With pleasure and appreciation we dedicate this book to our spouses and children, who made it possible for us to bring this text to fruition and who taught us what was really important in development over the lifespan. It is impossible to adequately describe the value of their encouragement and support. We also would like to express our appreciation to the Elsevier editorial staff who are extremely talented and helpful in keeping track of all the details necessary to complete such a task as well as their judicious editorial skills. Furthermore, no dedication of a book embracing this field would be meaningful without a tribute to the courage and perseverance of neurologically impaired children and their caretakers.

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Swaiman's Pediatric Neurology

Principles and Practice

SIXTH EDITION

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Preface to the First Edition

It is concurrently tiring, humiliating, and intellectually revitalizing to compile a book containing the essence of the information that embraces one's life work and professional preoccupation. For me, there is a certain moth-to-the-flame phenomenon that cannot be resisted; therefore this new book has been produced.

Pediatric neurology has come of age since my initial interest and subsequent immersion in the field. Concentrated attention to the details of brain development and function has brought much progress and understanding. Studies of disease processes by dedicated and intelligent individuals accompanied by a cascade of new technology (e.g., neuroimaging techniques, positron emission tomography, DNA probes, synthesis of gene products, sophisticated lipid chemistry) have propelled the field forward. The simultaneous increase of knowledge and capability of pediatric neurologists and others who diagnose and treat children with nervous system dysfunction has been extremely gratifying.

Although once within the realm of honest delusion of a seemingly sane (but unrealistic) devotee of the field, it is no longer possible to believe that a single individual can fathom, much less explore, the innumerable rivulets that coalesce to form the river of knowledge that currently is pediatric neurology. Streams of information in certain areas sometimes peacefully meander for years; suddenly, when knowledge of previously obscure areas is advanced and the newly gained information becomes central to understanding basic pathophysiologic entities, a once small stream gains momentum and abruptly flows with torrential force.

This text is an attempt to gather the most important aspects of current pediatric neurology and display them in a comprehensible manner. The task, although consuming great energies and concentration, cannot be accomplished completely because new conditions are described daily.

The advancement of the field necessitated that preparation of this text keep pace with current knowledge and present new and valuable techniques. My colleagues and I have made every effort to discharge this responsibility. Because of continuous scientific progress, controversies are extant in some areas for varying periods; wherever possible, these areas of conflict are indicated.

This book is divided into four unequal parts. Part I contains a discussion of the historic and clinical examination. Part II contains information concerning laboratory examination. Chapters relating to the symptom complexes that often reflect the chief complaints of neurologically impaired children compose Part III. Part IV provides detailed discussion of various neurologic diseases that afflict children.

Although every precaution has been taken to avoid error, bias, and prejudice, inevitably some of these demons have become embedded in the text. The editor assumes full responsibility for these indiscretions.

It is my fervent hope that the reader will find this book informative and stimulating and that the contents will provide an introduction to the understanding of many of the conditions that remain mysterious and poorly explained.

Kenneth F. Swaiman, MD
Autumn 1988

Preface to the Sixth Edition

In 1975, a little over 40 years ago, the first two-volume reference text concerning Pediatric Neurology was published. In 1971, Dr Swaiman was approached by an executive editor of C.V. Mosby to discuss publishing a book on pediatric neurology based on papers he had read from a University of Minnesota Continuing Medical Education Course. This was a year before the first Child Neurology Society meeting and thus before the formal organization of pediatric neurologists. Drs. Ken Swaiman and Frank Wright began this project, immediately facing the challenge of delineating the field. At that time, the importance of neurochemistry and genetics was being emphasized, there were questions as to whether learning disabilities or autism were legitimate components of child neurology, and recruiting authors for various chapters was difficult as sub-specialties of the discipline were in their infancy or under- or undeveloped.

The first edition preface of *The Practice of Pediatric Neurology* stated, "*We have aspired to create a well-illustrated book that stresses the mainstays of modern pediatric neurology—the staggering array of neuromuscular and metabolic diseases described in the past 30 years, the relationship of embryology to congenital malformations, the growing number of recognized but yet unexplained degenerative diseases of childhood, and higher cortical function as related to learning capabilities of the child.*" The book and the subsequent 1982 edition were internationally well received. *Pediatric Neurology* and subsequently *Swaiman's Pediatric Neurology* were first published in 1989 and then in 1994, 1999, 2005 and 2012. Elsevier succeeded C.V. Mosby as the publisher in 2012. The growth of the discipline is documented by the fact that the number of pages and chapters has grown greatly: from 40 chapters/1082 pages in 1975 to 108 chapters/2290 pages in 2012.

This sixth edition of *Swaiman's Pediatric Neurology: Principles & Practice* reflects the remarkable increase in knowledge and complexity of the field since the fifth edition. Keeping abreast of all new information required us to increase the size and scope of this book from 108 to 170 chapters. To avoid publishing an overwhelming and oversized tome, we meticulously curated the most immediately necessary information and guidance in the print book, while providing the full text, our most comprehensive edition ever, online. We are proud of our mobile-optimized, downloadable e-book, which is included with your print purchase, and which provides easy and complete searchable and annotatable access to the content. Between the portable print book and the expansive online text, this reference offers a remarkable collection of well written chapters on topics of importance to professionals around the world who care for children with neurological disorders.

To accomplish these goals, we have increased the number of editors from four in the last edition (Ken Swaiman, Stephen Ashwal, Donna Ferriero, Nina Schor) to eight by adding four new and accomplished individuals with expertise in specific areas: Andrea Gropman (neurogenetics and metabolic disorders); Richard Finkel (neuromuscular disorders); Phillip Pearl (pediatric epilepsy); and Michael Shevell (neurodevelopmental disabilities). We also had some unofficial expert guidance for specific sections of the book on neurodevelopmental malformations (Bill Dobyns); pediatric movement disorders (Jon Mink); and pediatric neurooncology (Roger Packer).

Major changes in the book that the reader will find of interest include:

- Completely new sections on pediatric immune mediated nervous system disorders (4 chapters), cerebrovacular diseases (6 chapters), neurooncology (13 chapters), neuromuscular disorders (18 chapters), and clinical care care of the child with neurologic disorders (11 chapters).
- Major expansions of the sections on perinatal acquired and congenital disorders (7 chapters), neurodevelopmental disabilities (11 chapters), pediatric epilepsy (23 chapters), and nonepiletiform paroxysmal disorders and disorders of sleep (7 chapters).
- Three new chapters for the section on emerging concepts in child neurology including topics related to the developmental connectome, stem cell transplantation, and cellular and animal models of neurological disease.
- Updates of all remaining chapters by an international group of authors who are experts in their respective fields.
- Other new chapters in different sections of the book include: neuropsychological assessement; development of a neonatal neurointensive care unit; neonatal traumatic brain, spine and peripheral nervous system injury; an overview of the conceptual framework of the developmental encephalopathies; an overview of how to evaluate patients with a suspected metabolic disorder; a review of conditions associated with vitamin metabolism; and a chapter on nutrition and malnutrition and the developing brain.

We hope that the reader will find this book a useful resource and that the information will benefit the many children who suffer from these conditions. It is our wish that the greater world community will increase support for the care of neurologically impaired children and the research necessary to provide further understanding of, and improved treatment and preventive measures for, neurologic diseases. This support will improve the survival and quality of life of these brave children and their families.

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PART I

Clinical Evaluation

1 General Aspects of the Patient's Neurologic History

Kenneth F. Swaiman and John Phillips

An expanded version of this chapter is available on www.expertconsult.com. See inside cover for registration details.

When presented with a challenging patient in 1885, with students Josef Babinski, Sigmund Freud, and others looking on, Dr. Charcot used the most important tools he had at his disposal: a careful history and a detailed examination. If he were alive today, Dr. Charcot would use those same tools.

There is no substitute for an accurate and thorough history. The patient or parent begins with an explanation of his or her concern. In most medical settings this opening statement lasts less than 60 seconds if not interrupted, as it unfortunately generally is, by the physician (Beckman and Frankel, 1984). More focused questions follow as a differential diagnosis is developed. In some cases language can be a barrier, particularly with the growing multiculturalism in the United States and other countries, and it is important that the interview be conducted in the native language of the patient. This may require the use of interpreters. Using a nonprofessional or poorly trained medical interpreter should be avoided because this has a much higher risk of causing clinically significant errors than using an experienced professional interpreter (Flores et al., 2012). In addition, an effective medical interview requires eye contact to help establish patient rapport (Cole and Bird, 2014), and the near-ubiquitous presence of computers in examination rooms, with an electronic medical record at the physician's fingertips, can be a problem. Compared with using paper medical records, there is now significantly more time spent looking at the medical record (now on a computer screen) and less time looking at the patient (Asan et al., 2014). However, although there is no turning back on the electronic medical record, and the pressure to see more patients may be increasing, the balance of new technology is certainly favorable. Once adopted, the electronic medical record improves physician's productivity and portable telemedicine equipment allows access to virtually any language needed in a medical setting (Cheriff et al., 2010).

Arriving at an appropriate differential diagnosis is an active process. As information is obtained suggesting specific disease categories, further questioning helps narrow the possible diagnostic possibilities. This process is assisted by considering general groups of neurologic diseases. Memory aides are often used to recall differential diagnostic categories—VINDICATE and VITAMIN C are two common examples (Table 1-1). Overlap exists between categories, and within each disease category are multiple subcategories and then specific diseases that become more or less likely as the patient interview proceeds. For example, recurrent strokes from MELAS (a disorder of energy metabolism) could be considered both a vascular and a metabolic process. The important issue is that, if focal weakness is the presenting concern, then the patient interview continues in an effort to discern whether the weakness is recurrent or triggered by fever or dehydration (which might make a metabolic disorder more likely). Further questioning may disclose a history of lactic acidosis that narrows the differential diagnosis further to a possible mitochondrial disorder such as MELAS, which can then be confirmed with genetic testing. The point is to begin broadly. Cast a wide net. Consider all disease categories until a more focused differential diagnosis is possible based on the detailed history.

Part of clarifying the history of the current illness is to answer four basic questions:

1. Is the process acute or insidious?
2. Is it focal or generalized?
3. Is it progressive or static?
4. At what age did the problem begin?

The order in which disease findings develop and the precise **age of onset** of symptoms and signs may be critical factors in the process of accurate diagnosis. Many degenerative disorders have specific ages of onset that can help narrow the differential diagnosis; cognitive regression in an 8-year-old may raise the question of adrenoleukodystrophy, whereas in a 2-year-old one might consider neuronal ceroid lipofuscinosis. The presence of repeated episodes or associated phenomena should also be determined. For example, although the clinical manifestations of cerebrovascular events such as an acute stroke normally develop over minutes to hours, the underlying process may be long-standing; therefore, acute onset of vascular symptoms may be the result of a subacute or chronic process. On the other hand, infections, electrolyte imbalances, and toxic processes (such as exposure to over-the-counter drugs, prescription medications, insecticides, and other toxins found around the home) usually progress over a day to several days before maximum symptoms occur. More chronic are degenerative diseases, inborn metabolic disorders, and neoplastic conditions that usually progress insidiously over weeks to months.

Evaluation of whether a condition is **focal or generalized** is central to the diagnostic process. A focal neurologic lesion is not necessarily one that causes focal manifestations but is one that can be related to dysfunction in a circumscribed neuroanatomic location. For example, a focal lesion in the brainstem may cause ipsilateral cranial nerve and contralateral corticospinal tract involvement. If the problem is not focal, it usually results from a generalized process or from several lesions (i.e., multifocal). Neoplastic and vascular diseases frequently result in focal processes; occasionally, trauma results in such abnormalities. Generalized or multifocal conditions are usually associated with degenerative, congenital, metabolic, or toxic abnormalities.

