An Introduction to Neurobehavioral Disorders

(Temporal Limbic Epilepsy)

John Matthew, M.D.
The Health Center, Plainfield, Vermont

Of the various neurobiological disorders which can occur in children, adolescents, or adults, and which may cause severe emotional disturbance, some of the more common but, often least well recognized and understood, are a group of disorders caused by abnormal electrical activity in the temporal and inferior frontal lobes of the brain.

Because many of the manifestations resulting from this electrical activity are "psychological" or behavioral in nature, and the more "neurological" symptoms may be subtle, overlooked, or nonexistent, this disorder may sometimes masquerade as, be misdiagnosed as, or "duplicate" classical psychiatric syndromes such as panic disorder, depression, schizophrenia, bipolar illness or attention deficit disorder. It may also coexist with these other disorders.

The temporal lobes and lower portion of the frontal lobes, which contain the limbic system, are the portions of the brain which are most susceptible to injury and have the lowest threshold or trigger point for having electrical brain impulses spread abnormally from cell to cell. Minor injuries from the birth process, falls, or blows to the head or more clear-cut injuries such as those sustained in automobile accidents or being knocked out, are thought to be the cause of the abnormal electrical activity in many persons suffering from this disorder. Protracted febrile convulsions, infections, and anoxia are also important causes.

There are a variety of symptoms which may result from this abnormal spread of electrical activity in the temporal lobe(s), depending upon the severity and location(s) of the electrical problem. The result may range from clear-cut temporal lobe epilepsy, with problems evident to any observer, to subtle variations in mood or temperament caused by more localized electrical events. The most common features of these subical ("below seizure") or "partial complex seizure" (pes) neurobehavioral disorders are moods which shift rapidly, either spontaneously or with little stimulus; irritability; and "dysphoria", which is a feeling of brooding, tenseness, or mild depression.

"Subical" really isn't non-seizure. This term describes any event which electrophysiologically involves seizure discharge but which causes symptoms that are below the threshold of recognition as a clinical seizure. Thus subical events are the results of abnormal electrical discharges, but the electrical seizure activity (ictus) is so localized, involving only particular brain regions, that the manifestations which result, are not seen as "seizures" as these are ordinarily understood.

While the classic ictal events tend to be highly stereotyped in a given patient, there is a striking difference between the classic seizure events, if these occur in an individual, and the interictal ("between seizures") and subical ("below seizure") manifestations. These tend to be pleomorphic, with one event not quite like another, involving paroxysmal or intermittent symptoms which are less stereotyped. Sometimes a rich and variable "psychopathology" results, at times ending abruptly if a generalized seizure event occurs.

An illustrative case contains many instructive elements:

An 11 year old boy presents with a history of rather suddenly having intervals of marked change in his usual behavior and personality. This has included explosive rages and withdrawal from most of his activities with fear of "spells" of "dizziness" and abrupt onset of intervals of remarkable depression. He has been in treatment, with no medication suggested and virtually no progress made, for nine months with a psychiatrist who diagnosed panic disorder. The clinical formulation explained his unusual behavior as an expression of the child's reaction to intrafamilial disturbance.

It is true that this boy's symptoms include panic and other symptoms often associated with panic disorder (complaints of dizziness, for example). It is also true that the family is "disturbed". His two siblings and his parents are in fact distraught to see the boy having explosive rages, spells of deep depression and intervals of "complete change of personality". Their world as well as his, is literally coming apart. Neither of his siblings
apparently has been nominated for this role of "expressing pathology of the disturbed family system", though they seem as well qualified for this role as is their brother.

A few minutes of informed inquiry puts the matter into a much more likely focus. The boy's problems began shortly after a pair of impressive head blows received in falls while playing the previous year. One involved running full force backward into a tree with "a sound like a coconut being hit" and a brief interval of loss of consciousness resulting.

