Clinical Electrophysiology

A Handbook for Neurologists

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Part 5: The Casebook of Clinical/Neurophysiology Consults

Preface

Clinical Electrophysiology was designed for residents, neurology attendings, and intensive care specialists. It was conceived as a bridging tool that enables the clinical electrophysiological investigation to be tied in with the neurological consultation. This helps the clinician to order the appropriate electrical test, understand the meaning of the interpretation, and then integrate these findings with the clinical question to arrive at a diagnosis. It may further provide information on the differential diagnosis, the prognosis (where warranted), further relevant investigations, and some brief comments on treatment. A brief clinical reference list is included.

In making this portable aid, we placed emphasis on the inpatient clinical setting, giving the appropriate symptoms and signs, and pertinent electrophysiology results that might be found. The discussion that follows is specific to the figure given. Hence, for example, confused patients may have any of a number of EEG findings, but the discussion and prognosis are directed only to the one pattern under discussion, for example, triphasic waves. Diagnostic questions (particularly on chronic conditions) that would largely be encountered in the outpatient clinic, or investigated after patients' discharge, are not included. Hence, chronic neuropathies, palsies, Parkinson's disease, and most genetic conditions are omitted. Similarly, conditions without electrophysiologic relevancies or those warranting other types of tests (CT, MRI, and ultrasound) are not included. Although a comprehensive tome addressing all neurological testing would clearly be useful, it would not be easily portable.

For immediate relevance to neurology consults, we avoided general discussions of the neurological examination, disease entities and electrophysiology in general, as there are a number of excellent books that address these issues in detail. We recommend, of course, supplemental use of these tomes as they are essential to the understanding of clinical neurology.

The book is organized by the presenting neurological problem, for example, confusion, coma, abnormal movements, or difficulty weaning off a respirator, limb numbness, or weakness. Within these topics, there may be some general diagnostic considerations, definitions of terms, but of principal importance, we provide a test result that may be encountered. For example in a comatose patient, we give an EEG showing an invariant alpha frequency pattern. There follows an interpretation of the illustrated finding, differential diagnosis, prognosis, and references. In this way, the "vignette" starts with a clinical problem and reaches a diagnostic, prognostic, or therapeutic end.

Because the handbook is "problem-oriented," it is not a comprehensive treatment of neurologic problems. It is briefer and covers mostly what a hospital clinician might encounter on neurology consultation rounds in a typical year. The last section, however, is a "casebook," which provides several rarer, but classic, clinico-neurophysiological problems. The casebook format provides more clinical information and leaves the reader to test him or herself as the case unfolds. More information on the electrophysiological findings can be found in the respective section in the handbook.

Please use the book, if helpful, in wording your consults and in providing references. Do give us feedback into any shortcomings and major areas that we failed to include. We hope you find it a useful aide-memoire as you address clinical challenges.

Peter W. Kaplan, MB, FRCP
Thien Nguyen, MD, PhD

Introduction

We have designed this handbook to accompany you on your rounds. We believe that the handbook works best in the "middle step" of the neurology consultation process. In the first step, historical data are collected and an examination is performed to arrive at an opinion, possibly then suggesting complementary tests. If electrophysiological tests are requested, it is at the next step that the handbook is helpful in addressing the significance of the findings, the differential diagnosis, prognosis, and in providing some brief therapeutic directions. In the final step, a concluding opinion can then be formulated. In other cases, the handbook can be used to review the meaning of a particular test result that has already been received, so as to be able to provide further information to the patient's treating physicians.

Too often, the nature and significance of test results can remain uncertain: do they represent a "red herring"? Are they helpful in eliminating or confirming a particular diagnosis among many? What do they tell us about prognosis?

Standard textbooks abound to help with taking the history of a neurological complaint, performing physical examinations, or discussing the many disorders that can be diagnosed. Other texts may discuss in detail the techniques and interpretation of EEG, evoked potentials, NCVs, and electromyography. The handbook bridges the gap between the electrophysiological laboratory and the bedside.

