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Authors: Heilbroner, Peter L.; Castaneda, Glenn Y.
Title: Pediatric Neurology: Essentials for General Practice: Essentials for General Practice, 1st Edition

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Dedication
To Irving Fish, MD, who showed us that clinical sense does not preclude common sense; to our patients; and to our families, without whom this book would not have been possible.
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Introduction
Many physicians feel uneasy approaching a young patient who has a complaint suggesting a disorder of the nervous system. The most obvious and oft-cited reason stems from their training, when they did not have an opportunity to evaluate or treat a large enough number of pediatric patients with a neurological problem. In addition, we have found that many physicians were not taught practical concepts that would enable them to feel more confident when seeing pediatric neurological cases.
Physicians in other fields who try to learn more about child neurology from textbooks generally do not find the approach they are seeking. These authoritative sources, indispensable for the pediatric neurologist, are too detailed and devote too much space to uncommon diseases to make it easy to learn the concepts and skills needed to treat the common disorders.

Pediatric neurology textbooks also are not designed to quickly answer the non-pediatric neurologist's questions: When should (MRI) be orderedâ or repeatedâ to evaluate a child's headaches? How concerned should I be about the arachnoid cyst noted in the MRI report? What should I do when a child diagnosed with attention-deficit/hyperactivity disorder (ADHD) is not responding to medication? And what does it really mean when his mother tells me that he has a â processing problemâ ? In a case of developmental delay, when should I suspect a degenerative disorder? And which tests should I order? Is an electroencephalogram (EEG) useful in evaluating a child who has frequent behavioral outbursts? What do I tell a parent who wants to know if her son's tics will lead to Tourette syndrome? A major reason for writing this book was to provide readily accessible answers to everyday questions.

Why Learn Pediatric Neurology?

Pediatric patients with headaches, suspected seizures, fainting spells, tics and head injuries are often evaluated in the office or emergency room. The parents of children with school-related and behavioral problems or suspected ADHD often seek medical guidance, as do parents of infants who are delayed in attaining motor or language milestones. Other patients will, occasionally, present with signs or symptoms of a more serious neurological disorder such as Guillain-Barre syndrome, transverse myelitis, or a central nervous system tumor that must be promptly diagnosed and appropriately treated.

Unless all of these patients can be quickly seen by a pediatric neurologist â something not possible in many places in the United States, and in many other countries â the physician responsible for their care must know how to proceed. He should recognize the differential diagnosis suggested by the history, use the physical examination to explore possibilities suggested by the differential diagnosis, know which tests will provide the most useful information, and be able to offer effective initial and when possible definitive treatment.

Pediatric neurological referral will ultimately be necessary for difficult or serious cases, and most parents would like their pediatrician to understand the neurologist's approach and perspective. To many parents, neurological problems are mysterious and frightening; therefore it is important that the primary care physician clearly understand these disorders and their treatment. Throughout this book we address the questions that parents most often ask. We also review a number of subjects that tend to cause confusion and impede communication with the family: the purpose and interpretation of the EEG, the terms used to characterize learning disabilities, and what causes headaches.

The Scope of this Book

Pediatric Neurology: Essentials for General Practice does not replace a traditional textbook. Our primary purpose is to teach useful clinical skills and thoroughly review the diagnosis and treatment of the more common disorders seen in the office or clinic. We also suggest an approach to neurological syndromes encountered in the emergency room. Such syndromes include the child with an altered mental status or whose gait has acutely deteriorated. Also included are overviews of the surgical management of epilepsy and the treatment of brain tumors that may be helpful for the pediatric resident caring for hospitalized patients.

We do not include some rare diseases, and we discuss other disorders generically (e.g., â Mucopolysaccharidoses,â rather than separate sections each devoted to a specific kind of mucopolysaccharidosis). Our approach is not intended to minimize the importance of rare diseases. To the children and families affected by a rare disorder, their condition is overwhelmingly important, and the physician who cares for these patients provides an invaluable service. That having been said, a heavy emphasis on rare diseases in an introductory book is distracting to the physician who needs to learn about the conditions he will see on a regular basis.

This book briefly discusses the management of neurological conditions encountered in the intensive care unit, such as coma, spinal cord injury and elevated intracranial pressure. If a more comprehensive discussion of ICU neurology is sought, the reader may consult definitive pediatric, medical and neurosurgical reviews and textbooks.

Chapter 1 presents guidelines intended to help the physician take the history of a neurological complaint. We then offer suggestions for a physical examination guided by the differential diagnosis suggested by the history. The doctor who seeks a general overview should start by reading through Chapter 1. The same chapter may also serve as a general reference for the physician who has more neurological experience. The remainder of the book is devoted to specific syndromes and disorders.

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1
Suggestions for History and Physical Examination

Approach to the Family

Parents may be frightened when their child develops a neurological symptom. The worst possibilityâ that their child has a life-threatening or disabling diseaseâ is feared, since everyone knows that the brain is a critically important, intimidatingly complicated, mysterious organ. Advice from â expertâ friends, the overdramatic tone of medical reporting on television, and the Internet tend only to exacerbate the parentsâ anxiety, and as a result, they may arrive in the office in a state of great consternation, expecting the worst. Assuming that the physician does not suspect a serious condition based on his evaluation, his most important role often becomes one of reassurance. It is important to remember that a neurological symptom is not by definition a serious symptom! Just as a typical case of fever is far more likely to result from a benign viral infection than from tuberculosis, a child's tremor is far more likely to be a sign of neurological immaturity than an early sign of Wilson disease, and a headache is much more often a migraine headache than a symptom of a brain tumor. Therefore, we should not jump to order invasive, expensive tests because a symptom happens to be neurological, just as we do not reflexively order a computed tomography (CT) scan of the abdomen for every child with a stomachache.

A calm manner and nontechnical language will usually help relax the family. It is helpful to directly address unspoken fears (i.e., â It's normal to worry about a brain tumor when your child has migraines. You'll be glad to know that a tumor virtually never causes migraine headaches.â).

The physician often can make parents feel better by demystifying med-ical terms. When a parent learns that epilepsy is defined as a history of two or more unprovoked seizures, or that Tourette syndrome is defined as history of motor and vocal tics of more than 1 year in duration, or that cerebral palsy is a general term that means a history of delayed motor development

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(in many cases, relatively mild), the parent is given perspective and will relax, even if the parent suspects that the child's problem may be significant.

