原版影印

国外经典医学教材改编/影印系列

供医学各专业本科生、研究生、长学制学生、留学生用

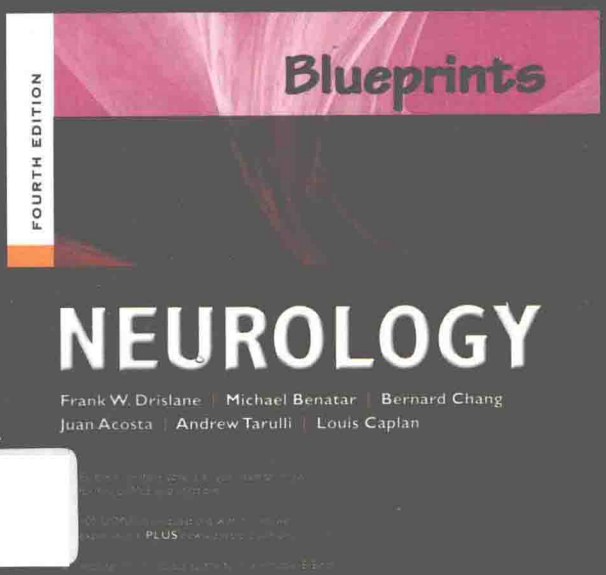
NEUROLOGY

(Fourth Edition)

神经病学

（第 4 版）

Frank W. Drislane
Michael Benatar
Bernard Chang
Juan Acosta
Andrew Tarulli
Louis Caplan



北京大学医学出版社

国外经典医学教材改编/影印系列

NEUROLOGY

(Fourth Edition)

神经病学

(第4版)

Frank W. Drislane, MD
Professor of Neurology
Harvard Medical School
Comprehensive Epilepsy Center
Beth Israel Deaconess Medical Center
Boston, Massachusetts

Michael Benatar, MBChB, MS, DPhil
Associate Professor of Neurology,
and Epidemiology & Public Health
Walter Bradley Chair in ALS Research
Chief, Neuromuscular Division
University of Miami
Miami, Florida

Bernard S. Chang, MD, MMSc
Associate Professor of Neurology
Harvard Medical School
Comprehensive Epilepsy Center
Beth Israel Deaconess Medical Center
Boston, Massachusetts

Juan Acosta, MD
Adjunct Assistant Professor of Neurology
Des Moines University
Neurologist, Clinical Neurophysiologist
McFarland Clinic
Ames, Iowa

Andrew Tarulli, MD
Assistant Professor of Neurology
Harvard Medical School
Division of Neuromuscular Disease
Beth Israel Deaconess Medical Center
Boston, Massachusetts

Louis R. Caplan, MD
Professor of Neurology
Harvard Medical School
Cerebrovascular Service
Beth Israel Deaconess Medical Center
Boston, Massachusetts

北京大学医学出版社
Peking University Medical Press

图书在版编目（CIP）数据

神经病学：第4版＝Neurology：英文/（美）德斯兰编.
—北京：北京大学医学出版社，2014.9
ISBN 978-7-5659-0886-6

Ⅰ.①神… Ⅱ.①德… Ⅲ.①神经病学-教材-英文
Ⅳ.①R741

中国版本图书馆CIP数据核字（2014）第144379号

北京市版权局著作权合同登记号：01-2014-5298
Frank W. Drislane, Michael Benatar, Bernard S. Chang, et al: Neurology. 4th ed
ISBN: 978-1-4511-1768-4

神经病学（第4版）

编　　写： Frank W. Drislane, Michael Benatar, Bernard S. Chang, et al
出版发行： 北京大学医学出版社
地　　址：（100191）北京市海淀区学院路38号　北京大学医学部院内
电　　话： 发行部 010-82802230；图书邮购 010-82802495
网　　址： http://www.pumpress.com.cn
E-mail： booksale@bjmu.edu.cn
印　　刷： 北京画中画印刷有限公司
经　　销： 新华书店
责任编辑： 冯智勇　　**责任印制：** 张京生
开　　本： 889mm×1194mm　1/16　　**印张：** 16.75　　**字数：** 456千字
版　　次： 2014年9月第1版　2014年9月第1次印刷
书　　号： ISBN 978-7-5659-0886-6
定　　价： 69.00元

Preface

Blueprints Neurology was first published more than 15 years ago as part of a series of books designed to help medical students prepare for USMLE Steps 2 and 3. Just as professional board evaluations have developed over time, and medical training continues to advance, so too has *Blueprints Neurology* evolved to assist practitioners and students across multiple evaluation settings.

Examination preparation remains the core component of the series; to that end, the authors review the subject matter of the examination before each edition. The authors and editors work together to organize the most important, current, and factually correct material into a complete but concise review guide. Our ultimate goal remains integrating depth of factual knowledge with breadth of practice information in order to optimize both understanding and retention. We have been pleased to hear from our readers that the book is utilized by many medical students during their clinical rotations, as well as in preparation for shelf and board examinations. Residents in Emergency Medicine and Family Practice, as well as nurse practitioners and physicians' assistants, have found Blueprints helpful during the Neurology portion of their training. We believe the book's applications have broadened with each edition due to the quality of our authors and their dedication to highlighting and clarifying a targeted range of basic yet important topics that must be mastered.

