which, however, is rarely maintained. On the contrary, as a rule, it partially recedes and confines itself to certain muscles, which then remain permanently paralyzed. According to Beevor (cf. lit.), the affection sometimes embraces groups of muscles corresponding to those which Ferrier in his experiments on monkeys saw contract after stimulation of the different cervical nerve roots. In the majority of cases the paralysis takes in one leg. The paralyzed muscles rapidly atrophy, and the electrical excitability undergoes quantitative as well as qualitative changes—reaction of degeneration. The whole extremity is stunted in its growth, and even the bones may be found several centimetres shorter than those of the other leg. The appearance of such an extremity, in which at first all passive motions are possible, is quite characteristic. The skin is pale, cyanotic, and feels cold, but retains its sensibility completely. Skin and tendon reflexes are lost, but there are no vesical symptoms. Later on secondary contractures develop, among which the so-called "paralytic clubfoot" is the best known. In consequence of the paralysis of the peroneal muscles, their antagonists, the calf muscles, become permanently contracted and cause the point of the foot to hang down. In the arms analogous conditions may be found, the non-paralyzed antagonists always assisting in the production of the contractures.

Roughly speaking, this is the course in most cases, only occasionally the initial fever may be slight enough to be overlooked and the paralysis develop without the child ever having taken to his bed. In rare cases the convulsions, instead of lasting for days, continue for weeks. In others, again, several months may pass before the onset of the actual paralysis; but all these are the exceptions, which need not confuse us in making a diagnosis. The further general development (with the exception of that of the paralyzed extremity) is perfectly normal, and neither, as happens in the cerebral infantile paralysis, does the mind become in any way impaired nor do the initial convulsions ever recur. The child grows up in good health, but always remains, especially if one leg is affected, a cripple. If, as often happens, contractures or a spontaneous paralytic luxation of the hip develop, the patient has for years to be under the care of the surgeon, and needs braces and the like. If an arm is affected, the capability of the patient for making his living is naturally considerably and permanently interfered with.



**Diagnosis.**—It is not difficult to avoid mistaking the disease for any other if we bear in mind the characteristic onset, the localization, the behavior of the paralysis itself, the flaccid condition of the muscles, the absence of the reflexes, and the cold and cyanotic skin. Where we find a hemiplegia—i. e., where the arm and leg of the same side are paralyzed—we should in children always first think of infantile spastic hemiplegia (page 271), as it is one of the rarest exceptions for the spinal paralysis to take on this distribution. Confusion with the syphilitic pseudo-paralysis, also known under the name of Parrot's disease, is avoided by remembering the fact that in this disease the paralysis makes its appearance immediately, or at least within a few days, after birth (Dreyfouss, Revue de méd., août 1885, v), while Heine's paralysis of children does not occur at such an early age.

**Prognosis.**—The prognosis, as soon as the initial acute symptoms have passed, is, so far as life is concerned, absolutely favorable; so far as the recovery of function in the affected extremity is concerned, equally unfavorable. Any notable improvement is very rare, complete cure out of the question. These points should be carefully considered before inducing a poor and struggling father to let his child undergo year after year an expensive and useless course of treatment.

**Ætiology.**—Of the ætiology of the disease we know nothing. It is doubtful whether cold is ever a causative factor. It is possible that infectious influences, the action of certain microorganisms, will at some time be proved to be the cause of the disease. For the present, however, this is nothing more than a hypothesis which has not gained any firmer ground from the report of Cordier of an epidemic of the disease (Lyon méd., 1888, I, 2). In a small village thirteen children were inside of two months taken ill with anterior poliomyelitis and four died. According to Cordier, the appearance of the disease in summer, the sudden onset, the similarity in course, speak for an infectious origin, the infection, as he supposes, taking place through the air passages.