The differential diagnosis is not for the parents to worry about

The physician should think twice before reviewing a â listâ (i.e., the differential diagnosis) with the family. He should be especially wary when the differential diagnosis is long and complicated, such as when a patient is being evaluated for a first seizure or for markedly delayed development. It is best to give the family a general sense of the diagnostic possibilities, order appropriate tests, and say that more will be known when the test results come back. Giving the family a long list of possible diagnoses will only exacerbate their anxiety. Every new medical term they hear will lead to 10 more questions, the answers to most of which will be of no concern to them once the diagnosis has been made.

Tell the family to beware of the Internet

The Internet can be a good source of information for families, but websites are inconsistent in quality and often misleading. Parents should try to stay off the Internet until a diagnosis has been made or at least until the differential diagnosis has been narrowed. Like hospital dramas on television, many websites distort the significance of medical symptoms and signs. For example, if the parent of a mildly hypotonic child enters the word â hypotoniaâ into an Internet search engine, she may be directed to sites devoted exclusively to serious neurological conditions such as spinal muscular atrophy, Rett syndrome, and muscular dystrophy. The fact that the great majority of mildly hypotonic children have no serious underlying disorder may not even be mentioned.

Explaining the cause, not using jargon, and acknowledging the limits of our understanding

We often must explain to parents that many neurological diagnoses [e.g., migraine, attention-deficit hyperactivity disorder (ADHD), and tic] are made primarily on the basis of recognizing a characteristic pattern of symptoms and signs, rather than by laboratory and diagnostic test [magnetic resonance imaging (MRI), electroencephalogram (EEG), etc.] results. A primary focus on the clinical presentation is disorienting to many people, who are used to relying on test results (e.g., a positive or negative throat culture) and who will persist in asking about the exact cause of their child's problem. When the cause is known, it should, of

course, be shared with the family. Our understanding
P.3

of the cause of many neurological disorders is incomplete, however, and this fact should not be disguised. Pseudoscientific terms (â a processing problemâ in a case of learning disability or â a chemical imbalanceâ in a case of depression) should generally be avoided in these conversations. These terms do not explain the child's condition to parents and, therefore, only lead to more questions (â What kind of processing problem?â â Can you test my child for a chemical imbalance?â). It is better to simply say, with respect to the previous two examples, that educational testing reveals that your child comprehends information that she reads better than information that she hears or that your child appears depressed. Theories about the underlying cause can then be presented, as long as they are characterized as theories.
When the parents ask why the exact cause of their child's problem cannot be demonstrated, it can be explained, if need be, that we obviously do not biopsy the human nervous system except in the most serious cases (and even if we could, the results would reveal little about the working brain). In this respect, neurology is different from many other medical specialties whose practitioners routinely obtain tissue for diagnosis, and this leads to a somewhat abstract quality to neurological diagnosis making. We cannot definitively demonstrate to a parent that the language area of the brain of their child with a learning disability has a pattern of synaptic connections that differs from some other children; that the brain of their child with a primary generalized seizure disorder contains neurons that discharge abnormally; or that their depressed son's raphe nucleus may not release enough serotonin. (Functional imaging technologies are potentially useful in this regard and one day may be more widely available.) Once again, this limitation does not prevent us from diagnosing our patients accurately and treating their condition to everyone's satisfaction.
Taking the History of a Neurological Problem
Red flags
Many pediatric patients who come to the office or clinic with a neurological complaint are ultimately diagnosed with a condition, such as benign developmental motor delay, tic disorder, migraine, or concussion, that is either treatable or, if left untreated, does not have grave implications for the patient's health. Intermixed with these cases, however, will be some patients whose condition is potentially much more serious. Although we emphasize that this list does not include every serious disorder and there is no simple formula that can be used to quickly determine that a patient has a serious problem, the following six clinical syndromes should always merit extra attention and prompt diagnostic testing (Table 1.1).
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TABLE 1.1 â Red flagsâ : serious syndromes in child neurology

Unexplained focal neurological symptoms or signs
Developmental regression or marked stagnation
Symptoms and signs of elevated intracranial pressure and hydrocephalus
Seizures with implications for the patient's development, such as infantile spasms
Symptoms and signs of meningitis
Unexplained, persistent loss of a neurological function (e.g., marked change in mental status, loss of the ability to walk)

Unexplained focal neurological symptoms or signs
A symptom (such as a headache) is a complaint. The doctor or another individual observes a sign (e.g., â the patient's reflexes are abnormally briskâ or â my 2-year-old child is falling down a lotâ).
A disease process affecting a localized region of the nervous system usually causes one or more focal neurological symptoms or signs. Focal neurological symptoms and signs, especially if not transient, should always be regarded as abnormal. Focal neurological signs and symptoms are often unilateral (noted on only one side of the body). Two classic syndromes are hemiparesis (weakness of the upper and lower extremity on one side) and hemianopsia (loss of a visual field). A lesion within the opposite cerebral hemisphere is the usual cause. Infarction (stroke) in the left hemisphere, for example, often causes weakness of the right arm and leg or a right visual field deficit. However, the physician must keep in mind that brainstem, cerebellar, and spinal cord lesions often cause ipsilateral (on the same side as the lesion) or bilateral symptoms. For example, weakness of both legs might be caused by a tethered spinal cord, incoordination of the right hand might be caused by a right cerebellar tumor, and loss of ability to move the eyes to the right might be caused

by demyelination in the right half of the pons.

A majority of pediatric neurological complaints, such as headache, learning problems, fainting spells, tremor, tic, delayed fine motor skills, and behavioral problems do not result from a localized disease process in the nervous system. Table 1.2 lists some typical pediatric neurological problems and suggests the likelihood that the problem is the result of a lesion in a specific area of the nervous system.

Transient focal neurological symptoms

Transient focal neurological symptoms lasting only a few seconds or minutes are differentiated from persistent focal neurological symptoms. Concussion, migraine, and partial seizure are the most common causes of transient

P.5

neurological symptoms. Concussion and migraine do not result from a structural lesion in the brain, and if the history suggests migraine or concussion, the chances are good that the patient does not have a serious underlying disorder. However, partial seizures can be caused by a structural lesion and should be evaluated promptly with MRI and EEG if they were not febrile seizures.

