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PROGRESSIVE LOCOMOTOR ATAXY:

ITS SYMPTOMS, DIAGNOSIS, AND TREATMENT.

BY

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The following Paper was read in November last before the Medical Society of London, and as the subject is one of much interest at the present moment, I now again submit my views of this disease to the Profession, hoping thereby to afford reliable data for further investigations.

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PROGRESSIVE LOCOMOTOR ATAXY.

The disease to which I purpose to draw attention in this paper is not a new one, for it was more than thirty years ago described by various authors as tabes dorsalis, and more recently, chiefly by French physicians, as progressive locomotor ataxy. Yet the nature of the complaint, and the connexion which exists between the symptoms manifested during life and the structural changes found after death, have, until quite recently, been misunderstood, and I therefore offer no futher apology for bringing this important subject under the notice of the Profession. Before, however, proceeding to a description of the disease as it is now known, I will say a few words about its name and history, showing the steps by which we have gradually attained to that knowledge of its character which we possess at present.

Tabes dorsalis is first spoken of in the works of Hippocrates, and was by the father of medical literature believed to arise from excesses in sexual intercourse, the chief symptoms of the disease being spermatorrhœa, marasmus, and hectic fever. This

meaning of the term, however, has gradually changed, and those authors who wrote on tabes in the first decennia of this century, understood by it atrophy of the posterior portion of the spinal cord, brought on, not merely by sexual exhaustion, but also by exposure to wet, rheumatism, gout, and other causes, the chief symptoms being a peculiar form of paraplegia. The disorder was chiefly investigated by English and German physicians, such as Abercrombie, Hufeland, Steinthal, Romberg, and others. Their descriptions, although in some instances most eloquent, were, however, to a certain extent, wanting in accuracy, inasmuch as several different affections of the cord were comprehended under the name of tabes, and a clear distinction was not drawn between tabes and paraplegia. It was only after a more careful clinical study of the symptoms had been made, and after pathological anatomy, aided by the microscope, had stepped in, that a peculiar disease of itself, and one characterized by uniform structural lesions, could take its place in our nosological system. The chief credit of the anatomical investigations is due to Professors Virchow, Türck, Rokitansky, and Leyden, and in this country to Dr. Gull and Mr. Lockhart Clarke, who have shown that, in well-marked cases of tabes, an actual waste of nerve-fibres of the posterior columns of the spinal cord takes place, together with the formation of amyloid corpuscles, and considerable proliferation of connective tissue.

The first who drew a distinction between this disease and paralysis was Dr. Todd. He said, in an article on the nervous system in his Cyclopædia, that two kinds of paralysis might be noticed in the lower extremities: the one consisting simply in the impairment or loss of voluntary motion; the other distinguished by a diminution or total absence of the power of co-ordinating movements. In the latter form, while considerable muscular power remained, the patient found great difficulty in walking, and the gait was so tottering and uncertain that his centre of gravity was easily displaced. In these few words we have a good description of the symptom of *ataxy*,* upon which lately so much stress has been laid by French physicians. The term "ataxy" is as old as that of "tabes," for it also originated with Hippocrates; and it has likewise entirely changed its meaning in the course of time. Some authors have applied it to chorea, others to fevers, others to various nervous disorders. At present, however, we understand by ataxy, not a disease of itself, but merely a symptom to which various disorders may give rise, and which essentially consists of a want of co-ordination of voluntary movements, and a tendency on the part of the patient to lose his balance, but without actual loss of power, and apart from tremor, chorea, and paralysis. This symptom may be observed in disease of the cere-

* From τάξις, order, and privative alpha (want of order).

bellum, and in poisoning by alcohol, lead, and mercury; but it is more especially connected with that disease which has been long familiar to us as tabes. The best clinical study of this symptom we owe to M. Duchenne de Boulogne, who from 1858 up to this time has published a number of memoirs, in which he described what he thought to be an entirely new disease, which he called "progressive locomotor ataxy," and which he believed to be a functional disorder of the cerebellum. His apparent discovery was hailed as a real one in France, and Professor Trousseau actually proposed to call the new stranger "Duchenne's disease;" but on looking more closely into the matter we find that Duchenne's description is altogether applicable to our old friend, tabes. I have not the slightest hesitation in acknowledging the great ability and originality of M. Duchenne's researches, which were perhaps more strikingly displayed in this case just on account of his being unacquainted with the previous literature on the subject; yet, if I thought it desirable to attach a proper name to this affection, I should prefer calling it "Todd's disease," as Todd first drew the distinction between ataxy and paralysis, eleven years previous to Duchenne. But the best plan is, perhaps, merely to drop the term "tabes," as being too vague, and to call the disease under consideration progressive locomotor ataxy, or wasting of the posterior columns of the spinal cord.

