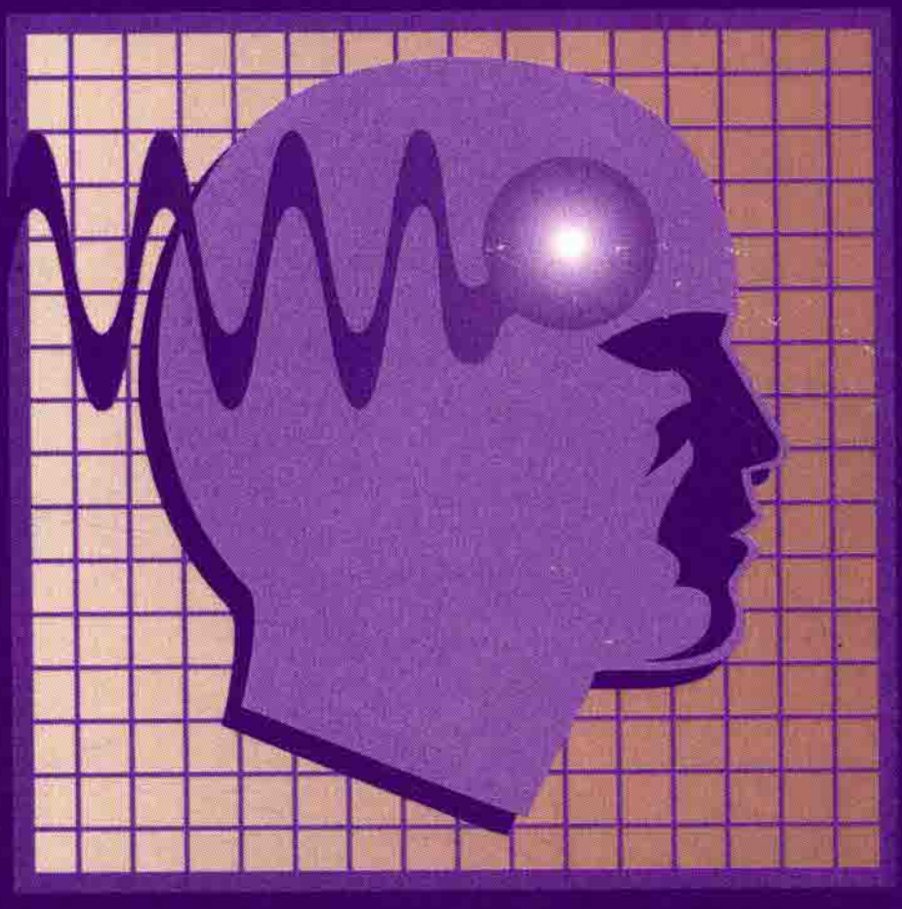
Contemporary Diagnosis and Management of the Patient With Epilepsy®



Ilo E. Leppik, MD

Fifth Edition

Contemporary Diagnosis and Management of the Patient With Epilepsy®

Fifth Edition 2000

By

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and

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Table of Contents

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This book has been prepared and is presented as a service to the medical community. The information provided reflects the knowledge, experience, and personal opinions of Ilo E. Leppik, MD, Director of Research, MINCEP Epilepsy Care, and Clinical Professor of Neurology and Pharmacy, University of Minnesota, Minneapolis, Minnesota.

This book is not intended to replace or to be used as a substitute for the complete prescribing information prepared by each manufacturer for each drug. Because of possible variations in drug indications, in dosage information, in newly described toxicities, and in other items of importance, reference to such complete prescribing information is definitely recommended before any of the drugs discussed are used or prescribed.

This Fifth Edition has been extensively revised, reflecting the most current clinical developments in seizures and epilepsy.

Acknowledgments

I would like to extend my thanks in preparing this book to Matthew T. Corso of Handbooks in Health Care Co., for his editing of the copy, and to my wife, Peggy, and our children, Peter, David, and Karina, for their patience and support.