PART 1
Central nervous system disorders

Section A: Altered consciousness: confusion, delirium, and unresponsiveness; agitation, hallucination, and abnormal behavior

These are some of the "altered states" that prompt neurology consults. Patient problems rather than specific, prepackaged "diagnoses" generate consults. Hence, clinical training rather than standard texts is the major source of learning the physician's approach to managing *problem-oriented* questions.

Unfortunately, the causes (or diagnoses) underlying a particular complaint are legion–-consider the potential causes of "dizziness," for example, low blood pressure or neurilemmoma, migraine or brainstem stroke, low blood sugar or otolith disease, and multiple sclerosis or Meniere's disease.

Clearly the constellation of symptoms and signs (and those absent) from the patient's description of clinical features (the syndrome) will pare down the possibilities and direct the diagnostic evaluation and investigation. Excellent texts are available that can address "lists" of probable alternatives to particular complaints. Maybe the future will lie in the use of a palm-held computer into which the complaint/symptom will be logged, followed by associated (or not) clinical features, resulting in the generation of a "probability list," which can be used even while one is rounding on patients.

In this section, we address certain states of altered consciousness or behavior that fall short of coma. Locked-in states, minimally conscious states, akinetic mutism, and vegetative states are a different order of "unresponsiveness," and are found in their own section further on. Those examples contained here involve acute or subacute global diminution in the level of consciousness, vigilance, memory, and cognitive processing in keeping with encephalopathies ("altered mental status") or "acute confusional states" due to toxic/metabolic, infectious, or ictal disturbances.

Some definitions in current use are as follows:

Delirium: An acute alteration in cognitive function with impaired short-term memory, sleep cycle inversion, sometimes with increased motor activity in the form of agitation and tremulousness (think withdrawal or delirium tremens), often with amnesia.

Confusion: A general term that usually needs further definition. Often, however, it is used to refer to a state of impaired language output, orientation, the ability to follow commands and to retain information.

Altered mental status: This could subsume the above. Also a non-specific term, which could apply to psychosis, coma, or dementia. It also needs further specification.

Encephalopathy: A Greek-derived term for diffuse brain dysfunction–-also non-specific. But then globally confused patients are often perforce "nonspecifically" cognitively impaired (a clue in itself).

Or there may be a clinical question at the outset: Is this nonconvulsive status epilepticus (NCSE)? This is specific and provable one way or the other. One might consider the variety of clinical features seen with NCSE and obtain an EEG.

So where to go? Once the probable type of higher cortical disturbance has been tested, for example, with a mini-mental status examination, more detailed testing of the patient's orientation, language, memory, ability to follow commands, to interpret events (the "cookie thief" picture), and then a probability list of diagnoses

can be produced. This might include a consult with the following:

> ***Possible toxic/metabolic encephalopathy. Suggest the exclusion of systemic infection in this patient with chronic diminished tolerance to the many causes of encephalopathy (e.g. cerebral atrophy; dementia). Consider also investigation of ictal/post-ictal possibilities (with an EEG).***

If in the course of investigating altered consciousness or abnormal behavior in a patient, the EEG reveals an epileptiform abnormality, turn then to the section on seizures (Part 2) for further electroclinical correlations and suggestions.

The easier questions to answer are often those centered on a request for *prognosis*. In particular instances such as after anoxia, "ball-park" answers can be provided, or even some highly exact ones. For example, the prognosis in a lethargic patient 3 days after CRA can be given with much support from the literature, and from EEG and SSEPs (somatosensory evoked potentials). For these types of questions and for those patients in coma, locked-in states, and vegetative states, please refer to Part 3 on these disorders. A brief overview on prognosis and evaluation can also be found in the section on Evoked Potentials in Consultative Neurology.

1. Diffuse and frontal fast activity—beta

MICU, CICU, NICU, SICU, WARD, ER

CLINICAL CORRELATES: A patient may have been referred to the electrophysiology laboratory for one of several clinical reasons, and the EEG reveals medium to high-voltage diffuse beta frequencies. In a patient with little history, it would suggest drug intoxication and the need for a toxin screen. The patient may be normal, drowsy, or rarely agitated.

