lesion. Charcot separated the two types—one with simple wasting, in which the anterior horns are alone involved; and the other in which, with degeneration of the cornua, the pyramidal tracts are affected, causing wasting *plus* a spastic condition. To this he gave the name of amyotrophic lateral sclerosis. There is but little evidence, however, to show that the anterior horns are ever affected without secondary changes in the pyramidal tracts, and Leyden and Gowers regard the two diseases as identical.

Etiology.—The cause of the disease is unknown. It is more frequent in males than in females. It attacks adults, developing after the thirtieth year, though occasionally younger persons are attacked. A large majority of all cases of progressive muscular atrophy under twenty-five years of age are of myopathic (i. e., muscular), not myelopathic (i. e., spinal) origin. Cold, wet, exposure, fright, and mental worries are mentioned as possible causes. Hereditary influences are present in certain cases. The father of the man whose case is referred to above died of progressive wasting of the muscles, but there have been no other cases in the family. It is highly probable that when many members of a family are affected the disease is not spinal, but an idiopathic muscular atrophy; and yet, in the Farr family, which I recorded a few years ago, in which thirteen members were affected in two generations, with the exception of two, the cases occurred or proved fatal above the age of forty, and the late onset speaks rather for a spinal affection. The amyotrophic form may develop late in life—after seventy—as a senile change.

Morbid Anatomy.—The following are the important anatomical changes: (*a*) The muscles waste and undergo fatty and sclerotic changes. The terminal branches of the motor nerves are degenerated. (*b*) The anterior roots are atrophied in those sections of the cord corresponding to the wasted muscles. (*c*) The gray matter shows the most marked alteration. The large ganglion cells of the anterior horns are atrophied, or, in places, have entirely disappeared, the neurogliar tissue is increased, and the fibres of the anterior nerve-root passing through the white matter are wasted. (*d*) In a majority of all the cases there is sclerosis in the antero-lateral tracts, but the direct cerebellar and the antero-lateral ascending tracts are spared. It was to this combination of atrophy of the anterior horns and sclerosis of the antero-lateral columns that Charcot gave the name amyotrophic lateral sclerosis. (*e*) The degeneration of the gray matter is rarely confined to the cord, but extends to the medulla; the motor nuclei are found extensively wasted in cases which have shown bulbar symptoms during life. (*f*) Cerebral changes also occur. The pyramidal tracts have been found degenerated through the pons and capsule, and in the motor cortex the large ganglion cells are wasted.

The essential anatomical change is a slow degeneration of the motor path, involving specially the nerve-cells of the anterior cornua and the anterior root-fibres, to which the loss of power and wasting in the muscles

are secondary. The upper segment is also involved, either simultaneously or at a later period.

Symptoms.—Irregular pains may precede the onset of the wasting. In one case the pains were about the hip and shoulder joints and the patient was treated for chronic rheumatism. The hands are first affected, and there is difficulty in performing delicate manipulations. The muscles of the ball of the thumb waste early, then the interossei and lumbricales, leaving marked depressions between the metacarpal bones. Ultimately the contraction of the flexor and extensor muscles and the extreme atrophy of the thumb muscles, the interossei, and lumbricales produces the claw-hand—*main en griffe* of Duchenne. The flexors of the forearm are usually involved before the extensors. In the shoulder-girdle the deltoid wastes first; it may waste even before the other muscles of the upper extremity. The trunk muscles are gradually attacked; the upper part of the trapezius long remains unaffected. Owing to the feebleness of the muscles which support it, the head tends to fall forward. The platysma myoides is unaffected and often hypertrophies. The arms and the trunk muscles may be much atrophied before the legs are attacked. The glutei, the vasti, and the tibialis anticus are first attacked when the disease begins in the legs. In the member of the Farr family who came under my notice (if this was really a myelopathic disorder) the wasting began in the gluteal and hamstring muscles of the left leg. The face muscles are attacked late. Ultimately the intercostal and abdominal muscles may be involved, the wasting proceeds to an extreme grade, and the patient may be actually "skin and bone," and, as "living skeletons," the cases are not uncommon in "museums" and "side-shows." Deformities and contractures result, and lordosis is almost always present. A curious twitching of the muscles (fibrillation) is a common symptom, and may occur in muscles which are not yet attacked. It is not, as was formerly supposed, a characteristic feature of the disease. The irritability of the muscle is increased. Sensation is unimpaired, but the patient may complain of numbness and coldness of the affected limbs. The galvanic and faradic irritability of the muscles progressively diminishes and may become extinct, the galvanic persisting for the longest time. In cases of rapid wasting and paralysis there may be the reaction of degeneration. The excitability of the nerve-trunks may persist after the muscles have ceased to respond. The loss of power is usually proportionate to the degree of wasting.

