Merritt's Textbook of Neurology

SEVENTH EDITION

Edited by

LEWIS P. ROWLAND, M.D.

Merritt's Textbook of Neurology

SEVENTH EDITION

Edited by

LEWIS P. ROWLAND, M.D.

Columbia University College of Physicians and Surgeons
Neurological Institute of Presbyterian Hospital
New York, New York

LEA & FEBIGER 1984 PHILADELPHIA

Lea & Febiger
600 Washington Square
Philadelphia, Pennsylvania 19106
U.S.A.

First Edition, 1955
Reprinted, 1955, 1957

Second Edition, 1959
Reprinted, 1959, 1961

Third Edition, 1963
Reprinted, 1964, 1966

Fourth Edition, 1967
Reprinted, 1968, 1969, 1970

Fifth Edition, 1973
Reprinted 1974, 1975, 1977

Sixth Edition, 1979
Reprinted, 1981

Seventh Edition, 1984

Library of Congress Cataloging in Publication Data

Merritt, H. Houston (Hiram Houston), 1902–1979
Merritt's textbook of neurology.

Rev. ed. of: Textbook of neurology. 6th ed. 1979.
Bibliography: p.
Includes index.
1. Nervous system—Diseases. 2. Neurology.
I. Rowland, Lewis P. II. Title. III. Title: Textbook of neurology. [DNLM: 1. Nervous system diseases. WL 100 M572t]
RC346.M4 1984 616.8 83–19550
ISBN 0–8121–0893–0

Print No. 5 4 3 2 1

PREFACE

The first edition of this Textbook was published in 1955; the sole author was H. Houston Merritt. He remained the only author until the 5th edition, when he began to accept the help of some of his former students. In 1977, at the time of his 75th birthday celebration, we suggested that he allow us to help more actively in the preparation of the sixth edition. However, once again, he wanted to do most of it himself, and he did, despite a gait disorder that was increasingly disabling. The book was published less than a month after he died in 1979, a few days before his 77th birthday.

Merritt had many talents—as clinician, clinical investigator, discoverer of phenytoin, editor, and administrator—but he also took his role as teacher seriously and he trained two generations of neurologists, first at the Boston City Hospital and Harvard Medical School between 1933 and 1945, then at Montefiore Hospital (1945–48) and the New York Neurological Institute, Presbyterian Hospital (1948–1967) as Professor of Neurology at Columbia University. In the decade after his formal retirement, he continued to teach medical students and house officers.

The textbook was another of his teaching achievements, and for many years, it was the one used by most students throughout the United States. Editions in Spanish, Italian, Portuguese, Turkish, and Japanese languages assured worldwide influence. When Dr. Merritt died, it therefore seemed natural to use the Textbook as the vehicle for an appropriate memorial, which would serve two purposes. First, it would allow his many students to participate in paying homage and it would be a historical document that would testify to the achievements of Merritt's students. Second, the memorial would not be merely decorative; it would be useful in a way that Houston Merritt would have appreciated.

The large number of participants in this volume departs from Merritt's own solo practice. Most contributors were selected because they had been either house officers or staff members at the Neurological Institute during Merritt's tenure and then had achieved outstanding careers of their own; a few came to NINY in the last decade of his life. A motley crew, they are nevertheless truly distinguished. Among them are 35 current or former heads of departments of neurology throughout the United States, and others who achieved renown in different clinical ways.

Naturally, this is not the total number. A few of Merritt's students have already died, including Saul Korey (who was Chairman of Neurology, first at Case-Western Reserve and then at The Albert Einstein College of Medicine) and Warren Kempinsky (Marquette University). And there are some who became heads of departments, not in Neurology but in Anatomy or Neuroscience (Malcolm Carpenter and Dominic Purpura), or Psychiatry (John Nardini and Robert Williams). Two Neurology Chairmen were not able to participate in the project (Robert McMasters, formerly Chairman at University of Texas, San Antonio, and Clark Randt, NYU).

Few individuals have ever had such an influence on the emerging neurology of a country. It is not likely to happen in the United States again; the period of expansion ended more than a decade ago and now there are many centers of excellence throughout the country, each with its own training program.

In asking Merritt's students to help to prepare this revision, we set some guidelines. The success of earlier editions was due in large measure to Merritt's style—direct, unadorned, clear, and without excess wordiness; we asked authors to recognize these qualities. They were asked to preserve as much as the original text as possible, bringing it up to date. Merritt preferred statements of fact to opinion; his text was prepared with tables of relative frequencies of manifestations of different diseases or actual outcomes of therapy, not merely opinion; we wanted to retain that characteristic. Opinions were to be clearly identified. He alway asked for new illustrations and we have provided more with this edition. Merritt always managed to include a few lines, at least, about "new"

diseases and to cite an appropriate reference or two. The reference lists have been selected carefully, another characteristic of previous editions.