His list of subsequent and current symptoms is quite long, including "terrible behavior" with explosive rages, spells of dizziness (spinning sensations); severe headaches; panics; intervals of abrupt brief, deep depression; withdrawal from his usual activities; "sensitivity to odors"; marked sensitivity to criticism; frequent 'deja vu'; intervals of slight confusion; brief episodes of motionless staring; memory gaps; intervals of "out of character" aggressive behavior; visual distortion ("faces look plastic"); perceptions of immobile objects appearing to move; micropsia and macropsia; visual image color changes; visual images taking on an artificial picture-like quality; memory gaps for portions of his anger spells; prickly paraesthesias of his limbs with intervals of fearfulness; and brief spells of "dizziness" with confusion, imbalance, lack of memory, and "out of touch" followed by 30 minutes of "not right" before return to normal.

This boy's EEG and brain study with magnetic resonance imaging (MRI) are both normal, but he has a clear case of temporolimbic or "neurobehavioral" disorder consequent to his head injury. Treatment with an antiepileptic drug causes "90%" improvement to be reported by his parents and by the boy. Adding low dose lithium helps control persistent angry outbursts. Within a few weeks of proper diagnosis he is doing well again in school, in sports, with friends, and in his family.

On reviewing the case, the boy's psychiatrist noted that neither the boy nor his family had informed him about his sensory and memory symptoms or other unusual experiences. The psychiatrist, following accepted theory and practice concerning psychiatric history taking, had not asked about such symptoms.

This case illustrates that questions about symptoms of partial complex seizures should probably be a routine part of the psychiatric evaluation. Careful inquiry may be necessary to uncover a history of characteristic events, which often are not spontaneously reported by the individual but which are clues to his or her having minor non-motor seizure events. The events are typically paroxysmal in onset and may occur only after a long latent period has passed since the occurrence of the presumed triggering injury, if there has been one. These symptoms and events may include any of the following; in subtle or clear-cut fashion:

- Minor interruptions of attention ("spaceouts") which may be from seconds to minutes in duration
- Minor interruptions of memory ("memory gaps") The "memory gaps" may include lack of recollection of activities, details, or states of mind
- Staring spells, decreased ability to concentrate or failure to "register" things in short term memory. The family may see these absences as "she doesn't pay attention when I talk with her." Also, "low motivation" or "daydreaming" in school are common descriptions.
- Brief automatic movements such as lip smacking, a twitch or tremor, licking, chewing, swallowing, sniffing, blinking, unusual laughter, or an automatic smile or other unusual facial expression
- Spontaneous flushing, sweating, looking very pale, or feeling short of breath, or feeling faint or off balance, or having prickling of fingers or toes
- Abdominal pains (without abdominal illness), unusual feelings in the stomach, a peculiar sensation in the top of the abdomen, or appetite increase or decrease.
- Headaches, binges of water drinking; vertigo (spinning sensation), nausea, diarrhea, insomnia, or sleep which is interrupted or fails to refresh. There is a correlation of this disorder with migraine type headaches, and, some experts report, stuttering in some individuals
- Frequent episodes of Deja-Vu ("I've been here before" in new circumstance) or Jamais-Vu (sense of unfamiliarity in a familiar/old circumstance) or having feelings of things not being real (dreamy or "twilight zone"); feeling cut off or at a distance from a situation one is in, as if observing from another point of view
- Periods, which may be quite brief, of confusion; of disorientation; of difficulty finding a familiar word; of loss of memory of a place, or of an event; or loss of the ability to do something or recall a concept. Having problems retrieving a visual memory or finding ones way in unfamiliar circumstances
Amnesia, or more often a hazy or partial memory, for all or part of an episode such as an anger attack or interval of altered mood, or temporary amnesia for some portion of one's past.

Smelling a smell which others don't smell, or tasting a taste with no source in the mouth; often these are bad smells or tastes such as the smell of burning rubber or hair, or rotten food, or the taste of metal, "musty" or the like. The smells or tastes may be difficult for the individual to describe.

Changes in vision such as seeing things magnified or smaller than reality, or appearing to recede or move toward the observer; seeing sparkling spots, flashing or colors; experiencing visual distortions such as spots or blurs; seeing things which aren't there; misinterpreting an object for something else.

