# Neurology in Clinical Practice

SIXTH EDITION

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**VOLUME 2 NEUROLOGICAL DISORDERS** 



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Volume II: Neurological Disorders
Sixth Edition

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# Foreword

The first ideas that led to Neurology in Clinical Practice (NICP) originated in Newcastle upon Tyne in the mid-1970s. Professor John Walton—now Lord Walton of Detchant, then professor of neurology and dean of the university's medical school-and several of us on the faculty believed we should write a Newcastle neurology textbook. We decided that the first section would describe how experienced neurologists approach common neurological conditions such as headache, walking difficulty, loss of vision, and so on. The second section would deal with neurological investigations such as neurophysiology and neuroimaging. The third section would provide an introduction to related neuroscience disciplines such as neurogenetics and neuroimmunology. The fourth section would outline the principles of management of neurological conditions, and the fifth would cover all the individual neurological diseases. The textbook would be divided into two volumes, with volume I containing the first four sections and volume II the neurological diseases.

The "Newcastle textbook" never got beyond the planning stage, and in 1977 I moved to Tufts New England Medical Center. There I started the journal, *Muscle and Nerve*, and was its founding editor for 10 years. However, the concept of an innovative practical textbook of neurology remained at the back of my mind. The opportunity to return to this project presented itself in 1987 when a small medical publisher approached me to write a book about neurology. A multiauthor textbook of the magnitude that I conceived needed at least four editors who were not only clinicians and research workers with expertise in the major neurological subspecialties, but who were also established leaders across the breadth of neurology. I approached Bob Daroff, Gerry Fenichel, and David Marsden—all giants in the field—and they agreed to join me in this project.

We chose the title, *Neurology in Clinical Practice*, because we wanted the book to be used not only by neurologists in training and practice but also by others whose specialties border upon neurology, such as internists and neurosurgeons. Together, Bob, Gerry, David, and I selected the authors for the 84 chapters that made up the first edition and laid out guidelines for the chapter, its content, and format. We set tough time schedules, and Bob Daroff, in particular, ensured that our authors met the deadlines. All four editors reviewed the manuscript for every submitted chapter to ensure uniformity of style and content.

During this time, the small medical publishing company was bought by Houghton Mifflin, which was then acquired by Butterworth (later Butterworth-Heinemann), which eventually became part of the Elsevier group. Nancy Megley was the publishing editor with Butterworth for the first edition. The fact that NICP was published at the end of 1990 with a 1991 copyright is proof of the support we had from our contributors and Butterworth.

We devoted a great deal of attention to the technical aspects of textbook production. For instance, we wished to have the highest quality reproduction of halftone illustrations and chose top-quality china clay paper for the book. The first edition, divided into Volume I, Principles of Diagnosis and Management, and Volume II, The Neurological Disorders, encompassed 1941 pages plus 88 pages of index and weighed 16 pounds; we may have been responsible for a number of hernias among our readers. The first edition of NICP received the Most Outstanding Book award for 1991 from the Association of American Publishers and was greeted with very favorable reviews by all the neurological journals. It soon established itself as a leading international textbook of neurology.

Wishing to keep NICP up to date, we published the second edition in 1996. We were fortunate to be joined by Susan Pioli, then director of medical publishing for Butterworth-Heinemann and later neurology publisher for Elsevier. Susan continued to work with us through the fifth edition. For the second edition, we selected a number of new authors, and the text was completely rewritten. In editing it, we embraced the digital age and went electronic with an added CD version. The five sections were merged into three: Part 1, Approach to Common Neurological Problems; Part 2, Neurological Investigations and Related Clinical Neurosciences; and Part 3, Neurological Diseases. By slightly reducing the grade of paper, we were able to produce a lighter book and accommodate much new material in 2128 pages plus a 117-page index. We also produced the Pocket Companion to Neurology in Clinical Practice, Second Edition, which was almost entirely the work of Gerry Fenichel. It became very popular with residents, who came to refer to it as "the Baby Bradley."

For the third edition (published in 2000), besides recruiting new authors and adding new material, we persuaded Butterworth-Heinemann to publish NICP online, and it became the first major neurology textbook to be available in that format. Our initial discussions had revolved around how much material we could get onto a CD—at that time, 500 MB was the maximum capacity—but that was enough space to include only the text and not the illustrations. In the end, we leapfrogged straight into online publishing with www. expertconsult.com, thereby allowing us to add much more content, particularly videos of electroencephalograms, electromyograms, and eye movements. Tragically, we were in the final stages of production on the third edition when David Marsden died; that edition was dedicated to his memory.

For the fourth edition, published in 2004, we invited Joe Jankovic to join us in David's place. Joe brought his expertise in movement disorders and was responsible for adding videos of these fascinating conditions to www.expertconsult.com. This unparalleled teaching tool greatly expanded the educational role of NICP. Following the publication of our fourth edition, in collaboration with Karl Misulis, we launched the *Review Manual for Neurology in Clinical Practice*, a book of questions and answers intended as an introduction to board examinations.

