

Symposium

NEUROLOGICAL CONDITIONS IN CHILDREN

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INTRODUCTION

RANDOLPH K. BYERS, M.D.

In our discussion this morning we will deal with certain neurologic conditions of childhood. Such conditions differ from those in adults in that they affect a more or less rapidly developing complex organ. You need only be reminded of the rapid increase in brain weight and of the functional maturation attending and continuing after this weight gain to realize this fact. Our various developmental and intellectual examinations are attempts at measuring the latter.

The results of disease processes may show themselves by disturbing in various ways tissues already functionally active, as for instance by suppressing respiration in the newborn, or they may injure tissue not yet mature in which case they tend to distort or suppress development. As Dr. Crothers has long maintained, recovery from acute cerebral insults in infancy and childhood differs from recovery in adult life. In the latter, approximate return to the status quo ante is a satisfactory result. If, however, a 3 year old regains his attributes as a 3 year old and then fails to continue to develop he will be feeble-minded at 6. Neurologic recovery in childhood therefore requires the resumption of orderly development, and can often not be determined for a period of years after an insult.

This is not the place to attempt an etiologic catalogue of cerebral disease in childhood. Suffice it to remind you that prenatal, natal and postnatal periods of life each has its own special developmental hazards. Because these affect the developing nervous system they tend in many instances to produce rather widespread changes resulting in the functional disabilities which we label mental retardation, cerebral palsy and epilepsy. Once such holistic difficulties are established we are powerless to effect radical care.

Effective plans for the care and treatment of these chronic injuries can be made only if we have a careful assessment of the deficits and especially the remaining assets. In the very early months of life severe deficits may exist with little or no observable functional abnormality. For example a neonate with a cortical cyst in the parietal region may well show no functional disturbance at birth but at 2 months he may use the involved arm less than its fellow, and when he reaches the upright stage he will have a typical hemiplegia with reflex changes and contractures. In many instances long periods of development are necessary before any true assessment of deficits is possible.

Chronic convulsions, it seems to me, behave similarly. When they become evident early in life gross changes in brain mass or structure are more likely to be found on histologic or gross examination than is the case with chronic convulsions beginning later in life. Dr. Gibbs will consider the philosophy of the convulsive state with you.

Since we are powerless to change damage to the nervous system once it has become established it is of the utmost importance that we make every effort to prevent or minimize it during acute crises. A very commonly recurring acute crisis is that of meningitis, and on its proper handling may rest the entire intellectual and physical future of the affected child. Dr. Alexander will tell you about guides to treatment aimed at expediting recovery and insuring as well possible future development.

Unless we have some insight into the natural history of cerebral palsy we will not know whether treatment has ameliorated the motor defect nor more importantly will we be informed as to its bearing on personality development. Dr. Crothers will show you what has happened in the way of changing motor patterns and personality development in this condition in patients who have attained adulthood.

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Finally Dr. Matson will call to your attention some of the very discrete disturbances of the nervous system found early in life. Again though they may be discrete, developmental and growth factors make their recognition and treatment on a surgical basis important.

I should now like to introduce to you Dr. Frederic Gibbs, Professor of Psychiatry at the University of Illinois, whose subject is "Convulsive Disorders in Childhood."

Following Dr. Gibbs, the speakers will be: Dr. Hattie E. Alexander, Associate Professor of Pediatrics at the College of Physicians and Surgeons, Columbia University, New York; Dr. Bronson Crothers, Clinical Professor of Pediatrics Emeritus, Harvard Medical School, Consultant in Neurology, Children's Medical Center, Boston; Dr. Donald Matson, Assistant Professor in Surgery, Harvard Medical School, Neurosurgeon, Children's Medical Center.

CONVULSIVE DISORDERS IN CHILDHOOD

FREDERIC A. GIBBS, M.D.

Epilepsy is what the brain does when it is slightly injured, but the type of injury seems to be of less importance in determining the clinical outcome than the stage of development (maturational state) of the brain at the time the injury was received.

The form of the epileptic disorder varies greatly with age. Clinical and electroencephalographic manifestations must be considered together; they are but different aspects of the same thing. In the first year very severe electroencephalographic abnormality with almost continuous high-voltage, slow waves interspersed with spike seizure activity is common; this has been given the term hypsarhythmia, meaning mountainous arrhythmia. The pattern denotes nearly total disorder. It is associated clinically with very frequent brief spasms, quivering spells or myoclonic jerks. Generalized convulsions occur in some cases but are not usually a prominent feature of the clinical picture. The function of the cortex appears to be so severely disturbed that there is insufficient organization to produce a series of full-blown tonic-clonic seizures. The hazard in cases with hypsarhythmia and infantile spasms is not epilepsy, but death or feeble-mindedness.