The contributors responded in good humor; some were asked to revise major sections of this volume because they had done so in previous editions; others, equally distinguished, were asked to provide only a few paragraphs. These tasks were never refused because they were either overwhelming or demeaning. Some authors retained large sections of the 6th edition, others preferred to prepare an entirely new revision. With 70 authors, variations in style and content could have led to much unevenness and it was the editor's responsibility to smooth out the chapters. Contributors have been generous in allowing modification. We hope that the result will be seamless, that it will be impossible for the reader to know whether a section was written mostly by Merritt, mostly by a student alone, equally by both Merritt and a student, or with much editing.

The general order of the chapters is much as it was in the 6th edition; a few have been shuffled around. Merritt thought that clinical neurology would have achieved a rational basis when he could arrange all the chapters in terms of etiology, but that is still impossible. We have therefore arranged the chapters according to principles that are stated in the table of contents, but the principles are not entirely consistent; some are based on etiology or pathogenesis and others are based on anatomy. We hope that the arrangement is useful; that is the final test.

We have added one new section that Dr. Merritt had deliberately avoided. He never included a review of anatomy or provided a guide to the neurologic examination because other books did that very well and were familiar to medical students before they appeared on the neurologic floors for the clinical clerkship. Reviewers, however, have criticized the book on the grounds that it provided excellent descriptions of each disease, but the student had to know which disease the patient had before the student could look it up in Merritt's. We have therefore provided a brief introduction on neurologic diagnosis, which could have been entitled *How to Use This Book*. Confronted with a particular patient who has particular symptoms, the reader can be guided from a description of that symptom in Chapter I to the several diseases that might cause the symptom. Subsequent chapters can then be consulted for more detailed discussion of the specific diseases, including treatments as well as diagnosis.

This arrangement, however, caused some problems because we found it necessary to discuss seizures and headache as "symptoms" in the introductory chapter and as "conditions" later. In contrast, the new chapter on sleep disorders was not divided. Other new chapters are found in a chapter on the neurology of "environmental" disorders. Drs. Jubelt and Harter provided the section on AIDS in a heroic eleventh-hour effort. The condition was unknown when this edition was planned, hence its placement at the end of the book rather than in the chapter on infections.

We were saddened by the death of one contributor, Elliott Weitzman. He was a pioneer in the study of sleep disorders and what he called "chronobiology." He had just started a new laboratory at the time of his death at age 54 on June 15, 1983. Fortunately, he had completed his contributions to this book.

The editor is indebted to Ms. Shirley Susarchick for retyping many of the chapters as they came into our office, and we are grateful to secretaries throughout the country for their help. Drs. Sadek Hilal, Michael Mawad, and S. R. Ganti were extremely helpful in providing new illustrations for this edition. Mrs. Emilene Lopat provided bibliographic assistance for this volume as she did for the first edition and every one that followed, and Ms. Edna Borea supervised the office staff for this volume as she had for preceding volumes. Mr. Kenneth Bussy and Mr. John Hicks-Courant of Lea & Febiger saw us through every stage involved in the production of the book.

Two women who influenced Houston Merritt should also be mentioned in gratitude. All preceding volumes were dedicated to Mabel Carmichael Merritt, and she continues to be interested in the affairs of this department and in this book. The other person is Lucy G. Moses, who has supported the department, and therefore this book, in many times of urgent need. Finally, we thank the spouses and children of all contributors for making the volume possible. None have been more forbearing than Esther E. Rowland and our children, Andrew, Steven, and Judith.

New York, New York Lewis P. Rowland

CONTRIBUTORS

Milton Alter, M.D., Ph.D.
Professor and Chairman, Department of Neurology, Temple University School of Medicine.
Chairman, Department of Neurology, Temple University Hospital.
Philadelphia, Pennsylvania

J. Lobo Antunes, M.D.
Associate Professor of Neurological Surgery, Columbia University College of Physicians and Surgeons.
Associate Attending in Neurological Surgery, Neurological Institute of Presbyterian Hospital.
New York, New York

Alan M. Aron, M.D.
Professor of Clinical Neurology, Professor of Clinical Pediatrics, Mt. Sinai School of Medicine.
Director of Child Neurology, Attending Neurologist and Pediatrician, Mt. Sinai Hospital.
New York, New York

James Henry Austin, M.D.
Professor and Chairman, Department of Neurology, University of Colorado Health Sciences Center.
Denver, Colorado

