THE
CHICAGO MEDICAL
RECORDER

AUGUST, 1894.

Original Articles.

CEREBRAL PALSY OF CHILDREN.

By ARCHIBALD CHURCH, M. D.

PROFESSOR OF MENTAL DISEASES, CHICAGO MEDICAL COLLEGE; PROFESSOR OF NEUROLOGY, CHICAGO POLYCLINIC; NEUROLOGIST TO ST. LUKE'S AND COOK COUNTY HOSPITALS.

It is only recently that text-books have in any systematic manner even mentioned the paralysis occurring in early life due to intracranial lesions, yet the great frequency of this serious misfortune and the fact that of paralyzed children about one-half as many suffer from cerebral disease as those who are crippled by spinal lesions, renders the importance of the subject at once self-evident. Of late years, especially since 1890, this condition has met with the attention of many observers and a mass of literature has resulted which is of the greatest importance. It is only necessary to mention the works of Drs. Sarah McNutt, Osler, Sachs, Peterson, Gibney, Sinkler and Smith, in this country; Cotard, Cruveilhier, Brechet and Marie in France; Kundrat, Struempel, Benedict, Bernhardt in Germany; Gowers, Ross, Abercrombie and others in England.

In every neurological clinic patients are being presented almost daily where the malady has escaped medical elucidation at the hands of family practitioners, who have been satisfied to class the cases as spinal paralysis, idiocy, or congenital defect, without seeking a more accurate designation, and attributing the condition to teething, infectious disease or some injury. Yet the characteristics of the affection are so marked that having been once pointed out their recognition is not a matter of much difficulty.

These children present three types of disability, dependent

upon the location and extent of the brain injury. They are hemiplegic, diaplegic and paraplegic, as one side, both sides or only the lower extremities are particularly affected.

In the hemiplegic type, which is perhaps the most common, the conditions of the paralyzed side are analagous to those which appear in the hemiplegic adult, with the modifications that might be expected from disease occurring in the brain of the developing child. Rigidity, retarded development, contractures, spasticity and increased reflexes are the prominent symptoms. As with

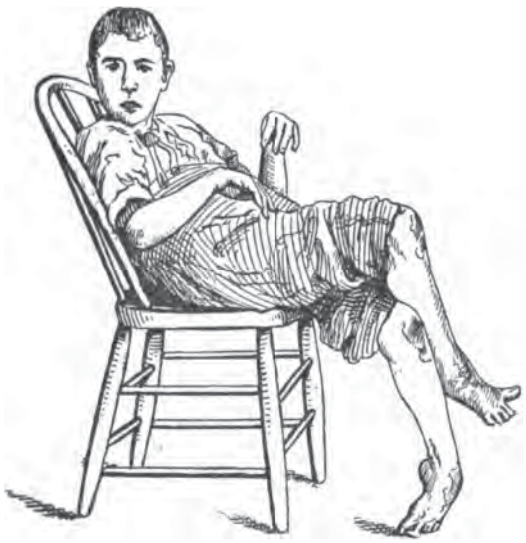
HEMIPLEGIC SINCE BIRTH.

Showing athetosis in right hand; overaction in right face and shoulder.

adults, the paralyzed members are sometimes the seat of associated movements; if the sound limbs be automatically or violently thrown into activity, especially in gestures of an emotional character, the paralyzed limbs are inclined to duplicate the movements, and in addition they are frequently the seat of spontaneous choreic or athetoid action, which in some instances persists during sleep, and which is as a rule intensified by volitional activity and emotional disturbance. In these cases of athetoid extremities, owing to the muscular activity, a part of the condition of athetosis, the muscles are frequently well developed in size and consistency. Indeed such limbs may be larger and firmer than those on the sound side, but the lack of synergism in their functions

is attended by great weakness, so that these individuals will register upon the dynometer but a small fraction of the pressure that would be normal, and the joints, as a rule, are capable of wider range and latitude of motion owing to the relaxed ligaments and plastic condition of the joint surfaces in childhood, which conduces to their deformation under the perverted muscular control.

In the diplegic and paraplegic cases the rigidity in the lower extremities is usually complicated by more or less adductor spasm, interfering greatly with walking and producing the characteristic cross-legged progression or position. The retraction of the posterior muscles of the legs overbalancing the anterior and peroneal



CONGENITAL DIPLEGIA.

Showing athetosis in all four extremities; adductor spasm; equino varus, mental defect.

groups gives rise to the equine position of the foot, usually associated with a tendency to varus, in many cases of high degree.

Where the limbs are not the seat of athetoid movements a position analagous to that of hemiplegia in adults but with a lack of development in the muscles and bones is the rule, so that when these affected individuals reach mature years there is a great discrepancy in the size and length of the limbs upon two sides of the body, and sometimes asymetry of the face.

If, previous to the onset of the paralysis, the child has learned

to talk, speech will be either abolished or greatly interfered with for a time, irrespective of the side of the brain involved, but subsequently in nearly every instance, if the intelligence is not too much impaired, articulate language will be more or less completely restored. The mental development of the child is also arrested, and depending upon the extent and character of the lesion, idiocy, imbecility, feeble-mindedness, or only a slight defect, may follow.