**Treatment.**—Little more is known about the treatment than about the ætiology. All measures to cure or even merely to improve this rapidly developed paralysis are more or less useless. Electrical treatment with the faradic or galvanic current, systematic massage, gymnastic exercises, together with rubbing with all sorts of salves—all these have been tried without any

noteworthy success. In a few cases I have seen the methodical use of heat, in the form of hot sand baths, warm packs, etc., bring about at least a perceptible improvement; but even here this was out of proportion to the care and trouble which had been taken. Certain it is that the influence of the different baths has been greatly overrated, whether it be the brine baths of Kreuznach, Reichenhall, Kolberg, or the chalybeate springs of Pyrmont, Flinsberg, Schwalbach, or the sodium waters of Rehme, Soden, or, finally, the non-medicated hot springs of Gastein, Johannisbad, and many others, each of which has its advocates. The most appropriate appear to be those last mentioned, but in most cases we shall even then find that while perhaps the child's general condition is improved and it becomes strong owing to the good hygiene and fresh air, the paralysis, for the sake of which all has been undertaken, remains absolutely unchanged and presents no improvement.

In view of these unsatisfactory results, the interesting but still scanty communications, according to which the growth of bone can artificially be increased, deserve our deep interest. In 1887 Helferich proposed to tie round the affected (paralyzed, atrophic) limb elastic rubber tubing so as to produce an artificial engorgement, and through this a more active nutrition of all the tissues, including the bones. Schüller also has reported before the Berlin Medical Society, November 28, 1888 (Deutsch. Med.-Ztg., 1888, 99, page 1182), several cases which were thus treated and which showed decided improvement. To judge from his communication, this treatment undoubtedly should be tried in all suitable cases. It is, however, a procedure which, as well as the orthopædic treatment so important for the prevention of deformities, should not be undertaken without consulting a surgeon.

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Though the lesions of the gray anterior horns when occurring in children are well understood, both in their anatomical and their clinical aspect, yet when the same process takes place in adults our knowledge becomes very limited. Here the material at our disposal is still so small that only in rare exceptions can we say definitely whether we are dealing really with an anterior poliomyelitis and not rather with a peripheral disease, a multiple neuritis. Clinically, the differential diagnosis between the two can only be made in the initial stage, as the peripheral disease is accompanied with pains and sensory disturbances which are absent in the central affection.

A patient is taken ill with grave general disturbances—fever, somnolence, convulsions, delirium, etc.—and within a short time, perhaps in one or two weeks, a widespread paralysis in all four extremities is developed. The paralyzed muscles become flaccid and atrophy, the tendon reflexes disappear; sensation, however, as well as bladder and sexual functions, present no abnormity. With a history like this we must think of a lesion of the anterior gray horns. This idea becomes more than a conjecture if on examination the affected muscles are found to be such as are supplied from ganglionic cells, which most probably lie in close proximity to one another in the spinal cord. In such cases, as Remak has shown so beautifully, certain types of paralysis are observed—the forearm type (paralysis of all the extensors without the supinator longus) and the upper-arm type (paralysis of the biceps brachialis

anticus, deltoid, and the supinator longus)—but unfortunately such instances are rare, and therefore even quite an experienced physician may feel uncertain about the diagnosis.

The difficulty becomes greater if the paresis or paralysis is not extensive and does not develop rapidly, but slowly and by fits and starts. In these cases not rarely a temporary improvement may be noted and arouse hopes of complete recovery, unfortunately never justified. These are the instances in which we find not complete but partial reaction of degeneration in the paralyzed muscles—intermediate form of chronic anterior poliomyelitis (Erb). It goes without saying that we must have the other symptoms, especially the loss of reflexes, even to justify a conjectural diagnosis. Moreover, it is necessary that there should be absolutely no sensory changes, and that bladder and sexual functions should be normal. Of the points of difference between anterior poliomyelitis and tabes we shall speak later.

We can hardly expect much from any treatment. Electricity, however, should be tried, if for no other reason than that something is done. Duckworth recommended, besides, belladonna, iron, quinine, and cod-liver oil, and claimed to have cured cases with these remedies.

With reference to the ætiology, nothing certain is known. Whether traumatism can ever cause anterior poliomyelitis remains doubtful, notwithstanding the report of Gibbons (Med. Times and Gazette, September 5, 1885). He had among his patients a boy nine years of age who after a fall on his knees developed the symptoms of an anterior poliomyelitis (and recovered completely!). In cases of chronic anterior poliomyelitis which came to autopsy, sometimes atrophy in the ganglionic cells of the anterior horns through the whole length of the cord, as well as atrophy of the anterior roots, was observed, while the peripheral nerves remained intact (Oppenheim).