Howard S. Barrows, M.D.
Professor of Neurology and Medical Education, Southern Illinois University School of Medicine.
Springfield, Illinois

Myles M. Behrens, M.D.
Associate Professor of Clinical Ophthalmology, Columbia University College of Physicians and Surgeons.
Associate Attending Ophthalmologist, Presbyterian Hospital.
New York, New York

Leonard Berg, M.D.
Professor of Clinical Neurology, Washington University School of Medicine.
Neurologist, Barnes Hospital, Jewish Hospital of St. Louis, St. Louis Children's Hospital.
St. Louis, Missouri

Patrick F. Bray, M.D.
Professor of Neurology and Pediatrics, University of Utah School of Medicine.
Associate Neurologist and Pediatrician, University of Utah Medical Center.
Salt Lake City, Utah

John C.M. Brust, M.D.
Professor of Clinical Neurology, Columbia University College of Physicians and Surgeons.
Director, Department of Neurology, Harlem Hospital Center, Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Rosalie A. Burns, M.D.
Professor and Chairman, Department of Neurology, The Medical College of Pennsylvania.
Director, Department of Neurology, Hospital of the Medical College of Pennsylvania.
Philadelphia, Pennsylvania

Sidney Carter, M.D.
Professor Emeritus of Neurology and Pediatrics, Columbia University College of Physicians and Surgeons.
Chief, Department of Neurology, Blythedale Children's Hospital.
Valhalla, New York

Abe M. Chutorian, M.D.
Professor of Clinical Pediatrics and Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Lucien J. Côté, M.D.
Associate Professor of Neurology, Associate Professor of Rehabilitation Medicine, Columbia University College of Physicians and Surgeons.
Associate Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Darryl C. DeVivo, M.D.
Sidney Carter Professor of Neurology, Professor of Pediatrics, Columbia University College of Physicians and Surgeons.
Director of Pediatric Neurology, Attending Neurologist, Attending Pediatrician, Neurological Institute of Presbyterian Hospital.
New York, New York

Salvatore DiMauro, M.D.
Professor of Neurology, Co-Director of the H. Houston Merritt Clinical Research Center for Muscular Dystrophy and Related Diseases, Columbia University College of Physicians and Surgeons.
New York, New York

Roger C. Duvoisin, M.D.
Professor and Chairman, Department of Neurology, University of Medicine and Dentistry of New Jersey—Rutgers Medical School.
Chief, Neurology Service, Middlesex General—University Hospital.
New Brunswick, New Jersey

Stanley Fahn, M.D.
H. Houston Merritt Professor of Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Robert A. Fishman, M.D.
Professor and Chairman, Department of Neurology, University of California Medical School—San Francisco.
San Francisco, California

John M. Freeman, M.D.
Professor of Neurology and Pediatrics, Johns Hopkins University School of Medicine.
Director, Child Neurology Service, Johns Hopkins Hospital.
Baltimore, Maryland

Arnold P. Friedman, M.D.
Adjunct Professor of Neurology, University of Arizona College of Medicine.
Consultant, Neurological Associates of Tucson.
Tuscon, Arizona

Sid Gilman, M.D.
Professor and Chairman, Department of Neurology, University of Michigan Medical School.
Chief, Neurology Service, University Hospital.
Ann Arbor, Michigan

Gilbert H. Glaser, M.D., Med. Sc.D.
Professor and Chairman, Department of Neurology, Yale University School of Medicine.
Chief of Neurology, Yale-New Haven Hospital and Medical Center.
New Haven, Connecticut

Arnold P. Gold, M.D.
Professor of Clinical Neurology, Professor of Clinical Pediatrics, Columbia University College of Physicians and Surgeons.
Attending Neurologist and Pediatrician, Neurological Institute of Presbyterian Hospital.
New York, New York

Mark A. Goldberg, Ph.D., M.D.
Professor of Neurology, Professor of Pharmacology, University of California School of Medicine—Los Angeles.
Chairman, Department of Neurology, Harbor-UCLA Medical Center.
Los Angeles, California

David Goldblatt, M.D.
Professor of Neurology, University of Rochester School of Medicine and Dentistry.
Attending Neurologist, Strong Memorial Hospital.
Rochester, New York

Eli S. Goldensohn, M.D.
Professor of Neurology, Albert Einstein College of Medicine, Professor Emeritus of Neurology, Columbia University College of Physicians and Surgeons.
Consulting Neurologist, Presbyterian Hospital, Attending Neurologist, Montefiore Medical Center, Bronx Municipal Hospital Center.
New York, New York

Melvin Greer, M.D.
Professor and Chairman, Department of Neurology, University of Florida College of Medicine.
Chief, Neurology Service, Shands Teaching Hospital.
Gainesville, Florida

