly, does not pronounce always the end of the words. The remission of symptoms is a peculiarity of his disease. Thus sometimes he speaks much more distinctly.

Muscular enfeeblement. Dynamometer, 40 in the right, 25 in the left. The patient when lying down cannot lift his leg. If it is raised for him, he allows it to fall again.

Reflexes; plantar and cremasteric, distinct, normal; abdominal, gluteal, epigastric, scapular, but slightly marked.

Patellar reflexes and Achilles tendon reflexes *very exaggerated.* No ankle clonus.

Sensation. Rheumatic-like pains in the extremities. Vertigo. The patient localises exactly the lightest touch and the position of his limbs in space.

Spinal irritability. Slight hypho-scoliosis. Electric reactions normal.

Psychical manifestations. The patient is habitually apathetic, often excitable, and ready to laugh causelessly. Memory good. Imagination unimpaired.

In the discussion following the case, M. Botkin maintains that the presence of pareses, of contractures, and of exaggerated patellar reflexes does not exclude Friedreich's disease. He adds, however, that there exist in his patient two symptoms which have not been hitherto observed in this disease, unilateral amaurosis and grinding of the teeth. In any case, says he, even when these two symptoms would indicate a focal lesion, they cannot be regarded as due to a multilocular sclerosis. If we admit, with Féré and Erb, that Friedreich's disease is an intermediate form between disseminated sclerosis and tabes, there is nothing astonishing, concludes Botkin, in the fact that the symptoms of both lesions may be met with in this intermediate form, as has occurred in my case.

It will be sufficient to refer to the comparative table that we have given above of the differential diagnosis beeween Friedreich's disease and disseminated sclerosis to be convinced that Dr. Botkin's case does not belong at all to Friedreich's disease. We think that similar cases will not henceforth be accepted under the heading of this disease.

With regard to my provisional clinical classification of the combined scleroses, M. Dejerine writes to me :—

"Your type No. 1 (ataxic paraplegia of Gowers), and your type No. 3 (ataxospastic tabes of Grasset) are only one and the same type, clinically and anatomically, as I have established in 1884 and 1886. It is nothing else than *ataxo-paraplegic tabes of Dejerine.* I have even, unless I am mistaken, shewn you the preparations of it."

Not only has M. Dejerine shewn me numerous preparations of combined scleroses, but he has admitted me with the greatest courtesy to his admirable practice at the

Bicêtre, where he has given me permission to examine several of his patients affected with combined scleroses. I am happy to express here my sincere gratitude to him. His words have been fully opened to me, as also those of Professor Charcot at the Salpêtrière, for the observations I was desirous of making on Friedreich's disease and the combined scleroses. I owe thanks also to Professor Prevost, of Geneva, who has had the kindness to show me the preparations of the cord of the patient whose case he has published, with the autopsy, in the Archives de Physiology, 1877, and Professor Schiff, who has shewn me the cord sections which he possesses, belonging to the first patient examined *post-mortem* by Friedreich.

I must state here that I have not touched upon the study of the combined scleroses, and that the historical investigation, in particular, of this disease remains quite untouched, and has nothing to do with my clinical classification. I have not occupied myself at all in this work of the examination of the history of the combined scleroses, and I had no occasion to occupy myself with it. It would have been a digression. I have mentioned the combined scleroses only incidentally in the chapter on diagnosis, and with the sole object of showing that they could not be confounded with Friedreich's disease, when the latter presents itself under one of the forms corresponding with the four types which I have described. Now, from this point of view, it was necessary for me to separate into two distinct clinical types, ataxic paraplegia properly called and spastic ataxy. My classification has no other significance, and especially would not attempt to trench upon a historical question. If I had had to treat of this question, I venture to say that I should have recognised openly the legitimate place, and the importance which ought to be attributed, in a historical study of the combined scleroses, first of all to the case recorded by M. Prevost which dates from 1877, afterwards to the works of Westphal, Kahler and Pick, Babesiu, Raymond, Damaschino, Hopkins, Ballet and Minor, &c.; finally, especially to the memoir published in 1884 by M. Dejerine, which marks a decisive epoch in this study, and where is found for the first time, unless I am

mistaken, the complete picture of ataxo-paraplegic tabes, two years in fact before the works of Gowers and Grasset, which were printed in 1886, which is seen clearly from the bibliographical notices which I have given.

In conclusion, I shall draw attention to the work of Dr. E. Huet, *on chronic chorea*, Paris, 1888-1889, where will be found (p. 190) an interesting page devoted to the differential diagnosis of Friedreich's disease and chronic chorea, both of which are family and hereditary affections, with motor inco-ordination.

But whilst in Friedreich's disease there is choreiform instability during rest, in hereditary chorea we observe small separate, multiplied, varied and irregular movements. Romberg's sign is not present in the latter disease, nor yet nystagmus. The affections of speech differ greatly in the two diseases. The knee jerks are rather exaggerated in chorea, which is especially an affection of adult age, whilst Friedreich's disease appears, as we know in childhood and youth.

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