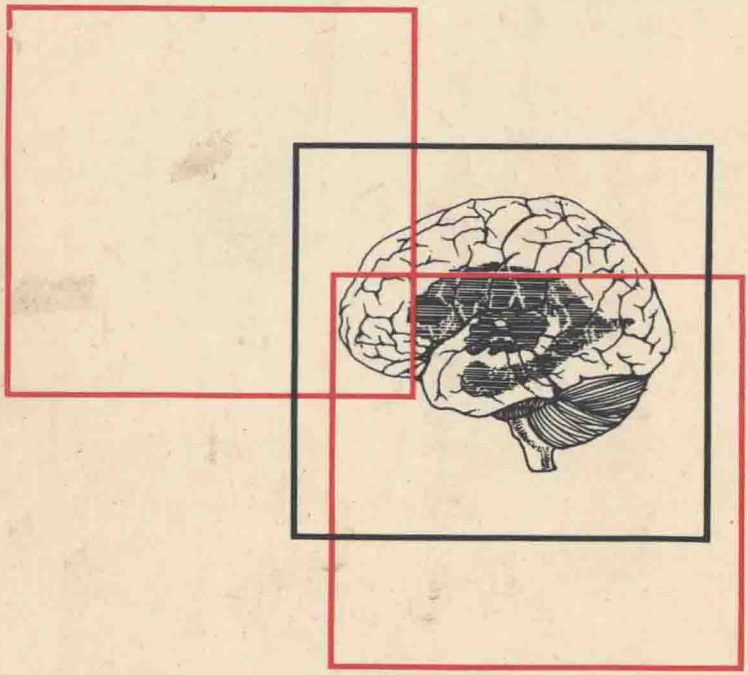
CHILDHOOD NEUROLOGICAL PROBLEMS

A Textbook for Health Care Professionals



Doris A. Trauner

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A Textbook for Health Care Professionals

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To my husband Dick –
for everything

FOREWORD

An understanding of pediatric neurology is essential to the understanding and management of a large number of children with a variety of problems. This follows from the central position of the nervous system in directing other functions and its vulnerability to the effects of disease, injury or toxin. All of us who take care of children need to know a certain amount of pediatric neurology. I have long been interested in having a concise source of information that covered the breadth of the field. Doctor Trauner has provided us with just this. This book was written with a broad audience of health care professionals in mind—nurses, physiotherapists, speech pathologists, anyone who deals with children with problems involving the nervous system. I believe that the book will also be useful to physicians in family practice and pediatrics.

Dr. Doris Trauner is a dedicated physician, concerned daily with the problems of children with neurologic disorders. She has done original work in pediatric neurology from the time she was a medical student at the Medical College of Virginia. Doctor Trauner is now Assistant Professor of Neurology and Pediatrics at the University of California San Diego and is one of the world's authorities on Reye's syndrome.

Among her many talents is Dr. Trauner's real skill as a teacher. She is good at making complicated things understandable to students and to the rest of us. I feel that in this book she has selected the important issues in pediatric neurology and that she has presented them with the clarity that will make this book useful to a broad audience of health care professionals.

WILLIAM L. NYHAN, M.D., PH.D.
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PREFACE

MANY PROFESSIONALS are involved in the care, education and rehabilitation of children with neurologic disorders. Physical, occupational and speech therapists, school nurses, teachers and educational psychologists – all must deal with the special problems that such children present. Understanding of neurologic dysfunction and its implications for the child's future life sometimes is incomplete. Many of the afflictions of the nervous system are rare and may be new to the person called on to deal with the results. Often, medications have been prescribed that may alter the child's educational or therapeutic response. A reference source that deals with these problems would be a useful guide to all those involved in the care of such a child.

During the time that I spent as a pediatric neurology consultant at the Brandecker-Ridge Easter Seals Treatment Center in Chicago, Illinois, I became aware of a problem. Personnel at the Center were dedicated and well trained and desired to know more about their patients' diagnoses in order to treat them most effectively. But we could find no single text that discussed neurologic disorders, treatment and outcome in a concise, readable and thorough manner. This book was undertaken partly through the impetus of these dedicated people.