Donald H. Harter, M.D.
Charles L. Mix Professor of Neurology, Chairman, Department of Neurology, Northwestern University School of Medicine.
Chairman, Department of Neurology, Attending Neurolgist, Northwestern Memorial Hospital.
Chicago, Illinois

William K. Hass, M.D.
Professor of Neurology, New York University School of Medicine.
Attending Neurologist, New York University Medical Center.
New York, New York

Christian Herrman, Jr., M.D.
Professor and Vice Chairman, Department of Neurology, University of California School of Medicine—Los Angeles.
Attending Neurologist, University of California Hospitals—Los Angeles, Consultant in Neurology, St. John's Hospital, Santa Monica, Wadsworth Veterans Administration Hospital.
Los Angeles, California

T.R. Johns II, M.D.
Alumni Professor and Chairman, Department of Neurology, University of Virginia School of Medicine.
Neurologist-in-Chief, Neurological Service, University of Virginia Hospital.
Charlottesville, Virginia

William G. Johnson, M.D.
Associate Professor of Clinical Neurology, Columbia University College of Physicians and Surgeons.
Associate Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Burk Jubelt, M.D.
Assistant Professor of Neurology, Northwestern University Medical School.
Associate Attending Neurologist, Northwestern Memorial Hospital.
Chicago, Illinois

Robert Katzman, M.D.
Professor and Chairman, Department of Neurology, Albert Einstein College of Medicine.
Attending Neurologist, Montefiore Medical Center.
New York, New York

Charles Kennedy, M.D.
Professor of Pediatrics and Neurology, Georgetown University of Medicine, Washington, D.C.
Guest Worker, Laboratory of Cerebral Metabolism, National Institute of Mental Health.
Bethesda, Maryland

M. Richard Koenigsberger, M.D.
Associate Professor of Neurosciences and Pediatrics, University of Medicine and Dentistry of New Jersey.
Director, Department of Pediatric Neurology, College Hospital, United Children's Hospital, Beth Israel Hospital.
Newark, New Jersey

Robert B. Layzer, M.D.
Professor of Neurology, University of California School of Medicine—San Francisco.
San Francisco, California

Niels L. Low, M.D.
Professor of Clinical Neurology and Clinical Pediatrics, Columbia University College of Physicians and Surgeons, New York, New York
Clinical Director, Blythedale Children's Hospital.
Valhalla, New York

Elliott L. Mancall, M.D.
Professor and Chairman, Department of Neurology, Hahneman University.
Philadelphia, Pennsylvania

J.T. Marotta, M.D.
Professor of Medicine (Neurology), Associate Dean of Clinical and Institutional Affairs, University of Toronto Faculty of Medicine.
Senior Neurologist, Wellesley Hospital.
Toronto, Ontario, Canada

John H. Menkes, M.D.
Clinical Professor of Pediatrics and Neurology, University of California School of Medicine—Los Angeles.
Attending Neurologist, UCLA Center for Health Sciences, Cedars-Sinai Medical Center.
Los Angeles, California

J.P. Mohr, M.D.
Sciarra Professor of Clinical Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Theodore L. Munsat, M.D.
Professor of Neurology, Tufts University School of Medicine.
Director, Neuromuscular Research Unit, New England Medical Center.
Boston, Massachusetts

Audrey S. Penn, M.D.
Professor of Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

David E. Pleasure, M.D.
Professor of Neurology, Pediatrics, and Orthopedic Surgery, University of Pennsylvania School of Medicine.
Attending Neurologist, Hospital of the University of Pennsylvania, Children's Hospital of Philadelphia.
Philadelphia, Pennsylvania

Charles M. Poser, M.D.
Professor of Neurology, Boston University School of Medicine, Lecturer on Neurology, Harvard Medical School.
Attending Neurologist, Boston University Hospital, Beth Israel Hospital, Veterans Administration Hospital.
Boston, Massachusetts

Leon D. Prockop, M.D.
Professor and Acting Chairman, Department of Neurology, University of South Florida College of Medicine.
Consultant, James A Haley Veterans Administration Hospital, Attending Neurologist, Tampa General Hospital.
Tampa, Florida

Isabelle Rapin, M.D.
Professor of Neurology and Pediatrics, Albert Einstein College of Medicine.
Attending Neurologist and Child Neurologist, Albert Einstein Affiliated Hospitals.
New York, New York

Neil H. Raskin, M.D.
Professor of Neurology, University of California School of Medicine—San Francisco.
San Francisco, California

