The clinical course of the disease is practically unknown. Probably there does not exist any well-defined constant clinical picture, but the symptoms vary according to the anatomical seat of the process. They are symptoms of paralysis or of irritation, and are partly "cortical symptoms" and partly to be referred to disease of the cerebral vessels (cf. Friedmann, Arch. f. Psych., 1889, xxi, 2, page 461). In the few cases in which a diagnosis could be made during life, apoplectiform attacks, rhythmical choreic movements, longer or shorter spells of unconsciousness, were observed. The difficulty of grouping and correctly interpreting the symptoms is chiefly owing to the impossibility of an early diagnosis. Hence it will be the chief task of future observers to direct their attention to the initial stage, for only after we have once become familiar with the development and the anatomical changes in this first stage can we hope to elaborate an efficient mode of treatment, which, we need not say, at present is absolutely wanting. The irrational trials with potassium iodide we can certainly not regard as such.

B. IN CHILDREN.

Cerebral Palsy of Children—Polio-encephalitis (Strümpell).

Pathological Anatomy.—In view of the comparatively frequent occurrence of cerebral palsies in children, it is rather to be wondered at that so extremely little is known about their pathogenesis and their initial stage, more especially with reference to the anatomical changes that occur. This may perhaps be accounted for by the difficulty, and sometimes even impossibility, of making an early diagnosis. At a time when we are able to recognize the disease we usually have to deal with a process which has already passed through all, or almost all, of its different stages. It is the same with the lesions which we find: they in no wise explain the exact nature of the disease, but only give us an idea of the many various ways in which the brain with its meninges may be altered in early childhood as a consequence of the disease, which was most probably intra-uterine. General cachexias of the parents —e. g., syphilis—may be the cause when the affection begins during intra-uterine life; in this case more than one child may have the disease. During the act of birth traumatisms may produce unilateral or bilateral cortical hæmorrhage. After birth, infectious diseases (pertussis, scarlatina, tuberculosis,

diphtheria, syphilis) play the most important *rôle* in the causation. The case which is briefly described on page 277 (Fig. 78) shows that injuries—from a fall, for example—may also precede the affection. We do not, however, know of what nature this process is, whether it is a sinus thrombosis, as Gowers claims, or an inflammation leading to atrophy, as in meningo-myelitis chronica. Neither can we tell whether the increase in the connective tissue which has been noted by many authors is a primary one, and what part the disease of the vessels, the thickening of their walls (Hayem, and others), plays in the process; but one thing is certain, that the disease is not confined to the gray cortex alone (as Strümpell has assumed, and for which reason he has proposed the name polio-encephalitis, analogous to polio-myelitis, cf. lit.), but that the white matter as well may be implicated. This is shown by the case published by Kast (cf. lit.), and also by the following observation, which was made in my wards, and which I propose to relate here in brief, as autopsies in cases of this class are rare:

Magdalena St., twenty-one years old, coming from a healthy family, was taken sick in her second year with violent fever. According to her mother's account, she had convulsions for four days and four nights. When she wanted to get out of bed after this her left side was found to be paralyzed. Inside of three months her condition was so far improved that she could walk, although with a limp. Gradually the left lower leg became smaller and somewhat curved, and she complained of pain in the whole limb. The upper extremity did not at first take part in the atrophy; it was, however, almost completely powerless. For two years the convulsions did not reappear; but for the last four years the patient had had, on an average, about one epileptiform attack every three weeks, in which she bites her tongue and passes her urine involuntarily. The following is an extract of the note made on October 25, 1885:

Head: Right parietal region painful to percussion; in the region of the left glabella and the hairy part of the scalp, on the same side, there are several areas of anæsthesia. Pupillary reactions and movements of the eye muscles normal. Nothing abnormal in the distribution of the facial and hypoglossal nerves. On the right side hearing is much below normal, on the left there is complete deafness. On the anterior third of the left half of the tongue taste is lost. Uvula straight and movements of the soft palate normal.