In the second to fifth year a more organized epileptic pattern is common. This consists of 2 per second spike and wave discharges; it is referred to as petit mal variant. It is usually associated with discharges of very high voltage spikes which are particularly numerous in sleep, and it is characterized clinically by frequent, short tonic and severe tonic-clonic convulsions (grand mal). Feeble-mindedness is common in cases with a petit mal variant and next to hypsarhythmia the prognosis is worse than for any other type of epilepsy.

From the fifth to the twelfth year the highly organized 3 per second spike and wave of petit mal epilepsy is common. This pattern is associated with pyknoleptic seizures which have, in addition to brief impairment of consciousness, one or more of the following manifestations: fluttering of the eyelids, rhythmic (3 per second) nodding of the head or jerking of the arms and trunk. High voltage discharges of the grand mal type may be present but they are less prevalent than in petit mal variant and grand mal is a less serious complication than in petit mal variant epilepsy.

All the patterns thus far described are diffuse and more or less generalized, but with increasing age the epileptic disorder not only becomes more organized and less devastating, but more localized. For example, at ages 10 to 20 years, 14 and 6 per second positive spikes are common; these are believed to denote an epileptic focus in the thalamus or hypothalamus. The clinical manifestations are attacks of dizziness, pain, rage and vegetative disturbances but tonic-clonic convulsions occur in approximately 50% of cases. In adult life the epileptic pattern that is most common is a focus of seizure activity in the anterior part of the temporal lobe. A high percentage of patients with this type of focus have major convulsions, but the pathognomonic clinical feature is confusional attacks or trance-like episodes with coordinated and more-or-less purposeful movements.

Although focal epilepsy is less common in infants and children than in adults, epileptic foci occur at all ages. In infancy the most common focus is in the occipital areas, in childhood in the mid-temporal areas and as previously stated the anterior temporal focus is the characteristic focus of adult life.

Discrimination and understanding are aided by viewing the clinical and electroencephalographic aspects of epilepsy in combination, over a time range that permits consideration of maturational changes. This view suggests that the form which epilepsy takes is largely determined by the age (maturational state) of the brain when the injury was sustained. Although certain types of injury are more common at certain ages than others a great variety of different types of injury are capable of producing the same electroencephalographic disorder and the same types of clinical epilepsy.

GUIDES TO OPTIMAL THERAPY IN BACTERIAL MENINGITIS

HATTIE E. ALEXANDER, M.D.

It is now apparent that the potential 100% recovery rate from the commonly occurring varieties of pyogenic meningitis is far from being realized. The limitations of therapy over the past 10 years are documented by a number of reports of a significant incidence of sequelae of neurologic and psychologic nature. The frequency of subdural collections of fluid as a complication during recovery from pyogenic meningitis has been reported from a number of institutions. These limitations are apparently the result of late diagnosis, both clinical and bacteriologic, and suboptimal use of the available therapeutic agents.

The goal—100% recovery—can be approached only if we aim for optimal therapy. This paper will present the guides which are of value in attaining optimal therapy. In the process 2 controversial subjects will be discussed: indications for intrathecal therapy, and the use of certain combinations of antibiotics. Four principles can serve as guides to optimal therapy.

I. For therapy to be optimal, it must be applied early in the course of the disease.

II. The organism must be eliminated in the shortest possible time. A number of guides are useful in attaining this aim.

A. Use of 2 agents which work through different mechanisms of action to prevent emergence of resistance. The available knowledge on this subject is summarized.

B. One of the agents used should, if possible, be primarily bactericidal.

C. In order to obtain the most rapid elimination of the infectious agent, bactericidal concentrations must be attained rapidly in the spinal fluid and constantly maintained over a long enough period to kill the organisms during their first exposure.

III. Intrathecal therapy should be kept at a minimum.

The aim to reduce intrathecal therapy to a minimum is occasioned not by evidence that patients are damaged by reasonable doses intrathecally, but by the fact that it is now unnecessary except under the circumstances outlined.

IV. Agents which may injure patients are to be avoided if others which are safe are equally good.

It is now possible to avoid to a large degree agents which may injure the patient, such as streptomycin, polymyxin B and bacitracin. Only in tuberculous meningitis are we justified in accepting the risk of damage by the usual streptomycin therapy.

THE LONG-TIME PROGNOSIS OF CEREBRAL PALSY

BRONSON CROTHERS, M.D.

For the past year we have been recalling individuals with cerebral palsy who have been seen on the neurologic service or in private practice, since 1925.

All cases have been seen by one physician or his close associates, all psychologic testing has been done under the supervision of one of 2 psychologists, and all physical therapy under the supervision of one individual.

Whatever is good or bad in this situation, one thing is clear. All mistakes are our own, and we know pretty adequately our own attitudes. We started off with a few convictions which we still cling to.

First we always recognized that we were dealing with a physiologic and psychologic problem in a growing and developing child. We were also aware that the family, the school and later the industrial opportunities had a very definite effect upon the patterns developed as years went on.

Furthermore we accepted very quickly the idea that psychologists could help us more than psychometrists.