TABLE 1.2 Various pediatric neurological syndromes and likelihood of a focal disturbance in the nervous system as the cause

Problem	Caused by a focal lesion?
Headache	Relatively unlikely but always must be considered
Learning disability	Possible in cases of previous brain injury; otherwise, almost never
Attention-deficit hyperactivity disorder	Virtually never
Mildly delayed gross or fine motor development with otherwise normal neurological examination	Virtually never
Autism	Virtually never
Blurred vision	Possible; other causes more common
Seizure	Must be considered in the case of a focal-onset or poorly characterized seizure
Hemiparesis	Likely
Ataxia	Likely (but can be caused by medication, migraine, other nonstructural causes)
Double vision	Likely

Chronic Focal Neurological Symptoms

The physician must also differentiate a focal deficit of recent onset from a chronic focal deficit. A chronic focal neurological deficit (such as chronic right hemiparesis) is usually the result of a previously diagnosed problem (for example, a brain injury many years in the past), whereas a recent-onset focal symptom or sign suggests a new problem warranting investigation.

Developmental Regression or Stagnation after Normal Development

A mild delay in attaining infantile and early childhood developmental milestones (especially motor milestones) is frequently reported and is not usually caused by a serious underlying disorder. However, a history of lost, previously attained developmental milestones, severe developmental delay, or marked

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stagnation of development is always cause for concern. Autism, a degenerative or neuromuscular disease, chronic infection (e.g., congenital human immunodeficiency virus), or a brain tumor may present with developmental stagnation or regression.

Symptoms and Signs of Elevated Intracranial Pressure and Hydrocephalus

Signs and symptoms of elevated intracranial pressure (ICP) are reviewed below (see exam of patient with headaches) and in Chapter 2. Brain tumor, pseudotumor cerebri, and hydrocephalus are important conditions that cause an elevation of the ICP in a child or adolescent. Hydrocephalus in a neonate and infant does not result in elevated ICP because the cranial sutures are not yet closed and, instead, causes progressive macrocephaly.

Seizure Disorders with Serious Implications

Infantile spasms in particular must be recognized.

Symptoms and Signs of Meningitis

A child or adolescent with a headache, a stiff neck or a Kernig or Brudzinski sign, and fever should be assumed to have meningitis and should promptly be sent for a lumbar puncture. Meningitis in an infant or toddler may present less dramatically. These patients may not have a stiff neck or exhibit other signs of meningeal irritation. The most characteristic signs of meningitis in very young patients are fever and altered mental status (decreased alertness, lethargy, or irritability).

A headache and a stiff neck unaccompanied by fever suggests cervical disease (the most common cause) or, although seen rarely in the pediatric population, a subarachnoid hemorrhage.

Marked, Unexplained Loss of Neurological Function

The patient who is rapidly losing the ability to walk, for example, must be immediately evaluated. Unexplained mental status changes always warrant close attention.

Suggestions for Taking the History

- The rate of onset of a patient's symptoms often helps define the differential diagnosis (Table 1.3). For example, if a patient has lost the ability to walk over a period of a few seconds or minutes, a traumatic injury, ischemic stroke (brain or spinal cord), or a hemorrhage is the most likely cause. If this problem has developed over the course of hours or days, an immunologically

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mediated disorder such as Guillain-BarrÃ© syndrome, transverse myelitis, or an infection in the nervous system (e.g., a brain abscess) should be suspected. The patient who has become unable to walk over a period of weeks to months may have a spinal tumor. If the patient's gait has been deteriorating over a period of years, a degenerative neuromuscular disease may be the cause. Finally, if the patient has never walked, the cause is probably a nonprogressive condition such as spina bifida or cerebral palsy. In addition, it is important to recognize that migraine, vasovagal syncope, breath-holding spells, seizures, and panic attacks are the most common causes of transient or intermittent neurological symptoms.

TABLE 1.3 Time course of neurological disorders

Acute onset (seconds to minutes)	**Subacute onset (hours to days)**	**Gradual onset (weeks to months)**	**Progression over months to years**	**Stable for years**	**Intermittent**
Trauma	Infection	Tumors	Degenerative disorders	Previous neurological injury/disease (prior stroke, cerebral palsy, spina bifida, etc.)	Migraine
Infarction	Inflammation	Some degenerative disorders	Some tumors		Seizure
Hemorrhage	Demyelination	Smoldering infection (abscess)	Chronic infection (e.g., human immunodeficiency virus)		Vasovagal syncope
	Toxin/drug				Breath-holding spells
					Panic attacks

- The physician should always inquire about nonneurological symptoms. For example, a patient may come to the office because of headaches, but it is only after she is asked if she has been feeling excessively fatigued or has a rash, arthralgias, or a sore throat that the cause of the headaches (Lyme disease, mononucleosis, or another viral syndrome) is suggested.
- The physician should always ask if the patient is currently taking a prescription medication that could cause the presenting complaint. The patient with a tremor may be taking an antidepressant, a stimulant drug for ADHD, or a beta-agonist drug for asthma. The ataxic patient may be taking an antiepileptic drug or a sedative-hypnotic. The patient with headaches may be taking methylphenidate, a vitamin supplement, or an antibiotic. Seizures can be provoked by several medications (see Chapter 6).
- The family history is important in neurological and psychiatric cases. Common conditions that often â run in the familyâ include ADHD, learning disability, migraine, tic/Tourette syndrome, mood and anxiety disorders, syncope, tremor, and several forms of epilepsy. In the much rarer case of a metabolic or degenerative disorder, the family history is often very important.

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- Psychogenic disorders are common. Indeed, the primary problem turns out to be psychogenic, or at least exacerbated by psychological factors, in an appreciable number of cases that, at first glance, suggested an organic disorder. A clue suggesting a psychogenic cause for the patient's apparently neurological problem is the occurrence of that problem only in a particular situation or setting. Some examples include headaches that bother a patient at one divorced parent's house but never at the other parent's house, bizarre behavior at home that is never noticed by a child's teacher, or trouble walking that started immediately after an emotionally traumatic event. Another clue suggesting a nonorganic problem is inconsistency of physical findings, such as a patient's inability to raise a lower extremity off of the examination table despite intact ability to support weight on the same extremity while standing. Some diagnostic testing is generally indicated, but the doctor should not continue to order tests if the results are normal and he strongly suspects a psychogenic disorder.