Trunk: On the left half of the chest touch and the prick of a pin are not perceived; temperature sense seems decidedly subnormal.

Upper Extremities: The whole left upper extremity, including the hand, is shorter and smaller than the right; motion of the wrist, especially extension, is impaired. The hand is flexed on the forearm, and only with force can the flexion be overcome. The thumb is drawn into the hollow of the hand, the rest of the fingers are slightly flexed. Motion in the shoulder joint normal; in the elbow joint extension is slightly impaired. There is a general decrease in the sensibility. Electrical reactions are found to be normal for both currents, on direct as well as on indirect stimulation. The right upper extremity does not show abnormity with regard to development, size, mobility, or sensibility.

The left lower extremity is considerably smaller and shorter than the right; sensibility is the same as in the corresponding upper extremity; the same holds for the electrical condition. Tendon and skin reflexes are retained on both sides.

The epileptic attacks continued, and occurred about every sixth or eighth day; the intelligence became more and more impaired. A tuberculous process in the left lung was superadded to the already existing trouble, and, in consequence of general failure of strength, the patient died on March 22, 1886.

Autopsy: Eighteen hours after death. Extract from the post-mortem record: After opening the skull the pia is seen to be considerably thickened at different places, especially over the right hemisphere. In volume the right hemisphere is not much smaller than the left; the anterior and posterior central convolutions on the right side, especially in their lower half, are markedly atrophic, the gyri are shrunken to about a third of their natural size; the marginal and the angular gyrus present the same atrophic condition. The upper right parietal lobule is less atrophic, nevertheless the gyri are here also remarkably narrow. The portions of the first and second frontal convolutions bordering on the central convolution appear also atrophic. On section the gray matter is seen to be considerably diminished.

The ventricles appear markedly enlarged. On frontal sections, after Pitres' method, no important changes, with the exception of the shrinking, either in the centrum ovale, or in the basal ganglia, can be observed macroscopically; on microscopical examination spider-cells and fat-granules are found in considerable numbers not only in the gray cortex, but also in the white matter.

This observation determines us in maintaining with Kast the old designation, "cerebral palsy of children," a name by which no definite pathological change is implied, and in abandoning the term polio-encephalitis, to which the pathological changes do certainly not always correspond, especially as the

latter name has already been proposed by Wernicke for the disease of the gray matter around the third and fourth ventricles.

Symptoms.—The symptoms of this disease-group differ according as the pathological process is confined to one hemisphere or attacks both.

In the former case the symptoms and the course are so characteristic that a correct diagnosis can almost always be made during life. The disease usually sets in brusquely, the symptoms are violent and can not be overlooked. The child is seized with a high fever; soon, sometimes only a few hours later, twitchings—at first only in one extremity, later in the whole side—appear; at times the whole body may be convulsed; this may last, with but slight interruptions, for from one to three or even four days, and be accompanied by persistently high temperature. The symptoms now abate, the convulsions become less frequent, but after their disappearance the child is found to have lost the use of the limbs of one side —hemiplegia infantilis spastica (Benedikt). If an early and careful examination be made, a moderate facial paralysis is noted, the condition of the extremities being very nearly the same as has been described on page 226. The condition of the facial nerve in this affection has recently been studied by W. Koenig (Deutsche med. Wochenschr., 1893, 42). Here, as in the common cerebral hemiplegia, the arm is pressed against the thorax, the forearm flexed at right angles with the upper arm, the hand flexed and adducted, the fingers bent. The leg is slightly flexed at the knee joint, the foot extended; not uncommonly the big toe is in marked dorsal flexion. The sensibility is, as a rule, not much altered. After several weeks the little patient regains enough power to perform the coarser movements with the leg, while for a considerably longer time the arm does not take part in the improvement. If the child was able to walk before the onset of the disease, it will generally regain this faculty after a time, but its gait will always be halting.

The further course of the disease is not the same in all cases, and it has been our experience that it differs according as the initial convulsions continue or cease. This, therefore, is an important point to consider in the prognosis for the relative recovery. It will also decide the question whether the child, while bodily more or less a cripple, is in addition to be men-

tally defective and totally useless to the community. What conditions determine the continuance of the convulsions, whether this is influenced more by the nature of the lesions or more by their seat, we are unable to say.

