On Progressive Locomotor Ataxia. By J. R. Gasquet, M.B. Lond.

The three works whose titles we have placed at the foot of this article* represent almost entirely our current English literature on the disease, 'progressive locomotor ataxia,' of which they all treat, and it is therefore the more unfortunate, and almost provoking, to find that all three of them, although most excellent so far as they go, are professedly incomplete in their purpose and scope. Thus Dr. Althaus has given us a lecture, which is admirably calculated to give a general notion of the disease to a beginner, but does not answer many questions which the more advanced practitioner would ask.

On the other hand, Mr. Clarke presents us with the most interesting collection of cases of ataxia yet made, connected by remarks of the highest practical value, although too desultory and detached to be of much use to any but those who have already studied the subject.

Lastly, Dr. Bazire, who is doing such good service by his excellent translation of Trousseau's 'Clinique Médicale,' has supplied, in his notes to that work, some very useful information on some points connected with this disease, but nothing more.

Our object in the next few pages will be, not to attempt to fill up the void which we have just pointed out; for this we have neither experience, learning, nor space enough at our command. We propose only to do what we have just quarrelled with Dr. Althaus for doing, and to endeavour to spread a general knowledge of the disease as widely as possible, in order that multiplied observation may clear up (as it alone can) much that is still obscure about its nature and treatment. This would probably be the result of calling general attention to the malady here just as much as in France, where so much that is interesting on the subject has been published in the last few years.

* 1. 'On Epilepsy, Hysteria, and Ataxy.' Three Lectures, by Julius Althaus, M.D., Physician to the London Infirmary for Epilepsy and Paralysis. (London, Churchill, 1866.)—2. 'St. George's Hospital Reports.' Vol. I, 1866. "On the Diagnosis, Pathology, and Treatment of Progressive Locomotor Ataxy," by J. Lockhart Clarke, F.R.S.—3. 'Professor Trousseau's Clinical Lectures,' translated, with Notes and Appendices, by P. Victor Bazire, M.D., &c. (London, Hardwicke, 1866.)

There are really no grounds for supposing that the disease is notably (if at all) rarer in England than in France. Every practical man, on first reading an account of ataxia, will recall cases which he had diagnosed to be intractable rheumatism, amaurosis, or paraplegia, but which, when reconsidered, seem to have been ataxic in their symptoms and character.

Nor could such a man take much shame to himself for his past opinions; for, up to the year 1843, when Dr. Todd (in the 'Cyclopædia of Anatomy and Physiology') first clearly pointed out that there were cases in which muscular power was not lost, and yet the movements of the body were very irregular, no scientific knowledge of the disease we are considering was possible, since its most important symptom was unrecognised. Its pathological anatomy had, however, been described by Jacoby (one of the school of Romberg) in 1842, and this was followed up, after an interval of ten years, by Türck and Rokitansky; but the disease itself was still undistinguished from paraplegia, muscular atrophy, and "tabes dorsalis,"* so that, in spite of Romberg's having distinguished between the symptoms ataxia and paraplegia,† Dr. Gull deserves full credit for originality in his description of the first well-observed case, in 'Guy's Hospital Reports' for 1858.

Duchenne followed closely upon this with a tolerably complete history of the disease; for which we may hardly be inclined to agree with Trousseau in naming the disease after him, but which we must admit to be the greatest advance made up to that time. This had the further advantage of being taken up by Trousseau, who gave a good idea of the symptoms and course of ataxia to the numerous readers who studied with eagerness the 'Clinique Médicale' of that great physician; consequently, in the last few years many efforts have been made in France to acquire a more correct knowledge of it, and although these have led their authors in some instances to very different conclusions, yet the progress made there has been on the whole very decided. In England less has been written, and less attention paid to the subject, but it is evident that a generally correct notion of it is widely diffused; while, on the contrary, in Germany the unfortunate association of the disease, on its first description by Romberg, with 'tabes dorsualis,' has led to its being confused, by some of the best-informed German physicians, with several other diseases.

We will now describe briefly the symptoms and course of the disease, premising that it is much more common in men than in

* "Dorsualis," as used by the Germans, is the more classical form of the adjective.

† In the description of "Tabes Dorsualis" in the edition of his great work for 1857.

women (M. Topinard gives the proportion as 81 males to 33 females; Eisenmann, 50 males to 20 females); that it is very rare in early youth and in old age; and that most of those who suffer from it have been frequently exposed to cold and damp. Either without any other obvious cause, or after a cold distinctly caught, the first symptoms of the disease occur.