We should be comfortable talking about psychogenic problems with our patients and their families. When a doctor does not honestly identify a patient's problem as stress or anxiety related, it is confusing to the family and contributes to the stigma associated with behavioral disorders. It should also be explained that saying that a psychiatric/psychological etiology exists for a patient's symptoms does not necessarily mean that the patient is not experiencing real symptoms.

Role of EEG and MRI

MRI and EEG are helpful in making the diagnosis of some, but by no means all, neurological disorders. These tests are often inappropriately ordered (for example, in cases of ADHD, tic disorder, and typical vasovagal spells). The usefulness of these tests is discussed throughout this book. Table 1.4 provides examples of appropriate and inappropriate clinical situations in which to order these tests.

Physical Examination

Focus on the Differential Diagnosis Suggested by the History

Many physicians make the mistake of performing the same physical examination when evaluating a patient with a neurological problem of any kind. We would recommend a more individualized approach. A history of a fainting spell, a history of back pain, and a history of a school-related problem are entirely different complaints, and each suggests a unique differential diagnosis and particular aspects of the examination that should be emphasized. A focused neurological examination is much more likely to provide useful information than a boilerplate examination.

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TABLE 1.4 Magnetic resonance imaging (MRI) and electroencephalogram (EEG) in common pediatric neurological cases

Condition	MRI	EEG
Headache	Ordered to exclude a mass lesion or arteriovenous malformation. Most causes of headache are *not* diagnosable by MRI.	Not indicated.
Learning disability, attention-deficit hyperactivity disorder	Not indicated in typical cases.	Not indicated.
Tic	Not indicated unless a different kind of movement disorder is suspected.	Not indicated unless a focal seizure is suspected.
Seizure (nonfebrile)	Usually indicated, other than in cases of idiopathic primary generalized epilepsy (see Chapter 6).	Generally indicated.
Syncope	Very low yield.	Not indicated unless seizure suspected.
Autism	May show nonspecific abnormalities; no value in making the diagnosis of autism.	Indicated if history of seizures (*not* tantrums/outbursts) or in *rare* cases of suspected Landau-Kleffner syndrome (see Chapter 9).
Unexplained focal	Usually indicated (except in clear cases of	Indicated if patient has a history of

neurological signs/symptoms	peripheral nerve injury).	seizures.
Developmental delay	In cases of mild/moderate motor delay, yield is low. Consider in cases of global (motor and cognitive) delay and especially developmental regression.	Indicated only if patient has a history of seizures.

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TABLE 1.5 Suggestions for focused neurological examination

Complaint	**Emphasis of examination**
Headache/head injury	Inspect the cranium in cases of trauma, in particular for signs of a basilar skull fracture. Evaluate neck for stiffness, tenderness, and adenopathy; assess tenderness to percussion of sinuses. Examine optic fundi for papilledema or hemorrhage. Perform full assessment of mental status in cases of head injury. If no specific neurological complaints, a concise examination of cranial nerves, motor system, and cerebellar function is often adequate.
School and behavioral problems	Behavioral and cognitive assessment is stressed; screen for learning disability. Look for behaviors suggestive of attention-deficit hyperactivity disorder (ADHD), anxiety, or depression. Check for â soft signs.â
Tic/movement disorder	Behavioral assessment should be included given comorbid disorders (ADHD, etc.). Observe abnormal movements. Assess tone for evidence of rigidity. In cases of tic disorder (most common), concise motor and cranial nerve exam is sufficient. In cases of other movement disorders, a comprehensive motor examination is required.
Seizures	Examine skin for signs of a neurocutaneous disorder. Observe for hyperventilation if absence seizures are suspected. Especially in cases of focal-onset seizures when no brain imaging study has yet been completed, a comprehensive neurological examination is recommended to search for focal signs.
Syncope	Take orthostatic blood pressure readings; examine skin, thyroid gland, heart, and lymph nodes. Concise cranial nerve, motor, and cerebellar assessment.
Hemiparesis	*Comprehensive* cranial nerve, motor, sensory, reflex, and cerebellar examination.
Ataxia	*Comprehensive* cranial nerve, motor, sensory, reflex, and cerebellar examination.
Neck or back pain (especially if accompanied by complaints of weakness or numbness)	Range of motion of neck, neck and back tenderness, straight leg-raising test; and *comprehensive* motor, reflex, and sensory examination.

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The examination of a child with a school-related problem should include screening for a learning disability and a careful assessment of behavior looking for signs of ADHD, anxiety, or depression. The examination of the patient who has fainted should include orthostatic blood pressure readings, cardiac auscultation, and evaluation of the skin, thyroid gland, and lymph nodes looking for signs of a nonneurological medical problem (e.g., hypothyroidism, mononucleosis) that might cause the patient to faint or feel lightheaded. The examination of a patient with back pain should include an assessment of strength, gait, sensation, and deep tendon reflexes in the lower extremities; spinal tenderness; and the straight leg-raising test to assess nerve root irritability. Conversely, it is generally unnecessary to spend 20 minutes testing the power of every muscle group of a child who was referred for a school or behavioral problem, to perform a comprehensive sensory examination if the patient has a history of a typical fainting spell, or to perform a lengthy cognitive evaluation when the primary complaint is back pain, assuming that the history does not suggest any additional concerns.

Am I going to Miss Something Important?

We also suggest a concise examination of all neurological systems, which is described in the following sections, that should always be performed to make certain that no important physical findings or occult problems are missed. In cases that seem unusual or atypical or when standard therapy does not appear to be

effective, we also urge that the physician â step backâ and perform (or repeat) a comprehensive examination that may reveal evidence of a more unusual disorder.
Table 1.5 includes examples of common complaints and aspects of the physical examination to be emphasized.
General Examination
This section reviews aspects of the general physical examination that are particularly important in neurological cases. An approach to the neurological part of the examination follows. Once again, we emphasize that the entire examination is rarely necessary. A sense of the likely differential diagnosis should guide the examiner.