Each chapter in the book consists of a single subject for review. Most can be read in under an hour. The topics contained in each chapter are grouped in an orderly fashion, with "Key Points" highlighted throughout, facilitating instant review of the concepts most frequently tested.

This edition includes important updates on topics such as multiple sclerosis, stroke, Pediatric Neurology, genetics and new drugs (for example, to treat narcolepsy, cataplexy and other sleep disorders).

As you would expect, we have included the popular end-of-book review questions that so many of you have told us you count on, PLUS more Clinical Vignette questions. These questions are written in the "clinical vignette" style used on USMLE and Board examinations. Thus, readers not only can evaluate their grasp of the material but also begin to acclimate themselves to the expected testing environment.

We are proud to offer this fourth edition of *Blueprints Neurology*. It incorporates suggestions we have received from medical students, faculty, clinicians, and program directors with regard to content and organization. Virtually all of the chapters are coauthored by at least one expert in the respective content area. Utilizing authors with dual backgrounds in academic medicine and private practice for each chapter permits incorporation of the most recent information and practice parameters available and accepted at publication.

We hope you find *Blueprints Neurology* to be a beneficial investment, regardless of how you use it.

Frank W. Drislane, MD
Michael Benatar, MBChB, MS, DPhil
Bernard S. Chang, MD, MMSc
Juan Acosta, MD
Andrew Tarulli, MD
Louis R. Caplan, MD

Acknowledgments

We thank our patients for the opportunity of working with them and learning Neurology, our colleagues and teachers in the Beth Israel Deaconess Medical Center Neurology department for teaching us more fascinating concepts about the nervous system, and our families for tolerating the many hours spent writing and revising this book.

Abbreviations

A(ß)	amyloid-beta
ABP	abductor pollicis brevis
Abs	antibodies
AßPP	amyloid–beta protein precursor
ACA	anterior cerebral artery
ACE	angiotensin converting enzyme
AD	Alzheimer disease
ADEM	acute disseminated encephalomyelitis
ADHD	attention deficit–hyperactivity disorder
ADM	abductor digiti minimi
AED	antiepileptic drug
AICA	anteroinferior cerebellar artery
AIDP	acute inflammatory demyelinating polyradiculoneuropathy
AIDS	acquired immunodeficiency syndrome
AION	anterior ischemic optic neuropathy
ALS	amyotrophic lateral sclerosis
ANA	antinuclear antibody
APP	amyloid precursor protein
APS	antiphospholipid syndrome
AVM	arteriovenous malformation
AZT	zidovudine
BMD	Becker muscular dystrophy
BPPV	benign positional paroxysmal vertigo
CBC	complete blood count
cGMP	cyclic guanosine monophosphate
CIDP	chronic inflammatory demyelinating polyradiculoneuropathy
CJD	Creutzfeldt-Jakob disease
CK	creatine kinase
CMAP	compound muscle action potential
CMT	Charcot-Marie-Tooth disease
CN	cranial nerve
CNS	central nervous system
COMT	catechol O-methyl transferase
CP	cerebral palsy
CPAP	continuous positive airway pressure
CSF	cerebrospinal fluid
CT	computed tomography
DH	detrusor hyperreflexia
DI	detrusor instability
DLB	dementia with Lewy bodies
DM	dermatomyositis
DMD	Duchenne muscular dystrophy
DSD	detrusor-sphincter dyssynergia
DTRs	deep tendon reflexes
DWI	diffusion-weighted imaging
EA	episodic ataxia
ED	erectile dysfunction
EEG	electroencephalogram
EMG	electromyography
ER	emergency room
ESR	erythrocyte sedimentation rate
ET	essential tremor
EWN	Edinger-Westphal nuclei
FDI	first dorsal interosseus
FEV1	forced expiratory volume in 1 second
FLAIR	fluid-attenuated inversion recovery
FTA	fluorescent treponemal antibody
FTD	frontotemporal dementia
FVC	forced vital capacity
GAD	glutamic acid decarboxylase
GBS	Guillain-Barré syndrome
GCS	Glasgow Coma Scale
GTC	generalized tonic-clonic
HD	Huntington's disease
HIV	human immunodeficiency virus
HNPP	hereditary neuropathy with liability to pressure palsies
HS	Horner's syndrome
HSAN	hereditary sensory and autonomic neuropathy
HSV	herpes simplex virus
IBM	inclusion body myositis
ICA	internal cerebral artery
ICP	intracranial pressure
ICU	intensive care unit
IIH	idiopathic intracranial hypertension
INO	internuclear ophthalmoplegia
INR	international normalized ratio
IVIg	intravenous immunoglobulin
LEMS	Lambert-Eaton myasthenic syndrome
LGN	lateral geniculate nucleus
LMN	lower motor neuron
LND	light-near dissociation
LP	lumbar puncture
MAG	myelin-associated glycoprotein
MCA	middle cerebral artery
MELAS	mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke

MERRF	myoclonic epilepsy with ragged red fibers
MFS	Miller Fisher syndrome
MG	myasthenia gravis
MLF	medial longitudinal fasciculus
MMN	multifocal motor neuropathy
MND	motor neuron disease
MRA	magnetic resonance angiography
MRC	Medical Research Council
MRI	magnetic resonance imaging
MRV	magnetic resonance venography
MS	multiple sclerosis
MSA	multiple system atrophy
MSLT	multiple sleep latency test
MuSK	muscle-specific kinase
nAChR	nicotinic acetylcholine receptor
NCS	nerve conduction studies
NCV	nerve conduction velocity
NFTs	neurofibrillary tangles
NIF	negative inspiratory force
NMDA	N-methyl-D-aspartate
NMJ	neuromuscular junction
NMS	neuroleptic malignant syndrome
NSAIDs	nonsteroidal anti-inflammatory drug
OCD	obsessive-compulsive disorder
ON	optic neuritis
PANDAS	pediatric autoimmune neurologic disorders associated with streptococcal infection
PAS	periodic acid–Schiff
PCA	posterior cerebral arteries
PCD	paraneoplastic cerebellar degeneration
PCNSL	primary central nervous system lymphoma
PCR	polymerase chain reaction
PD	Parkinson's disease
PDC	paroxysmal (nonkinesigenic) dystonic choreoathetosis
PEO	progressive external ophthalmoplegia
PET	positron emission tomography
PICA	posteroinferior cerebellar artery
PKC	paroxysmal kinesigenic choreoathetosis
PM	polymyositis
PML	progressive multifocal leukoencephalopathy
PN	peripheral neuropathy
PNS	peripheral nervous system
POTS	postural orthostatic tachycardia syndrome
PP	periodic paralysis
PPD	purified protein derivative
PPRF	paramedian pontine reticular formation
PS1	presenilin 1
PS2	presenilin 2
PSP	progressive supranuclear palsy
PT	prothrombin time
PTT	partial thromboplastin time
PVR	post-void residual
QSART	quantitative sudomotor axon reflex test
RAPD	relative afferent pupillary defect
REM	rapid eye movement
RF	radiofrequency
riMLF	rostral interstitial nucleus of the MLF
RPR	rapid plasma reagin
rt-PA	recombinant tissue-type plasminogen activator
SAH	subarachnoid hemorrhage
SCA	spinocerebellar ataxia
SCA	superior cerebellar artery
SE	status epilepticus
SLE	systemic lupus erythematosus
SMA	spinal muscular atrophy
SNAP	sensory nerve action potential
SPECT	single-photon emission computed tomography
SSRI	selective serotonin reuptake inhibitor
STT	spinothalamic tract
TB	tuberculosis
TCD	transcranial Doppler
TE	time to echo
TIA	transient ischemic attack
TORCH	toxoplasmosis, other agents, rubella, cytomegalovirus, herpes simplex
TR	time to repetition
TSC	tuberous sclerosis complex
UMN	upper motor neuron
VA	visual acuity
VDRL	Venereal Disease Research Laboratory
VOR	vestibulo-ocular reflex
VP	venous pulsation
VPL	ventroposterolateral
WD	Wilson's disease

Contents

Chapter

1 The Neurologic Examination

To practicing neurologists, the neurologic exam reflects the uniqueness of the specialty. In a world of technology, it remains a purely clinical tool still unmatched in its ability to identify and localize abnormalities of the nervous system. To students, however, the exam can be both mystifying and bemusing, an endless series of maneuvers designed to elicit seemingly obscure and inexplicable findings.

When its principles and elements are presented simply, though, the exam is logical and elegant, reflecting the rational diagnostic process that characterizes not just Neurology but all of medicine.

PRINCIPLES

1. **The neurologic exam is not a standardized checklist.** Part of the intimidation of performing the exam is its sheer length; hours could be spent on examining the mental status alone. In reality, however, the exam is used in a focused and thoughtful way, depending on what hypotheses have been generated about the patient's illness from the history. A patient presenting with confusion may need quite a comprehensive mental status exam, whereas a patient presenting with a left foot drop may need detailed motor, sensory, and reflex testing of the left leg. In both cases, general screening elements of the remaining parts of the exam might be sufficient.
2. **Observation is more important than direct confrontation testing.** Most abnormalities of the nervous system manifest themselves in ways visible to the observant examiner. A significant anomia becomes evident when a patient uses circumlocutions to relate the history, and proximal weakness is obvious when a patient has difficulty rising from a chair. It is often more useful to describe a patient's observed activities and capabilities than to describe the findings obtained upon formal testing. Confrontation testing is subjective and variable; the grading of muscle strength depends in part on the examiner's effort and expectations of what the patient's "normal" strength should be. The observation of a pronator drift, for example, is less subjective.
3. **The object is to localize.** The extent and complexity of the nervous system require that any attempt to formulate a concise differential diagnosis must begin with an accurate localization of the problem to a specific region of the nervous system. Left hand weakness may stem from carpal tunnel syndrome, a brachial plexus injury, cervical spondylosis, or a right middle cerebral artery stroke, all of which have different diagnostic workups, treatments, and prognoses. The alert physician thinks, "What signs would be present in a carpal tunnel problem that would not be present in a brachial plexus problem (and vice versa)?" Those signs are then sought and the exam further refined if necessary.
4. **Not all findings have equal importance.** A common difficulty facing medical students is that completion of the exam results in a long list of many minor abnormalities of questionable importance,

such as a 20% decrease in temperature sensation over a patch on the left thigh. Though certainly in some cases incidental findings may be the clue to a previously unsuspected diagnosis, in most cases the highest importance must be given to findings directly related to the patient's symptoms and to "hard" findings that require definitive explanation, such as a dropped reflex or a Babinski sign.

