BRUCE O. BERG

CHILD NEUROLOGY

A CLINICAL MANUAL

Maruzen Asian Edition

Child Neurology

A CLINICAL MANUAL

Edited by

Bruce O. Berg, MD

Professor of Neurology and Pediatrics; Director of Child Neurology, University of California, San Francisco

With Associate Authors

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Foreword

Child neurology has grown and has become firmly established as a specialty during the last 30 years. This period has seen an improved understanding of the nervous system and of many of the disorders that affect it. New diagnostic tools and therapeutic measures have been devised. Many neurologic diseases, once poorly understood and classified under the vague heading of degenerative disease, have now been recognized as being the result of inborn errors of metabolism and, in many of these, the enzymatic defects have been defined.

New diagnostic methods have made it possible to more readily and more accurately define abnormalities in the nervous system. Computerized axial tomography (CT scanning) and ultrasonography are noninvasive techniques that have revolutionized the localization of lesions in the brain and spinal cord. Neurologically impaired children have been benefited by some of the new methods of medical management that have been developed. For example, the ability to monitor and influence intracranial pressure has improved the survival rate of children with brain swelling secondary to trauma and to illnesses such as Reye's syndrome.

With the development of new drugs, infections of the nervous system including one viral illness can now be treated more effectively. New anticonvulsant agents and the development of drug monitoring have resulted in better seizure control in children with convulsive disorders. The judicious use of amniocentesis and prenatal diagnostic methods have led to the prevention of a number of neurologic disorders for which there are still no cures.

It is important that all physicians involved in the care of children be made aware of the dramatic changes that have resulted from this explosion of knowledge in neurology. In this book, Dr. Bruce Berg, a distinguished child neurologist and teacher, has made his specialty with much of its new information, available to these physicians by carefully selecting for discussion a limited number of topics encompassing both acute and chronic neurologic problems. Utilizing his own vast experience and that of some of his colleagues he has described in an informative, lucid, and concise manner with the liberal use of carefully conceived tables, a practical, clinical approach to the child with a suspected neurologic disorder.

Dr. Berg has pointed up the importance of the medical history and of the physical and neurologic examinations in making a diagnosis of neurologic disease at a time when there is a tendency to replace these basic tools and medical judgment by excessive and, at times, unthinking reliance on expensive diagnostic measures. All physicians who are responsible for the care of children will find this book extraordinarily informative and useful.

Sidney Carter, MD
March, 1984

To Linda,
Kate, and Sarah

Preface

This book is designed to be a readily available first reference on the practical matters of diagnosis, differential diagnosis, and treatment of disorders of the nervous system in infants and children. It is hoped that the book will be a useful guide for all physicians and house officers in pediatrics and neurology.

The book is organized into two interrelated parts. The first ten chapters deal with fundamental topics in child neurology; with neuroanatomic information included for a better understanding of the neurologic examination. The second part of the book is problem oriented and deals principally with major symptoms. Throughout, detailed tables are included for brevity and clarity of presentation. Key references are placed at the end of each chapter.

I thank the contributing authors for their ready enthusiasm and interest in the preparation of the text. Thanks also to all of my colleagues, from whom I have learned so much. I am particularly grateful to my wife, whose enduring patience and support enabled me to work during those precious hours when I should have been with my family.

I am indebted to Drs Robert A. Fishman and Michael J. Aminoff for their critical advice and continued support, to Dr Henry J. Ralston, III for his critical review of the sections on neuroanatomy, and to Mrs Lynda Levy and Ms Trish McGrath for their superb secretarial skills and remarkably good humor during the preparation of the manuscript.

Finally, I am indebted to Richard Jones of Jones Medical Publications for his knowledge, wisdom, and quiet patience in guiding the manuscript to its completion.

