PROGRESS IN PEDIATRICS

REVIEW OF LITERATURE FOR 1920 ON NEUROLOGY OF CHILDREN *

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CHICAGO

BLOOD VESSELS

The tendency toward hemorrhage in the new-born, according to J. A. Foote¹ is frequently first manifested as intracranial hemorrhage and it is important to recognize that this may be and often is a local manifestation of a general condition. The early resort to lumbar puncture and the hypodermic administration of substances that tend to increase blood coagulability are rationally indicated in every case of suspected intracranial hemorrhage of the new-born, even prior to operation.

It was found by F. C. Rodda² that 50 per cent. of infants dying during delivery or during the first few days following, died of hemorrhage; apart from gross trauma other factors existed in the production of this hemorrhage and not always was the intracranial hemorrhage caused by obstetric procedures, many occurring after normal labor and in children with hemorrhagic tendencies. For this reason, blood coagulation tests are stressed.

The cause of intracranial hemorrhages are divided by J. B. Sidbury³ into two general heads. First, spontaneous, or hemorrhage due to a general condition, as hemorrhage of the new-born; and, second, traumatism. Under the latter head the most common causes are (1) prolonged, tedious or hard labor, with or without instruments; (2) precipitate labor with injury to the child's head; (3) injudicious use of pituitary extract; (4) breech extraction of the after-coming head; (5) premature babies, who usually have very fragile blood vessels, which are not strong enough to undergo the amount of pressure necessary even in a normal delivery.

Among the symptoms described by the author, the following are mentioned as signs to bear in mind: Convulsions or twitching of one or more extremities, bulging fontanel (not a constant sign), nystagmus,

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1. Am. J. Dis. Child. **20**:18 (July) 1920.

2. J. A. M. A. **75**:452 (Aug. 14) 1920.

3. Arch. Pediat. **37**:545, (Sept.) 1920.

strabismus, which is more or less constant, increased reflexes, which may be more marked on one side. Stiff neck and Kernig may be present if the irritation is confined more to the base of the brain. The pulse is strong, full and at times slow. The respirations are irregular and at times simulate the Cheyne-Stokes phenomenon.

The author advises repeated daily lumbar punctures until the spinal fluid is clear of blood when the intracranial pressure is found to be increased on measurement. If this procedure fails to remove such signs as papillitis or twitching in any group of muscles, a decompression operation should be considered.

Cranial sinus thrombosis in children is described by S. Oppenheimer⁴ who thinks that the symptomatology is similar to that of adults. Two forms of thrombosis are described: (1) Primary or marasmic; and (2) secondary or infective, sometimes termed inflammatory. Marasmic thrombosis occurs less frequently than the infective and is most frequently located in the longitudinal, rarely in the lateral and still more rarely in the cavernous sinuses. It is met with in the young most frequently during the first two years of life and more especially during the first six months of infancy. Exhausting diarrhea is one of its most potent causes, although acute and chronic pulmonary affections play an important etiologic part. The diagnosis of primary sinus thrombosis in children is difficult and is seldom made during life. In view of the impossibility of a definite diagnosis in most cases, the treatment is wholly symptomatic.

Secondary or infective sinus thrombosis is much more frequent and follows extension of inflammation from parts contiguous to the sinus wall. It is the term usually applied to thrombosis which arises from invasion of the sinuses by pathogenic microorganisms. In children, infective sinus thrombosis may follow skull fractures, scalp wounds, anthrax of the lip, mouth, nasal and orbital cavities, erysipelas of the face and forehead, furunculosis of the lips, face or neck. By far the most frequent cause is suppurative middle ear disease. It may also occur as a complication of a diphtheritic infection of the middle ear or of scarlet fever, but more often from measles and influenza.

The most important general symptom of lateral sinus thrombosis is fever which rises excessively high in young children. In older children there may be headache, pains in the occipital region and tenderness on palpation along the jugular vein, due to lymph node enlargement. Dilatation of the cervical veins and the possible hardening of the internal jugular vein, with or without edema behind the mastoid process, are signs of diagnostic significance.

4. Arch. Pediat. **37**:65 (Feb.) 1920.

In establishing the diagnosis of sinus thrombosis pneumonia, typhoid fever, acute endocarditis, malaria and certain cases of scarlatinal infection must be excluded. The author emphasizes the importance of early diagnosis because operative procedures are attended by the cure of a great many cases which otherwise would terminate fatally.

MENINGES

An early diagnostic sign in basilar meningitis is described by G. Gingold⁵ as follows: By flexing the head on the chest either a bilateral or a unilateral strabismus develops which will last as long as the head is kept in a flexed position, and disappears as soon as the head is relaxed. In many cases the strabismus is accompanied by retraction of the upper eyelids. In some cases he noticed a contraction of the pupils. This "reflex" strabismus was present in almost every case that came under the author's observation.

