

The Neurologic Diagnosis

Jack N. Alpert

The Neurologic Diagnosis

A Practical Bedside Approach

Second Edition



Springer

Jack N. Alpert
Department of Neurology
University of Texas Medical
School at Houston
Houston, TX
USA

ISBN 978-3-319-95950-4 ISBN 978-3-319-95951-1 (eBook)
<https://doi.org/10.1007/978-3-319-95951-1>

Library of Congress Control Number: 2018956294

© Springer Nature Switzerland AG 2019

This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

The publisher, the authors, and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, express or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

This Springer imprint is published by the registered company Springer Nature Switzerland AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

In memory of Morris B. Bender, a superlative clinical neurologist of the twentieth century, who stimulated my lifelong interest in the manifestations, evaluation, and diagnosis of patients with neurologic diseases. His influence permeates this text.

Foreword

Assessment of neurological complaints, a high percent of a family practitioner and internist's practice, requires an accurate history and a careful neurologic examination. As Dr. Alpert expertly discusses in detail, the history provides clues to the clinical diagnosis, while the complementary neurologic examination localizes the lesion(s). Dr. Alpert is able to distil out from his over four decades of a busy practice and from his excellent teaching of medical students and neurological trainees a logical, readable, and provocative approach to each neurological complaint. In a chapter of 42 cases of *Diagnostic Dilemmas*, one's clinical acumen is challenged with practical questions and astute observations.

By discussing neurological disorders in terms of ten *Neuroanatomic Diagnoses*, Dr. Alpert compartmentalizes neurologic diseases into convenient and manageable discrete entities. By building upon the unique anatomy and physiology of each unit, greater logic is made in one's deductive reasoning for a diagnostic conclusion. Special emphasis is given to the *Six Major Decussations* with clinical correlations, as with strokes, autonomic disorders, neuromuscular diseases, and the poorly responsive patient. A separate chapter on *Common Symptoms in the Neurology Clinic* is a potpourri of frequently seen and rare cases which will challenge the beginner and the experienced practitioner of neurology. This textbook is highly recommended to the serious student of clinical neurosciences, be they medical students, trainees, or practitioners.

Frank M. Yatsu
Chairman, Emeritus, Department of Neurology
University of Texas Medical School at Houston
Houston, TX
USA

Preface

The second edition of this book retains the same emphasis on the simplification of making a neurologic diagnosis. The unique diagnostic principles of neurology are based on the requirement of an anatomic definition of the disease before a differential diagnosis can be offered. This process often disrupts the thinking of medical students and residents and certainly adds to the confusion of the practicing physician, thus providing the necessity for neurologic expertise and consultations. The explosion of efficacious treatment options for common neurologic diseases and disorders such as headache, epilepsy, multiple sclerosis, Parkinson's disease, sleep disorders, and stroke mandates an accurate diagnosis which depends on a thorough history and meticulous neurologic examination. This initial, critical, first step is the focus of this text.

The first eight chapters provide core knowledge of neuroanatomy and diagnostic principles. They include several updates on clinical-neuroanatomic correlations so that a rereading, although repetitive to some extent, should remain useful. Chapter 9 now includes a brief review of the muscular dystrophies with several added challenging cases. A new Chap. 10 on myelopathies not provided in the first edition adds brief discussions of a group of diseases which uniquely target the spinal cord. Although some are uncommon or rare, it is important to be mindful of them. As multiple sclerosis often begins with myelopathic features, the discussion of one case furnishes an opportunity for elaboration of new diagnostic criteria according to the recent (2017) revisions of McDonald's criteria. These are briefly summarized. A full review will require a return to source material.