Bruce O. Berg, MD
San Francisco
April, 1984

Authors

Bruce O. Berg, MD *Professor of Neurology and Pediatrics, Director of Child Neurology, University of California, San Francisco.*

Suzanne L. Davis, MB, ChB *Assistant Professor, Neurology and Pediatrics, University of California, San Francisco.*

Michael S. B. Edwards, MD *Associate Professor, Neurological Surgery, University of California, San Francisco.*

Richard M. Flower, PhD *Professor and Vice Chair, Otolaryngology, University of California, San Francisco.*

Bryan D. Hall, MD *Associate Professor of Pediatrics, University of Kentucky, Lexington, Kentucky.*

Creig S. Hoyt, MD *Professor of Ophthalmology and Pediatrics, University of California, San Francisco.*

Thomas K. Koch, MD *Instructor, Neurology and Pediatrics, University of California, San Francisco.*

Seymour Packman, MD *Assistant Professor, Pediatrics, University of California, San Francisco.*

Lawrence H. Pitts, MD *Chief, Neurosurgery Service, San Francisco General Hospital; Associate Professor, Neurosurgery, University of California, San Francisco.*

James F. Schwartz, MD *Professor of Pediatrics and Neurology, Emory University School of Medicine, Atlanta, Georgia.*

Roger P. Simon, MD *Associate Professor, Neurology, University of California, San Francisco; Chief, Neurology Service, San Francisco General Hospital.*

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Neurologic Examination

Bruce O. Berg, MD

The clinical evaluation of children with symptoms referable to the nervous system demands complete attention to eliciting and interpreting historical details and performing a careful, complete neurologic examination. The history suggests the nature of the pathophysiologic process, and the examination localizes the site of the lesion.

There is a variety of clinical methods available, but each physician must develop his own method of performing the neurological assessment. One must be able to accommodate to and control each clinical situation—the loquacious historian as well as those of parsimonious prose. Above all *it is essential to listen to what the mother and child are telling you.*

The prenatal, perinatal, and developmental histories as well as the present illness are important to understand the nature of the patient's problem, and the wise clinician will not overlook the medical history of the biological family. The value of an accurate history in solving the clinical problem cannot be overemphasized.

The method of assessing the **mental status** of older children is similar to that of adults. In the case of very young patients, however, a developmental history will lend much pertinent information.

The patient's clothing should be removed so that abnormal physical findings are less likely to be overlooked. Allow the child to disrobe privately with his parents and have him wear an examining gown or remain in his underwear. A gentle but firm approach can be amazingly successful.

THE SKULL

The skull is readily available to inspection, palpation, auscultation, percussion, and measurement. The head circumference is a reliable indicator of intracranial volume. During the first 3 months of life, the full-term infant's head circumference should increase by 2 cm each month; from the third through sixth months, 1 cm each month; and from 7-12 months 0.5 cm each month (see Appendix).

Percussion of the skull is a useful diagnostic maneuver for in the presence of increased intracranial pressure an abnormally tympanitic sound may be elicited (McEwen's sign, 'cracked pot sound'). **Auscultation** of the skull may also provide useful information; cephalic bruits may be heard over the region of vascular malformations, tumors, or other space-occupying lesions that may compress large vessels. One should listen for bruits over the closed eye, the temporal fossae, and carotid arteries. Cepha-

lic bruits are commonly heard in skulls of normal infants and young children; 60% of 4 to 5-year old and 10% of 10-year old children. One should be cautious, however, in their interpretation, particularly if the bruit is localized to one area of the skull or is obliterated by compression of the ipsilateral carotid artery.

Transillumination is a valuable examination technique that is performed with a Chun gun transilluminator or a flashlight utilizing two type D batteries. The Chun gun is preferred because it emits a standard light source of high intensity. Rubber adaptors are used with each of the light sources so that good contact is made between the rubber ring and the scalp. The procedure should be carried out in a darkened room after the examiner has accommodated to the darkness. Circumstances that may alter transillumination include the gestational age of the infant, extracranial and intracranial factors.

The degree of transillumination is greater in infants of decreased gestational age and there is about 1 cm decrease in frontal transillumination from about 26 weeks' gestation to term.