The diagnosis and treatment of hydrocephalus resulting from strictures of the aqueduct of Sylvius is described by W. E. Dandy.⁶ He concludes that cicatricial stenosis of the aqueduct is the most frequent lesion in congenital hydrocephalus (about 50 per cent.) and is found in a large percentage of cases of hydrocephalus occurring in infancy and early childhood. The stenosis may occupy the entire length of the aqueduct or varying parts; it may be only a thin, even transparent membrane. Again, the stricture may be only partial. Strictures of the aqueduct of Sylvius can be diagnosed and localized accurately. The indigo-carmin test will indicate that an obstruction is present; ventriculography will be the means of precisely locating the obstruction. Spontaneous relief is not possible. Surgical attempts to drain the fluid from the third ventricle to the exterior of the brain have all proved futile. The openings invariably close and the fluid cannot absorb in the subdural space.

He suggests a new surgical procedure by which a new aqueduct of Sylvius is constructed. A tube is left in place for from two to three weeks, whereby it is hoped the epithelium will regenerate and establish a new canal. This operation has been performed on two patients, both of whom recovered from the operation. One patient died of pneumonia several weeks later; the second seems to be well one year after the operation.

Although tuberculous meningitis is considered as being almost universally fatal, A. W. Hollis and I. H. Pardee⁷ report four cases

5. Arch. Pediat. **37**:19 (Jan.) 1920.

6. Surg. Gyn. Obst. **30**:324 (April) 1920.

7. Arch. Int. Med. **26**:49 (July) 1920.

of recovery following the intraspinal administration of antimenin-gococcic serum. Two of these patients showed tubercle bacilli in the spinal fluid. They believe that the intraspinal injection of the serum has two distinct actions: first, by adding to the spinal fluid certain antibodies which it is able to develop itself; and, second, by the introduction within the dura of a foreign protein in the form of horse serum.

The cerebrospinal fluids of forty-eight cases were studied by N. Novick.⁸ In order to ascertain whether the percentage of bovine infection in tuberculous meningitis is appreciably greater, perhaps, than is commonly accepted; whether the bovine type of virus has a special predilection for the meninges. Of the forty-eight strains of tubercle bacilli isolated from the cerebrospinal fluid of forty-eight clinical cases of tuberculous meningitis, three of the strains proved to be of the bovine type. In the analysis of 10,040 cases by Rosenau, whom the author quotes, an alarming percentage of bovine infection in children under 5 years (49 per cent.) is found. Milk entering as the chief element in the diet of children probably serves as a means of transmission of tuberculous disease.

Puncture of the cisterna magna is described by James B. Ayer⁹ for the diagnosis or treatment of postmeningitic subarachnoid block: this results from adhesions which cut off the communication between the different parts of the cerebral and spinal subarachnoid space. A case of staphylococcus meningitis is related in which lumbar puncture yielded only 7 c.c. of clear fluid rich in albumin, but almost without cells and with no micro-organisms, while the cistern fluid was purulent, under high pressure, with numerous cocci. The cistern was washed out with salt solution and at a subsequent puncture autoserum was injected. The patient recovered.

ENCEPHALITIS

Epidemic or lethargic encephalitis in children is discussed by J. B. Neal.¹⁰

There is a gradual onset, followed by marked lethargy and asthenia; headache; low, irregular temperature; frequently cranial nerve palsies. Other paralyses may occur. A masklike expression is often present. The onset may be sudden and is so more frequently in children than in adults. Other striking features that may be present are, catatonia, marked tremors, choreiform movements, profuse sweating, insomnia, delirium, and a marked twitching of certain groups of muscles. A

8. J. M. Research **41**:239 (Jan.) 1920.

9. Arch. Neurol. & Psych. **4**:529 (Nov.) 1920.

10. Arch. Pediat. **37**:321 (June) 1920.

slurring, hesitating speech, with a very slow response is quite characteristic. Disturbances of vision, either diplopia or blurring are fairly common, and are very diagnostic.

The course of the disease is often remarkable for the transient changes in the condition of the patient either for the better or the worse. One of the most striking features of the disease in adults is the prolonged course, which in children is unusual. A differential diagnosis in children must be made from tubercular meningitis, brain tumor, meningism with some unknown underlying cause, syphilitic involvement of the central nervous system, and poliomyelitis or poli-encephalitis and meningitis. The diagnosis from tuberculous meningitis is difficult. In a case of two or three weeks' standing, the patient does not seem to be as ill when suffering with encephalitis. Vomiting is a less constant feature and the pulse is more likely to be regular in encephalitis than in tuberculous meningitis. The spinal fluid examination may serve to differentiate too.

Brain tumor is comparatively rare in children, and usually shows a more protracted course than does encephalitis. Choked disc does not occur in encephalitis although some edema may be present. Meningism may be ruled out by a normal spinal fluid. When changes are found in the spinal fluid, some underlying cause, such as whooping cough, inflammation near the meninges, mastoid or sinus involvement or otitis media must be ruled out before assuming that it is a simple case of meningism. Syphilis of the central nervous system is less common in children and, therefore, is seldom a problem in differential diagnosis. The diagnosis from the encephalitic form of poliomyelitis is very difficult. From various forms of acute purulent meningitis, encephalitis may be differentiated by the examination of the spinal fluid which in the former will show varying degrees of cloudiness, excess of polymorphonuclears, and a diminished or absence Fehlings' reaction.

