ELSEVIER MOSBY

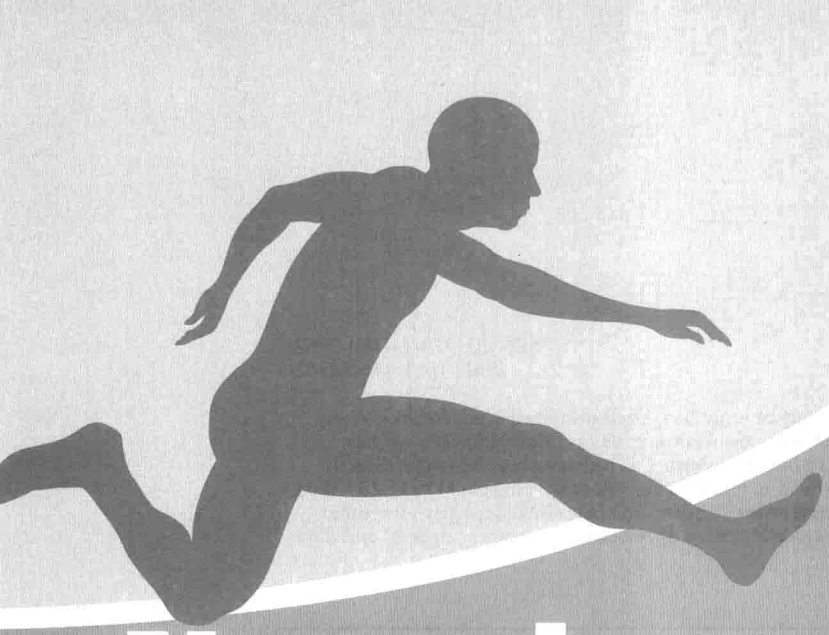
CRASH COURSE

Neurology

Joyce Liporace
Anish Bahra
Katia Cikurel

- For students taking the USMLE
- New review series to speed you through your clinical years
- Concise text and clear illustrations help you to absorb information fast
- Online USMLE questions

online access + interactive extras
studentconsult.com



Neurology

Joyce Liporace, MD

Director, Women's Health in Epilepsy Program,
Center for Neuroscience,
Riddle Health Care Center,
Media, Pennsylvania

UK edition authors

Anish Bahra and Katia Cikurel

ELSEVIER
MOSBY

ELSEVIER
MOSBY

1600 John F. Kennedy Blvd.
Suite 1800
Philadelphia, PA 19103-2899

CRASH COURSE: NEUROLOGY
ISBN-13: 978-1-4160-2962-5
ISBN-10: 1-4160-2962-1

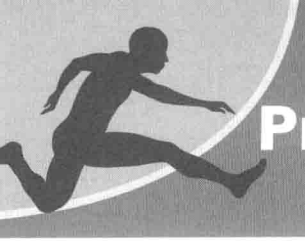
Library of Congress Control Number: 2005052281

Acquisitions Editor: Alex Stibbe
Project Development Manager: Stan Ward
Publishing Services Manager: David Saltzberg
Designer: Andy Chapman
Cover Design: Antbits Illustration
Illustration Manager: Mick Ruddy

Printed in China

Last digit is the print number:
9 8 7 6 5 4 3 2 1

CRASH COURSE
Neurology

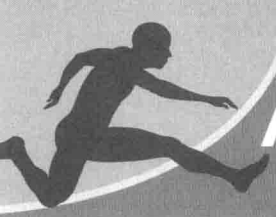


Preface

Neurology is often viewed as extremely complicated and difficult to grasp. Graduating medical students identify the study of neurology as overwhelming. *Crash Course: Neurology* is meant to provide a concise but comprehensive study guide for medical students and residents. Part I of the book presents an analysis and differential diagnosis of common disease symptoms such as dizziness, headache, or weakness. Part II provides a practical guide to taking a history and completing an examination. Part III discusses the presentation and management of the most common neurological diseases affecting the central and peripheral nervous systems.

Medical students should find *Crash Course: Neurology* a handy single source guide during their neurology rotations and also when studying for board examinations. Residents should find that it contains relevant information when caring for patients. I hope that it serves them well.