Butterworth-Heinemann completely revamped the fifth edition of NICP, published in 2008. It was printed in color with completely redrawn figures to bring it into line with standard textbook format. Again, with rigorous editing we incorporated much new material and removed out-of-date work. Despite the major explosion of knowledge in the clinical and basic neurosciences in the previous 17 years, the NICP fifth edition had expanded to only 2488 pages.

In the 22 years since the first publication of NICP, it has become the major international textbook of neurology and been translated into Spanish, Italian, Polish, and Turkish. When making academic visits to medical centers in other countries, I have found myself lauded as an editor of "the bible, *Neurology in Clinical Practice*." I know that Bob Daroff, Gerry Fenichel, and Joe Jankovic have had the same experience.

When I stepped down as chair of the Neurology Department at the University of Miami in 2007, I decided it was time to move on to other interests and retire from the editorship

of NICP. It had been an exciting and satisfying 20 years, and editing each new edition provided me personally with a complete neurological update course. For this, the sixth edition of NICP, my editorial colleagues and the publishers have been fortunate to persuade John Mazziotta to take my place. He brings a wealth of knowledge about the expanding field of functional imaging of the nervous system. The NICP sixth edition retains the structure of the textbook that was conceptualized nearly 40 years ago in Newcastle, but the clinical and scientific contents remain ever new. I have no doubt of the continuing success of our textbook and wish it well.

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# Preface

Neurology in Clinical Practice is a practical textbook of neurology that covers all the clinical neurosciences and provides not only a description of neurological diseases and their pathophysiology but also a practical approach to their diagnosis and management. In the preface to the 1991 first edition of this book, we forecasted that major technological and research advances would soon reveal the underlying cause and potential treatment of an ever-increasing number of neurological diseases.

The 20 years that have passed since that prediction have been filled with the excitement of new discoveries resulting from the blossoming of neurosciences. Clinical neuroscience has taken on the important and challenging problems of neuroprotection in both neurodegenerative disorders and acute injuries to the nervous system, such as stroke, multiple sclerosis, and trauma. In line with this effort, basic science progress in areas of neuroplasticity and neural repair are yielding important results that should translate into clinical utility in the near future, Advances in the genetics of neurological diseases have not only facilitated genetic testing but also provided important insights into the pathogenesis of diseases and helped identify potential therapeutic targets. Significant advances have taken place in the management of patients with both ischemic and hemorrhagic stroke. When the first edition of this textbook was published, there was essentially no effective means of treating acute ischemic stroke. Today we have numerous opportunities to help such patients, and a campaign has begun to educate the general public about the urgency of seeking treatment when stroke symptoms

The advent of teleneurology is also beginning to provide treatment for patients who lack access to neurological specialists or whose problems are too complicated for routine management in the community. Teleneurology consults are beginning to be provided nationwide across all subspecialties of our discipline, with a particular emphasis on patients who need intraoperative monitoring, critical care neurology, and stroke interventions.

To the benefit of patients, clinical neuroscience has partnered with engineering. Neuromodulation has become an important part of clinical therapy for patients with movement disorders and has applications in pain management and seizure control. Along these same lines, brain-controlled devices will soon help provide assistance to individuals whose mobility or communication skills are compromised. Recent advances in optogenetics have led to development of techniques that allow exploration and manipulation of neural circuitry, which may have therapeutic applications in a variety of neurologic disorders.

Finally, a search for biomarkers that reliably identify a preclinical state and track progression of disease is a promising goal in many neurodegenerative disorders.

Neurodegenerative disease, Alzheimer disease (AD) in particular, continues to be a worldwide crisis. The financial aspects associated with AD alone are staggering and have the capacity to bankrupt the modern world. For example, if no treatment or means to delay AD is found by 2050, the annual cost of care for such patients in the United States will exceed \$1 trillion, and the 40-year interval aggregate cost will exceed \$20 trillion. The costs in terms of suffering and hardship for patients and their families is too immense to quantify. As such, there is an urgent need for basic and clinical neuroscience to make progress in finding ways to delay the onset of neurodegenerative disorders and, ultimately, prevent them.

There is evidence of some startling new advances in neuroscience that are only just being considered today. The engineering of nanotechnologies into strategies to treat patients with neurological disorders is just beginning. One can envision a future that includes smart nanoimaging agents, nanopumps that can help regulate deranged circuitry on a local basis, and nanostimulators to participate in the growing field of neuromodulation. In addition, other partnerships with nanoengineers will produce sensors that can monitor not only the external condition of a patient by tracking movements, vital signs, and sleep behaviors but also internal states when such sensors are developed on a nano scale.