Duchenne has divided these (and his division has been followed by most subsequent writers on the subject) into three stages.

The first stage is characterised especially by pain, and by symptoms which point to the encephalon. The pains, which, according to Mr. Lockhart Clarke and most observers, are the most constant and earliest phenomena, are generally neuralgic in character, very sharp, lancinating, and only momentary, like a frequently repeated succession of electric shocks. Or, again, they may be dull, aching, rheumatic, and are then usually confined to one spot, and relieved by pressure or rubbing.

Of whichever kind they may be, they very often begin in the lower extremities, wander over the body, and finally settle in one leg or thigh, whence they proceed upwards; in many cases, however, their progress is quite irregular. They sometimes assume the form of painful constriction of the thorax, abdomen, or thighs. They are generally increased by excess in walking, drinking, or venery, or by constipation; and, still more notably, by cold and damp weather. So marked is the influence of the state of the atmosphere, that these patients dread the autumn and winter, and are very much better in summer and spring.

Generally, after these pains have been in existence for some time, but sometimes without their having been noticed at all, symptoms may be observed which show that the encephalon is involved. In very rare cases the disease has begun by violent headache, vertigo, photopsia, and tinnitus aurium (M. Carre's "cerebral" variety); in others (as in a case recorded by Trousseau) temporary hemiplegia may occur.

But, much more frequently, mischief is indicated by paresis of some of the cranial nerves. The second, third, and sixth nerves are those most often affected, the commonest symptoms being therefore, in their usual order of occurrence, strabismus (generally internal), ptosis, diplopia, amblyopia, and amaurosis.

On examining the affected eye with the ophthalmoscope at an early stage of the disorder, the capillaries are observed to be congested, the whole fundus being darker than is natural, but by degrees the retina becomes atrophied, is of a greyish colour, and surrounded with a white circle.

With regard to the state of the pupils, Romberg says that he has frequently found them contracted to the size of a pin's point in

"tabes dorsualis;"* and it would appear from Mr. Clarke's cases that this is the rule when the ocular nerves are otherwise unaffected; but ptosis and strabismus are more generally accompanied with dilated pupil.

A curious alteration of vision has been occasionally noticed; the patient has been found to see two images with one eye only open, or three with both open.

The other cranial nerves are much less frequently affected. Dr. Althaus says that the olfactory never is, but Mr. Clarke gives one case in which the sense of smell was almost entirely lost.

Almost all these affections of the cranial nerves disappear after a short time, with or without treatment, except ptosis and amblyopia, which usually go from bad to worse.

Spermatorrhœa, ending generally in anaphrodisia and impotence, is a very common symptom, but in a few rare cases (as Trousseau has especially noted) sexual desire and power are morbidly great. The bowels and bladder are generally sluggish; indeed, in one case which came under our own observation, retention and involuntary evacuation of urine was one of the most prominent symptoms, but this is rare. In all other respects the health seems to be unaffected.

With regard to the proportion of cases in which the early symptoms appear, statistics are as yet very imperfect. In 28 cases out of 125, collected by Topinard,† the cranial symptoms were entirely absent; in 14 out of 63, according to M. Carre, they preceded the pains, while in the remaining 49 we may suppose that the pains occurred first.

The average duration of this stage of the disease is from four to five years, but this is very variable, and a very few cases are recorded in which all its phenomena were absent, and the disease began abruptly.

The second stage is marked by the occurrence of the most important symptom of the disease, viz. irregularity (or "ataxia") of movement. Of this, as Mr. Clarke has pointed out, there are two distinct forms. In the earlier form there is mere unsteadiness of the limbs affected; the patient staggers and totters, especially on first beginning to walk or on turning round; he adopts various expedients to maintain his balance; he cannot stand with his eyes shut and his feet placed together, and, as the disease advances, cannot stir without keeping his eyes fixed on his feet. When the upper extremities are attacked, no delicacy in the use of the fingers or arms is possible.

In the second form the motor disturbance is exhibited in spas-

* 'Nervenkranheiten,' Bd. i, Abth. 3, p. 684.
† 'Union Médicale,' Mars, 1865.

modic movements of those muscles which the will intends to put in action. The limbs are flexed, or extended with a sudden violent jerk; they are strangely thrown about; the patient can no longer walk without support, and, when the disease reaches the muscles fixing the pelvis, can no longer stand at all, though he may be capable of exerting great force with his legs while sitting or lying down.

