rect in stating that "acute ascending paralysis—defined so as to exclude all cases in which the sensory symptoms are prominent, or in which well-marked bulbar symptoms are not present—must therefore be regarded as a clinical entity for which no corresponding lesion has as yet been discovered." It is not improbable that some toxic agent is responsible for the symptoms.

Symptoms.—Weakness of the legs, gradually progressing, often with tolerable rapidity, is the first symptom. In some cases within a few hours the paralysis of the legs becomes complete. The muscles of the trunk are next affected, and within a few days, or even less in more acute cases, the arms are also involved. The neck muscles are next attacked, and finally the muscles of respiration, deglutition, and articulation. The reflexes are lost, but the muscles neither waste nor show electrical changes. The sensory symptoms are variable; in some cases tingling, numbness, and hyperæsthesia have been present. In the more characteristic cases sensation is intact and the sphincters are uninvolved. Enlargement of the spleen has been noticed in several cases. The course of the disease is variable. It may prove fatal in less than two days. Other cases persist for a week or for two weeks. In some instances recovery has occurred, but in a large proportion of the cases the disease is fatal.

The *diagnosis* is difficult, particularly from certain forms of multiple neuritis, and if we include in Landry's paralysis the cases in which sensation is involved, distinction between the two affections is impossible. We apparently have to recognize the existence of a rapidly advancing motor paralysis without involvement of the sphincters, without wasting or electrical changes in the muscles, without trophic lesions, and without fever—features sufficient to distinguish it from either the acute central myelitis or the polio-myelitis anterior. It is doubtful, however, whether these characters always suffice to enable us to differentiate the cases of multiple neuritis.

IV. CHRONIC AFFECTIONS OF THE SPINAL CORD.

I. Spastic Paraplegia.

Definition.—Loss of power with spasm of the muscles of the lower extremities.

While clinically spastic paraplegia, or, as it is sometimes called, *tabes dorsalis spasmodique*, is a well-defined, readily recognizable affection, etiologically and anatomically it presents marked differences, and various groups must be separated, all of which present, however, the combination of spasm with loss of power. As the pyramidal tracts are involved, the term lateral sclerosis is sometimes used as the equivalent of spastic paraplegia. I shall consider the following forms:

(1) **Secondary Spastic Paralysis.**—After any tranverse lesion of the cord, whether the result of slow compression (as in caries), chronic myelitis, the pressure of tumor, chronic meningo-myelitis, or multiple sclerosis, degeneration takes place in the pyramidal tracts below the point of disease. The legs soon become stiff and rigid, and the reflexes increase. It happens occasionally, as Bastian has shown, that in compression paraplegia the limbs may be flaccid without increase in the reflexes—*paraplegie flasque* of the French. The condition of the patient in these secondary forms varies very much. In chronic myelitis or in multiple sclerosis he may be able to walk about, but with a characteristic spastic gait. In the compression myelitis, in fracture, or in caries, there may be complete loss of power with rigidity.

(2) **Primary Spastic Paraplegia.**—This is believed to depend upon a primary sclerosis of the lateral or pyramidal tracts. Clinically it is common to meet with cases in adults, particularly in syphilitic subjects, who have pains in the back, perhaps a girdle sensation, and a gradually developing, progressive spastic paraplegia. It may be impossible from the history or the physical examination to determine whether the condition is secondary to a transverse myelitis or a meningo-myelitis, or whether the lesion is a primary degeneration of the pyramidal tracts. The question is still debated whether a primary lesion of the lateral tracts ever takes place, or whether, in such instances, there is not always some lesion of the motor cells in the anterior horns. Cases may persist for years without any atrophy. In other instances there are signs of involvement of the posterior columns as well, forming the condition of ataxic paraplegia, which will be considered separately. So far as I know, the only case which is claimed to demonstrate the existence of a primary lateral sclerosis is that of Dreschfeld's, which occurred in 1881.

