Symptoms.—The disease, which is of slow development, makes its appearance, as a rule, about adolescence, and may persist for fifteen or twenty years. There are irregular pains, chiefly in the cervical region; muscular atrophy develops, which may be confined to the arms, or sometimes extends to the legs. The reflexes are increased and a spastic condition develops in the legs. Ultimately the clinical picture may be that of an amyotrophic lateral sclerosis. The tactile sensation is usually intact and the muscular sense is retained, but painful and thermic sensations are not recognized, or there may be in rare instances complete anæsthesia of the skin and of the mucous membranes (Déjérine). This combination of loss of painful and thermic sensations with paralysis of an amyotrophic type is regarded as pathognomonic of the disease. The special senses are usually intact and the sphincters uninvolved. Trophic troubles are not uncommon. Owing to the loss of the painful and heat sensations, the patients are apt to injure themselves. A man aged seventy, whom I saw with Déjérine at the Bicêtre, had had the symptoms for over twenty-five years. Loss of sensation had preceded the atrophy, and the terminal phalanx of the middle finger was charred, as he experienced no sensation whatever when the hot end of the cigarette neared his finger. Scoliosis also may be present in these cases. The loss of painful and thermic impressions is due to the fact that these pass to the brain in the peri-ependymal gray matter, particularly that portion in the posterior roots, which is almost constantly involved in syringo-myelia. The tactile sensation is retained because the postero-external column is uninvolved.

In typical cases the *diagnosis* is easy. The combination of an amyotrophic paralysis, the picture of progressive muscular atrophy of the Aran-Duchenne type, with retention of tactile and loss of thermic and painful sensation, is probably pathognomonic of the disease. Of affections with which it may be confounded, anæsthetic leprosy is the most important, since the anæsthesia and the wasting may closely simulate it; but, as a rule, in leprosy trophic changes are more or less marked. There is often loss of phalanges and there is no characteristic dissociation of sensory impressions.

There is a remarkable affection confined to a district of Brittany and known as Morvan's disease, after the physician who described it. The disease is chronic and characterized by neuralgic pains, cutaneous anæsthesia, and painless and destructive whitlows. In Gombault's autopsy neuritis was found, but it could not be decided, owing to the state of the cord when examined, whether cavities existed or not. Joffroy reports a case in which syringo-myelia was present and claims the affections are identical. The curious distribution of the disease and the fact that at least 20 cases have occurred in a population of 5,000, suggest that it is possibly a peripheral neuritis of infectious origin. Church, of Chicago, has reported case in which, with features believed to be characteristic of syringo-myelia, the patient had the painless and destructive whitlows which form so special a feature in Morvan's disease.

V. Compression of the Spinal Cord (*Compression Myelitis*).

Definition.—Interruption of the functions of the cord by slow compression.

Etiology.—Caries of the spine, new growths, aneurism, and parasites are the important causes of slow compression. Caries, or Pott's disease, as it is usually called, after the surgeon who first described it, is in the great majority of instances a tuberculous affection. In a few cases it is due to syphilis and occasionally to extension of disease from the pharynx. It is most common in early life, but may occur after middle age. It follows trauma in a few cases. Compression occasionally results from aneurism of the thoracic aorta or the abdominal aorta, in the neighborhood of the cœliac axis.

Malignant growths frequently cause a compression paraplegia. A retroperitoneal sarcoma or the lymphadenomatous growths of Hodgkin's disease may invade the vertebræ. More commonly, however, the involvement is secondary to scirrhus of the breast.

Of parasites, the echinococcus and the cysticercus occasionally occur in the spinal canal.

Symptoms.—These may be considered as they affect the bones, the nerves, and the cord.