Chapter 11 remains focused on the practical diagnosis of common neurologic diseases and disorders. The *International Classification of Headache Disorders (ICHD)*, third edition reported in *Cephalalgia* (2013) supplies the substrate for a reexamination of these entities. Positron emission tomography (PET) imaging of the brain during migraine, relatively unknown among most medical practitioners and some neurologists, is now included. The section on vertigo has been enlarged by the addition of new cases as well as modified by the clarification and explication of relatively new clinical signs. The section on epilepsy incorporates the International League Against Epilepsy (ILAE) classification of epilepsies published in *Epilepsy*

(2017). The transition from the old terminology will undoubtedly be resisted but, nevertheless, eventually accepted, likely over many years since the prior classification in 1981 held force for decades. Discussion of psychogenic factors has been expanded and incorporates principles of differentiating psychogenic non-epileptic seizures (PNES) or pseudoseizures from seizures and psychogenic pseudosyncope (PPS) from syncope. The section on sleep disorders has broadened and will include elements of the most recent International Classification of Sleep Disorders 3rd Edition 2014. Understanding these conditions is increasingly recognized as furnishing important clinical clues for diagnoses, such as recognizing rapid eye movement (REM) sleep behavior disorder as a prelude to Parkinson's disease, as well as providing treatment opportunities for the maintenance of overall health.

In Chap. 12, Diagnostic Dilemmas, recently discovered neurologic diseases are introduced such as autoimmune encephalitis, chronic traumatic encephalopathy, and the neurologic complications of Zika virus infections. Advances in the neurologic armamentarium for the diagnosis of Parkinson's disease, particularly the non-motor manifestations, are explored. This chapter is also expanded by several new challenging case reports of rare diseases. In Chap. 13, additional syndromes have been added, always intriguing for many neurologists, since they add an element of historical interest.

The relevance of the neurologic examination has been questioned by many physicians, including some experienced neurologists. There have even been statements that simply reviewing an MRI scan with a neuroradiologist could easily be more valuable. Forgotten, astonishingly, is the reality that the majority of patients visiting most neurologic clinics have normal studies. Just think of migraine, epilepsy, Parkinson's disease, sleep and neuromuscular disorders, as well as neuro-otologic conditions whose diagnoses depend on a thorough history and neurologic examination. Additionally, the benefit of a soothing touch which may convey interest and empathy by the caring, thoughtful examiner provides a form of treatment by reducing the discomfort of anxiety. Abandonment of this gift which is bequeathed to all physicians is a developing tragedy of modern medicine.

A video has been provided which focuses on the standard neurologic examination methods as well as special techniques which may have been forgotten or omitted from current neurological training. (It is available as electronic supplementary material on this book's page on the Springerlink.com website. Those without a subscription to Springerlink.com can still view the ESM material for free.)

Certainly, superb training programs which focus on stroke may differ significantly from those which concentrate on neuromuscular diseases.

In summary, it is my hope that this text will expand the horizons of the reader beyond that with which he is so familiar.

Acknowledgments

Without the patience and encouragement of my wife, Ruth, this book could never have been completed. Donna J. Williams was my invaluable, conscientious, and superb transcriptionist. A number of neurologists, who provided excellent advice for the first edition, which is an integral element of this text, include Drs. Frank Yatsu, Stanley Appel, Randolph Evans, James Grotta, Ernesto Infante, Frank Perez, Victor Rivera, and Loren Rolak. Michael Griffin, my editorial advisor, insisted on a well-disciplined, correctly documented manuscript and prodded me to complete the text without excessive delay. I appreciate everyone's influence and guidance.

Contents

1	Introduction	1
2	The Ten Neuroanatomic Diagnoses	3
	Common Neurologic Signs: Localizing Value	4
	Central Nervous System Disease (CNS), Peripheral Nervous System (PNS) and Muscle Disease (M)	4
	Cerebral	5
	Extrapyramidal	9
	Brainstem/Cerebellum	11
	Spinal Cord Lesions (Myelopathies)	15
	Radiculopathy	20
	Plexopathy	22
	Neuropathy	25
	Neuromuscular Junction	27
	Myopathy	28
	Meningeal Disease	29
	Synopsis	33
	Multiple Choice Questions	35
	Answers	37
	Bibliography	39
3	The Neurologic History Holds the Diagnostic Keys	41
	Neurologic Symptoms in Psychiatric Disease	50
	Bibliography	52
4	Neurologic Examination	53
	Vital Signs	54
	Mental Status Examination	55
	Visual Observations	55
	Auditory Observations	56
	Dysarthria	56
	Aphasia	57