Chapter 1

Introduction

"Epilepsy is an illness of various shapes and horrible."—Aretaeus[1]

Epilepsy and seizures are the most common serious neurologic symptoms, affecting all ages. A person experiencing a seizure and his or her family are often very frightened by seizures. Moreover, epilepsy has many socioeconomic consequences (loss of driving, social embarrassment, changes in employment). Thus, appropriate treatment must include educating the patient, counseling about emotional impact, and providing appropriate information to employers and regulatory agencies.

A critical concern to patients with epilepsy is the unpredictability of seizures; they may occur at work, while driving, or in social settings. This makes it difficult for even a person who has had only a few seizures to lead a life free of constant fear. Also, many persons are afraid of sustaining serious injury or dying during seizures, adding to their anxiety. A physician must not only prescribe medications, but must be aware of the total impact of this disorder on a person's life,

Table 1: Active Cases of Epilepsy (prevalence) per 1,000[2]

Country	Age-Adjusted Prevalence
USA	
• Rochester, Minnesota	6.0
• Copiah County, Mississippi	7.0
Iceland	3.7
England	4.4
Nigeria	5.0
India (Bombay)	3.6
Guam	6.1
Poland	8.1

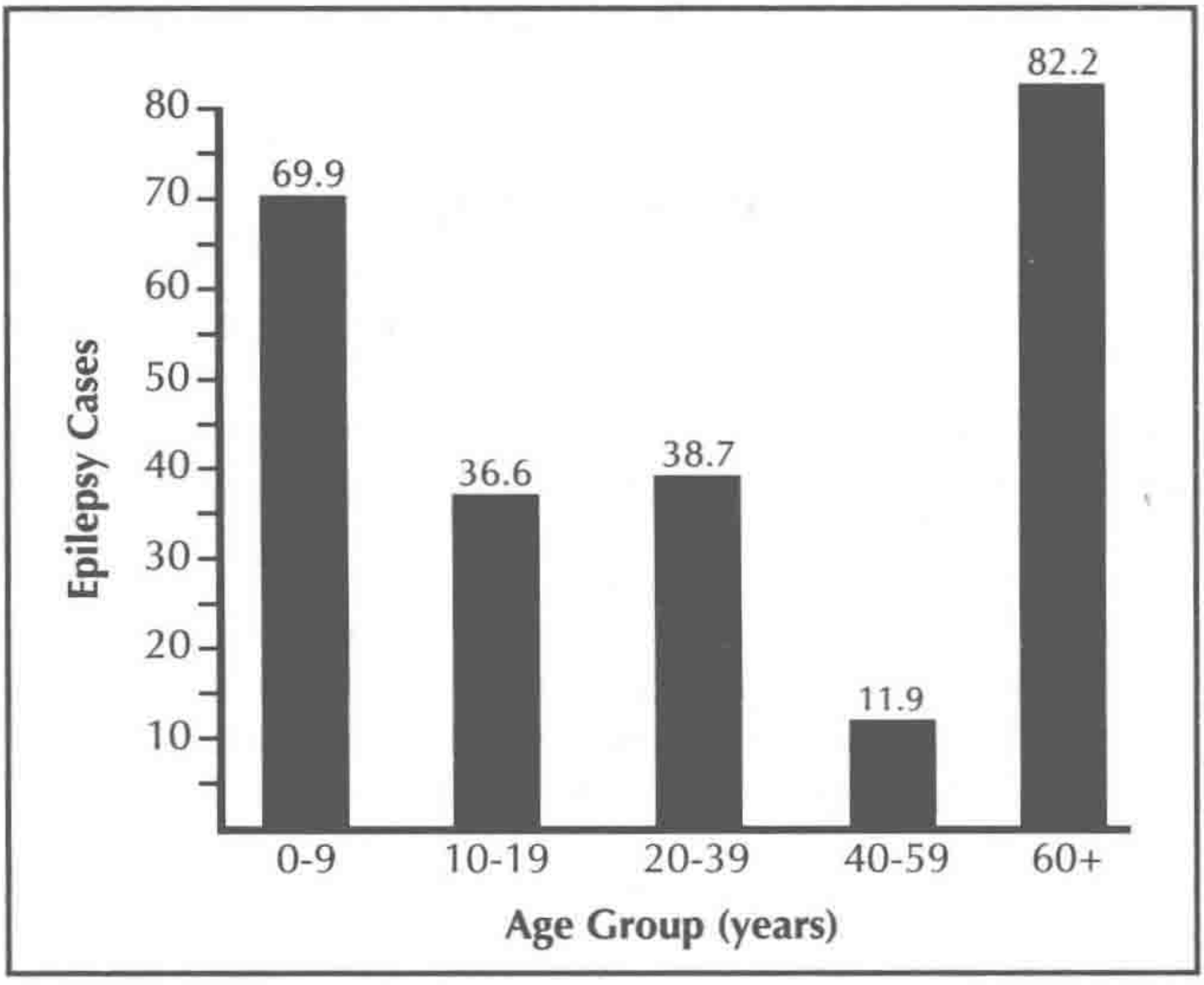

***Figure 1**—Histogram of average annual incidence of epilepsy by age; new cases per year per 100,000. Data from Rochester, Minnesota. Adapted from Hauser.[2]*

and facilitate use of appropriate community resources to help the person with epilepsy deal effectively with this condition. Epidemiologic studies have shown that the cumulative adjusted lifetime incidence of epilepsy in Rochester, Minnesota, is 3.1% by age 80.[2] In other words, if all persons lived to age 80 in that community, 31 in 1,000 will have or have had a diagnosis of epilepsy sometime in their lives. Seizures are even more common. The lifetime cumulative incidence of seizures approaches 11%, or 110 in 1,000.[2]

Thus, most persons who experience seizures do not have epilepsy. Epilepsy is a disorder of the central nervous system (CNS) whose symptoms are seizures, but many persons will have provoked seizures (febrile convulsions, alcohol withdrawal seizures, etc) without primary CNS pathology. The clinical manifestations of a seizure, whether in a person with epilepsy or provoked, are similar, making the evaluation and treatment of a person with a single seizure particularly challenging.