The *symptoms* of spastic paraplegia are very distinctive. The patient complains of feeling tired, of stiffness in the legs, and perhaps of pains of a dull aching character in the back or in the calves. There may be no definite loss of power, even when the spastic condition is well established. In other instances there is definite weakness. The stiffness is felt most in the morning. In a well-developed case the gait is most characteristic. The legs are moved stiffly and with hesitation, the toes drag and catch against the ground, and, in extreme cases, when the ball of the foot rests upon the ground a distinct clonus develops. The legs are kept close together, the knees touch, and in certain cases the adductor spasm may cause cross-legged progression. On examination, the legs may at first appear tolerably supple, perhaps flexed and extended readily. In other cases the rigidity is marked, particularly when the limbs are extended. The spasm of the adductors of the thigh may be so extreme that the legs are separated with the greatest difficulty. In cases of this extreme rigidity the patient usually loses the power of walking. The nutrition is well maintained, the muscles may be hypertrophied. The reflexes are greatly

increased. The slightest touch upon the patellar tendon produces an active knee-jerk. The rectus clonus and the ankle clonus are easily obtained. In some instances the slightest touch may throw the legs into violent clonic spasm, the condition to which Brown-Séquard gave the name of spinal epilepsy. The superficial reflexes are also increased. The arms may be unaffected for years, but as a late manifestation rigidity may develop.

The *diagnosis* is readily made, but it is often very difficult to determine accurately the nature of the underlying pathological condition. A history of syphilis is present in many of the cases. The course of the disease is progressively downward. Years may elapse before the patient is bed-ridden. Involvement of the sphincters, as a rule, is late; occasionally, however, it is early. The sensory symptoms rarely progress and the patients may retain the general nutrition and enjoy excellent health. Ocular symptoms are rare.

(3) **The Spastic Paraplegia of Infants** (*Paraplegia Cerebralis Spastica* —Heine).—This is usually a birth palsy, often the result of difficult labor. In twenty-three of the twenty-four of Little's cases, there was either difficult labor or premature delivery. Several children may be affected in a family. Gee reports two cases in one family, Schultze three, and with Latimer I saw a brother and a sister with the disease. In this connection it is interesting to note that Bernhardt has recently described a family form of spastic paraplegia, in which four brothers were affected, the disease developing in each about the thirtieth year. The stiffness of the legs may not be noticed for some months after birth, but usually on dressing the child the mother notices the rigidity. When attempts are made to walk the stiffness and awkwardness then become apparent. On standing, the attitude is very characteristic. There is talipes equinus, varying from the slightest raising of the heel to a condition in which the child stands on tiptoe. In older children, as they walk, the toe-cap of the shoe is usually much worn. The strong adductor action may produce typical cross-legged progression, in which each foot is dragged over and planted in front, or even on the other side of its fellow. In attempting to flex the legs there is a marked resistance, which gradually yields—the lead-pipe contraction, as Weir Mitchell calls it. The reflexes are increased, though in some children it is not an easy matter to obtain them. The ankle clonus, as a rule, is not obtainable. Sensation is unimpaired, and the bladder and rectum are not involved.

The symptoms of this affection in children are almost identical with the spastic paraplegia of adults. The arms may be involved—spastic diplegia. The disease is probably of cortical origin. There are frequently symptoms indicating cerebral defects, such as idiocy, imbecility, and nystagmus. Some of the cases depend, no doubt, upon bilateral meningeal hæmorrhage occurring during delivery. Others are probably due to arrest of development of the pyramidal tracts. This condition in children must not be confounded with tetany or with the pseudo-paralytic rigidity so often associated with rickets.

(4) **Ataxic Paraplegia.**—This name is applied by Gowers to a disease characterized clinically by a combination of ataxia and spastic paraplegia, and anatomically by involvement of the posterior and lateral columns.

The disease is most common in middle-aged males. Exposure to cold and traumatism have been occasional antecedents. In striking contrast to ordinary tabes a history of syphilis is rarely to be obtained.