Agnosias	60
Apraxias	61
Disorders of Attention and Recognition	62
Neuropsychological Evaluation	65
Episodic Memory	65
Semantic Memory	66
Working Memory	66
Procedural Memory	67
Cranial Nerve Examination	67
Olfactory Nerve (I)	67
Optic Nerve (II)	67
Optic Nerve: Evaluation of Central Vision	69
Visual Field Examination	71
Ophthalmoscopic Findings and Visual Field Defects with Lesions of the Visual System	76
3rd, 4th, and 6th Cranial Nerves (Oculomotor, Trochlear, Abducens)	82
Eyelids: A Critical Initial Observation	82
Extraocular Muscles	84
Actions of Extraocular Muscles with the Eye in Primary Position	84
Saccades and Pursuit	88
Dysconjugate Gaze and Abnormal Eye Positions	89
Gaze Paresis	92
Pretectal Syndrome	92
Eye Deviations	92
Ocular Oscillations: Nystagmus	93
Special Tests	98
Ocular Oscillations: Dyskinesias	99
Pupils: Observations	99
Trigeminal Nerve	102
Seventh Cranial Nerve (Facial)	105
Vestibulocochlear Nerve (8th Cranial Nerve)	109
Cochlear Nerve: Background Anatomy	109
Cochlear Nerve: Clinical Evaluation	109
Vestibular Nerve: Background Anatomy	110
Vestibular Nerve: History	110
Vestibular Nerve: Physiology	111
Vestibular Nerve: Examination	111
Glossopharyngeal Nerve (9th Cranial Nerve)	116
Vagus Nerve (10th Cranial Nerve)	117
Spinal Accessory Nerve (11th Cranial Nerve)	118
Hypoglossal Nerve (12th Cranial Nerve)	119
Motor Examination	120
General Observations	120
Close Inspection	126

Coordination	127
Strength	128
Tone	130
Three Motor Systems and Neurologic Signs	132
Gait and Station Examination	133
Common Gait Disorders	133
Uncommon Gait Disorders	136
Reflex Examination	138
Method of Examination	139
Enhancement Technique	144
Reflex Aberrations	145
Superficial Reflexes	145
Abnormal Reflexes	146
Reflexes of Questionable Significance	148
Synopsis	148
Major Principles	149
Sensory Examination	149
Sensory Abnormalities According to Lesion Site	152
Nonorganic Sensory Loss	155
Sensory Terminology	156
Dermatome Patterns	156
Mechanical Signs	157
Meningeal Disease	157
Cervical and Lumbar Radiculopathy	158
Questions (True or False)	160
Answers	161
Bibliography	162
5 Evaluation of the Poorly Responsive Patient	163
Clouding of Consciousness (Lethargy)	164
Delirium	164
Obtundation	164
Stupor	164
Coma	164
Neurologic Examination	165
Inspection	165
Respiration	165
What Is the Respiratory Pattern?	165
Mental Status Examination	166
Cranial Nerves	169
Utility of Caloric Testing	170
Spontaneous Eye Movements in a Poorly Responsive Patient	171
Eye Deviations	172
Pupils	173
Corneal Reflexes	173