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## CHAPTER II.

### ATROPHIA MUSCULARIS PROGRESSIVA SPINALIS—PROGRESSIVE MUSCULAR ATROPHY.

PROGRESSIVE muscular atrophy was first described by Duchenne and Aran in 1849 and 1850, and was recognized by Cruveilhier in 1855 as a spinal affection. Thanks to the work of Lockhart Clarke, and especially that of Charcot, the occurrence of a pathological process restricted to the gray substance of the spinal cord, which is accompanied by a muscular atrophy of typical distribution, is now established beyond the slightest doubt.

**Pathological Anatomy.**—The process, which is usually most pronounced in the cervical cord, consists again of an atrophy and transformation of the gray anterior horns into a fine fibrous tissue containing spider cells. The large ganglionic cells are partly or wholly destroyed, or at any rate are diminished in number and perceptibly smaller. Here, too, the lesion extends to the anterior nerve roots and corresponding fibres of the motor nerves. On microscopical examination we find that the muscles supplied by them retain their transverse striation, but the fibres are decidedly diminished in size. Some fibres also show the so-called degenerative atrophy—that is, a fatty, wax-like degeneration, with increase of the interstitial connective tissue and multiplication of the muscle nuclei. Which of the described processes has to be regarded as the primary one, in other words, whether the disease actually does start in the gray matter of the cord, and not perhaps in the peripheral nerve endings; whether both processes may occur at the same time, or whether they may succeed each other in the same individual, and at what age they occur, all these points have recently given rise to much controversy, as has also the question of the importance of hereditary influences. Those who wish to inform themselves more thoroughly on this subject are referred to the articles by Hoffmann (Deutsche Zeitschr. f.





Nervenheilk., 1893, iii, 6, p. 427), Strümpell (ibid., p. 471), Bernhardt, Ueber die spinal-neurotische Form der Muskelatrophie (Virch. Arch., 1893, cxxiii, Heft 2), and others.

**Symptoms.**—The onset of the disease is in many cases very characteristic. The patient begins to complain of weakness in the arms, sometimes more in the right than in the left, which soon interferes to some extent with his ordinary actions. Sensory changes and pains are absent—a point which is of vast diagnostic importance. Not many weeks after these symptoms have appeared the competent observer will notice a peculiar flatness, a sunken-in condition of the ball of the thumb, while at the same time the thumb is more than usually approximated to the second metacarpal bone ("ape hand," Fig. 138). The interosseal spaces on the back of the hand are sunken in and

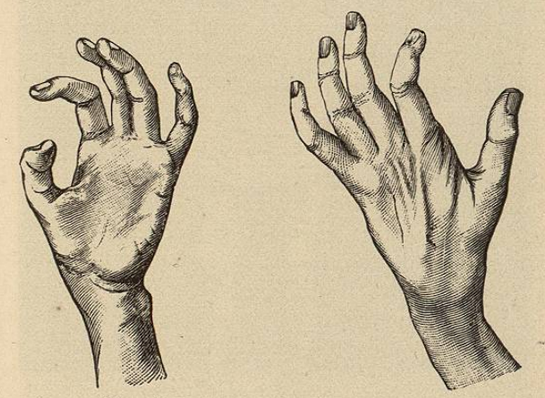

Fig. 138. Fig. 139.

Figs. 138, 139.—PROGRESSIVE MUSCULAR ATROPHY. (After EICHHORST.) Fig. 138, ape hand. Fig. 139, sunken-in interosseal spaces on the back of the hand.

the terminal phalanges of the fingers are in incomplete extension (Fig. 139). The hollow of the hand seems flattened (atrophy of the lumbricales), and the atrophy of the muscles of the thenar and hypothenar becomes more and more apparent. As the function of the interossei becomes disturbed to a greater extent, the same claw-like position of the fingers develops which has been described on page 349 as occurring in affections of the ulnar nerve ("claw hand," "*main en griffe*").