The following details of a case of this affection which I have had under my observation for about five years, are well suited to illustrate the symptoms and progress of the disease:—

R. B——, a commercial traveller, aged thirty-seven, a tall and rather spare man, with a sallow complexion, first consulted me in February, 1860, when he gave me the following history. His mother had always been healthy, but his father had for the greater part of his life suffered from epilepsy, and died in a fit. The patient was brought up to the law; but in consequence of a disappointment he left that profession, and enlisted as a soldier. He served in Australia and Canada, and during that time suffered much from rheumatism. He never exceeded in drinking, but occasionally in sexual intercourse. He twice had gonorrhœa, but no syphilis. He suffered for a long time from hæmorrhoids, for which, in 1855, he underwent the operation by ligature, and he ascribed the commencement of his present illness to that operation, saying that he never felt quite the same man after the hæmorrhoidal flow had ceased. He left the army in 1856, and married. He is now father of a healthy child. For the last eighteen months he has been a commercial traveller, and as such he is always on the move, and frequently exposed to cold, damp, and great fatigue. In January, 1857, he first noticed that his sight became weak, and he

had some difficulty in writing and reading small print. Soon after he felt pains of a peculiar character, which he described as electric shocks through the legs, and as if the muscles were being rent asunder. These shocks came every two or three minutes. He underwent treatment by liniments and other external applications, but without relief. His gait now became tottering, and he had considerable difficulty in walking. He never goes out without a stick, and sometimes he is obliged to use two. In summer 1859 he consulted the late Dr. Todd, who told him that his case was incurable, and that he would have to lie on the sofa for the rest of his life. He prescribed strychnine and iron, and after having taken it for some time the patient felt rather stronger, but there was no improvement in the special symptoms of the disease.

Present state, February 14, 1860:—The patient's intellect, memory, and speech, are quite normal. He does not suffer from headache, giddiness, strabismus, or ptosis. Both pupils are enlarged, the left more so than the right. He complains of weak sight, and the opthalmoscopic examination shows the arteries of the optic nerve smaller than usual. His senses of hearing, smell, and taste are natural. Respiration and heart-sounds normal; pulse at the wrist rather feeble, but quite regular. His digestion is tolerably good. Tongue clean, appetite satisfac-

tory, but bowels rather costive. He complains of a sensation as if a net were tightly drawn round the abdomen. He is occasionally troubled in passing the urine, but there is no stricture. The urine is of 1030 specific gravity, and contains a sediment of urates, but no excess of phosphates, and no albumen or sugar. The sexual power has not notably diminished of late. On examination of the back by pressure, percussion, galvanism, and hot sponges, no place can be discovered which is particularly sensitive. The patient complains of numbness in the hands, more especially in the third and fourth fingers of the left hand. He can distinguish heat and cold, and feels the prick of a pin and pressure distinctly; yet the æsthesiometer shows a considerable diminution of tactile sensibility in the fingers. The upper extremities are pretty well nourished, and the muscles answer well to a galvanic current of moderate power. He can bend and stretch the arms with force, but he finds it difficult to button his shirt and to feed himself. The lower extremities are more affected than the upper ones. The patient has had sensations of "pins and needles" in the feet, but these have for some time past given place to numbness. He says that in walking he has a sensation as if he trod on cotton or springs, and when going upstairs he feels as if the steps rose under his feet. He must continually look at his limbs in order not to lose his balance, and can scarcely walk at all in the dark.

If told to shut the eyes, or stand with both feet together, he begins to stagger. In walking, he throws the legs forwards with a jerking motion; and, as he cannot measure his distance from the ground, he puts his foot down with great force. If lying down, he can bend and stretch the legs with considerable power, but he seems to exaggerate every movement, all muscular contractions being, not slow and equable, but violent, sudden, and jerking.

I ordered the patient thirty minims of the syrup of iodide of iron three times a day, sulphur baths twice a week, and a pill of aloes and myrrh at bed-time. After having used these remedies for about a month, there was a marked improvement in every respect. The pains were nearly gone, the sight was rather better, the walking decidedly steadier and less fatiguing, and the feeling of numbness slighter. He went on favourably until March, 1861, when, having had a long and very laborious journey to perform, he returned considerably worse, and from that time the disease gradually gained upon him, in spite of all treatment. In 1863 he had a course of nitrate of silver, but the affection was then evidently too far advanced for any medication to do good. The sight got rapidly worse, and he became at last completely amaurotic. The sensation in the lower extremities was also entirely lost. The patient was now confined to his room, and during the last six months of his life he never

left his bed. His intellect remained unimpaired to the last, and his disposition was always cheerful. He died in January, 1865, of a sharp attack of bronchitis.

Pathological Anatomy.

On opening the spine, the vertebræ and the vertebral canal appear healthy. The sac of the dura mater often contains a somewhat considerable amount of clear or slightly turbid liquid. The membranes themselves may be normal, but in some cases, the *posterior* part of the dura has been found thickened, and adherent to the pia by thin false membranes. Nothing of this kind is found at the *anterior* part of the dura. The posterior part of the pia is almost always less transparent than it should be; and presents a yellowish or milky appearance. It is often so firmly adherent to the substance of the posterior columns that it cannot be separated from them without tearing off some portions of medullary matter. These changes have, however, only been noticed in about one-half of the cases examined, and we must, therefore, consider them rather as incidental, than as pathognomonic appearances. The latter are found in the cord itself, which shows in its posterior columns, a peculiar grey coloration which is not superficial, but embraces their entire depth, and constitutes the characteristic anatomical feature of the disease, being always connected with a definite alteration of

the intimate structure of the cord. In cases where the pia is opaque, it is necessary to remove it to show the grey coloration, but where that membrane is transparent, it becomes visible immediately upon the removal of the dura. We then see, instead of the white matter of the posterior columns, either one or two grey bands proceeding from the lower end of the cord to the middle of its dorsal portion, and occupying the whole space between the opposite insertions of the posterior roots. As we proceed higher up, these bands become narrower, and often separate into smaller stripes, which run up to the calamus scriptorius, and the floor of the fourth ventricle.

