"It may be said, without reserve, that this work is the most clear, concise, and complete textbook upon diseases of the nervous system in any language. And when the large number of such works which have appeared in Germany, France, and England within the past ten years is considered, this implies high praise."—The American Journal of the Medical Sciences.

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VOLUME I
DISEASES OF THE SPINAL CORD AND NERVES
in spite of the fact that the unsteadiness in walking is very similar in the two diseases; the common affection of the arms in the one, and the conspicuous head symptoms (severe pain, optic neuritis, &c.) of the other, sufficiently distinguish them. It is very common for the tremor of the head to be at first ascribed to mere "nervousness," and also for the isolated form, in girls, to be regarded as hysterical. The presence of nystagmus should at once decide the question; it is absolute proof of definite disease.

**Prognosis.**—The prognosis in every case is very serious, since the disease, being a developmental affection, is essentially progressive; but life may be prolonged for many years, and in slight cases the malady may interfere comparatively little with the patient's occupation. Thus I have known a man, with very marked symptoms, follow his business as a tradesman for many years. The only guide to individual prognosis is the observed rate of progress, which has little relation to the age at which the symptoms commence.

**Treatment.**—As in other diseases that depend on a congenital tendency, treatment is almost powerless. The measures recommended for ordinary locomotor ataxy (apart from those suggested by the special symptoms of tabes) are those most suitable to the hereditary form. Arsenic, phosphorus, and nitrate of silver deserve a trial; now and then we seem to check the progress of the disease for a longer or shorter time, but its individual tendency determines its course, almost irrespective of treatment.

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**Simple Senile Paraplegia.**

This name seems the most unobjectionable for a condition, somewhat rare, which appears hitherto to have been unrecognised.* It is confined to late life, occurring in those over 40, and especially over 50. Its characteristic is simple weakness of the legs, with some slowness of movement, but without wasting, sensory disturbance, or reflex alteration. The knee-jerk is normal, and there is no foot-clonus. The malady develops very gradually and is slowly progressive, although it seems seldom to attain such a degree as to prevent standing. The condition of the legs resembles that in cases of paralysis agitans without tremor, in which the malady is manifested only by weakness, and stiff slowness of movement of the limbs, face, and trunk. Cases are met with in which the condition of the legs above described is associated with slight symptoms in the arms and face, such as characterise these cases of paralysis agitans. This senile paraplegia is probably a partial development of the morbid process of that disease; instead of being general, it is limited to the structures for the legs. If, as the condition seems to suggest, paralysis agitans depends on peculiar changes in the

* A note upon it by myself appeared in the 'Centralbl. f. Nervenkr.,' 1890.
that of the spinal cells, and "progressive bulbar paralysis" is then associated with "progressive muscular atrophy:" it often comes on after the disease has existed for some time, and ends life; occasionally it initiates the malady.

Two varieties of the disease have been distinguished by Charcot, who has been followed in this by most subsequent writers. In the one variety the disease is manifested only by the wasting of the muscles; in the other there are, in parts other than those much atrophied, the indications of degeneration of the pyramidal tracts of the cord. In the latter cases, Charcot regarded the degeneration of the pyramidal tracts as the primary lesion, and the affection of the cornea as secondary. He therefore termed the cases of the first class "protopathic," and those of the second class "deuteropathic," and gave to the latter the name "amyotrophic lateral sclerosis." In Germany, however, and especially by Leyden, doubts have been expressed as to the validity of the distinction between the two classes of cases, and as to the sequence of the lesions in the second class. These doubts are not without foundation. The subject will be discussed when the pathology of the disease is considered, and reasons will then be given for the course here adopted of considering the two varieties together as essentially one disease.

But cases are also met with (although not frequently) that present gradation to subacute polio-myelitis in one direction, and perhaps also to polynévritis in another. Attempts are being made to classify these, but types may be multiplied indefinitely from the intermediate forms, and many more pathological and clinical observations are necessary before a useful classification of the cases can be made. In the following description the common forms are chiefly considered.

Causes.—The disease is more frequent in males than in females, the proportion being about three to one. It is chiefly a disease of adult life, commencing usually between twenty-five and forty-five. I have known it to begin at fourteen and as late as seventy, and a case is recorded in which it began at twelve;* there was wasting and rigidity in the limbs, tongue, lips, and lower face. But most cases of muscular atrophy in early life are idiopathic, and not spinal. Heredity is to be traced only in less than half the cases, and generally as an indirect neuropathic disposition. Rarely there is direct inheritance of the disease. Among instances that I have met with are a lady whose mother died from a similar atrophy, and another case (from which Fig. 135 is taken) in which a brother had died from some chronic disease of the cord, attended with wasting; but three cases with bulbar symptoms have been recorded, two of whom were cousins, the children of two sisters: the third, who had had syphilis, was a cousin, but his affection was atypical.† Direct inheritance seems to be most common in cases that occur late in life. When many members of a family suffer from

muscular atrophy, the malady is nearly always idiopathic and not spinal. The affection occurs in all classes of society, and it is doubtful whether workers with the muscles furnish a larger proportion of the cases than can be accounted for by their greater exposure to certain exciting causes.

Of these more direct causes one of the most frequent is mental distress and anxiety, and this is especially met with in females and in late life. A severe fright has been thought to excite it. Another cause is exposure to wet cold, which is also a cause of many other chronic spinal diseases. Sometimes the exposure has been habitual; sometimes a single exposure has been effective, and some neuralgic pains, indicative of the deleterious influence on the nervous system, have followed the exposure and connected it with the later wasting. The pain has been either in the part afterwards wasted, or in some other, more common, seat of neuralgia. Thus one patient, after remaining in wet clothes for twelve hours, suffered for six weeks from severe neuralgia in the left side of the face, and then the muscles of the left shoulder began to waste. Although excessive use of individual muscles may cause them to waste, it is doubtful whether this influence produces general muscular atrophy. Injury to the cord, such as results from concussion of the spine, is a rare cause. It often produces disseminated myelitis, which may be manifested by muscular wasting combined with other symptoms.* In a few instances I have known progressive atrophy, of typical characters, to slowly follow a concussion, as if this had set up a perversion of the nutrition of the nerve-elements. Still more rarely a fall, injuring one limb, has been followed by muscular atrophy commencing in this limb and becoming general. Thus a woman fell downstairs, and pitched on her left hand and wrist; she had pain in the arm for a long time; two years after the fall this arm began to waste, and the atrophy ultimately became general. The relation might be passed as an accidental coincidence, were it not that in other central diseases—even, for instance, in paralysis agitans—the symptoms may commence in an injured limb. In rare cases, the disease develops, during adult life, in a subject of old infantile palsy (polio-myelitis), and may start from the most affected part (see p. 304).

The disease sometimes succeeds syphilis, and no other cause may be traceable. As with other degenerative diseases, an interval of years elapses between the primary disease and its nervous sequel. The cases that I have seen after syphilis have been typical in course, and evidently degenerative in nature. That syphilis has some share in the causation of these cases is probable from its relation to other diseases, such as tabes; but it is noteworthy that in one case the wasting commenced during, and in another directly after, an energetic

† 'Virchow's Archiv,' 1889, p. 116.
mercurial course. This relation is intelligible if the disease is due to a product of the syphilitic virus, and not to the organism themselves (see Tabes). Occasionally syphilis and a neurotic heredity can both be traced. In the cases in which atrophy has followed an acute specific disease, especially measles, and has begun in early life, it is probable that the malady is a chronic neuritis and not the central affection now under consideration. Such cases are known as of the “Peroneal type,” from the muscles in which wasting is first obtrusive, and are separately described at a later page. General muscular atrophy may result from lead poisoning, but this form is not, as a rule, progressive in character when its cause has ceased to act. It resembles the ordinary form of progressive muscular atrophy, however, in seat and features, and thus differs from the common atrophic palsy of the extensors that is produced by lead. In many cases of progressive muscular atrophy no cause for the disease can be traced. When obtrusive symptoms have followed some adequate cause, it is not uncommon for this to have been preceded by slight symptoms, which show that the malady already existed. A careful inquiry for such symptoms should never be neglected.