The foregoing description applies to the group of cases in which the atrophy and paralysis are flaccid—*atonic*, as Gowers calls it. In other cases, those which Charcot describes as amyotrophic lateral sclerosis, with the wasting there is more or less spasm, which may exist from the outset. This *tonic* atrophy may involve the legs chiefly or is present in the arms and legs. The reflexes are greatly increased. The most typical condition of spastic paraplegia may be produced. On starting to walk, the patient

seems glued to the ground and makes ineffectual attempts to lift the toes; then four or five short, quick steps are taken on the toes with the body thrown forward; and finally he starts off, sometimes with great rapidity. Some of the patients can walk up and down stairs better than on the level. The wasting is never so extreme as in the atonic form, and the loss of power may be out of proportion to it. The sphincters are unaffected. Sexual power may be lost early.

As the degeneration extends upward an important change takes place from the development of bulbar symptoms, which may, however, precede the spinal manifestations. The lips, tongue, face, pharynx, and larynx may be involved. The lips may be affected and articulation impaired for years before serious symptoms occur. In the final stage there may be tremor, the memory fails, and a condition of dementia may develop.

Gowers gives the following useful classification of the varieties of this affection: (1) Atonic atrophy, becoming extreme; (2) muscular weakness with spasm, but without wasting or with only slight wasting; and (3) atonic atrophy, rarely extreme in degree, with excess of the reflexes. These conditions may "coexist in every degree and combination—between universal atonic atrophy on the one hand and universal spastic paralysis without wasting on the other."

Diagnosis.—The affection must be distinguished from the primary muscular atrophies which usually occur in younger persons, often affect many members of a family, and have a different distribution, beginning either in the muscles of the shoulder girdle—sparing the hands or involving the face and upper-arm muscles—or the peroneal group. Muscular atrophy in the adult, beginning in the muscles of the thumbs, gradually involving the interossei and lumbricales, as a rule is of myelopathic origin.

Treatment.—The disease is incurable. I have never seen the slightest benefit from drugs or electricity. The downward progress is slow but certain, though in a few cases a temporary arrest may take place. With a history of syphilis, mercury and iodide of potassium may be tried, and Gowers recommends courses of arsenic and strychnine. Probably the most useful means is systematic massage, particularly in the spastic cases.

Bulbar Paralysis (*Glosso-labio-laryngeal Paralysis*).

An affection of the motor nuclei of the medulla oblongata, rarely primary, more commonly a part of a general degenerative affection of the nuclei of the motor path. The disease is sometimes called by the name of Duchenne. Acute and chronic forms may be recognized.

(1) *Acute bulbar paralysis* may be due to (*a*) hæmorrhagic or embolic softening in the pons and medulla; (*b*) acute inflammatory softening, analogous to polio-myelitis, occurring occasionally as a post-febrile affection.

The onset is usually sudden, hence the term apoplectiform. The cases

are almost invariably bilateral. As the nuclei presiding over the muscles of the tongue and lips are involved the speech is almost or entirely lost. The saliva drools, the lips are flabby and flaccid, swallowing may be difficult, and there may be loss of power in the laryngeal muscles. Usually these cases rapidly prove fatal, but occasionally a case with a sudden onset, like that figured by Gowers, may become chronic. In these acute cases there may be loss of power in one arm, or hemiplegia, sometimes alternate hemiplegia, with paralysis on one side of the face and loss of power on the other side of the body.

(2) *Chronic bulbar paralysis* is an affection of adult life, rarely beginning under the fortieth year, and in a great majority of the cases it is only part of a general degeneration of the motor nuclei. The disease usually begins with slight defect in the speech, and the patient has difficulty in pronouncing the dentals and linguals. The paralysis starts in the tongue, and the superior lingual muscle gradually becomes atrophied, and finally the mucous membrane is thrown into transverse folds. In the process of wasting the fibrillary tremors are seen. Owing to the loss of power in the tongue, the food is with difficulty pushed back into the pharynx. The saliva also may be increased, and is apt to accumulate in the mouth. When the lips become involved the patient can neither whistle nor pronounce the vowels *o* and *u*. The mouth looks large, the lips are prominent, and there is constant drooling. The food is masticated with difficulty. Swallowing becomes difficult, owing partly to the regurgitation into the nostrils, partly to the involvement of the pharyngeal muscles. The muscles of the vocal cords waste and the voice becomes feeble, but the laryngeal paralysis is rarely so extreme as that of the lips and tongue.

The course of the disease is slow but progressive. Death often results from an aspiration pneumonia, sometimes from choking, more rarely from involvement of the respiratory centres. The mind usually remains clear. The patient may become emotional. In a majority of the cases the disease is only part of a progressive atrophy, either simple or associated with a spastic condition. In the latter stage of amyotrophic lateral sclerosis the bulbar lesions may paralyze the lips long before the pharynx or larynx becomes affected.

The *diagnosis* of the disease is readily made, either in the acute or chronic form. The involvement of the lips and tongue is usually well marked, while that of the palate may be long deferred. A condition has been described, however, which may closely simulate bulbar paralysis. This is the so-called *pseudo-bulbar* form or bulbar palsy of cerebral origin. Bilateral disease of the motor cortex in the lower part of the ascending frontal convolution may cause paralysis of the lips and tongue and pharynx, which closely simulates a lesion of the medulla. Sometimes the symptoms appear on one side, but in many instances they develop suddenly on both sides. A bilateral lesion has usually been found, but in several instances the disease was unilateral.