The anatomical features are a sclerosis of the posterior columns, which is not more marked in the lumbar region and not specially localized in the root zone of the postero-external columns. The involvement of the lateral columns is diffuse, not always limited to the pyramidal tracts, and there may be an annular sclerosis.

The *symptoms* are well defined. The patient complains of a tired feeling in the legs, not often of actual pain. The sensory symptoms of true tabes are absent. An unsteadiness in the gait gradually develops with progressive weakness. The reflexes are increased from the outset, and there may be well-developed ankle clonus. Rigidity of the legs slowly comes on, but is rarely so marked as in the uncomplicated cases of lateral sclerosis. From the start, incoördination is a well-characterized feature, and the difficulty of walking in the dark or swaying when the eyes are closed may, as in true tabes, be the first symptom to attract attention. In walking the patient uses a stick, keeps the eyes fixed on the ground, the legs far apart, but the stamping gait, with elevation and sudden descent of the feet, is not often seen. The incoördination may extend to the arms. Sensory symptoms are rare, but Gowers calls attention to a dull, aching pain in the sacral region. The sphincters usually become involved. Eye symptoms are rare. Late in the disease mental symptoms may develop, similar to those of general paresis.

In well-marked cases the *diagnosis* is easy. The combination of marked incoördination with retention of the reflexes and more or less spasm are characteristic features. The absence of ocular and sensory symptoms is an important point.

(5) **Hysterical Spastic Paraplegia.**—There is no spinal-cord disease which may be so accurately mimicked by hysterical patients as spastic paraplegia. There is wasting in the hysterical paraplegia, the sensory symptoms are not marked, the loss of power is not complete, and there is not that extensor spasm so characteristic of organic disease. The hysterical contracture will be considered later.

The reflexes are, as a rule, increased. The knee-jerk is present, and there may be well-developed ankle clonus. Gowers calls attention to the fact that it is usually a spurious clonus, "due to a half-voluntary contraction in the calf muscles." A true clonus does occur, however, and there may be the greatest difficulty in determining whether or not the case is one of hysterical paraplegia.

(6) **Primary Combined Sclerosis** (Putnam).—In addition to the ataxic paraplegia just mentioned, here may be considered certain cases which are characterized anatomically by a relatively chronic sclerosis of the posterior columns, of the lateral columns, chiefly the pyramidal tract, and also of the cerebellar tract. With these are usually associated more acute changes in adjoining areas, either diffuse or systemic, some grade of degeneration in the gray matter, and involvement of the nerve roots. This form has been studied by J. J. Putnam and Dana. The cases are usually in women —seven out of nineteen collected by Dana; the ages, from forty-five to sixty-four. The disease runs a rather rapid course. Neuropathic inheritance is present in some instances. Putnam thinks that possibly both lead and arsenic play a part in the etiology.

The *symptoms* are both sensory and motor. The onset is usually with numbness in the extremities, progressive loss of strength, and emaciation. Paraplegia gradually develops, before which there have been, as a rule, spastic symptoms with exaggerated knee-jerk. The arms are affected less than the legs. Mental symptoms similar to dementia paralytica may develop toward the close.

The *diagnosis* of this mixed sclerosis rests upon the combination of sensory and motor symptoms with the presence of exaggerated reflexes. As stated, the sensory features consist chiefly of paræsthesia, and there may be difficulty in distinguishing the condition from multiple neuritis. The frequency of the disease in more or less enfeebled or anæmic women past middle life is also an important feature.

Treatment of Spastic Paraplegia.—In the majority of cases spastic paraplegia is incurable. The cases which result from transitory compression, as in caries, may get well; but in the other forms the disease is uniformly progressive, and remedies have little or no control. When syphilis is suspected a thorough course of mercury and iodide of potassium should be given. Scrupulous attention should be paid to the bladder symptoms, and the same measures may be used as will be advised in locomotor ataxia. In the infantile form of paraplegia much may be done by the orthopædic surgeon to overcome rigidity and contracture. In several instances I have known persistent friction with forcible flexion and extension and the application of proper apparatus enable a patient to get about comfortably.