In child neurology, it is particularly important that the clinician always attempt to determine whether the condition

TABLE 1-1 Disease Category Acronyms

Vindicate	
V	Vascular
I	Inflammatory
N	Neoplastic
D	Degenerative
I	Infectious
C	Congenital
A	Allergy or autoimmune
T	Trauma or toxin
E	Endocrine
Vitamin C	
V	Vascular
I	Infectious
T	Trauma
A	Autoimmune
M	Metabolic
I	Idiopathic
N	Neoplastic
C	Congenital

is **progressive or static**. This is best accomplished by taking a detailed developmental history. Documenting the age of acquisition of major motor, language, and social milestones allows characterization of a child's developmental progress compared with age-based norms. To address the critical issue of developmental regression, questions are posed that determine whether the child is no longer capable of motor or intellectual activities that were previously performed. This information is essential to the diagnosis of progressive disease, which is usually preceded by a period of normal development. Occasionally, prior formal neurologic and psychometric evaluations may be available that help enormously in providing objective documentation of prior developmental status. Reviewing family photographs, videos, baby books, or old Facebook postings can be helpful. In progressive conditions, such as those caused by metabolic or neoplastic disorders, documentation of increasing loss of normal function or an increase in any symptoms is essential. Static conditions can be the result of traumatic or anoxic injury, a congenital abnormality, or perhaps a resolving acute toxic ingestion.

Reviewing the **medical history** is an essential part of any interview and often provides information that is critical in arriving at an appropriate diagnosis, extending information gleaned from the history of current illness. In child neurology, medical history begins with the moment of conception. Was conception achieved with reproductive technology, the long-term consequences of which are not clear? Was the mother older than 35 years at the time of conception, a possible risk factor for adverse outcome? During gestation, were there exposures to toxins such as alcohol, nicotine, or prescription or illicit drugs? Was the pregnancy planned and was the mother healthy throughout gestation? At what point was fetal movement first noted (i.e., quickening), which should be at approximately 4 to 5 months' gestation, and did it continue throughout the entire pregnancy? Were there problems with poverty, nutrition, or exposure to violence and stress? Obtaining some of this important information could be difficult because of privacy concerns, and may require a confidential interview with mother.

Details of the delivery are critical and may not be entirely recalled by parents; therefore, reviewing birth records if available is important. Information such as duration of labor, medications or assistive devices (forceps and vacuum extraction) used, presence of meconium at delivery, and gestational age may have direct relevance to later brain development. The general status of the newborn immediately after delivery should also be understood. Objective information is important such as birth weight, Apgar scores, head circumference, and any neonatal complications encountered. Obtaining accurate information may require interviewing all caregivers, particularly with children whose mothers had pregnancy-related health problems and may have been sick. An enormously important piece of information that needs to be confirmed and not just assumed is the status of neonatal screening. Depending on the state or country of birth, most children are screened as a public health measure for certain treatable metabolic conditions. In the United States (http://www.babysfirsttest.org/newborn-screening/states) and Canada (http://raredisorders.ca/documents/CanadaNBSstatusupdatedNov.112010.pdf), it varies by state and province, but all screen for at least some disorders of amino acid metabolism, fatty acid oxidation diseases, organic acid conditions, and hemoglobin and endocrine disorders.

Family history is particularly important as many disorders encountered by the clinician may have a genetic basis. Begin by asking if any family members suffer from the same problems that affect the patient. Autosomal-dominant traits may be present in successive generations, although the degree of expressivity may vary. Autosomal-recessive traits often do not manifest in successive generations but may be present in siblings. Consanguinity must be considered when autosomal-recessive disease is part of the differential diagnosis, even if it is not forthcoming from interviewing parents (the incidence of false paternity has been reported to be from 1% to 30% depending on the population studied).

Cognitive and behavioral development is influenced by the child's home environment. A social history helps identify whether risk factors such as exposure to poverty, violence, parental depression, or bullying are present. Some clinics find the SEEK parent screening questionnaire helpful to identify potential challenges in the home environment (Fig. 1-2).

Caregivers should be questioned carefully about the nature and results of previously performed tests, including electrodiagnostic tests, brain-imaging studies, biochemical studies (e.g., quantitative assays of amino acids, organic acids, lactic acid, and lysosomal enzymes), biopsies, and chromosomal or gene studies. It is particularly important to review the full report of prior comprehensive genetic tests such as microarray analysis, exome sequencing, or genome-wide association studies (GWAS). Even when these tests fail to establish a clear diagnosis, they usually identify abnormalities of unknown clinical significance, and what was once unclear may have over time been found to have clear clinical implications. Therefore it may be helpful to perform an updated review of abnormalities identified on prior genetic testing. Use of prior medication should also be documented, including those medications that may have been prescribed but not taken, with note made of results of such therapies. If prior imaging has been performed, reviewing the study and not simply relying on the report is helpful to confirm findings (this is particularly important if the radiology interpretation is by someone without pediatric experience, or whose experience is not known as may occur with an on-call teleradiology service).

Thus, history taking is an active process. Beginning with a presenting complaint, the clinician broadly considers multiple diagnostic categories, which are narrowed as open-ended questions and followed by more specific queries. An exhaustive, all-encompassing neurologic history is impossible to obtain, particularly under the time constraints most clinicians face in today's environment. Therefore a skilled clinician is able to focus the interview on relevant information, often following up details that are more important than the patient or caregiver is aware of, and likewise, gently steering the conversation

SEEK Parent Questionnaire
A Safe Environment for Every Kid

No. ___

Dear Parent or Caregiver: ***Being a parent is not easy.*** *We want to help families have a safe environment for kids. We are asking everyone these questions. Please answer the questions about your* ***child being seen today*** *for a check-up. They are about issues that affect many families. If there's a problem, we'll try to help.*

Today's Date: ____/____/200_
Child's Date of Birth: ____/____/____
Sex of Child: □ Male □ Female

PLEASE CHECK

□ Yes	□ No	Do you need the telephone number for **Poison Control**?
□ Yes	□ No	Do you need a **smoke alarm** for your home?
□ Yes	□ No	Does **anyone** smoke **tobacco** at home?
□ Yes	□ No	Is there a **gun** in your home?
□ Yes	□ No	In the last year, did **you** worry that your food would **run out** before you got money, or food stamps to buy more?
□ Yes	□ No	Do you worry that your **child** may have been **physically** abused?
□ Yes	□ No	Do you worry that your **child** may have been **sexually** abused?
□ Yes	□ No	Lately, do **you** often feel **down, depressed,** or **hopeless**?
□ Yes	□ No	Do **you** often feel **lonely**?
□ Yes	□ No	During the past month, have **you** felt **little interest** or **pleasure** in the things you used to enjoy?
□ Yes	□ No	Do you often feel your **child** is **difficult** to take care of?
□ Yes	□ No	Do you wish you had more **help** with your **child**?
□ Yes	□ No	Do **you** feel so **stressed** you can't take another day?
□ Yes	□ No	Do **you** sometimes find you need to **hit/spank** your child?
□ Yes	□ No	In the past year, have **you** or **your partner** had a problem with **drugs** or **alcohol?**
□ Yes	□ No	In the past year, have **you** or **your partner** felt the need to cut back on **drinking** or **drug use**?
□ Yes	□ No	Have **you ever** been in a relationship in which you were physically **hurt** or **threatened** by a partner?
□ Yes	□ No	In the past year, have **you** been **afraid** of a partner?
□ Yes	□ No	In the past year have **you** thought of getting a **court order** for protection?
□ Yes	□ No	**Are there any problems you'd like help with today?**

Please give this form to the doctor or nurse you're seeing today. Thank you

_____ _____
Provider's name, PRINTED Provider's Signature Date

Figure 1-2. SEEK questionnaire. *[With permission from Dubowitz et al. (2007).]*

away from trivial discussion. Carefully listening to what is stated and how it is stated is important. Directly questioning the child when possible provides unique information, particularly if done at the onset of the interview before the adults start talking. Documenting exact quotes, using the vocabulary of the child or caregiver, improves accuracy of the medical record and avoids relying on an observer's interpretation of events when first-hand information is available. Bringing this information together is then the job of the clinician, who assembles all relevant facts into a cogent story that characterizes the neurologic process.

A child's developmental status is critically important in any neurologic assessment, and using a valid developmental screening test is helpful. Several assessment tools are available. These can be divided into provider-administered tools, such as the **Denver developmental screening test-II** (DDST-II) (see Fig. A-1ab in Appendix A), and parent questionnaires (Tervo, 2005), such as the **ages and stages questionnaire** and the **parents' evaluation of developmental status**.

Using the DDST-II, development is plotted over four broad domains of gross motor, fine motor, personal-social, and language skills from birth through 6 years of age. The age distribution for passing at the 25th, 50th, and 90th percentile is noted for each of 125 items. The DDST-II can be performed in a busy office setting; however, it is only a screening test, and any concerns should be followed up with more extensive developmental assessments. Also, adaptations must be made based on the cultural context. For example, the DDST-II items "using a spoon and fork" and "playing board games" are not relevant in cultures in which no one does these activities, and may need to be substituted by more appropriate developmental items. Concerns have been raised about the lack of validity of the DDST-II, and in the United States some states do not recommend using the DDST-II as a result of poor sensitivity and specificity (Minnesota Department of Health, http://www.health.state.mn.us/divs/fh/mch/devscrn/).

The **ages and stages questionnaire** is a preferred screening tool of many pediatric clinics and public health departments for children from 4 months to 5 years old, utilizing the insight that parents and caregivers offer regarding their child's development (Thompson et al., 2010). It requires responses from parents and caregivers to answer questions regarding whether specific developmental skills are demonstrated (yes, sometimes, and not yet). Five broad developmental domains are covered: communication, gross motor, fine motor, problem solving, and personal-social. Similarly, the **parents' evaluation of developmental status** is used for children from birth to 8 years old and relies on parent report through a 10-item standardized questionnaire covering expressive and receptive language, fine motor, gross motor, behavior, socialization, self-care, and learning, which identifies risk based on parental assessment. Both the ages and stages questionnaire and the parents' evaluation of developmental status are appropriate screening tools for child development and can be completed in a busy office setting.

Often it is not development but behavior that is of concern to parents. Clarifying as much as possible the nature of the behavioral concern helps. More helpful than hearing "He's acting out" is knowing if there is a problem with attention, impulsivity, aggression, or mood swings. Is there social withdrawal? Are there compulsions? Is the maladaptive behavior triggered by something or does it occur without warning?

Assessment tools are available that can help clarify problem behavior. For young children, one of the few available standardized measures is the **infant toddler social emotional assessment** or its abbreviated version, the **brief infant toddler social emotional assessment**, both of which are designed for children from 1 to 3 years old. The Brief Infant Toddler Social Emotional Assessment includes separate forms for parents and for childcare providers, each of which can be completed in less than 10 minutes. Two general types of behavior are assessed: 1) social emotional problems, including aggression, anxiety, dysregulation, and atypical behavior, and 2) social emotional competence such as attention, motivation, empathy, and positive peer relationships. For older children, the **child symptom inventory** provides parent and teacher checklists to help screen for a broad number of behavioral concerns in children from 5 to 18 years of age such as anxiety, attention deficit hyperactivity disorder, depression, oppositional defiant disorder, and conduct disorder. Likewise, the **Swanson, Nolan and Pelham qestionnaire** (SNAP) for children from 6 to 18 years old has forms available for teachers or parents that help distinguish between attention deficit hyperactivity disorder and other childhood behavioral disorders. Other general behavior assessment tools commonly used are the **Achenbach system of empirically based assessment** for children from 6 to 18 years old (this includes the **child behavior checklist**, which is a parent-report questionnaire, and the **youth self-report** to be completed by the child), and the **behavioral assessment system for children, second edition**, for age 2 to 25 years (this also includes questionnaires for parents and teachers, as well as a self-report questionnaire). In all instances in which behavior is a concern, it is helpful to use assessment tools that characterize behavior across environments (i.e., home, school, and daycare).

