affections. A hemiplegia due to cerebral hæmorrhage can in most cases be excluded, owing to its rarity in childhood. Such, moreover, would usually not be associated with any muscular

Fig. 79.

The patient, now eighteen years of age, had, when six months old, an "apoplectic stroke," and never learned how to walk properly, since the left half of the body was paralyzed up to her second year. The power of motion has improved to a certain extent; the left leg, however, and the left arm have remained behind in development, so much so that the arm is eight centimetres, the leg twenty-five centimetres, shorter than the corresponding limb of the right side. The shortening of the leg is partly due to a resection of the knee joint performed thirteen years ago (the reason for which procedure could not be made out). Patient suffers from epileptiform attacks, occurring once a month; they last from a quarter to three quarters of an hour, and consist of more or less violent convulsions. During these, consciousness is sometimes completely retained. There is no trace of dementia.

atrophy. In the diagnosis of the bilateral affection we must take into consideration the possibility of a multiple sclerosis, Friedreich's disease, brain tumor (especially tubercles), meningitis, and cerebral syphilis. In many cases it is impossible to come to a satisfactory conclusion.

Prognosis.—The prognosis *quoad valetudinem* is absolutely, *quoad vitam* relatively, unfavorable. Complete recovery is impossible, and has never been observed. If the patient does not succumb during the first days of the disease, he will remain a cripple all his life, his mental condition being good only in the most favorable cases. Under unfavorable conditions he may be epileptic and weak-minded, and to a greater or lesser extent deprived of the use of his limbs. The utmost we can expect is that the diseased side may atrophy only to a moderate degree,

Fig. 80.

The family form of spastic paraplegia: *a*, fourteen years old; *b*, sixteen years old; *c*, thirteen years old. In the first the disease began at the age of seven and a half, in the second at one and a half, in the third at nine. The mother had eleven children, eight of whom are living (among them the three patients). (After Newmark, San Francisco.)

that the patient may be sound enough in other respects, bodily and mentally, and thus be capable of making his own living (Figs. 77 and 78).

Treatment.—The treatment is, on the whole, entirely unsatisfactory. Even by the light of an early diagnosis, we are

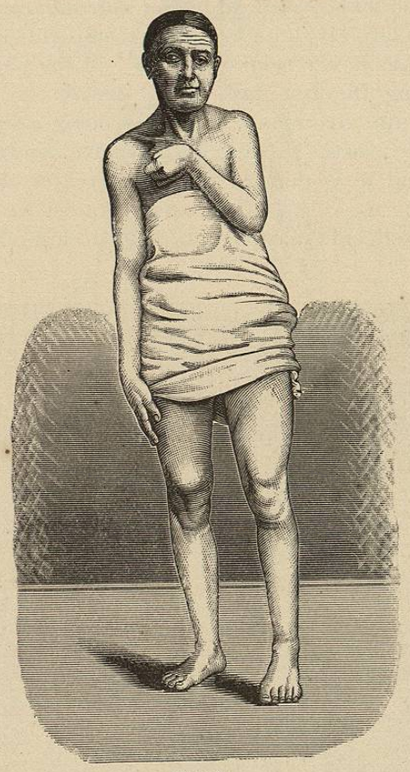

Fig. 81.

The patient is now forty-four years old. The date of onset of the disease can not be definitely determined. She suffered from epileptiform convulsions from early childhood up to her tenth year; these have now entirely disappeared. At times, however, a "*tic convulsif*" (in the distribution of the left facial) is noted. The development of the left half of the body has been retarded, the upper extremity being two centimetres, the lower three centimetres, shorter than the corresponding limb of the right side. There is also a dfference of from four to five centimetres in the circumference of the limbs of the two sides. The shoulder, elbow, and wrist joints are contracted, the first being in a position of adduction, the second in one of flexion, and the third in extension. Marked degree of dementia.