The purpose of this text is to provide professionals involved in the care of the neurologically disabled child with a comprehensive guide to childhood neurologic problems; what they mean to the patient, family and professional; and a discussion of treatments used for the more commonly encountered disorders. It is hoped that with the expertise of the professionals who read this book, such children will have a brighter future.

Numerous people have been involved in the preparation of

this book, either directly or indirectly. Dr. William Nyhan, Chairman of the Department of Pediatrics at the University of California, San Diego, has been a continuing source of wisdom, guidance and enthusiasm ever since my internship in his department. Dr. Peter Huttenlocher, my mentor in pediatric neurology, shared willingly his tremendous knowledge of this field. His dynamic teaching and love for pediatric neurology have attracted many of his students into this specialty.

My good friends and professional associates in the Communicative Disorders Center at UCSD, with Mrs. Carol Grote as Director, have taught me much about rehabilitation of children with neurologic handicaps. The importance of their work is too often underestimated, but the children they treat are well aware of the benefits of rehabilitative therapy.

Ms. Valerie Lockhart spent many long hours in manuscript preparation, managing to do an excellent job despite constant interruptions. Mrs. Beverly Gonsowski donated her time to this effort as well.

I want to thank Drs. William Nyhan, Hector James, Marjorie Seybold, Ray Skoglund and James Connor, Mrs. Carol Grote and Susan Lingle for reviewing certain chapters and for their constructive criticisms. A very special note of thanks goes to Dr. Paul Schultz for his thorough and constructive review of the entire manuscript.

All the drawings in the text were done by Ms. Penelope Roberts.

DORIS A. TRAUNER, M.D.

CONTENTS

1

NORMAL NEUROLOGIC DEVELOPMENT

ANY UNDERSTANDING of the child with neurologic problems must begin with some knowledge of normal neurologic development. This chapter will offer a brief discussion of brain anatomy. With that background, we then will turn to normal development from birth to early school years.

The brain is a complex structure, both anatomically and functionally. Anatomic examination of a brain reveals that it is divided into several parts (Fig. 1–1). The earliest area to develop is the brainstem, which is located at the base of the brain. This structure contains fiber tracts connecting other parts of the brain with the spinal cord; groups of cells called cranial nerve nuclei, which are responsible for such functions as smell, movement of eye and facial muscles, pupillary responses and hearing; and vital centers for control of heart rate and respiration.

The spinal cord projects downward from the brainstem and is encased in a bony covering, the vertebral column or spine. The spinal cord sends out projections from nerve cells to muscles and joints, and carries nerve fibers which control bowel and bladder function as well as sensation and strength, under control from cells inside the brain.

Immediately adjacent to and partially covering the brainstem is the cerebellum, which is a center for muscle tone and coordination.

The cortex is located above the brainstem, and in humans makes up the largest part of the brain. It is divided into right and left cerebral hemispheres and contains many folds. Each indentation on the surface is called a *sulcus*, and the section of cortex between two sulci is called a *gyrus*.

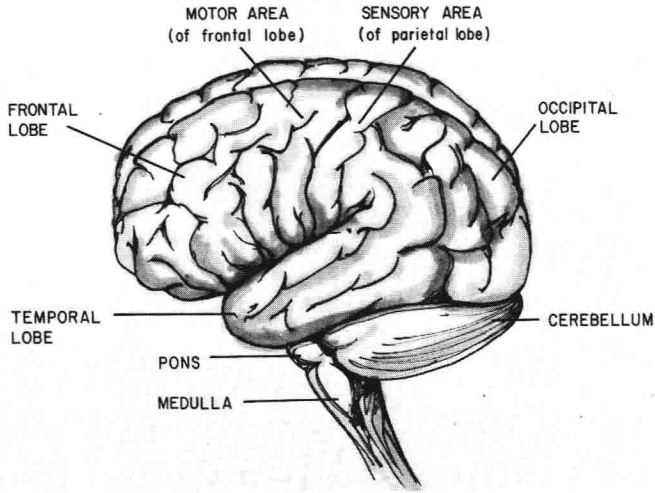


Fig. 1–1.—Diagrammatic representation of the brain and its major divisions in lateral view.