KEY POINTS

- The neurologic exam is not a standardized checklist.
- Observation is more important than confrontation.
- The object is to localize.
- Not all findings have equal importance.

TABLE 1-1 Commonly Performed Elements of the Neurologic Examination

Mental status	
Attention	Serial backward tasks (months of the year, digit span)
Language	Fluency of speech, repetition, comprehension of commands, naming objects, reading, writing
Memory	Three words in 5 minutes
Visuospatial function	Drawing clock, copying complex figure
Neglect	Line bisection, double simultaneous stimulation
Frontal lobe function	Generating word lists, learning a motor sequence
Cranial nerves	
II	Visual acuity, fields, pupils, funduscopic exam
III, IV, VI	Extraocular movements
V, VII	Facial sensation and movement
IX, X, XII	Palate and tongue movement
Motor	
Bulk	Palpation for atrophy
Tone	Evaluation for rigidity, spasticity
Power	Observational tests (pronator drift, rising from chair, walking on heels and toes), direct confrontation strength testing
Reflexes	
Muscle stretch reflexes	Biceps, brachioradialis, triceps, knee, ankle
Babinski sign	Stroking lateral sole of foot
Sensory	
Pinprick and temperature	Pin, cold tuning fork
Vibration and joint position sense	Tuning fork and moving digits
Coordination	
Accuracy of targeting	Finger-to-nose, heel-to-shin
Rhythm of movements	Rapid alternating movements, rhythmic finger, or heel tapping
Gait	
Stance	Narrow or wide base
Romberg sign	Steadiness with feet together and eyes closed
Stride and arm swing	Assessment for shuffling, decreased arm swing
Ataxia	Ability to tandem walk

ELEMENTS OF THE EXAM

As discussed earlier, the specific features to include in the neurologic exam should vary with each patient; commonly performed elements of the exam are described in this section and listed in Table 1-1.

MENTAL STATUS

Neurologists use the mental status exam to identify cognitive deficits that help to localize a problem to a specific region of the brain. Thus the exam differs from that used by psychiatrists, whose objectives in performing the exam are different.

The first step in mental status testing is to assess the level of consciousness. This may vary from the alert wakefulness of a clinic outpatient to the coma of a patient in the intensive care unit. There is a tendency to use "medical" terminology—such as **stuporous**, **obtunded**, or **lethargic**—to describe the level of consciousness, but these have variable meanings; it is more useful to describe how well patients stayed awake or what stimulation was required to arouse them.

Next, assuming the level of consciousness allows for further testing of cognitive functions, attention is tested, typically with serial forward and backward tasks. These include digit span, reciting the months of the year, or spelling the word "**world**," forward and backward. Attention is usually tested early, because significant inattention compromises the ability to perform subsequent cognitive tests and may render their interpretation difficult.

Next, language is assessed. As noted previously, listening to patients tell their histories may be all that is necessary to gauge language ability. Formal testing, however, includes assessing the fluency of spontaneous speech, the ability to repeat, the ability to comprehend commands, the ability to name both common and less common objects, and the ability to read and write.

For memory testing, most often the patient is given three words and asked to recall them several minutes later, with the aid of hints if necessary. More information can be gained by giving longer lists of words and charting the patient's learning (and forgetting) curve. Visual memory can be tested with three simple shapes for the patient to draw from memory in several minutes.

Visuospatial function can be tested in a variety of ways. Patients can be asked to draw a clock, a cube, or another simple figure; alternatively, they can be asked to copy a complex figure drawn by the examiner (Fig. 1-1).

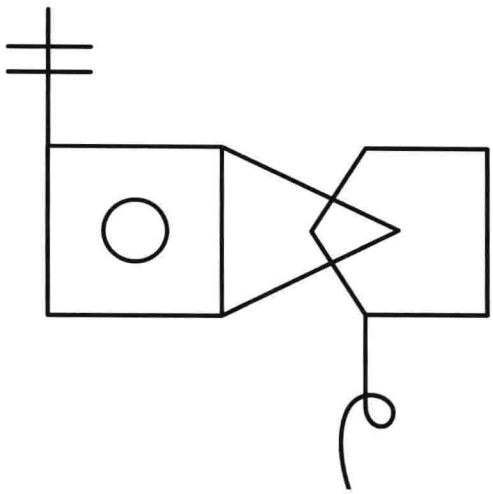


Figure 1-1 • Example of a complex figure to be copied by the patient as test of visuospatial function.