After this condition has thus for weeks or months undergone no marked change, the disease begins to attack either the muscles of the forearm, or, passing over these, implicates the

muscles of the shoulders and with special preference the deltoid. In the former case the extensors are attacked earlier and more seriously than the flexors. The muscles of the trunk and legs are either later or never affected, but if invasion of the diaphragm and other respiratory muscles occur this may prov-

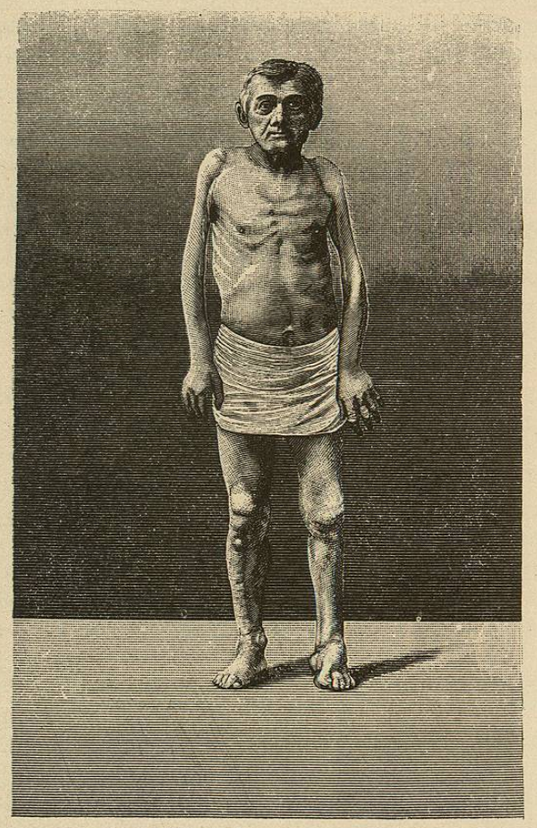

Fig. 140.—PROGRESSIVE SPINAL MUSCULAR ATROPHY (personal observation).

fatal, as may also an extension of the process from the cord to the medulla oblongata, in which case the symptoms of progressive bulbar paralysis are superadded (page 154). If this does not take place and the respiratory muscles are spared, the disease may last for years and tens of years, and death is only caused by an intercurrent acute malady.



Apart from the characteristic onset, the following signs help to make the diagnosis certain: (1) Fibrillary twitchings in the affected muscles, which can at times be produced by tapping the muscles, but which are often seen to appear of their own accord and continue without interruption. (2) The con-

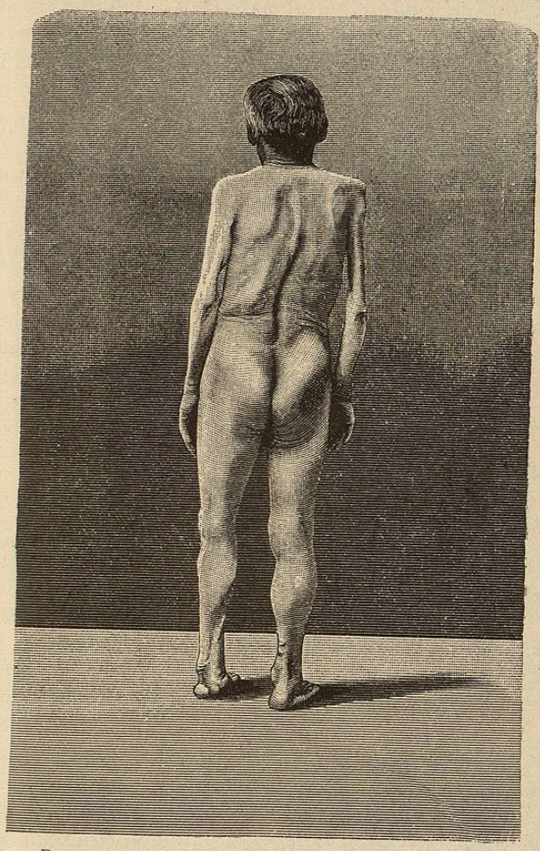

Fig. 141.—PROGRESSIVE SPINAL MUSCULAR ATROPHY (personal observation).

dition of the electrical excitability, which depends directly on the number of muscle fibres left. If the greater number of the fibres are wasted, then the excitability for both currents is equally decreased. If all the fibres of a muscle have disappeared and only fat and connective tissue remain, the excitability of the muscle is completely lost. It is only exceptionally