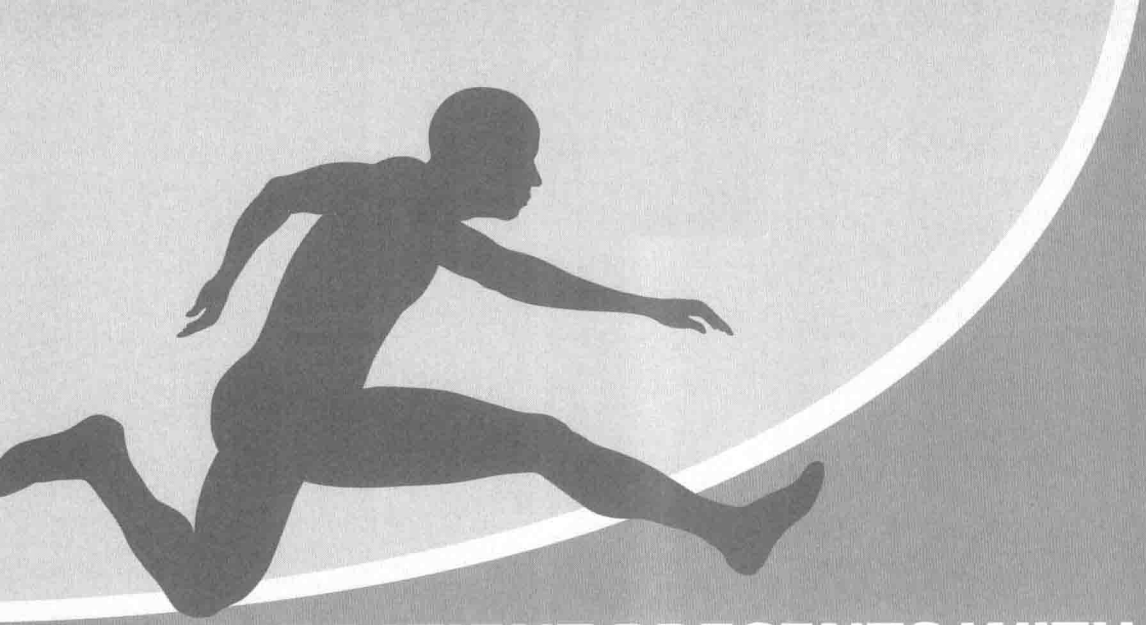
Joyce Liporace, M.D.



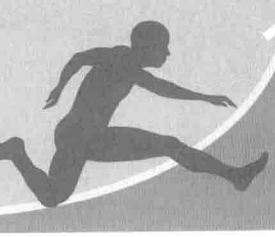
Acknowledgments

I am grateful to the original editors of the UK edition, Anish Bahra and Katia Cikurel. Special thanks to my children, Mike and Matt, and my husband, Tim, for their encouragement and support, and to the Jefferson residents for their inspiration and quest to learn everything about the nervous system.

Contents



THE PATIENT PRESENTS WITH



1. Disorders of Higher Cerebral Function

Impairment of cognitive (intellectual) function is often a prominent manifestation of cerebral hemisphere disease. The common diffuse or multifocal pathologies in the brain—Alzheimer's or generalized vascular disease, respectively—cause the syndrome of dementia, in which several aspects of cognitive function are impaired. Single localized lesions will predictably cause more focal cognitive deficits, such as aphasia or agnosia. For this reason it is important to be aware of the function of the different lobes and anatomical areas within the cerebral hemispheres. The four lobes—frontal, parietal, temporal, and occipital—are shown in Fig. 1.1. The cerebral cortices have specialized functions. Certain functions are attributes of either the right or left hemisphere; one hemisphere is therefore termed "dominant" and the other "nondominant." The left hemisphere is dominant in over 90% of right-handed people and in about 70% of left-handed people.

Cortical function and clinical manifestations of dysfunction of each lobe are considered below.

The frontal lobe

Function

- The motor cortex. The primary motor cortex is concerned with motor function of the opposite side of the body; the corticospinal and corticobulbar fibers are topographically represented in Fig. 1.2 (termed the homunculus).
- The supplementary motor cortex. This area is concerned with turning of the eyes and head contralaterally.
- Broca's area (dominant hemisphere). Broca's area is the motor center for the production of speech. This is a function of the dominant hemisphere.
- The prefrontal cortex. Personality, emotional expression, initiative, and the ability to plan are governed by the anterior part of the frontal cortex.
- The cortical micturition center. There is normally a cortical inhibition of voiding of the bladder and bowel.

The blood supply to the frontal lobe is from the anterior and middle cerebral arteries, branches of the internal carotid artery.