The number of *active cases* (prevalence) in any given year in most countries where valid prevalence statistics are available is 5 to 8 per 1,000 (or 0.5% to 0.8%). Preliminary statistics also show that epilepsy may be more common in some underdeveloped countries (Table 1).[2] The difference between the 3.1% by age 80 incidence and the 0.5%-to-0.8% range in any given year is explained by the fact that many patients outgrow their epilepsy. The 3.1% lifetime figure includes all persons who continue to experience epileptic seizures as well as those who experienced such seizures at some time in their lives but no longer do so.

Epilepsy, by definition, is a condition in which an individual is predisposed to recurrent seizures because of a central nervous system (CNS) disorder. A seizure is a sudden, involuntary, time-limited alteration in behavior, including a change in motor activity, in autonomic function, in consciousness, or in sensation, accompanied by an abnormal electrical discharge in the brain.[3]

Many clinicians had believed that the onset of epilepsy is most common in children. But recent studies have shown that epilepsy's incidence is highest among the elderly (Figure 1).[2] One survey of nursing home residents revealed that 10.1% (4,573 of 45,405) were receiving at least one antiepileptic drug.[4]

The causes of epilepsy are many; any disease that affects the central nervous system is capable of producing epilepsy. Yet, the most common cause remains that nettlesome category, "unknown" or cryptogenic. Thus, epilepsy is not a single disease, but rather a disorder with many causes. In the young, genetic syndromes and birth trauma are the most common causes. Cerebrovascular diseases are the most common causes of epilepsy in the elderly.

Epilepsy is also variable in its degree of severity. Some persons have mild epilepsy with seizures that are completely controlled with medicine. Others continue to have a few seizures, and some patients have intractable epilepsy with multiple seizures despite appropriate treatment.

In addition, epilepsy is an episodic and paroxysmal condition. Most persons with epilepsy are completely normal between seizures, but are subject to unpredictable episodes of loss of consciousness and motor control. Thus, epilepsy is a

frightening condition, misunderstood by many, including health-care professionals. This has placed an extra burden on the patient with epilepsy, who often faces discrimination in the workplace and in society.

Physicians caring for patients with epilepsy must not only have the medical knowledge to properly control seizures, but must also be sensitive to the impact this disorder has on the person as a whole. Simply controlling seizures without attending to the social and emotional issues will often lead to a less than ideal outcome.[3]

This handbook is intended to serve as an introduction to this relatively common and fascinating disorder, which can so profoundly affect the mind and consciousness.