Symptoms.—The definite symptoms of the disease are often preceded by aching pain in the part afterwards wasted, rarely severe. Sometimes there is such pain in the spine or elsewhere, especially, as already stated, in the cases that are due to exposure to cold. Rarely some sensation other than pain precedes the local symptoms. Thus in one case a sense of coldness preceded the atrophy in each limb that was attacked. Weakness and wasting usually come on together, but either may first attract the attention of the patient. In the shoulder and back, the loss of power is usually first noticed, and in such covered parts the wasting may become considerable before it is observed. In the hand the wasting is often first noticed, but sometimes it is the impairment of some delicate action, such as writing, that draws attention to the part. The affected muscles lose their proper shape, and there is flattening, or even a depression, where there should be a prominence. If a patient is fat, however, the wasting may cause at first very slight alteration in the external aspect.

The disease commences in the arm in nine tenths of the cases, and as frequently in one arm as in the other. It begins with almost equal frequency in the hand and in the shoulder muscles. From the part first affected the disease spreads to other parts of the same limb. Before it has attained a considerable degree in one limb, it usually shows itself in the corresponding limb on the other side; often in the muscles corresponding to those in which it commenced, sometimes in those affected second in order of time. As the muscles waste, paralysis results of various character and degree, corresponding to the atrophy. In the hand, the thenar muscles and interossei are usually the first to suffer. The thenar eminence becomes flattened, and the base of the first metacarpal bone becomes prominent. Of the interossei,

the atrophy of the abductor indicis is especially conspicuous; the normal prominence gives place to a hollow beside the metacarpal bone when the thumb is abducted. Depressions form between the metacarpal bones on the back of the hand, and also between the flexor tendons in the palm in consequence of the wasting of the lumbricales. The forearm muscles may be next involved, the flexors usually before the extensors; and with the flexors the supinators may suffer, or they may escape until the biops is involved. Occasionally the disease begins in the forearm, and then especially in the extensor muscles of the fingers, sometimes in those of the thumb, especially of its phalanges. The several parts of the long finger muscles may suffer unequally; the ulnar extensors usually suffer most. Of the shoulder muscles, the deltoid is generally the first to manifest the disease, and in some cases which begin in the hand the deltoid suffers before the forearm muscles. The rounded contour of the shoulder becomes changed (Figs. 7, 134, 135), and the head of the humerus can be recognised beneath the acromion. It is not rare for part of the del-
toid to suffer and part escape: we have seen (p. 30) that the several portions have different functions and associations, and this probably involves a relation to separate groups of nerve-cells. The wasting of the deltoid is soon followed by that of the other muscles of the upper arm and of the scapula. The triceps usually suffers less and later than the biceps, but sometimes the reverse is the case. The supra- and infra-spinati are often affected with the deltoid. In exceptional instances the deltoid or hand muscles escape, and there are cases (perhaps a special group) in which the upper arm and shoulder muscles waste even to an extreme degree, and the forearm and hand muscles escape entirely.

In most cases, the wasting early involves the muscles of the back, and it sometimes begins in them. The middle and lower parts of the trapezius usually suffer first; the rhomboids and erectors of the spine at a later date. The affection of the trapezius is readily recognised if the patient tries to put the shoulder back (Fig. 136). The highest part of the trapezius presents a remarkable indisposition to atrophy; it often remains intact to the last, and then may contrast with the wasting below it, standing out on each side, like a cord passing from the occiput to the shoulder. Hence Duchenne termed this part the alium moriens. I have, however, seen several otherwise typical cases (beginning either in the deltoids or in the hands) in which the highest parts of the trapezius suffered before the middle parts. The levator anguli scapulae also generally escapes, even when all the muscles about it are wasted. The serratus, latissimus, and pectoralis major are usually affected later. They may escape wholly or in part; isolated bundles of the pectoralis may waste, the intervening parts escaping, or either the upper or lower part of the muscle may atrophy alone. According to the affection of the muscles that are attached to the scapula, the position of the bone changes, and it becomes rotated under the influence of the muscles that are unaf-fected and unopposed (Fig. 7, p. 28). The muscles that extend the head on the spine often suffer in considerable degree, and from this there results a difficulty in the carriage of the head (Fig. 136). It is habitually inclined backwards, so as to balance it on the spine with but little muscular exertion; if moved forwards, it falls so that the chin touches the chest. It can be brought back into its former position only with difficulty; the patient has to incline the trunk backwards, so as to bring the head nearly into the vertical position, and then, with a sudden contraction of the sterno-mastoids, and a jerk, the head goes back into its former posture. The increased innervation of the weak extensors of the head often causes a synergic over-action of the frontalis muscles, which are normally associated with the extendors (so that the eyebrows are raised when the head is put back to look upwards). The skin at the back of the neck lies in transverse folds when the neck is extended. The patient may be unable, when lying in bed, to move the head from side to side. The sterno-mastoids also are often wasted; either the sternal or clavicular part may be most affected. In striking contrast to the general wasting of the neck is the condition of the platysma myoides, which always escapes, and may become hypertrophied in a vain attempt at compensation.

The muscles of respiration suffer in the majority of cases, and their impairment constitutes a grave source of danger to life. The intercostals rarely escape altogether; the diaphragm is involved in many cases, and respiration is then carried on by the intercostals and superior thoracic muscles. On the other hand, the intercostals may suffer much and the diaphragm remain free; respiration is then purely abdominal, and the walls of the thorax may be almost motionless in breathing, or there may be merely a slight movement of elevation of the upper ribs. The chest becomes flattened in front, and narrow from before back, from the influence of atmospheric pressure on the ribs, unopposed by the intercostals. Either the upper or lower intercostals may suffer most. When the diaphragm is paralysed, the upper chest muscles often suffer more than the lower, and a compensatory increased movement of the lower ribs carries forward the abdominal wall, and may suggest that the diaphragm is acting when it is not. A careful examination will always prevent the error. The muscles of the abdominal wall occasionally waste, but far less frequently than those of the thorax.

Wasting in the legs is much less common than in the arms, and if it occurs is usually slighter in degree; but occasionally the disease first manifests itself in the legs, and is more intense in them than elsewhere. The glutei, extensors of the knee, and the muscles in the front and on the outer side of the lower leg, are those that are most affected. We shall presently see, however, that the legs are often paralysed when they are not wasted, and sometimes they are the seat of wasting which differs in certain features from that which is the special characteristic of the disease.