(1) **Vertebral.**—In malignant disease and in aneurism erosion of the bodies may take place without producing any deformity of the spine. In caries, on the other hand, it is the rule to find more or less deformity, amounting often to angular curvature. The compression is largely due to the thickening of the dura and the presence of caseous and inflammatory products between this membrane and the bone. The compression is rarely produced directly by the bone. Pain is a constant and, in the case of aneurism and tumor, agonizing feature. In caries, the spinal processes of the affected vertebræ are tender on pressure, and pain follows jarring movements or twisting of the spine. There may be extensive tuberculous disease without much deformity, particularly in the cervical region.

(2) **Nerve-root Symptoms.**—These result from compression of the nerve-roots as they pass out between the vertebræ. It is remarkable how frequently, even in extensive caries, they escape and the patient does not complain of radiating pains in the distribution of the nerves from the affected segment. Pains are more common in cancer of the spine secondary to that of the breast, and in such cases may be agonizing. There may be acutely painful areas of hyperæsthesia of the skin or anæsthesia—the *anæsthesia dolorosa*. Trophic disturbances may occur, particularly herpes. In the cervical or lumbar regions pressure on the anterior roots may give rise to wasting of the muscles supplied by the affected nerves.

(3) **Cord Symptoms.** (*a*) *Cervical Region.*—Not infrequently the caries is high up between the axis and the atlas or between the latter and the occipital bone. In such instances a retropharyngeal abscess may be

present, giving rise to difficulty in swallowing. There may be spasm of the cervical muscles, the head may be fixed, and movements may either be impossible or cause great pain. In a case of this kind in the Montreal General Hospital movement was liable to be followed by transient, instantaneous paralysis of all four extremities, owing to compression of the cord. In one of these attacks the patient died.

In the lower cervical region there may be signs of interference with the cilio-spinal centre and dilatation of the pupils. Occasionally there is flushing of the face and ear of one side or unilateral sweating. Deformity is not so common, but healing may take place with the production of a callus of enormous breadth, and complete rigidity of the neck. The nerves of the upper extremities may be involved, and shooting pains may occur in the arm.

(*b*) *Dorsal Region.*—The deformity is here more marked and pressure symptoms are more common. The time of onset of the paralysis varies very much. It may be an early symptom, even before the curvature is manifest. More commonly it is late, occurring many months after the curvature has developed. The paraplegia is slow in its development; the patient at first feels weak in the legs or has disturbance of sensation, numbness, tingling, pins and needles. The girdle sensation may be marked, or severe pains in the course of the intercostal nerves. Motion is, as a rule, more quickly lost than sensation. Finally, there is complete interruption with the production of paraplegia, usually of the spastic type, with exaggeration of the reflexes. This may persist for months, or even for more than a year, and recovery still be possible.

(*c*) *Lumbar Region.*—In the lower dorsal and lumbar regions the symptoms are practically the same, but the sphincter centres are involved and the reflexes are not exaggerated.

Diagnosis.—Caries is by far the most frequent cause of slow compression of the cord, and when there are external signs the recognition is easy. There are cases in which the exudation in the spinal canal between the dura and the bone leads to compression before there are any signs of caries, and if the root symptoms are absent it may be extremely difficult to arrive at a diagnosis. Janeway has called attention to persistent lumbago as a symptom of importance in masked Pott's disease, particularly after injury. Brown-Séquard's paralysis is more common in tumor and in injuries than in caries. Pressure on the nerve-roots, too, is less frequent in caries than in malignant disease. The cervical form of pachymeningitis also produces a pressure paralysis, the symptoms of which have already been detailed. Pressure from cancer is naturally suggested when spinal symptoms follow within a few years after an operation. In paraplegia following tumor of the vertebra secondary to cancer of the breast, and in the erosion of the spine by retroperitoneal growths, the suffering is most intense. The condition has been well termed *paraplegia dolorosa*.