A common behavioral question is whether there is attention deficit hyperactivity disorder, in which case the **revised Conners parent rating scale** (age 3 to 17 years) or the **Vanderbilt** (age 6 to 12 years) is commonly used. Another common question that arises is whether a child has autistic spectrum disorder, which can be screened for using the **modified checklist for autism in toddlers** (Fig. 1-5) completed by a caregiver or via physician interview. Screening all children for autism is now recommended by the American Academy of Pediatrics at age 18 and 24 months.

Developmental screening is an enormously important part of the neurologic evaluation, but it must be interpreted carefully. For example, language delay may be caused by a congenital brain malformation or could be entirely the result of an impoverished environment (Fernald et al., 2013). Also, a single developmental screening is not as accurate as repeated assessments over time, and any abnormal screening results should prompt an immediate referral for more complete developmental testing by an appropriate professional.

Thus, a careful and informed history is at the core of the neurologic assessment of a child. The presenting complaint is explored in detail, followed by relevant aspects of the medical history, family history, and review of prior investigations or treatments. Assessment of the developmental status of the child is an essential component of this process, which eventually guides the examination to follow.

Child neurology is a unique and inherently complex endeavor, and at times the initial history uncovers information requiring clarification through a follow-up literature search. Commonly used general medical search engines include PubMed (http://www.ncbi.nlm.nih.gov/pubmed) or Google Scholar (http://scholar.google.com/). Specific websites might be considered such as Treatable Intellectual Disorder (http://www.treatable-id.org/) for metabolic conditions, or the Online Mendelian Inheritance in Man (http://www.omim.org/) for genetic disorders. As always, a tincture of humility is helpful for even the experienced clinician, knowing that a diagnosis can be frustratingly elusive at first. But what is initially confusing may be clarified as a child is followed over time and new information obtained, and through a partnership with the family and careful attention to neurologic detail, the correct diagnosis can be identified.

M-CHAT

Please fill out the following about how your child usually is. Please try to answer every question. If the behavior is rare (e.g., you've seen it once or twice), please answer as if the child does not do it.

1.	Does your child enjoy being swung, bounced on your knee, etc.?	Yes No
2.	Does your child take an interest in other children?	Yes No
3.	Does your child like climbing on things, such as up stairs?	Yes No
4.	Does your child enjoy playing peek-a-boo/hide-and-seek?	Yes No
5.	Does your child ever pretend, for example, to talk on the phone or take care of a doll or pretend other things?	Yes No
6.	Does your child ever use his/her index finger to point, to ask for something?	Yes No
7.	Does your child ever use his/her index finger to point, to indicate interest in something?	Yes No
8.	Can your child play properly with small toys (e.g. cars or blocks) without just mouthing, fiddling, or dropping them?	Yes No
9.	Does your child ever bring objects over to you (parent) to show you something?	Yes No
10.	Does your child look you in the eye for more than a second or two?	Yes No
11.	Does your child ever seem oversensitive to noise? (e.g., plugging ears)	Yes No
12.	Does your child smile in response to your face or your smile?	Yes No
13.	Does your child imitate you? (e.g., you make a face-will your child imitate it?)	Yes No
14.	Does your child respond to his/her name when you call?	Yes No
15.	If you point at a toy across the room, does your child look at it?	Yes No
16.	Does your child walk?	Yes No
17.	Does your child look at things you are looking at?	Yes No
18.	Does your child make unusual finger movements near his/her face?	Yes No
19.	Does your child try to attract your attention to his/her own activity?	Yes No
20.	Have you ever wondered if your child is deaf?	Yes No
21.	Does your child understand what people say?	Yes No
22.	Does your child sometimes stare at nothing or wander with no purpose?	Yes No
23.	Does your child look at your face to check your reaction when faced with something unfamiliar?	Yes No

Figure 1-5. Modified checklist for autism in toddlers. *[With permission from Robins et al. (1999).]*

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The complete list of references for this chapter is available in the e-book at www.expertconsult.com.
See inside cover for registration details.

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E-BOOK FIGURES AND TABLES

The following figures and tables are available in the e-book at www.expertconsult.com. See inside cover for registration details.

Fig. 1-1 Patterns of onset and courses of neurologic conditions.
Fig. 1-3 Abbreviated Swanson, Nolan and Pelham Questionnaire (SNAP) rating scales.
Fig. 1-4 Vanderbilt assessment scales.
Table 1-2 Major available screening tools.

2 Neurologic Examination of the Older Child

Kenneth F. Swaiman and John Phillips

The neurologic examination provides critical and unique information that cannot be acquired otherwise (Campbell, 2013). Regardless of patient age, the essence of this information is the same: mental status, cranial nerves, motor, reflexes, sensory, and coordination/cerebellar testing. *How* this information is obtained is very age-dependent, however (Egan, 1990).

OBSERVATION/MENTAL STATUS

Observation during history-taking is helpful, even when a child isn't being directly questioned (Menkes et al., 2005). Abnormal movements might suggest epilepsy, motor tic disorder, or a behavioral diagnosis such as attention deficit hyperactivity disorder (Pina-Garza, 2013). Caregiver–child interactions during the interview may offer clues into what the home environment is like. During this period the examiner surreptitiously assesses mental status, making note of language, attention, affect, and general developmental status (see Table 2-1).

SCREENING GROSS MOTOR FUNCTION

Consider beginning with a rapid screening examination in case the child later becomes uncooperative. Start with the child standing. Ask the child to hop in place on each foot, tandem-walk forward and backward, toe-walk, and heel-walk. Then, checking for Gowers' maneuver, the child is asked to rise quickly from a squatting position, followed by asking the child to stand with the feet close together, eyes closed, and arms and hands outstretched. This maneuver allows simultaneous assessment of Romberg's sign and adventitious movements. Finally, finger-nose-finger movements help assess cerebellar function.

PHYSICAL EXAMINATION

Cranial Nerve Examination

Examination of the cranial nerves in infants and younger children usually requires some modification of the sequence and may need some ingenious improvisation of the procedure, according to the degree of cooperation of the child (Volpe, 2008). As is the case with all examinations of infants and young children, the less threatening portions of the examination should be performed first.

Olfactory Nerve: Cranial Nerve I

Cranial nerve I can be evaluated by having the child smell pleasant aromas (e.g., chocolate, vanilla, peppermint) through each nostril while the other is manually occluded. Anosmia may occur after head trauma, with a severe upper respiratory tract infection, or in the rare instance of a frontal lobe mass involving the cribriform plate region.

Optic Nerve: Cranial Nerve II

Begin with formal visual acuity testing using a Snellen chart or a "near card" in older children. Younger children are more difficult and many times only gross vision can be evaluated. Beyond 4 years of age, the E test is useful. The child is taught to recognize the E, and to discern the direction in which the three "arms" are pointing and point a finger accordingly.

Peripheral visual field testing is accomplished using a small (3-mm) white or red test object, a toy, or in a pinch, the examiner's fingers can be used. The test object is moved from the temporal to the nasal fields and then from the superior and inferior portions of the temporal and nasal fields while the child looks directly at the examiner's nose. Finger counting can be used if acuity is grossly distorted. In cases of extreme impairment, perception of a rapidly moving finger can be used.

The optic disc (i.e., optic nerve head) of the older child is sharply defined and often salmon-colored, which differs from the pale gray color of the disc in an infant. In the presence of a deep cup in the optic disc, the color may appear pale, but the pallor is localized to the center of the disc. The pallor of optic atrophy occurs centrally and peripherally, and is accompanied by a decreased number of arterioles in the disc margins. Most commonly, papilledema is associated with elevation of the optic disc, distended veins, and lack of venous pulsations. Hemorrhages may surround the disc. Before papilledema is obvious, there may be blurring of the nasal disc margins and hyperemia of the nerve head.

The presence or absence of the pupillary light reflex differentiates between peripheral and cortical blindness. Lesions of the anterior visual pathway (i.e., retina to lateral geniculate body) result in the interruption of the afferent limb of the pupillary light reflex, producing an absent or decreased reflex. Anterior visual pathway interruption can cause amblyopia in one eye. In this situation, the pupil fails to constrict when stimulated with direct light; however, the consensual pupillary response (i.e., response when the other eye is illuminated) is intact. The deficient pupillary reflex is revealed by alternately aiming a light source toward one eye and then the other. In the eye with decreased vision, consensual pupillary constriction is greater than the response to direct light stimulation (Marcus Gunn pupil); the pupil of the affected eye may dilate slightly during direct stimulation (Haymaker, 1969).

Oculomotor, Trochlear, and Abducens Nerves: Cranial Nerves III, IV, and VI

The oculomotor, trochlear, and abducens cranial nerves control extraocular motor movements; these nerves must operate synchronously or diplopia ensues. Cranial nerve III innervates the superior, inferior, and medial recti; the inferior oblique; and the eyelid elevator (levator palpebrae superioris). Cranial nerves IV and VI innervate the superior oblique muscle and the lateral rectus muscle, respectively. Unfortunately for

TABLE 2-1 Emerging Patterns of Behavior from 1 to 5 Years of Age

15 Months	
Motor:	Walks alone; crawls up stairs
Adaptive:	Makes tower of two cubes; makes line with crayon; inserts pellet into bottle
Language:	Jargon; follows simple commands; may name familiar object (ball)
Social:	Indicates some desires or needs by pointing; hugs parents
18 Months	
Motor:	Runs stiffly; sits on small chair; walks up stairs with one hand held; explores drawers and waste baskets
Adaptive:	Piles three cubes; initiates scribbling; imitates vertical stroke; dumps pellet from bottle
Language:	Ten words (average); names pictures; identifies one or more parts of body
Social:	Feeds self; seeks help when in trouble; may complain when wet or soiled; kisses parents with pucker
24 Months	
Motor:	Runs well; walks up and down stairs one step at a time; opens doors; climbs on furniture
Adaptive:	Makes tower of six cubes; circular scribbling; imitates horizontal strokes; folds paper once imitatively
Language:	Puts three words together (subject, verb, object)
Social:	Handles spoon well; tells immediate experiences; helps to undress; listens to stories with pictures
30 Months	
Motor:	Jumps
Adaptive:	Makes tower of eight cubes; makes vertical and horizontal strokes but generally will not join them to make a cross; imitates circular stroke, forming closed figure
Language:	Refers to self by pronoun "I"; knows full name
Social:	Helps put things away; pretends in play
36 Months	
Motor:	Goes up stairs alternating feet; rides tricycle; stands momentarily on one foot
Adaptive:	Makes tower of nine cubes; imitates construction of "bridge" of three cubes; copies circle; imitates cross
Language:	Knows age and gender; counts three objects correctly; repeats three numbers or sentence of six syllables
Social:	Plays simple games (in "parallel" with other children); helps in dressing (unbuttons clothing and puts on shoes); washes hands
48 Months	
Motor:	Hops on one foot; throws ball overhand; uses scissors to cut out pictures; climbs well
Adaptive:	Copies bridge from model; imitates construction of "gate" of five cubes; copies cross and square; draws man with 2–4 parts besides head; names longer of two lines
Language:	Counts four pennies accurately; tells a story
Social:	Plays with several children with beginning of social interaction and role playing; goes to toilet alone
60 Months	
Motor:	Skips
Adaptive:	Draws triangle from copy; names heavier of two weights
Language:	Names four colors; repeats sentences of ten syllables; counts ten pennies correctly
Social:	Dresses and undresses; asks questions about meanings of words; domestic role playing

(Adapted with permission from Behrman RE, et al. Nelson Textbook of Pediatrics, 14th edn. Philadelphia: WB Saunders, 1992.)

purposes of understanding, the function of extraocular muscles depends somewhat on the direction of gaze. The lateral and medial recti are abductors and adductors of the globe, respectively. The superior rectus and inferior oblique are elevators, and the inferior rectus and superior oblique are depressors. The oblique muscles act in the vertical plane while an eye is adducted. The recti muscles serve this function when an eye is abducted (Figure 2-2). When directed forward (i.e., primary position), the oblique muscles effect torsion around the anteroposterior axis (rotation) of the globes.