The face almost always escapes the general atrophy, and its normal appearance may present a striking contrast to the rest of the body. In many cases, however, the lips are paralysed as part of the bulbar palsy that so often accompanies the spinal disease. A remarkable case in which the face shared the atrophy of the limbs, and the tongue
escaped, has been recorded by Langer;* on the other hand, the tongue has been the first part to suffer, the arms being affected a month later.†

As the wasting progresses, the appearance of the parts in which it is most advanced becomes extremely changed. All trace of muscle may disappear from parts of limbs or even from an entire limb (Fig. 134); and there is usually a wasting of the adipose tissue as well as of the muscle, so that the bone seems to be covered only by fascia and skin. The transverse processes of the vertebrae may be felt in the hollow beside the spine, and the bony prominences about the shoulder may be almost as conspicuous as in a skeleton, so that beneath the acromion there may be a groove, into which the finger can be placed, in consequence of the descent of the head of the humerus from the glenoid cavity. The unequal affection of antagonistic muscles leads to various contractions and deformities. In the hand, especially, distortion is apt to occur; from preponderant paralysis of the interossei and contraction of the long flexor and extensor muscles, the "claw-like hand" develops in extreme degree (Figs. 19 and 20, p. 36). If all the muscles of a part are equally and simultaneously affected, no deformity results unless an unsupported part yields to the influence of gravitation (Fig. 134). Lordosis is very common in cases in which the trunk and hip muscles are involved. It is often the indirect result of the weakness of the extensors of the hip-joint, in consequence of which the pelvis is unduly inclined forwards, and carries with it the lower lumbar vertebrae; the upper part of the trunk has then to be inclined backwards, to bring the centre of gravity over the base of support; it disappears when the patient sits, and the pelvis, resting on the ischial tuberosities, is no longer inclined. This was the case, for example, in a woman, with extreme lordosis from progressive muscular atrophy, in whom, when standing, a plumb-line from the seventh cervical spine fell three inches from the sacrum and an inch and a half from the buttok. Sitting, her spine was perfectly straight.

The electrical irritability of the wasted muscles presents changes which vary in character in different cases. When the wasting is slow, there is usually a diminution in both faradic and voltaic irritability, similar in character to each current. The irritability fails with the muscular nutrition, and when the wasting is great only a slight contraction can be obtained, even with a strong current. When the wasting is extreme, irritability at last becomes extinct, but the voltaic irritability of the muscular fibres persists long after the faradic irritability has disappeared. It may be normal in degree, or lowered, but is seldom increased in such cases. The quality of the voltaic irritability may also be normal, but sometimes AOC (anodal closure contraction) occurs as readily, or more readily than

KCC. Tetanic contraction during the passage of the current is produced with undue readiness compared with the closure contractions, and opening contractions are often caused by currents but little stronger than those that cause closure contractions. Thus there may be the qualitative change of the reaction of degeneration without the voltagic increase (see p. 56). When there is the rapid and considerable paralysis described on p. 483, either at the onset or during the course of the disease, the palsy being followed by rapid wasting, there may be quick extinction of faradic with actual exaltation of voltaic irritability, and the reaction of degeneration may be present in all its characteristics. Between these forms every intermediate condition may be met with, including the mixed reaction described on p. 26.

The mechanical irritability of the muscles is considerably increased; a tap causes a local contraction of the fibres struck. Moreover, spontaneous flickering contractions of parts of the muscles are very common, now of one bundle, now of another, conspicuous to the eye, although scarcely felt by the patient. This "fibrillation," as it is termed, is so frequent as to be characteristic, but it is not invariable, nor is it confined to this disease. It may sometimes be observed in muscles that are not yet invaded by the wasting, but where it is observed atrophy usually follows.

In the parts affected by the characteristic wasting, all reflex action is abolished, clearly in consequence of the damage to the motor part of the reflex arc; the afferent portion of the arc is unaffected, for, as we shall see, there is no loss of sensation. The myotatic irritability ("tendon-reflex action") is also lost, and lost early. The knee-jerk, for instance, disappears as soon as there is even a slight diminution in the bulk of the thigh muscles, in cases in which the legs are the seat of primary wasting. The muscles are flaccid and toneless—a condition that may be conveniently termed "atonic atrophy." To this common rule, however, exceptions are sometimes met with; there may be from the first rigidity of the affected muscles. When this is the case, the wasting does not go on to the entire destruction of the muscle; it is often trifling, but may be considerable. With this rigidity there is a preservation of myotatic irritability. We may term this, "tonic atrophy." It is generally associated with aspastic paralysis, without wasting, in some other part. For instance, in one case the forearms and hands were the seat of this tonic atrophy, and in the legs there was simple aspastic paraplegia. We shall presently consider the pathology of this condition.*

Sensory symptoms are usually slight and always subordinate. They

* It should be noted, however, that, in the muscles that are the seat of toneless wasting, when the atrophy has attained an extreme degree, slight rigidity may develop, accompanied by considerable tenderness of the muscle. It is probable that this rigidity is idiopathic, and due to the structural changes in the muscles presently to be described. This condition, which should be distinguished from the initial rigidity, is rare: the early relaxation is usually maintained to the last.
are confined to pain, usually dull and rheumatoid in character, felt chiefly in the limbs in which the malady is most active. Such pains are common at the onset, as we have already seen, and they may recur from time to time during the course of the disease. Sometimes vague feelings of “numbness” or “deadness” are complained of, but cutaneous sensibility is never impaired. (When anesthesia accompanies muscular wasting, both symptoms are due either to chronic meningitis, damaging the nerve-roots, or to disseminated focal myelitis or multiple neuritis.) Nor do the muscles lose such sensitivity as they normally possess. Perception of posture persists, and the muscles are not only sensitive to pressure, but are often more sensitive than in health both to pressure and to extension.

When the arms are the seat of such atrophy as has been described, the legs, if not also wasted, may be normal, or they may be slightly affected, the thighs thin, and the knee-jerks lost. Often they are the seat of gradual loss of power without wasting, and with an increase of myotatic irritability. The knee-jerk is excessive; a clonus can readily be obtained, and the reflex over-action of the muscles may increase to actual rigidity and spasm, so that there is the condition of spastic paraplegia described in a preceding chapter. It is rare, however, for the spasm to reach the higher degree of intensity, and for the muscles to present the massive firmness that characterises the simple form of spastic paraplegia. On the other hand, there is often some diminution in the size of the muscles, and thus we have a gradation to the condition of tonic atrophy just described. In this condition, moreover, there is usually only a moderate diminution in electrical excitability, which does not go on to extinction. The one condition does not seem to pass into the other; the state of rigidity and myotatic excess does not give place to muscular relaxation; atomic atrophy does not succeed the tonic wasting. In very rare cases, of which I have seen a few instances, there is a similar condition in the hands when the shoulders are the seat of simple atomic atrophy. The forearms are then rigid, moderately wasted, with myotatic excess, while the muscles of the shoulders are extremely wasted and absolutely flaccid. In still more rare cases the whole arms are thus affected, are thin and rigid, and in no part is there atomic atrophy. In one remarkable case of this character, the muscles of the trunk participated in the spasm; after coughing or yawning, respiration would be checked for a few seconds by general spasm, and whenever the patient was raised from the bed, the arms, legs, back, and neck became stiff in tetanoid rigidity, the head being bent backwards.

The combinations of the ordinary atomic wasting and loss of power, without wasting and with increased myotatic irritability, are exceedingly numerous and variable. The most frequent, as already stated, is flaccid atrophy in the arms and spastic paralysis in the legs; but I have twice seen atomic atrophy below the knees, with weak but well-nourished thigh muscles and much increased knee-jerk. The

variable combination of these conditions gives rise to differences that are more apparent than real, and may easily be allowed undue weight in classification.