As a rule, the attacks, even if they should have a tendency to continue, do not recur for months, for one, two, or even four years, after the acute period of the disease has passed off. Then, however, they may return on any provocation—after a fright, maltreatment, sometimes during the second dentition—at first at long intervals of months, then more frequently. At first they may be slight and of short duration, then more severe, until finally they resemble in every point the classical attacks of epilepsy—in other words, the hemiplegic or hemiparetic patient has now become an epileptic. As has been stated, the influence which these attacks have upon the mental development of the child is very detrimental. Much more often than is the case in idiopathic epilepsy does the patient become weak-minded. The condition of speech found in this disease is interesting. If the patient had fully acquired speech previous to the attack, it is only affected if the lesion is on the left side of the brain, in which case the symptoms do not differ from those which we have described under left-sided cerebral hæmorrhage. If, on the other hand, the patient has not yet learned to speak, he will, in case the fits continue, either not learn at all or only very imperfectly, and his talk will, even if his mind is only slightly impaired, be quite unintelligible; but often the attacks do not recur, so that the mental development progresses normally. In such cases speech likewise reaches a gratifying degree of development even if it had not yet been fully acquired or had been lost. The healthy hemisphere takes on vicariously the work of the injured one (cf. page 182, remarks on aphasia of children).

Independently of the epileptiform attacks, there may occur changes in the extremities which are in a way analogous to those described above. An especially characteristic symptom is the pronounced spastic condition which manifests itself in an increase of the reflexes, rigidity and spasm of the muscles—hemiplegia infantilis spastica. This rigidity is especially well marked in the muscles of the hand and the calves, and leads, preferably in the former location, to contractures, which, however, differ from others, inasmuch as they cease during rest and sleep and only appear on voluntary motion. Benedikt, above others, has pointed out that at one time one, at another time another, group of muscles may be affected; that, e. g., in walking, the foot may be held normally, while again in the same foot we may see a talipes calcaneus, or at another time a talipes equinus. Similar conditions are observed in the hands; thus the fingers, which appear to be in a state of immobile flexor contraction, may at other times present a remarkable degree of mobility. We shall shortly discuss carefully the entirely involuntary movements of the affected hand which are noted in the course of this disease. E. Remak has shown that such a spastic paralysis with contracture may lead to a luxation; in the case which he reports a retroglenoid subacromion luxation developed (Berlin. klin. Wochenschr., 1893, 52).

In almost all cases of infantile cerebral paralysis an arrest in development or growth becomes apparent in the affected extremities. This may be only insignificant, so as to be hardly appreciable. On the other hand, the limbs may in all their dimensions be considerably smaller than the corresponding ones of the sound side. Occasionally the whole half of the body, trunk and head as well, share in this arrest, and we have what is called a general hemiatrophy. Borgherini has observed this to occur a few days after the onset of the disease (Deutsch. Arch. f. klin. Med., xl, 5, 6).

The following illustrations of cases from my clinic represent different types of the cerebral paralyses of children:

Figs. 75 and 76: Hemiatrophy of the whole left side of the body. Epileptiform attacks. Dementia.

Figs. 77 and 78: Hemiatrophy of the whole left side of the body. Cause: Traumatism. No fits. Intelligence normal.

Fig. 79: Atrophy of the left upper and lower extremity (resection of the knee joint). Epileptiform attacks, with a moderate degree of dementia.

Figs. 81 to 84: Atrophy of the paralyzed side, very slight, but perceptible. All three patients suffer from epileptiform attacks and are demented. All three present contractures on the affected side, either in the wrist (Fig. 81, and also Fig. 75) or in the ankle joint (Figs. 83 and 84).

All these eight cases, which came under my observation, depended upon disease of the right hemisphere. Whether this side is altogether more frequently attacked, and, if so, how the fact is to be explained, I dare not at present decide.