A follow-up study of this sort ought to be concerned with several aspects of the problem:

1. The validity of the original appraisal. In this connection we have been interested in the possibility that unexpected changes may be due to anatomic changes as time went on.

2. The possibility is obvious that a reappraisal might throw light on the validity of various methods of treatment.

3. The emotional, economic and educational costs can be correlated with the efficiency of the individual in adult life.

4. Revision of methods of examination might be suggested.

In this symposium 3 moving pictures were shown. In one, a series of pictures at various ages

show the difficulties of an alert child with severe cerebral palsy and tonic neck patterns. At first the child appears to share the optimism of the therapist. As time goes on, less enthusiasm for the imposed activity is evident.

The persistent tonic patterns finally convinced the medical part of the group that the child would not benefit by further pressure. Finally her status in young adult life is shown with her assets and deficits clearly defined.

It is our hope that a series of such observations may clarify some of the problems we all see.

Another aspect of the study was illustrated by 2 pictures, showing methods of establishing the pressure of hemianopsia in children. We were lucky enough, a year ago, to enlist the interest of Dr. J. P. M. Tizard, of St. Mary's Hospital, in London. He found hemianopsia in a large number of the hemiplegics that returned for study. Many of these children had been recognized as hemiplegics in infancy, but sensory studies had not been done then, or later.

The hospital has under observation a considerable group of young children so that it was possible to find appropriate hemiplegics at various ages. The method of Dr. Tizard is illustrated by a film showing the co-operation of a borderline defective at 12 years. A second film, of a defective restless child of 5 years, shows another method of demonstration which involves selection of toys presented at equal distances from each side. These films shown, of course, are of interest only because they show the relevant activities of children at different ages.

As we go on with this study, we will attempt to continue the rather laborious follow-up of cases and hope for further disconcerting experiences which will lead to review of our technics with younger children. Certainly we have been taught to look for sensory disturbances and to appreciate their importance.

DEVELOPMENTAL ANOMALIES OF THE NERVOUS SYSTEM OF SURGICAL SIGNIFICANCE

DONALD D. MATSON, M.D.

There is a new type of surgery practiced today in an ever increasing degree; this is prophylactic surgery directed toward the prevention of the complications of congenital defects. Two types of congenital disorders involving the central nervous system are selected for discussion because they are rather poorly understood and because, when promptly and properly diagnosed, they are subject to surgical treatment with gratifying results.

The first of these consists of congenital dermal sinuses with persistent intracranial or intraspinal extension. When there is failure of both mesodermal and ectodermal structures to fuse properly in the midline, dermal sinus tracts may extend all the way into the central nervous system, particularly in the suboccipital and lumbo-sacral regions. Here the sinus may become attenuated in the meninges or it may expand to form an epidermoid or a dermoid cyst. Such a cyst may act as any other expanding lesion to interrupt function by local compression or by obstruction of spinal fluid circulation. It also acts as a persistent portal of infection from the skin surface.

The results of surgical treatment prior to infection have been uniformly gratifying; the results of treatment when the proper diagnosis was made only after meningitis or abscess formation occurred have been extremely poor. This lesion should be suspected whenever there is a midline skin dimple or sinus tract opening and particularly if there is an underlying defect in the bone of the skull or spine on x-ray examination. A history of meningitis or physical signs of increased intracranial pressure or spinal cord compression associated with such a skin defect are practically pathognomonic.

Such a sinus tract should not be probed or injected. The treatment of choice is complete excision of the entire dermal sinus tract together with all cystic expansions wherever they may extend, before infection has occurred or at least at a time when no active infection is present. This usually means posterior fossa exploration for the intracranial lesions and laminectomy for the spinal lesions.

The second congenital disorder is an unusual but constant type of spina bifida occulta in which the distinguishing feature is the projection of a discrete bony spicule from the posterior surface of a vertebral body through the center of the spinal canal, thus dividing it into compartments and transfixing the spinal cord or cauda equina. These patients are designated by the term diastematomyelia, signifying a cleft or division of the spinal cord. It is important to correct this disorder during the period of active growth of a child to prevent progressive loss of function.

These bony spicules have been seen at various levels from T₆ to L₄. They may be suspected by

an overlying cutaneous abnormality such as excessive hair growth, a skin dimple, a subcutaneous fat pad, or cutaneous vascular malformation. The lesion is usually discovered during investigation of a gait disturbance, lower extremity deformity or impairment of bladder or rectal sphincter control. Roentgenographic findings are characteristic, showing a widened spinal canal, spina bifida, and the midline bony spicule.

Treatment consists in surgical excision of the bony spicule, division of the numerous fibrous adhesions between the septum and the split cord, and conversion of the reduplicated spinal canal into one channel. This has repeatedly resulted in clinical improvement and almost always in an arrest of any advancing neurologic abnormality.

These 2 lesions are illustrative of conditions in which well conceived and carefully performed surgical procedures can do much to prevent progressive disability and perhaps disastrous complications and should, therefore, be recommended strongly as elective procedures.

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