The extreme emaciation of the most affected parts shows that the adipose tissue wastes as well as the muscles. The atrophied limbs are usually cold, and may be livid or pale, but there is no tendency to acute vaso-motor disturbance in the ordinary form of the disease. I have known the skin of the face to become thin and smooth, so that, in one instance, the dark iris could be seen through the closed eyelids, but such a change is quite exceptional. In another patient there was a very remarkable form of local atrophy. Certain areas of muscles underwent wasting, the rest being normal; the affection commenced by a livid discoloration of the skin, and the wasting seemed to involve the skin, subcutaneous tissue, and muscle, causing local depressions. Considerable tracts of the trapezius were thus affected, and smaller spots in the arms and legs. The patient was a single woman of thirty-five, and the disease, which did not show a strongly progressive tendency, is probably a special affection, perhaps of local nature.

The functions of the sympathetic are not, as a rule, deranged. Dilatation or contraction of one pupil has been frequently observed, chiefly in association with atrophy of the muscles that are supplied from the lower part of the cervical region; and it no doubt depends on the disease of the spinal cord, and not of the sympathetic itself. The reflex action of the iris is usually normal, and optic nerve atrophy never occurs.* Nystagmus is present in rare cases.

The visceral functions are usually little disturbed. Sexual power is often lost. The sphincters rarely suffer, even when the wasting is general and extreme; occasionally they are involved, and they may even suffer early. In other cases, in which the legs become weak after the wasting sets in in the arms, the affection of the sphincters may coincide with that of the legs. In the composition of the urine, slight changes have been found, but not constantly. Urea has been increased in some cases, lessened in others.† The quantity of lime has been

* I have once met with a remarkable reflex fixation of the eyeballs in a case of advanced progressive muscular atrophy. If the patient, looking to one side, was suddenly told to look at an object on the other side, his head was instantly turned towards the second object, while the eyes remained fixed on the first, by a movement corresponding to that of the head but in the opposite direction, and then, after a few seconds, they were slowly moved towards the second object. The phenomenon continued to the end of the patient’s life. As I pointed out in an account of this curious condition (‘Brain,’ vol. 1), it is interesting evidence of a normal reflex mechanism in the fixation of the eyes; this was, as it were, isolated by disease, which lessened voluntary control over it.

† In a patient who weighed 7st. 13lbs., and whose urine varied between 700 and 900 cubic centimetres, I found the daily excretion of urea to vary between 10? and 15 grammes, the average being 13 grammes. This is just half the normal average for a man of that weight, which would be (according to Parkes), 24 grammes.
found to be increased (Fromman), that of kreatin diminished, even to one tenth of the normal (Rosenthal, Langer). Glycosuria has been observed in association with bulbar symptoms. The lungs may suffer from the impairment of breathing power, when the intercostals and diaphragm are weakened.

Varieties.—The chief varieties of the disease depend on the relative distribution of the three conditions: (1) atonic atrophy, becoming extreme; (2) muscular weakness with spasm, but without wasting, or with only slight wasting; and (3) tonic atrophy, rarely extreme in degree, with myotatic excess. The commonest condition is to have atonic atrophy in the arms and upper part of the trunk, with simple weakness and spasm in the legs. Atonic atrophy in both arms and legs is less common, and the least common is tonic atrophy alone, in the arms, or universal. The last is, indeed, extremely rare. It is important to note, however, that these conditions may coexist in every degree and combination; between universal atonic atrophy on one hand, and universal spastic atrophy without wasting on the other, there is every gradation. The latter does not come into the category now under consideration, but similar cases are met with in which there is atrophy of a few muscles (as, for instance, of the hands only), which complete the series. The varieties due to the different combinations of atrophy and paralysis have been already alluded to.

A peculiar form of muscular atrophy has been mentioned, first conspicuous in the peroneal regions and spreading thence to other muscles of the leg and to the hands. It is characterised by occurrence in early life, and often in several members of the same family, and has been termed the "peroneal type" by Dr. H. Tooth. It is probably not dependent on the spinal cord, and is considered more fully at a subsequent page.

Complications.—Progressive muscular atrophy is occasionally accompanied by the symptoms of some other degenerative disease of the spinal cord. The paralysis with spasm, already described, can scarcely be regarded as a complication; it is rather part of the disease, and its relation to the other symptoms will be considered when we discuss the pathology of the affection. In rare cases, muscular atrophy in the arms is accompanied by the symptoms of locomotor ataxy in the legs. By far the most frequent complication is bulbar paralysis, weakness of the lips, tongue, pharynx, and often of the laryngeal muscles. It is the expression of a degenerative process in the medulla, similar to that which, in the spinal cord, causes the affection of the limbs. It may come on at any stage of the disease, may precede the spinal symptoms, or only come on when these have attained a considerable degree. In characters, the bulbar palsy may resemble perfectly that which occurs in isolated form, and will be described among the diseases of the brain. Frequently, however, there is but little paralysis of the tongue, even when swallowing and articulation are much impaired. There may also be slight interference with articulation when there is no marked bulbar palsy. A minor complication is the neuraltic pain, which, as already stated, is sometimes troublesome in the early stage of cases that are due to cold. In several patients I have known headache to be frequent and severe throughout the course of the disease. General paralysis of the insane has been met with as an exceptional complication.

Course.—The malady, in most cases, is steadily progressive, as its name implies, but in the rate of its progress it varies in different cases, and in the same case at different periods. It may, moreover, become stationary (sometimes as the result of treatment) at any period in its course, and when once actually arrested, it does not usually again become active. Unfortunately, the tendency to cessation is greatest in the later stages of the disease, when there is little except life to be preserved. Sometimes progress ceases at an earlier stage, and chiefly, I think, in those cases in which the atrophy is strictly symmetrical, and develops simultaneously, or almost simultaneously, on the two sides. Thus I have met with several cases in which there was symmetrical wasting in the two hands, or in certain muscles of the two forearms or of the two upper arms, and in which the atrophy, after attaining a considerable degree in its limited seat, had become stationary and continued so. Occasionally some symptom (as weakness of the sphincters) may pass away, although the atrophy progresses.

When the progress at the commencement is rapid, it usually continues rapid, until the disease has attained a wide extent, although the acute local onset mentioned below may be followed by slow extension. When it begins slowly, it is usually slow throughout.

Although the disease sometimes commences in the second arm very soon after its onset, it more commonly makes some progress in its primary seat before beginning on the second side, and the interval that elapses varies with the rate of extension. It often happens that a year intervenes between the affection of the two arms, and I have known, in a very chronic case, the atrophy to slowly progress in one arm for seven years before it showed itself in the other. It is not common for the arms to be reduced to practical helplessness in less than two or three years, but the hands may become useless in as short a time as six months, while in one case the wasting, commencing at the shoulder, had invaded the whole arm in the course of a month. The shortest time in which I have known a patient to reach the last stage of the disease is nine months.

With any rate of general progress, the otherwise uniform course of the disease may be broken by the occurrence of almost sudden palsy in a certain group of muscles. Considerable loss of power, it may be absolute paralysis, comes on in a few days, or even in a few hours. It may occur at the onset of the affection. The extensors of the wrist

and fingers are the muscles most commonly thus affected. The weakness is usually followed by a well-marked degenerative reaction in the muscles. I have seen several cases which began by such subacute or even acute paralysis of the extensor muscles, first in one arm and soon afterwards in the other; the initial condition closely resembled the paralysis from lead-poisoning, even to the escape of the supinator longus and ext. ossis met. pol. This cause was, however, excluded, and soon the muscles of the shoulder and back presented commencing slow progressive atrophy, followed by wasting of the interossei until a typical state was reached. In another case, the quick loss of power was confined to the extensors in one arm, which were already weak and slightly wasted, most of the other muscles of the upper limbs having been long atrophied. When there is weakness of the legs, without wasting, the onset of this may coincide with the atrophy of the arms, or may succeed it at any interval. In one case five years elapsed, after the arms began to waste, before the legs became weak. It is very rare for the paraplegic weakness to occur first.