- Vital signs should always be recorded. Fever suggests an infection either directly involving the nervous system or, more often, resulting in a secondary neurological complaint, such as the headache associated with a nonspecific viral syndrome. Low blood pressure may explain fainting.
- The head circumference of neonates, infants, and toddlers should be measured. The head circumference should also be measured in cases of premature birth, perinatal complications, and delayed development, or when the patient's head appears large, small, or unusually shaped.
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  Measurement of the head circumference is always from glabella (between the eyebrows) through inion (external occipital protuberance). Poor measurement technique is among the most common causes of a â largeâ or â increasingâ head circumference (see Chapter 11 and Fig. 11.1).
- The patient's general appearance should be noted. Obesity often causes sleep apnea, which can lead to fatigue, headaches, and difficulty concentrating. Obesity may also suggest a behavioral or genetic disorder (e.g., Prader-Willi syndrome or an eating disorder). An emaciated appearance may result from a large number of underlying medical and psychiatric disorders.
- The skin always must be examined. CafÃ©-au-lait spots, melanotic whorls, port wine stains, and hypopigmented macules are often signs of a neurocutaneous disorder (Figs. 1.1, 1.2 and 1.3; see Chapter 14) associated with seizures, tumors of the nervous system, developmental delay, and behavioral problems.

Figure 1.1 Neurofibromatosis type I: cafÃ©-au-lait spots and axillary freckling.

- Dysmorphic facial features, including hyper- or hypo-telorism, low nasal bridge, and deformities of the face, ears, mandible, teeth, or palate, suggest a syndrome of congenital malformation. Other physical features suggestive of a congenital syndrome include an unusual appearance or distribution of scalp
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  hair; small, malformed, extra, or too few digits; simian crease; and abnormally formed limbs. Other patients will have a history of malformed internal organs. Such children often have a history of

delayed motor and language development, cognitive impairments, and behavioral problems.

Figure 1.2 Tuberous sclerosis: adenoma sebaceum.

- The heart, lungs, and abdomen must always be examined. A cardiac murmur, while often â innocent,â may result from a congenital heart defect that is associated with neurological or behavioral problems (in cases of Williams syndrome, for example) or could be a source for cerebral emboli predisposing to stroke. Hepatosplenomegaly is a rare but important finding that, to a pediatric neurologist, suggests a lysosomal storage disease.
- Signs of a congenital spinal defect (tuft of hair, lipoma, and sacral dimple) are especially important if the presenting complaint is an abnormal gait or loss of sphincter control. These external stigmata of spina bifida are also often noted in asymptomatic patients, and an approach to diagnostic testing is suggested in Chapter 11.
- Orthopedic abnormalities (e.g., scoliosis, lordosis, or pes cavus) can be an early sign of a spinal tumor or a progressive neuromuscular disease such as Charcot-Marie-Tooth disease. Spinal deformities also develop in the later stages of Rett syndrome, muscular dystrophy, and other degenerative disorders.

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Figure 1.3 Tuberous sclerosis: large hyperpigmented lumbar shagreen patches and hypopigmented macules (arrow).

- Asymmetrical development of the extremities can be a sign of an underlying neurodevelopmental or acquired disorder. Potentially significant signs include leg-length discrepancy, atrophy of muscles in one extremity or on one side of the body, and asymmetry of digits.

- Nuchal rigidity is assessed in cases of headache. If the neck of an older child, teenager, or adult flexes easily and without pain, meningitis or subarachnoid hemorrhage is unlikely to be the diagnosis. If there is resistance to neck flexion, however, other tests of meningeal irritation should be performed. Kernig sign is a reflexive flexion of the thigh by the patient to relieve pain caused by flexion of the neck by the examiner, and Brudzinski sign is pain elicited by extension of the leg after the thigh is first flexed to 45 degrees above the horizontal (all by the examiner).
  It is important to be aware that these signs of meningeal irritation often cannot be elicited from infants less than 1 year of age, when neck and axial muscles are not well developed. Therefore, the diagnosis of meningitis in an infant cannot be excluded by the finding of a supple neck.
- The examination of the patient with a history of headaches must always include inspection of the optic fundi to make certain that there is not papilledema. In cases of headache, maxillary sinus (and frontal sinus,
  P.15

  in adolescents) tenderness to percussion also should be assessed. The examiner may listen over the orbits and mastoids for a cranial bruit caused by an arteriovenous malformation (AVM). In many cases, however, cranial bruits are benign.
- If the patient has had a head injury, the cranium should be palpated for signs of a skull fracture (marked tenderness, swelling, step-off). The examiner should recognize the signs of a basilar skull fracture: purplish â raccoon eyes,â discoloration over the mastoids (Battle sign), and blood behind the tympanic membrane. A cerebrospinal fluid leak is suggested by a complaint of fluid dripping from the nose or ears. Cerebrospinal fluid, and not nasal mucus, will give a positive result for glucose when tested with a urine dipstick.
- In cases of back injury or back pain, the straight leg-raising test is used to help determine whether there is nerve root involvement (radiculopathy). With the patient supine, the lower extremity is passively elevated to 45 degrees above the horizontal. An increase in pain, especially pain radiating to the lower extremity, caused by this maneuver suggests nerve root involvement. In contrast, a fracture of a lumbar vertebra will often cause pain or tenderness that is exaggerated by spinal percussion and that often does not radiate to the thigh or leg.

Neurological Examination

The standard neurological examination is divided into six parts: behavioral/ mental status, cranial nerves, motor system, reflexes, sensory system, and cerebellar system (1).

The neurological examination should be a useful means of exploring the differential diagnosis and not a tedious chore. Therefore, we suggest a concise version of the examination, which is adequate for many situations, as well as (in some cases in later chapters) suggestions and indications for a more comprehensive assessment. We suggest that the examiner think carefully about the differential diagnosis, use parts of the concise examination to evaluate aspects of the nervous system unlikely to be involved, and perform a comprehensive examination when appropriate.

Behavior

Concise assessment

The patient's level of consciousness (alert, drowsy, lethargic, obtunded, unresponsive), affect (calm, agitated, distractible, hyperactive, depressed, anxious, hostile, etc.), social interactiveness (presence of eye contact, quality of interaction with the examiner and parents), and language skills (direction following ability; comprehension; speech quantity, content, and articulation) are always assessed. Observation and conversation are often an
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adequate assessment, especially if the parent or patient has no specific complaint of a cognitive or behavioral problem.