The blood picture in encephalitis is not characteristic. A slightly leukocytosis may be present. Blood cultures are negative and the urine usually is negative.

The spinal fluid is clear and usually increased in amount. The cells are slightly or moderately increased, the great majority not running higher than in poliomyelitis. The albumin and globulin are increased. Although Neal states that the reduction of Fehlings' solution is normal, it has been found by other workers that a characteristic feature of the spinal fluid is the excess of reduction. The gold chlorid curve duplicates those obtained in poliomyelitis fluid.

E. Wieland¹¹ reports four cases of epidemic encephalitis in children. In three of these cases the pandemic of grippe played no rôle, and in

11. Schweiz. med. Wehnschr. 50:581, 1920.

the fourth case, it probably did not act as an etiologic factor. All of his cases showed the pathognomonic lethargy. The first case showed after a four day prodromal afebrile period, the onset of a choreic delirious stupor with high fever, tachypnae and meningism, following which there developed a brachiofacial paralysis of crossed type. At the beginning, the diagnosis lay between meningism and purulent meningitis, the results of the lumbar puncture ruling these out. Brain abscess was thought of and an exploratory brain puncture was performed with negative results. The second case showed an afebrile lethargy with rigidity of the neck and Kernig sign. It was an abortive case. The third patient, after a period of somnolence and stupor developed marked motor restlessness with hallucinations with choreiform movements in the arms and legs. The fourth patient showed during the prodromal stage, headache, diplopia, ptosis, rigidity of the pupil and lethargy. In the treatment he advises large doses of potassium iodid.

Some advance has been made in the study of the cause and pathology of lethargic encephalitis in the adult form and this knowledge is applicable to the condition occurring in children. C. V. Economo¹² believes that the disease is always produced by a diplostreptococcus.

Lethargic encephalitis and influenzal encephalitis are regarded as identical by H. Weiss¹³ and a similar view is held by W. Lowenthal¹⁴ and O. Speidel.¹⁵

Boccolari¹⁶ found a hemispherical diplococcus slightly larger than the meningococcus and G. Gabri¹⁷ obtained a coccus resembling the micrococcus tetragenus. A gram-positive diplococcus was obtained by blood culture in three cases by A. Maggiora, M. Mantovani and T. Tombolato.¹⁸

The globoid organism of Strauss and Loewe¹⁹ is discussed further by these investigators. They have been able to produce in rabbits and monkeys lesions typical of this disease by Berkfeld filtrates of brain material, nasopharyngeal mucous membrane and nasal washings. The virus has been passed through many series of animals. By means of the ascitic tissue culture method of Noguchi they have been able to cultivate a minute, filterable organism which has been recovered from animals that have been inoculated with virus and which have succumbed

12. *Neurol. Centralbl.* June 1, 1920; *Policlinico (sez. med.)* **27**:93, 1920.

13. *Orvosi Hetilap*, 1920, No. 7.

14. *Deutsch. med. Wchnschr.*, 1920, No. 11, **46**:289.

15. *Münch. med. Wchnschr.* **67**:630 (May 28) 1920.

16. *Policlinico* **27**:100 (Jan. 26) 1920.

17. *Policlinico* **27**:106 (Jan. 26) 1920.

18. *Riforma med.* **36**:114 (Jan.) 1920.

19. *J. Infect. Dis.* **27**:250 (Sept.) 1920.

to the experimental disease. The cultures thus obtained from animals have produced the disease when injected into other animals and the organism again recovered. As a method for differentiation of epidemic encephalitis from epidemic poliomyelitis, they suggest that rabbits are susceptible to infected material from epidemic encephalitis and not from poliomyelitis.

The topic of contagion is discussed by A. Netter²⁰ who cites the case of a child taken to Paris four weeks after the onset of the disease and transmitting it to his sister. He quotes the observations of MacIntosh and Turnball of twelve cases of undoubted encephalitis among the eighteen pupils and three employes of an English boarding school for young girls: twelve of the twenty-one residents of the house were taken ill and four died.

P. Marie and C. Tretikoff²¹ in a new article confirm the correctness of their previous communication relative to the pathology of the disease. They emphasize as the principal features of the disease, the early occurrence of perivascular infiltrations, principally located about the gray matter. The infiltrations consist chiefly of lymphocytes but plasma cells are fairly common while polymorphonuclear cells are rare.

In two cases of a delirious type of encephalitis with marked choreic movements, Peter Bassoe²² reports that the pial infiltrations as well as satellitosis and neuronophagia in the cerebral cortex were relatively more marked than in the usual types of the disease. However, even in these cases the maximum changes were found in the usual locations.

SYMPTOMATOLOGY

The encephalitis epidemic of 1920 in Austria is discussed by C. V. Economo,²³ who points to the three stages of the hyperkinetic cases: an initial stage with fever, severe pains, delirium tremens and herpes which is followed by myoclonic twitching and severe movements, and then the lethargic stage.