The chief danger to life is from pulmonary maladies, rendered grave from the weakness of the muscles of respiration. The common complication of bulbar paralysis is another frequent cause of death, either by the interference with swallowing and nutrition, or by the laryngeal paralysis. Less commonly, death results from bedsores and septicemia, or from intercurrent maladies.

**Pathological Anatomy.**—The wasting of the muscles is as evident after death as during life. They are reduced in size and pale in colour. Sometimes there is little in the tint of what remains to suggest muscular tissue. Parts of a muscle may be hardly distinguishable from adjacent fat. On the other hand, the bulk of the muscle may be dark, and pale streaks in it may mark the position of local degeneration. Under the microscope, the fibres present various changes, and of these four are well defined. (1) There may be simple narrowing of the fibres, without any considerable change in their striation (Fig. 137), although the strie often seem to be further apart than normal, and sometimes the fibrillary segmentation is unusually distinct. (2) Simple fatty degeneration, in which the transverse striation gives place to a granular appearance (Fig. 138), the granules becoming larger and fewer (Fig. 137) until ultimately distinct globules are scattered through the sheath. Where the muscle resembles fatty tissue to the naked eye, the microscope may show only sarcolemma sheaths containing groups of globules. (3) Muscular fibres are seen in which the sheath contains only a clear material enclosing a few fatty globules, and a few transverse strie, faint, as if fading away. It is probable that this is not the result of fatty degeneration, but of a different process, which has been termed "vitreous degeneration,"—a sort of dissolution of the strie, indicated by the appearance of such fibres as are shown in Figs. 137, 139, 140. (4) A longitudinal striation develops in the fibre, and at first coexists with the transverse striation; but ultimately the latter becomes indistinct, and the fibre looks like a fasciculus of longitudinal connective-tissue fibres (Fig. 140). Sometimes, with the longitudinal striation, the fibre presents a transverse striation very much finer than normal, the strie being narrower and nearer together, as if from a division of the "sarcous elements." This change may sometimes be seen alone, and
of reddish-brown pigment. There is often also an increase of the nuclei (Fig. 138), and sometimes of the fibres of the interstitial tissue. Two or three rows of nuclei may lie between the fibres. The capillaries may be dilated and distended. It is very common to see muscular fibres that are much altered side by side with others that present a nearly normal appearance. Ultimately the sheaths become empty, and shrink, and may be scarcely distinguishable from the interstitial fibrous tissue.

The peripheral nerves contain many degenerated nerve-fibres, and the terminal branches for the muscles a still larger number. If the nerves are traced up to the cord it is found that the degenerated fibres come only from the anterior roots. These are conspicuously changed to the naked eye, small and grey. They may resemble fine threads of connective tissue, and under the microscope no nerve-fibres may be found in them, or only a few may remain of normal aspect, the rest being in various stages of degeneration, or represented only by their empty sheaths. The degree of affection of the anterior roots corresponds to the wasting in the parts supplied by them. The posterior roots are normal.

The spinal cord is often softer than natural at the affected part, and the white substance of the lateral columns may be grey and translucent in aspect, especially in the cervical enlargement. Under the microscope morbid changes are seen in the anterior cornua, and also, in most cases, perhaps in all, in the antero-lateral white columns. The change in the anterior cornua corresponds in its intensity to the origin of the nerves to the most affected parts, and since the latter are usually the arms, the disease is generally most intense in the cervical enlargement. In stained sections the grey matter of the horn is less deeply tinted than normal, but in some parts it may stain more deeply, especially in the circumference of the cornu and the processes of grey matter which project into the white column. Occasionally, larger areas have a dense aspect and stain deeply, but the central part of the cornu is generally pale and wasted in aspect. Most of the large nerve-cells have disappeared; many entirely, while others are represented only by small angular bodies. Not a single large cell may be seen in a section which, in a normal cord, would contain a considerable number. Frequently, however, a few large cells can still be seen, but most of these have lost their processes and are more globular than normal. The interstitial tissue is also changed. The nerve fibrils waste with the cells, and there is an increase of the small, angular, and stellate cells and other connective-tissue elements. The larger vessels are dilated and surrounded by unduly wide spaces, but there is no considerable diminution of the capillaries. The cornu as a whole is not usually changed either in size or shape. Similar alterations may be traced through the dorsal region. In the lumbar enlargement, the grey matter may be normal, even when it is much altered in the cervical region; but if the legs are wasted, there are changes similar to those already described. When the atrophy begins in the legs, the disease may be more intense in the lumbar than in the cervical enlargement; but more often any degeneration that exists is slighter, and a larger number of normal or slightly

FIG. 141.

FIG. 142.

FIG. 141.—A normal anterior cornu for comparison with the next figure.
FIG. 142.—Progressive muscular atrophy; anterior cornu, cervical region.
(From drawings by Dr. H. R. Spencer.)

changed cells can be seen. Sometimes certain groups of cells are little affected while others are much atrophied. The degeneration may involve to some extent the intermediate grey matter between the cornus, but here it ceases; the posterior horn is always normal.
There is distinct degeneration of the anterior root-fibres passing from the cornu through the anterior column. A few fibres may remain, but whole fasciculi appear to be replaced by fibrous tissue. There is also degeneration of the fibres of the anterior commissure, in consequence of which it stains much more deeply than normal, in the regions in which the grey matter is considerably diseased.

In the white columns there is usually considerable, and often almost complete, degeneration of the pyramidal tracts, anterior and lateral.

The area of sclerosis, resulting from the degeneration of the two tracts, varies in extent according to the size of the anterior tract, and to the distance which it extends down the cord. In Fig. 144 this is illustrated in an unusual manner, in consequence of an inequality in the decussation of the pyramids at the medulla, fewer fibres of the left pyramidal tract having crossed than of the right.

Fig. 144.—Progressive muscular atrophy. A, medulla oblongata; degeneration of the anterior pyramids A P; B, cervical, C, first dorsal, D, first lumbar, E, mid-lumbar sections. The nerve-cells have disappeared from the anterior cornu, excep. pt in E, in which a few remain. There is complete degeneration of the pyramidal tracts, anterior and lateral. The degeneration at the medulla was unequal in this case; of the left pyramidal fewer fibres crossed than of the right, and hence the left anterior pyramidal tract is larger, while the right lateral tract is smaller than normal, and, moreover, has ceased at the middle of the lumbar enlargement, to which the left anterior tract extends.

* In this case the arms were greatly wasted and facies; the legs were paralysed, rigid, with moderate wasting. The muscles of the back were also atrophied. In the arms the weakness and wasting came on at the same time, in the right arm some time before the left. The shoulder-muscles suffered first, but the atrophy quickly spread, and was ultimately extreme in the muscles of the shoulders, forearms, and hands. The disease ran a rapid course; death occurred two years after the gradual onset.

+ I have not yet met with a single case of progressive muscular atrophy in which the pyramidal tracts were unaffected. Such cases are extremely rare, but one has been published by Strümpell ("Zenker's Zeitschrift," 1887, and "Neur. Centrallbl.," 1888, p. 139) which was characterized by the general wasting of the arm, a very slow course, and the absence of leg symptoms. The spinal nerve-cells and motor nerves were degenerated. Strümpell suggests that the affection may have begun in the peripheral motor nerves, and have ascended to the cord. If so, the case belongs to a separate class.