- Eyelids 174
- Palate 174
- Motor Function 175
- Reflexes 175
- Sensory 176
- Meningeal Signs 176
- Metabolic and Hypoxic-Ischemic Encephalopathies 176
 - Metabolic Encephalopathy 176
 - Respiratory Disorders 178
 - Eye Signs 179
 - Abnormalities of Motor Function 180
 - Differential Diagnosis of Metabolic Disorders 181
 - Specific Diseases Which Mimic the Confusion of Metabolic Encephalopathy 182
 - Case Reports 184
 - Hypoxic-Ischemic Encephalopathy 187
- Prognostic Factors in Comatose Survivors After CPR 188
- Traumatic Brain Injuries (TBI) 190
 - Glascow Coma Scale 190
- Herniation Syndromes 191
 - Falcine Herniations 192
 - Central Transtentorial Herniation 192
 - Uncal Herniation 195
 - Infratentorial Herniations 196
- Chronic Disorders of Consciousness 197
 - Coma 197
 - Vegetative State 197
 - Akinetic Mutism 198
 - Minimally Conscious State 198
 - Locked-in State 200
- Brain Death 200
- Questions (True or False) 204
- Answers 205
- References 205
- 6 The Six Major Anatomic Decussations with Clinical Correlation 207**
 - The Corticospinal Tract 207
 - The Corticobulbar Pathways 209
 - The Oculomotor Decussation and Associated Pathways 213
 - The Visual Pathways 217
 - The Sensory Systems 222
 - Questions (True or False) 225
 - Answers 226
 - Bibliography 227

7	Cerebrovascular Anatomy with Clinical Correlation	229
	Ischemic Stroke (Cerebral Infarction) and Transient Ischemic Attack (TIA).	229
	Summary.	243
	Venous Sinus Disease	252
	Intracerebral Hemorrhage.	254
	Subarachnoid Hemorrhage	258
	Questions (True or False)	264
	Answers.	265
	References.	266
8	Autonomic Nervous System Anatomy with Clinical Correlation	269
	Pupil	269
	Case Reports	275
	Blood Pressure	279
	Genitourinary System	283
	Anatomic and Physiologic Overview	283
	Neurogenic Bladder with Clinical Correlation	284
	Sexual Function	285
	Bowel Function.	286
	Questions (True or False)	286
	Answers.	287
	References.	288
9	Neuromuscular Diseases: Neuroanatomic and Differential	
	Diagnoses	289
	Case Reports	290
	Appendix A	342
	References.	344
10	Myelopathies	347
	Case Reports	348
	Answers.	360
	References.	375
11	Common Symptoms in the Neurology Clinic	377
	Headache.	377
	Migraine	403
	Trigeminal Neuralgia	405
	Temporal Arteritis	405
	Cluster Headache	405
	Chronic Paroxysmal Hemicrania	406
	Hypnic Headache	406
	Short-Lasting, Unilateral Neuralgiform Headaches with Conjunctival Injection and Tearing (SUNCT)	406
	Glossopharyngeal Neuralgia.	406

- Vertigo 408
 - Epilepsy 421
 - Epilepsy Syndromes 422
- Near-Syncope, Syncope, and Seizure 425
 - Psychogenic Non-Epileptic Seizures (PNES). 427
 - Psychogenic Pseudosyncope (PPS) 427
- Sleep Disorders 438
 - Sleep Apnea 440
 - Parasomnias 446
 - Chronic Primary Insomnia 448
 - Central Disorders of Hypersomnolence 450
 - Physiology of Sleep and Narcolepsy 451
 - Epworth’s Sleepiness Scale (ESS) 452
 - Idiopathic Hypersomnia 453
- Other Transient Neurologic Symptoms 454
 - “Drop Attack” (The Sudden Inexplicable Fall). 455
 - Confusion/Amnesia. 458
 - Transient Visual Impairment. 459
 - Transient Visual Illusions and Hallucinations. 462
 - Transient Motor and Sensory Disturbances 465
 - Paroxysmal Motor Phenomena. 468
- References. 470
- 12 Diagnostic Dilemmas 473**
 - References. 530
- 13 Neurologic Terminology 533**
 - Glossary 533
 - Acronyms 533
 - Signs 535
 - Syndromes. 545
- Case Report Index by Disease 559**