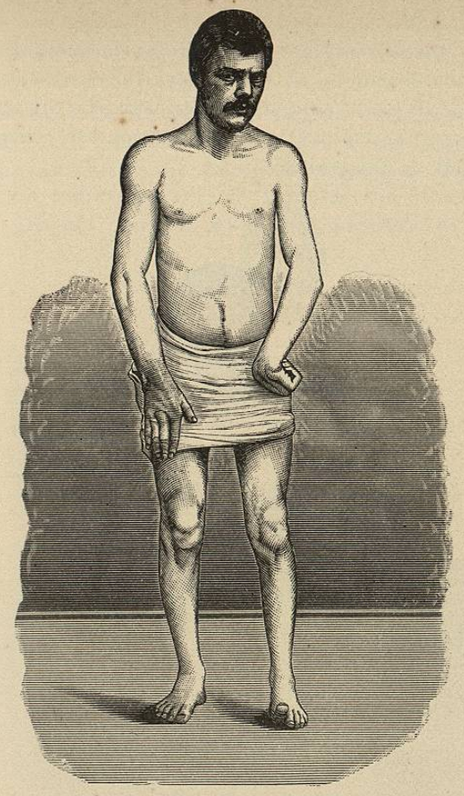

Fig. 82.

The patient, who is now twenty-two years of age, was taken ill with convulsions in early childhood. They ceased, but after an interval of ten years reappeared in his fourteenth year, and have continued up to the present time, being quite severe and recurring frequently. From childhood he has suffered from a severe motor speech disturbance, and is only able to utter a few unintelligible syllables, and that with great effort; at such times almost all the muscles of the body are affected with associated movements. Atrophy of the left side is to be noted. The circumference of the left upper arm measures three centimetres, that of the left forearm two centimetres, that of the thigh four centimetres, and that of the leg two centimetres less than the corresponding measurements on the right side. The left arm is one centimetre, the left leg one centimetre and a half, shorter than the right arm and right leg respectively. The left hand and fingers are in flexor contraction. Patient is moderately demented.

not in a position either to prevent the continuance of the epileptiform attacks or to ward off the changes in the affected extremities, the symptoms of irritation, the atrophy, etc. The symptomatic treatment of the epileptiform attacks by the different bromides and the galvanization of the atrophic parts is all that lies in our power, and, unfortunately, little enough is accomplished by these means.

While we do not attempt to give a detailed account of the pathology of cerebral diplegias (Freud, Leipzig und Wien,

Fig. 83.

1893), two questions must be discussed, namely, (1) under what conditions do contractures, (2) under what conditions do certain movements, which are independent of the will of the patient, develop? Unfortunately, we are not able to answer these

questions satisfactorily. With regard to the first, the idea deserves to be mentioned that the extent of the cerebral lesion and the secondary degeneration depending upon it are of some significance.

The same uncertainty exists in regard to the second question. We are not acquainted with the immediate conditions which, in the course of the cerebral palsies of children, give

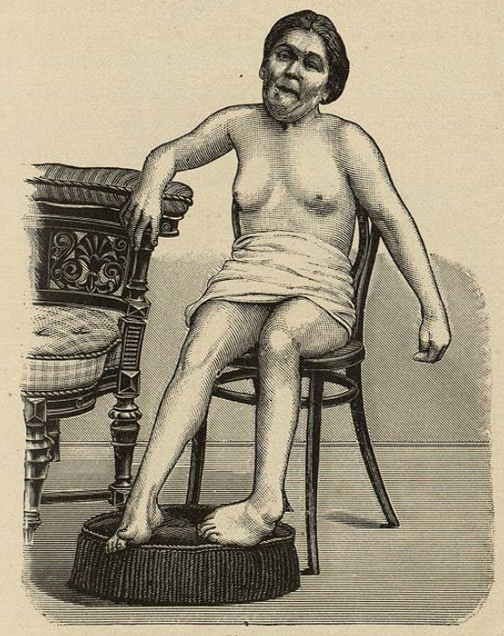

Fig. 84.