In heterophorias, also called phorias, both globes are directed normally on near or far objects during fixation; however, one or both deviate when one eye is occluded while the other eye fixes. Forcing fixation of the uncovered eye by alternately covering each eye confirms the diagnosis of heterophorias. Exophoria is a predisposition to divergence, whereas esophoria is a predisposition to convergence.

Eye deviations detectable during binocular vision are heterotropias, also called tropias. Adduction tropias are esotropias; abduction tropias are exotropias. Tropias are most often caused by compromised extraocular muscle innervation. Extraocular palsies can frequently be detected by observation of eye movements. A red glass is placed in front of an eye, and a focused, relatively intense white light is aimed at the eyes from various visual fields while the child fixes on the light. A merged, solitary, red–white image is perceived when extraocular movements are normal; however, when muscle paresis is present, the child reports a separation of the red and white images when looking in the direction of action of the affected

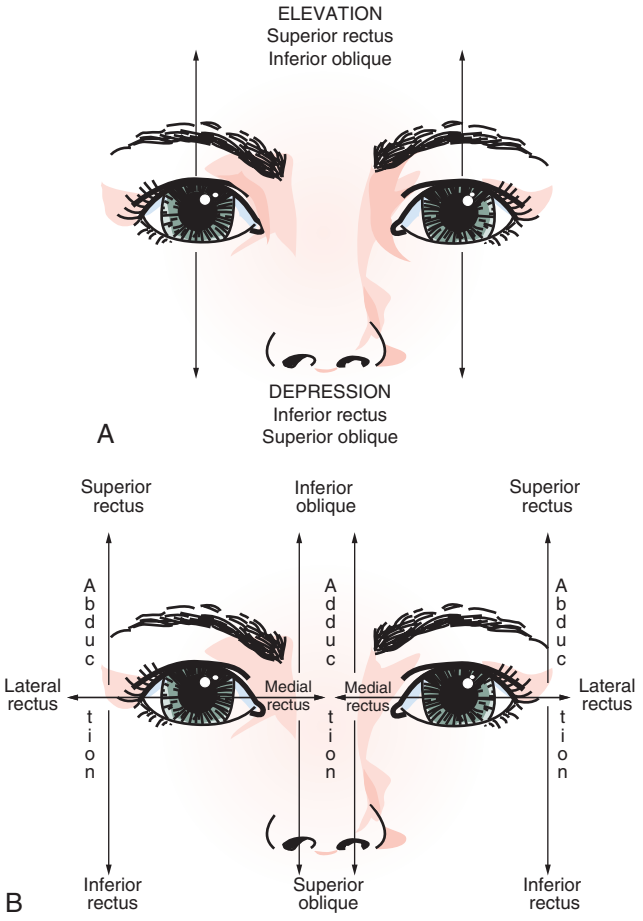

Figure 2-2. Extraocular muscle movement. A, In primary position. **B,** In abduction and adduction. *(Courtesy of the Division of Pediatric Neurology, University of Minnesota Medical School.)*

muscle. The farthest peripheral image is the one perceived by the abnormal eye; this eye can be identified by the color of the image. Volitional turning of the head accompanies paresis of the lateral rectus muscle to forestall diplopia; the head is deviated toward the paretic muscle, and the eyes are directed ahead. In superior oblique or superior rectus muscle palsies, tilting of the head toward the shoulder opposite the side of the paretic eye muscle occurs.

Extraocular muscle dysfunction is associated with many conditions that affect the brainstem, cranial nerves, neuromuscular junction, or muscles. Cranial nerve VI function may be impaired by increased intracranial pressure, irrespective of cause. Squint, usually esotropia, often accompanies decreased visual acuity in infants and young children.

Ptosis and extraocular muscle paralysis accompany dysfunction of cranial nerve III. Ptosis resulting from oculomotor nerve compromise is usually more pronounced than is the malposition of the lid associated with Horner syndrome. Complete oculomotor nerve paralysis, although uncommon, causes the eye to position downward and outward. Poor adduction and elevation are also evident.

Eye deviation is often the harbinger of a serious neurologic problem. Destructive lesions of the brainstem nuclei cause conjugate eye deviation toward the opposite side. Destructive cerebral hemispheral lesions cause eye deviation toward the side of the lesion; conversely, an irritative cerebral hemispheral lesion (such as a seizure focus) causes the eyes to turn away from the side of the lesion. Hence for a cortical lesion, "the patient looks at their stroke but away from their seizure." Vertical gaze paresis results from dysfunction of the tectal area of the midbrain. Patients with a pineal tumor or hydrocephalus may be unable to elevate the eyes for upward gaze.

Brainstem lesions, especially those in the midbrain or pons, may disrupt the medial longitudinal fasciculus. The resultant impairment of conjugate eye movement is referred to as an internuclear ophthalmoplegia. There is weakness of medial rectus muscle contraction of the adducting eye, which is accompanied by a monocular nystagmus in the abducting eye. Occasionally, paresis of lateral rectus muscle movement in the abducting eye may occur. Medial longitudinal fasciculus involvement may be unilateral or bilateral.

Internal ophthalmoplegia consists of a fully dilated pupil that is unreactive to light or accommodation. Extraocular muscle function is normal when each muscle is tested separately. The oculomotor nerve, nucleus, or the parasympathetic ciliary ganglion may be a site of involvement.

External ophthalmoplegia results in ptosis and paralysis of all extraocular muscles. Pupillary reactivity is normal.

Opticokinetic nystagmus is a useful test in evaluating the eye movements of children. A drum or tape with stripes or figures is slowly rotated or drawn before the child's eyes in horizontal and vertical directions. With fixation, the child should visually track the object in the direction the tape is being drawn, with a rapid, rhythmic movement (refixation) of the eyes in the reverse direction to enable fixation on the next figure or stripe. Absence of such a response may result from failure of fixation, amaurosis, or disturbed saccadic eye movements.

The child who appears clinically blind because of a conversion reaction usually exhibits a normal opticokinetic nystagmus response. Children who manifest congenital nystagmus and have an opticokinetic nystagmus response in the vertical plane likely have adequate functional sight.

Spontaneous nystagmus (i.e., involuntary oscillatory movements of the eye) may be horizontal, vertical, or rotary; a patient can exhibit all three types. The movements may consist of a slow and a fast phase, giving rise to the term jerk nystagmus. However, the phases may be of equal duration and amplitude, appearing pendular. In general, vertical nystagmus is associated with either medication or brainstem dysfunction. While a few beats of horizontal nystagmus with extreme lateral gaze are normal, persistent horizontal nystagmus indicates dysfunction of the cerebellum or brainstem vestibular system components; the nystagmus is coarser (i.e., the amplitude of movements are greater) when the direction of gaze is toward the side of the lesion. Seesaw nystagmus is characterized by disconjugate (alternating) movement of the eyes, which move upward and downward in a seesaw motion. This type of nystagmus may accompany lesions in the region of the optic chiasm (see Chapter 6).

Trigeminal Nerve: Cranial Nerve V

Cranial nerve V, the trigeminal nerve, has motor and sensory functions. The motor division of the trigeminal nerve innervates the masticatory muscles: masseter, pterygoid, and temporalis. Temporalis muscle atrophy manifests as scalloping of the temporal fossa. The masseter muscle bulk may be assessed by palpation while the patient firmly closes the jaw. Pterygoid muscle strength is evaluated by having the patient open the mouth and "slide" the jaw from one side to the other while the examiner resists movements with the hand to assess muscle strength. The jaw reflex is elicited when the examiner places a finger on the patient's chin while the mouth is slightly open and taps the finger to stretch the masticatory muscles. A rapid muscle contraction with closure of the mouth is the reflex response. This stretch reflex receives its afferent and efferent nerve control from cranial nerve V; the segmental level is located in the midpons. The expected reflex reaction is absent with motor nucleus and peripheral trigeminal nerve compromise. Conversely, this reflex is overactive in the presence of supranuclear lesions; rarely, jaw clonus may be evident. Because of weakness of the ipsilateral pterygoid muscles, unilateral impairment of the trigeminal nerve causes deviation of the jaw toward the side of the lesion. Cranial nerve V is also responsible for sensation involving the face, including eye and the anterior half of the scalp.

Facial Nerve: Cranial Nerve VII

Taste sensation over the anterior two-thirds of the tongue, secretory fibers (parasympathetic) innervating the lacrimal and salivary glands, and innervation of all facial muscles are accomplished by cranial nerve VII. Complete motor dysfunction on one side of the face ensues when the cranial nerve VII pathway is disrupted in the nucleus, pons, or peripheral nerve. The patient is unable to move the forehead upward, close the eye forcefully, or elevate the corner of the mouth on the side of the affected nerve.

Central (supranuclear) facial nerve impairment produces only paresis of the muscles involving the lower face, with resultant drooping of the angle of the mouth, disappearance or diminution of the nasal labial fold, and widened palpebral fissure. The muscles of the forehead, which are innervated bilaterally, are unaffected.

Taste sensation in the anterior two-thirds of the tongue is in part provided by the chorda tympani nerve, which traverses the path of the facial nerve for a short distance. Testing of taste sensation is difficult. Evaluation of taste requires that the patient extend the tongue and that the examiner hold the tip of the tongue with a piece of gauze and place salty, sweet, acidic, and sour and bitter materials, usually represented by salt, sugar, vinegar, and quinine, on the anterior portion of the tongue. The patient's tongue must remain outside of the mouth until the test is completed. An older patient should be able to identify each substance.

Auditory Nerve: Cranial Nerve VIII

Function and evaluation of cranial nerve VIII are discussed in detail in Chapters 7 and 8. Although cranial nerve VIII is known as the auditory nerve, it has auditory and vestibular functions.

Patients who fail to develop speech or who have slow speech development, as well as those who have difficulty with fluency and articulation, may have hearing impairment. Older children can cooperate with formal audiometric testing. Such testing may not be possible in younger infants, but brainstem auditory-evoked potentials may provide the necessary information concerning hearing impairment and the level of dysfunction within the nervous system.

Clinical evaluation and caloric testing can be used for gross assessment of vestibular function. To perform caloric testing, the patient is in the supine position, with head flexed at 30 degrees. Ice water (10 mL) is injected over 30 seconds into one external auditory canal at a time. The conscious patient develops coarse nystagmus toward the ipsilateral ear; no eye deviation occurs. If the patient has some degree of obtundation, there is a modification of the response. The eyes become tonically deviated ipsilaterally, with accompanying nystagmus occurring contralaterally. If the patient is comatose, cold water stimulation usually causes tonic deviation ipsilaterally and no nystagmus; if the coma is profound or the patient is brain-dead, no eye changes occur.

Glossopharyngeal and Vagus Nerves: Cranial Nerves IX and X

Examination of the larynx, pharynx, and palate provides most of the desired information concerning the function of cranial nerves IX and X. Unilateral paresis of the soft palate causes an ipsilateral droop, even when the patient is expelling air through the open mouth or gagging in response to a tongue blade. Bilateral involvement causes a flaccid soft palate bilaterally.