The excitomotor phenomena which persists after the acute stage of the disease has subsided are divided by P. Marie and G. Levy²⁴ into choreic movements, bradykinetic oscillations, which are slow, regular movements of large amplitude involving the proximal part of one or both limbs of the same side, myoclonic movements, simple tremors, movements limited to the face, and a Parkinsonian symptom-complex.

20. *Presse méd.* **28**:293 (April 7) 1920.

21. *Ann. d. méd.* **7**:1 (Jan.) 1920.

22. *J. A. M. A.* **74**:1009 (April 10) 1920.

23. *Wien. klin. Wchnschr.* **33**:324 (April 15) 1921.

24. *Rev. Neurol.* **36**:513 (June) 1920.

In the myoclonic form of encephalitis, R. C. Hamill²⁵ showed that the movements had a definite time relation to the respiratory movements, being either equal to or double their rate.

Under the name of acute infections myoclonus multiplex, J. Ramsey Hunt²⁶ gives a full description of the symptoms of this type of disease. LaSalle Archambault²⁷ described the choreic athetoid and choreic psychotic syndromes in encephalitis.

OTHER TYPES OF ENCEPHALITIS

A. S. Root²⁸ reports a case of what he supposes to be focal hemorrhagic encephalitis in an infant 15 months old who for three weeks had been in a state of profound somnolence and who improved strikingly after a transfusion of blood from her mother.

CONVULSIONS IN WHOOPING COUGH

Convulsions in whooping cough were observed in twenty-nine cases out of a series of 342 cases by F. Reiche.²⁹ The greatest number of convulsions were observed in children under 3 years of age. The direct cause of the convulsions varied. In one case there was a tonsillitis; in three cases there was an infection of the meninges; in one case there was an otitis media, in five cases the whooping cough was complicated by tuberculosis, one of these being tuberculous meningitis and one miliary tuberculosis; in three cases there was a sinus thrombosis and in seventeen cases no cause other than the whooping cough could be found. All of the twenty-nine cases terminated fatally.

MISCELLANEOUS

A case of congenital athetosis is reported by Maas.³⁰ This patient was operated on by Borchart. The right cortex was exposed and a strip about 3x1x2 cm. excised. The athetoid movements disappeared and a weakness developed in the thumb. Shortly after power returned but unfortunately when it did the involuntary movements returned as well.

TUMOR

Neuroblastoma Sympathicum.—A case occurring in a boy, 13 years old, is reported by P. C. Gunby.³¹ Failure of vision, weakness, slight headache and bilateral papilledema were the chief general symptoms.

25. Arch. Neurol. & Psych. **4**:44 (July) 1920.

26. Neurol. Bull. **2**:391 (Nov.-Dec.) 1919.

27. Arch. Neurol. & Psych. **4**:484 (Nov.) 1920.

28. Arch. Pediat. **37**:538 (Sept.) 1920.

29. Ztschr. f. Kinderh. **25**:28 (May) 1920.

30. Neurol. Centralbl. **39**:584, (Nov.) 1920.

31. Am. J. M. Sc. **160**:207, 1920.

No local signs were found. The entire vertex was nodular with small tumors. At necropsy a large tumor supposed to be a malignant hypernephroma was found in the left suprarenal and multiple metastatic growths in the skull and sternum were found which on microscopic examination did not show the characteristics of a hypernephroma and the diagnosis of a neuroblastoma was confirmed.

A solitary tubercle of the midbrain is reported by J. H. M. Knox.³² His case was that of a colored boy, 3 years old, who had been perfectly well until six months previously when he stopped crying. Four months later, it was noticed that he trembled when he fed himself. At this time, drooping of both eyelids was noticed, and on examination, a lateral deviation of the eyeball to the right was noted; the right eye frequently showed nystagmus. The gait was normal, and the deep reflexes with the exception of the knee jerks, which were slightly increased, were normal. Within two weeks he had two attacks of weakness, became drowsy and had difficulty in standing and walking. A slight secondary optic atrophy was present and roentgenologic examination showed moderate internal hydrocephalus. The necropsy revealed a tubercle the size of a hickory nut in the midbrain.

A case of blastomycosis of the brain with abscess formation occurring in a boy, 17 years old, is reported by J. T. Moore.³³ The infection resulted from the habit of holding a stick or splinter in the mouth. He developed an abscess behind and above the left ear. Blastomycetes were found in the pus. At necropsy four abscesses were found in the frontal and parietal regions of the left hemisphere.

THE BASAL GANGLIA

Dystonia musculorum deformans has been studied by H. W. Frauenthal and C. Rosenbeck³⁴ who describe and illustrate a case in a Russian Jewess, 17 years old. The following conclusions were drawn:

A summary of the clinical features of this case shows that the patient at a very early age manifested peculiar, involuntary movements—at first limited to an extremity and then, gradually and definitely over a period of several years, various muscle units affected a disposition to behave in this abnormal manner. At present—ten years after the onset—the entire body seems to be convulsed in a relentless dystonia. Evidently no further progress is possible unless it be the muscles innervated by the cranial nerves which thus far have escaped. This is in accord with most writers on the subject who have emphasized the absence of cranial nerve involvement. That these, however, may also be implicated in their supranuclear pathways has definitely been established by

32. *Am. J. Dis. Child.* **20**:436 (Nov.) 1920.