Ralph W. Richter, M.D.
Professor of Neurology, Oral Roberts University School of Medicine.
Director, Neurology Division, City of Faith Medical Center, Chairman, Neurology Section, St. John Medical Center.
Tulsa, Oklahoma

Roger N. Rosenberg, M.D.
Professor of Neurology and Physiology, Chairman, Department of Neurology, University of Texas Health Sciences Center, Southwestern Medical School.
Chief, Neurological Services, Parkland Hospital, Children's Medical Center.
Dallas, Texas

Allen D. Roses, M.D.
Professor and Chief, Division of Neurology, Duke University Medical Center.
Durham, North Carolina

Lewis P. Rowland, M.D.
Henry and Lucy Moses Professor, Chairman, Department of Neurology, Columbia University College of Physicians and Surgeons.
Director, Neurology Service, Neurological Institute of Presbyterian Hospital.
New York, New York

Labe C. Sheinberg, M.D.
Professor of Neurology and Rehabilitative Medicine, Director, Medical Rehabilitation Research and Training Center for Multiple Sclerosis, Albert Einstein College of Medicine.
New York, New York

Edward B. Schlesinger, M.D.
Professor Emeritus of Neurological Surgery, Columbia University College of Physicians and Surgeons.
Consultant in Neurological Surgery, Neurological Institute of Presbyterian Hospital.
New York, New York

Donald L. Schotland, M.D.
Professor of Neurology, University of Pennsylvania School of Medicine.
Attending Neurologist, Hospital of the University of Pennsylvania.
Philadelphia, Pennsylvania

James F. Schwartz, M.D.
Professor of Pediatrics and Neurology, Emory University School of Medicine.
Chief, Division of Pediatric Neurology, Grady Memorial Hospital, Henrietta Egleston Hospital for Children.
Atlanta, Georgia

Daniel Sciarra, M.D.
Professor of Clinical Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

William A. Sibley, M.D.
Professor of Neurology, University of Arizona College of Medicine.
Attending Neurologist, University Hospital, Tucson Medical Center Hospital, Consultant, St. Joseph's Hospital, St. Mary's Hospital.
Tucson, Arizona

Bertram E. Sprofkin, M.D.
Clinical Professor of Neurology, Associate Clinical Professor of Neuropathology, Vanderbilt Medical School.
Nashville, Tennessee

Bennett M. Stein, M.D.
Byron Stookey Professor, Chairman, Department of Neurological Surgery, Columbia University College of Physicians and Surgeons.
Director of Neurosurgery, Neurological Institute of Presbyterian Hospital.
New York, New York

Hartwell G. Thompson, M.D.
Professor and Head, Department of Neurology, University of Connecticut School of Medicine.
Chief of Neurology, John Dempsey Hospital, University of Connecticut Health Center
Farmington, Connecticut

James F. Toole, M.D.
Walter C. Teagle Professor of Neurology, Wake Forest University Bowman Gray School of Medicine.
Winston-Salem, North Carolina

Carmine T. Vicale, M.D.
Professor Clinical Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

Leon A. Weisberg, M.D.
Professor of Neurology, Tulane Medical School.
Director of Neurology-Neurosurgery Service, Charity Hospital of New Orleans, Section Director of Adult Neurology, Tulane University Hospital.
New Orleans, Louisiana

Elliott Weitzman, M.D.†
Director, Institute of Chronobiology, New York Hospital—Cornell Medical Center.
White Plains, New York

†Deceased

Harry H. White, M.D.
Attending Neurologist, Mercy Medical Center, La Plata Community Hospital.
Durango, Colorado

Melvin D. Yahr, M.D.
Chairman, Department of Neurology, Mt. Sinai College of Medicine.
New York, New York

Frank M. Yatsu, M.D.
Professor and Chairman, Department of Neurology, University of Texas Medical Center at Houston.
Chief, Neurology Service, Hermann Hospital, Consultant, St. Luke's Hospital.
Houston, Texas

Dewey K. Ziegler, M.D.
Professor and Chairman, Department of Neurology, University of Kansas College of Health Sciences and Hospital School of Medicine.
Attending Neurologist, University of Kansas Medical Center.
Kansas City, Kansas

Earl A. Zimmerman, M.D.
Professor of Neurology, Columbia University College of Physicians and Surgeons.
Attending Neurologist, Neurological Institute of Presbyterian Hospital.
New York, New York

CONTENTS

NEUROLOGIC DISORDERS OF UNCERTAIN PATHOGENESIS

IX. Cerebral Degenerations of Childhood

X. Neurocutaneous Disorders

XI. Cranial Nerve Disorders

XII. Peripheral Nerve Disorders

XIII. Ataxias and Dementia

XIV. Movement Disorders

XV. Spinal Cord Diseases

Chapter 1

Symptoms of Neurologic Disorders

1. DELIRIUM AND DEMENTIA

Robert Katzman

Delirium and dementia are two of the most common disorders of elderly patients, but both conditions may occur at any age. *Delirium* is the acute confusional state that may accompany infections, fever, metabolic disorders, and other medical or neurologic diseases. *Dementia*, in contrast, is usually chronic and progressive, and is usually caused by degenerative diseases of the brain or by multiple strokes. The most significant difference, however, is that delirium is manifested by a fluctuating mental state, whereas patients suffering from dementia are usually alert and aware until late in the course of the disease.