Progressive bulbar paralysis is an incurable affection. Transient improvement may occur. Strychnine may be tried. Electricity is of doubtful benefit. Special care must be taken in feeding these patients, and when deglutition becomes much impaired the stomach-tube should be employed.

III. DISEASES OF THE BRAIN.

I. AFFECTIONS OF THE MENINGES.

Diseases of the Dura Mater (*Pachymeningitis*).

(*a*) **Pachymeningitis Externa.**—Hæmorrhage often occurs as a result of fracture. Inflammation of the external layer of the dura is rare. Caries of the bone, either extension from middle-ear disease or due to syphilis, is the principal cause. In the syphilitic cases there may be a great thickening of the inner table and a large collection of pus between the dura and the bone. In a remarkable case of this kind at the Montreal General Hospital the frontal lobes were so compressed by the thickened skull, and the purulent effusion between the bone and the dura, that the anterior vertical measurement of the brain was only 2·5 cm., while that of the posterior part was 8 cm.

Occasionally the pus is infiltrated between the two layers of the dura mater or may extend through and cause a dura-arachnitis.

The symptoms of external pachymeningitis are indefinite. In the syphilitic cases there may be a small sinus communicating with the exterior. Compression symptoms may occur with or without paralysis.

(*b*) **Pachymeningitis Interna.**—This occurs in three forms: (1) Pseudo-membranous, (2) purulent, and (3) hæmorrhagic. The first two are unimportant. Pseudo-membranous inflammation of the lining membrane of the dura is not usually recognized, but a most characteristic example of it came under my observation as a secondary process in pneumonia. Purulent pachymeningitis may follow an injury, but is more commonly the result of extension from inflammation of the pia. It is remarkable how rarely pus is found between the dura and arachnoid membranes.

Hæmorrhagic Pachymeningitis (*Hæmatoma of the Dura Mater*).

This remarkable condition, first described by Virchow, is very rare in general medical practice. During ten years no instance of it came under my observation at the Montreal General Hospital. On the other hand, in the post-mortem room of the Philadelphia Hospital, which received material from a large almshouse and asylum, the cases were not uncommon, and within three months I saw four characteristic examples, three of which came from the medical wards. On the other hand, the frequency

of the condition in asylum work may be gathered from the fact that Wigglesworth found 42 examples in a series of 400 unselected post-mortem examinations.

The disease is found chiefly in males and in persons over fifty years of age. It is most frequent in forms of chronic insanity and in chronic alcoholism. It has also been found in profound anæmia and other blood conditions, and is said to have followed certain of the acute fevers.

The morbid anatomy is interesting. Virchow's view that the delicate vascular membrane precedes the hæmorrhage is undoubtedly correct. Practically we see one of three conditions in these cases: (*a*) Subdural vascular membranes, often of extreme delicacy; (*b*) simple subdural hæmorrhage; (*c*) combination of the two, vascular membrane and blood-clot. Certainly the vascular membrane may exist without a trace of hæmorrhage—simply a fibrous sheet of varying thickness, permeated with large vessels, which may form beautiful arborescent tufts. On the other hand, there are instances in which the subdural hæmorrhage is found alone—in 15 out of Wigglesworth's 42 cases—but it is possible that in some of these at least the hæmorrhage may have destroyed all trace of the vascular membrane. In some cases a series of laminated clots are found, forming a layer from 3 to 5 mm. in thickness. Cysts may occur within this membrane. The source of the hæmorrhage is probably the dural vessels. Hugenin and others hold that the bleeding comes from the vessels of the pia mater, but certainly in the early stage of the condition there is no evidence of this; on the other hand, the highly vascular subdural membrane may be seen covered with the thinnest possible sheeting of clot, which has evidently come from the dura. The subdural hæmorrhage is usually associated with atrophy of the convolutions, and it is held that this is one reason why it is so common in the insane; but there must be some other factor than atrophy, or we should meet with it in phthisis and various cachectic conditions in which the cerebral wasting is as common and almost as marked as in cases of insanity.

The symptoms are indefinite, and the diagnosis cannot be made with certainty. Headache has been a prominent symptom in some cases, and when the condition exists on one side there may be hemiplegia. Extensive bilateral disease may exist without any symptoms whatever.

Diseases of the Pia Mater.

(*a*) **Acute Leptomeningitis.**—In this form the exudation is between the pia and the arachnoid membranes.

Etiology.—Acute inflammation of the pia mater occurs under the following circumstances: (1) As a result of an eruption of tubercles, most frequently in the basal meninges, forming the basilar or tuberculous meningitis which has been already considered (see tuberculosis). (2) In the epidemic cerebro-spinal fever. (3) Secondary to acute general diseases,