More comprehensive assessment

A more comprehensive behavioral/cognitive assessment is necessary in the case of a patient referred for a developmental, behavioral, or school-related problem. In cases of head injury or change in mental status and if there has been general neurological deterioration, the patient's cognitive status is of primary importance.

Chapters 3, 5, 9, and 12 offer guidelines for a cognitive, learning, and mental status assessment.
If the examiner suspects that a toddler is late to develop speech, the child's hearing must be assessed. This must be done by means of formal audiometric testing. In many other cases, delayed speech is an early sign of autism. Speech apraxia is a general term for difficulty articulating sounds. It is a fairly common developmental problem that is often associated with other neurodevelopmental delays and sometimes behavioral problems. Dysarthric (uncoordinated) speech is noted in cases of cerebellar disease; it is an acquired problem in most cases. Aphasia is an acquired language disorder that is caused by an injury to the cerebral cortex, usually in the left hemisphere, and that is rarely diagnosed in children and adolescents.

Cranial Nerves

Concise Assessment for Children and Adolescents

For patients who appear generally well and have no focal neurological complaints or signs, the cranial nerve examination can be limited to pupillary reaction to light, the optic fundus, eye movements through all gaze positions, and assessment of facial, palatal, and tongue movement. This series of tests should take only a few moments for the experienced doctor. However, the funduscopic examination may require practice before it can be done quickly and accurately. At a minimum, the physician should learn to assess the optic disc and vessels for signs of papilledema, identify a macular cherry red spot, and recognize retinal hemorrhages. Until the physician has become proficient, it is suggested that a funduscopic examination be practiced in every case, even if the patient is being evaluated for a nonneurological complaint.

More Comprehensive Cranial Nerve Assessment

If the presenting complaint involves vision, facial weakness, or swallowing; if any focal neurological signs are noted during the rest of the examination; or if there has been general neurological deterioration, a more comprehensive assessment of the cranial nerves becomes necessary.

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The visual fields are evaluated by confrontation testing, and visual acuity is assessed with a vision card (â near cardâ) held at about 14 inches from the patient. The near card tests the integrity of the neurological systems underlying vision (pathway leading from the retina to the occipital lobes) and not the patient's visual acuity. If the patient complains of double vision but the extraocular movements appear normal, the reflection of a pocket light off the right and left cornea should be checked for symmetry in all gaze positions. Double vision always results from misalignment of the two eyes. The reflection of the light should be symmetrical (with respect to the midpoint of the pupils) in all gaze positions. Double vision is one of the more potentially serious symptoms in neurology and always merits close attention. If the extraocular movements appear disconjugate or the corneal light reflex is asymmetric, an MRI should be ordered to look for a brainstem, cranial nerve, or orbital tumor unless an ophthalmological cause has been established.
If there is a recent history of weakness of all the muscles of one side of the face, Bell palsy is almost always the cause. The patient with Bell palsy should not be able to raise her eyebrow on the side of the palsy. A cerebral hemispheric lesion, in contrast to Bell palsy, causes paresis of only the lower half of the face (see Chapter 14) as well as, in the majority of cases, weakness of the hand or arm on the same side as facial weakness.
Asymmetric movements of the palate and tongue are rarely noted in children. When present, these findings suggest a brainstem lesion and are an indication for MRI.
In the older child or teenager, hearing is tested if there is a complaint of hearing loss, ringing in the ears, or vertigo. Office tests that may better define the hearing problem are reviewed in Chapter 14. In cases of suspected hearing loss, formal audiometric testing is necessary. Unilateral hearing loss is rare but should raise the possibility of a vestibular neuroma, usually a manifestation of neurofibromatosis type II (see Chapter 14).

Cranial Nerve Examination in Younger Patients

The cranial nerve examination of infants, toddlers, and young or uncooperative children primarily relies on observation. The physician can assess the extraocular movements as the child looks about the room or follows a light and the movements of the face, tongue, and uvula when the child smiles or cries. When it is difficult to observe how the pupils react to light (as when the patient has dark irises), examination with a flashlight in a dark room is often successful. Visual fields can be assessed by bringing novel objects slowly from behind the patient's head across the child's field of vision, first on one side and then the other. Another method may be by simultaneously bringing similar objects in each of the examiner's hands into the child's peripheral field of vision. A child with intact visual fields would look at one and then the other object.

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The examiner must possess considerable skill if he wishes to properly examine the optic fundus of an infant or toddler. In practice, the â funduscopicâ examination of these patients is often limited to checking for the red reflex. If the examiner is lucky, he will glimpse the optic disc. The patient should be referred to an ophthalmologist for a dilated funduscopic examination if the presence of a macular cherry red spot needs to be determined (as in the case of a degenerative disorder), if retinal hemorrhages are suspected, or if the infant has a problem involving vision.

Motor System

Concise Assessment for Children and Adolescents

A thorough motor examination is always indicated if the presenting complaint involves strength, coordination, or gait; if focal neurological symptoms are reported or focal signs are noted on examination; or in cases of general neurological deterioration.

In many other cases, however, such as when the patient comes to the office because of a school or behavioral problem, syncope, or migraine, the motor examination is unlikely to contribute to the diagnosis. In such cases, it is reasonable to make the motor examination concise, concentrate on more relevant aspect of the examination, and leave plenty of time for a talk with the family.

A concise motor examination should include biceps and grip strength; ability to stand, walk, toe and heel walk, jump or hop, and run; and observation for abnormal posturing. Resistance to passive limb movement, or muscle tone, should also be noted. An adequate evaluation of fine motor ability in such cases is accomplished by asking the patient to wiggle the fingers of both hands (â play the pianoâ), rapidly touch the index finger and thumb together, and â walkâ his or her hand rapidly across the table top using the index and middle fingers. The young patient should be asked to perform these movements with both hands simultaneously and each hand independently. Fine motor testing often will demonstrate â soft signsâ (see Soft Signs section) in developmentally delayed children. Pencil skills are evaluated, when necessary, by asking the patient to write a few sentences about a subject of her or the examiner's choice. Left handedness is often relevant to the diagnosis of learning disability and should be noted.

If there is no complaint of weakness or incoordination and these tests reveal no abnormality, further testing of the motor system is often unnecessary, and the examiner may feel comfortable moving to the next phase of the examination.

