

Preface

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 pathophysiologic 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 neurology 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 categories. 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, diagnoses, 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 beneficial. 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 literature and rich personal clinical experience. I have enjoyed learning from their writings—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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