Treatment.—In compression by aneurism or tumor the condition is hopeless. In the former the pains are often not very severe, but in the latter morphia is always necessary. On the other hand, compression by caries is often successfully relieved even after the paralysis has persisted for a long period. When caries is recognized early, rest and support to the spine by the various methods now used by surgeons may do much to prevent the onset of paraplegia. When paralysis has developed, rest with extension gives the best hope of recovery. It is to be remembered that restoration may occur after compression of the cord has lasted for many months, or even more than a year. Cases have been cured by rest alone; the extradural and inflammatory products are absorbed and the caries heal. The most brilliant results in these cases have been obtained by suspension, a method introduced by J. K. Mitchell in 1826, and pursued with remarkable success by his son, Weir Mitchell. During my association with the Infirmary for Nervous Diseases I had numerous opportunities of witnessing the really remarkable effects of persistent suspension, even in apparently desperate and protracted cases. Mitchell's conclusions are that suspension should be employed early in Pott's disease; that used with care it enables us slowly to lessen the curve; that in these cases there must be, in some form, a replacement of the crumpled tissues; that unless there is great loss of power the use of the spine-car or chair of J. K. Mitchell enables suspension, especially in children, to be combined with some exercise; that no case of Pott's disease should be considered desperate without its trial; that suspension has succeeded after failures of other accepted methods; that the pull probably acts more or less directly on the cord itself, and that the gain is not explicable merely by obvious effects on the angular bony curve; that the methods of extension to be used in carious cases may be very varied, provided only we get active extension; that the plan and the length of time of extension must be made to conform to the needs, endurance, and sensation of the individual case. It may be months before there are any signs of improvement. In protracted cases, after suspension has been tried for months, laminectomy may be considered, and has in some instances been successful.*

The general treatment of caries is that of tuberculosis—fresh air, good food, cod-liver oil, and arsenic. Counter-irritation in these instances is of doubtful value.

Unilateral Lesions of the Spinal Cord (Brown-Séquard's Paralysis).—Tumors, stab wounds, and less frequently fracture or caries, may destroy one half of the cord, causing a peculiar and definite palsy, which was first recognized by Brown-Séquard, after whom it has been named. In a unilateral lesion the motor fibres are interrupted after their decussation in the medulla, consequently there is paralysis of the leg, or, if the lesion is in the cervical cord, of the arm and leg on the same side—spinal hemi-

* See full discussion of the subject by J. William White, Therapeutic Gazette, 1891.

plegia. As the sensory fibres, entering the cord through the posterior roots, decussate at once and ascend in the opposite half of the cord, there is loss of sensation on the side opposite to the lesion, so that in hemisection of the cervical cord above the brachial enlargement there is motor paralysis of the arm and leg of the same side and anæsthesia of the arm and leg of the opposite side. The anæsthesia may be only to painful and to thermic sensation. In many cases the tactile sensation is unimpaired. The muscular sense is diminished on the same side as the lesion, and on this side also the skin is hyperæsthetic, so that a slight irritation is felt very acutely. Of this phenomenon, which may persist for years, no satisfactory explanation has been given. Just above the level of the hyperæsthesia there is a narrow zone of anæsthesia, which is at the exact physiological level of the lesion and corresponds to the fibres coming from the same side, which are involved at once on entering the cord. Above this again there is a narrow zone of hyperæsthesia. The reflexes are usually increased on the side of the lesion and the temperature is slightly raised.

The following table of Gowers illustrates the distribution of these various symptoms in a hemi-lesion of the cord:

Cord.

Zone of cutaneous hyperæsthesia. Zone of cutaneous anæsthesia.	Lesion.	
Motor palsy. Hyperæsthesia of skin. Muscular sense impaired. Reflex action first lessened and then increased. Temperature raised.		Muscular power normal. Loss of sensibility of skin. Muscular sense normal. Reflex action normal. Temperature same as that above lesion.

It is only in exceptional cases that all these features are met with in a case of Brown-Séquard's paralysis, and the condition may be transitory and rapidly replaced by paraplegia.