The grey colour of the posterior columns is sometimes uniform throughout, in other cases it is dark in the median line, and light laterally. Occasionally the grey merges into amber, pink or reddish yellow, according to the stages of the degenerative process. Generally, however, it is so similar to that of the healthy grey matter, that Olivier believed the whole process to be one of hypertrophy of the latter substance. Laterally the grey coloration is mostly limited by the posterior horns, and centrally by the commissure. This commissure and the central part of the grey matter, is sometimes also affected, and in far advanced cases the disease may even extend to the lateral columns; while the anterior horns, columns and roots are always healthy.

As regards the *shape* of the cord, it appears

flattened from before backwards, so that at first sight it would seem to be actually enlarged. Such, however, is not the case, for the flattening results from the diminution of the bulk of the posterior columns, which are always smaller than usual. Where the disease has been severe, the posterior columns may be entirely wanting, being replaced by a thin band of connective tissue. The *consistence* of the grey matter and of the antero-lateral columns is normal, which may also be the case with the posterior columns; occasionally, however, these latter are found softened, and even semi-fluid.

The posterior roots are, in most cases, similarly affected to the posterior columns. Sometimes the whole substance of the root is in a state of degeneration, in other instances the grey stripes alternate with healthy white bands. The lower roots are generally more affected than the upper ones.

If we now ask ourselves which of the two is the primary and essential change, that in the *cord* or in the *roots?* the answer cannot be doubtful. The affection of the roots must be secondary to that of the columns:—

1stly. Because in a certain number of cases the posterior *columns* have been found diseased, while the *roots* were healthy.

2ndly. Because in other instances where both were diseased, the columns were in a far more advanced stage of degeneration than the roots.

3rdly. Because in most cases, although columns

as well as roots may be diseased at a *lower* part of the cord, yet, further upwards, the columns may show extensive disease, while the roots appear healthy; and lastly, because in no case has there been atrophy of posterior roots without simultaneous atrophy of posterior columns.

In the upper portion of the cerebro-spinal axis the disease is always less severe than in the lower part. The calamus scriptorius occasionally shows traces of it, but the cerebellum which has in all cases been examined with the greatest care, has always been found healthy. This latter circumstance has caused considerable disappointment to certain pathologists who concluded *à priori*, from physiological premises, that structural changes of the cerebellum *must* be found in progressive ataxy. Such, however, is not the case, and this shows again how necessary it is to be cautious in the application of physiological theories to pathological processes.

The cerebral nerves have only been examined in a few cases, and consequently our knowledge regarding the structural changes in them is still very deficient. The optic nerves have several times been found softened or entirely destroyed, only a few fibrous strings being seen in place of nervous matter. The ulceration may spread to the chiasma, but stops short at the corpora geniculata. In the retina it proceeds from the papilla to the periphery of that membrane. The other cerebral nerves are

only seldom diseased. In one case, the olfactory nerves, although apparently healthy, were, on being examined with the microscope, found to be almost smothered by amyloid corpuscles. The motor oculi, the hypoglossus, and the vagus have been found injured in a few cases. These results correspond entirely with the symptoms observed during life; for while the cord and the optic nerves, when once thoroughly diseased, are generally permanently disabled, the symptoms referable to the other cerebral nerves almost always disappear after a short time.

I shall now describe the more minute structural changes found in the nervous matter by the aid of the microscope. In conducting such investigations, it is advisable first to harden the parts artificially by putting them for a day or two into alcohol, and afterwards for a few days into a weak solution of chromic acid. If the specimens have been thus prepared, fine sections can easily be made. The first thing which is then observed is that, whereas a section of healthy nervous matter taken from the anterior columns is dark, one from the diseased posterior columns is transparent. This results from the fact that in the anterior columns healthy nerve-tubes are crowded together, while in the posterior columns most of them have been destroyed by the disease, and are replaced by a clear and nearly homogenous mass, which contains a number of small granules, and connective tissue.

In order to show distinctly the relations between

nerve-tubes and connective tissue, the specimens should be put into a mixture of alcohol and turpentine, whereby the myeline contained in the nerve-tubes becomes so transparent that it seems to have almost disappeared, and offers no impediment to a clear view of the connective tissue, with all its ramifications, more especially if we add a solution of carmine, whereby the meshes of the connective tissue acquire a red colour. Again, if we wish to know what has become of the nerve-tubes, the specimens should be left for a long time in chromic acid, and then treated with alcohol and turpentine, but not with carmine. By this proceeding, the nerves, and especially the myeline contained in them, acquire a slightly yellow or greyish tinge, and form a striking contrast to the uncoloured connective tissue. The polarised light may also be made use of in such researches. For this, specimens are put into glycerine, and in a dark field we may then easily recognize the myeline by its milky mother-of-pearl appearance.

By using these means, the following circumstances are elicited:—The nerve-tubes are either diseased or entirely destroyed. They often appear granular, varicose, more narrow than usual, and nearly or entirely devoid of myeline and the cylinder axis. Besides these diseased nerves we find proliferation of connective tissue, slender fibres, round cells whose membrane nearly touches the nucleus, a more considerable number of free nuclei, and

from one to four nucleoli in one nucleus. Capillary vessels may also be discovered, which are either healthy or diseased. The adventitia is often very much thickened, and the vessels appear surrounded by oil-globules. Finally, we meet with a large number of amyloid corpuscles, presenting the well-known mother-of-pearl appearance. They are most numerous along the course of the blood vessels, and seem to abound chiefly where the degeneration is not very far advanced, while they are less frequent where an entire destruction of nervous matter has taken place. The process may, therefore, be characterized as one of destruction of nervous matter, proliferation of connective tissue, degeneration of blood-vessels, and formation of corpora amylacea and oil-globules. It resembles both chronic inflammation and simple atrophy, but neither of them altogether, and should therefore be looked upon as one "*sui generis*."