Lesions of the frontal lobe

Lesions of the frontal lobe give rise to:

- Contralateral mono- or hemiparesis and facial weakness of upper motor neuron type. The pattern of weakness depends on the area of cortex damaged.
- Paralysis of contralateral eye and head turning.
- Broca's expressive aphasia. This consists of nonfluent, hesitant speech with intact comprehension. The patient knows what he or she wants to say but has difficulty finding the correct words, often producing the wrong word. The ability to repeat words is better than spontaneous speech.
- Behavioral change. Features of altered behavior include social disinhibition, loss of initiative and interest, inability to solve problems and loss of abstract thought, and impaired concentration and attention without intellectual or memory decline. This is more common in bilateral lesions. Severe bilateral pathology may result in akinetic mutism, in which the patient is not paralyzed and has the ability to speak but lies still and silent.
- Elicitation of primitive (grasping and sucking) reflexes. These reflexes originate from the parietal cortex and are usually inhibited by the prefrontal cortex.
- Apraxia of gait. This is the inability to walk normally despite preservation of motor and sensory function.
- Incontinence of urine and/or feces. This results from loss of cortical inhibition. There is no desire to micturate. Milder symptoms are frequency and urgency of micturition.

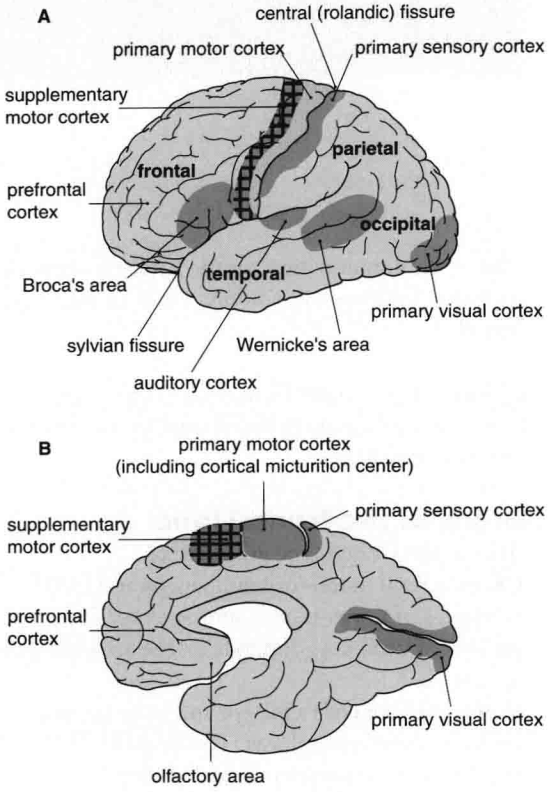


Fig. 1.1 Functional regions of the cerebral cortex. (A) Lateral left hemisphere. (B) Medial right hemisphere.

Focal seizures arising from the frontal cortex give rise to clonic movements of the contralateral lower face, arm, and leg, and congugate deviation of the head and eyes toward the convulsing side (i.e., away from the side of the lesion).

By virtue of their proximity, lesions of the frontal lobe may be accompanied by disturbance of the olfactory and visual pathways.

The parietal lobe

Function

- The sensory cortex. The parietal cortex receives afferent projections via the thalamus from the somatosensory pathways. The fibers are represented topographically, like the motor pathways.
- Language (dominant hemisphere). Pathways within the arcuate fasciculus connecting Broca's area (frontal) with Wernicke's area (posterior temporal) pass through the inferior parietal region.
- Use of numbers, as for calculation (dominant hemisphere).
- Integration of somatosensory, visual, and auditory information (mainly nondominant). This allows awareness of the body and its surroundings, appropriate movement of the body, and constructional ability.
- Visual pathways. The upper part of the optic radiation (subserving the lower quadrant of the contralateral visual field) passes deep within the parietal lobe and may be affected in lesions of the deeper white matter.

The blood supply to the parietal lobe is from the middle cerebral artery.

Lesions of the parietal lobe

Lesions of the parietal lobe give rise to:

- Discriminative sensory impairment of the opposite side of the face and limbs. There is impairment of position sense and two-point discrimination, and inability to recognize objects by form and texture (astereognosis) or figures drawn on the hand (agraphesthesia). Pain, temperature, touch, and vibration are intact; however, their localization when applied to the body may be impaired.

Syndromes of the dominant hemisphere

Syndromes of the dominant hemisphere include:

- Gerstmann's syndrome. This consists of confusion of the right and left sides of the body, inability to distinguish the fingers of the hands (finger agnosia), and impairment of calculation (dyscalculia) and writing (dysgraphia). Difficulty with reading (dyslexia) may also occur; this is a function of the dominant parieto-occipital cortex. This lesion occurs with damage to the angular gyrus that spares Wernicke's area.
- Bilateral ideomotor and ideational apraxia. This is the inability to carry out a task on request or by imitation, with normal comprehension and without disturbance of motor or sensory function.