Delirium

The word *delirium* comes from the Latin word meaning "to leave the furrow (or track)." It is used to refer to a mental disturbance that is marked by clouding of consciousness and inability to maintain attention; there is often restlessness and incoherence. Sometimes there are misinterpretations, illusions, and hallucinations. Characteristics that lead to the classification of a mental state as delirium include impairment of memory and intellectual function; the presence of a medical or neurologic condition to which the mental impairment is secondary; the disappearance of mental impairment if the primary medical or neurologic disorder is reversed; and the effect of the primary disorder on the brain, which is diffuse rather than focal.

Delirium is involved in a wide range of clinical states. In elderly patients with severe bronchopneumonia, delirium takes the form of lethargy and confusion; such patients lie quietly in bed despite the concomitant tachypnea. Delirium is a common secondary disorder in previously healthy patients who are suffering a severe febrile illness. *Delirium tremens* differs from other forms of delirium in that it occurs only in persons who are addicted to alcohol. It develops 24 to 48 hours after withdrawal from alcohol. The onset is marked by confusion, agitation, and hyperactivity. Memory is affected and there are hallucinations. Autonomic hyperactivity results in tachycardia and high fever. If untreated, delirium tremens can be fatal.

Delirium is treated as a medical emergency because the disease or drug intoxication to which it is secondary may be fatal if untreated. The following drugs have been known to cause delirium.

Atropine and related compounds
Barbiturates
Bromides
Chlordiazepoxide (Librium)
Chloral Hydrate
Clonidine
Diazepam (Valium)
Digitalis
Flurazepam (Dalmane)
Glutethimide (Doriden)
Haloperidol
Lithium
Meprobamate
Mephenytoin
Methyl Dopa
Phenytoin
Propanolol
Tricyclic antidepressants

The fluctuating state of awareness in delirium is accompanied by characteristic electroencephalographic (EEG) changes. The varying level of attention parallels slowing of the background EEG rhythms. Appropriate treatment of the underlying disease improves both the mental state and EEG of the patient. A patient with bronchopneumonia shows less confusion and a more normal EEG when given an oxygen mask. This finding has been

incorporated into medicine so firmly that the steps needed to improve confusional states and altered levels of awareness in patients arriving in emergency rooms are among the first practical aspects of medicine learned.

Mental Status Examination. The mental status evaluation is an important part of every neurologic examination. It includes evaluation of awareness and consciousness, behavior, emotional state, content and stream of thought, and sensory and intellectual capabilities. Specific aspects of intellectual activity are most often impaired in organic disease of the brain. Evaluation of these aspects constitute the mental status examination, which is important in the diagnosis of both delirium and dementia. Intellectual impairment is obvious in such florid conditions as delirium tremens or advanced dementia, but a cognitive deficit may not be evident in early cases of delirium or dementia unless the physician specifically tests mental status. Traditionally, mental status examinations test *information* (e.g., where were you born? what is your mother's name? who is the President? when did World War II occur?), *orientation* (what place is this? what is the date? what time of day is this?), *concentration* is tested by using serial reversals (e.g., spell "world" backwards, name the months of the year backwards beginning with December); *calculation* (e.g., simple arithmetic, making change, counting backwards by 3's or 7's); *reasoning* (e.g., interpretation of proverbs); *judgment and memory* (e.g., identify these three objects, please try to remember their names, I will state a name and address, please repeat after me and try to remember for a few minutes)—(Table 1-1). The most important and sensitive items are probably orientation to time, serial reversals, and a memory phrase.

In addition to testing the mental status, it is necessary to test higher intellectual functions, including disorders of language (dysphasias), constructional apraxia (Fig. 1-1), right-left disorientation, inability to carry out complex commands, especially those requiring crossing the midline (e.g., touch your left ear with your right thumb), inability to imagine acts (ideomotor apraxia; e.g., pretend that you have a book of matches and show me how you would light a match); unilateral neglect, or inattention on double stimulation. These abnormalites are often associated with more focal brain lesions, but may also be impaired in delirium or dementia.