33. *Surg. Gyn. & Obst.* **31**:590 (Dec.) 1920.

34. *J. Nerv. & Ment. Dis.* **52**:134 (Aug.) 1920.

several authenticated observations. The racial predilection, i. e., its limitation to offspring of Polish or Russian Jews is not without interest. The vulnerability of the nervous system in this race to certain types of degenerative disease is thus accentuated.

E. W. Taylor³⁵ also contributes an article on this disease, reporting two cases in a brother and sister, Russian Jews. He prefers the term *dystonia lenticularis*. He believes that there exists a type of motor disorder characterized by hypertonia and hypotonia giving rise to a destructive form of gait and motor disturbances in general, sufficiently definite to demand a provisional classification. Although occurring with frequency in children of the Jewish race, it is not confined to that race nor does it always occur in childhood. It is sporadic rather than truly familial. It is not to be sharply differentiated from various other forms of motor disorder of presumable extrapyramidal origin. It is due to structural alterations of the lenticular nucleus, has no defined pathologic anatomy but its association with alterations of the liver is probable.

Acute cerebocerebellar ataxia is described by J. P. C. Griffith,³⁶ who reports three new cases consequent to acute cerebellar encephalitis. With this are always combined symptoms indicating an involvement of the large brain as well and the title of "cerebro cerebellar encephalitis" or "cerebro-cerebellobulbar encephalitis" is consequently to be preferred. The cause varies decidedly. In the majority of cases previously reported in medical literature, some infectious disease had preceded the attack. Among the symptoms most frequently seen are unconsciousness usually of but short duration, ataxia, affection of speech, disturbance of mentality which is usually temporary, nystagmus, and loss of power. The prognosis would appear good as to life and of the seventeen cases collected from literature, complete recovery occurred in a considerable number of cases and it would appear that the disease will leave no traces in the majority of instances.

SPINAL CORD

Hemorrhage of the spinal cord at birth is discussed by Charles W. Burr³⁷ who reports two cases with cervical segment lesions, both being breech presentations. He refers to the report of Spencer who in forty-four cases in which the spinal cord was examined found in eighteen edema and congestion and in thirty-six hemorrhage of the cord. In one of Burr's cases there was hemorrhage, swelling and

35. Arch. Neurol. & Psych. **4**:417 (Oct.) 1920.

36. Am. J. Dis. Child. **20**:82 (Aug.) 1920.

37. Am. J. Dis. Child. **19**:472 (June) 1920.

myelitis. He is inclined to believe such hemorrhage spontaneous and not caused by direct violence but by rupture of overfull vessels from pressure of soft parts during birth.

Rupture of the spinal cord in dystocia is discussed by F. H. Kooy.³⁸ His case is unique as the child lived for nearly nine years. The child was born by version and extraction, the shoulders being delivered by the method of Mueller. From her birth until her death, about nine years afterward, she suffered from paraplegia (except for flexion of the legs at the hip joint), from total or almost total loss of sensibility of the legs and from incontinence of urine and feces and a decubitus.

The significance of xanthochromia of the cerebrospinal fluid is discussed by Isaac A. Abt and I. Harrison Tumpeer.³⁹ They report an infant born in the eighth month of gestation who lived thirty-seven days and manifested the signs of an internal hydrocephalus, pyelitis and a bronchial pneumonia. They feel that the xanthochromia was due to subpial hemorrhage as a result of meningo-encephalitis.

SYPHILIS OF THE NERVOUS SYSTEM

Nerve deafness due to congenital syphilis is reported by W. B. Kay⁴⁰ occurring in three children, 7, 5 and 4 years old, respectively. In each of them the disease began between the first and second year. The Wassermann reaction was positive in the oldest child and in the parents. Kay believes these children to show the manifestation of late hereditary syphilis.

P. Hutinel⁴¹ discusses the relation of hereditary syphilis and dystrophy. He states that hereditary syphilis produces two types of groups, the first in which specific alternations are produced by the treponema or reactions of this organism, and the second class in which nutritional difficulties occur. Such dystrophic changes may be local, partial, frequently multiple and in some general. They attack chiefly the endocrine glands and the offspring of such patients may show hereditary dystrophies.

A case of juvenile tabes is reported by Kerr⁴² occurring in a child 14 years old and characterized by lightning pains, primary optic atrophy and absent knee jerks. The examination of the blood was negative.

Toni Schmidt Krapelin⁴³ details thirty-nine certain cases of juvenile paretics among the 2,184 admissions to the psychiatric clinic

38. *J. Nerv. & Ment. Dis.* **52**:1 (July) 1920.

39. *Am. J. Dis. Child.* **20**:153 (Sept.) 1920.

40. *J. A. M. A.* **74**:1162 (April 24) 1920.

41. *Arch. de méd. d. enf.* **23**:5, 145, 205 (Jan.-April) 1920.

42. *Lancet* **1**:598 (March 13) 1920.

