ON CEREBRAL DIPLEGIAS IN CHILDHOOD
(LITTLE'S DISEASE).

R. Massalonga (Wiener medicinische Blatter, 1898, xxi, 7, 12) divides all forms of cerebral diplegias in children into the following groups:

(1) General typical muscular rigidity; general muscular rigidity, slight form, in which the symptoms are less prominent.

(2) General muscular rigidity, in which the symptoms are still less marked, where the muscular rigidity, so to speak, is in a latent condition, as it only appears in certain movements.

(3) Transition forms between general muscular rigidity and paraplegic muscular rigidity.

(4) Absence of paraplegic muscular rigidity.

(5) Paraplegic muscular rigidity, accompanied by hemiparesis.

(6) General muscular rigidity, accompanied by hemiparesis.

(7) General muscular rigidity, with bilateral spastic hemiplegia.

(8) Bilateral athetosis.

(9) General spastic chorea.

(10) Spastic muscular rigidity, more or less extensive, combined with chorea or athetosis.

(11) Chorea and athetosis combined.

The name of Little's disease is proposed by Massalonga for this group, in place of the term "cerebral diplegias of childhood" advocated by Freud, as the muscular rigidity preponderates in the great majority of cases, while paralysis is only very exceptionally present.

As against the etiology of the disease described by Little, Massalonga from his own and the experience of others emphasizes:

(a) That the cerebral diplegias of childhood may be congenital as well as acquired.

(b) That they may be produced by difficult labor and asphyxia of the new-born, but in all probability are caused by febrile diseases of an infectious nature.

(c) That an irregular pregnancy, disturbed by nervous affections, seems to predispose to cerebral diplegias, even though labor proceeds normally.

(d) That no connection can be found between the etiological factor and the clinical form of the disease.

The primary condition, according to Massalonga, is a meningoencephalitis brought about by an extra- or intrauterine affection (toxemia); the other findings at the post-mortem, hemorrhages, cysts, sclerosis and atrophies of the brain, porencephaly, pachymeningitis, etc., are only phases of evolution or degeneration of the primary meningo-cerebral condition, no matter whether they are caused by primary lesions of the nervous elements, or by affections of the vessels which could be ascribed to the same cause, namely, infection or intoxication. The spinal cord lesion is always secondary, even in cases where distinct cerebral symptoms could not be demonstrated clinically. That this secondary degeneration of the pyramidal tracts is not found in all sections is due to the late development of these tracts which frequently only become fully developed.
in the second or third year of life. Van Gehuchten's view that spastic muscular rigidity in prematurely born infants is dependent on an imperfect development of the spinal cord, and the total absence of the pyramidal tracts, being therefore of spinal origin, is met by Massalonga with the assertion that the imperfect development of the pyramidal tracts was probably the most important cause of cerebral diplegias; that it was not of a primary nature, but the result of anatomical changes of an inflammatory nature, of a toxic-infectious origin, of the cerebral membranes. Massalonga reports seven cases observed by him personally:

Case I.—General spastic rigidity. A child 6 years of age. Labor and birth were normal. Three months ago the child began to loose flesh, vomited, and had diarrhea. A few months later the mother noticed that the extremities were rigid and difficult to move; she observed also rolling of the eyeballs, and little intelligence. Condition, three years later: normal mind, voluminous skull, nystagmus horizontalis, and internal strabismus of the left eye. Speech consists in inarticulate sounds. Ataxia of the upper extremities, and spasm of the upper and lower extremities are present. Bilateral pes varo-equinus, atactic-spastic gait, increased reflexes of the upper and especially of the lower extremities. Mind and sensory organs normal. Very slight improvement during stay in the hospital.

Case II.—Paraplegic rigidity and spastic hemiparesis of the left side. Patient, 17 years of age. Mother is hysterical; pregnancy and birth were normal. Patient was well until 13 months of age. A high fever then suddenly came on, lasting several days, with epileptiform attacks which recur for years, decreasing however in intensity and frequency. The patient only learned to walk and talk at 5 years of age; the lower extremities always maintained a position of flexion and abduction, the feet were in a varo-equinus position, the right arm was also flexed at the elbow joint. The right extremities suffered most. Irritable, angry temperament. Speech improved in the course of years, always remained imperfect however. Memory good, writing undecipherable. Present condition: poorly developed, dolichocephalic skull, ears standing out, mouth half closed, the right side of the face is less developed than the left; gait skipping, the legs touching each other and the right foot being dragged; the right arm is abducted, the arm and hand in a position of flexion; speech difficult, with a guttural sound; intelligence good; slightly myopic; there is a general increase of all the tendon reflexes.