If both hemispheres are affected, the symptoms are very different, and the diagnosis is much more difficult. Freud has classified all these affections as instances of cerebral diplegia, a term which may be accepted without reserve, as it is purely

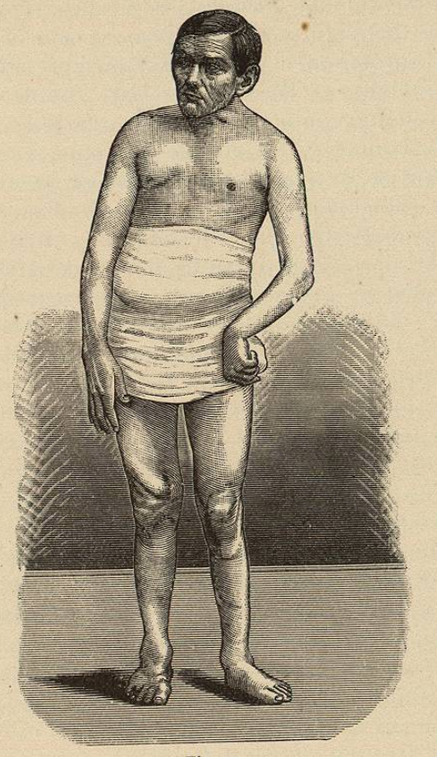

Fig. 75.

descriptive, and is noncommittal so far as regards the pathological process. He divides the cases into four groups: 1. The general cerebral spasticity first described by Little and called by the English authors Little's disease. 2. The paraplegic spasticity (so-called spastic paralysis). 3. The bilateral hemiplegia. 4. The bilateral chorea and athetosis. The latter will be referred to again (page 284). Little's disease and spastic paralysis are frequently congenital; in the latter affection especially predisposing and hereditary influences play some part, and several members in the same family may develop the disease (Newmark,

A Contribution to the Study of the Family Form of Spastic Paraplegia, Amer. Journ. Med. Sci., April, 1893). (Cf. Fig. 80.)

Diagnosis.—The diagnosis of the unilateral affection is, as a rule, easy, as the acute onset with the consequent hemiplegia

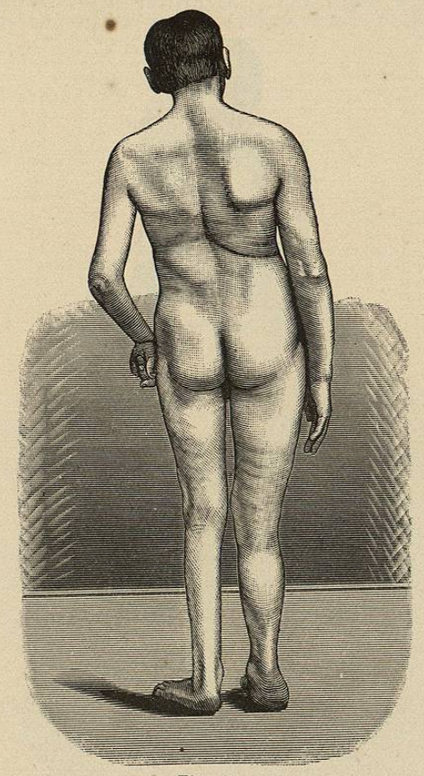

Fig. 76.

The patient, who is now thirty-six years old, was taken ill in early childhood with an acute violent fever and convulsions; the latter lasted for several days, but after that disappeared. From that time the left side did not develop as well as the right, so that now the left upper extremity, which can be moved, with difficulty only, in the shoulder and elbow joints, is seven centimetres shorter than the right, while the left lower extremity is three centimetres shorter than its fellow. The whole half of the body has shared in the atrophy, which is also well marked in the nates. The circumference of the left upper arm measures four centimetres and a half, that of the left leg three centimetres less than that of the corresponding extremity. When the patient was fourteen years old the convulsions reappeared, and he has still one or two epileptiform attacks a week. He is quite demented.