Though "neurological" events listed above such as brief staring episodes or visual phenomena may often be observed or discovered with careful questioning, all of the apparent characteristics of the disorder may be confined to what appears to be the "psychological" sphere.

The disorder is quite often phasic, at least in its more florid manifestations. Intervals of relative normalcy may alternate with phases during which more characteristics or symptoms occur. Between more symptomatic periods these individuals often report feeling restless, tense, irritable, or easily frustrated, but they may be quite normal.

The time course of the phasic events may be quite variable, with the more obvious disorders of mood, for example, occurring every few days to every few months, usually characterized by rapid onset and termination or sudden immediate appearance and fading. Periodic recurrences are the rule, though the emotional events may vary from mated and subtle forms to florid, or deep depression.

The characteristic mood swings may be most of the obvious clinical picture. A frequent presentation is of rapidly cycling bipolar mood disorder, with substantial alterations of mood occurring 4 or more times a year. An individual may have swings of variable intensity at various times, some subtle and some marked, some short lived and others longer lasting. Other "psychological" events, which may occur for intervals of seconds, minutes, hours or days, include:

-- Transient or short lived (seconds to hours), sometimes intense, emotions which are spontaneous or easily triggered, possibly including feelings of anxiety or restlessness, elation, panic, frustration, depression, loss of pleasure, anger, irritability, fear, paranoia, sense of impending doom or sense of impending ecstasy, moodiness, apprehension, being "uptight", testy or bitchy or feelings of being threatened or attacked.

-- Marked emotional lability, which may include the sudden explosive release of pent up or crude emotions, with paroxysms or states of anger, hate, jealousy, envy, or vengefulness.

-- Rage attacks or violence, unprovoked or easily stimulated, out of proportion to the stimulus. Sometimes these may involve destroying property or hurting someone. This category of effects may range from seeming to "just have quick temper" to storms of emotions during which one's anger is out of control and they become very much unlike their usual selves.

-- Extreme anger, which does not have to be aimlessly expressed or occur with some alteration of normal consciousness. Quite purposeful behavior may occur and the fury may be quite directed, for example at one individual or group.

-- Impulsive suicide attempts made in a fit of rage or anger.

-- Intervals of apparent "change of personality" or "forced thinking", rushing thoughts, or intrusion of an idea into one's thoughts. Others may note a change from good humor to marked seriousness, or a startlingly abrupt change in personality, or even very brief episodes of an unusual laugh or a sneer which is uncharacteristic. "Gelastic seizures" may cause intervals of uncontrollable laughter.

-- Intervals of unusual emotional intensity, impulsiveness or frustration, overemphasis and/or satisfactions in conversing, accentuated sensitivity to slights, selfishness, a tendency to become angry about trivial events, increased sense of justice and/or heightened sense of outrage.

-- Intervals of "manic" or somewhat manic thought and behavior, with heightened intensity of feelings and ideas, inflated self-confidence, high energy, and/or unbridled enthusiasm. These "highs" may alternate with intervals of depression.

-- "Hypomanic" presentations including high spirited roguery, grandiose thoughts, tales of adventurous experiences, and cheerful, ebullient, affable, good-natured, or hypersocial intervals.
Mixed states involving "energized dysphoria" or other mixtures of depressive and hypomanic or manic characteristics are often seen, particularly in adolescents and young adults.

Impulsivity and a feeling of "internal unrest"

Periods of incredibly stubborn, obstinate, markedly selfish, surly, sullen, and/or quarrelsome disposition. The individual may perceive mistreatment by others or feel that others are being cross with them.

"Dissociative episodes" which may involve alteration of personality, sense of personal identity and/or speech pattern, sometimes with amnesia reported afterwards.

The less desirable emotional states may alternate with intervals of relative normalcy, during which the individual may exhibit a strong sense of conscience, tolerance, justice, altruism, and/or piousness. The contrast with other states of mind such as angry outbursts or episodes of abusiveness may be quite striking.

An individual may experience quite a few symptoms from the above lists or may have only quickly changing moods which vary for little apparent reason or change abruptly.