The gag reflex is mediated through cranial nerve IX and is elicited by touching the posterior pharyngeal mucosa with a tongue blade. Normal individuals may have absence or a seemingly disproportionately violent response; assessing the importance of changes in the gag reflex is difficult in the absence of other findings. The integrity of cranial nerves IX and X is necessary for a gag response; sensation of the soft palate and uvula travels via cranial nerve IX, and motor function is carried by cranial nerve X. Thus unilateral weakness causes deviation of the uvula *away* from the weak side (unlike unilateral weakness of cranial nerve XII, discussed later, which causes deviation of the tongue *toward* the weak side).

Spinal Accessory Nerve: Cranial Nerve XI

Cranial nerve XI provides innervation for the trapezius and sternocleidomastoid muscles. Cranial nerve XI comprises some fibers from C1 and C2, and some from the motor nucleus in the brainstem, and is unique in combining brainstem and cervical cord origins. The trapezius muscles are assessed when the patient is asked to shrug the shoulders against resistance while the sternocleidomastoid muscle is tested by asking the child to rotate their head to one side against resistance. Weakness of the sternocleidomastoid muscle results in an inability to rotate the head to the contralateral side.

Hypoglossal Nerve: Cranial Nerve XII

The tongue muscle is the primary responsibility of cranial nerve XII. Atrophy and fasciculation of the tongue occur when the ipsilateral hypoglossal nucleus or hypoglossal nerve is involved. The protruded tongue deviates toward the involved side because contraction of the normally innervated tongue muscle causes protrusion and is unopposed.

Skeletal Muscles

Tone, bulk, and strength of the skeletal muscles should be determined during this portion of the examination. Motor functions of the spinal nerves are described in Table 2-3.

The strength of limb muscles is assessed, when possible, by testing the child's ability to counteract resistance imposed by the examiner on proximal and distal muscle groups or individual muscles.

Muscle Testing

The following scoring system is useful for recording muscle power:

5: normal power
4: inability to maintain position against moderate resistance
3: inability to maintain position against slight resistance or gravity
2: active movement with gravity eliminated
1: trace of contraction
0: no contraction.

Arm and shoulder strength can also be assessed by asking the child to lean against a wall with legs placed a foot or two from the wall edge and arms outstretched with the palms against the wall. Testing lower extremity strength can be assessed by asking the child to sit on the floor then rapidly stand; the normal child will spring erect. With weakness of the hip extensors, however, Gowers' maneuver will be engaged, and the patient climbs up their own legs, pushing themselves into the erect position.

Muscle bulk is evaluated by gentle palpation and observation. Muscle tenderness, nerve tenderness, and nerve hypertrophy can also be assessed by palpation. Myotonia can be elicited by tapping over the thenar eminence and deltoid muscles.

Muscle tone is evaluated when the child is relaxed so that resistance to passive movement can be monitored. Aside from passive movement of limbs at joints, the examiner also assesses the extensibility of muscles by shaking the limbs and determining the range of motion.

Tone may be decreased in the presence of cerebellar disease and anterior horn cell disease. Tone may be increased because of the rigidity associated with basal ganglia disease and spasticity associated with corticospinal tract dysfunction.

Deep Tendon Reflexes

Standard deep tendon reflexes (i.e., muscle stretch reflexes) are elicited: biceps, triceps, brachioradialis, patellar, and Achilles reflexes.

The response to elicitation of deep tendon reflexes can be characterized as follows:

0: absent
1: hyporeflexic (trace, or only seen with reinforcement)
2: normal
3: hyperreflexic
4: unsustained clonus
5: sustained clonus.

Enhancement of tendon reflex responses when reflexes are seemingly absent can be promoted by having the child squeeze an object such as a block or ball or perform the more

TABLE 2-3 Extraocular Muscle Paralysis

Nerves	Muscles*	Function
Cervical Plexus (C1–C4)		
Cervical	Deep cervical	Flexion, extension, and rotation of neck
Phrenic	Scalene	Elevation of ribs (inspiration)
	Diaphragm	Inspiration
Brachial Plexus (C5–T1)		
Anterior	Pectorales major and minor	Adduction and depression of arm downward and medially
Long thoracic	Serratus anterior	Fixation of scapula on raising arm
Dorsal scapular	Levator scapulae	Elevation of scapula
	Rhomboid	Drawing scapula upward and inward
Suprascapular	Supraspinatus	Outward rotation of arm
	Infraspinatus	Elevation and outward rotation of arm
Subscapular	Latissimus dorsi	
	Teres major	Inward rotation and abduction of arm toward the back
	Subscapularis	Inward rotation of arm
Axillary	Deltoid	Raising of arm to horizontal
	Teres minor	Outward rotation of arm
Musculocutaneous	Biceps brachii	Flexion and supination of forearm
	Coracobrachialis	Elevation and adduction of arm
	Brachialis	Flexion of forearm
Median	Flexor carpi radialis	Flexion and radial deviation of hand
	Palmaris longus	Flexion of hand
	Flexor digitorum sublimis	Flexion of middle phalanges of second through fifth fingers
	Flexor pollicis longus	Flexion of distal phalanx of thumb
	Flexor digitorum profundus (radial half)	Flexion of distal phalanges of second and third fingers
	Pronator quadratus	Pronation
	Pronator teres	Pronation
	Abductor pollicis brevis	Abduction of metacarpus I at right angles to palm
	Flexor pollicis brevis	Flexion of proximal phalanx of thumb
	Lumbricals I, II, III	Flexion of proximal phalanges and extension of other phalanges of first, second, and third fingers
	Opponens pollicis brevis	Opposition of metacarpus I
Ulnar	Flexor carpi ulnaris	Flexion and ulnar deviation of hand
	Flexor digitorum profundus (ulnar half)	Flexion of distal phalanges of fourth and fifth fingers
	Adductor pollicis	Adduction of metacarpus I
	Hypothenar	Abduction, opposition, and flexion of little finger
	Lumbricals III, IV	Flexion of first phalanx and extension of other phalanges of fourth and fifth fingers
	Interossei	Same action as preceding. Also spreading apart and bringing together of fingers
Radial	Triceps brachii	Extension of forearm
	Brachioradialis	Flexion of forearm
	Extensor carpi radialis	Extension and radial flexion of hand
	Extensor digitorum communis	Extension of proximal phalanges of second through fifth fingers
	Extensor digiti quinti proprius	Extension of proximal phalanx of little finger
	Extensor carpi ulnaris	Extension and ulnar deviation of hand
	Supinator	Supination of forearm
	Abductor pollicis longus	Abduction of metacarpus I
	Extensor pollicis brevis	Extension of proximal phalanx of thumb
	Extensor pollicis longus	Abduction of metacarpus I and extension of distal phalanges of thumb
	Extensor indicis proprius	Extension of proximal phalanx of index finger
Thoracic Nerves		
Thoracic	Thoracic and abdominal	Elevation of ribs, expiration, abdominal compression, etc.
Lumbar Plexus (T12–L4)		
Femoral	Iliopsoas	Flexion of leg at hip
	Sartorius	Inward rotation of leg together with flexion of upper and lower leg
	Quadriceps femoris	Extension of lower leg
Obturator	Pectineus	Adduction of leg
	Adductor longus	
	Adductor brevis	
	Adductor magnus	
	Gracilis	
	Obturator externis	Adduction and outward rotation of leg

Continued on following page

TABLE 2-3 Extraocular Muscle Paralysis *(Continued)*

Sacral Plexus (L5–S5)		
Superior gluteal	Gluteus medius, Gluteus minimus	Abduction and inward rotation of leg; also, under certain circumstances, outward rotation
	Tensor fasciae latae	Flexion of leg at hip
	Piriformis	Outward rotation of leg
	Gluteus maximus	Extension of leg at hip
Inferior gluteal		
Sciatic	Obturator internus, Gemelli, Quadratus femoris	Outward rotation of leg
	Biceps femoris, Semitendinosus, Semimembranosus	Flexion of leg at hip
Peroneal	Tibialis anterior	Dorsiflexion and supination of foot
Deep	Extensor digitorum longus	Extension of toes
	Extensor hallucis brevis	Extension of great toe
Superficial	Peroneus	Pronation of foot
Tibialis	Gastrocnemius, Soleus	Plantar flexion of foot
	Tibialis posterior	Adduction of foot
	Flexor digitorum longus	Flexion of distal phalanges II–V
	Flexor hallucis longus	Flexion of distal phalanx I
	Flexor digitorum brevis	Flexion of middle phalanges II–V
	Flexor hallucis brevis	Flexion of middle phalanx I
	Plantar	Spreading, bringing together, and flexion of proximal phalanges of toes
Pudendal	Perineal anal sphincters	Closure of sphincters of pelvic organs; participation in sexual act; contraction of pelvic floor

*Various muscles may receive still other nerve supplies than those mentioned. The following are the principal accessory nerve supplies: the brachial muscle receives fibers from the radial nerve; the flexor digitorum sublimis, from the ulnar; the adductor pollicis, from the median; the pectineus, from the femoral; the adductor magnus, from the tibial.

(With permission from Haymaker W. Bing's Local Diagnosis in Neurological Diseases, 15th edn. St. Louis: Mosby, 1969.)

traditional Jendrassik maneuver (i.e., hooking the fingers together while flexed and then attempting to pull them apart). Hyperreflexia may be indicated by an abnormal "spread" of responses, which includes contraction of muscle groups that usually do not contract when a specific reflex is being elicited (i.e., crossed thigh adductor or finger flexor reflexes).

Other Reflexes

A flexor (plantar) toe sign response is normal in children. The Babinski reflex is elicited by firm, steady, slow stroking from posterior to anterior of the lateral margin of the sole with an object such as a key or a tongue blade. The stimulus should not be painful. A positive response is a slow, tonic hyperextension of the great toe. A similar response is elicited using maneuvers such as the Chaddock (firmly stroking the lateral aspect of the foot) or Oppenheim (downward pressure on the medial aspect of the tibia).

Flicking the patient's nail (second or third finger) downward with the examiner's nail (i.e., the Hoffmann reflex) results in flexion of the distal phalanx of the thumb. No response or a muted response occurs in normal children; a brisk or asymmetric response occurs in the presence of corticospinal tract involvement.

Abdominal reflexes are obtained by stroking the abdomen from lateral to medial with strokes beginning just above the umbilicus, lateral to the umbilicus, and just below the umbilicus directed toward the umbilicus. Unilateral absence of the reflex can be associated with acquired corticospinal tract dysfunction.

The cremasteric reflex is elicited in males by stroking the inner aspects of the thigh in a caudal–rostral direction and observing the contraction of the scrotum. The reflex is normally present and symmetric. Absence or asymmetry may indicate corticospinal tract involvement.

Sensory System

Cooperation is necessary for a successful sensory examination. Vibration and proprioception can be assessed in all four limbs. Touch may be assessed by a single stimulus or by double simultaneous stimulation of two skin areas, which involves touching two parts of the body simultaneously (i.e., double simultaneous stimulation test). Extinction is the term used to denote failure of the child to perceive both stimuli. The contralateral parietal lobe to the side on which the unidentified stimulus was applied is the site of dysfunction. Pain, as tested with a pinprick, must be assessed gently, rapidly, and in a nonthreatening and playful manner. Segmental sensory innervations of the arm and leg should be noted. For example, the nipples are at approximately the T5 level and the umbilicus at the T10 level.

Cortical sensory function can be tested in the older child. Stereognosis is the recognition of familiar objects by touch. After the patient closes the eyes, objects are placed by the examiner in one of the child's hands and then the other. The patient should recognize the objects by size, texture, and form. Objects may include a button, coins, safety pin, or key. Absence of stereognosis is astereognosis. Astereognosis usually results from lesions of the parietal lobe.