Extracranial conditions that increase transillumination include paucity of hair, decreased pigment of skin and hair or a collection of fluid between the skull and the light. Conditions that decrease transillumination include an abundance of hair with increased pigment of hair and skin, increased thickness of skull, or a subgaleal hematoma.

Intracranial conditions which increase transillumination include increased collections of fluid in the subdural or subarachnoid space or in those patients with thin cortical mantles and large ventricles.

Configuration of the skull may often lend a clue to the diagnosis, especially in patients with large or macrocephalic heads. The skull of a hydrocephalic child is commonly globular shaped; biparietal widening is often seen in patients with chronic subdural hematomas, and in patients with Dandy-Walker syndrome there is usually a ballooning of the posterior aspect of the skull. Other forms of skull irregularities are found in different types of craniostenoses, depending upon what sutures are prematurely fused.

Microcephaly is secondary to abnormally decreased brain growth and has been arbitrarily defined as a head circumference greater than 2 standard deviations below the mean for age, sex, and gestation. It does not always presage mental retardation, for about 2-7% of children who are considered microcephalic by definition have normal intelligence.

THE SPINE

The spine should be inspected for its configuration, such as abnormal curvature of scoliosis, khyphosis, or lordosis, and also for the presence of dermal sinus tracts. Dimples or dermal sinuses may overlie a vertebral anomaly that is otherwise asymptomatic. Areas of localized tenderness to percussion and/or palpation may indicate an intraspinal lesion.

Table 1-1 The Clinical History

Complaint (reason for referral) What is the problem as perceived by the parent and/or child.

Family history (biological family)

Mother's gestational history List each pregnancy in order; abortions still births, living children (ages, sex, medical status; if deceased, what was the cause of death).

Mother
Father
Maternal grandmother and grandfather
Mother's siblings (brothers, sisters)
Paternal grandmother and grandfather
Father's siblings (brothers, sisters)

} Age, medical status, occupation; if deceased, what was the cause of death.

Identify and describe any family member with neuromuscular disease, convulsive disorder or migraine, visual or hearing problems. Describe any family member who is mentally ill, mentally retarded, or who has a specific learning disability.

Past medical history

Birthdate. Birthweight

Describe gestational history: note duration of pregnancy — if there were trauma, infection, or vaginal bleeding. List medications taken during the pregnancy, including alcohol, and use of tobacco. Describe the labor, its duration, and whether the delivery was vaginal or by cesarian section.

Neonatal period Describe the infant's status at birth; note the Apgar score, condition while in nursery, and the duration of hospitalization.

Developmental history Age at which the following were achieved:

Head support; smiled; rolled over; sat alone; stood/cruised; walked alone; acquisition of language.

Diseases Describe illnesses; age at occurrence, duration, and possible sequelae.

Surgery List any operative procedure, the age of child when it occurred, and possible complications.

Trauma Describe what happened, the child's age when it occurred, and whether or not there was an alteration of consciousness, and if there were sequelae.

Present illness Describe the problem in appropriate chronology and detail.

Table 1-2 The Neurologic Examination

General Describe the physical habitus of the patient.

Weight: Height: Head circumference:
Blood pressure: Respiration: Pulse: Temperature:

Speech Describe the quality of speech — presence of dysarthria.

Skull Configuration, auscultation, percussion.

Spine Describe configuration, deformity, tenderness, or limitation of movement. Nuchal rigidity usually implies meningeal irritation. *Kernig's sign* is characterized by the involuntary flexion of the knees when the examiner gently flexes the thigh while the leg is in extension. *Brudzinski's sign* is the flexion of the knees and hips following the flexion of the head on the chest.

Mental status Note the method of assessment.

Cranial nerves

I. Olfaction Note the method of assessment.

II. Visual fields Note the method of assessment.

Visual acuity O.D. O.S.

Fundi Describe media, disks, vessels, retina, and macula.

III, IV, VI. Pupils — size, configuration, reactivity to light and accommodation.

Extraocular motility — abnormal eye movements.

V. Corneal reflex Motor and sensory components — describe method of testing.

VII. Facial mobility Symmetry of facial mobility.

Taste Anterior 2/3 of tongue, note method of assessment.