+ The patient was a man forty-eight years of age. The disease followed a severe exposure to cold. The wasting of the arms was extreme, and they were absolutely powerless. The legs were much less wasted than the arms, but very weak, the loss of power being out of proportion to the wasting.
the left pyramid having crossed than of the right, so that the left half of the cord contains more than its proper share of the fibres, and the anterior tract extends into the lumbar region even further than does the lateral tract. Where the direct cerebellar tract exists, this, always unaffected, limits the sclerosis of the lateral tract on the outer side. On the inner side, the "lateral limiting layer" (see p. 182) is usually much less degenerated than the pyramidal tract, and intervenes between the latter and the grey matter; but this layer suffer in some degree, perhaps by the affection of outlying pyramidal fibres contained in it. The sclerosis does not usually cease at the anterior extremity of the pyramidal tract, but extends forward in the "mixed zone" of the lateral column, lessening in intensity, and ceasing usually opposite the outer part of the anterior cornu. It is here most intense close to the grey substance, and ceases before it reaches the surface of the cord, the antero-lateral ascending tract being unaffected. This sclerosis may be due to the degeneration of the short vertical fibres that pass between adjacent regions of the anterior cornu, and which share the degeneration of the nerve-cells. Occasionally it extends, in slighter degree, into the anterior columns. The posterior columns are always free from definite degeneration, but there may be some general increase of connective tissue throughout the whole cord. The degeneration of the pyramidal tracts is usually considerable in degree; often most of their fibres seem to have perished. If it is incomplete in the cervical region, it may be slight in the lower part of the cord, the degenerated fibres being chiefly those that end in the cervical enlargement and dorsal region. When the sclerosis is intense, it is not confined to the cord; it can be traced up through the decussation (Fig. 143, n) and through the medulla oblongata, in which, as Figs. 143 and 144 show, the whole of both pyramids may be degenerated, so that they stain deeply, and no nerve-fibres can be distinguished in them. The degeneration of the pyramids has been found in several cases to extend through the pons and crus to the internal capsule, and even through the white substance to the cortex. Above the capsule, however, the pyramidal fibres are so mingled with others that their degeneration is no longer recognizable by the deeper staining of a definite tract; but it has been detected by the presence of abundant products of degeneration in the path of the fibres. In the motor cortex, the large ganglion-cells have been found fewer than normal, and distinctly degenerated, many having lost their processes, and, in the interstitial tissue, the spider cells and other connective-tissue elements are increased in number. But the degeneration of the pyramidal tracts has also been found to cease at the crus and at the decussation.†

* Kosswiökoff, 'Archives de Neurologie,' 1888, No. 18; and 'Centralbl. f. Nervenkrk,' 1885, p. 409; Marié, 'Neur. Centraalbl.,' 1884, p. 61.
‡ Moli, 'Arch. f. Psych.,' 3, 718.

In cases in which the symptoms of bulbar paralysis were present during life, the motor nuclei of the medulla have presented changes corresponding to those of the grey matter of the spinal cord, with a similar degeneration of the nerve-cells. In other instances, the degeneration of these nuclei has been slight, but in such cases the degeneration of the pyramids has been intense, and doubtless involved the fibres connecting these nuclei with the cortex of the brain.

The sympathetic nerves and ganglia, when examined, have been found normal, or have only presented such changes as are common apart from symptoms of disease of the nervous system.

Pathology.—The theories, according to which the disease was regarded as primarily one of the muscles or of the sympathetic nerves, have now only an historical interest. The constancy of the changes in the ganglion-cells of the spinal cord, the degeneration of the motor root-fibres, and the analogous effects of acute lesions of the anterior cornu, leave no doubt of the relation of the muscular wasting to the disease of the grey matter, and essentially to that of the ganglion-cells, and the fibres proceeding from them. The slowness of the change causes the impairment of the nutrition of the muscular fibres to proceed pari passu with that of the nerve-elements, and the interference with motor conduction to be proportional to both. We are thus able to understand the condition of electrical excitability, and its slow failure, as nerve and muscle degenerate together. It is only when the slow degeneration is varied by a more acute process of destruction of cells and more rapid degeneration of fibres that the muscular tissue is for a long time less damaged than the nerve-fibres, and presents paralysis in excess of the wasting, with a voluntary irritability in excess of the faradic irritability of the nerve-endings (p. 24). Thus the essential lesion of the disease is a slow decay of the lower segment of the motor path, the segment which consists of the ganglion-cells and their prolongations in the axis-cylinders of the nerve-fibres (see p. 176). To this the conspicuous symptom, the muscular wasting, is secondary. It is perhaps better thought of as a degeneration of the whole segment than as simply a lesion of the ganglion-cells, although the latter being the element on which the nutrition of the segment depends, the two views are merely different modes of stating the same fact.*

But the disease is rarely limited to the lower segment of the motor path. The case mentioned in the note on p. 489 is an almost isolated example of such limitation—an "exception that proves the rule."†

* In a few anomalous cases, degeneration of the motor cells of the cord has been met with when no degeneration could be detected in the peripheral mixed nerves (see Kroghal, 'Neur. Centraalbl.,' 1891, p. 133). The interpretation of these cases is, at present, uncertain. Possibly, complete disappearance of some fibres left the rest apparently normal, although the skill of the investigators renders this explanation difficult to accept. Many more observations, however, are needed to justify a modification of current views.†

Moreover, the fact that these cases may not be primarily spinal must be borne in mind.
We have seen that the pyramidal tracts are commonly degenerated, and it is probable that the degeneration often extends through their entire extent, and involves the motor cells of the cortex. Thus the upper segment is often degenerated as well as the lower segment. In the presence of that complete degeneration of the lower segment which causes the atonic atrophy of the muscles, the degeneration of the upper segment can cause no symptoms. The loss of power that it would produce is also caused by the degeneration of the lower segment, and the latter abolishes the myotatic irritability, excess of which is the characteristic indication of disease of the upper segment. Hence the degeneration of the upper segment of the motor path for the muscles that present the atonic atrophy cannot be recognised during life, for it cannot produce its characteristic symptoms.

What is the relation between the degeneration of the two segments, between the lateral sclerosis and the affection of the ganglion-cells? We cannot assume (as some have been inclined to do) that the affection of the upper segment is secondary to that of the lower, because a primary lesion of the grey matter, such as occurs in polio-myelitis, does not cause ascending degeneration of the related pyramidal fibres. Moreover, even a complete interruption of the pyramidal tracts is followed by no degeneration of their upper parts. Hence the intense degeneration met with in progressive muscular atrophy cannot be regarded as secondary. Neither, when there is atonic atrophy, can we consider the affection of the upper segment to be the primary lesion, and to be the cause of that in the lower. In the seat of slight atrophy there are, as a rule, no indications of the preceding degeneration of the upper segment, which would certainly exist if this lesion were the primary change. Moreover, degeneration of the upper segment does not necessarily cause any degeneration of the lower. It is a matter of every-day observation that intense degeneration of the termination of the upper segment may occur, from dorsal myelitis, for instance, without any considerable wasting of the legs, and such degeneration has never been known to excite the complete degeneration of the lower segment which causes atonic atrophy. Hence the only adequate explanation of the facts is that the degeneration of the upper and lower segments is simultaneous, or if not simultaneous, at least so far independent that neither is the cause or consequence of the other; both are the results of the same tendency to degeneration of the motor path. Atonic muscular atrophy is thus, at least in many cases, the visible expression of a tendency to decay of the whole motor path from the cortex of the brain to the muscles.