There are secondary effects on the individual's social and psychological development. She or he may be ashamed of, or confused or embarrassed by their words or behavior or may become convinced that they are not the sort of person they had thought themselves to be, or that they simply cannot get along with their families, in school, or in a particular situation or location. The disorder can also continue at a low level between bigger episodes, causing ongoing disturbances which the individual—and those around him or her—may tend to see as their "real" character or personality rather than as manifestation of this illness. They often have a good natured, helpful attitude alternating with stubbornness, egocentricity, argumentativeness and/or vindictiveness.

The treatment of these disorders is anti-epileptic medication, of which there are a variety. Sometimes one way may work better than another for a particular individual. These medications are entirely different from major tranquilizers (neuroleptic), antidepressants, or antianxiety medications. They are not meant to tranquilize or sedate the person who takes them. They help control the abnormal electrical activity, restoring control of emotions and stopping the space outs, memory gaps, and so forth.

Often co-medications with small amounts of tricyclic antidepressant medications or lithium added to anti-epileptic medication, is necessary to help the individual with depression or unstable moods which may not be alleviated by the anti-epileptic medication.

When left unattended, these disorders may wreak havoc in the lives of the individuals and of their families. Volatile moods and outbursts of anger, defiance, aggressiveness, or rage imperil relationships with friends, family, and coworkers and often hurt loved ones. School failure is frequent, since poor concentration, memory gaps, and/or irritable/explosive moods interfere with normal learning. Untreated individuals often leave behind a trail of lost jobs, dropped interests, broken romances or marriages, disrupted family relationships, and bridges burnt behind them. An interval of overly optimistic enthusiasm may land him or her in financial trouble or cause involvement in schemes unlikely to succeed. Often the individual, impelled by his or her impulsive restlessness, moves from place to place, or job to job, or to friend to friend, generating a chain of personal and social catastrophes. Dangerous driving and other risk-taking behavior is common.

At worst, perhaps, some of these volatile, irritable unhappy people end up in prisons for such crimes as assault. Over 20 years ago, in fact, anti-epileptic medications were offered on a trial basis to prisoners in a Massachusetts County prison who reported such characteristics as irritability, unusual tenseness or restlessness, temper outbursts, fearfulness, or difficulty focusing and learning. Almost all reported rapid and desirable changes in mood and concentration, and reduced tendency to get into arguments or fights. Quite similar results were obtained in similar trials with adolescents in detention.

The untreated individual with a neurobehavioral disorder is also particularly prone to drug abuse. The unpleasant emotional states described above are uncomfortable and the individual does not wish to be feeling or acting so negatively. He or she in part "self medicates" with drugs, including alcohol, and tobacco. Cigarette smoking helps calm restlessness and anxiety and moderate irritable moods, but at the cost of the deadly consequences of tobacco use. Alcohol temporarily decreases dysphoria, but it also lowers the seizure threshold, making the electrical events more likely. Once set off, the electrical events can be stimulated for days, long after the alcohol is gone from the body. Various other street drugs including marijuana have similar effects, as do certain prescription medications such as antihistamines, some asthma medications, cortisone, and some antidepressants. These individuals are also prone to "pathological intoxications", in which alcohol...
induces bizarre or uncharacteristic, often explosive, behavior. Using Cocaine may induce the partial complex seizure state in a perfectly normal individual or strongly aggravate the tendency. This characteristic mobilization of the disorder by certain medications and drugs makes it more important than ever that these people not use street drugs and alcohol and that care be taken in choosing prescription medications.

It has been reported that the use of Cocaine or LSD can induce permanent temporolimbic epilepsy changes in the brains of individuals who previously demonstrated no evidence of such a disorder, either causing the disease or at least unmasking it. These individuals continue to experience the disorder long after discontinuing drug use.

Other than certain medications, alcohol, and street drugs stimulating the disorder, the seizure threshold is lowered when the individual is fatigued, sleep deprived or overtired and by strong emotion. Thus the disorder may be triggered by being poorly rested or by normal emotions such as strong anger or anxiety arising from circumstances, then snowballing into outbursts of emotionality.