These two forms are evidently produced by different degrees of one morbid state—absence of co-ordination of the muscles.

In the former case the motor irregularity is owing to an inability to combine properly the numerous muscular contractions which are requisite for even the simplest movement; in the latter the natural harmony between the antagonist muscles, which is one of the elements of normal co-ordination, is also abolished; the flexors or extensors alone act, whence the jerking and violent movements.

In the immense majority of cases this striking symptom begins in the lower limbs, and spreads upwards from them; in only two instances (one recorded by M. Carre, the other by M. Vernay) it began in the upper extremities, in the latter case remaining confined to them, but in the former spreading downwards.

A certain loss of muscular power is sometimes complained of by patients, although it cannot be detected by the physician; and involuntary twitching (especially of the fingers) is not uncommon.

Cutaneous hypæsthesia (numbness) is an almost invariable symptom of this stage of the disease. It generally begins, and continues to be most noticeable, in the soles of the feet and the legs, whence it spreads to the thighs; in the upper extremities only the third and little fingers are affected. As the disease advances the numbness goes on, in most cases, to absolute anæsthesia; or sensibility to pain may be entirely lost, the sense of touch remaining intact, or sometimes sensation may be very tardy or obtuse, or the patient may be unable to tell in what part of the body he is touched; for all these varieties have been noticed.

The sense of pressure or weight is also frequently very blunt; while, on the contrary, that of temperature is seldom affected.

In rare instances there is great hypæsthesia, the slightest touch then producing extreme pain.

Reflex movements are usually excited with difficulty; but, in one remarkable case of Mr. Clarke's, "although the feet and legs were almost completely deprived of the sense of touch and pain, yet their surfaces were so susceptible to excito-motor impressions, that the slightest touch or brush threw the whole body into motion, and caused the patient to jump almost from his chair."

The last of the three periods into which Duchenne has divided the disease is simply the termination of the second. All the symptoms become worse, and the patient is hopelessly bedridden. From this

cause, and to a less degree from slight paraplegia, the muscles begin to lose their power; they become atrophied, and partially degenerated into fat. Painful spasms frequently occur in the affected limbs, and are among the most troublesome symptoms of this stage of the disease. The urine is retained, or is passed involuntarily; and either this, or sloughing of the back, is the most common cause of death.

The duration of the disease, from the first occurrence of ataxia of movement until the fatal termination, is very variable; it is often ten years, or even more.

The diagnosis of the disease is generally easy enough when it has passed into its second stage; but, before motor disturbance has been perceived, it is difficult, and very often impossible. The character, seat, intractability, and progress of the pains, will guide us to a suspicion of their nature, which will be heightened if ocular symptoms, especially strabismus and amblyopia* combined, be also present.

Even when ataxic symptoms are fully developed, it may be difficult to know whether these signify the existence of the disease we are speaking of, or that of some one of the other numerous affections in which unsteadiness of gait occurs. The following points will be the most important for a correct diagnosis. Unless the early history of the disease be quite unknown, we may at once set aside hysteria (which can make an excellent imitation of the symptom ataxia) and chronic alcoholism. General paralysis (in England at least) is almost always accompanied by its characteristic mental disturbances, and the motor irregularity extends to the face, tongue, and lips. There is also apparently real loss of muscular power, although Bouillaud, Wunderlich, and Dr. Skae consider that there is no actual paralysis, but only ataxia.† Syphilis may bear a closer resemblance to many of the symptoms of ataxia; but in a doubtful case the iodide of potassium would of course be tried.

Disease of the cerebellum is also attended with a tottering gait, which might easily be mistaken for ataxia, with strabismus and amblyopia; but these symptoms do not generally preserve the same order of appearance as in ataxia. The irregularity of movement in cerebellar disease is also shown by epileptiform convulsions, and "mouvements de manège;" there is generally difficulty of speech,

* Dr. Hughlings Jackson stated, and Mr. Clarke has repeated, that the ophthalmoscopic appearances of amaurosis from ataxia are different from those of amaurosis from cerebral disease; but this would appear not to be the case. See a letter from Dr. Althaus in the 'Lancet' of June 17th, 1865.