VIII. Hearing Describe method of testing: Rinne test; Schwabach test; Weber test.

IX, X. Position and movement of palate and uvula; gag reflex. Describe sensation of posterior pharyngeal wall; phonation.

XI. Describe the bulk and power of sternocleidomastoid and upper fibers of trapezius muscles.

XII. Tongue — position, mobility, power, presence or absence of fasciculations.

Table 1-2 (cont'd) The Neurologic Examination

Motor system Describe the muscle bulk; presence or absence of fasciculations. Describe the muscle tone and power.

Tendon reflexes

Biceps: C5-6

Supinator: C5-6

Triceps:C6-8

Superficial abdominal:
Upper: T7-9
Lower: T11-12

Patellar: L2-4

Achilles reflex: L5-S2

Plantar response (sign of Babinski): L4-S2

Station and gait Describe the quality of movement — whether there is truncal and/or limb ataxia or a specific disorder of movement.

Sensory system Pain and touch. Temperature. Vibration. Joint position sense. Higher cortical function: two-point discrimination; stereognosis; graphesthesia.

Sphincters

General examination

Cardiovascular system

Respiratory system

Abdomen

Skin

CRANIAL NERVES

I. OLFACTORY NERVE

Neural pathways The ability to perceive the vast array of smells in the environment is a primitive function dependent upon this first cranial nerve. First-order neurons are bipolar cells with ciliated distal processes located in the superior aspect of the nasal cavity. The axons penetrate the cribriform plate of the ethmoid bone and synapse within the olfactory bulbs. The axons of the next-order neurons traverse the olfactory tract to the olfactory tuberculum, dividing into medial and lateral striae with some fibers crossing the anterior commissure to the contralateral olfactory bulb. Fibers of the medial striae pursue a course to the medial hemispheric surface; whereas, those fibers of the lateral striae follow an oblique course around the anterior perforated substance to terminate in the prepiriform, periamygdaloid and entorhinal cortex. Because of bilateral hemispheric innervation, lesions proximal to neuronal decussation do not result in the loss of smell (anosmia). Olfactory impulses that reach the amygdala are thought to be related to maternal and sexual behavior but the full importance of these neurobehavioral relationships is not known.

Clinical evaluation of the olfactory nerve requires an unobstructed nasal cavity, free of mucus or inflammation. The common cold is the most frequent cause of impaired olfaction (hyposmia). Each nostril should be tested separately by gently compressing one nostril and presenting to the other a nonirritating volatile substance such as cinnamon, cloves, oil of wintergreen, or lavendar. Irritating substances such as camphor, ammonia, or formaldehyde should be avoided, for they may stimulate gustatory end-organs or fibers of the trigeminal nerve.

The ability to perceive smell appears in early infancy, as demonstrated by an infant's recognition of the odor of his mother's used breast pads, compared to breast pads of other mothers or unused pads. Infants also appear to recognize and sense differences between pleasant and unpleasant smells.

Causes of anosmia include trauma to the ethmoid bone or cribriform plate which may also result in a CSF leak. Other causes of impairment of smell include meningitis, hydrocephalus, the uncommon brain tumor of childhood located in this region of the olfactory neural pathway, and illicit drug sniffing.

II. OPTIC NERVE

This cranial nerve subserves the sense of vision and is the one cranial nerve that can be examined directly, at least in part by ophthalmoscopy. Primary modalities of optic nerve function include the visual fields, visual acuity, color vision as well as day and night vision. Each eye should be examined separately.

Neural pathway The site of the lesion causing a visual field defect is best understood by reviewing the neuroanatomy of the visual pathways (see Figure 1-1).