We still have a long way to go to reach the ultimate goal of being able to understand and treat all neurological diseases. Neurology remains an intellectually exciting discipline, both because of the complexity of the nervous system and because of the insight that the pathophysiology of neurological disease provides into the workings of the brain and mind. Accordingly, we offer the sixth edition of *Neurology in Clinical Practice* as the updated comprehensive and most authoritative presentation of both the art and the science of neurology.

For this edition, the text has been completely rewritten, and almost a fifth of the chapters have been prepared by authors new to the cadre of contributors. The layout of the pages has been completely redesigned to provide a user-friendly environment for accessing the material. The companion website, www.expertconsult.com, has been refined and expanded and includes video and audio material, additional illustrations and references, and chapters on key related material from other established neurology texts. It also is regularly updated with minireviews of important new publications in the neurological literature.

A work of this breadth would not have been possible without the contributions of many colleagues throughout the world. We are deeply grateful to them for their selfless devotion to neurological education. We are also grateful to our Elsevier counterparts, Lotta Kryhl, content strategist, and Lucia Gunzel, content development manager, who were

key in drawing this project together. Additionally, we thank Cindy Thoms, project manager, without whose energy and efficiency the high quality of production and rapidity of publication of this work would not have been achieved. Finally, we gratefully acknowledge the contributions of our readers, whose feedback regarding *Neurology in Clinical* 

Practice and the website has been invaluable in enhancing our educational goals.

Robert B. Daroff, MD Gerald M. Fenichel, MD Joseph Jankovic, MD John C. Mazziotta, MD, PhD

# Video Contents

32A	Electrod 32A-1	cephalography and Evoked Potentials Seizure 1		32B-27	Complex Repetitive Discharge—Abruptly Stopping
	32A-2	Seizure 2		32B-28	
	32A-3	Seizure 3			Change in Configuration
	Clinical Electromyography 32B-1 End-Plate Noise			32B-29	Complex Repetitive Discharge—Multiple Quantal Jumps
	32B-2	End-Plate Spikes		32B-30	3
	32B-3	Fibrillation Potential			Complex
	32B-4	Positive Sharp Wave		32B-31	3-
		Fibrillation Potential Changing to a Positive Sharp Wave with Needle			Cramp Discharge
				32B-33	
		Movement		32B-34	The state of the s
	32B-6	Fibrillations / Positive Sharp Waves 1+		32B-35	Polyphasic
	32B-7	Fibrillations / Positive Sharp Waves 2+		32B-36	Early Reinnervation
	32B-8	Fibrillations / Positive Sharp Waves 3+		32B-37	
	32B-9	Fibrillations / Positive Sharp Waves 4+			Polyphasic
	32B-10	Fasciculation Potential		32B-38	
	32B-11	2B-11 Fasciculation Potential—Reinnervated		32B-39	and Increased Amplitude Slightly Decreased Recruitment
		Potential 12 Fasciculation Potential—Very Slow Frequency			Moderately Decreased Recruitment
	32B-12			32B-40	
	32B-13	Fasciculation Potential—Multiple Potentials		32B-42	
	32B-14	Myotonic Discharges		32B-43	Satellite Potentials
		Myotonic Discharges and Positive Sharp		32B-44	Two Satellite Potentials
		Waves		32B-45	
		Myokymic Discharge		32B-46	Satellite Potential with Intermittent
	32B-17	Myokymic Discharge—Two Overlapping Discharges		32B-40	Blocking
	32B-18	Discharge		32B-47	
	adamer.			32B-48	Polyphasic and Unstable
	32B-19	2B-19 Myokymic Discharges, Doublets, and Multiplets		32B-49	Nascent Units
	32B-20	Myokymic Discharges and Fasciculation Potentials		32B-50	Brief Duration, Short Amplitude, and Polyphasic Motor Unit Action Potentials
	32B-21			32B-51	Poor Activation
		Myokymic Discharges—Multiple Discharges	35	Neuro-o 35-1	phthalmology: Ocular Motor System Internuclear Opthalmoplegia
	32B-22	Myotonia/Myokymia—Translational Waveform		35-2	Gaze-Evoked Nystagmus
	32B-23	Rest Tremor		35-3	Forced Ductions
		Doublets	37	Neuro-otology: Diagnosis and Management of Neuro-otological Disorders	
	32B-25	Doublets, Triplets, and Singlets		37-1	Acute Peripheral Vestibular Nystagmus
	32B-26	Complex Repetitive Discharge		37-2	Ocular Flutter