43. *Ueber die Juvenile Paralyse*, Berlin, 1920, Julius Springer.

at Munich in the last fifteen years. Twenty-six were males, thirteen were females. There were eight doubtful cases, or cases in which an erroneous diagnosis was made, and seven cases of "preparalysis" in children who showed no clinical signs of metasyphilitic affection of the central nervous system, but in whom serologic findings characteristic of general paresis were more or less accidentally discovered.

As regards the serology in ancestry, results are given of forty-eight persons of twenty-one families. A positive blood Wassermann occurred in five out of eleven fathers, fourteen out of eighteen mothers and in nine out of nineteen brothers and sisters. Alcoholism was found in 20 per cent. of the cases. Over one-fourth of the cases showed general arrest of development, one-third were mentally deficient and among the early signs was the change in character. An acute onset with hemiplegia and other focal symptoms and major epileptiform attacks were relatively frequent, suggesting a combination of this disease with brain syphilis. Optic atrophy without other tabetic symptoms is comparatively frequent. Total immobility of the pupil is more common than simple loss of the light reflex. As a forerunner of the characteristic speech disorder there is sometimes observed in paralytic and preparalytic children a peculiar hurried, toneless and puffing way of speaking. Periods of excitement are common but an agitated condition persisting throughout was seen only in about 10 per cent. of the cases. A majority of the cases showed a typical Wassermann reaction in the blood serum and spinal fluid. No therapeutic results worth mentioning were obtainable by any known treatment.

MUSCULAR ATROPHIES

Amyotonia congenita is described by N. O. Pearce.⁴⁴ He reports five cases, two appearing in twins. In reviewing the literature he states that 132 cases have now been reported. In his cases the symptoms were present at birth, in four affecting the upper extremities and in all the lower extremities. The knee jerks were absent in three, very weak in one and sluggish in one case. Pad feet were present in three cases. In the first of his cases an exhaustive metabolic study was carried out and a lowered creatinin excretion was found in addition to the excretion of creatinin, on a low protein diet. A normal uric acid excretion, and increased rest nitrogen accompanied by an increased neutral sulphur and a normal phosphorus excretion was present.

A case with complete histologic examination of the nervous and muscular systems is reported by J. B. Holmes.⁴⁵ The spinal cord was relatively large and the anterior roots were diminished in size. The

44. *Am. J. Dis. Child.* **20**:393 (Nov.) 1920.

45. *Am. J. Dis. Child.* **20**:405 (Nov.) 1920.

large cells of the anterior horn were strikingly few in number and in part were represented by cells that are much smaller in size but otherwise of the same appearance. Only infrequently was a cell of normal size found, but save in size, such cells are of perfectly normal appearance. There was a complete absence of acute or inflammatory condition, myelinization was normal, and there was no evidence of recent degeneration. In the muscles are found fibers of small, apparently healthy muscle fibers, intermingled with bundles of hypertrophied fibers. The small fibers appeared to represent less fully developed bundles of muscle tissue. The component muscle cells are embryonic in appearance, resembling most closely muscle cells of a human fetus in the third month. "The histopathologic findings are best explained, on an assumption of a delayed or retarded embryonic development affecting certain motor cells of the anterior horns and certain groups of developing muscle cells (fibers)."

The relationship of amyotonia congenita and the congenital familial spinal muscular atrophies is discussed by K. H. Krabbe⁴⁶ who reports seven cases. He believes that cases which have been described as amyotonia congenita really represent two different diseases. One of these, true amyotonia congenita, is a benign disease consisting of hypotonia, hyperflexibility and weakness, but no muscular atrophy. From this condition the patient recovers if an intercurrent disease does not produce death. Other cases, including six of his own, are to be considered cases of congenital familial progressive muscular atrophy, resembling the Wernicke-Hoffmann type of muscular atrophy.

On the other hand, E. J. Huenekens and E. T. Bell⁴⁷ believe the two conditions to be extreme types of the same disease, represented by Erbs' juvenile form of muscular dystrophy and the hereditary form of Leyden and Moebius.

PERIPHERAL NERVES

Obstetrical palsies are considered to be due to stretching and tearing of the roots of the brachial plexus by J. W. Sever.⁴⁸ Girls and boys are equally affected. The upper arm type is the most frequent; both arms are very infrequently affected. They are practically always associated with difficult labor involving either forceps or force. They are more frequent in head deliveries, and they may be associated with fracture of the clavicles. He states that it is not the result of posterior dislocation or fracture of the humerus. On the other hand, J. Turner Thomas⁴⁹ states that in most cases such paralysis disappears, and in

46. Brain **43**:166 (July) 1920.

47. Am. J. Dis. Child. **20**:496 (Dec.) 1920.

48. Canadian M. A. J. **10**:141 (Feb.) 1920.

49. Am. J. M. Sc. **154**:207 (Feb.) 1920.

all cases in which serious crippling of the limb was found, a posterior subluxation of the shoulder joint was present. He argues that the basic causal lesion was in the shoulder joint and not in the brachial plexus.