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Chapter 2

Seizures

Over the ages, many different terms have been used to describe epilepsy and seizures. This proliferation of descriptive words has led to unnecessary confusion among patients and health-care providers. This chapter will review the terminology and classification of seizures; in Chapter 3 we will examine the classification of the epileptic syndromes.

The word *seizure* is sometimes used vaguely to refer to a sudden, catastrophic event, especially if the precise nature of the event is unknown. Many other words have been used in this context, including *fit*, *spell*, and *attack*. In this handbook, a seizure is defined as a paroxysmal, time-limited event that results from abnormal neuronal activity in the brain. However, physicians may face a situation in which the precise nature of the event being evaluated has not been determined. In this circumstance, 'possible seizure' would be a better description to avoid labeling someone before a definitive diagnosis is made.

Most seizures are not epileptic (nonepileptic), that is, not generated primarily by the brain. Nonepileptic seizures may be either physiologic or psychogenic. Nonepileptic physiologic seizures are a response to some disturbance external to the central nervous system, such as hypoxia, toxins, or fever (Figure 1). Nonepileptic psychogenic seizures are often a reaction to psychic stress. Epileptic seizures can arise from distinct regions of the brain (localization-related), or be caused by a general dysfunction of the biochemical mechanisms (generalized).

Behavioral Features

The major behavioral features of seizures that distinguish them from usual activity are that they are stereotypical and repetitive. They lack the typical modulation seen with volitional behavior. For example, a clonic seizure involves maximal contraction of skeletal muscles, followed by relaxation, with the cycle usually repeated a few times per second. This very primitive movement pattern accomplishes no useful func-