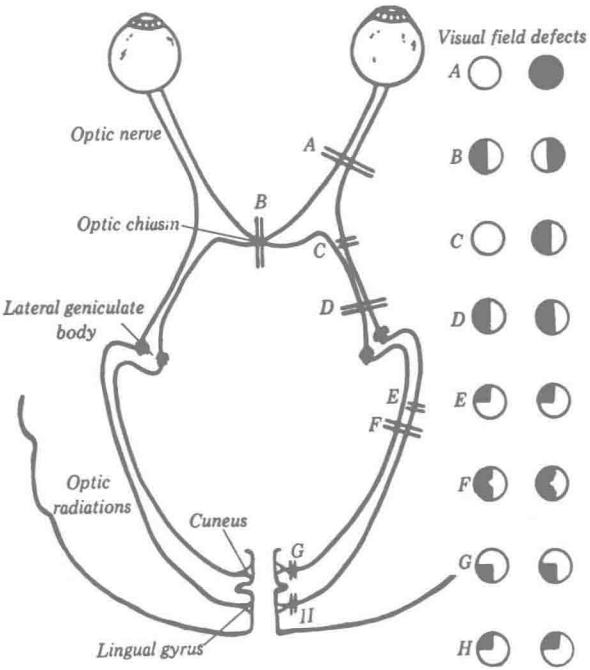

Fig. 1-1 The visual pathways. A-H show sites of potential lesions and associated deficits in the visual fields (black areas)

Clinical evaluation The *visual field* represents the perceived space subtended by one eye when that eye is fixed on some point. That point of fixation is projected on the macula while the other objects within that visual field are also percieved, particularly if in motion. In addition to white test objects, color test objects should be used to determine visual fields, for changes of the color field perimeter usually precede obvious changes of the visual fields (color desaturation).

The visual fields are examined by one of several methods including confrontation, using a tangent screen or by perimetry. Overlapping of the right and left visual fields enables one to have binocular vision; there is a crescentic monocular segment in each temporal field.

The method of confrontation compares the visual field of patient with that of the examiner, each of which is seated about 3 feet (1 meter) from the other. The patient closes the right eye while the examiner closes his left eye; each person gazes into the other's open eye. The examiner then introduces a white-headed pin, or similar object, into the peripheral visual field in a plane equidistant from both parties, documenting the point when the patient first sees that test object. This method, though somewhat crude, is very useful and can be readily accomplished at the bedside. The blind spot can also be demonstrated by this method.

When using a tangent (Bjerrum) screen, the three-dimensional visual field is projected upon a flat surface and that portion of central vision is examined. This method of examination is particularly useful in determining the overall dimensions of the visual fields, the configuration and size of the blind spot, as well as central visual defects (scotomata).

A variety of perimeters is available to determine visual fields. The patient sits before the instrument, looking at the fixation point; test objects are introduced into the visual field in one of the multiple meridians. When that object is first perceived by the patient, that point is recorded on a graph; the points of each meridian are connected for each test object and the perimeter of the visual field is established (the isopter). There are different isopters for each size of test objects.

Older infants and young children may be tested by introducing a dangling bright object, such as a shiny tape measure, from behind the patient's head and slowly introducing it into the field of vision. The child's eyes will quickly dart to the test object when initially seen. A normally intelligent child of 5 or 6 years can be tested by the confrontation method but it is usually not until the latter part of the first decade that perimeters are reliably used.

Visual acuity is a measure of the resolution power of each eye and should be evaluated for near and distant vision. Near vision is determined by presenting standard characters (Jaeger test) at a distance of 14 inches (35.5 cm) from the eyes. Distant vision is examined by presenting to the patient a set of standard letters of varying sizes (Snellen chart) that can be normally read at distances ranging from 10-200 feet. This examination is carried out by presenting the stimulus (letters) at 20 feet (6 meters) from the patient. Normal vision is determined arbitrarily as the ability to read standard letters at a distance of 20 feet; namely, 20/20 (6/6). The numerator is the distance of the patient from the chart and the denominator is the distance at which the smallest letter as read by the patient would be read by one with normal vision.

Color blindness, a heritable disorder primarily affecting males, may be total or partial. The usual method of examination of color vision is by presentation of pseudoisochromatic plates of Isahara, or other available standard test color cards. Day blindness (hemeralopia) and night blindness (nyctalopia) are considered in Chapter 13.