More Comprehensive Motor Assessment

If the primary complaint is weakness, a problem walking, or incoordination or if there are focal complaints or signs, a more comprehensive motor evaluation becomes necessary. In these cases, the power of the trapezius (shoulder

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shrug), sternocleidomastoid (turning of the head), deltoid (abduction of the arm at the shoulder), biceps (flexion of the forearm), triceps (extension of the forearm), wrist flexors and extensors, and hand intrinsic muscles should be tested. In the pronator drift test, the patient is asked to extend both arms with palms facing upwards and eyes closed. If one arm drifts downward and pronates, weakness on that side is suggested. Asking the patient to shake his head from side to side as if saying â noâ may elicit pronator drift if it does not occur with eye closure alone.

Power of the psoas (hip flexion) and gluteus (hip extension), hamstrings (leg flexion) and quadriceps (leg extension), tibialis anterior (dorsiflexion of the foot) and gastrocnemius (plantar flexion), and the extensors and inverters and everters of the foot should be assessed with the patient lying in the supine position.

The standard grading scale for muscle power is:

- 0/5â no movement of the muscle tested
- 1/5â trace movement of the muscle
- 2/5â insufficient power to overcome gravity
- 3/5â power sufficient to overcome gravity but not resistance from the examiner
- 4/5â power that can overcome gravity and some resistance from the examiner
- 5/5â full power

Pluses and minuses (e.g., 4+, 5-) are often added for the sake of nuance but are of little value unless an examiner is doing serial assessments for his own record and comparison.

Testing for a Gower sign, suggesting hip and thigh muscle weakness typical in cases of myopathy, should be included if there is a history of a delay in or regression of motor development. A Gower sign means that a patient can only transition from a sitting position on the floor to a standing position by using his arms to help push off the floor and straighten his torso. As a general rule, diseases of muscle produce proximal weakness (of the shoulders, hips, and thighs), whereas diseases of the nerve cause distal weakness (of the hands, legs, and feet).

Soft signs

A pediatric neurologist always keeps an eye out for â soft signsâ when he examines young children. The finding of a soft sign never identifies a specific disorder but does suggest general neurodevelopmental delay or immaturity. Children exhibiting soft signs often have a history of premature birth, delayed motor or language milestones, or neurobehavioral problems. Commonly noted soft signs include: (a) clumsy fine finger movements; (b) spread (e.g., the

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entire upper extremity moves during an attempted movement of only the hand); (c) mirror movement (unintentional movement of the opposite hand or arm when the child makes a hand or arm movement, such as waving goodbye); (d) overflow (an unrelated body part, such as the tongue, moves during a hand movement); and (e) clumsiness while hopping or running. Marked asymmetry of soft signs, although rare, should be noted because this finding is equivalent to a focal neurological sign and may suggest a structural abnormality in the brain.

Gait

The assessment of gait is a very important part of the neurological examination. A normal gait usually is evidence of adequate lower extremity strength and coordination, although subtle lower extremity weakness may only be evident when the patient rises from a chair, climbs stairs, or tries to jump or hop. Typical pathological gait patterns should be recognized. A circumducting gait often accompanies spastic hemiparesis or paraparesis (as in cases of cerebral palsy or prior stroke); a slap-foot gait is usually caused by neuropathy (e.g., Charcot-Marie-Tooth disease); a waddling gait is a sign of myopathy (e.g., muscular dystrophy); and an ataxic gait suggests a disease of the cerebellum.

A clumsy gait in a young child is often a sign of nonspecific and ultimately benign developmental motor delay; this diagnosis should be suspected if the child's gait has always been clumsy and there is no history of regression of motor skills. An antalgic gait refers to gait or posture assumed in order to lessen or avoid pain due to muscle, joint, bone, or other organ disease. In a toddler or young child, this may not clearly appear as a limp. The child will try movements that will minimize the weight applied to the affected limb or joint and the amount of time that weight is placed on the limb. This is often mistaken as a sign of weakness rather than of pain.

An odd-appearing gait that does not correspond to one of the previously mentioned patterns is often psychogenic (hysterical gait). Ataxia abasia (lunging from side to side), a psychogenic gait, is often mistaken for true ataxia.

The tightness of the heel cords should be checked if a toddler walks clumsily or frequently falls. The examiner should be able to dorsiflex the foot to about 10 degrees above the horizontal. Tight heel cords, preventing dorsiflexion of the foot, are commonly noted in cases of cerebral palsy (see Chapter 10) and can be the only abnormal finding in mild cases. Tight heel cords also can result from muscular dystrophy and other neuromuscular disorders (e.g., Charcot-Marie-Tooth disease) but are not an early sign of these diseases, and unlike in cerebral palsy, the deep tendon reflexes are often decreased rather than increased.

Children with tight heel cords often walk on their tiptoes. Toe walking not caused by tight heel cords is a common habit of children with autism, but it is also often a habit of normal children.

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Motor Assessment of the Infant and Toddler

The evaluation of the motor system in infants and very young children is, again, largely based on observation. The young infant should move all four extremities equally and, when older, should use either hand to reach for and manipulate objects. The gait of young children should be symmetrical, and

use of the hands should be symmetrical as well, although preferential use of the right hand often develops by about 3 years of age.

Figure 1.4 Vertical and horizontal suspension tests.

The history of infants should always include a developmental assessment of the age at which head control and ability to roll over, sit, and pull to a standing position were achieved. The toddler should learn to walk by 16 months of age and to run by 2â 3 years of age. However, it is important to recognize that there is considerable variability in the ages at which normal children achieve their motor milestones.

Both axial (truncal) and extremity muscle tone should be assessed in the neonate and infant. The examiner seeks evidence of either â floppiness,â suggesting hypotonia, or stiffness, suggesting hypertonicity. Truncal tone is evaluated by the pull-to-sit maneuver, in which the infant lying on his back is pulled gently by his hands so that his back and head no longer rest on the table. If this demonstrates head-lag at an age of more than 2 months, the infant is probably hypotonic. The hypotonic infant will often slip through the examiner's hands when held in the position of vertical suspension (Fig. 1.4; the infant is held in a vertical position, supported only by the examiner's fingers under the axillae) or â drapeâ when held in horizontal suspension (the infant is supported only by the examiner's hand under the abdomen). In cases of hypertonicity, the same maneuvers often produce scissoring of the legs and arching of the neck and back.