Symptoms and Progress.

I now proceed to analyse the symptoms and progress of the disease. Duchenne has distinguished three stages of ataxy, and although these are by no means always so well defined as this author would lead us to believe, yet his division may, with certain modifications, be adopted for the sake of convenience. The first stage is marked by certain affections of the cerebral nerves, pains of a peculiar character, and diminution of sexual power; it gene-

rally lasts from four to five years, sometimes for a much shorter, sometimes for a longer period. In the second stage the symptom of ataxy supervenes, together with loss of sensibility; this may last ten years and more. In the third stage the symptoms of the first and second stage become more severe, complications, such as paralysis and spasms, arise, and death results from exhaustion or from intercurrent diseases.

The commencement of progressive ataxy is either slow or subacute. One or more of the cerebral nerves are generally the first to suffer; those most frequently affected being the optic, and the third, fourth, and sixth, pairs. The chief symptoms are, therefore, amblyopia, double vision, strabismus, and ptosis. Sometimes there is even treble vision, or two images may be observed by one eye, while the other is closed—symptoms which have not yet found a satisfactory physiological explanation. The ophthalmoscopic examination of the fundus occuli shows, at first, symptoms of congestion; the capillaries are diseased, and the whole fundus has a violet colour. Amongst the symptoms mentioned, strabismus and double vision have the tendency to disappear within a few months, with or without treatment, ptosis is liable to continue much longer, and amblyopia almost always, in the course of time, merges into amaurosis. The ophthalmoscope then shows evident symptoms of atrophy of the retina; the diameter of the blood vessels is diminished,

the papilla is of a greyish or mother-of-pearl hue and excavated, and a white circle is seen at its margin. These facts are important as regards prognosis, for while we may reasonably hope that strabismus and double vision will disappear of themselves, ptosis and amblyopia offer less favourable prospects. From this it is easy to understand why double vision and strabismus have not been mentioned in the description of tabes by the older observers, these symptoms having escaped their attention from not being present at an advanced stage of the disorder.

The other cerebral nerves may also show signs of paralysis, with the only exception of the olfactory; there may be loss of taste, deafness, difficulty of mastication, dysphagia, and numbness or loss of sensation of the face, lips, tongue and gums. These latter symptoms are, however, comparatively rare.

Almost all diseases of the cord are accompanied by *pain*, but progressive ataxy more frequently than any other. Pain of a peculiar character constitutes, indeed, one of the most distressing symptoms of this affection. The sensations are short, sharp, and sudden, similar to electric shocks. After a second or two the pain is gone, and the patient has a short interval of rest; but soon there is another pang, and this may go on for two or three days consecutively, after which there is a free interval of a few weeks or even months. During such

paroxysms the patient is mostly obliged to be in bed, and their effect on the general health is prejudicial. The pains often begin in the feet, then migrate about the body, sparing only the head, and finally settle in one of the legs, from where, as the disease advances, they gradually proceed upwards. During the attacks neither swelling nor redness is perceptible in the parts affected, but after some time considerable hyperæsthesia sets in, so that the patient is exceedingly sensitive to touch or even a slight draught of air. In other cases there is no hyperæsthesia, but numbness, and strong pressure relieves the pain. If the eyes are attacked, a flow of tears, heat, and dilatation of the pupils are caused; if the bladder is invaded, catarrh of that organ may be produced.

As time wears on, the pains generally increase in severity, and appear at shorter intervals. They are most liable to come on when sudden atmospheric changes occur, and after exposure to wet, or after excesses in walking, drinking, or sexual intercourse. The patients generally dread winter. As spring advances they frequently improve, and this is often believed to be due to the remedies which happened to be employed about that time. Some patients are, by an increase in the severity of the pains, able to predict, with considerable certainty, an impending change in the weather.

Spermatorrhœa is another important symptom, but it is absent in a number of cases. Where it is

present, it seems to accelerate the progress of the disease. Emissions occur first at night, and with erections; after a time they likewise occur in the day time, and without erections, more especially on voiding the bowels. If this condition is allowed to go on for some years, impotence is the final result, but under the influence of a suitable *régime* and treatment it is often improved or cured. In exceptional cases the disease is ushered in by priapism and satyriasis. Eisenmann has recorded a case in which these latter symptoms continued more or less for thirty years, and were only relieved by large doses of opium.

The bladder and rectum may also be affected in the first period of progressive ataxy, but they are more generally so in the second. Constipation is the rule, while involuntary fæcal discharges are rare. The bladder is not sufficiently emptied, and the urine is passed tardily. Incontinence may also be present, but this is often promptly relieved.