Graphesthesia is the ability to recognize numbers, letters, or other readily identifiable symbols traced on the skin. This ability can be determined best by tracing the symbols in a preliminary trial while the child's eyes are open. When the patient's eyes are closed, the figures are traced over the palm or forearm. Failure to identify the symbols is dysgraphesthesia. By 8 years of age, most children are able to identify all single digits correctly.

The ability to distinguish between closely approximated stimulation at two points is two-point discrimination. Normal findings have been reported for children 2 to 12 years old.

Testing of this modality is frequently performed over the fingertips. Absence or impairment of two-point discrimination results from parietal lobe dysfunction.

Cerebellar Function

Cerebellar function is assessed in a number of ways. Hand patting (i.e., alternating pronation and supination of the hand on the thigh while the other hand remains stationary on the other thigh) is a good method for assessing dysdiadochokinesis. The maneuver is repeated with each hand separately to assess the presence of mirror movements (i.e., synkinesis). Other cerebellar tests include repetitive finger tapping (thumb to forefinger), foot tapping, and finger-to-nose, finger-to-finger (examiner's)-to-nose, and heel-to-knee-to-shin stroking. Several signs of cerebellar disease include head tilt, tremor when approaching a target (intention tremor), overshooting or undershooting a target (dysmetria), speech with an unusual cadence or prosody (cerebellar speech), or gait ataxia.

GAIT EVALUATION

The evaluation of gait is discussed in detail in Chapter 5.

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The complete list of references for this chapter is available in the e-book at www.expertconsult.com.
See inside cover for registration details.

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E-BOOK FIGURES AND TABLES

The following figures and tables are available in the e-book at www.expertconsult.com. See inside cover for registration details.

Fig 2-1 Bilateral oculomotor nerve paralysis.
Fig 2-3 Facial sensation supplied by the trigeminal nerve.
Fig 2-4 Right facial paralysis of the peripheral type.
Fig 2-5 Möbius' syndrome is manifested by bilateral palsy of cranial nerves VI and VII.
Fig 2-6 Fasciculation of the tongue, especially of the right lateral border, in a patient with group 2 Werdnig–Hoffmann disease.
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3 Neurologic Examination after the Newborn Period Until 2 Years of Age

Kenneth F. Swaiman and John Phillips

An expanded version of this chapter is available on www.expertconsult.com. See inside cover for registration details.

There is no one way to organize the examination of an infant. Experienced examiners develop individual techniques and sequences that are flexible depending on the child's level of cooperation (Jan, 2007). The technique presented here, which has worked for many clinicians, is a four-stage examination sequence, beginning with the least intrusive maneuvers. Developmental assessment is an integral part of this examination; norms are well established (Box 3-1), and when development lags, referral for more detailed testing may be considered.

The **first stage** of the examination is observation. In the **second stage**, the head, muscle tone, superficial and deep sensation, gross response to sound, and visual fields can be evaluated while the child remains on the caregiver's lap. The **third stage** becomes more invasive, and may require help from a caregiver or assistant. At this point, a general examination is performed, including measurement of the occipitofrontal circumference and optic fundi. In the **fourth stage** of the examination, the child is placed on the floor and encouraged to crawl, walk, and run, if possible.

EVALUATION OF THE PATIENT

Stage 1

Because many children at this age need a few minutes to feel comfortable with a stranger, it is preferable not to rush into the examination. It is often helpful to have the child sit on the caregiver's lap facing the examiner during this history-taking session to encourage familiarity. The point is to try to help the child become comfortable with the examination room and the examiner.

The sequence of examination should be flexible and determined by the child's comfort level and temperament, although eventually a complete examination must be conducted (Campbell, 2013). Most importantly, it is imperative that the clinician comprehensively conduct that aspect of the examination related to the chief complaint.

Observations done at this stage include an assessment of the child's level of alertness, awareness of surroundings, and affect. Communication skills can be noted and compared with age-appropriate expectations (Egan, 1990). Movement should be evident as well, particularly of the face, eyes, and extremities, and the examiner should look specifically for any asymmetry or abnormalities of control or posture.

Head

Examination of the head is done systematically, looking for asymmetry, indentations, and protuberances. Evaluation of the fontanels and cranial sutures should be performed with gentle palpation. Hair color, distribution, texture, and pattern, including unusual whorl patterns, also should be assessed.

The occipitofrontal circumference should always be measured. If the child becomes agitated, this can be deferred until later in the examination (stage 3), but it must be taken at some point. Change over time, rather than a single measurement, provides the most useful information. In addition, the size of the anterior fontanel, which is typically closed by 12 months of age, should be recorded, along with any tenseness when the child is sitting comfortably in an upright position. Other fontanels are usually difficult to palpate, except in pathologic states. Finally, the head should be auscultated for the presence of unusual intracranial bruits.

Cranial Nerves

Most of the examination of cranial nerve function of the infant and toddler can be completed by observation with minimal invasive procedures. Details concerning examination of each cranial nerve can be found in Chapter 2. Toys or colorful objects can facilitate the assessment of extraocular movements in young children. If the child appears uninterested in bright objects, the possibility of a visual defect or an underlying intellectual defect must be considered. Double simultaneous stimulation (i.e., simultaneously bringing two bright objects into both temporal fields) normally causes the child to look from one object to the other; failure to take notice of one object may indicate homonymous hemianopsia. An opticokinetic tape (with repetitive bars or objects) should be drawn horizontally and then vertically across the child's field of vision. An absent response may result from lack of visual fixation or from gross impairment of vision.

A beam from a small flashlight directed at each eye allows evaluation of pupil size, pupillary responses, and the red retinal reflex. Eye features to be noted include symmetry of the palpebral fissures, relative size of the two globes, angulation of the eyes compared with other facial components (i.e., mongoloid or antimongoloid slant) and with the ears, cataracts, conjunctival telangiectases, colobomas of the iris, ptosis, proptosis, and malformed or eccentrically placed pupils.

Observing the child's facial movements throughout the entire examination is helpful (Nelson and Eng, 1972). Widening of the ipsilateral palpebral fissure or inability to bury the limbus when crying is indicative of facial nerve weakness. In the younger infant, sucking and rooting reflexes should be obtained. Sometimes, the child can be induced to protrude the tongue if the examiner urges the child to imitate the examiner's tongue movements. Deformity, atrophy, or abnormal positioning of the tongue can be observed. Tongue fasciculations should be evaluated with the tongue in the resting position, and by gently elevating the tongue with a depressor and examining the undersurface.

Basic responses to the sound made by a tuning fork, rubbing fingers together, ringing a bell, or using a toy noisemaker that generates noise at a modest volume may provide much information. The examiner must be careful not to confuse response to a visual cue (e.g., the movement needed to elicit noise from a toy) with response to the sound.

BOX 3-1 Child Development from 2 Months through 2 Years

2 MONTHS
- Keeps hands predominantly fisted
- Lifts head up for several seconds while prone
- Startles in response to loud noise
- Follows with eyes and head over 90-degree arc
- Smiles responsively
- Begins to vocalize single sounds

3 MONTHS
- Occasionally holds hands fisted
- Lifts head up above body plane and holds position
- Holds an object briefly when placed in hand
- Turns head toward object, fixes and follows fully in all directions with eyes
- Smiles and vocalizes when talked to
- Watches own hands, stares at faces
- Laughs

4 MONTHS
- Holds head steady while in sitting position
- Reaches for an object, grasps it, brings it to mouth
- Turns head in direction of sound
- Smiles spontaneously

5–6 MONTHS
- Lifts head while supine
- Rolls from prone to supine
- Lifts head and chest up in prone position
- Exhibits no head lag
- Transfers object from hand to hand
- Babbles
- Sits with support
- Localizes direction of sound

7–8 MONTHS
- Sits in tripod fashion without support
- Stands briefly with support
- Bangs object on table
- Reaches out for people
- Mouths all objects
- Says "da-da," "ba-ba"

9–10 MONTHS
- Sits well without support, pulls self to sit
- Stands holding on
- Waves "bye-bye"
- Drinks from cup with assistance
- Uses pincer grasp

11–12 MONTHS
- Walks with assistance
- Uses two to four words with meaning
- Creeps well
- Assists in dressing
- Understands a few simple commands

13–15 MONTHS
- Walks by self, falls easily
- Says several words, uses jargon
- Scribbles with crayon
- Points to things wanted

18 MONTHS
- Climbs stairs with assistance, climbs up on chair
- Throws ball
- Builds two to four-block tower
- Feeds self
- Takes off clothes
- Points to two or three body parts
- Uses many intelligible words

24 MONTHS
- Runs, walks up and down stairs alone (both feet per step)
- Speaks in two- to three-word sentences
- Turns single pages of book
- Builds four- to six-block tower
- Kicks ball
- Uses pronouns "you," "me," and "I"

(Data from Frankenburg WK, Dodds J, Archer P, et al. Pediatrics. 1992;89:91; Illingsworth RS. The development of the infant and young child. 9th ed. Baltimore: Williams & Wilkins; 1987; and Knobloch H, Stevens F, Malone A. The revised developmental screening inventory. Houston, Texas: Gesell Developmental Test Materials; 1980.)

Motor Evaluation

As with all other parts of the evaluation, the motor examination begins with observation. Even before touching the child, general posture and the symmetry of movements of the arms and legs are observed, with note made of any gross discrepancies in muscle bulk or limb length. Definite hand preference (such as reaching across the midline to avoid using the contralateral hand) before 24 months is abnormal.

Decreased muscle bulk may not be appreciated because of the large amount of subcutaneous fat at this age, and muscle atrophy may be undetected. Careful palpation helps distinguish between fat and muscle.

The next step is evaluation of muscle tone, which is defined as resistance of muscle to passive stretch. This also requires palpation. Muscle tone and range of motion of the arms and legs are best assessed when the child is in the relaxed state by gently shaking and moving the hands and feet in flexion and extension. Pronation and supination of the hands and forearms provide further information about range of motion and the presence of spasticity or rigidity. Greater-than-normal resistance to passive movement indicates hypertonia, whereas less-than-normal resistance indicates hypotonia. It is important to distinguish increased *tone* from limitation of movement due to joint *contracture.* It is also important to note when the child is actively resisting the examiner, which is a reflection of *strength*. Spontaneous muscle movements, particularly those against gravity, provide the most information concerning muscle strength. Further assessment of strength is provided by judging the degree of resistance that occurs when active movement is attempted against the examiner. Experience helps in this part of the examination, particularly in cases of mild abnormalities, but even a novice should be able to make a reasonable judgment about both tone and strength in most instances.

Upper motor neuron unit involvement may cause decreased movement of an entire extremity or more focal abnormalities, such as limited flexion of the arm at the elbow, persistent fisting, or adduction of the thumb against the palm. Erb's brachial plexus injury is a lower motor neuron disorder commonly causing internal rotation and adduction at the shoulder, often with the "waiter's tip" posture (Pina-Garza, 2013). Interacting with the infant using toys and other interesting objects may facilitate the evaluation of limb strength, range

of motion, and coordination. In the older cooperative child, individual muscle testing should be carried out when appropriate.

There is a normal developmental sequence of fine motor control as the child becomes more adept at reaching for objects. Grasping things with both hands and holding the object before the face or immediately placing it in the mouth is later superseded by transferring the object from hand to hand and manipulating the toy. The infant's grasping skills are best demonstrated in response to small objects. The 4- to 5-month-old infant is able to grasp an object with the entire hand, at 7 months the thumb and the neighboring two fingers are used, and the pincer grasp (using only the thumb and forefinger) should be present by 9 to 11 months. The palmar grasp reflex (i.e., obligate grasp reflex) should gradually diminish from 3 to 6 months of age. The persistence of the obligate grasp reflex beyond 6 months of age may signal corticospinal tract dysfunction. Observation of the child's ability to raise the arms while reaching for an object helps assess proximal muscle strength. Congenital malformations of the fingers and hands from webbing to clinodactyly can be readily determined during this portion of the examination.

