Status epilepticus is a wonderful field of study and of clinical work. Its tremendously varied presentations offer insights into the workings of the human brain. Basic science and clinical studies of generalized convulsive status alone have taught us enormous amounts about brain processes, from cellular function to neuronal morphologic changes and cell death. The electrophysiology of status in both experimental and clinical cases is instructive about neuronal connections and helps to explain brain function in illness and in health.

Clinically, status epilepticus is worthy of intensive study. One of the primary values of the neurologist to his or her patients is the wise application of specialized knowledge and powers of observation in making accurate diagnoses of bizarre or baffling behavior that does not necessarily appear epileptic to others—or appears epileptic but is not. Focus on accurate diagnosis helps the neurologist to initiate appropriate and potentially beneficial treatment for serious illness.

Status epilepticus was recognized in antiquity but only became the subject of medical writings in the late nineteenth century and of informative laboratory studies in the last 50 years. The existence of nonconvulsive status was posited by Charcot but only became clearly diagnosable after Berger developed the EEG in the mid twentieth century. In Shorvon’s 1994 monograph, Status Epilepticus: Its Clinical Features and Treatment in Children and Adults, he lamented the fact that there were just 370 publications related to status in his review of a large database through 1978. Now, there are several hundred helpful papers on status every year.

The great clinical neurophysiologist Niedermeyer said of epilepsy, “There is no disease named ‘epilepsy.’” Rather, there are very many illnesses that cause epileptic seizures. Similarly, over the last few decades, neurologists have learned to ask “Which type of status?” when asked how to treat it. This book emphasizes the recognition and diagnosis of the very many different forms of status epilepticus and the necessarily different evaluation, management, and treatment of each. Here, those types are organized into status with convulsions or major abnormal movements (Chaps. 7–18) and those considered nonconvulsive (Chaps. 19–25).

There has been a dramatic increase in useful knowledge about the treatment of convulsive status since the previous edition of this book, discussed in several chapters (especially Chaps. 16–18, and 29). There has been a similar growth in recognition of the remarkably varied forms of nonconvulsive status, the huge range of underlying illnesses that can cause both convulsive and nonconvulsive status (e.g. several autoantibody syndromes), and also of entirely new and different types of treatment, whether dietary, new drugs, or stimulation. The field has become much more complicated, but better understanding of these complex illnesses and new insights and approaches to them may help to achieve the goal of better treatment for patients with this often very threatening illness.

Of necessity, discussion of these many types of status epilepticus and many old and new treatments will occasion some overlap or duplication among chapters. Those on status in the very young (Chaps. 26–28) offer an illustration. One focuses on the youngest patients (neonates), another on unusual status syndromes in children, and the third on the most difficult cases of refractory status in pediatric ICUs. The chapters overlap, particularly when discussing
medications, but it should be apparent that these are different areas of expertise. In all chapters, different approaches, controversies, and speculation were not proscribed, but rather encouraged. Indeed, different perspectives and opinions are worth considering, as many can be educational. Different chapters may offer different approaches to the same problem, but in the end, we are confident that most of our authors would have very similar approaches to similar cases, and also that seeing different styles and then making an independent decision is a good way of refining one’s own approach to the management of status epilepticus.

While this volume focuses on a clinical perspective, all neurologists confronting status epilepticus will want to understand better its underlying biology and pathophysiology. Information on basic studies appears in many chapters, but especially in those dealing with the clinical and pathologic consequences of generalized convulsive status on the one hand and nonconvulsive status on the other (Chaps. 9, 10, and 25).

Over the last decade, the booming practice of continuous EEG monitoring, especially in critically ill patients, has changed the field, with better identification of status, while simultaneously generating controversies about what is and what is not status epilepticus (see Chaps. 19 and 23).

One of the advantages and pleasures of working in a field of academic Neurology over decades is the opportunity to meet and work with individuals in the enlarging international community of scholars interested in status epilepticus, many of whom have given generously of their time and wisdom to this volume. We owe a tremendous debt of gratitude to our many wonderful co-authors on this project—whom we believe to be among the finest neurologists, epileptologists, and clinical neurophysiologists in the world. The range of their expertise and ability to inform us is impressive, and the range of their backgrounds similarly diverse, with their homes in at least a dozen countries. Much of what we know about status epilepticus has come from reading their papers, listening to their lectures, and discussing status epilepticus at national and international meetings or other conferences, and in the hallways and conference rooms of our home institutions, or theirs. In reviewing their contributions here, we have enjoyed learning even more.

It is important and a pleasure to thank our editors at Springer, Mr. Greg Sutorius, senior editor, for initiating the 2nd edition and helping to shepherd it through, and developmental editor, Ms. Katherine Kreilkamp, for offering help with far more than customary production editing—everything from assistance with permissions to guidance on formatting and even syntax, with mastery of the process, gracious author communication and encouragement to finish, and shockingly rapid responses to our queries.

Some of our best experiences in Medicine have been in helping get an individual patient through the threatening illness of status epilepticus, and especially in the most refractory cases. We remember the successes gladly, and we remember the failures very sadly, and we hope that the wisdom of our co-authors will help increase the number of successes for our readers and their patients.

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