The weakness of some parts, as the legs, with excessive myotatic irritability, often going on to spasm, is explained by the degeneration of the pyramidal fibres for the legs, the lower segment being unaffected. It is easy to understand that the affection of the upper segment, and escape of the lower, or the affection of both, may vary much in relative extent, and give rise to the multiform varieties already mentioned. In such a case, the nerve-cells for the part thus paralysed are normal. This weakness of the legs usually succeeds the wasting in the arms, and in most cases in which spastic paraplegia is followed by atrophy in the arms, the latter have not shared the spastic palsy. When this condition is associated with slight wasting of the legs, without considerable change in electrical irritability, the condition exists that we have more than once considered, in which we must assume that the motor nerve-cells of the cord, while structurally intact, undergo slight changes in nutrition. In this condition there are many nerve-cells of normal appearance in the grey matter; the alterations in nutrition are too slight to cause changes in aspect, or if there are such changes we have not yet learned to detect them. They are perhaps results of the degenerative changes in the termination of the upper segment; when such degeneration is secondary to a focal lesion of the cord or brain, the changes in nutrition of the cells seldom attain such a degree as to arrest the myotatic irritability, or to cause wasting such as attends the destruction of these cells.

In some cases, however, the muscular wasting may be great, although the increase of myotatic irritability persists. In these cases, as we have seen, there is considerable rigidity of the muscles throughout the whole course of their wasting, the condition that we have termed "tonic atrophy." In such a condition it is common to find that many nerve-cells have disappeared or are very small, but others remain normal or slightly changed in aspect. Apparently, in addition to the degeneration of the upper segment and to the nutritional changes just mentioned, we have then a considerable degeneration of many, but not destruction of all, the elements of the lower segment. We cannot regard this as simply secondary to the degeneration of the upper segment, for the reasons already given. It must be the expression of a distinct pathological tendency similar to that which elsewhere causes the atonic atrophy and total wasting, but lighter in extent and later in time—insufficient to prevent the less affected cells from causing rigidity under the influence of the degeneration of the upper segment. It is doubtful whether the tonic atrophy ever goes on to atonic atrophy. Theoretically conceivable, it is certain that if it ever occurs it is extremely rare. Nor does it seem that atonic atrophy ever gives place to tonic atrophy, with excessive myotatic irritability. The rigidity of tonic atrophy is due to the degeneration of the upper segment, but the effect is not produced if the lower segment is already the seat of such extensive degenerative changes as abolish myotatic irritability. The pyramidal fibres for the parts that are the seat of atonic atrophy are constantly found degenerated,

* At the same time, we have seen (p. 213) that the nutritional stability of nerve-fibres is less in their lower than in their upper parts, and that their isolated degeneration may be greatest in the lower parts. That this may sometimes also be the case in these pyramidal fibres is suggested by the fact already stated that the degeneration may extend no higher than the destruction of the or area cerebri.
although the muscles have been flaccid to the last. In the rare cases (mentioned on p. 480) in which muscles, with atonic atrophy, become rigid towards the end of the process, it is probable, as the tenderness suggests, that the rigidity is idiopathic, due to the changes in the muscles, and is not dependent on the central nervous system. It may be the result of the increase in the interstitial connective tissue, and the longitudinal division and fibration by which the muscular fasciculi come to resemble bundles of connective-tissue fibres. It is also possible that similar idiopathic muscular changes may ultimately, in tonic atrophy, maintain and increase the rigidity that is primarily dependent upon the spinal cord.

It has been mentioned that the cases in which the legs present the simple palsy and spasm, or the tonic atrophy, which indicate degeneration of the pyramidal tracts, have been separated by Charcot, and termed "amyotrophic lateral sclerosis," the separation being based on the assumption that in such cases the primary lesion is the degeneration of the pyramidal tracts, and that the affection of the grey matter is secondary or "deuteropathic," even where the atrophy is atonic. We have seen also that this assumption is unwarranted so far as the atonic atrophy is concerned. It is probable that the pyramidal tracts are degenerated, if not constantly, at any rate in such a very large proportion of the cases of progressive muscular atrophy, that Charcot's distinction is, in effect, giving a new name to an old disease, and that the sequence is not that indicated by the name. Whether there are indications of lateral sclerosis or not, depends on the circumstance whether the degeneration of the pyramidal fibres is or is not more extensive than the complete degeneration of the nerve-cells that causes atonic atrophy. If the latter is universal, the pyramidal tracts may be totally degenerated, and yet there may be none of the characteristic indications of such degeneration. On the other hand, both arms and legs may be the seat of the spastic paralysis that indicates pyramidal degeneration, and atonic atrophy may be limited to a few muscles of the hands. Between these we have every gradation, in degree and distribution, of atonic atrophy, spastic paralysis, and tonic wasting.

The process in the grey matter has been regarded by some as a chronic inflammation. The occasional rapid increase in the symptoms may be thought to be justification for this view; but the process in general is at the degenerative extremity of the series of nerve-lesions. The principle involved in this question has been already discussed, and we have seen that, whatever be the nature of the primary process, we must recognize secondary tissue changes of independent energy, and that a distinct process of inflammation may occasionally form part of these (see p. 398). The significance of the occurrence of inflammation may therefore easily be overrated, so far as concerns the question of the process, and the tissue-elements in which the disease begins.

Diagnosis.—The simultaneous and gradual onset of weakness and wasting, the slow but progressive increase and extension of the symptoms, render the diagnosis of the developed malady simple and easy. At the onset, when only a single muscle or group of muscles is affected, the question arises whether the atrophy is local or is the commencement of a wider affection. Local atrophy is said sometimes to occur from great over-use of a muscle, especially one of the small muscles of the hand, but such a case is extremely rare, and only to be suspected on the clearest indications.

As a rule, the local atrophy from which the affection has to be distinguished is that due to disease of the nerves. The wasting from disease of single nerves or at a plexus (as the brachial) is sufficiently distinguished by its limitation, coupled with its rapid onset and associated sensory symptoms. Much more difficult is the distinction of some forms of multiple neuritis. The difficulty presents itself chiefly in two forms:—(1) When the spinal affection begins as sub-acute atrophic palsy (see p. 483). (2) When neuritis affects chiefly motor branches, as in the arms in lead-poisoning, or in the legs in alcoholism, &c. In the first case it is necessary to wait for signs of slower wasting in other parts than those first affected before a diagnosis can be made. In the second, a careful search will generally reveal other symptoms of neuritis, and a known cause is usually obtrusive.

In pachymeningitis of the cervical region, with considerable damage to the nerve-roots, the wasting in the arms may resemble that of progressive muscular atrophy, and there is often weakness with rigidity in the legs, but the wasting is less chronic in onset, and is always accompanied by distinctive sensory symptoms,—by acute pains and usually by anaesthesia, irregular in distribution. The same distinctions suffice for the diagnosis in diseases of the nerve-roots of the cauda equina, as by a tumour. This may cause slow wasting in the legs, but there is always severe pain and loss of sensibility. Chronic disseminated myelitis may cause wide-spread muscular atrophy, but is distinguished by the presence of symptoms of irregular damage to other structures in the cord. The diagnosis from syringo-myelia will be considered in the account of this disease.