VI. Lesions of the Cauda Equina and Conus Medullaris.

The spinal cord extends only to the second lumbar vertebra. Injury, tumors, and caries at or below this level involve not the cord itself, but the bundle of nerves known as the cauda equina and the terminal portion of the cord, the conus medullaris. Much attention has been given recently to lesions of this part. The whole subject is admirably discussed in a recent work by Thorburn.* Fractures and dislocations are common in the lumbo-sacral region, tumors not infrequently involve the filaments of the cauda equina, and some of the nerves may be entangled in the cicatrix of a spina bifida.

* A Contribution to the Surgery of the Spinal Cord. By William Thorburn. London, 1890.

In a fracture or dislocation of the first lumbar vertebra the conus medullaris may be compressed with the last sacral nerves given off from it. In a case recently reported by Kirchhoff there was laceration of the conus with complete paralysis of the bladder and rectum, a case which is held to favor the view that the ano-vesical centre in man is situated in this region of the cord. There are several instances on record in which injury of the cauda equina has produced paralysis of the bladder and rectum alone, sometimes with a slight patch of anæsthesia in the neighborhood of the coccyx or the perinæum. More commonly branches of the sacral or lumbar nerve roots are involved producing an irregularly distributed motor and sensory paralysis in the legs. When the lumbar nerve-roots from the second to the fifth are compressed there is paralysis of the muscles of the legs, with the exception of the flexors of the ankles, the peronæi, the long flexors of the toes, and the intrinsic muscles of the feet, and loss of sensation in the front, inner, and outer part of the thighs, the inner side of the legs, and the inner side of the foot. The sacral roots may alone be involved. Thus in a case which I have reported the patient fell from a bridge and had paralysis of the legs and of the bladder and rectum. When seen sixteen years after the injury, there was slight weakness, with wasting of the left leg; there was complete loss of the function in the ano-vesical and genital centres, and anæsthesia in a strip at the back part of the thigh (in the distribution of the small sciatic), and of the perinæum, scrotum, and penis. The urethra was also insensitive. In a second case, in a young man with a healed spina bifida there was, with a small area of anæsthesia, involvement of the bladder and rectum, but retention of the sexual power.

Starr's table, given in the section on motor localization, will be found useful in determining the nerve fibres and segments involved in these cases of injury of the cauda equina.

VII. Tumors of the Spinal Cord and its Membranes.

New growths may develop in the cord or in its membranes, or may extend into them from the spine. The first two alone will be considered. Occasionally lipoma and parasites occur in the extradural space. Within the dura fibromata, sarcomata, and syphilitic and tuberculous growths are most common. In the cord itself, and attached to the pia mater, the tuberculous, syphilitic, and gliomatous growths are most frequent. Of 50 cases of tumor of the spinal cord and its envelopes analyzed by Mills and Lloyd, only 3 were parasitic. Of these 26 were some form of neoplasm, of which sarcomata were most common, 5 were gummatous, and 4 tuberculous. Herter has recently reported 3 cases of solitary tubercle in the cord, and has analyzed others from the literature. Of 24 cases in which the age was given, 15 occurred between the ages of fifteen and thirty-five, and 5 before the fifth year. The tumor is most common in

the dorsal and lumbar regions, and is usually met with in connection with tuberculous lesions elsewhere.

The anatomical effects of tumors are very varied. Slow compression is usually produced by growths external to the cord, and it is remarkable what a high grade of compression the cord will bear without serious interference with its functions. In cases of prolonged interruption ascending and descending degenerations occur. Tumors developing within the cord may lead to syringo-myelia. And, lastly, tumors not infrequently excite intense myelitis.

Symptoms.—These will naturally vary a good deal with the segment involved and with the degree of pressure and the extent of implication of the nerve-roots.