ETIOLOGY: Benzodiazepine, chloral hydrate, or barbiturate treatment or intoxication. Occasionally, sedative withdrawal states. With high medication doses, the patient may be sedated to the point of unarousablility (beta coma, usually >30 μV on EEG). It can occur with brainstem injury [4].

CLINICAL EVALUATION: Record all medications to which the patient has access. Look for medication/sedative effects; alternately, the patient may be agitated rarely with delirium.

ANCILLARY TESTING: Toxin screen for barbiturates or benzodiazepines. MRI of brainstem structures.

DIFFERENTIAL DIAGNOSIS: For the EEG pattern, it may occur with benzodiazepines, barbiturates, sedative withdrawal, childhood mental retardation and cerebral palsy, brainstem injury.

PROGNOSIS: There is little dependable literature on the significance of this finding. The prognosis/reversibility, when this is due to medications, is excellent. In children there is a report of continuous beta spindling in cerebral palsy and mental retardation (extreme spindles). The spindle beta patterns are associated with a good prognosis regardless of etiology, with the exception of children not on barbiturates or benzodiazepines.

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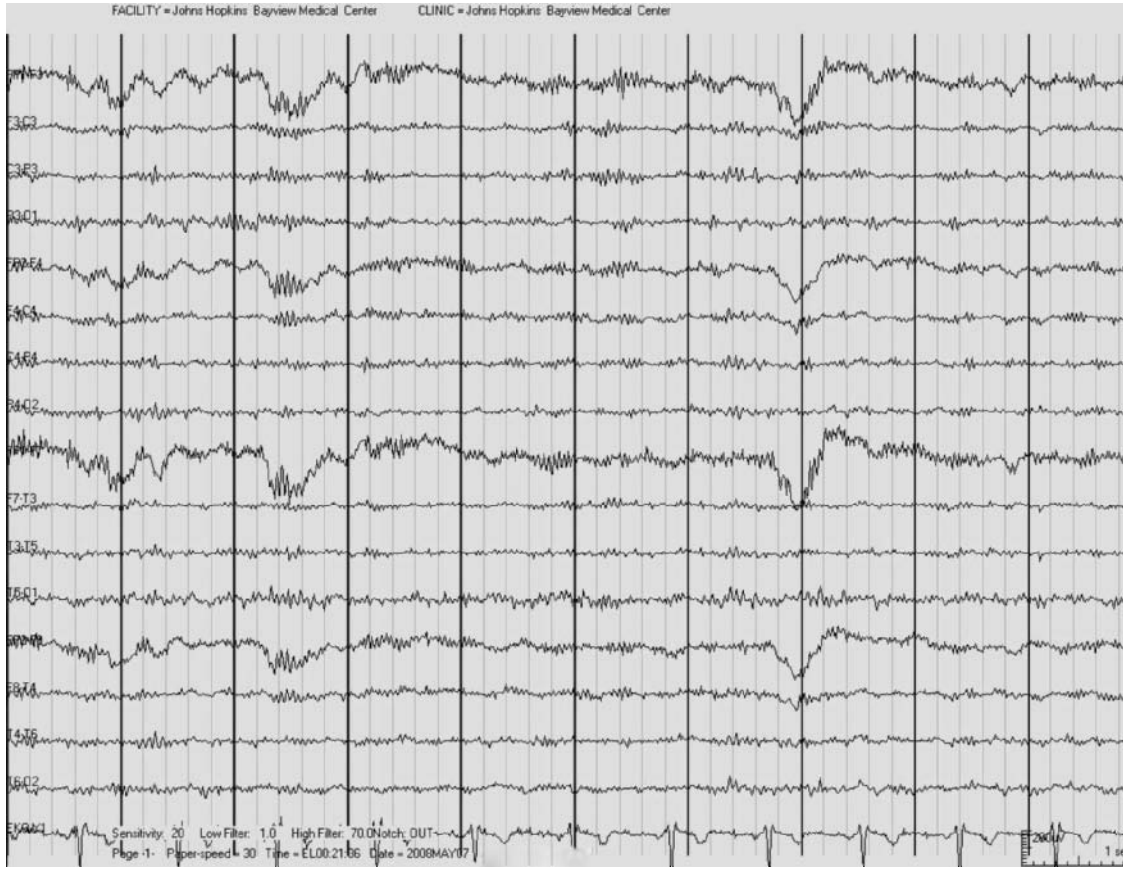