The onset of the disease can not definitely be fixed, since the mother of the patient does not remember it, and the patient herself, who is now eighteen years old, is demented and completely deprived of the power of speech. The fact, however, that the illness began in early childhood with convulsions is unquestioned; it is, however, not known how long they lasted nor what followed them. When the girl was five years old she was not yet able to walk, because the left leg was moved only with difficulty, and the foot gradually assumed an equino-varus position, which can still be noted. Patient now walks on the outer edge of her foot, and the leg is scarcely moved at the knee joint. The left upper extremity can be moved voluntarily in the shoulder and elbow joints; the fingers and the hand present athetoid movements, while in the facial muscles of the left side a marked "*tic convulsif*" is noted. Marked dribbling of saliva. Patient no longer suffers from epileptic attacks, but has from time to time periods of excitement, during which she becomes aggressive.

rise to peculiar (unilateral or bilateral) motions in the affected extremities, which present the following characteristics:

The patients are absolutely unable to keep the fingers and the toes of the affected side still; they are in constant motion day and night, during waking and sleeping, without interruption. If we observe these movements more closely, we find them to be relatively slow, rhythmical, and monotonous. The fingers seem to be directed with a definite aim, as if they were attempting to seize something, and it is easily remarked that the normal limits of the movements are exceeded—the fingers are hyper-extended, the toes are elevated almost at right angles or fasten themselves to the floor like claws, etc. (cf. Fig. 85). All this is only possible in consequence of an unusual stretching of the ligaments, which also admits of positions of distinct subluxation. The will of the patient has hardly any influence over these movements, and only in light cases, and then but temporarily, may the patient succeed, by firm pressure of the affected hand upon the body, or by fixing the fingers with the unaffected hand, in restricting a little the abnormal excursions; as soon as the mechanical impediment is removed, they will, however, begin again with increased vigor.

The muscles of the forearm present a firmer consistence, a certain degree of hypertrophy. The arm feels hard, and the surface temperature is 0.5° to 1° C. (0.9° to 1.8° F.) higher than on the opposite, sound, side; not but what the muscular strength is materially lessened and sometimes so much diminished that the examination with Duchenne's dynamometer yields astonishing results. With the affected arm the patient can hardly lift five kilogrammes, notwithstanding the apparently good development of the muscles, while with the well arm five to eight times as much work can easily be done. In the muscles of the lower extremity a similar condition may be noted; not infrequently the ankle joint takes part in these movements of the toes, and, in exceptional cases, the knee joint as well. Other muscles than those of the extremities are not affected.

The first who studied these movements carefully was Hammond, of New York, in 1871. He gave them the special name *athetosis* (*α-τίθημι*) and raised them thus to the dignity of a separate disease, which, in our opinion, they never deserve. Athetosis—and by this we mean the athetoid spasms—does not constitute a disease, but merely a symptom. It is the expression of cerebral affections, the anatomical basis of which is variable.

Only in the rarest instances, one could almost say never, do athetoid movements occur alone without any other symptoms. Almost always they are associated with other disturbances,

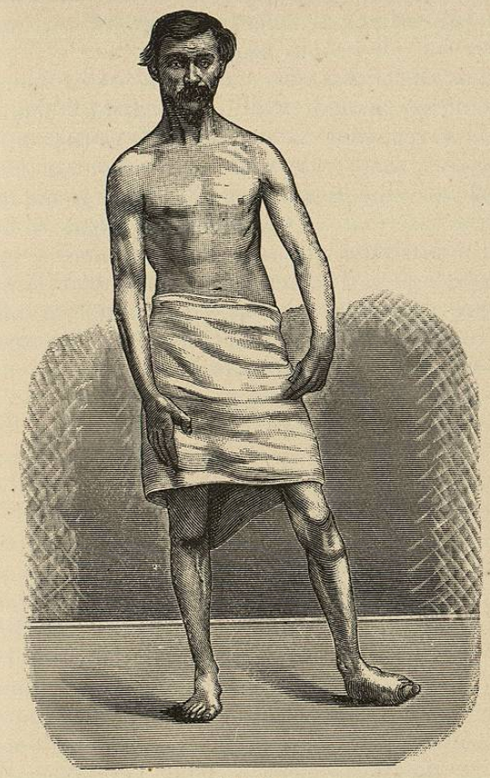

Fig. 85.