III, IV, VI. (OCULOMOTOR) NERVES

Neural pathways These cranial nerves are closely related functionally and neuronatomically. The *third nerve nuclei (oculomotor)* are located just below periaqueductal grey matter of the mesencephalon ventral to the aqueduct of Sylvius. This nerve innervates the inferior, medial, and superior recti muscles as well as the inferior oblique and levator palpebrae muscles. It contains parasympathetic fibers that innervate the intrinsic muscles of the eye, the ciliary muscle and sphincter of the pupil via the ciliary ganglia. The *fourth cranial nerve (trochlear) nuclei* are ventral to the aqueduct in the lower mesencephalic grey matter just rostral to the pons. This nerve, the smallest of all cranial nerves, innervates the superior oblique muscle. The *sixth nerve nuclei (abducens)* originate in the grey matter ventral to the fourth ventricle within the dorsal tegmentum of the

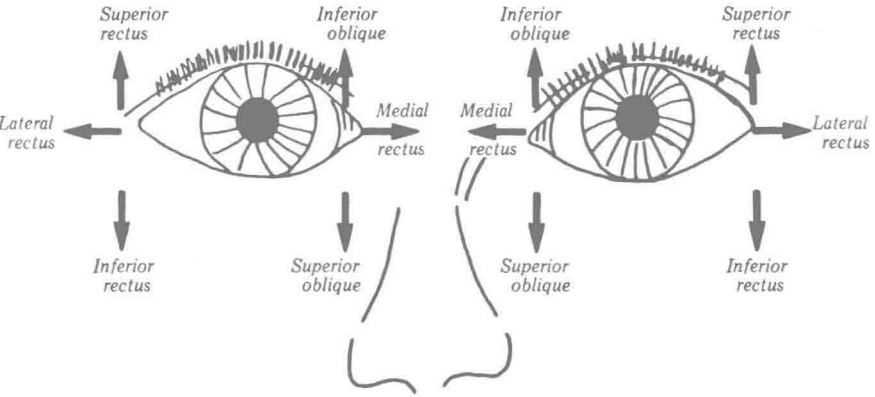

Fig. 1-2 The action of extraocular muscles attached to the globe in primary position, abduction, and adduction.

lower pons and innervate the lateral rectus muscles. These three cranial nerves regulate ocular motility, pupillary constriction, and some lid function (see Chapter 13).

V. TRIGEMINAL NERVE

Neural pathways The fifth and largest of all cranial nerves carries both sensory and motor fibers and has complex connections with other cranial nerves. The Gasserian ganglian, the cellular origin of sensory fibers, lies adjacent to the apex of the petrous bone in the middle fossa. Proximal fibers follow a course into the pons; the distal fibers are divided into three main sensory divisions — the ophthalmic, maxillary, and mandibular, which leave the skull through the superior orbital fissure, the foramen rotundum, and the foramen ovale, respectively. The motor root emerges from the pons below the Gasserian ganglian and leaves the skull through the foramen ovale, joining the mandibular root for a short distance and then divides to supply the muscles of mastication.

Sensory fibers carry impulses to three primary nuclei within the pons and medulla. Large diameter fibers primarily carry tactile information to the main sensory nucleus where, after synapse, axons ascend in the dorsal ascending trigeminal tract to the thalamus. The nucleus of the descending root (trigeminal spinal tract) extends caudally from the main sensory nucleus to a level of C4. Smaller diameter pain, temperature, and tactile fibers terminate on the cells of this nucleus. Fibers of the ophthalmic division are ventrolateral, mandibular fibers are dorsomedial, and maxillary fibers are in between. Exteroceptive fibers of VII, IX, and X join this descending tract at the levels at which they enter the brain stem, descend for varying distances, synapse, and neuraxons of the next order cross the midline to ascend in the ventral ascending trigeminal tract, and terminate in the thalamus. The course of the mesencephalic sensory root of the trigeminal nerve is closely associated with the motor root and carries proprioceptive impulses from muscle. Muscles innervated by the trigeminal nerve