Neglect is a mental status finding typically not sought by nonneurologists, yet its presence can be a very important sign. Patients with dense neglect may fail to describe items on one side of a picture or of their surroundings, or may fail to bisect a line properly. Subtle neglect may manifest as extinction to double simultaneous stimulation, in which a patient can sense a single stimulus on either side but when bilateral stimuli are presented simultaneously will sense only the one on the nonneglected side.

Tests of frontal lobe function include learning a simple motor sequence of hand postures, inhibiting inappropriate responses when following a "go/no-go" paradigm, or generating lists of words beginning with a particular letter or belonging to a particular category.

KEY POINTS

- The mental status exam should begin with assessment of level of consciousness and attention, because these can affect the interpretation of subsequent tests.
- Language, memory, visuospatial function, neglect, and tests of frontal lobe function are other key elements of the mental status exam that can suggest focal brain lesions.

CRANIAL NERVES

It is usually easiest to test the cranial nerves (or at least to record the results) in approximate numerical order (Table 1-2).

TABLE 1-2 The Cranial Nerves

Nerve	Name	Exit through Skull	Function
I	Olfactory	Cribriform plate	Olfaction (test using nonnoxious substance)
II	Optic	Optic canal	Vision (acuity, fields, color), afferent limb of pupillary reflex
III	Oculomotor	Superior orbital fissure	Superior rectus, inferior rectus, medial rectus, inferior oblique, levator palpebrae, efferent limb of pupillary reflex
IV	Trochlear	Superior orbital fissure	Superior oblique
V	Trigeminal	Superior orbital fissure (V1), foramen rotundum (V2), foramen ovale (V3)	Muscles of mastication, tensor tympani, tensor veli palatini, facial sensation, afferent limb of corneal reflex
VI	Abducens	Superior orbital fissure	Lateral rectus
VII	Facial	Internal auditory meatus	Muscles of facial expression, stapedius, taste on anterior two-thirds of tongue, efferent limb of corneal reflex
VIII	Vestibulocochlear	Internal auditory meatus	Hearing, vestibular function
IX	Glossopharyngeal	Jugular foramen	Movement of palate, sensation over palate and pharynx, taste over posterior one-third of tongue, afferent limb of gag reflex
X	Vagus	Jugular foramen	Movement of palate, sensation over pharynx, larynx, and epiglottis, efferent limb of gag reflex, parasympathetic function of viscera
XI	Accessory	Jugular foramen	Sternocleidomastoid and trapezius movement
XII	Hypoglossal	Hypoglossal foramen	Tongue movement

Olfaction (cranial nerve I) is rarely tested, but when this is important, each nostril should be tested separately with a nonnoxious stimulus, such as coffee or vanilla.

Tests of optic nerve (II) function include visual acuity (using a near card), visual fields (tested by confrontation with wiggling fingers or with a small red object, which is more sensitive), and the pupillary light reflex, the afferent limb of which is mediated by this nerve. Funduscopic examination is the only means by which a part of the central nervous system (the retina) can be directly visualized.

Extraocular movements (III, IV, and VI) are usually tested in three ways: by having the patient pursue a moving target, that is, a drawing of the letter "H" in front of the face (pursuit), by directing the patient's gaze rapidly to various stationary targets (saccades), and by fixating on an object while the head is being turned passively (vestibulo-ocular movements). The presence of nystagmus should be noted.

Muscles of mastication (V) are tested by assessing strength of jaw opening and palpating over the masseters bilaterally while the jaw is clenched. Facial sensation can be tested to all modalities over the forehead (V1), cheek (V2), and jaw (V3). The afferent limb of the corneal reflex is mediated by this nerve.

Muscles of facial expression (VII) are tested by having patients raise the eyebrows, squeeze the eyes shut, or show the teeth. Though uncommonly tested, taste over the anterior two-thirds of the tongue is mediated by this nerve and can be evaluated with sugar or another nonnoxious stimulus.

Hearing (VIII) may be evaluated in each ear simply by whispering or rubbing fingers; more detailed assessment of hearing loss may be accomplished with the Weber and Rinne tuning fork (512 Hz) tests. Vestibular function can be tested in many ways, including evaluation of eye fixation while the patient's head is rapidly turned or by observation for drift in one direction while the patient is walking in place with the eyes closed.

Palatal elevation should be symmetric and the voice should not be hoarse or nasal (IX and X). Failure of the right palate to elevate implies pathology of the right glossopharyngeal nerve. The gag reflex is also mediated by these nerves.

Sternocleidomastoid strength is tested by having the patient turn the head against resistance; weakness

on turning to the left implies a right accessory nerve (XI) problem. The trapezius muscle is tested by having patients shrug their shoulders.

Tongue protrusion should be in the midline. If the tongue deviates toward the right, the problem lies with the right hypoglossal nerve (XII).

KEY POINTS

- Cranial nerve testing is most easily performed and recorded in approximate numerical order.
- Key elements of the cranial nerve exam include assessment of vision and eye movements, facial movement and sensation, and movements of the palate and tongue.