The organic process lays these individuals liable to the onset of epilepsy. About one-half of them that outlive the shock of the original brain disease sooner or later develop epileptoid attacks,

PARAPLEGIA FROM BIRTH.

Showing spastic lower extremities, adduction and cross-leg walk.

which, as might be expected, are in many instances of a marked Jacksonian character with a tendency to become more and more generalized, and in some instances are of a general epileptic character from the first. Under this protracted epileptic state dementia is likely to ensue. The outlook for the mental strength of the child so attacked is a gloomy one at best.

As in the adult, the tendency is for the lower to recover more rapidly than the upper extremity, and the face, if affected on one side only, will in conditions of repose regain something of the appearance of symmetry, but the overactivity which marks the

hemiplegic side and is associated with the spasticity will manifest itself during emotional expression, such as laughing or crying, even where all the other symptoms have practically disappeared.

These cases fall into three groups, as has been clearly pointed out by various writers: First, those dependent upon conditions present before birth; second, to the accidents of parturition; third, to brain lesions acquired subsequent to birth. Sachs and Peterson make the following classification: First, palsies of intrauterine onset, marked by large cerebral defects, hemorrhages and cerebral agenisis; second, paralysis occurring during labor, due to meningeal hemorrhage; third, paralysis acquired after birth from hemorrhage, embolism, thrombosis, chronic meningitis, hydrocephalus and perhaps the primary encephalitis of Struempel.

The developmental defects of the first class frequently result in cyst-like cavities constituting true porencephaly, in some extreme cases a large portion of the hemisphere being wanting. Fetal cerebral hemorrhages may follow injury to mother and child, or marantic conditions affecting the fetus. Meningeal hemorrhages occurring during labor are very much more frequent than has usually been supposed. The instructive report of Herbert Spencer regarding the conditions found in stillborn children shows that out of 130 such instances there were 53 cases of hemorrhage into the cerebral pia and arachnoid, one case of intracerebral hemorrhage and 30 cases of hemorrhage into the meninges and substance of the cord. It is evident that if the tendency to meningeal hemorrhage be so great as here indicated in the stillborn, many children in whom the hemorrhage is of less severity may escape immediate death to present subsequently the palsy under consideration. Peterson very clearly points out in a recent article that the spinal meninges being so frequently involved, some of the paraplegic cases may be due, not to cerebral conditions, but result from the interference with the conducting elments of the cord itself, especially some of those cases that are marked by a high grade of mental intelligence, and present no symptoms above the waist.

The relation between protracted labor and cerebral palsy is an intimate one, a majority of such cases being of primiparous birth and often assisted into the world by forceps. Without trying to overlook the danger forceps may do the brain, the injuries, especially in the way of meningeal hemorrhage, from the compression of protracted labor, are of much more significance and indicate early instrumental interference rather than the opposite course if

the element of severe compression is present and a skillful use of forceps is assured. On the other hand in the writer's experience some of the cases have been due to precipitate birth where the pressure upon the head has been suddenly and forcibly applied by a vigorous uterus.

The post-natal acquired palsies of a cerebral character are due in a large proportion of cases to cortical or subdural hemorrhage, embolism causing but a few, and thrombosis being responsible for even a less number. Chronic meningitis and hydrocephalus in these cases are usually mixed processes, the nature and origin of which are difficult to elucidate on the post-mortem table where terminal conditions have served to confuse the situation. Struempel believes that many of these cases are due to a primary encephalitis, a process analagous to the spinal disease, involving the anterior horns of gray matter in anterior poliomyelitis, and he designates it polioencephalitis. In substantiation of his views he points to the occasional febrile onset, the acute manifestations of the disease and the subsequent diffuse sclerosis of the cortex. Here, however, as elsewhere, post-mortem opportunities have been rare, as these cases only come after some years to the investigation of the anatomist, and the theory of Struempel is not supported by early anatomical evidence. Osler, however, leans to the idea that there is some foundation for the hypothesis.

The terminal conditions from hemorrhage, embolism and thrombosis are somewhat similar, and result in about one-half the cases, according to Sachs and Peterson, in cysts, atrophy and sclerosis, with evidence of hemorrhage in about one-quarter of this number. The cystic condition is sometimes confused with true porencephalus which is a developmental defect. Embolism and thrombosis are so rare that except in the presence of constitutional conditions strongly predisposing to such processes, they may usually be left out of the argument. In the paralysis following acute rheumatism, endocarditis, scarlet fever, diphtheria, or in the subjects of hereditary syphilis, they should be taken into prominent consideration.

The diagnosis of this condition can usually be made out with readiness. The congenital cases are likely to present the greatest difficulty as very little is expected of an infant up to the age of nine months or a year. A hemiplegic state, however, would be soon made out. In the conditions acquired after birth a previously active child is suddenly taken with convulsions and these convulsions being followed by more or less paralysis such investigation would

follow as to leave little doubt regarding the trouble in hand. Later on, when such cases are brought for examination the rigidity in the extremities, the increased reflexes, the spasticity, frequently the morbid movements, the retarded development and the preserved electrical responses should distinguish the paralysis from that of a spinal or peripheral character in which true atrophy would be found, with lost reflexes and the reaction of degeneration or the refusal of the muscles to respond to all forms of electrical stimulation.

