Status Epilepticus
CURRENT CLINICAL NEUROLOGY

Daniel Tarsy, MD, SERIES EDITOR

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Dedication

To my parents, Betty Kay and Edward Drislane, who gave me all I could want to start life, and to Rose, Catherine, Helen, and Edward, who help to sustain its enjoyment.

—Frank W. Drislane, MD
Series Editor’s Introduction

There can be little doubt that status epilepticus is one of the most serious and challenging acute nervous system disorders to face the neurologic clinician. The mortality and morbidity of this disorder is high and the urgency of swift response is critical. Chapters describing its diagnosis and treatment have appeared in four previous volumes in the Current Clinical Neurology series including Seizures: Medical Causes and Management by Delanty, Critical Care Neurology and Neurosurgery by Suarez, Handbook of Neurocritical Care by Bhardwaj, Mirski, and Ulatowski, and Seizures in Critical Care by Varelas. The discussion of status epilepticus has now been greatly expanded in Status Epilepticus: A Clinical Perspective by Frank Drislane. The explosion of new information concerning the definition, classification, pathophysiology, epidemiology, electroencephalography, pathologic consequences, and treatment of “status” justifies this very impressive monograph on the subject. Given the urgency of prompt treatment of this potentially devastating disorder and the inherent difficulty of collecting reliable and useful clinical data, it is actually remarkable how much information has accumulated on the subject over the last several decades.

This is largely a clinical volume and a proper approach to the subject therefore begins with an all-inclusive classification. Whereas generalized convulsive and focal status epilepticus are familiar and have long been recognized, nonconvulsive status epilepticus is a more recent arrival on the scene that is all too often misdiagnosed and must first be appreciated in order to be properly recognized and treated. This very important topic is strongly emphasized in this volume with several chapters devoted to classification, differential diagnosis, descriptions of seizure “imitators,” electroencephalographic patterns, clinical presentations, unusual behavioral and cognitive manifestations, and treatment as it specifically applies to nonconvulsive status epilepticus. Because “seizures beget seizures” the urgency of prompt diagnosis and treatment is obvious. This begins with the identification and management of underlying structural and metabolic causes and continues with the broad knowledge of the array of anticonvulsant treatments currently available for treatment of this disorder in all of its manifestations.

In recent years clinical neurology has evolved from a specialty emphasizing classical description and phenomenology to one that includes the management of the critically ill patient. There is no better example of this than the management of status epilepticus. In this volume, Dr. Drislane has brought together an impressive array of experts in the field that will serve to advance the state of the art of this critically important discipline.

Daniel Tarsy, MD
Status epilepticus is a wonderful field of study and of clinical activity. Its fantastically 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 clinical and experimentally induced cases is instructive about neuronal connections and helps to explain brain function in pathologic conditions and in health. The many other forms of status epilepticus also illustrate brain mechanisms in widely varying ways.

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 to combat serious illness.

The study of status epilepticus is undergoing exponential growth. It was recognized in antiquity, but only became the subject of medical writings in the late 19th century and of scientific laboratory studies just more than 30 years ago. The existence of nonconvulsive status was likely deduced by Charcot, but it only became clearly diagnosable after Berger developed the EEG in the mid 20th century.

It has been just a decade since the publication of Shorvon’s monograph, *Status Epilepticus: Its Clinical Features and Treatment in Children and Adults*. Professor Shorvon lamented the fact that there were just 370 publications related to status in his review of a large database through 1978. Before his book, there was just the colloquium from Drs. Delgado-Escueta, Wasterlain, Treiman, and Porter (*Advances in Neurology Volume 34: Status Epilepticus: Mechanisms of Brain Damage and Treatment*) that served as a text for more than ten years. In the past decade, however, studies and writing on this subject have exploded, with hundreds of papers each year now. Consequently, over a thousand references are cited in this text.

*Status Epilepticus: A Clinical Perspective* attempts to bring together developments in the study of status epilepticus through 2004. It cannot cover all areas of investigation (especially the basic scientific) and stresses a clinical perspective. The book is organized along the lines of different forms of status epilepticus as encountered by the clinician “in the field.” The underlying genetic, biological, and developmental background, as well as the precipitating factors that lead to an episode of status, are discussed within each of these areas. One sees a similarity but also a difference from one clinical form of status epilepticus to another, demonstrating that status is not a single disease, but rather arises from different substrates and comprises multifaceted illnesses.
Any understanding of status epilepticus requires its placement in a larger context, including its history, its differentiation from other neurologic and psychiatric illnesses, and its statistical delineation by epidemiologists. My gratitude goes to Drs. Kaplan, Dworetzky and Bromfield, and Waterhouse for their own perspectives tying the study of status epilepticus to the larger world.