MOTOR EXAM

The motor exam includes more than just strength testing—in fact, power should usually be the portion of the exam performed last.

First, bulk is assessed by observing and palpating the muscles and comparing each side to the other and the patient's overall muscle bulk to that expected for age. The presence of fasciculations or of adventitious movements such as tremor or myoclonus should also be noted.

Tone is one of the most important parts of the motor exam. In the upper limbs, tone is checked by moving the patient's arm at the elbow in both flexion-extension and circular movements, by moving the wrist in a circular fashion, and by rapidly pronating and supinating the forearm using a handshake grip. Abnormalities of tone such as spasticity and

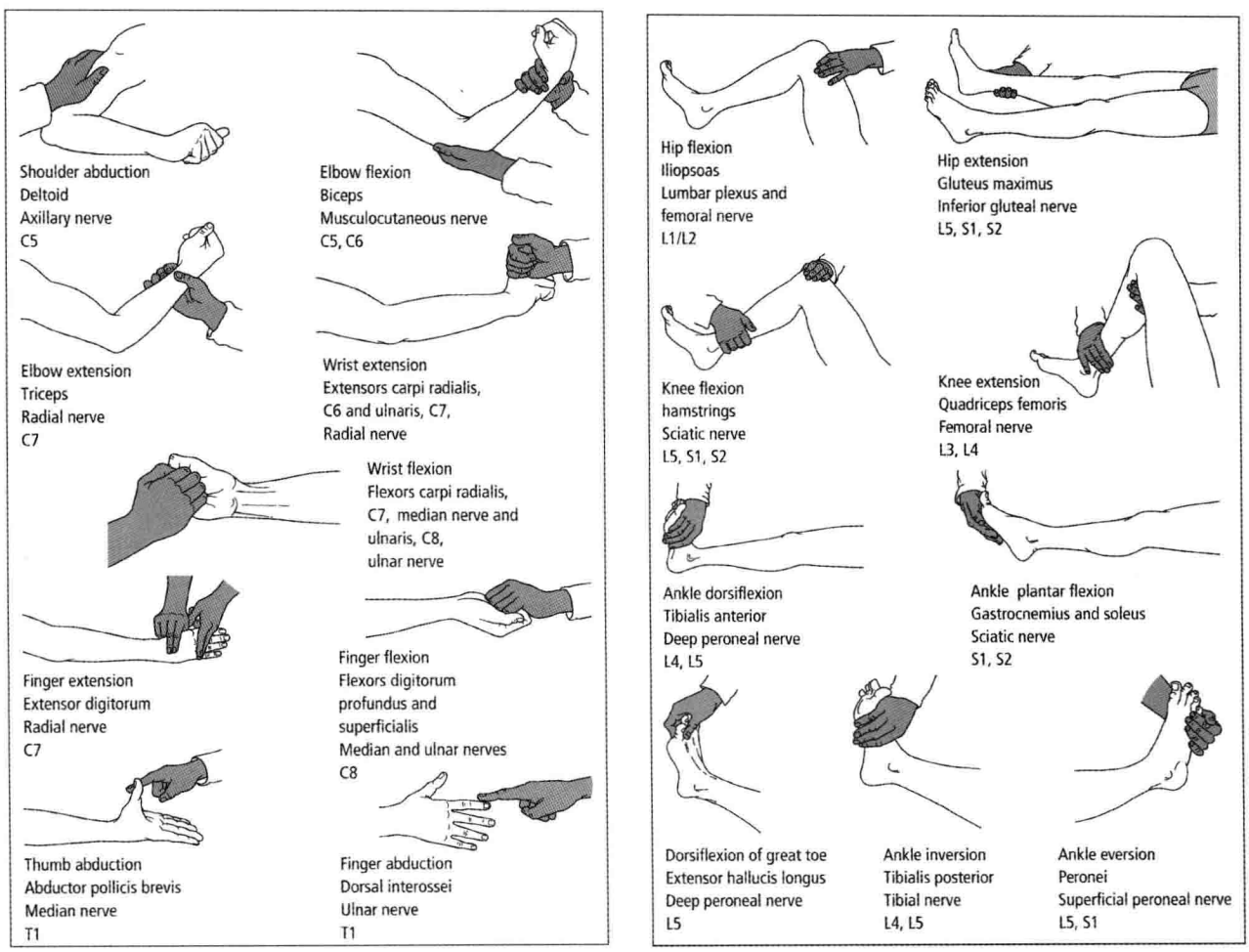


Figure 1-2 • Power testing of individual movements. For each movement, the predominant muscle, peripheral nerve, and nerve root are given.
(Reproduced with permission from Ginsberg L. *Lecture Notes: Neurology*. 8th ed. Oxford: Blackwell Publishing; 2005:40–41.)

TABLE 1-3 Medical Research Council Grading of Muscle Power

0	No contraction of muscle visible
1	Flicker or trace of contraction visible
2	Active movement at joint, with gravity eliminated
3	Active movement against gravity
4	Active movement against gravity and some resistance
5	Normal power

rigidity are discussed in subsequent chapters. Tone in the lower limbs can be tested well only with the patient supine. The examiner lifts the leg up suddenly under the knee; in the presence of increased tone, the heel comes off the bed.