In some instances the cranium does not develop as might be expected, and microcephalus results. The question may arise whether or not the mental retardation may be due to the microcephaly or the microcephaly only a part of the maldevelopment. Here the history of early closure of the fontanelles, or the fact that the fontanelles were obliterated at birth, may be a guiding factor. Many cases of early synostosis of the cranial bones present spastic symptoms, increased reflexes and mental retardation, without showing post-mortem any evidence of cerebral disease, except in very late cases some cortical atrophy or slight sclerosis.

The prognosis is usually good as to life, but the life of such individuals is a short one, very few such cases reaching the age of thirty.

As regards motor power, it may usually be promised in hemiplegics that the face will practically recover and the lower extremities serve the purpose of locomotion, but for the upper extremities the prognosis must be guarded. Speech will usually be regained, and after a few months the prospects as to mental development may be estimated with some certainty.

It has been stated that about one-third of these cases become subsequently epileptic, and the development of epilepsy with a tendency to epileptic dementia must be carefully borne in mind. It may usually be stated with positiveness that the individual will be defective, that the muscular power in the involved limbs will be lessened and that mentally, in all probability, there will be some defect. In the rare cases where the condition is one of true paraplegia the development of a higher grade of mentality is possible.

The treatment of these unfortunate cases is varied and it may not be amiss in the first place to urge the skillful use of forceps in protracted labor attended by great compression to prevent meningeal hemorrhages, so frequently the result of that process. In paralysis acquired after birth the treatment at first is very similar to that instituted with adults: quiet, purgation, cold to the head if

hemorrhage is diagnosed, and the withdrawal of all excitement. The later treatment consists in general measures, such as massage, and electricity to the extremities, by which much can be done to prevent distortions and to maintain the nourishment of the muscles, thereby assisting in the vicarious control of the members by the uninjured side of the brain in hemiplegics, and to maintain for the patient the greatest possible muscular strength under all circumstances. The mental state can be greatly modified for the better by careful instruction and in some institutions for the feeble-minded it is simply wonderful what is accomplished by conscientious, scientific and assiduous effort in this direction.

The presence of epilepsy, constituting as it does a grave element both as to the present health of the individual and his future prospects, will call for solicitous attention. These patients as a rule do not stand the bromides well and they are particularly prone under prolonged bromide exhibition to develop the depressed features that go with the continuous use of that drug. By carefully grading it to their tolerance and seeking with it merely to hold the epileptic tendency in abeyance, sometimes much good can be accomplished, but the question will often arise here, as in general epilepsy, whether it is better to stupify the patient constantly with bromide, perhaps thereby eliminating fits, or to let him have a fair degree of mental and physical activity without bromides, and an occasional epileptic seizure.

The surgical treatment consists in attention to the contractured extremities, and is principally orthopedic. By braces and apparatus, associated with tenotomies and sometimes with a correction of joint surfaces, very frequently helpless patients may be put upon their feet; but athetoid limbs are rebellious to all forms of mechanical appliances. Many of the cases bring forward the problems of brain surgery, many of them present Jacksonian fits, but unfortunately, the early favorable prospects of operation for Jacksonian epilepsy have not been fulfilled by wider experience, and it is probable that if a small portion of the widely sclerosed cortex be removed, thereby destroying the initial symptoms of seizure in a given case, the scar tissue laid down and the epileptic tendency elsewhere present in the brain will lead to a recurrence of the fits, only slightly modified in character. Cysts, to be sure, may be evacuated, but it is doubtful that very much good will be accomplished. Porencephalic cases are not to be benefited by increasing the porencephaly. There are probably some of these cases that can be greatly benefited by cerebral surgery. Some

cases of athetoid spasm may be cured as far as the spasm is concerned by the ablation of the associated cortex. In New York, in one case where the spasm was so severe as to cause endless discomfort to the patient, an amputation was done, with great relief, and the operator expresses himself as justified. A similar case appeared in my practice, where an arm was so seriously convulsed that the entire time of the patient was occupied in holding with the disengaged hand the unruly member, which as soon as released would fly behind her back, or into her face or behind the neck, and resist very vigorous efforts to dislodge it. The cortex was exposed, the arm centers verified by the electric current and this portion of the cortex removed. Relaxation and paralysis in the athetoid member resulted, thereby liberating and making available for use the sound hand of the patient, and subsequently, slight voluntary control returned in the formerly athetoid but now partly paralyzed extremity.

In cases of microcephaly where there is a distinct history of closed fontanelles at birth and where the general strength and health are good, the operation of linear craniotomy still promises something for improvement, but the linear exsection should be sufficiently extensive to give the cranium an opportunity to expand, which is not accomplished by the incisions laid down by early operators. It is also a fact that these defective individuals do not bear surgical interference with the brain at all well, and the mortality from such operations is consequently high.

The management of these cases means years of patience on the part of friends and physicians, but in many instances much effort will be rewarded by a fair degree of improvement.

PULLMAN BUILDING.