Neurological problems commonly occur in the context of an underlying systemic disease, and these neurologic presentations are a frequent source of inpatient and outpatient neurological consultation. In many patients, the neurological disorder is a manifestation of a previously diagnosed systemic illness or its treatment, but in still many others the neurological disorder is the presenting manifestation of a medical condition that has not yet been diagnosed. The aim of this book is to provide the physician with an overview of the clinical presentation, pathophysiology, diagnosis, and treatment of the various neurological syndromes that occur in the setting of systemic illnesses.

In this book, “systemic disease” and “medical disease” are used interchangeably, and refer to the kind of disease or syndrome in which the primary dysfunction involves an organ or system other than the nervous system, with the nervous system disorder occurring as a secondary—though in many cases, a potentially major—consequence. This book, therefore, focuses mostly on the neurological illnesses that occur in the setting of those primary illnesses that are typically considered to be under the purview of general internal medicine or its subspecialties. In addition, this book discusses the neurological complications that occur due to medications and other therapies typically used to treat these systemic illnesses. Conversely, this book does not focus on diseases—such as many genetic disorders—with multisystem manifestations that include both neurological and systemic complications, but where the neurological disease is not considered a complication of the systemic disease.

Unlike most books on this subject, the chapters of this book are organized and defined by neurological clinical scenarios, rather than by medical diseases. Specifically, each chapter focuses on a particular category of neurological presentation (e.g. movement disorders) and discusses the various systemic illnesses, or their treatment, that can cause dysfunction within that category of neurologic disorders. This organizational scheme, I propose, especially parallels the very common clinical scenario where the medical illness underlying the neurological syndrome is unknown; in these scenarios, the clinician needs to have some knowledge and understanding of the various systemic illnesses that can lead to these neurologic presentations.

The following major neurologic presentations define the chapters of this book: headache, encephalopathy, dementia, stroke, seizures, neuroophthalmologic disorders, neurootologic disorders, movement disorders, spinal cord disorders, peripheral nerve disorders, neuromuscular junction disorders, disorders of skeletal muscle, autonomic nervous system disorders, and sleep disorders. Each chapter, in turn, is subdivided into major categories of systemic illness that can lead to neurologic dysfunction: endocrine disorders; electrolyte and other metabolic disorders; systemic autoimmune disorders; organ dysfunction and failure; systemic cancer and paraneoplastic disorders; systemic infectious diseases; complications due to transplantation, complications of critical medical illness; drugs, alcohol, and toxins; and vitamin and mineral deficiencies. In individual chapters, some of these subheadings are excluded when they are not particularly relevant to that chapter’s neurologic topic.

The book begins with the chapter on headache (Chapter 2) by Kevin Kahn, MD, from the Carolina Headache Institute, who discusses secondary headache syndromes that can be associated with systemic
disease, as well as the interface between systemic illness and primary headache syndromes. Chapter 3, written by Allison Weathers, MD, from Rush University Medical Center provides an overview of the diffuse encephalopathy (delirium) syndromes that (by definition, arguably) occur within the setting of systemic dysfunction. In contrast, in Chapter 4, Jennifer Molano, MD, and Brendan Kelley, MD, from the University of Cincinnati tackle the interaction of systemic dysfunction and neurologic syndromes that more resemble dementia than typical toxic-metabolic encephalopathies; these authors also provide additional insights into the interface between the primary degenerative dementias and systemic illnesses.

In Chapter 5, Sarkis Morales-Vidal, MD, and José Biller, MD, from Loyola University Medical Center review the many systemic disorders that can be associated with, and potentially cause, cerebrovascular disease and stroke that the clinician should keep in mind in addition to the usual and well-known medical stroke risk factors. In Chapter 6, Matthew Hoerth, MD, and Joseph I. Sirven, MD, from the Mayo Clinic, Scottsdale, discuss the many medical problems that can lead to seizures; typically, recognition of these systemic causes of seizures can avoid unnecessary, or prolonged, antiepileptic drug therapy in these patients.

In Chapter 7, Matthew Thurtell, MBBS, from the University of Iowa and Janet Rucker, MD, from the Mount Sinai School of Medicine review and illustrate the many neuroophthalmological signs and symptoms that occur due to, and give clue to, the presence of an underlying potentially serious and sometimes vision-threatening systemic illness. In Chapter 8, Terry Fife, MD, from the Barrow Neurological Institute in Arizona discusses the many—and probably underrecognized by many neurologists—auditory or vestibular neurootologic syndromes that can occur due to medical illness.

In Chapter 9, Brandon Barton, MD, and Christopher Goetz, MD, from Rush University Medical Center review the many movement disorders (including parkinsonism, dystonia, tremor, chorea, myoclonus, ataxia, and tics) that can occur due to systemic disease or its treatment. In Chapter 10, Sital Patel, MD, and I, also from Rush, discuss myelopathies (whether from extrinsic compression of the spinal cord or intrinsic noncompressive spinal cord dysfunction) that can occur as a complication of an underlying medical disorder.

In Chapter 11, Michelle Mauermann, MD, from the Mayo Clinic Rochester and Ted Burns, MD, from the University of Virginia review the many neuropathic syndromes, and their characteristic clinical patterns, that can occur due to systemic disorders. Extending the discussion further, in Chapter 12, Jaffar Khan, MD, from the Emory University School of Medicine discusses presynaptic and postsynaptic neuromuscular junction disorders and their association with underlying systemic illness. In Chapter 13, Hannah Briemberg, MD, FRCP, from the University of British Columbia reviews the many myopathic disorders that can occur as a consequence of medical illness and certain medications.

In Chapter 14, Brent Goodman, MD, and Eduardo Benarroch, MD, from the Mayo Clinic, Rochester, review autonomic nervous system manifestations that can occur—with or without other signs of neurologic dysfunction—in the setting of systemic disease; the authors also review how to assess for these autonomic disorders. Finally, in Chapter 15, Erik St. Louis from the Mayo Clinic, Rochester, discusses the association of disorders of sleep, including the parasomnias, and underlying systemic illness.

Each chapter concludes with a list of the authors’ suggestion of “Five things to remember about” that particular neurologic topic and its relation to systemic disease; these can be construed as suggested minimum “take home” points that provide some additional overall clinical perspective for the reader.

Although written primarily with the neurologist (generalist neurologist, subspecialist neurologist, or neurologic trainee) in mind, the material in this book should also be of interest and accessible to internal medicine physicians, other primary care providers, internal medicine subspecialists, and even interested medical students. It is my hope that the reader of this text will find that the unique neurologic syndrome-based approach in the following pages will provide clinically useful insight into the wide variety of neurological disorders that occur in the context of systemic disease, and provide practical clinical clues to both the neurological diagnosis and the underlying medical diagnosis and management of these patients.