Finally, strength or power is assessed, by both functional observation and direct confrontation (Fig. 1-2). A pronator drift may be observed in an arm held supinated and extended in front of the body. The patient may be asked to rise from a chair without using the arms or to walk on the heels or toes. The power of individual muscles assessed by direct confrontation testing is graded according to the Medical Research Council (MRC) scale (Table 1-3), although refinements of the scale (such as the use of 4−, 4, and 4+) or the use of a 10-point scale will increase precision.

KEY POINTS

- The motor exam begins with assessment of bulk and tone.
- Abnormalities of increased tone include both spasticity and rigidity.
- Strength testing involves both functional observation as well as confrontation testing of individual muscle power.
- Strength is graded on the MRC scale from 0 to 5.

REFLEXES

Muscle stretch (or "deep tendon") reflexes can be useful aids in localizing or diagnosing both central and peripheral nervous system problems (Fig. 1-3).

In the arms, the biceps, brachioradialis, and triceps reflexes are the ones commonly tested. Pectoral and

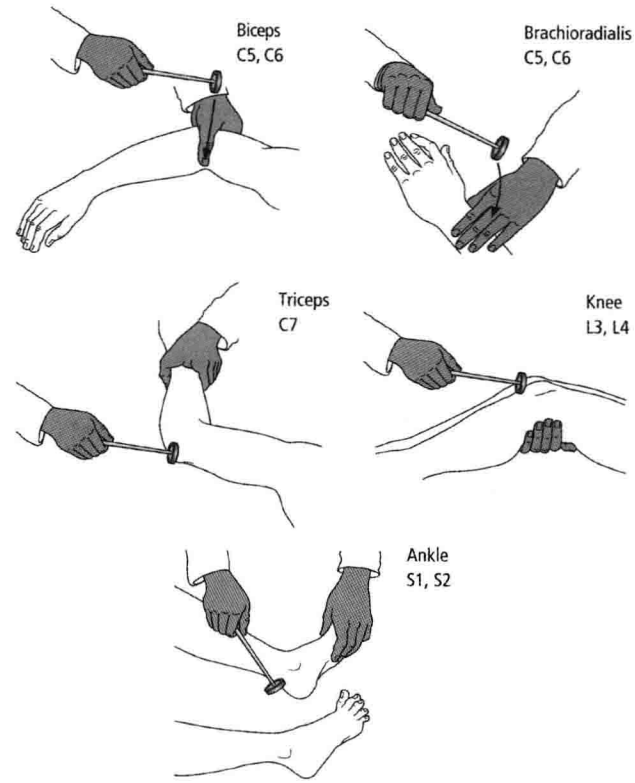


Figure 1-3 • Muscle stretch ("deep tendon") reflexes. (Reproduced with permission from Ginsberg L. *Lecture Notes: Neurology*. 8th ed. Oxford: Blackwell Publishing; 2005:44.)

finger flexor reflexes can also be tested. Hoffmann's sign is sought by flicking the distal phalanx of the middle finger while observing for flexion of the thumb.

In the legs, patellar (knee jerk) and ankle reflexes are the ones commonly tested. The adductor reflex can also be tested. The Babinski sign is sought by stroking the lateral sole of the foot while observing for extension of the great toe. Clonus, if present, can be elicited by forcibly dorsiflexing the ankle when it is relaxed.

SENSORY EXAM

The sensory exam can be frustrating to perform because of the tedium of potentially examining the entire body surface (see dermatome map in Chapter 6) as well as the inherent subjectivity and all-too-frequent inconsistencies in patients' responses.

In general, sensation should be tested in detail in areas relevant to a patient's complaints, especially if the complaints are sensory in nature. Otherwise, screening elements of the sensory exam that are

targeted at the distal lower extremities, where most asymptomatic sensory abnormalities are likely to be found, may be sufficient.

Pinprick sensation is tested with a safety pin, the sharp edge of a broken-off cotton swab, or special pins designed for the neurologic exam.

Temperature sensation, mediated by the same pathway, is most easily tested with the side of a tuning fork, which, if freshly retrieved from an instrument bag, will be quite cold on the skin.

Vibration is tested by striking the 128-Hz tuning fork and placing its stem against the joint being tested, typically beginning at the toes.

Joint position sense, or proprioception, is tested beginning most distally by holding the patient's great toe by its sides and moving it slightly upward or downward.

Light touch is often not useful to test in isolation, because it is carried by a combination of pathways and by itself is unlikely to provide clues to localization or diagnosis.

KEY POINTS

- The sensory exam is usually the most subjective portion of the neurologic exam.
- In a patient without sensory complaints, screening elements of the sensory exam that are targeted at the distal extremities may be sufficient.
- Pinprick and temperature are carried in one pathway; vibration and joint position sense in another.

COORDINATION

This portion of the exam, often imprecisely referred to as "cerebellar" testing, in fact serves to test coordinated movements whose successful completion requires the interaction of multiple components of the motor system, not just the cerebellum.