From primary muscular atrophy, "idiopathic atrophy," "muscular dystrophy," as it has been termed, the diagnosis is sometimes easy, sometimes very difficult. It is easy in the pseudo-hypertrophic form, and often also in the atrophic variety (the cases in which no muscles are large), on account of its characteristic distribution, its course, the age at which it begins, and the tendency to affect many members of a family, and males more than females—features that will be presently described. Whenever several cases of muscular atrophy occur in a family, or during childhood or youth, the probability is great that they are idiopathic and not spinal. Indeed, the idiopathic form is to be suspected whenever muscular atrophy begins under twenty, unless there are distinctive spinal symptoms. But cases are sometimes met
with in which idiopathic atrophy begins in adult life, and the distinction of such cases may be very difficult, and will be better understood after a perusal of the account of this disease.

**Prognosis.**—The progressive character of the malady renders the prognosis, in every case, grave and uncertain. The chief guide is the observed tendency of the morbid process, both the fact of its actual advance and the energy it manifests. At the same time, there is a possibility of arrest, greater in middle life than in old age. Some increase of atrophy in the parts already affected may occur for a short time after the process in the cord has ceased to spread—the muscular wasting going on until it corresponds to the changes in the nerves that have already taken place. The prospect of arrest seems to be greater in the cases in which the wasting is strictly symmetrical and nearly simultaneous on the two sides, than in those in which it is irregular, and attacks the second side when it has attained a marked degree in the first. Spontaneous cessation unfortunately seldom tends to occur until an advanced stage is reached; but, as the result of treatment, arrest may take place at any stage. The danger to life is chiefly proportioned to the interference with the muscles of respiration, and to the indications of implication of the medulla. Bulbar symptoms increase the gravity of the prognosis, especially when definite weakness can be recognized. Slight vague difficulty in articulation may remain stationary for years, and does not necessarily render the prognosis worse, especially if the atrophy elsewhere does not increase rapidly. If the malady ceases to advance, the prospect of any recovery depends on the rate at which the disease has progressed. Recent rapid loss of power may be to some extent recovered from, especially when the muscles present the degenerative reaction. Wasting that has existed for six months will probably persist unchanged. In a typical chronic case there is little hope of any actual recovery of tissue or power. The effects depend on a slow destruction of nerve-elements, the renewal of which seems to be impossible.

**Treatment.**—The first important element is to secure favorable conditions of life, and to maintain the general health as perfect as possible. Fresh air and gentle exercise are important, but all fatiguing exertion should be avoided, and likewise all mental strain. When the patient becomes helpless, great care is necessary. Bedsores in this disease mean inattention, and may always be avoided.

Only one method of treatment has, in my own experience, shown itself capable of arresting the disease, not indeed in all, but in more than half the cases in which it has been employed. * It is the admin-

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* At first I regarded the apparent result with doubt, but careful and repeated observations have made it impossible to consider the arrest of the disease as other than the direct effect of the treatment. It may be asked, Why should not a larger dose by the mouth be equally effective? A large dose involves risk of over-stimulating the nerve-elements, and doing harm rather than good; it is quite different from a small dose acting suddenly.

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**CHRONIC MUSCULAR ATROPHY.**

Administration of strychnia by hypodermic injection. In seven almost consecutive cases, in middle life, this treatment has been followed by arrest within a month of its commencement, and the arrest has been permanent in all the cases but one. In the senile cases the treatment has failed, but in most of them the disease was in an advanced stage, and the lumbar cord had begun to suffer. In some of the cases in which the result was prompt and distinct, strychnia given by the mouth had failed. It is conceivable that the different result is due to the fact that the agent is brought into more rapid contact with the nerve-elements, perhaps in purer form. Its action may possess greater momentum, as it were, and may thus exert an influence on the nutrition of the nerve-elements much more considerable than when it is slowly absorbed from the alimentary canal. One injection daily has been given, at any convenient place. The nitrate is the most convenient salt, one hundredth of a grain at first, quickly increased to one fortieth. It is not wise to give a larger quantity, lest the nerve-elements should be over-stimulated; although, in their damaged state, they may be unable to manifest the influence, it may yet do harm. When the malady is apparently arrested, it is well to intermit the injections for one week in three or four. Other nerve tonics seldom exert a distinct influence, but those that are useful in other degenerative diseases, as tabes, may be given by the mouth at the same time as the injections are employed. In a malady so grave, it is desirable to neglect nothing that may possibly exert a beneficial influence.

Local treatment of the muscles has very little influence on the wasting, as may, indeed, be expected from its nature. The most sedulous and skilful use of electricity, voltaic or faradic, fails, as a rule, to produce any effect on the course of the disease. If the malady is progressing at the same rate in each arm, and the muscles of one arm are regularly treated with electricity, while those in the other arm are left alone, no difference can be detected in the rate of wasting on the two sides. It is possible, nevertheless, that electricity sometimes does a little good. In cases in which a rapid loss of power has occurred, and weakness is out of proportion to the wasting, some recovery is possible, and there is no doubt that the excitability of the muscular tissue is maintained for a longer time by galvanism, although the bulk of the muscle may not be influenced. In other cases, all that can be said is that the influence of electricity, properly applied, is in the right direction. Moreover, the disease is one of those in which patients find it hard to believe that electricity cannot help them, and the probability is that their conviction will be fostered by some medical adviser. It is one of the diseases in which unjustifiable assertions are too often made that early electrical treatment would have been successful. It may be well, therefore, if only to satisfy the patient that nothing has been left untried, that a careful course of electrical treatment should be adopted. Paradism may be used.
if the muscles are sensitive to it, but if they present any greater irritability to volatiaim, it is better to use this. It is immaterial whether the application is confined to the muscles or whether one electrode is placed over the affected part of the spinal cord. The latter method has no disadvantages, but my own observations have failed to confirm the confident statements sometimes made regarding its superiority. It is very important that the current-strength employed should be moderate. Strong applications often cause much subsequent pain, and even increased disability, and should be carefully avoided. I have known a rapid increase of weakness follow a strong application, in a way that convinced the patient at least that the two were connected.

Rubbing and massage of the muscles have been frequently employed, and of this treatment also it may be said that its influence is in the right direction, although usually inappreciable so far as the muscular atrophy is concerned. Combined with passive movement, the influence of rubbing in preventing and diminishing deformities is more distinct. No special bath treatment is of service. When the disease occurs in the subjects of syphilis, specific treatment invariably fails, and I have even known the progress of the disease to be distinctly accelerated by an energetic course of treatment both by iodide and by mercury. It is important that all treatment should be pursued in moderation, and that its effects should be carefully watched.

ARTHRITIC MUSCULAR ATROPHY.

Arthritic inflammation is almost invariably attended with rapid wasting of the muscles that move the joint. This occurs equally, whatever be the cause of the inflammation, and whether this is spontaneous or traumatic. It attends chronic as well as acute inflammations, and occurs in animals if joint-inflammation is produced in them.*

The muscles that waste are chiefly those which extend the affected joint. The atrophy is well seen in the muscles in front of the thigh, when the knee is inflamed. If the ankle is affected, the calf muscles chiefly waste; if the hip, the glutæi; if the wrist, the extensor muscles of the forearm; the triceps when the elbow is affected; the deltoid when the shoulder-joint is inflamed. In rheumatoid arthritis of the finger-joints, such wasting is usually very conspicuous in the interossei, and especially in the abductor indicis. The atrophy, however, sometimes involves the flexors as well as the extensors, and rarely muscles of the limb that are near but do not move the affected joint. In very rare cases all the muscles of a limb have presented some wasting. Cases