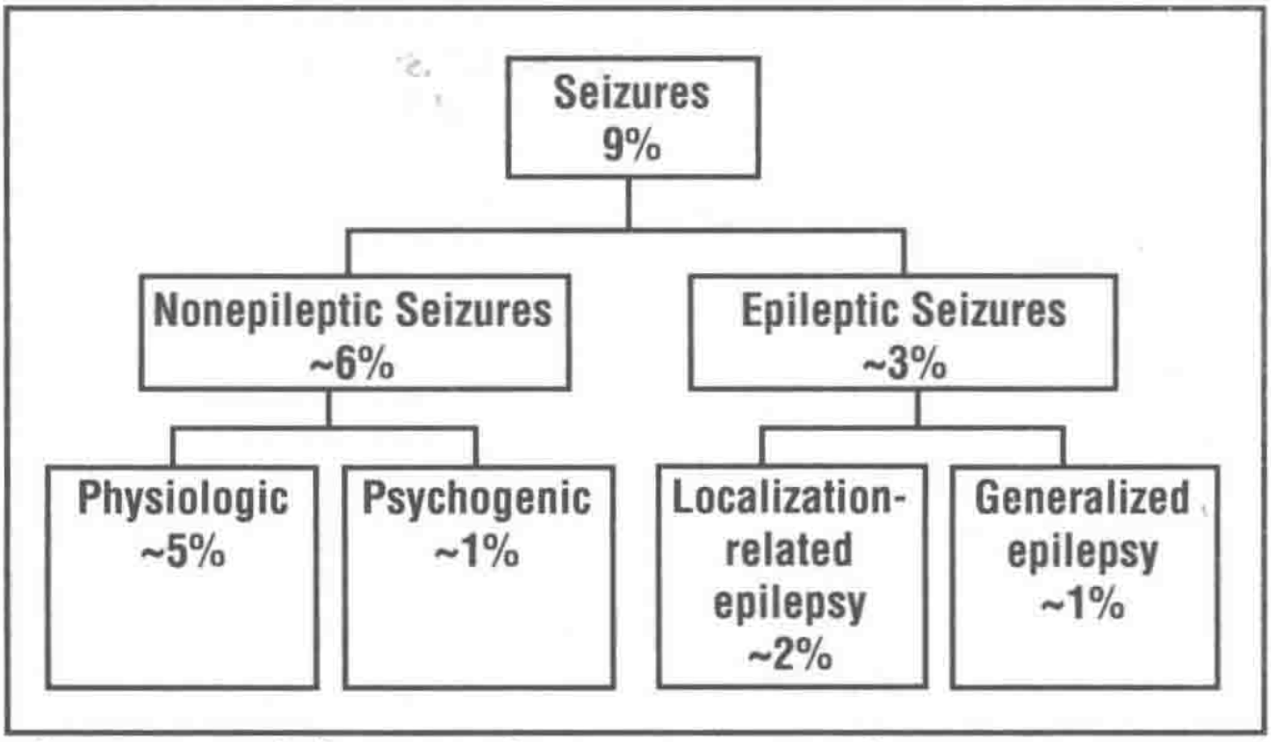

Figure 1—*Subdivision of seizures into etiologic categories and their appropriate lifetime incidences.*

tion and is in marked contrast to the usual complex, modulated activity that our muscle groups are capable of performing.

Generalized tonic-clonic seizures can evolve from partial seizures (**I** C in Table 1) or can be generalized from the onset (**II** E in Table 1). Although generalized tonic-clonic seizures of either type are indistinguishable in the clinical setting, it is critical for appropriate treatment that the correct classification be made.

More common than the dramatic generalized tonic-clonic seizures (GTS) are the partial seizures. These may be simple or complex and may evolve into secondarily generalized tonic-clonic seizures (Table 1). In 1954, Penfield used the term *automatism* to describe the unconscious, meaningless behavior exhibited by persons having a complex partial seizure.[1] A patient with complex partial seizures may perform activities that superficially appear normal, but that are done without seeming comprehension of the social setting. For example, one of my patients was described by his friends as experiencing the following: while playing cards, he developed a blank look on his face, put down his cards, walked slowly to the refrigerator, opened its door, urinated, and then appeared confused. Upon recovering awareness, he had no recollection of this event. Fortunately, his friends were aware of his epilepsy, and had seen other episodes of his unusual behavior. Imagine the re-

Table 1: Epileptic Seizures: Classification and Characteristics, as Proposed by the ILAE[2]

I. Partial Seizures (Focal Seizures)

A. Simple partial seizures
   1. with motor signs
   2. with somatosensory or special sensory symptoms
   3. with autonomic symptoms
   4. with psychic symptoms

B. Complex partial seizures
   1. simple partial onset followed by impairment of consciousness
   2. with impairment of consciousness at the onset

C. Partial seizures evolving to secondarily generalized seizures
   1. simple partial seizures
      (a) evolving to generalized seizures
   2. complex partial seizures
      (b) evolving to generalized seizures
   3. simple partial seizures evolving to complex partial seizures evolving to generalized seizures

II. Generalized Seizures (Convulsive or Nonconvulsive)

A. 1. typical absence seizures (*petit mal*)
   2. atypical

B. Myoclonic seizures

C. Clonic seizures

D. Tonic seizures

E. Tonic-clonic seizures (*grand mal*)

F. Atonic seizures

III. Unclassified Epileptic Seizures

• Includes all those seizures that cannot be classified because of incomplete data or because they defy classification into the above catagories; for example, neonatal seizures with swimming movements.

IV. Status Epilepticus

Used with permission, *Epilepsia 1981;22:489-501.*

action to this behavior from persons not familiar with the manifestations of complex partial seizures.

One of the major developments in epileptology has been the adoption of the International Classification of Epileptic