Syndromes of the nondominant hemisphere

Syndromes of the nondominant hemisphere include:

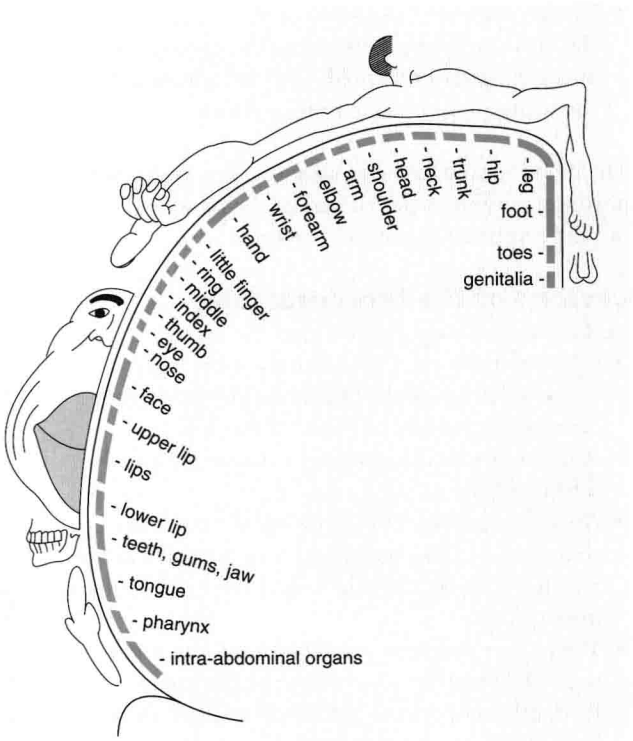


Fig. 1.2 Topographical distribution of the sensorimotor pathways (homunculus).

- Constructional apraxia (visuospatial dysfunction). There is difficulty in drawing simple objects (e.g., a house) and with construction (e.g., using building blocks).
- Dressing apraxia. There is difficulty with putting on clothes.
- Contralateral sensory inattention. There is neglect of the opposite side of the body; this may be motor, sensory, or visual (e.g., a hemiplegic patient may ignore the paralyzed side or there may be denial of the hemiplegia). Sensory and visual neglect are discussed in Chapter 14.
- Visual disturbances. If the deeper fibers of the parietal lobe are involved, a contralateral homonymous inferior quadrantanopia (one-fourth of the visual field) and ipsilateral loss of optikokinetic nystagmus may arise. Smooth pursuit eye movements may become "broken."

Focal seizures of the parietal cortex manifest as sensory symptoms of the contralateral side of the body. Descriptions of various sensations may be given (e.g., "pins and needles," tingling), and the symptoms often "march" to adjacent areas of the body.

The temporal lobe

Function

- Wernicke's area (dominant hemisphere). This area is concerned with comprehension of written and spoken language.
- The auditory and vestibular cortex. The primary auditory cortex receives fibers arranged in order of frequency of tone. The auditory pathways from each ear project to both auditory cortices. The dominant temporal lobe is important for the comprehension of spoken words, and the nondominant for the appreciation of sounds and music. Vestibular fibers terminate just posterior to the auditory cortex.
- The limbic lobe. The olfactory and gustatory cortices lie in the medial temporal lobe. The limbic system is important in memory, learning, and emotion.

- Visual pathways. The fibers of the lower part of the optic radiation (subserving the upper quadrant of the contralateral visual field) pass deep through the white matter of the temporal lobe.

The blood supply to the temporal lobe is from the posterior cerebral (medial part of the lobe) and middle cerebral (lateral part) arteries.

Lesions of the temporal lobe

Lesions of the temporal lobe give rise to:

- Cortical deafness. This will only occur with bilateral lesions of the primary auditory cortices. The patient may be unaware of the deficit. Auditory hallucinations may occur in temporal lobe epilepsy.
- Auditory agnosia. This is the inability to recognize sounds (e.g., ringing of a bell, whistling of a kettle, a melody). It occurs in lesions of the nondominant hemisphere.
- Wernicke's receptive aphasia (temporoparietal region). Wernicke's receptive aphasia arises from inferior parietal and superior temporal lesions (Wernicke's area and above). There is impaired comprehension of speech and written language without difficulty with expression. The speech is fluent but words are replaced with partly correct words (word salad), incorrect words related to the words intended (paraphrasia), or newly created meaningless words (neologisms). Thus the speech does not make sense, but the patient has poor insight into the problem. Repetition is poor.

Note that with auditory agnosia and Wernicke's receptive aphasia the function of hearing is normal.