The patient, now twenty-nine years old, was taken at the age of six months with an "apoplectic stroke" followed by convulsions, which at first occurred at long intervals, later more frequently, viz., about once every two weeks; they presented all the characteristics of epileptiform seizures. Quite early, peculiar involuntary movements appeared in the left extremities, more particularly in the left arm, which must be considered as athetoid. At regular intervals the fingers are extended and again drawn into the hollow of the hand, this being repeated about fifty times a minute. In the left foot similar, although, of course, less pronounced movements, occurring especially in the ankle joint, are noted. At about the age of five the convulsions reappeared, although occurring with diminished frequency, i. e., from three to five times a year. The patient is excitable, irascible, and at times even violent. Intelligence is normal.

either psychical (the patients are mentally undeveloped, demented, sometimes of a changeable, irritable disposition) or somatic, such as paralyses or spasms in the distribution of different nerves—for instance, the facial—contractures, etc. Again, the patient may be subject to epileptiform attacks which recur at intervals of various lengths.

If we thus affirm that every athetosis—be it the much rarer bilateral form (see above), be it unilateral, the "hemiathetosis"—is only to be regarded as a symptom, we are, on the other hand, willing to admit that there are individual cases where the athetoid movements are such a prominent and dominating feature of the case that we may overlook others, or at least not be inclined to attribute any importance to them. So it is in an instance reported by Gnauck, who speaks of a primary—that is, idiopathic—athetosis, but who has noted a simultaneous paresis of the facial and a hemianæsthesia of the affected side. We can hardly call this an idiopathic affection, but must rather look upon it as a prehemiplegic phenomenon (cf. page 218); and, similarly, some explanation can be found for the few remaining cases published as "idiopathic" athetoses, some of which were congenital. These movements are always a symptom of cerebral disease. That they are occasionally met with in the course of other diseases—e. g., spinal affections, especially tabes—there can be no doubt. The pathological changes observed in cases which had presented athetoid movements during life, in addition to those found in cases of cerebral palsies of children, consisted in small foci of softening in the basal ganglia, the thalamus (Lauenstein), the corpus striatum (Schulz), and in the temporal lobes (Ewald), although we can in none of these instances be certain that the lesions found were actually the cause of the movements. After cerebral hæmorrhage where we have a lesion of the internal capsule, in old hemiplegias therefore, hemiathetoid movements are occasionally seen, yet, in comparison with the frequency of cerebral hemiplegias in adults, these are very rare, certainly much rarer than in the so-called infantile hemiplegias. We see, therefore, that cortical lesions and lesions of the cortico-muscular tract as well as of the basal ganglia may give rise to athetoid movements, although we do not understand the *nexus causalis*, if indeed such exist. In our opinion, disease of the cortex undoubtedly plays the principal part in the causation of athetosis, and we can all the more reckon upon the occurrence of athetoid movements

if the cortical disease has appeared in early childhood and has been either entirely confined to or has affected more particularly the motor region, the central convolutions, and the adjacent portions. In lesions of the other parts of the brain, especially of the basal ganglia, the thalamus, the lenticular nucleus, and the caudate nucleus, athetoid movements are only exceptionally developed, the conditions which favor their occurrence being then wholly unknown. That there is a cerebral lesion which produces no other symptom, whether psychical or somatic, than these movements is unlikely, and consequently, as we said, the name "athetosis," as indicating a separate disease, can not be held to be justifiable.

Keeping well in mind, then, the characteristics of the movements which have just been described, and especially after having had occasion to study their peculiarities, one can hardly mistake them for anything else. A good point to remember is that they continue during sleep, so that the patients have to stop or at least impede them by mechanical appliances.

We shall give up the idea of chorea or hemichorea which we might entertain should the athetoid movements be accompanied by facial spasm, if after observation of the patient we have been convinced that the movements persist when he is asleep. Furthermore, the duration of the disease and the fact that it resists all therapeutic measures, more especially the protracted use of arsenic, are facts not reconcilable with the diagnosis of chorea. Other points of difference will be found in the chapter on the latter disease.

We possess no specific which will put a stop to these athetoid movements; their treatment is that of the primary disease, and, as this is usually beyond our reach, the outlook in athetosis is necessarily very gloomy. If Hammond claims to have effected a cure by stretching the median nerve, we may be pardoned for asking how long this cure lasted; and if Gnauck has seen the movements disappear after the use of the galvanic current and the internal administration of potassium bromide, we are justified in assuming that in his case the affection was due to a functional disturbance of the motor area. What lasting good results can be accomplished by hyoscine, a drug which has been used by Erb, I have not as yet been able to establish with the material at my disposal.