Within the cord the symptoms are those of a gradually progressing paraplegia, which may at first have the picture of a Brown-Séquard paralysis. Atrophy follows the involvement of the anterior cornua, and vaso-motor disturbances may be marked. The reflexes are lost at the level of the lesion, but if in the dorsal cord, the reflexes are retained in the legs. The symptoms are apt to be complicated with those of acute or subacute myelitis, which may completely alter the clinical picture. Tumors of the spinal membranes are characterized by the early onset and persistence of the root symptoms, which consist of radiating pains, girdle sensation, hyperæsthesia, or anæsthesia in various portions of the trunk. There may even be severe pain in the anæsthetic areas. Irritation of the motor roots may cause spasm of the muscles supplied, or wasting with paralysis. The paraplegia supervenes some time after the occurrence of the root symptoms. In the dorsal region the level of the growth is usually accurately defined by the level of the pain and the condition of the reflexes.

The diagnosis of tumor within the cord is sometimes easy, the characteristic features being the constancy and severity of the root symptoms at the level of the growth and the progressive paralysis. Caries may cause identical symptoms, but the radiating pains are rarely so severe. Cervical meningitis simulates tumor very closely, and in reality produces identical effects, but the very slow progress and the bilateral character from the outset may be sufficient to distinguish this.

In chronic transverse myelitis the symptoms may, according to Gowers, simulate tumor very closely and present radiating pains, a sense of constriction, and progressive paralysis.

The nature of the tumor can rarely be indicated with precision. With a marked syphilitic history gumma may naturally be suspected, and with coexisting tuberculous disease a solitary tubercle.

Treatment.—If the possibility of syphilitic infection is present the iodide of potassium should be given in large and increasing doses. For the severe pains counter-irritation is sometimes beneficial, particularly the thermo-cautery; morphia is, however, often necessary.

In a few instances tumors of the cord or of the membranes are amenable to surgical treatment. The removal by Victor Horsley of a growth from the membranes of the cord in a patient of Gowers' was one of the most brilliant operations of modern surgery.

VIII. Progressive (Spinal) Muscular Atrophy

(*Chronic Degeneration of the Motor Nuclei—Poliomyelitis Anterior Chronica*).

Definition.—A disease characterized by degeneration of groups of the motor nuclei in the cord and medulla, with wasting of the corresponding muscles. The pyramidal tracts are usually involved, and the paralysis may have a spastic character. In some cases the degeneration has been traced to the ganglion cells of the motor cortex.

Three affections, as a rule described apart, belong together in this category: (*a*) Progressive muscular atrophy of spinal origin; (*b*) amyotrophic lateral sclerosis; and (*c*) progressive bulbar paralysis. A slow atrophic change in the motor nuclei is the anatomical basis, and the disease, as Charcot states, is one of the whole motor path, involving, in many cases, the cortical, bulbar, and spinal centres. There may be simple muscular atrophy with little or no spasm, or progressive wasting with marked spasm and great increase in the reflexes. In others, there are added symptoms of involvement of the motor nuclei in the medulla—a glosso-labio-laryngeal paralysis; while in others, again, with atrophy (especially of the arms), a spastic condition of the legs, and bulbar phenomena, tremors develop and signs of cortical lesion. These various stages may be traced in the same case. I have for ten years had under observation a man whose illness began with weakness and atrophy of the hand muscles. Gradually the legs began to get stiff and the gait spastic; the arms subsequently wasted and the reflexes were increased. After these symptoms had persisted with increasing intensity for six or seven years, certain of the motor nuclei of the medulla became involved, the speech became thick, and the movements of the lips and tongue were impaired. Tremor has developed of late in the arms and hands. With these chronic changes the visceral functions have remained unimpaired and the mind unaffected. It has been a lesion of the motor segments, beginning in the lower and gradually extending upward. The disease began as progressive atrophy, and gradually assumed a typical picture of amyotrophic lateral sclerosis, and now the bulbar features are well marked and the tremor would indicate that the cortex is also involved.

For convenience, bulbar paralysis will be considered separately, and I shall here take up together *progressive muscular atrophy* and *amyotrophic lateral sclerosis*.

The disease is known as the Aran-Duchenne type of progressive muscular atrophy, after the French physicians who early described it, and as Cruveilhier's palsy. Lockhard Clarke demonstrated that it was a spinal