Dementia

Dementia is characterized by progressive intellectual deterioration that is sufficiently severe to interfere with social or occupational functions. Memory, orientation, abstraction, ability to learn, visuospatial perception, and constructional praxis are all impaired in dementia. In contrast to patients with delirium, subjects with dementia are alert and aware until late in the course of the disease. Delirium is most often associated with intercurrent systemic diseases or drug intoxication, but dementia is usually due to a primary degenerative or structural disease of the brain. Alzheimer disease (see Article 106, Presenile Dementia) accounts for over 50% of cases in both clinical and autopsy series (Table 1-2). Huntington disease (see Article 108, Huntington Disease and Other Forms of Chorea) is much less common, but is still an important cause in the preseneium. Parkinsonism (see Article 113, Parkinsonism) is sometimes associated with dementia. Less common degenerative diseases include Pick disease (see Article 106, Presenile Dementia), progressive supranuclear palsy (see Article 114, Progessive Supranuclear Palsy), and the hereditary ataxias (see Article 104, Hereditary Ataxias)

Twenty to 25% of cases of dementia are due to vascular disease. Contrary to earlier beliefs, dementia correlates less with the degree of cerebral arteriosclerosis than with the extent of destruction of cerebral hemisphere by multiple strokes; hence, the term *multi-infarct dementia* has come into use. Cognitive impairment becomes evident when 50 g to 100 g of cerebral hemisphere is destroyed. The strokes may be due to thrombosis, emboli, or hemorrhages. Minute infarcts *(lacunae)* occur in the basal ganglia in the presence of hypertensive disease that involves brain arterioles; the resulting "lacunar state" sometimes gives rise to dementia. *Binswanger dementia* occurs when hypertensive vascular disease of brain arterioles results primarily in white-matter disease including lacunae, cysts, and demyelination.

Intracranial mass lesions, including brain tumors and subdural hematomas, cause dementia without focal neurologic signs in as many as 5% of cases of dementia in some series. With the use of CT, these patients are rapidly identified and treated; future series of dementia cases will probably include fewer subjects with dementia caused by intracranial mass lesions.

The frequency of chronic communicating hydrocephalus *(normal pressure hydrocephalus)* as a cause of dementia in adults varies from 1% to 5% in different series. Diagnosis is usually straightforward when the hydrocephalus follows intracranial hemorrhage, head injury, or meningitis, but in idiopathic cases there is often difficulty in differentiating communicating hydrocephalus from ventricular enlargement due to brain atrophy.

At the turn of the century, the most common

TABLE 1-1. Mental Status Examination.

*OMC**	*MSQ***	*Mini-Mental State****
1. What *year* is it now?	1. What is the name of this place?	ORIENTATION What is the (year) (season) (date) (day) (month)? Where are we: (state) (county) (town) (hospital) (floor)?
2. What *month* is it now?	2. Where is it located? (address)?	
3. Memory Phrase, Repeat phrase after me: John Brown, 42 Market Street, Chicago	3. What is today's date?	REGISTRATION Name three objects: take 1 second to say each. Then ask the patient to name all three after you have said them. Give 1 point for each correct answer. Then repeat them until he learns all three. Count trials and record.
4. About what *time* is it? (Within 1 hour)	4. What is the month now?	
5. *Count* backwards 20 to 1	5. What is the year?	
6. Say the months in reverse order	6. How old are you?	
7. Repeat the memory phase	7. When were you born (month)?	ATTENTION AND CALCULATION Serial 7s. 1 point for each correct. Stop after 5 answers. Alternatively spell "world" backwards.
	8. When were you born (year)?	
	9. Who is the President of the United States?	RECALL Ask for the three objects repeated above. Give 1 point for each correct.
	10. Who was the President before him?	LANGUAGE Name a pencil and a watch (2 points) Repeat the following "No ifs, ands, or buts." (1 point) Follow a 3-stage command: "Take a paper in your right hand, fold it in half, and put it on the floor" (3 points) Read and obey the following: Close your eyes (1 point) Write a sentence (1 point) Copy design (1 point)

*Orientation Memory Concentration. (From Katzman R, Brown T, Fuld P, et al. Am J Psychiatry 1983; 140:734–739.)
**Mental Status Questionnaire. (From Kahn RL, Goldfarb AI, Pollack M, Peck A. Am J Psychiatry 1960; 117:326–328.)
***(From Folstein MF, Folstein S, McHugh P. J Psychiatr Res 1975; 12:189–198.)