Case III.—Bilateral athetosis and general muscular rigidity. Child, 4 years old. Father addicted to liquor. While pregnant the mother met a young person suffering with athetosis and was deeply impressed; she was subject to great excitement on account of her husband's drinking habit. Labor was difficult, the child was born asphyxiated. In the early part of the second week the mother noticed a condition of muscular rigidity of the neck, of the upper, and later also of the lower extremities of the infant; also slow but violent movements of the extremities (which ceased during sleep), particularly of the fingers and toes. Present condition: the skull is somewhat depressed in the left occipital region; mouth is half closed and frequently makes sucking motions in which the tongue also participates. The patient has at no time emitted articulate sounds.
Dysphagia accompanied by coughing paroxisms and suffocative symptoms is present. The arms are adducted, the forearms flexed, athetosis of the fingers is present; the legs are adducted and slightly extended. When excited the abnormal positions as well as the athetosis of the extremities are markedly increased, so also the rigidity of the muscles of the limbs and neck which is nearly always present. During sleep all these symptoms cease. The child is unable to stand alone; when assisted he walks with a spastic gait, the head and shoulders pointing backward. The reflexes of the soles of the feet are prompt, the others cannot be determined on account of the rigidity. Two months after admission the patient died of a broncho-pneumonia. Beside the latter, the autopsy showed the presence of enlargement of the heart, hyperemia and hypertrophy of the liver, hypertrophy of the spleen, hyperemia of the cerebral membranes, the pia being adherent in some of the convolutions, the convolutions flattened; the frontal and prerolandic convolutions discolored to a whitish-gray, somewhat firmer than normal, wrinkled, their white matter of a gray tint, slight hyperemia of the spinal membranes, the subarachnoid space containing quite a quantity of exudation. Microscopic findings: the nerve fibrillae of the pyramidal tracts very thin, and only distinctly degenerated in the cervical portion.

Case IV.—Bilateral athetosis and paraplegic muscular rigidity. Patient, 38 years of age. Mother suffered from hemiplegia for a long time; pregnancy normal, labor tedious and severe. The child remained well up to 10 months of age. At this time a high fever, restlessness, convulsions of the body and face made their appearance. These soon ceased, and were followed by paraplegia and athetosis of the upper and to a lighter degree also of the lower extremities. Intelligence is normal, speech inarticulate; irritable, passionate temperament. Later addicted to masturbation. Present condition: irregular movements of the muscles of the face, mouth and tongue; the eyeballs also are in continual motion, the head is carried forward, to the side or backward. Athetosis of the upper extremities and the toes is present. Legs in a position of adduction and flexion. Walking is impossible. A few muscles of the upper extremities are hypertrophic. Tendon reflexes increased.

Case V.—Absence of bilateral athetosis. Child, 6 years old, no hereditary taint. Pregnancy normal, labor tedious, child not born asphyxiated. After 10 days the child began to emaciate, the skin took on a yellowish color; sleep became restless, there is continued crying. A few days later irregular movements, at first of the arms and hands, then of the lower extremities occurred, accompanied by a slight degree of muscular rigidity. In the course of a few months this increased, extending to the muscles of the mouth, eyes and tongue. Intelligence slight, speech is confined to a few words. Present condition: beside the symptoms already mentioned, there is an apathetic condition, opisthotonus, the arms are adducted, forearms and hands flexed, the legs adducted, varo-equinus, gait difficult, even when supported, intelligence good.

Case VI.—Bilateral chorea-athetosis. The mother of the patient suffered with hysterical seizures during pregnancy. The father is a
drunkard and suffers from syphilis. Labor normal, the child remained well up to the fourth year of age. Then suddenly fever and delirium appeared. Since then he was unable to walk. The lower extremities are flexed and adducted. Speech is lisping; the child is apathetic, presents a motor weakness of the upper extremities. This is followed by athetoid and choreic movements of all extremities, the thorax, the neck and face. There is no improvement with increasing age. During the day the patient would lie on the floor, crawling like a snake, throwing about the extremities, flexing or extending the neck, with eyes rolling and distorting the mouth. All the muscles are spastically contracted, but moderately so. The tendon reflex is increased.

Case VII.—Congenital spastic chorea. Mother healthy; father suffers from articular rheumatism; pregnancy and labor normal. Seven days after birth fever and icterus made their appearance. At the same time choreic movements of the head and extremities occurred which increased later in force and frequency. The patient learned to walk and stand comparatively late. The intelligence is normally developed, speech is rudimentary. The choreic movements are much more marked in the upper extremities in the lower, as a strong muscular spasm is present here. At times there are also involuntary movements of the eyeballs and the mouth. Tendon reflex increased.

A RARE CASE OF TERATOMA COLLI.

H. Munker (Ungarische Medicinische Presse, 1898, iii, 44). The patient was a child, 2½ months of age, in whom the rapidly-growing tumor obstructed deglutition, so that an operation was unavoidable. The child died three days later. The soft fluctuating tumor, consisting of three protuberances, was situated on the right side of the neck, and a few isolated firm spots could be felt on it; the skin covering it was tense, traversed by large veins and small arterial vessels. It reached upward on the face as far as the right arcus zygomaticus, and also to the anterior border of the left sterno-cleido-mastoideus, downward to the third rib, and extended to the mastoid process, at which location it presented two glands the size of a pea. The skin of the upper lobe was continued to the integument covering the tumor. The voice was somewhat hoarse, respiration free.

Section of the extirpated tumor presented the following peculiar picture: Various large cavities of different shapes are found imbedded in the basic tissue in an irregular manner, partly side by side, partly in groups. These cysts are of the size of lemons to that of the head of a pin, some round, some in the form of a slit, others present diverticuli, etc.; the largest cysts have the thinnest walls, while the smaller ones as a rule have thick walls.

The histological examination revealed the following: Embryonic tissue, embryonic cerebral matter, fat, cartilage, muscular fibers arranged cross-wise in certain localities, cavities which looked like cysts, and others which resembled glandular tissue.