This EEG shows a medium- to high-voltage diffuse fast beta pattern. In this case, it is prominent anteriorly, particularly in light sleep and following arousal. Occasionally, it may show a spindling pattern. On EEG, in general, there are beta frequency bands typically seen at 18–25 Hz, less frequently at 14–16, and in one report at 35–40 Hz. It is considered high voltage when it exceeds 25 μV [1–4]. It was originally, probably incorrectly, believed to be associated with epilepsy, minimal brain dysfunction, dyslexia, hyperactivity, or other behavioral dysfunction. Conversely, this pattern is typical of a medication effect.

REFERENCES:

1. Frost JD, Carrie JRG, Borda RP, Kellaway P. The effects of Dalmane (flurazepam hydrochloride) on human EEG characteristics. *Electroencephalogr Clin Neurophysiol* 1973; 34:171–175.
2. Kellaway P. Orderly approach to visual analysis: Elements of the normal EEG and their characteristics in children in adults. In: Ebersole JS, Pedley TA (eds.), *Current Practice of Clinical Electroencephalography*, 3rd edn. Philadelphia, PA: Lippincott/Williams and Wilkins 2003;100–159.
3. Kellaway P. The development of sleep spindles and of arousal patterns in infants and their characteristics in normal and certain abnormal states. *Electroencephalogr Clinc Neurophysiol* 1952;4:369.
4. Otomo E. Beta activity in the electroencephalogram in cases of coma due to acute brainstem lesion. *J Neurol Neurosurg Psychiatry* 1966;29:383–390.

2. Diffuse slow activity–theta [1–4]

MICU, CICU, NICU, SICU, WARD, ER

Acute encephalopathies—frequently the elderly, multiorgan failure. Static encephalopathies, mild diffuse cortical dysfunction.

CLINICAL CORRELATES: Psychomotor slowing, confusion, clouding of sensorium. Brainstem function is intact.

ETIOLOGY: In the ICU, causes typically include toxic and metabolic dysfunction, and systemic infection. Often seen in elderly patients with cerebral atrophy with the above causes, as well as in dementias, static encephalopathies, mental retardation, and learning disability.

CLINICAL EVALUATION: Higher cortical function, general neurological examination.

ANCILLARY TESTING: CT or MRI may show subcortical atrophy; evidence of head injury; chronic encephalopathy. Test for organ failure—hepatic, renal, respiratory, or other organ dysfunction.

DIFFERENTIAL DIAGNOSIS: From the EEG perspective, check that the patient is not just drowsy or asleep during this EEG segment (normal drowsy pattern), and ensure that the EEG recording contains adequate noxious stimuli to ensure full arousal during the EEG.

PROGNOSIS: Due to static encephalopathy, it reflects a chronic state of cortical dysfunction and has no particular prognostic import. If seen with organ dysfunction, then the electroclinical picture may be reversible. Even after anoxia, patients with this theta pattern often improve clinically and on EEG [3, 4].