After one or several of these symptoms have continued for some time, other morbid signs present themselves, either suddenly or gradually, and by which the second period of the complaint is marked; the most important one of these, and from which the disease has received its name, is the locomotor ataxy, which consists of a peculiar disturbance of locomotion, as well as of equilibration. Volition loses its influence over the muscles, which, although still possessed of great intrinsic

force, are nevertheless unable to execute complex movements, or preserve the equilibrium of the body in its erect position. And here I would direct attention to a fact which was first prominently dwelt upon by Duchenne—viz., that co-ordination is composed of two several kinds of muscular action, that is, of the harmony between the antagonists, and of instinctive or voluntary associations of muscles. In voluntary movements the antagonists are not, as is generally believed, inactive, but every such movement is the result of a two-fold nervous action, by means of which both flexors and extensors are made to perform simultaneously, so that one set produces, and the other moderates and tempers, the movements. Without this co-operation of antagonists, all movements become devoid of certainty and precision. Now, in progressive ataxy, the patient first loses his instinctive faculty of regulating the kind and extent of his muscular actions, although he may still be able to associate muscles to contract in order to produce certain movements. In Nature isolated muscular contractions do not occur; they may be obtained by artificial means, such as Faradisation, but not physiologically. Most muscular functions, in fact, require a large number of simultaneous movements, which are again in their turn only the resultant of several forces. Most of these complex muscular contractions are learned in early life by daily, nay, hourly practice. Every child that first begins to

walk, may be said to show locomotor ataxy, and does not learn to walk or stand without having often fallen down, and thus received many practical lessons of the importance of a judicious co-ordination of movements. In after life these complex muscular contractions occur instinctively, almost mechanically, and without an effort of volition; and the loss of this faculty is what we have to understand by the term locomotor ataxy.

This generally begins in the lower extremities. The patient first notices an awkwardness in his movements when he walks in the dark, or in the morning while he is dressing. He soon takes to a stick when out of doors, but even with such aid he finds that he has to make considerable efforts to prevent himself from falling. In order to appreciate the degree of ataxy which may be present, we must examine the patient in all positions, whilst standing, walking, and lying down. If he is told to stand with both feet close together, he can seldom keep his balance. He staggers from one side to the other, and manœuvres desperately with his arms, almost like a rope-dancer; but unless he clings to some support, he nevertheless falls down at last. If told to stand with his eyes closed, his struggles to maintain himself are equally distressing. But this alone is not sufficient to enable us to diagnose progressive ataxy, because the same symptoms may be found in persons who have just recovered from acute diseases, or who suffer from

certain affections of the brain, or who are reduced by bad living, and in a weakly condition.

The ataxy becomes much more apparent if the patient is told to walk. He throws the legs forward with a jerking motion, and puts the feet down with great force. In turning round, he is especially awkward. At the commencement of the affection the patient can still walk a considerable distance, and feels the difficulty chiefly on first starting, or changing his direction. But as the disease advances, walking becomes almost or quite impossible. The patient is still able to make strong muscular exertions, and on trying to walk, flexors as well as extensors feel hard and contracted; yet he does not succeed in doing that which other persons accomplish without an effort. At first the ataxy is most striking in the pelvic and femoral muscles. But after a time it becomes also apparent in the leg and foot. The sole seems continually to search for support, one leg is crossed over the other, or jerked about in a disorderly manner, without the slightest intent or purpose. All efforts to check these movements are ineffectual and only serve to increase them. The patient soon becomes exhausted by the expenditure of so large an amount of muscular force, and is glad to get back into bed.

If we examine the patient while he is lying down, and tell him to flex or extend his limbs, he often does so in an abrupt and sudden manner. In a somewhat advanced stage of the disease no graceful

or easy movement is possible; he does not know what force to use, or where to stop, and he cannot continue a given movement for a certain length of time. How great is the difference between this condition and that of a paralysed person! The atactic patient has a great deal of power, and is able to make Herculean efforts to do what he is told, but he does not know how to do it, and expends his force in vain. In the paralytic, on the contrary, the power is lost, or greatly diminished; he cannot move, or if he succeeds in doing so, it is a feeble motion, although one not devoid of purpose.

In the upper extremities ataxy is not so well marked, nor so frequent as in the lower ones; and it mostly appears only at a later period of the disease. If the patient is told to touch his nose with the tip of the forefinger, to pick up a pencil or a piece of money, to describe a circle in the air, &c., the ataxy becomes apparent, more especially if he closes the eyes. He cannot write in a straight line, and is very awkward in dressing and feeding himself. The muscles of the face are only rarely affected; sometimes, however, there is facial spasm and difficulty of articulation.

Disturbances of not less importance are at the same time observed in the sphere of sensibility. When questioned, the patient generally mentions a feeling of numbness or heaviness in a limb or part of a limb, which may exist without any loss of sensation in the skin or other parts. It generally

commences in the toes, or the soles of the feet, and from there gradually spreads upwards to the abdomen, and the chest, which feel as if constricted by a circular band, a net, or a tight string. Where this feeling invades the chest, dyspnœa is also present. In the upper extremities the numbness is generally confined to the third and little finger, and only seldom spreads higher up. Numbness in the legs, and especially in the soles of the feet, is one of the most constant symptoms of the disease, and if absent, must make us doubtful whether the case is really one of ataxy or not; and when it diminishes or disappears, we may say with certainty that an improvement has taken place.

Anæsthesia is a frequent, but by no means constant symptom of ataxy, and generally appears only at a somewhat later period of the disease. We have here to distinguish several forms of anæsthesia, viz.—loss of the sensation of pain, which is also called analgesia; loss of the sense of touch, which is anæsthesia properly so called; loss of the sense of locality, loss of the sensation of temperature, and finally, loss of the sensation of pressure.