Direct examination of the hips should include assessment of the range of motion; decreased excursion may signify spasticity or subluxation of the hip joints. Galeazzi's sign is performed in a supine child by flexing the hips 90 degrees with feet on the examination table, noting any asymmetry of femur length. Hip disease such as subluxation often results in a shorter leg and may exist separately or as a result of spasticity. Conversely, increased excursion may represent hypotonia or ligamental laxity.

Initial examination of the legs consists of assessment of muscle symmetry and mass. Spontaneous motor movements are also evaluated, making note of the quality and symmetry of any movement. Assessment of tone is similar to that done with the arms and hands; one should gently shake the feet and passively move the joints of the lower extremities from hip to knee to ankle.

Deep tendon reflexes that are excessively brisk may indicate upper motor neuron unit disease, especially when associated with clonus. Asymmetry is particularly worrisome because of the association with pathologic conditions. Absent deep tendon reflexes are seen with anterior horn cell disease or peripheral neuropathy. The crossed adductor reflex is elicited when the patellar reflex is stimulated and resultant contraction of the adductor muscles occurs in the opposite leg. This response can be normal until approximately 1 year of age. However, persistence of the response, particularly unilaterally, suggests the presence of corticospinal tract involvement.

The plantar response can be as important in infants as in adults. There is no consensus about when an extensor response is a normal finding, although an asymmetric extensor toe sign is always abnormal, as is an extensor toe sign that persists beyond 12 months of age (Hogan and Milligan, 1971). A pathologic extensor plantar sign is indicative of upper motor neuron unit disease. Several beats of ankle clonus are often present in the neonatal period and should disappear by 2 months of age. The persistence of ankle clonus and extensor plantar responses in an older child suggests upper motor neuron unit disease even in the absence of hyperreflexia.

Cerebellar function is difficult to assess in infants; it is easiest when a cooperative child can be observed sitting, standing, walking, or reaching for objects. The examiner can also observe the child during play to see resting or intention tremor, dysmetria, titubation or truncal sway while sitting, and fine motor coordination. Decreased tone may accompany other signs of cerebellar dysfunction.

Sensory Testing and Cutaneous Examination

Light touch can be tested by gently stroking the extremities; this should lead to a reaction, with signs of recognition ranging from eye deviation and facial response to anxious withdrawal of the limbs (Figure 3-4). Application of a tuning fork often causes arrest of motion and a wide-eyed look of wonder in the child who cannot otherwise describe the feeling.

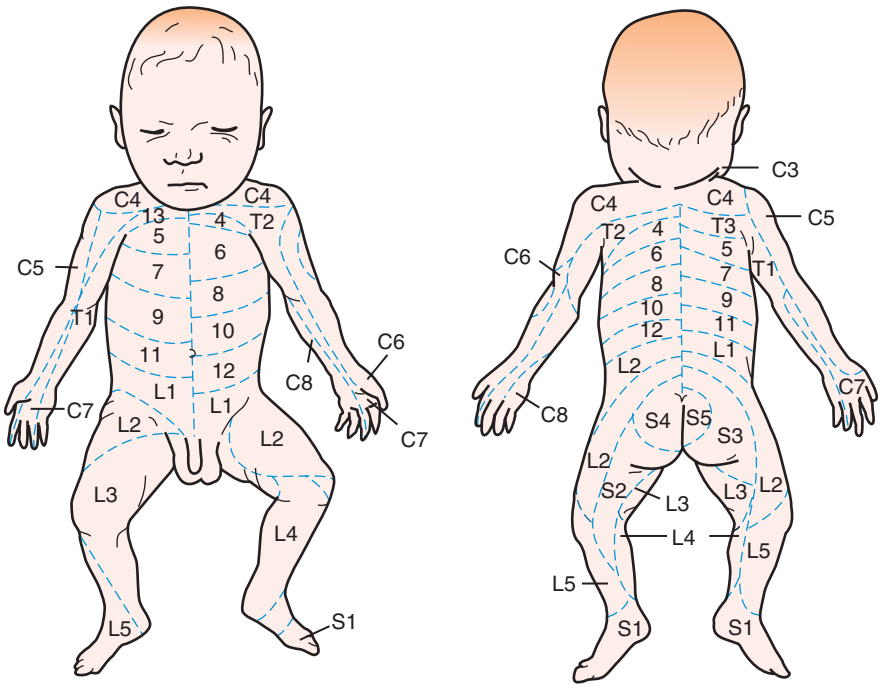

Figure 3-4. Segmental distribution of the cutaneous nerves of an infant. *(Modified from Fanaroff AA, Martin RJ. Neonatal-perinatal medicine: diseases of the fetus and infant, 5th ed. St. Louis: Mosby, 1992.)*

Proprioception cannot be directly evaluated at this age, but observations of sitting positions, gait, and posture may provide some clues. Pain response from light application of a pin or gentle pinching should be reserved until late in the examination, lest causing pain confirms a child's suspicions about the examiner, and upsets an already worried parent.

During the sensory examination, careful observation of the child's skin is important to rule out a neurocutaneous disorder. Are there hypopigmented macules of tuberous sclerosis present, café au lait spots as seen in neurofibromatosis, or a port wine stain in the area of the first division of the trigeminal nerve that might suggest Sturge-Weber syndrome? Particular examination of the spine is necessary to check for scoliosis, sinus tracts, scars, dimples, and hemangiomas. Unusual skin lesions or hair growth over the spine suggest the presence of an underlying mesodermal defect, such as diastematomyelia or spina bifida occulta. The spine should be palpated along its entire course for defects.

Abdominal and cremasteric reflexes are present at birth. The abdominal reflex is elicited by stroking the skin of the upper, middle, and lower portions of the abdomen laterally from the midline. Each stroke elicits a muscle contraction mediated by a different group of thoracic nerves from T8 to T12. The response results in the retraction of the umbilicus toward the stimulated side. The cremasteric reflex is elicited by upwardly stroking the inner thigh, beginning 3 to 5 cm below the inguinal crease. The cremasteric reflex results in an elevation of the testicles due to contraction of the overlying smooth muscles. Cremasteric reflexes are mediated by spinal nerves L1 to L2.

Stage 2

For stage 2 of the evaluation the child should be placed on an examination table with the caregiver close by to provide reassurance to the child and assistance to the examiner, if necessary. Motor evaluation of the older child can also be carried out on a larger, carpeted surface. By 3 months of age, an infant in the prone position should be able to hold the head and chest off the table. Good head control when held in the sitting position should be evident by 4 months of age. The child should be able to sit unsupported and maintain adequate balance by 8 to 9 months of age. Independent achievement of the sitting position should occur by 10 months of age. The child should crawl by 10 months, pull to a standing position by 10 months, and creep by 11 months. The child should walk with support by 12 months and without support by 13 to 14 months.

Trunk, shoulder, and pelvic girdle tone and strength are directly evaluated. The child is observed while held in vertical and horizontal suspension. A hypotonic infant often droops over the examiner's arm when held in horizontal suspension. In vertical suspension, the hypotonic child may slide through the examiner's hands (Volpe, 2008). The child may be unable to maintain a standing posture when the feet are placed on the table surface; this must be distinguished from active withdrawal of the legs that may also prevent successful standing. If spasticity is present, there may be arching of the extended head, neck, and back while in horizontal suspension. Spasticity may also cause extension of the lower extremities with "scissoring" (excessive abduction) and toe walking.

Motor Performance Instruments

Through the years, several instruments have been devised that are useful for evaluating motor performance in relation to chronologic age. These instruments have provided norms for evaluating the expected rate of motor development for a number of different assessments and maneuvers.

Developmental Reflexes

Developmental reflexes represent maturational stages of the developing nervous system (Prechtl, 1997). Occasionally, developmental reflexes can have localizing value, but usually they are nonspecific. Abnormal findings include the absence or poor manifestation of the expected response, persistence of a reflex that should have disappeared, or an asymmetric response (Table 3-1).

Correctly eliciting the **Moro reflex** requires holding the infant in the supine position, lifting the head, and then allowing the head to fall approximately 30 degrees while cradling the head in the examiner's hands. The expected response is initial extension and abduction of the arms with extension of the fingers, followed by adduction of the arms at the shoulder. Asymmetry at any age, or persistence beyond 5 to 6 months, is always abnormal.

The **asymmetric tonic neck reflex** (ATNR) may be detected in the neonatal period but reaches its peak at 2 months and is absent by 6 months of age. To elicit the reflex, the head is turned to one side while the infant is lying in the supine position. There is extension of the arm and leg on the side toward which the face is turned, while the contralateral extremities flex ("fencer's posture"). A normal infant should not maintain the position beyond a few seconds (i.e., obligate ATNR).

The **palmar grasp reflex** is elicited by placing an object or the examiner's finger in the palm of the infant's hand; this leads to an involuntary flexion response. This reflex subsides by 3 to 6 months of age and is replaced by voluntary grasping, which is necessary to allow transfer of objects from hand to hand.

In slightly older infants, the **Landau reflex** can first be elicited between 5 and 10 months of age, and can usually be seen up to 2 years of age. With one hand supporting the abdomen in the prone position, the examiner flexes the infant's head with his or her other hand. The normal response is flexion of the legs and trunk.

The **placing reflex** response can be demonstrated by holding the upright infant in a manner that causes the dorsal surface of the infant's feet to touch the underside of a tabletop. The infant flexes the legs at the hips and knees so that contact with the underside of the surface ceases.

One of the most useful maneuvers is the **traction response** (Zafeiriou, 2004). This is elicited with the infant in the supine position; the examiner grasps both hands and pulls the infant gently and slowly upward, to a sitting position. Marked head lag with little resistance to the examiner's pulling efforts characterizes the newborn response. By 1 month, the infant's head shows transient neck flexion followed by extension as the infant is pulled forward. Usually, by 3 to 5 months of age at the latest, the infant is able to participate actively with arm flexion at the elbow, and by holding the head and trunk in a straight line as the examiner pulls the child to the upright position. At this point there should be no head lag, and little or no forward motion of the head as the child reaches the upright position.

A valuable measure of vestibular function in the newborn can be obtained by holding the infant in a supine position with the feet closest to the examiner. As the examiner rotates the infant laterally in each direction, the eyes of the infant deviate in the direction of rotation, accompanied by intermittent nystagmus to the opposite side. This maneuver also allows extraocular movements to be assessed.