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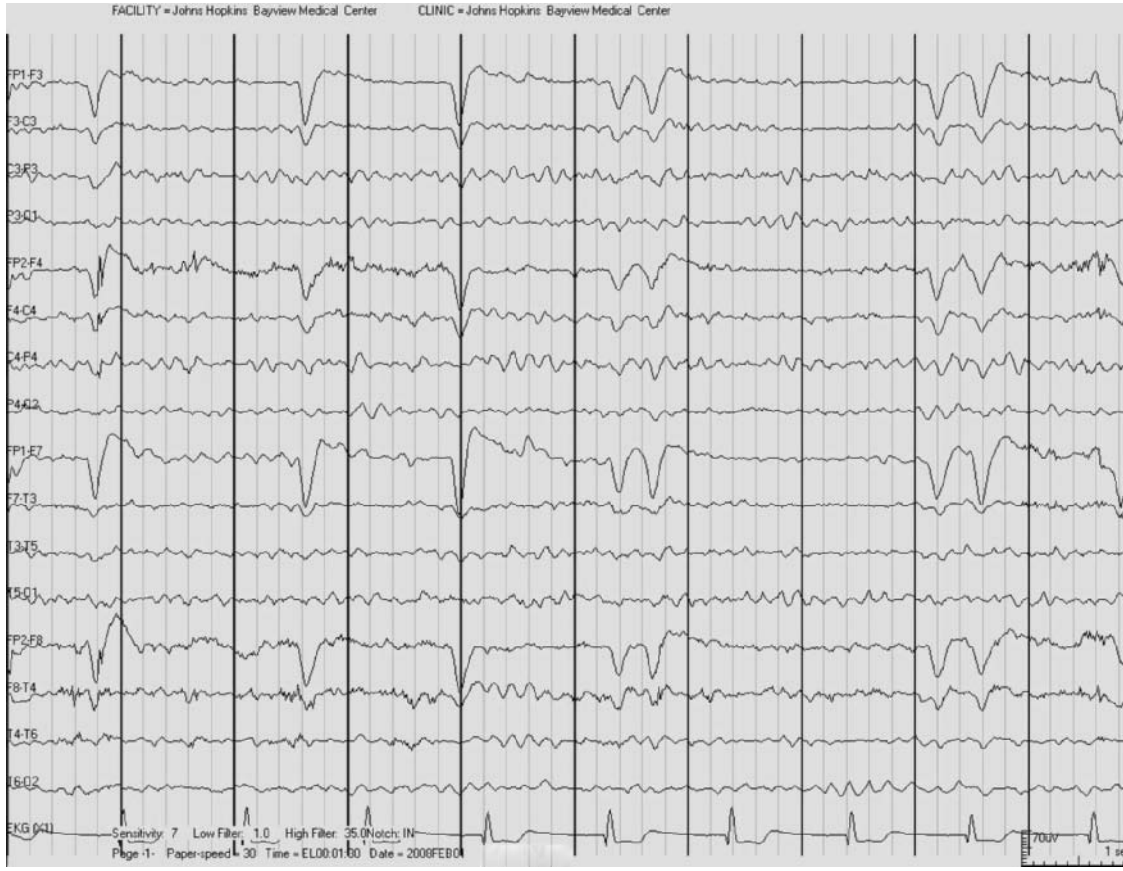

This EEG shows widespread theta activity. There is variable intrusion of alpha and delta frequencies; alpha can be seen with maximal arousal. The eye blink artifact seen every several seconds bifrontally indicates the awake state of the patient. Diffuse theta is onlyless frequent than other EEG patterns seen in confusion/encephalopathic states (possibly due to ascertainment bias).

REFERENCES:

1. Chatrian G-E, Turella GS. Electrophysiological evaluation of coma, other altered states of diminished responsiveness and brain death. In: Ebersole JS, Pedley TA (eds.), *Current Practice of Clinical Electroencephalography*. Philadelphia, PA: Raven Press 2003:405–462.
2. Gloor P, Kalabay O, Giard N. The electroencephalogram in diffuse encephalopathies: EEG correlates of grey and white matter lesions. *Brain* 1968;91:779–802.
3. Silverman D. Retrospective study of the EEG in coma. *Electroencephalogr Clin Neurophysiol* 1963;15:486–503.
4. Yamashita S, Morinaga T, Ohgo S, et al. Prognostic value of EEG in anoxic encephalopathy after CPR. Relationship among anoxic period, EEG grading and outcome. *Intern Med* 1995;34:71–76.