Sensibility to pain is generally diminished or even entirely destroyed. You may prick the patient with a pin or needle, or you may pinch his skin, and he will not feel it at all, or he will merely feel it as a touch. Cruveilhier mentions the case of such a patient who had suffered a fracture of the leg, and neither at the time of the accident, nor afterwards,

had felt any pain whatever. The sensibility to galvanism is also diminished, as the patient is able to bear the application of a very powerful current without the slightest inconvenience. The sense of touch is affected in a like manner. If he is told to close the eyes, and something is then given him to touch, he is not able to distinguish the nature of the object. This loss of the sense of touch is not confined to the upper or lower extremities, but may extend to the neck, tongue, and soft palate. A very common symptom is tardy sensation, so that when touched, the patient only feels it five or ten seconds afterwards. If the soles of the feet are tickled, there are scarcely any or no reflex movements, and the perception of it is dull or none.

The sense of locality is often wanting, therefore the patient, if touched in a particular part of the body, cannot tell you where he is touched. At the same time, the distance at which two separate sensations are perceived as such, is increased, as is well shown by examining the patient with Weber's pair of compasses. In the legs, where the normal distance for two separate sensations is an inch and a half, the patient is sometimes altogether unable to feel them as such, while in the face and the fingers he may still be able to do so.

A curious fact is, that the *sense of temperature* is only seldom deficient. Persons in good health are able to distinguish with certainty a difference of one, or even half a degree. Objects which are

warmer than 90° give a sensation of heat, and such as are below 90° feel cold. Now, patients suffering from ataxy, although they may have entirely lost all other sensations, are yet often able to distinguish between different temperatures. Thus, M. Topinard relates a case which was observed in Trousseau's wards in the Hôtel-Dieu, and where the patient was affected by double amaurosis, absolute anæsthesia as regards touch, pain, and locality, ataxy of motion to such an extent that he had not been out of bed for two months; who had completely lost his muscular sense, and who had only the sensation of heat and cold to tell him that he still had his limbs. Sometimes his legs would be jerked out of bed by spasms which he did not feel, and which, being blind, he could not see. Then, after a time, a sensation of cold would creep upon him, and the poor fellow would ask whether anything was the matter with his legs. This patient existed, as it were, only by his memory, as he had lost the consciousness of his body. But even the sense of temperature may be wanting. Leyden mentions the case of a patient who prepared a warm bath for himself, and not being able to distinguish between heat and cold, he made it very hot, and was severely scalded on going into it.

The last disturbance of sensibility we have to notice is that of the *sense of pressure*, which resides in the nerves of the skin and the deeper parts—viz., the cellular tissue, the muscles, and periosteum.

The sensation of pressure is produced by weights resting on certain parts of the body, more especially on bones. M. Eigenbrod, who had studied the changes which this sense undergoes in various affections, has found that there is a considerable diminution of it in ataxy. Persons in good health are generally able to distinguish a weight of thirty from one which is only twenty-nine pounds, but patients suffering from ataxy lose this faculty to a great extent. It has been shown by experiments that, if the sense of pressure in the soles of the feet is diminished by the application of ice or chloroform, the gait becomes tottering: it is, therefore, permitted to suppose that the uncertain walk of atactic patients is partly due to the diminution of the sense of pressure, a firm gait being only possible where there is a proper sensation of the resistance offered by the ground on which we walk.

In the third period of the disease we observe all the symptoms which are present in the second, only in a more marked degree. Sensation becomes more and more impaired, the ataxy more striking, and muscular force, which was previously intact, also begins to suffer. A semi-paralytic condition is gradually induced. The patient cannot grasp any objects with sufficient force; he cannot offer any resistance to movements imparted to his limbs by others; he has difficulty in raising his legs when in bed; and if he attempts to walk, the feet drag on the ground. Occasionally the muscles become atrophied, and

undergo fatty degeneration. Spasms may supervene, which are most troublesome at night and when the weather is damp. Where there has been amblyopia, it merges into complete amaurosis; there is paralysis of the bladder, incontinence or retention of urine, involuntary evacuations of fæces, and at last decubitus on the sacrum, unless great care is taken to prevent it. Death takes place either from this latter complication, or from inflammation of the bladder and kidneys, or from intercurrent diseases, such as bronchitis, pneumonia, and phthisis.

A curious symptom remains to be mentioned—viz., the state of mind of such patients. Unless they suffer from severe pains, they are mostly, if not happy, at all events resigned to their fate. They do not complain much, and are inclined to think lightly of their affection. In this respect progressive ataxy resembles pulmonary consumption, in which the mind is also often composed and cheerful, while in diseases of the brain and the liver there is almost always great depression or irritability, or both combined.

Causes.

The causes of progressive locomotor ataxy now remain to be mentioned. As regards age, we find that most patients are between thirty and fifty years. The disease is very rare in old people, and is not seen before the age of fifteen. The male sex is

more disposed to it than the female. Romberg says that scarcely one-eighth of the cases are females. In the observations recorded by French authors and tabulated by M. Topinard, the proportion is of one female to four males. Amongst the nine cases which I have observed, not one occurred in a woman. As regards occupation, it appears that persons who are much exposed to cold, damp, and fatigue, are more liable to it than others. Two of my patients were commercial travellers, who were continually on the move. Another patient, a young man who had been in good health before the disease broke out, attributed his illness to his having been obliged, after a ball in which he had taken active part, to walk home in thin boots in a pelting rain, having been unable to get a conveyance. In this case the symptoms supervened with great rapidity.

Soldiers are also very liable to it. "The malady, is rife," says Romberg, "when the strength is much taxed by continued standing in a bent posture, by forced marches, and the catarrhal influences of wet bivouacs, followed by drunkenness and debauchery, as is so often the case in campaigns; and this is the reason why tabes dorsalis was so frequent during the first decennia following the great French wars of the present century."