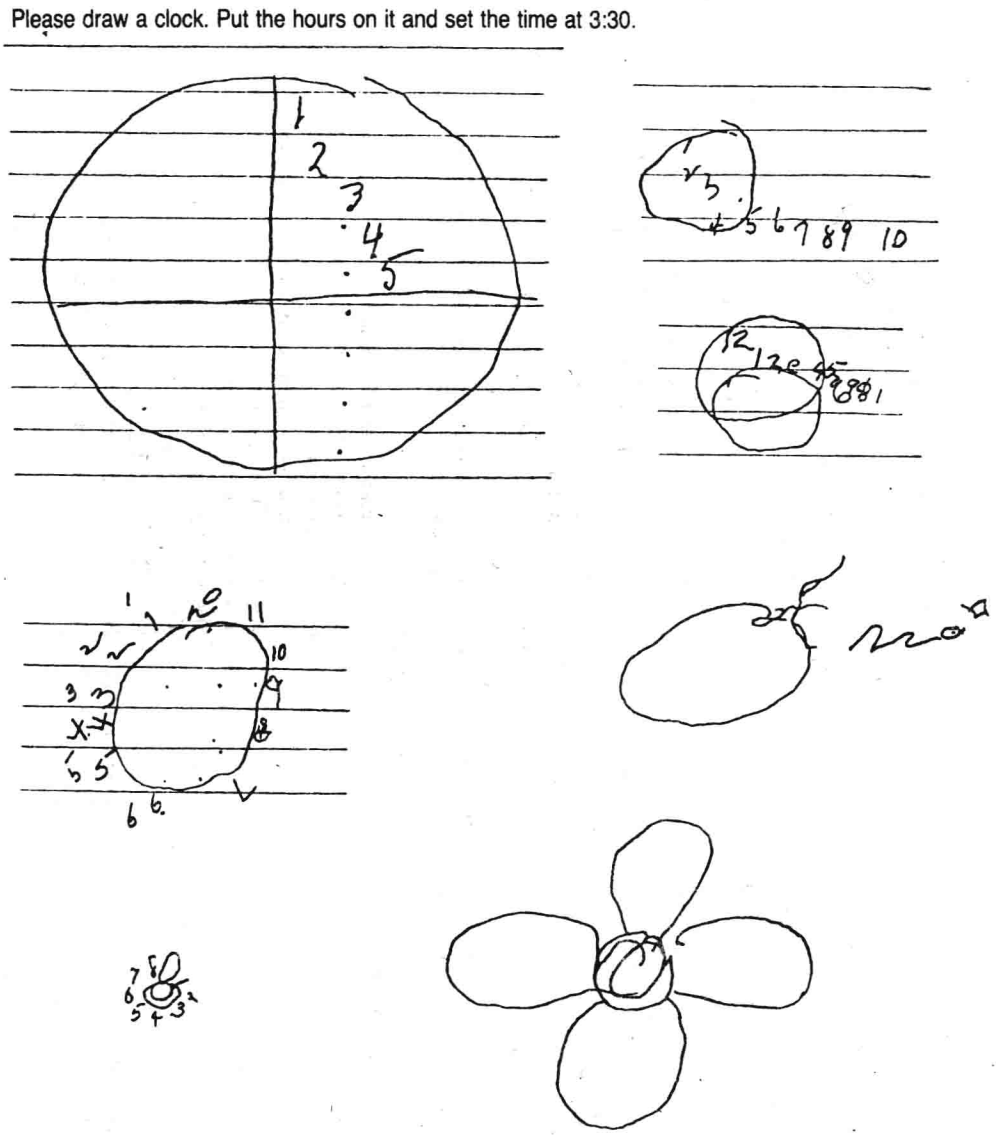

Fig. 1-1. A mental status examination question and examples of how a delirious patient might respond.

cause of dementia was neurosyphilis. Today, however, general paresis and other forms of neurosyphilis are rare. Creutzfeldt-Jakob disease has become the most common infectious cause of progressive dementia. Viral infections rarely present as chronic rather than acute encephalopathy. Fungal meningitis may occasionally present as dementia.

Nutritional, toxic, and metabolic causes of dementia are particularly important because they may be reversible. Korsakoff psychosis, usually found in alcoholics and attributed to thiamine deficiency, remains an important problem in our society. In contrast, the dementia of pellagra, a disorder produced by niacin and tryptophan deficiencies, has been almost entirely eliminated in the United States. Vitamin B_{12} deficiency occasionally causes dementia without anemia or spinal cord disease. Among the metabolic disorders that may present as dementia, hypothyroidism is the most important. Finally, prolonged administration of drugs may cause chronic intoxication (due to inability to metabolize the drug or to idiosyncratic reactions) that may be mistaken for dementia.