II. Locomotor Ataxia

(*Tabes Dorsalis; Posterior Spinal Sclerosis*).

Definition.—An affection of the nervous system characterized clinically by incoördination, with sensory and trophic disturbances and involvement of the special senses, particularly the eyes. Anatomically there are found sclerosis of the posterior columns of the cord, foci of degeneration in the basal ganglia, and sometimes chronic degenerative changes in the cortex cerebri.

Etiology.—It is a wide-spread disease, more frequent in cities than in the country. The relative proportion may be judged from the fact that of 1,816 cases in my neurological dispensary in two years there were 25 cases of locomotor ataxia. Males are attacked more frequently than females, the proportion being at least ten to one. Mitchell has called attention to the fact that it is a rare disease in the negro. Of 25 cases at my clinic, 3 were in negroes. It is a disease of adult life, a majority of the cases occurring between the thirtieth and fortieth years. Occasionally cases are seen in young men. The form of ataxia which occurs in children is a different disease. Of special causes syphilis is the most important. According to the figures of Erb, Fournier, and Gowers, in from fifty to seventy-five per cent of all cases there is a history of this disease. Erb's recent figures are most striking; of 300 cases of tabes in private practice 89 per cent had had syphilis.

Excessive fatigue, overexertion, exposure to cold and wet, and sexual excesses are all assigned as causes. There are instances in which the disease has closely followed severe exposure. James Stewart has noted that the Ottawa lumbermen, who live a very hard life in the camps during the winter months, are frequently the subjects of locomotor ataxia. Trauma has been noted in a few cases. Alcoholic excess does not seem to predispose to the disease. Among patients in the better classes of life I do not remember one in which there had been a previous history of prolonged drunkenness.

Morbid Anatomy.—When a patient has died in the advanced stage of the disease the following are the most important changes:

(*a*) The peripheral nerves may show signs of degeneration. Neuritis may indeed be present even when there have been no special symptoms indicating it. In other instances there is not only neuritis, but muscular atrophy.

(*b*) The posterior roots of the spinal cord are small, gray, and atrophic.

(*c*) The meninges of the posterior and lateral columns are thickened, more firmly adherent than normally, and the blood-vessels usually show signs of arterio-sclerosis.

(*d*) The changes in the spinal cord are as follows: (1) In advanced cases the posterior columns are uniformly sclerotic and the dorsal and lumbar regions are most extensively involved. In long-standing cases there is generally an increase of connective tissue throughout the cord and there may be degeneration (2) of the ascending antero-lateral tract; (3) of the direct cerebellar tract; (4) of the pyramidal tract.

(*e*) In early cases the course of the anatomical changes may be traced. The steps in the process are as follows: The posterior root-zone of Charcot is first involved, often with the fibres of the posterior root, so that it has been thought to begin perhaps as a neuritis of these roots within the vertebral canal. The narrow strip which lies between the pyramidal tract

and the posterior cornu, known as Lissauer's tract, is early involved, together with the nerve-cells of the adjacent Clarke's vesicular column. In what is known as the pre-ataxic stage these may be the only alterations. Subsequently the sclerosis extends widely in the postero-external, and subsequently in the postero-median columns.

(*f*) The cerebral changes—of less consequence than the spinal—may consist of (1) sclerosis in the restiform bodies, in the inferior peduncles of the cerebellum, and of certain of the cranial nerves, particularly the third, the optic, and the auditory; (2) cortical changes, consisting in some cases of a diffuse meningo-encephalitis.

Symptoms.—These are best considered under the three stages of pre-ataxic, ataxic, and paralytic.

Pre-ataxic Stage.—The following are the most characteristic features of this period:

Pains, usually of a sharp stabbing character; hence the term, lightning pains. They last for only a second or two and are most common in the legs. They may be associated with a hot, burning feeling. Occasionally herpes may develop at the site of the pain. They may occur at irregular intervals, and are more prone to follow excesses or to come on when the health is impaired.