The malady breaks out more frequently in autumn and winter than in spring and summer. Most patients who have the disease improve during the summer months; and we must, therefore, be

cautious to ascribe any beneficial results which may appear then to the treatment employed; while if they get better in winter, there is far more probability of the remedies used having been of actual service.

We have at present no sufficient data to say whether sexual excesses may cause the complaint. On the whole, experience goes far to prove that such excesses tend more to produce cerebral than spinal affections. At all events, I think it erroneous to put down such excesses as the chief cause of progressive ataxy, as was done in former times.

Emotion and anxiety, combined with overwork and bad living, are no doubt potent causes; but syphilis seems to have little or nothing to do with the disease.

Suppression of habitual perspiration, especially of the feet, has been mentioned as a cause in several instances. In the case related above, the patient himself believed that the suppression of the hæmorrhoidal flow was the cause of it. Sometimes the affection is hereditary. Children of parents who have suffered from epilepsy and other nervous disorders, are more liable to it than children of healthy parents. Accidents have often seemed to cause the outbreak of the complaint.

Diagnosis.

A decided opinion of the nature of the disease can scarcely be formed in the first period. We

may certainly have our suspicions of ataxy threatening the patient if we find that he has been much exposed to cold, damp, and fatigue, that he has committed excesses of various kinds, and suffers from the pains described above, together with strabismus, amblyopia, and anaphrodisia; but all such symptoms may be due to syphilis, rheumatic diathesis, and other diseases. Diagnosis is very easy in the second stage of the distemper, but it must be borne in mind that with the only exception of the symptom of ataxy itself, all other functional disturbances previously described as belonging to that stage may be absent. In the third period, diagnosis may become difficult in consequence of complications which arise, such as paralysis; the affection can then no longer be called ataxy proper, as the degeneration has already spread to the grey matter and the lateral columns of the cord.

There are few diseases which could be confounded with ataxy. In *chronic myelitis* we have, as a rule, no ataxy, but paralysis; moreover, the cerebral nerves do not suffer in that affection, and all symptoms are referable to simple inflammation. There is fixed pain at a certain point of the back, which is increased by pressure; besides which we have muscular spasms, diminution of muscular contractility, and atrophy of muscular substance.

From *disease of the cerebellum* ataxy may be distinguished by the absence of the fixed and permanent pain in the back of the head, and of the

vomiting, both of which symptoms are generally present where the cerebellum is diseased. The cerebral nerves may suffer alike in both affections; but the progress of these disturbances is not the same, for while in ataxy the double vision and strabismus come on suddenly, and generally disappear within a few weeks or months, in disease of the cerebellum they continue after having once commenced. In these affections the symptom of ataxy is by no means constant; sometimes there is, instead of it, an impulse to rush forwards or backwards; or the patient may be simply unable to keep his balance. Vertigo, convulsions of the face and limbs, and epileptiform seizures, which are common in diseases of the cerebellum, scarcely ever occur in progressive ataxy. With *softening of the brain* ataxy can scarcely be confounded, as in that affection we have mostly hemiplegia, and early impaired intellect and memory. In *general paralysis* there is weakness of the intellect and loss of muscular force, while in ataxy neither the intellect nor the muscles suffer.

Chronic poisoning by *alcohol* and *brain-syphilis* may give rise to the symptom of ataxy, but in most cases there are other characteristic indications which cannot fail to lead us on the right track. Where the affection of the brain is the only sign of venereal disease, which it is in exceptional cases, the effect of the treatment may guide us in our diagnosis. If nitrate of silver and hypophosphite

of lime should fail to retard the progress of the disease, we may try iodide of potassium; and if this should prove curative, the affection was most likely of a syphilitic kind.

Paralysis of the muscular sense is an affection quite different from ataxy. In the former, the guiding power of the muscular sense is lost, so that, in moving his limb, the patient has to trust exclusively to the sense of sight as a guide. The power of the muscles is as great as ever, but the patient is unable to make the slightest movement in the dark. In the daytime he can move the limbs quite well, provided he keeps his eyes steadily fixed on the parts; but as soon as anything else attracts his attention, and he turns the eyes in a different direction, his movements are arrested. The difference between this affection and ataxy may be easily perceived; I will merely add that paralysis of the muscular sense is not a disease of itself, but only a symptom, which may supervene in several nervous affections.

Prognosis and Treatment.

The prognosis is not favourable, for up to the present time not a single case is on record in which perfect recovery has ensued. Indeed, as late as 1851, Romberg wrote that there was no hope for patients of this class, that a fatal issue was inevitable, and that it was but common humanity to inform them that therapeutic interference could

only injure. This sad confession of impotence need no longer be made, and we may certainly congratulate ourselves on having far more control over the disease than we had formerly. Moreover, its progress is seldom rapid, although acute intercurrent disorders, such as erysipelas, bronchitis, and pneumonia, are grave complications which may carry the patient off suddenly. Much must depend upon the period at which the case comes under treatment. If all the symptoms of the disorder are fully developed, the hope of a cure may be slight, although even then much may be done to alleviate the patient's suffering. The case is different if the patient presents himself in an early stage of the disorder. The fact that the cerebral nerves, with the exception of the optic, generally recover from their affections in the course of the disease, goes far to prove that previous to the structural changes in the cord there is a functional stage, in which much may be done by medicines. Moreover, we must bear in mind that Messrs. Charcot and Vulpian have found nervous fibres in the process of reparation in the cord of a man who had died of ataxy; and that therefore even at a later time we must not give up hope altogether, especially if the patient is placed in favourable circumstances.