The cortex is further divided into lobes: frontal, parietal, temporal and occipital. Each lobe has certain primary functions to carry out, e.g., the frontal lobe is responsible for motor control.

In the interior of the brain there is a connecting system of fluid-filled spaces called *ventricles*. These contain cerebrospinal fluid, which bathes the brain.

The entire brain is surrounded by a thin layer of fluid and then by several thin coverings called *meninges*. Blood vessels course throughout the brain itself as well as over the surface, carrying necessary nutrients to brain cells.

The brain is encased in a bony covering—the *cranium*, or skull. The cranium is made up of several bones that are separate from one another at birth but which fuse over the first few years of life to form a solid bone.

The points at which two cranial bones meet are called *sutures*. In the infant there are two roughly diamond-shaped gaps between the bones called the *anterior* and *posterior fontanelles*. The posterior fontanelle closes soon after birth but

the anterior fontanelle ("soft spot" on top of the head) remains open until 12–18 months of age.

The major part of brain growth occurs in the first year of life. By the end of the first year, the brain is approximately two-thirds of adult size and by the end of the second year is about four-fifths that of the adult brain.

Any abnormality (such as infection, biochemical imbalance, trauma) that upsets the brain's development and growth during this period can exert profound effects on eventual brain function. Therefore, early neurologic evaluation of potential difficulties is important in assessing the nature and extent of any problem and in instituting treatment, when available, to prevent further damage to the growing brain.

Head size reflects brain size; if the brain is damaged and stops growing, the bones have no stimulus to grow and the individual bones of the skull will fuse prematurely, causing a small head, or *microcephaly*. Conversely, if there is increased pressure in the brain, as from an obstruction to fluid reabsorption or an increase in production of fluid, the head will grow more rapidly than normal and produce hydrocephalus. The normal head circumference for the full-term newborn infant is 35 centimeters (13¾ inches).

BIRTH TO 6 MONTHS

In the newborn, higher cortical function has not yet developed and most of the neurologic examination is directed toward assessment of the more primitive parts of the nervous system. The normal newborn can perform most basic functions, but at times to a limited degree. For example, the newborn infant blinks at bright lights, and pupils will constrict if a bright light is held near the eyes. The infant will also turn toward a light and blink at a loud sound.

The newborn infant is able to yawn, sneeze, cough and hiccup. All of these reflex functions require an intact brainstem. The infant responds to pain by withdrawal of the stimulated area and issues a cry of discomfort. In a prone position he can lift his head slightly and turn it from side to side.

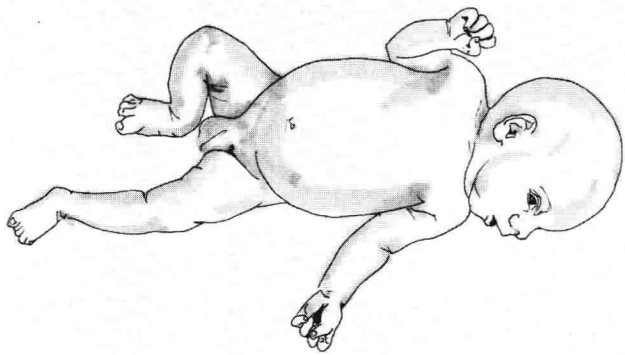
Certain reflexes are present in the normal newborn infant

that disappear as the brain matures. These reflexes are conducted through the more primitive parts of the brain and are inhibited as the more advanced cortical functions and voluntary control take over. Persistence of these immature reflexes after the normal age of disappearance of each reflex indicates some cortical dysfunction. Three of these reflexes are described here along with the method used to elicit them.