Ocular Symptoms.—(*a*) Ptosis, which may be single or double and is by no means uncommon either alone or (*b*) in association with external strabismus. The first complaint may be of double vision. Occasionally there may be paralysis of all the external muscles of the eye, producing ophthalmoplegia externa. (*c*) Argyll-Robertson pupil, in which, as already mentioned, there is loss of the iris reflex to light, but contraction during accommodation. The pupils are usually small—spinal myosis. (*d*) Optic atrophy. This is often an early, or even the first symptom. The loss of vision progresses, and in a large majority of cases leads to total blindness.

Loss of the Knee-jerk.—This is one of the earliest symptoms, and may occur years before there is ataxia. Taken alone it is of no moment, as there are individuals in whom the knee-jerk is absent; but in connection with the lightning pains and the ocular symptoms, it is of special importance. These are the most common symptoms of the pre-ataxic stage, and may persist for years without the development of incoördination. The patient may look well and feel well, and be troubled only by occasional attacks of lightning pains; or there is persistent ptosis, external strabismus develops, or, what is more serious, a progressive atrophy of the optic nerve. There is often a gradual loss of sexual power.

The disease may never progress beyond this stage, and when optic atrophy develops early and leads to blindness, the ataxia rarely, if ever, supervenes. There is a sort of antagonism between the ocular symptoms and the progress of the ataxia. Charcot lays considerable stress upon this, and Déjérine assured me that of the enormous tabetic material at the

Bicêtre in not a single instance in which optic atrophy had come on early and progressed to blindness was the patient ataxic, although there were cases which had had the lightning pains and lesions of the optic nerves for twenty-five years.

Ataxic Stage.—*Motor Symptoms.*—The ataxia develops gradually. One of the first indications to the patient is inability to get about readily in the dark or to maintain his equilibrium when washing his face with the eyes shut. When the patient stands with the feet together and the eyes closed, he sways and has difficulty in maintaining his position. This is known as Romberg symptom. On turning quickly he is apt to fall. Gradually the characteristic ataxic gait develops. The patient, as a rule, walks with a stick, the eyes are directed to the ground, the body is thrown forward, and the legs are wide apart. In walking, the leg is thrown out violently, the foot is raised too high and is brought down in a stamping manner with the heel first, or the whole sole comes in contact with the ground. Ultimately the patient may be unable to walk without the assistance of two canes. This gait is very characteristic, and unlike that seen in any other disease. The incoördination is not only in walking, but in the performance of other movements. If the patient is asked, when in the recumbent posture, to touch the knee with one foot, the irregularity in the movement is very evident. Incoördination of the arms is less common, but usually develops in some grade. It may in rare instances exist before the incoördination of the legs. In the large number of ataxics which frequented the Infirmary for Nervous Diseases at Philadelphia, there was only one, so far as I remember—at Weir Mitchell's clinic—in which the arms were first affected. It may be tested by asking the patient to close his eyes and to touch the tip of the nose or the tip of the ear with the finger, or with the arms thrust out to bring the tips of the fingers together. The incoördination may early be noticed by a difficulty which the patient experiences in buttoning his collar or in performing one of the ordinary routine acts of dressing.

One of the most striking features of the disease is that with marked incoördination there is no loss of muscular power. The grip of the hands may be strong and firm, the power of the legs, tested by trying to flex them, may be unimpaired, and their nutrition, except toward the close, may be unaffected.

Sensory Symptoms.—The lightning pains may persist. They vary greatly in different cases. Some patients are rendered miserable by the frequent occurrence of the attacks; others escape altogether. In addition, common symptoms are tingling, pins and needles, particularly in the feet, and areas of hyperæsthesia or of anæsthesia. The patient may complain of a change in the sensation in the soles of the feet, as if cotton was interposed between the floor and the skin. Sensory disturbances occur less frequently in the hands. Retardation of tactile sensation is common, and a pin-prick on the foot, instead of being instantaneously felt, is not per-