† Jaccoùd quotes three remarkable cases, in which, shortly before death, patients who had long been suffering from ataxia presented symptoms of general paralysis ("délire ambitieux"). Had we more details given, these cases might lead to clear up some of the obscurity attending the form of general paralysis, where motor signs precede the mental disturbance.

vertigo, fixed pain in the back of the head, and vomiting; none of which symptoms form part of ataxia.*

On opening the vertebral canal after death the membranes are often found to be thickened and opaque, and the pia mater adherent to the posterior columns; these appearances are observed in about one half of the autopsies, according to Dr. Althaus. The back of the cord looks flattened, and the posterior median fissure is generally obliterated. The posterior columns, instead of being white, are either entirely grey and semitransparent, or are streaked with bands of that colour, running up and down the cord. This change is most common and most distinct in the lumbar region, and in advanced cases has been seen to extend as far forward as the lateral columns, but the anterior ones are never involved. If a section of the cord be made, this grey colour is perceived to be not merely superficial, but to affect the posterior columns, wherever it is apparent on the surface, in their whole depth. It is also noticed that they are decidedly smaller than natural. When the cord examined is in an early stage of the disease, the affected portion is found of the normal consistence, or even softened, but in the great mass of more advanced cases it is unnaturally hard, almost as much so as a brain steeped in alcohol; hence the name "sclerosis," which has been given to the disease.

The posterior roots of the spinal nerves in the part diseased, and the nerves composing the cauda equina are almost always similarly affected; and, in twenty-one cases out of forty-eight, the posterior cornua of grey matter have been found in the same state. This much, at any rate, would seem to be invariably the rule in the vertebral canal, that the disease spreads from the posterior columns of the cord as a centre. On the other hand, as Mr. Clarke has especially pointed out, such changes as are remarked in the encephalon seem to spread from the distal extremities of the cranial nerves towards their centre. Thus, the optic nerve, chiasma, and tracts are frequently sclerosed; sometimes the corpora geniculata, and, more rarely, the corpora quadrigemina. The other cranial nerves are but seldom affected. The cerebellum, owing to the part it has been supposed to play in the co-ordination of muscular movement, has been almost invariably examined, and found healthy.

Microscopical examination of a portion of diseased cord shows that, in the early stages, the nerve-tubes are diminished in number and size, and granular bodies, produced by their disintegration, appear. As the disease advances, and the atrophy of the nerve-tubes becomes more and more marked, their place is taken by fibres of ordinary connective tissue, by nuclei, and by small cells (Robin's "myélocythes"). The capillaries are often thickened or atheroma-

* See papers on "Cerebellar Disease," by MM. Leven and Ollivier, in 'Archives Gén. de Méd.,' 1862 and 1863. Mr. Clarke mentions a case of ataxia, in which vomiting was a prominent symptom.

tous, and they are frequently surrounded, in long-standing cases, by oil-globules or corpora amylacea. The anatomical change, therefore, consists essentially of two parts—1. Diminution of the nerve-tubes, in both number and size. 2. Development of adventitious products. The former of these changes only is noticed in the posterior roots of the spinal nerves; but the cranial nerves have been found generally to contain new products, the corpora amylacea being especially abundant in the optic and hypoglossal.

When the grey matter of the cord, or of the ganglia in the posterior roots, is involved, the nerve-cells in it are almost invariably found healthy.

As to the connection between the anatomical state we have just described and the symptoms, the following is the most probable explanation. It would appear that by the destruction of the posterior fibres of the cord the sensory nerves which supply the muscles are cut off from the spinal motor centres, which are now believed to be the centres of muscular co-ordination, and that thus ataxia is produced. The hypæsthesia which is so generally a symptom of the disease is probably owing mainly to the destruction of the nerves themselves, and only partly to the sclerosis of the cord; for we have every reason to suppose that sensory impressions are conducted up the grey matter of the cord, which is generally intact in this disease.

The cause of this remarkable anatomical change, which affects at the same time the cranial nerves and the lower part of the spinal cord, is as yet unknown to us. We may compare locomotor ataxia, on the one hand, with cases of paraplegia produced by exposure to cold and wet, or by urinary disease; and, on the other, with instances collected by Dr. Handfield Jones (in the appendix to his 'Functional Nervous Disorders') of retinitis, amaurosis, and paralysis of various cranial nerves caused by remote irritation. The same author has also shown it to be very probable that symptoms of the same kind as those we have been considering may be produced, without any organic disease of the nervous centres, by the diatheses syphilis and rheumatism. These analogies would lead us to ask whether the cold caught (which most ataxic patients speak of as the cause of all their ills) is not really capable, given some special predisposition, of producing this disease, either by checking the secretions, or (to borrow Dr. H. Jones's phrase) by its directly "inhibitory" influence.