- Vestibular dysfunction. Vestibular dysfunction from a lesion of the vestibular cortex is uncommon, but vertigo may occur as part of the aura of temporal lobe seizures.
- Olfactory and gustatory hallucinations. Olfactory hallucinations and, less commonly, gustatory hallucinations may arise from lesions within the medial temporal lobe, particularly during seizures.
- Learning difficulties. Learning difficulties with auditory information occur in dominant hemisphere lesions; learning difficulties with visual information occur in nondominant hemisphere lesions.
- Memory impairment. This occurs with lesions of the medial temporal lobe involving the hippocampus and parahippocampal gyrus. Bilateral damage results in marked impairment of retention of new information.
- Emotional disturbances. Emotional disturbances from damage to the limbic system may include aggression and rage, apathy, and hypersexuality.
- Visual disturbances. A lesion involving the deeper fibers within the temporal lobe will cause a contralateral superior homonymous quadrantanopia. Complex (or formed) visual hallucinations can occur in temporal lobe seizures.

Temporal lobe seizures may begin with a prodrome of auditory, olfactory, gustatory, or visual hallucinations, a sensation of anxiety or fear, and often a rising epigastric sensation. There may be disturbances of memory, with feelings of familiarity (déjà vu) or unfamiliarity (jamais vu). Behavioral changes may occur; aggression and hypersexuality are reported but are uncommon.

Aphasia

Aphasia is a disorder of spoken and written language; it occurs with damage of the frontal, parietal, or temporal cortices. Broca's expressive and Wernicke's receptive aphasias occur with damage of the dominant hemisphere and have been discussed (see p. 3 and p. 6). The cortical areas subserving these functions are linked by the arcuate fasciculus, which runs in the subcortical white matter. This enables the comprehension of language with subsequent production of speech in response.

- Conduction aphasia occurs with damage to the arcuate fasciculus. The speech is fluent but "jargon" with paraphrasia and neologisms as in Wernicke's aphasia. However, comprehension of language is intact, the patient is aware of the problem, and repetition is markedly impaired.
- Global aphasia occurs with lesions of both Broca's and Wernicke's areas. There is a combination of nonfluent speech and impaired comprehension of language.
- Anomia is an inability to name objects and arises from a lesion of the dominant temporoparietal cortex. It may occur during recovery from the aforementioned aphasias.

The occipital lobe

Function

The function of the occipital cortex (Fig. 1.3) is the perception of vision and recognition of whatever is visualized. The blood supply is from the posterior cerebral artery, but the occipital poles have additional supply from a branch of the middle cerebral artery.

Lesions of the occipital lobe

Lesions of the occipital lobe give rise to:

- Contralateral homonymous hemianopic field defect. If this arises from a lesion of the posterior cerebral artery, there will be sparing of the macular area. A lesion of the occipital pole will affect the macular fibers only and result in a contralateral homonymous hemianopic macular field defect.
- Cortical blindness. Bilateral occipital lesions render the patient blind, with retention of the pupillary reflexes. The patient may deny the blindness (Anton's syndrome).
- Visual agnosia. Lesions of the visual association cortices cause impairment of recognition of faces and objects.
- Visual illusions. Objects may appear larger (macropsia) or smaller (micropsia); there may be disturbances of shape, color, and number. This is more common with lesions of the nondominant hemisphere.

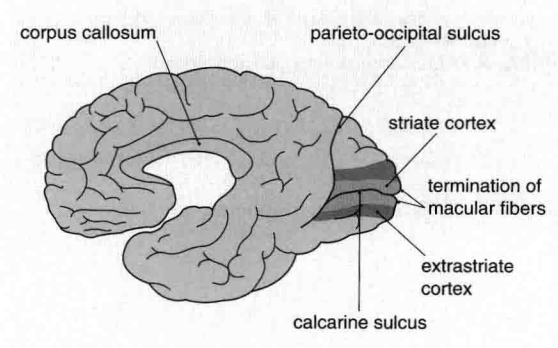


Fig. 1.3 The occipital lobe. The striate (primary visual) and extrastriate (visual association) cortices are shown, and termination of the macular fibers at the poles.

Visual hallucinations in seizures of the primary visual cortex are unformed (flashes of light and geometric shapes); those due to seizure activity from the visual association cortex or its connections with the temporal cortex are formed (objects, people).

Summary

Focal damage to the cerebral hemispheres usually results from vascular events (infarction or hemorrhage), tumors, trauma, or localized inflammatory lesions (e.g., abscess, tuberculoma). Generalized or multifocal cerebral dysfunction results most often from degenerative dementias (Alzheimer's disease, Pick's disease), multiple infarcts, demyelination, or diffuse infections (encephalitis, meningitis).

Fig. 1.4. summarizes the symptoms that may arise from focal lesions of the cerebral hemisphere.