is characteristic enough; but if the patient come from a phthisical family and is himself tuberculous, some doubt may arise. We may have a case of tuberculosis of the brain to deal with, which sometimes resembles in its onset the cerebral palsy of children. High fever and convulsions are not absent, and se-

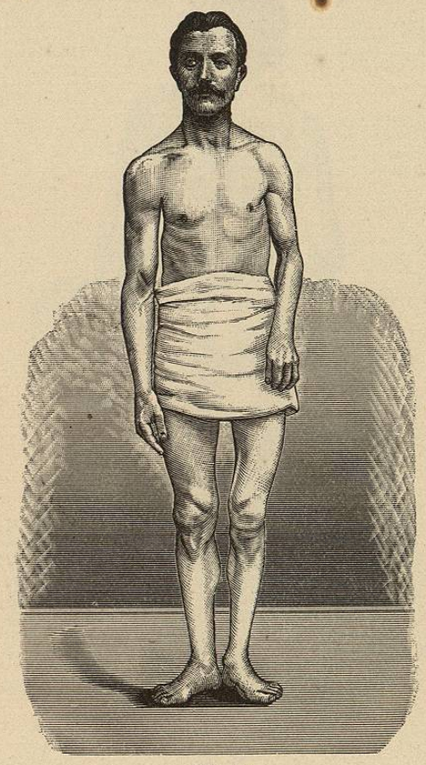

Fig. 77.

vere motor disturbances occur also. The fact, however, that in cerebral tuberculosis generally the base of the brain with its nerves, especially the oculo-motor and abducens, are implicated, and, further, that it runs a rapid and fatal course, will enable us to make a correct diagnosis.

Spinal and cerebral infantile paralyses can not be confounded with each other if we keep in mind that, in the latter, one whole side of the body is affected; that the muscles are rigid, the reflexes increased; that convulsions occur not only

at the onset, but also in the further course of the disease; that the mind becomes impaired, etc. In the spinal form, either one limb alone—arm or leg—is affected or both arms or both legs, and the reflexes in the paralyzed extremities are lost—signs enough to enable us to differentiate between the two

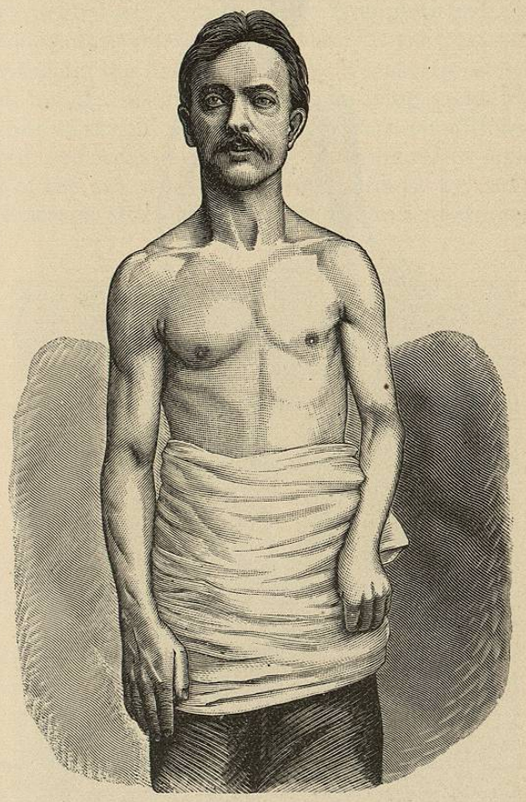

Fig. 78.

The patient, now thirty-four years old, fell, at the age of three, from a high foot-stool and injured the right side of his head. He lost a considerable amount of blood, and was unconscious for quite a long time. Six months after the injury the atrophy of the left side of the body became apparent, first in the upper, then in the lower extremity. With the exception of this atrophy, which has now taken in the whole half of the body, including the thorax (compare the left with the right mamma), the patient is perfectly healthy. He has never had epileptiform attacks, there are no hemiathetoid movements, and no psychical alterations whatever.