TABLE 3-1 Eliciting Primitive Reflexes

Reflex	Position	Method	Response	Age at Disappearance
Palmar grip	Supine	Placing the index finger in the palm of the infant	Flexion of fingers, fist making	6 months
Plantar grip	Supine	Pressing a thumb against the sole just behind the toes	Flexion of toes	15 months
Galant	Prone	Scratching the skin of the infant's back from the shoulder downward, 2–3 cm lateral to the spinous processes	Incurvation of the trunk, with the concavity on the stimulated side	4 months
Asymmetric tonic neck	Supine	Rotation of the infant's head to one side for 15 seconds	Extension of the extremities on the chin side and flexion of those on the occipital side	3 months
Suprapubic extensor	Supine	Pressing the skin over the pubic bone with the fingers	Reflex extension of both lower extremities, with adduction and internal rotation into talipes equinus	4 weeks
Crossed extensor	Supine	Passive total flexion of one lower extremity	Extension of the other lower limb, with adduction and internal rotation into talipes equinus	6 weeks
Rossolimo	Supine	Light tapping of toes 2–4 at their plantar surfaces	Tonic flexion of the toes at the first metacarpophalangeal joint	4 weeks
Heel	Supine	Tapping on the heel with a hammer, with the infant's hip and knee joints flexed and the ankle joint in neutral position	Rapid reflex extension of the lower extremity in question	3 weeks
Moro	Supine	Sudden head extension produced by a light drop of the head	Abduction followed by adduction and flexion of upper extremities	6 months
Babinski	Supine	Striking along the lateral aspect of the sole, extending from the heel to the head of the fifth metatarsal	Combined extensor response: simultaneous dorsiflexion of the great toe and fanning of the remaining toes	Presence always abnormal

(Data from multiple sources: Futagi Y, Tagawa T, Otani K. Brain Dev. 1992;14:294; Vojta V. Die cerebralen Bewegungstoerungen im Kindesalter, 4te Auflage. Stuttgart: Ferdinand Enke Verlag; 1988; Zafeiriou DI, Tsikoulas IG, Kremenopoulos GM. Pediatr Neurol. 1995;13:148; Zafeiriou DI, Tsikoulas I, Kremenopoulos G, et al. Brain Dev. 1999a;21:216; Zafeiriou DI, Tsikoulas I, Kremenopoulos G, et al. J Child Neurol. 1999b;14:514; Zafeiriou DI, Tsikoulas I, Kremenopoulos G, et al. Brain and Development. 1998;20:307; and Zafeiriou DI. Pediatr Neurol. 2000;22:75.)

There are other developmental reflexes, but those discussed here appear to be the most often evaluated and the most useful.

Stage 3

Examination of the optic fundi should be performed with the infant supine, possibly lying in the caregiver's lap or held over the caregiver's shoulder with the infant's head held tightly against the caregiver's head. Abnormalities of the fundi, including vascular changes, elevation of the optic disc, and retinal changes, along with abnormalities of the lens and media, should be assessed. Mydriatic agents and sedation are rarely employed in the office evaluation, although they are both occasionally necessary. During the first few months of life, the optic discs may be somewhat gray. This normal finding should not be confused with optic atrophy.

The general portion of the examination follows. A heart murmur may signify congenital structural anomalies more widespread than just in the heart. Stridor heard on auscultation may accompany weakness of the upper respiratory musculature. The presence of hepatosplenomegaly should be determined because many storage diseases, which also affect the brain, may be the cause of organ enlargement. When spinal lesions are suspected, the anal sphincter should be examined for tone and the presence of an anal cutaneous reflex (the so-called "anal wink"). Congenital anomalies of the genitalia should be noted. The remainder of the general examination, particularly the intrusive aspects, such as evaluation of the auditory meati, tympanic membranes, mouth, and teeth, can be done at this time.

Stage 4

Spontaneous motor abilities are assessed in this stage of the examination. The crawling child can be put on a carpeted floor or a suitable pad; if the child stands, or walks, he or she should be placed on the floor. The child should be allowed to ambulate or encouraged by rolling a ball across the room or having the child follow a parent across the room. Spastic diparesis, hemiplegia, waddling, footdrop, limp, or ataxia may be evident. The manner in which the child stoops and bends to retrieve a ball or block may show premature hand dominance, athetosis, tremor, or weakness of the legs. Whenever there is a question of proximal weakness, the child should be observed when arising from the floor to a standing position to determine the presence of Gowers' maneuver.

Unlike in the examination of older children, the testing of individual muscle groups in infants is usually impracticable. Nevertheless, evaluation of spontaneous movements and use of some specific maneuvers (e.g., traction response, wheelbarrow maneuver, standing from the floor or a seated position)

can provide information about spasticity, weakness, and incoordination. As always, a comparison of the examination findings must be made with expected age-related norms.

Further examination of muscle strength can be accomplished by using the parachute response; the examiner holds the child in the prone position over an examination table and gently thrusts the patient toward the table surface. A fully developed response (expected at 8 months) consists of arm and wrist extension, allowing the outstretched palms to make contact with the table as the infant supports his or her body weight with the arms and shoulders. Formal individual muscle testing can be used in the older child whenever necessary.

The sensory examination can be difficult. Begin with examination of touch, position sense, and vibration sense. A tuning fork placed on the appropriate bony prominence may elicit a look of surprise or bemusement. Evaluation of pain should be done last and only after the examiner demonstrates to the child the method that will be used.

During this phase of the examination, the Romberg maneuver can be performed. The older child is asked to stand in one place with the feet together and close the eyes; a positive Romberg's sign occurs when the child sways or falls with the eyes closed. The examiner should also observe the child for titubation, nystagmus, and dysmetria while reaching for objects. Cooperative children older than 3 years should be able to perform finger-to-nose testing with the eyes closed. The heel-shin test is frequently not possible in children younger than 4 years.

Assessment of the deep tendon reflexes is best carried out with the infant or toddler in the caregiver's lap. The biceps response in most infants can be difficult to elicit, but the triceps and brachioradialis reflexes are usually readily detected. The patellar and Achilles responses are typically present and easy to elicit. Toe signs can be evaluated as in older children.

GENERAL CONSIDERATIONS

Throughout the examination, the clinician should evaluate the child's alertness, interest in the surroundings, and ability to learn during the examination. The child's speech pattern should also be assessed. By 15 months of age, the child should have a consistent vocabulary of 2 to 6 words, and by 18 months, up to 20 words. Short phrases consisting of two or three words are usually part of the child's repertoire by 21 to 24 months. By 2 years of age, most children have a vocabulary of up to 50 words. Using specific scales to evaluate intelligence and development levels is of some help, but a single office assessment may not be reliable. It is therefore important that the examiner become proficient in informal means of evaluating these characteristics (Maria and English, 1993).

Young children are not always easy to examine. However, taking a staged approach as suggested here, and being sensitive to the child's temperament, often results in a successful examination.

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The complete list of references for this chapter is available in the e-book at www.expertconsult.com.

See inside cover for registration details.

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E-BOOK FIGURES AND TABLES

The following figures and tables are available in the e-book at www.expertconsult.com. See inside cover for registration details.

Fig. 3-1 Entire hand grasp of a 4-month-old infant.
Fig. 3-2 Use of two fingers and thumb in the grasp of a 7-month-old infant.
Fig. 3-3 Pincer grasp with the thumb and forefinger of an 11-month-old infant.
Fig. 3-5 Extended legs, scissoring, toe stance, and fisting in an infant with spastic quadriplegia.
Fig. 3-6 The Moro response to rapid extension of the neck in a 2-day-old infant. The abduction phase of arm movement is illustrated. A cry usually accompanies the response, and the leg position varies.
Fig. 3-7 The traction maneuver causes little response in a 2-day-old infant. There is little or no perceptible flexion of the neck or the arms at the elbows.
Fig. 3-8 Abnormal parachute response.
Box 3-2 Most Commonly Used Motor Performance Tools
Box 3-3 Tips for Examining Noncooperative Children
Table 3-2 Eliciting Postural Reactions

4 Neurologic Examination of the Term and Preterm Infant

Kenneth F. Swaiman and John Phillips

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THE TERM INFANT

The essence of the newborn neurologic examination, whether for a premature or a term-born child, remains exactly the same as with older children. It begins with observation, followed by an examination that may need to be done "out of order" depending on circumstances. Several examinations done over time may be required to most accurately characterize an infant's neurologic status.

Observation

Careful observation begins the examination, noting any congenital abnormalities that are present and the general level of alertness. Cranial nerve assessment can be largely obtained via observation, making note of spontaneous eye movement, facial symmetry, and response to sounds and light. Observation also provides much information regarding the motor system. For example, term infants have predominantly flexor tone with frequent flexion at the knees and elbows. Intermittent fisting of the hands, including adduction and infolding of the thumbs (i.e., cortical thumbs), is often present. Limb position and posturing should be roughly symmetric. While supine, a healthy infant may have spontaneous limb movements that are asymmetric with a jerking quality—this is normal. Excessive jitteriness or tremulousness, however, particularly of the hands or jaw may suggest hyperexcitability of the central nervous system (CNS).

Cranial Vault Evaluation

The occipitofrontal circumference should be plotted on a graph standardized for gender, race, and gestational age to determine whether the measurement falls within the normal range (i.e., two standard deviations above or below the mean). Variances within two standard deviations may be due in part to head shape; for example, the same volume will require a larger circumference in an oblong head compared with a round head. Significant deviation from normal always requires further evaluation.

Some deformities of the cranium are related to the birthing process. Vaginal deliveries may be associated with scalp and subcutaneous edema causing *caput succedaneum*, particularly if vacuum extraction is used. *Cephalohematomas* are hemorrhages within the periosteum of individual cranial bones and therefore do not cross suture lines. *Subgaleal hematomas* result from bleeding under the scalp aponeurosis and are often preceded by forceps or vacuum-assisted delivery space. The scalp may be edematous and boggy because of underlying blood. Although most subgaleal hematomas are benign, if large enough they can cause hypovolemic shock. Infants delivered by cesarean section usually have relatively round heads.

The anterior fontanel, readily palpable at birth and often pulsating with the infants heart rate, is concave or flat in relation to the surrounding cranium. The fontanel should be assessed with the child held in the sitting position if there is any question of increased pressure. A bulging fontanel without the child crying raises a concern regarding increased intracranial pressure. The anterior fontanel varies in size but usually ranges from 1 to 3 cm in its longest dimension (Popich and Smith, 1972). The posterior fontanel in the neonate usually is open but admits only a fingertip.

Cranial sutures (e.g., sagittal, metopic, lambdoidal, and squamosal) are readily palpable in the newborn. Overriding sutures, often the sagittal and lambdoidal, are sometimes seen in the first week of life. Gentle palpation should demonstrate that sutures readily separate from one another unless premature closure has occurred, which may cause asymmetric skull growth.

Auscultation over the infant skull, particularly the anterior fontanel and neck vessels, usually reveals a venous hum in a number of locations. Rarely, systolic-diastolic bruits, particularly those that are focal and asymmetric, indicate the presence of an arteriovenous malformation; however, at times these bruits may be heard in normal infants.

Developmental Reflexes

Developmental reflexes are primitive reflexes with complex responses, and largely reflect the integrity of the brainstem and spinal cord; the role of higher centers, although of importance, is not fully known. This includes the Moro, rooting, grasping, tonic neck, stepping, and placing reflexes. Many of these reflexes are present at birth and undergo modification during the first 6 months of life. Detailed discussion of these reflexes is presented in Chapter 3.

Motor Function

Gentle manipulation of the infant's limbs allows for assessment of muscle tone and strength. *Tone* is resistance to passive movement and should be evaluated while the infant is awake but at rest. *Strength* is resistance to active movement, which can be assessed in an infant by noting resistance to spontaneous movements. The optimal position to assess tone and strength is supine with the head in the midposition so that the tonic neck reflex does not augment tone unilaterally.

Horizontal and vertical suspensions are helpful maneuvers when assessing infant motor function. When held in the vertical position, the hypotonic and weak infant tends to slide through the examiner's hands. In the horizontal position, the hypotonic infant droops over the examiner's arms without raising head or legs. Conversely, increased tone may cause opisthotonus, with persistent extension in both vertical and horizontal positions. Scissoring (i.e., crossing of the legs because of excessive, involuntary adductor magnus contraction) may also be evident with increased tone, but usually does not occur until after the neonatal period. The most common cause of generalized decreased tone is depression of CNS function, congenital malformations, or neuromuscular disorders. Increased muscle tone may be seen in a variety of conditions that cause neonatal encephalopathy. Indeed, there