- 37-3 Gaze-Evoked Nystagmus and Imparied Smooth Pursuit 37-4 Gaze-Evoked Downbeating Nystagmus 37-5 Hypermetric Saccades 37-6 **Head-Thrust Tests** 37-7 Benign Paroxysmal Positional Vertigo 37-8 **Epley Maneuver** 70 Cranial Neuropathies 70-1 Large Left Hypertropia Secondary to Right Oculomotor Nerve Palsy 70-2 Left Appendicular Ataxia 70-3 **Prominent Left Ptosis** 70-4 Impaired Adduction, Elevation, and Depression with Intact Abduction of the Left Nerve 70-5 Bilateral Abduction Deficits Secondary
- History of Right Facial Palsy 2 Years ago and Current Left Facial Palsy

  74 Disorders of the Upper and Lower Motor Neurons 74-1 Patient 2 Showing Fasciculations in

to Demyelinating Bilateral Abducens

Facial Nerve Function in a Patient with a

74-2 Patient 3 (Babinski Sign)

Quadriceps

- 74-3 Walking Showing Footdrop Because of Dorsifelxor Weakness
- Supp Video 1 Parkinson's Dis

**Palsies** 

Esotropia

70-6

70-7

- Supp Video 1 Parkinson's Disease: Marked Flexion of the Trunk (Camptorcormia Because of PD-Related Skeletal Deformity
- Supp Video 2 Parkinson's Disease: Marked Neck and Trunk Flexion (Camptocormia) Due to PD-Related Dystonic Neck and Trunk Flexion, Improved after Botulinum Toxin Injections into Rectus Abdominus Bilaterally
- Supp Video 3 Parkinson's Disease; Striatal Hand Deformity
- Supp Video 4 Parkinson's Disease; Patient with Young-Onset Parkinson's Disease and Gait Difficulty Due to Freezing (Motor Blocks)
- Supp Video 5 Parkinson's Disease; Patient
  Describes Levodopa-Induced Visual
  Hallucinations (e.g., Seeing and Picking
  Worms)
- Supp Video 6 Parkinson's Disease; Levodopa-Induced Dyskinesia
- Supp Video 7 Progressive Supranuclear Palsy;
  Typical Worried, Frowning Facial
  Expression (Procerus Sign), Apraxia of
  Eyelid Opening, Although Vertical
  (Downward) Gaze is Preserved, Vertical
  Optokinetic Nystagmus is Absent, When
  Walking Patient Pivots on Turning

- (in Contrast to Patients with Parkinson's Disease Who Turn En Bloc)
- Supp Video 8 Progressive Supranuclear Palsy; Marked Vertical Ophthalmoparesis, Perseveration of Gaze to Left even though the Body Faces Forward
- Supp Video 9 Progressive Supranuclear Palsy;
  Typical Facial Expression with Deep Facial
  Folds, Square Wave Jerks on Primary
  Gaze, Slow Saccades, Inappropriate
  Laughter (Pseudobulbar Palsy), Right
  Arm Levitation
- Supp Video 10 Progressive Supranuclear Palsy;
  Deep Facial Folds, Vertical
  Ophthalmoplegia, Marked Postural
  Instability, Slumps into a Chair
- Supp Video 11 Progressive Supranuclear Palsy;
  Deep Facial Folds, Apraxia of Eyelid
  Opening, in Addition to Vertical
  Ophthalmopareses, Patient Demonstrates
  Evidence of Internuclear Ophthalmoplegia,
  the Presence of Right Arm Tremor (Atypical
  for Progressive Supranuclear Palsy)
  Suggests the Co-Existence of Parkinson's
  Disease
- Supp Video 12 Multiple System Atrophy; Patient Describes Symptoms of Dysautonomia, Demonstrates Flexion of the Neck and Apraxia of Eyelid Opening, Typical of MSA
- Supp Video 13 Corticobasal Degeneration; Patient Describes Apraxia of Left Leg, Demonstrates Ideomotor Apraxia in Left More than Right Hand and Marked Left Leg and Foot Apraxia
- Supp Video 14 Corticobasal Degeneration; Patient
  Describes Alien Hand Phenomenon in the
  Right Arm, Demonstrates Marked Apraxia
  in the Right More than Left Hand,
  Spontaneous and Evoked Myoclonus in
  the Right Hand, Markedly Impaired
  Graphesthesia
- Supp Video 15 Corticobasal Degeneration; Evoked Hand and Arm Myoclonus
- Supp Video 16 Corticobasal Degeneration;
  Patient Describes Right Alien Hand
  Phenomenon, Right Hand Myoclonus,
  Marked Ideomotor Apraxia in the Right
  More than Left Hand
- Supp Video 17 Vascular Parkinsonism; Broad-Based Gait, Freezing on Turning (Lower Body Parkinsonism) Associated with Binswanger's Disease (See Head MRI)
- Supp Video 18 Vascular Parkinsonism; Gait Initiation Failure (Pure Freezing)
- Supp Video 19 Essential Tremor; Marked Improvement in Right Hand Tremor with Contralateral Deep Brain Stimulation of the VIM Thalamus