M. Clauss⁵⁰ in discussing multiple neuritis in childhood states that aside from the neuritis following diphtheria there are only forty cases recorded. Most of these have followed acute infectious diseases particularly mumps, typhoid fever, scarlet fever, influenza, whooping cough, sore throat, acute articular rheumatism and appendicitis. The prognosis in multiple neuritis in childhood is good. He gives a history of six cases, one of particular interest associated with or followed by a tuberculous iridocyclitis which had been treated for six weeks with tuberculin.

DeLavernne⁵¹ reports in diphtheritic paralysis a meningeal reaction characterized by an excess of glucose, albumin and a normal cell count in the spinal fluid. An interesting case of post diphtheritic paralysis of the respiratory muscles occurring in a girl ten years old who recovered after prolonged artificial respiration is reported by W. M. Marriott.⁵²

The treatment of postdiphtheritic neuritis is discussed by H. Gödde.⁵³ He reports two cases in which steady improvement occurred following the administration of serum in large doses and he is inclined to agree with Paessler and Heubner in their administration of antitoxin.

Acrodynia, a polyneuritic syndrome resembling pellagra, and occurring in children under four years is discussed by Albert H. Byfield,⁵⁴ who reports seventeen cases. The chief features are:

1. *Nervous Symptoms*.—Paresthesias of the hands and feet leading to incessant scratching or chewing of the extremities. The reflexes are diminished in most cases. Dulness and apathy alternate with extreme restlessness and sleeplessness.

2. *Skin and Hair*.—The hands and feet are cyanotic and cold and become the seat of an erythematous rash which is confluent and most marked on the fingers and toes. Desquamation is present and most marked distally. Falling and pulling out of hair is common.

3. Loosening and falling out of the teeth.

50. Deutsch. Ztschr. f. Nervenhe. **65**:169 (March) 1920.

51. Bull. et mém. Soc. méd. d. hôp. de Par. **44**:1246, 1920.

52. J. A. M. A. **75**:668 (Sept. 4) 1920.

53. Neurol. Centralbl. **39**:532 (Aug. 16) 1920.

54. Am. J. Dis. Child. **20**:347 (Nov.) 1920.

4. *Anorexia and Resulting Emaciation*.—The rather common acetoneuria is probably the result of starvation superinduced by the anorexia.

5. *Leukocytosis* (11,000-30,000).

The duration of the disease varied from two to eight months. The mortality appears to be low. Two of the first cases seen were fatal but forced feeding had not been employed.

As to etiology, it appears that dietary error is not responsible as most of the patients had received proper feeding. An infectious origin is more probable and it has been suggested that we are dealing with a sequel of influenza. In this connection it is interesting to note that the term *acrodynia* was first used in 1828 to designate a similar affection also known as epidemic erythema. An influenza epidemic existed at that time.

ANOMALIES OF GROWTH

An endocrine origin and inclusion in the heredo-degenerations for facial hemiatrophy is suggested by F. Boenheim,⁵⁵ who reports several cases and analyzes the subject very thoroughly. His first patient, a man, aged 32 years, was a veritable giant, measuring 198 cm. in height. He was left-handed. In early childhood atrophy of the right side of the face began and came to a standstill at 15 years of age. At 29 or 30, a similar process began on the left side of the face and was still progressing at the time of examination. The growth of hair on the trunk, as well as on the most atrophic parts of the face was deficient. The right upper teeth were smaller than the left, as was the right ear. There were no acromegalic features and the sella turcica was normal. There were no signs of a lesion of the sympathetic. A supernumerary nipple was present on the right side. The subsequent involvement of the other side of the face is not very unusual as Coenenberg found it in eighteen of 250 cases.

Congenital Hemihypertrophia Totalis.—H. R. Coston⁵⁶ reports a case of hemihypertrophia totalis in a male child, aged 22 months. He was the fourth child of healthy parents of rather low mentality. The other three children have been mentally superior to their parents. The patient had never been ill and seemed normal mentally. The birth followed a quick and easy labor. The mother was well during pregnancy. There was available a history through six generations with only one cousin in any way defective. This child, a second cousin on the mother's side, had six toes on the right foot. There was no idiocy, insanity, epilepsy or deformity other than the one mentioned in the

55. Deutsch. Ztschr. f. Nervenhe. **65**:219 (May) 1920.

56. Med. Rec. **97**:222 (Feb. 7) 1920.

family. Immediately after birth, it was observed that the right side was "that of a 10 pound baby while the left side was that of a 7 pound baby."

Coston remarks that cases of hypertrophy are usually on the right side and there are many cases of partial hypertrophy confined to a single limb or one side of the face. Occasionally cross cases are seen.

Edward A. Park and Grover F. Powers⁵⁷ report two cases of scaphocephaly with malformation of the extremities. They point out that the deformities were not limited to the hands and feet in a number of the cases recorded.