Although *Status Epilepticus: A Clinical Perspective* focuses on the clinical presentations, diagnosis, and management of the many different forms of status epilepticus, all epileptologists are curious about its underlying biology. A better understanding of those topics is worthwhile for its own sake. We also hope and expect that treatment of status will be much better in a few decades; it will be the scientific discoveries in the field that make this come to pass. The chapter on cellular physiology and processes occurring during status epilepticus by Drs. Hope and Blumenfeld and that on the cellular damage and neuropathology of status by Dr. Fountain represent integral parts of the modern clinician’s understanding of status, even when all of one’s workday experience is in the clinical realm. References to pathophyslogic processes are also essential parts of other chapters.

Convulsive status epilepticus, that with uncontrolled and usually rhythmic and often violent movements, is recognized readily by most physicians and indeed probably by most citizens. The chapter on generalized status by Dr. Chang and that on the remarkably varied forms of focal status by Dr. Schomer show that this is a rich, complex field of study. These illnesses can be devastating, and even the treatment can be harmful, so a better understanding of their presentations and management is important for patients. If convulsive status (whether generalized or focal) has varied presentations, nonconvulsive status epilepticus might make the earlier categories appear relatively simple. Drs. Kaplan and Benatar show the extensive overlap of epilepsy and behavioral neurology in their chapters on the presentation of different forms of nonconvulsive status.

A book on status epilepticus without extensive reproductions and descriptions of EEG correlates would be nearly hemianopic. In her chapter, Dr. Herman offers a comprehensive view of the EEG and its use in status with striking illustrations that I believe seasoned electroencephalographers will find to be the classic examples one can keep in mind when reviewing less clear-cut or dramatic EEGs from patients who are in status epilepticus—or are thought to be in (or to have been in) status. Her chapter could serve as an atlas on the topic.

Treatment of status epilepticus is sometimes easier than diagnosis, but we and the patients are not always so fortunate. Happily, one of the major explosive areas of growth in publication and knowledge on status over the past decade has been on its management. Drs. Shih and Bazil review the treatment of generalized convulsive status epilepticus by the meticulous detailing of careful clinical studies. Traditions, myths and examples of dogma abound in this area. This chapter allows one to state what is actually known with any certainty. Fortunately (for the patients, if not for the up-to-date nature of this book) these studies will clearly have many companions soon. More will also be learned about the treatment of refractory status epilepticus, but at this point, the chapter by Drs. Smith and Bleck is as current as
possible and from authors with as much experience as an editor can find. Studies on
the treatment of nonconvulsive status have been far less rigorous, but I have
attempted to summarize them.

Status epilepticus in very young children and neonates often appears to those of
us in adult neurology to be a totally separate and mysterious illness. Drs. Riviello
and Gaitanis bring a coherent perspective to the field while describing tremendously
different syndromes and problems. Their review of the varied presentations and
their extensive clinical experience should serve as useful guides to evaluation of
children with status; treatment is discussed in detail. Differences from adult neurol-
ogy in approach to children and neonates are emphasized.

There are obvious areas of this book to which some readers could fairly object—
including overlapping material among the chapters and the lack of a completely
consistent dogma in both diagnosis and treatment. The overlap is hard to escape
when individual clinical problems are difficult to fit into occasionally arbitrary cat-
egories. I hope and believe that the overlap is acceptable, particularly noting that
each author has slightly different perspectives on the same questions, and those
different perspectives can enrich one’s appreciation for the large world of status
epilepticus.

Similarly, with the many authors’ clinical expertise, it can be helpful to see the
same problem or question addressed from different viewpoints and with somewhat
different approaches. I suspect that nearly every author would tackle an individual
patient’s clinical problem in a very similar way, but often there is no one right
answer and nothing unusual about finding partially conflicting recommendations
for an individual situation. In 2004, there are still debates about definitions, diag-
noses, and treatments of different forms of status epilepticus. Controversy and
speculation were not proscribed, but rather encouraged. Different opinions persist
and are reasonable. The authors have varied insights and styles. These multiple
approaches constitute not so much inconsistency, as different perspectives based
on rich experience, and it is hoped that those variations will be educational and
possibly even stimulate new and better studies.