TONIC NECK REFLEX (Fig. 1-2).—With the infant lying on his back, the examiner turns the head briskly to one side. On the side to which the face is turned, the arm and leg extend, while the limbs on the opposite side flex at the same time. This reflex usually is incomplete in the mature newborn and disappears at about the fourth month of life. It is necessary to be able to overcome this reflex before the infant can roll over and sit up. An asymmetry in the reflex or persistence past 4-6 months of age, or a complete reflex in a mature infant, suggests a cerebral abnormality.

MORO REFLEX (Fig. 1-3).—This reflex is induced by making a loud noise near the infant or by supporting the back and dropping the head slightly. It is characterized by generalized extension of all four limbs followed by flexion of the limbs. The fingers fan out and the infant cries. This reflex usually is present in the full-term newborn and gradually disappears over the first 4-6 months of life.

Fig. 1-2.—Tonic neck reflex in a normal infant.



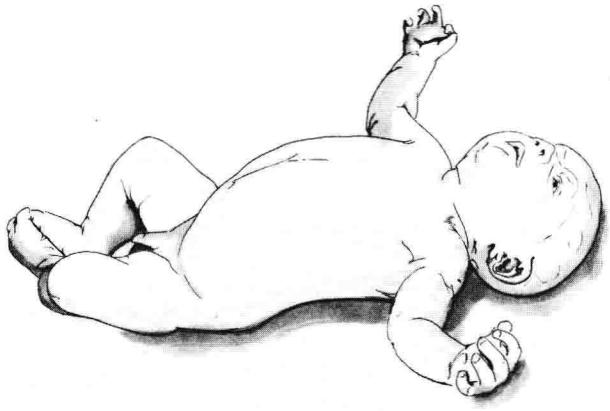


Fig. 1–3.—Moro reflex.

PLACING AND STEPPING REFLEXES.—If the infant is held upright and the feet are allowed to touch the table, a newborn infant will have automatic stepping reflexes. If the infant is lifted suddenly and the feet again are placed on the table, the baby will have an extensor thrust of the legs to support him-

TABLE 1-1.—ACQUISITION OF SKILLS DURING FIRST YEAR OF LIFE

Newborn	Fixes on light
	Grasp reflex
	Moro reflex
3 months	Social smile
	Good head control
6 months	Rolls over
	Reaches for and transfers object
	Sits with minimal support
	Laughs out loud
9 months	Pincer grasp
	Sits alone
	Stands with support
	Crawls
	Makes repetitive sounds ("da-da")
12 months	Walks with minimal support
	Crawls
	Uses 3-4 words
	Imitates behavior (waves "bye-bye")
	Plays peekaboo

self. These reflexes disappear as the child begins voluntary walking during the first year of life.

The acquisition and disappearance of these reflexes and other developmental milestones now will be discussed in chronologic order (Table 1-1).

DEVELOPMENTAL MILESTONES FROM BIRTH TO 6 MONTHS OF LIFE

If the newborn infant is placed face down (prone) on the table or bed he is able to turn his head from side to side. By 4 weeks of age he can lift his head above the surface of the bed. The newborn is also able to fix on a light or a bright object in the first days of life and is able to follow it with his eyes for a few degrees. By the end of the second month he can follow the light all the way to the right or left. In the first 4-8 weeks of life, if the infant is pulled from lying to sitting, the head lags (Fig. 1-4); in the upright position, head control is poor. By 12 weeks of age there is some head control while being pulled to sitting, but the head may not be fully upright when the infant is placed in the sitting position.

A normal infant displays a grasp reflex; that is, if an object is placed in the palm of his hand, he will automatically grasp it. This reflex persists until about 8 weeks of age, after which voluntary grasp begins to take over. By 12 weeks of age, the child should attempt to grasp an object that is held near him and may hold it briefly if contact is made with the hand.

By 8 weeks of age, most infants have a social smile; by 3 months, the child makes some noises indicating pleasure on social contact. A social smile should be present by 12 weeks of age.