What the special predisposition may be is as obscure in this as in most other diseases. At any rate this seems to be established, that sexual excess or abuse has little or nothing to do with it. Probably the only reason why some connection is still presumed to exist, by most men, between these vices and progressive ataxia, is because there is still a confusion in their minds between this disorder and

"tabes dorsalis," a name which includes all cases of exhaustion of the cord by venereal excess. The great majority of ataxic cases present no history of anything of this kind, but, on the other hand, point very decidedly to habitual exposure to cold and wet as the predisposing cause.

Many German, and some few French writers, of whom Jaccoud is the most prominent, have laid undue stress upon the anatomical character of the disease, and have looked upon it as being essentially a sclerosis of the spinal cord; but this view is generally condemned by those who have observed the cranial symptoms which, in a great majority of cases, are part of the evolution of the disease. This much only can be said in its favour, that the cord may be sclerosed in many other different affections,* especially in the disease known as "spedalsked," of which motor ataxia is an early symptom.

We have fortunately passed the time when Romberg could write with truth these despairing words:—"No patient suffering from this disease can be cured; death awaits them all; and the only consolation which can be given to those who love life, is the long duration of the disease." We condemn as emphatically as he does the useless cruelty of repeated issues, setons, and bleedings; but these are plans which no enlightened practitioner at the present day is likely to adopt, and we are firmly convinced that judicious treatment can do much, generally to relieve, sometimes to cure, those suffering from ataxia. Unfortunately the disease is seldom or never diagnosed until extensive destruction of nerve-tissue has taken place; but in this respect we may hope for improvement as it becomes better known.

Under the head of remedies which have been tried and been found of no avail, we may mention opium, bromide of potassium, and secale cornutum. But at least they have done no harm, while strychnia has never done any good, and, in one case recorded by M. Carre, produced violent pain; iodide of potassium, too, has appeared in some cases to accelerate the course of the disease.

The vapour, lamp, and Turkish baths have also disappointed the well-grounded hopes which had been placed in them; but sulphurous baths and electricity are especially commended by Dr. Althaus as palliatives. The severe pains of the first stage are relieved by belladonna, cannabis indica, and oil of turpentine, internally, and by dry-cupping over the neck and back.

Eisenmann found decided benefit in four cases of ataxia from the

* Thus M. Charcot, besides those cases in which the disease has spread from the posterior columns of the cord, has found it in the lateral columns only (in two cases of permanent muscular contractions in hysterical patients), and in circumscribed patches, distributed through the cord irregularly.—('Union Médicale,' 9 Mars, 1865.)

regular use of gymnastics; this is at any rate worth remembering as an adjuvant to more active treatment.

But the only remedy which has ever obtained any considerable reputation in the treatment of this disease is the nitrate of silver, introduced by Wunderlich. It is difficult, as in all other questions of therapeutics, to make out its precise value; but it would appear to have every chance of being successful when employed at an early period; and, when it fails, it seems to do so either from being given too late or without sufficient perseverance. The good effects it produces are too frequently only temporary; hence probably its prolonged exhibition is advisable. Of course the usual precautions will be taken to prevent its colouring the skin, or disturbing the stomach, bowels, or bladder. Dr. Althaus combines with it the hypophosphite of soda, which he considers beneficial.

Arsenic, which would on theoretical grounds be recommended, has been tried several times, but with partial success in one case only, recorded by M. Teissier; we confess that we should ourselves be very strongly disposed to give it a further trial.

M. Carre conjectures that possibly the internal administration of the Calabar bean might do good. We do not see grounds for putting any faith in it, and, if we may ourselves hazard similar guesses, would rather suggest aconite as likely to relieve the pains, and conium as being possibly a curative agent.

It is needless to add that the general health should be kept up by good food and tonics, cod-liver oil being especially valuable, considering the relation of fatty bodies to the nutrition of nervous tissue. The bowels should be kept well open, for this alone will frequently relieve the pains of the first period of the disease. In spite of the truth of Romberg's remark that long journeys are injurious to these patients, we should be inclined, seeing the improvement in the first stage effected by warm weather, to send those whose circumstances would allow it to winter in some tropical or semi-tropical climate.