Passive movements of the limbs evaluate muscle tone in the extremities. Hypotonic limbs offer decreased resistance to movement; hypertonic limbs are stiff. Spasticity, which is noted in most cases of hypertonicity, refers to velocity-dependent increased muscle tone and a â clasp-knifeâ response when an extremity is moved at a joint (for example, during elbow flexion).

The examiner should be aware that extremity tone and truncal tone may differ markedly in the same patient (see Chapters 8 and 10).

Deep Tendon Reflexes

Concise assessment

A brisk or diminished deep tendon reflex is rarely the only sign of a neurological disease. Therefore, the physician should not fret too much about a single hard-to-elicit or brisk reflex when the patient does not describe and/or has no other sign of a motor, sensory, gait, or balance problem. The reflex examination also can be concise in these cases; the examiner may test one deep tendon reflex in each extremity (e.g., brachioradialis in the upper extremity and quadriceps in the lower extremity) and move to another phase of the examination.

More comprehensive assessment

In the case of a patient with a focal motor deficit or marked generalized weakness, the examination of deep tendon reflexes combined with the motor examination helps to localize the disorder to the brain; cervical,

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thoracic or lumbar spinal cord; nerve; or muscle. The biceps, triceps, brachioradialis, quadriceps, ankle, and plantar reflexes should be tested in these cases. A crossed adductor reflex (percussion of the medial thigh causing adduction of the opposite thigh) is sometimes elicited in cases of a lesion in the brainstem or cervical or thoracic spinal cord. Clonus is a self-sustaining deep tendon reflex, most often obtained at the ankle, suggesting spasticity or an upper motor neuron lesion (see next section).

The standard grading scale for deep tendon reflexes is:

- 0â no obtainable reflex.
- Tr.â trace reflex
- 1â low normal reflex
- 2â normal reflex
- 3â brisk reflex
- 4â clonus
- 5â sustained clonus (not always used)

Once again, pluses and minuses are often added (e.g., 1+) but are of limited usefulness.

A Babinski sign (upgoing toe and fanning of the other toes on one foot elicited by gently stroking the sole) suggests an upper motor neuron disease (see next section) but is a normal finding in a young infant.

Primitive reflexes (Moro, grasp, tonic neck, stepping, placing, and parachute reflexes) should be assessed in the infant. It is often easier to elicit a grasp reflex or an asymmetric or obligate tonic neck reflex than to elicit deep tendon reflexes in a crying inconsolable infant or toddler. Asymmetry of a primitive reflex, especially the Moro reflex, suggests focal weakness, as in the case of a hemispheric lesion or Erb palsy. The delayed appearance of a primitive reflex or the persistence of a primitive reflex as an infant grows older is associated with syndromes of developmental delay or regression.

Putting the Examination Together: Upper and Lower Motor Neuron Syndromes

The cell body of an upper motor neuron is a tiny part of the motor cortex of a cerebral hemisphere (Fig. 1.5). The axon of this cell travels caudally (downwards) through the cerebral white matter into the brainstem, crosses the midline in the medulla, and joins the corticospinal tract. The distal end of the axon synapses on a spinal anterior horn cell. The anterior horn cell, or lower motor neuron, sends its axon, as part of a motor nerve that does not cross the midline, to supply a muscle. Cervical anterior horn cells project

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to muscles in the upper extremity, and lumbar anterior horn cells project to muscles in the lower extremity.

Figure 1.5 Upper and lower motor neurons.
Diseases (e.g., stroke, cervical or thoracic tumor) affecting the upper motor neuron ultimately cause weakness, spasticity, and exaggerated deep tendon reflexes. Examples would include the paretic, spastic, and hyperreflexic left arm and leg of a patient who has a history of a stroke or tumor in the right cerebral hemisphere (Fig. 1.6) and the paretic, spastic, and hyperreflexic left leg of a patient who has a tumor compressing the left side of the thoracic spinal cord (Fig. 1.7). (Note that these signs are contralateral in cases of a lesion above the brainstem and ipsilateral in cases of a spinal cord lesion. This is because the upper motor neuron crosses the midline in the medulla.)
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Figure 1.6 Upper motor neuron lesion: right hemisphere tumor.
Patient has left hemiparesis (arm and leg) with spasticity and brisk deep tendon reflexes
The physician should be aware that an upper motor neuron syndrome usually takes several weeks to develop following an acute neurologic event; in the interim, there may be decreased tone and diminished reflexes in the affected limb. For example, a patient with a stroke may initially present with flaccid left hemiparesis, which evolves over the course of a month into spastic left hemiparesis.
In contrast, diseases of the lower motor neuron cause weakness, decreased muscle tone, and diminished reflexes. These signs are present immediately following an injury and persist indefinitely. The site of the lesion may be the anterior horn cell body (i.e., the spinal cord), the motor nerve root, the nerve, or the neuromuscular junction. Typical presentations would include the flaccid lower extremities of a child with a history of lumbar meningomyelocele (Fig. 1.8), the weakness and areflexia associated with Guillain-BarrÃ© syndrome (Fig. 1.9), and the flaccid arm of a baby born with a brachial plexus injury (Erb palsy; Fig. 1.10).
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Figure 1.7 Upper motor neuron lesion: tumor compressing left side of spinal cord.
Patient has weakness of left lower extremity with spasticity, brisk reflexes. Upper extremity unaffected.

Sensory Examination

In many cases, the sensory portion of the neurological examination can simply be omitted. Certainly, a time-consuming sensory examination is almost never a necessary part of the evaluation of a patient who has a nonfocal disorder such as tension-type headaches and ADHD. In such cases, the physician's time is better spent obtaining a more complete history, concentrating on more relevant aspects of the physical examination, and discussing the diagnosis with the family and answering their questions.

When and how to perform a sensory examination

When a child or adolescent patient reports persistent sensory loss or paresthesias (tingling or burning), these symptoms should be taken seriously because they are relatively unusual complaints at this age. Persistent sensory complaints often are caused by neuropathy; less often, they are the result

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of a mass lesion or demyelination in the brain or spinal cord. (Transient sensory symptoms are often related to migraine headaches and, less often, to focal seizures.) Other complaints that suggest the need for a sensory examination include unexplained focal or generalized weakness, loss of the ability to walk, incontinence, and pain that radiates to an upper or lower extremity.