By 3 months of age, the child who is lying face down can lift his head and chest off the surface of the bed with his arms extended in front of him. By 4 months of age he is able to raise his head to an upright position and turn it from side to side. Between 3 and 4 months, the infant begins to bring his hands to the midline or to the mouth. At this time, the child also begins to make contact with objects brought within his reach and may bring these to the midline or to his mouth to explore

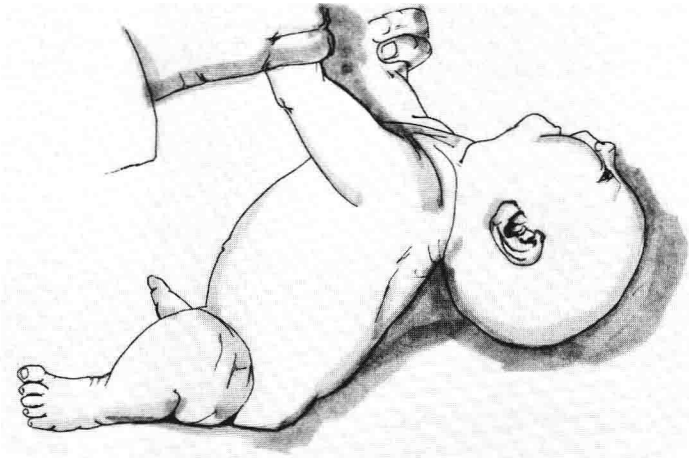


Fig. 1–4.—Normal head lag in a 2-week-old infant.

them. There no longer is any head lag when the child is brought to a sitting position, and the head is held steady and upright without bobbing when the child is standing or sitting. The infant also begins to enjoy being supported upright and is much more attracted to objects in his environment. He is beginning to be able to look around and to grasp an object of moderate size, such as a rattle or a ball.

By 5–6 months, the child has begun to roll over, first from front to back and then in the reverse position. By 6 months he often is able to sit alone, leaning forward on his hands or with only slight support at his back. At this age he can also use his thumb to oppose his fingers and can grasp an object and transfer it from one hand to the other. He now can be pulled from a sitting to a standing position and will support his weight on his extended legs.

A child of this age is much more responsive to social contact and he will laugh out loud when given attention or he may show signs of displeasure if pleasant contact is terminated. He begins to show preference for the person giving him most of his care.

At the end of 6 months, the normal infant no longer has tonic neck reflexes or a Moro reflex and no longer has a reflex grasp.

The child now can roll over and sit, at least with support, has good head control and reaches for and transfers objects from hand to hand. He is social and laughs out loud and at times makes cooing noises.

6–12 MONTHS

By this time, the normal infant is much more interested in his immediate environment and in his own body. The child will begin to show interest in his legs and feet and will play with them when lying on his back. Between 6 and 9 months, the hand movements become much more coordinated in that the infant can use his thumb and forefinger in a pincer grasp to get smaller objects. By 8–9 months he can sit up without assistance and by 9–10 months is beginning to creep and crawl. At this age he may be able to stand for a few seconds if the hands alone are supported and by 9 months he can take a few steps with the hands held.

Speech development progresses rapidly at this age also. The infant of 8 months makes repetitive sounds such as ma-ma and da-da. At this age he recognizes his own name and by 1 year of age he can show by his behavior that he knows the names of some objects. By 1 year of age he may meaningfully use 1–4 words other than ma-ma and da-da.

During the second 6 months of life, the infant shows some degree of social differentiation, preferring his mother and reacting negatively to strangers. There is some degree of dependence on his mother, although as the infant becomes more mobile there is less dependence on her physical presence. At this age, the child also becomes aware that an object that is covered up still is there, and peekaboo becomes an enjoyable game.

Imitative behavior begins during this time. The child may begin to imitate waving bye-bye and throwing a ball.

By 1 year of age, the infant has made considerable progress in gaining independence. He can walk or at least crawl, use a few words, laugh and play. The nervous system has matured tremendously and the child is ready to acquire the more sophisticated motor skills of the second year.