Finger-to-nose testing can identify the presence of dysmetria (inaccuracy of targeting) or intention tremor.

Heel-to-shin testing can elicit incoordination in the legs.

Rapid alternating movements, rhythmic finger tapping, and heel tapping are particularly sensitive to coordination problems. Patients may have trouble with the timing or cadence of these movements. *Dysdiadochokinesis* is the term used to describe difficulty with rapid alternating movements.

GAIT

Aside from orthopedic surgeons, neurologists are among the only doctors to routinely test a patient's gait, yet the "normal" function of walking requires the proper functioning of so many different aspects of the nervous system that it is frequently a sensitive way to detect an abnormality. In addition, certain diseases, such as Parkinson disease, have quite distinctive gaits associated with them.

The patient with a normal stance maintains the feet at an appropriately narrow distance apart; a wide-based stance is abnormal.

A Romberg sign is present when the patient maintains a steady stance with feet together and eyes open but sways and falls with feet together and eyes closed. Its presence usually implies a deficit of joint position sense, not cerebellar function.

Stride length should be full. Short-stepped or shuffling gaits are characterized by a decrease in stride length and clearance off the ground.

Ataxia of gait results in an inability to walk in a straight line; patients may stagger from one side to the other or consistently list toward one side. Ataxia is typically associated with a wide-based stance. Ataxia can be brought out most obviously by having the patient attempt tandem gait, walking heel to toe.

The arms normally swing in the opposite direction from their respective legs during ambulation. Decreased arm swing is often a feature of extrapyramidal disorders.

Finally, difficulty initiating ambulation or understanding the appropriate motor program for walking, leaving the feet "stuck to the floor" despite intact motor and sensory function, characterizes the gait of frontal lobe dysfunction, sometimes referred to as *gait apraxia*. Hydrocephalus is one etiology of such a gait disorder.

KEY POINTS

- Gait is one of the most important elements of the neurologic exam because it is sensitive for many deficits, and certain diseases have characteristic gait disorders.
- Stance, stride length, arm swing, ability to tandem walk, and initiation of walking should all be assessed in the gait exam.
- The Romberg sign suggests a deficit in joint position sense.

Chapter

2 Neurologic Investigations

CEREBROSPINAL FLUID ANALYSIS

Cerebrospinal fluid (CSF) bathes the internal and external surface of the brain and spinal cord. It is produced by the choroid plexus of the ventricles and absorbed through the villi of the arachnoid granulations that project into the dural venous sinuses. CSF is produced continually at a rate of about 0.5 mL/minute; the total volume is approximately 150 mL. The entire CSF volume is thus replaced about every 5 hours. Lumbar puncture (LP) via the L3-4 or L4-5 interspace is the most commonly used means of obtaining CSF for analysis. LP is contraindicated by the presence of a space-occupying lesion that is causing mass effect with raised intracranial pressure, local infection or inflammation at the planned puncture site, or a significant and as yet uncorrected coagulopathy.

TECHNIQUE

Optimal positioning is the key to a successful and atraumatic LP. LP is best performed with the patient in the lateral recumbent position with the legs flexed up over the abdomen. Ideally, a pillow should be placed between the legs, and the patient should lie on the edge of the bed where there is better support to keep the back straight. The anterosuperior iliac spine is at the level of the L3-4 vertebral interspace. The LP may be performed at this level, one interspace higher, or one to two interspaces lower. (Remember that the spinal cord ends at the level of L1-2.) The needle is inserted with the bevel facing upward (when the patient is in the lateral decubitus position), so that it will enter parallel to the ligaments and dura that it pierces rather than cutting them transversely. The needle is directed slightly rostrally to coincide with the downward angulation of the spinous processes. The needle is advanced gently until CSF is obtained. To measure the opening pressure reliably, the patient's legs should be extended slightly and note should be made of fluctuation of the CSF meniscus within the manometer with respiration.

INTERPRETATION OF RESULTS

CSF is a clear, colorless fluid. The glucose content is about two-thirds that of blood, and it contains up to 40 to 50 mg/dL protein. Fewer than 5 white cells per cubic mm are present, and these are lymphocytes. The opening pressure measured by LP in the lateral recumbent position is about 60 to 150 mm H_2O.

Xanthochromia refers to the yellow discoloration of the supernatant of a spun CSF sample. Its presence helps to distinguish an *in vivo* intrathecal hemorrhage from a traumatic tap (in which red blood cells (RBCs) have not lysed, so the supernatant is still colorless).

The implications of various CSF findings are summarized in Table 2-1. CSF findings in a variety of common conditions are summarized in Table 2-2. Special tests may be performed as indicated. Some examples include cytology for suspected malignancy, oligoclonal banding for suspected immune-mediated processes such as multiple sclerosis, 14-3-3 protein for Creutzfeldt–Jakob disease, and a variety of polymerase chain reaction and serologic tests for various infections.

SAFETY, TOLERABILITY, AND COMPLICATIONS

Cerebral or cerebellar herniation may occur when LP is performed in the presence of either a supratentorial or infratentorial mass lesion. A computed tomography