A study of achondroplasia is reported by Thomas Foster Wheelton⁵⁸ who states that achondroplasia is characterized by symptoms of dwarf growth (shortness of extremities, vertebral column and chest, and looseness of the skin) and symptoms of infolding (kyphosis basoes crania, sagittal narrowing of the foramen magna, depression of the nasal bridge, narrowing of the choanae, reduction in size of the sella turcica with consequent reduction in size of the pituitary body, and changes in the spine). The changes of achondroplasia are probably due to the smallness of the amnion, except in rare cases and are produced between the third and eighth weeks of intrauterine life. They consist mainly in a retardation of ossification of the epiphysis and the diminished production of endocranial bone, while the periosteum produces a comparatively normal amount of bone. Roentgenographic examination shows that near the epiphysis, the diaphysis become cup shaped producing a T-shaped outline without any disturbance of the epiphyseal line or zone of proliferation, though these may be uneven and take on more or less phantastic shapes. Two of the cases presented showed a new symptom, a wedge-shaped vertebra, which falls in the group of symptoms of infolding and fortifies the belief that the state of achondroplasia is produced by smallness of the amnion.

The enlargement of the head in these dwarfs has recently been studied by W. E. Dandy⁵⁹ and attributed by him to slight hydrocephalus.

A case of dwarfism attributed chiefly to general infantilism is reported by C. Sternberg.⁶⁰ His patient was a boy, aged 17, and was 36¼ inches tall. He was said to have been of normal size at birth but at the age of 1½ years he developed curvature of the spine and retardation of growth. He died as the result of tuberculous spondylitis.

57. *Am. J. Dis. Child.* **20**:235 (Aug.) 1920.

58. *Am. J. Dis. Child.* **19**:1 (Jan.) 1920.

59. *Bull. Johns Hopkins Hosp.* **32**:5 (Jan.) 1921.

60. *Beitr. z. path. Anat. u. z. allg. Path.* (Oct. 15) 1920.

He was classified as a case of true dwarfism. The bones other than the spine were normal and all the epiphyseal lines wide open. The appearance of the body was decidedly infantile. The genitalia were small and there was no trace of beard, axillary or pubic hair. The testicles showed primary defective development and for this reason, it was felt that the dwarfism could not be contributed to hypophyseal deficiency which would have caused atrophy of the testicles.

Mongolianism is regarded by E. Apert⁶¹ as one of the extremely varied group of abnormal infants which with congenital myxedema occupy two special groups each one different from idiocy, imbecility, retardation of evolution or mental anomaly. He has found that in the great majority of cases the Mongolian is frequently the last of a numerous family, the mother relatively advanced in age and weakened by previous pregnancies. In general, the Mongolians will always need guidance, they will lack intuition and judgment. They will remain below standard but can be sufficiently restored to escape feeble-minded institutions.

MENTAL DEFICIENCY

The effect of influenza on feeble-mindedness is studied by K. A. Menninger⁶² who concluded that the usual effect of influenza on the brain is not the production or precipitation of feeble-mindedness, and if it ever produces feeble-mindedness it is probably by means of an encephalopathy. On the other side, of those already manifesting mental lack, certainly a few are influenced adversely by influenza, and the symptoms of feeble-mindedness are aggravated, an aggravation which may be predominately in the intellectual sphere, in the emotional sphere or in the volitional sphere. Psychoses of an indeterminate type are occasionally precipitated by influenza in the feeble-minded, even as in normal persons. At least occasionally, though rarely, the effect of influenza on feeble-mindedness may be symptomatic amelioration.

Edward A. Strecker,⁶³ in a study of fourteen mentally defective and eighteen mentally retarded cases concluded that in 56 per cent. of all cases, congenital mental deficiency did not exist, and that in 84 per cent. of the retarded cases there were significant underlying physical factors on which the mental retardation depended. There were six cases of congenital syphilis, one of tonsillitis and organic heart disease, one of rachitis, one of organic brain disease, one of hypopituitarism and five who, although they had no definite organic disease, were undernourished and underdeveloped as the result of economic or environmental conditions.

61. Bull. méd. **34**:563 (June) 1920.

62. J. A. M. A. **75**:1044 (Oct. 16) 1920.

63. J. A. M. A. **75**:659 (Sept. 4) 1920.

William Healy⁶⁴ discusses the occurrence of certain signs and symptoms in delinquents which are significantly correlated with etiologic facts of great importance in the interpretation of the cases. These symptoms are indicative of some disturbance in the mental life of the patient and in the cases cited, consisted chiefly of disturbing sex experiences. From an intensive study of a large number of young delinquents, he has been led to the conclusion that about 10 per cent. are sufferers from the inner turmoil that we call mental conflict. He calls attention to the importance of this field and the remarkable accomplishments of therapeutic accomplishments.

The importance of an outpatient clinic in connection with an institution for the feeble-minded is pointed out by Walter E. Fernald⁶⁵ who described the system that has been developed for the examination of patients applying to this department. Among the types of advice given may be included institutional care, home care, special class, private school, private teacher, change of teacher, let-up school pressure, take out of school and go to work, change of work, treat as delinquent, special medical or surgical treatment, country life, vocational training, add new interests and recreation, place in selected family, modify home environment, etc. It is a striking fact that of the thousands of cases diagnosed as feeble-minded at this clinic since 1891, the majority have not subsequently applied for admission and are not known to have misbehaved seriously.

64. *Arch. Neurol. & Psych.* **4**:680 (Dec.) 1920.

65. *Am. J. Insanity* **77**:227 (Oct.) 1920.