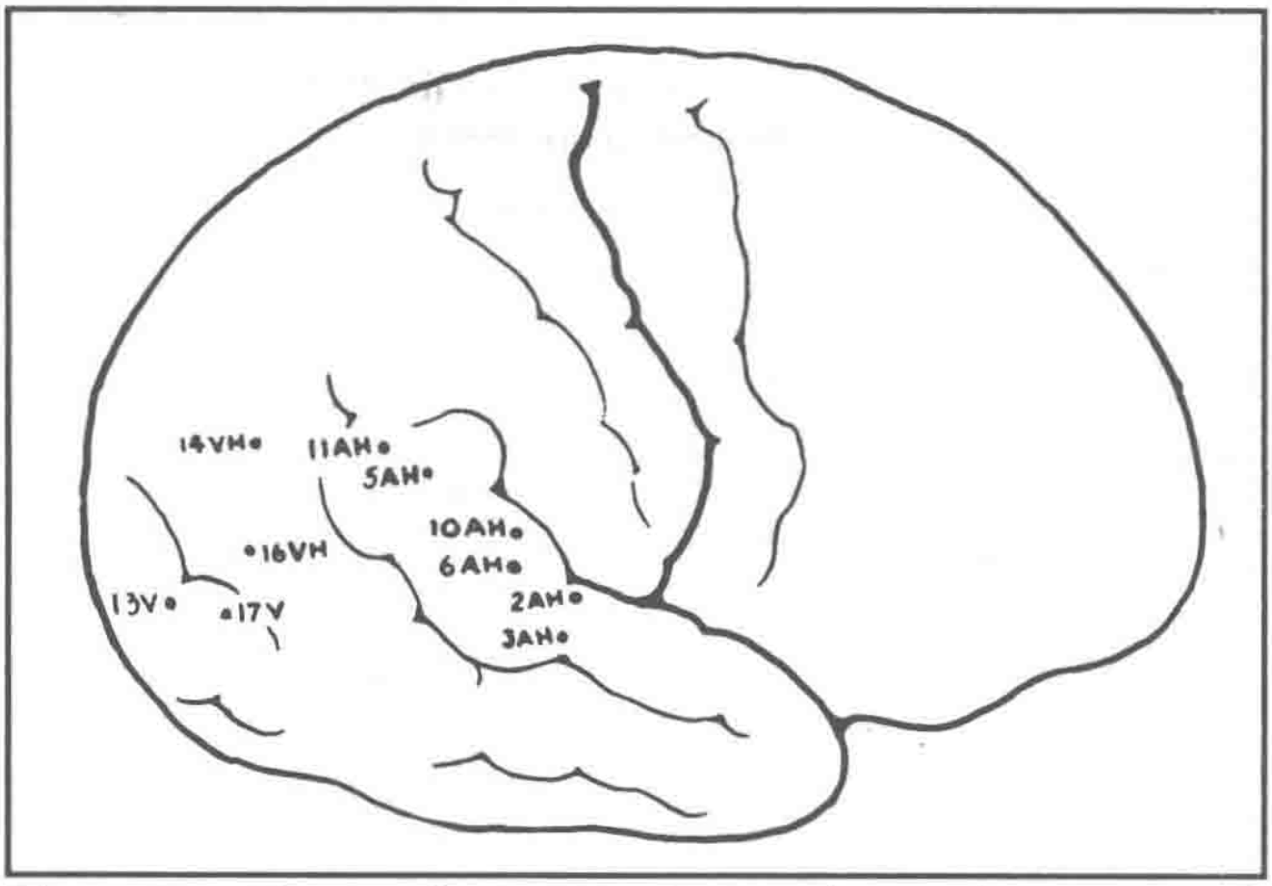

Figure 2—*Classic depiction of partial stimulation record. V=visual response; VH=visual hallucinatory seizure; AH=auditory hallucinatory seizure. Reprinted with permission from* Epilepsy and the Functional Anatomy of the Human Brain. *1st ed. Boston, Little, Brown and Co, 1954, p 464.*

Seizures (ICES).[2] At the core of this scheme, illustrated in Table 1, is the recognition of two major categories of seizures: those that begin locally in a specific region of the brain and then spread (partial seizures), and those that are generalized at onset (generalized seizures). Much of the basis for this classification is the recognition that the brain is highly organized, with specific functions represented in discrete anatomical regions. Thus, epileptic seizures that have their genesis in a discrete region of the brain (focus) are classified as partial seizures (Table 1). Conversely, those seizures for which there is no identifiable focal origin are classified as generalized. This division by physiologic criteria has proven useful in understanding epilepsy and in determining therapeutic strategies. This concept was elegantly discussed in a seminal book, *Epilepsy and the Functional Anatomy of the Human Brain*, by Penfield and Jasper.[1] This work was important in our understanding of the brain's organization (Figure 2). Their book provides numerous case reports of patients who had epilepsy surgery under local anesthesia, and those patients' responses to direct