I now come to the treatment of ataxy, and will first say a few words about diet and regimen. This must depend a good deal upon the condition in

which the patient is at the time he comes under treatment. If he is in a weakly state, plain and nourishing diet, with iron, quinine, and cod-liver oil, should be prescribed. I have never seen a case in which lowering did any good. Hippocrates has recommended milk diet in erotic tabes, and Eisenmann speaks highly of the same in progressive ataxy. I have often given milk-and-brandy, two and even three times a day, with decided benefit to the general health; but have never found it expedient to insist on an exclusive milk diet. Exercise should be very moderate; and for those who have undergone great fatigue, rest is most beneficial.

Counter-irritants to the spine have been used by many physicians, but, as a rule, the benefit obtained has not been proportionate to the sufferings inflicted by their application. The moxa, the hot iron, issues, blisters, and leeches have been employed. I give the preference over all of these to the continuous galvanic current, applied to the lower and middle portion of the spine. It has in several cases seemed to me of decided benefit in lessening the pains, and also the disagreeable feeling of constriction which is often felt at the abdomen and the chest. The action of this agent is as rapid as that of the iron; its application entails far less trouble and suffering than this latter; and its efficacy is superior to that of issues, blisters, and leeches. Dry cupping on both sides of the spine is also useful.

Iodide of potassium has been recommended by Duchenne and others; but no cases have been published in which this remedy has proved successful. On the contrary, several are on record in which it seemed to accelerate the progress of the disease. I have given it in two cases, but without any effect, and am certainly not in favour of its administration. Iodide and bromide of iron are useful, but do not effect a cure. Mineral waters have been very frequently employed, both externally and internally; and it seems that for a time they do good. Amongst the French spas, Barèges is the one most recommended; amongst the German waters, Marienbad and Wiesbaden have a special reputation; while chalybeates and indifferent thermal springs, which often prove efficacious in certain forms of paralysis, have entirely failed in ataxy. On the whole, I should be averse to sending atactic patients on a journey to some distant spa, as rest at home, with certain remedies to be mentioned hereafter, is more beneficial than all the mineral waters in the world. If, however, there should be costiveness and abdominal plethora, Carlsbad, Marienbad, or Kissingen water may be drunk at home with benefit. Sulphur baths may also be taken at home; and I think highly of them. I have never employed them alone; but they certainly seemed, in conjunction with other remedies, to do much in relieving the pains and diminishing the numbness. The patients feel more brisk and

supple after the baths, and are almost always desirous of repeating them as often as possible. A sulphur bath may be prepared by dissolving from three to six ounces of the sulphuret of potassium in two pints of warm water, and adding this to the bath. Vapour baths and Turkish baths should be avoided.

Nux vomica and strychnine have frequently been used, but generally seemed to do harm. Duchenne recommends faradization, and the late Professor Remak, of Berlin, praised galvanization as a curative agent. From faradization I have never seen any benefit; galvanization has proved useful at my hands not only in the pain and feeling of constriction, but also in the affections of cerebral nerves which are so frequent in the commencement of ataxy; on the other hand, it has been powerless against the disease itself, more especially against the symptom of ataxy.

Amongst the other remedies which have been used in this affection without much result, I will mention opium, bromide of potassium, secale cornutum, essence of turpentine, and arsenic. The only remedy which seems as yet to have done good, in a very large proportion of cases, is the nitrate of silver, given in doses of one-tenth to a half grain, two or three times a day. Professor Wunderlich, of Leipzig, was the first who employed silver in seven cases of this disease, in none of which, it is true, he obtained an actual cure, but in most of

them considerable improvement. In 1862, Messrs. Charcot and Vulpian, in France, took up Wunderlich's idea, and used the nitrate in five other cases, and in each of them there was much amendment. Since then this remedy has been employed in most cases of ataxy, and with somewhat variable success. In some it has so disagreed with the patients that it was necessary to discontinue its use; in others it had little or no effect; while in the majority of cases the remedy has proved, if not curative, at least very useful, and it is the one upon which most reliance can be placed in the treatment of this disease. I am in the habit of giving the silver together with the hypophosphite of soda, and this combination seems to answer much better than either of these remedies singly. I have, indeed, now a case of ataxy under my care in which the improvement has, for the last six months, been so considerable under this medication that I am hopeful of a cure. Certain precautions should, however, be taken in administering the nitrate. I generally employ it for four or six weeks consecutively, and then discontinue it for a fortnight or three weeks, giving in the meantime a slightly aperient mineral water. After this the use of the remedy may be safely recommenced, and continued for a month or so. The gums must be inspected from time to time, as the peculiar coloration which silver produces in the long run first appears in the mucous membrane, and only afterwards in the skin.

With the precautions mentioned, however, no disfigurement of the patient need be feared. I have never gone beyond the dose of half a grain, and perhaps this is another reason why in my cases the remedy has been borne without any inconvenience. I should, however, not recommend all cases of ataxy to be treated alike; in this affection we must, as in every other, study each individual case by itself, and advise for it what seems, under the special circumstances, most likely to do good. Thus hysterical or hypochondriacal patients in whom ataxy may supervene, will require a different treatment from plethoric persons, or such as have long suffered from rheumatism, or have been subject to privations and anxiety. Much is, therefore, left to the tact and discrimination which, together with knowledge, should, in all cases brought before him, guide the doings of the physician.

THE END.

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