What I believe to be the high quality of the syndrome descriptions, clinical
insights, and guidance on treatment is owed to my colleagues and coauthors. This is
largely a group of academic neurologists and epileptologists from the Northeastern
United States (which includes Virginia if one looks topographically rather than
historically). Much wonderful work on status epilepticus comes from California,
other parts of the United States, Britain, France, and many other lands around the
world. This is a field that has evoked, by its intrinsic interest and clinical urgency,
a productive collegial international cooperation that is both enjoyable and benefi-
cial. The work of others is referred to extensively, but the authors were solicited
from personal contacts through the world of neurology and epilepsy and chosen for
their academic achievements and because I knew that they could not fail to write
outstanding, clinically oriented chapters based on both careful study of the litera-
ture and rich personal clinical experience. I have enjoyed learning from their writ-
ings—and am confident that I will not be the last to do so.
It is a pleasure to thank Ms. Nicole Furia and her colleagues at Humana Press for their patience and encouragement and the invitation to organize and write this book. All the authors are clearly indebted to our patients, residents, and colleagues who share and collaborate in our clinical experience that has led to our learning about status epilepticus. They have helped to clarify our thinking. We hope that Status Epilepticus: A Clinical Perspective will help kindle an interest in status epilepticus in our younger colleagues and an interest in investigating it further. We believe that a better understanding of basic and clinical science is not only enjoyable, but will also help the many patients who will have status epilepticus in the future.

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Contributors

CARL W. BAZIL, MD, PhD • Department of Neurology, Columbia Comprehensive Epilepsy Center, Columbia University, New York, NY
MICHAEL BENATAR, MBChB, DPhil • Department of Neurology, Emory University School of Medicine, Atlanta, GA
THOMAS P. BLECK, MD, FCCM • Departments of Neurology, Neurological Surgery, and Internal Medicine, Neuroscience Intensive Care Unit, University of Virginia School of Medicine, Charlottesville, VA
HAL BLUMENFELD, MD, PhD • Department of Neurology, Yale University School of Medicine, New Haven, CT
EDWARD B. BROMFIELD, MD • Department of Neurology, Brigham and Women’s Hospital, Harvard Medical School, Boston, MA
BERNARD S. CHANG, MD • Comprehensive Epilepsy Center, Department of Neurology, Beth Israel-Deaconess Medical Center, Harvard Medical School, Boston, MA
FRANK W. DRISLANE, MD • Comprehensive Epilepsy Center and Department of Neurology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA
BARBARA A. DWORETZKY, MD • Department of Neurology, Brigham and Women’s Hospital, Harvard Medical School, Boston, MA
NATHAN B. FOUNTAIN, MD • Comprehensive Epilepsy Program, Department of Neurology, University of Virginia School of Medicine, Charlottesville, VA
JOHN N. GAITANIS, MD • Department of Neurology, Hasbro Children’s Hospital, Brown University, Providence, RI
SUSAN T. HERMAN, MD • Department of Neurology, University of Pennsylvania School of Medicine, Philadelphia, PA
OMOTOLA HOPE, MD • Department of Neurology, Yale University School of Medicine, New Haven, CT
PETER W. KAPLAN, MB, FRCP • Department of Neurology, Johns Hopkins Bayview Medical Center, Baltimore, MD
JAMES J. RIVIELLO, JR., MD • Division of Epilepsy and Clinical Neurophysiology, Critical Care Neurology Service, Department of Neurology, Children’s Hospital, Professor of Neurology, Harvard Medical School, Boston, MA
DONALD L. SCHOMER, MD • Clinical Neurophysiology Lab, Department of Neurology, Comprehensive Epilepsy Center, Beth Israel-Deaconess Medical Center, Harvard Medical School, Boston, MA
TINA SHIH, MD • Department of Neurology, Columbia Comprehensive Epilepsy Center, Columbia University, New York, NY
TERESA L. SMITH, MD • Departments of Neurosurgery and Neurology, University of Michigan, Ann Arbor, MI
ELIZABETH J. WATERHOUSE, MD • Department of Neurology, Virginia Commonwealth University School of Medicine, Richmond, VA