Left untreated, the disorder may become progressively worse. Through a process known as kindling, electrical activity that is on a "microscopic seizure" level becomes, over time, more easily triggered and more widespread. When left untreated the epilepsy in one hemisphere may induce mirror foci of seizure disorder in the opposite hemisphere.

Some sufferers, over years of uncontrolled PCS/mood disorder events, don't just lose the opportunities for normal emotional, school, work, family and friendship aspects of life—some become progressively worse to the point of becoming psychotic. Others develop distinctive behavioral syndromes that can include intense religious or philosophical interests, compulsive writing or graphic arts activity, alterations in sexuality, intermittent aggressiveness, and a stubborn tendency not to "let go" of arguments or a particular point of view in conversation. Many others don't progress to increasing difficulties, but the disorder takes a continuing toll on their lives and the lives of those around them, so long as it is untreated.

Just as there is a "kindling" phenomenon which may occur in the untreated individual, there may be a "quenching" phenomenon as well. When seizure medication is used an interval of improved control may at times be observed after medications is temporarily withdrawn. Just as in motor (Grand Mal) epilepsy, a proportion of individuals who are treated can, after several years treatment, be taken off the seizure medication and not have the disorder reoccur. These individuals appear to have healed, at least in so far as the more obvious manifestations are concerned.

Diagnosis of this disorder does not depend on any particular test. The EEG may be abnormal, either showing clear-cut "seizure" electrical events or less clear-cut changes. But the EEG may be perfectly normal even during a PCS event, because the EEG only records from the outside 5mm of the brain surface, while the structures involved in the PCS/mood events are often those deeper inside the brain. So a normal EEG does not mean this problem does not exist. Other biological tests characteristic of depression or mania are normal as well.

In even the most severe cases, images obtained by magnetic resonance imagery (MRI), our most sensitive brain imaging technology, are normal in almost half of instances. The most common abnormality seen is "mesial temporal sclerosis" caused by scarring in the medial portion of the temporal lobe cortex. Most often CT scans are normal, though an occasional cyst or other abnormality is found.

Similarly, the usual psychological tests are useless for finding this disorder. Even special "neuropsych" testing is abnormal in only a bit more than 20% of instances. The diagnosis depends upon a high index of suspicion in a physician familiar with the possibility of the problem occurring. Careful questioning for the characteristics is a necessity, since the changes can be subtle. Also family members must be asked to contribute their observations since the afflicted individual often does not--can not--recall all the details or see him or herself clearly. This is said to be particularly true for those with an epileptic "focus" or problem in the right temporal lobe, since they may tend to "polish" their stories or deny their difficulties. Perhaps they truly do not record all of the details in memory, since the misrepresentation does not appear willful, or contrived.

One writer described a characteristic of the disorder as "lack of insight and a hypertrophied (over-grown) sense of denial". So input from family members is essential.
The disorder tends to build up in spirals of moodiness, explosiveness, behavioral changes, and even altered thinking ("ideation"), with profound changes in insight, misinterpretations of reality and altered judgement, then disappear, sometimes after a "staring spell" which can be quite brief, even lasting only a couple of seconds. The affected individual tends to credit his or her altered feelings to the circumstances of life, and, since he may not remember portions of his behavior, he may feel falsely accused when confronted with his behavior.

Even when there are no obvious manifestations of the disorder apparent, subclinical epileptiform activity may interfere with learning and scholastic performance. Additionally, there are often associated neuropsychological deficits which must be taken into account in approaching the individual's education and training. These are elucidated by neuropsychological testing undertaken after medications have been employed to bring the abnormal electrical activity under control.

A common error for psychologist, patient, family, or friends to make is to confuse the illness with personality or the result of circumstances of life, which may result in failure to get help for this sometimes subtle but often distinctive disorder of brain electrical activity.

On the other hand, a trial of the medications which help this problem is of extremely low risk and can rapidly return control of his or her life and emotions to the individual who is otherwise an unwitting victim with no chance of exerting control or will power to control the disorder him or herself, except to avoid drugs, alcohol, and other potential triggers.

It is usually a great relief to the individual to find that the problem is biological and to be able to get it under control with rather simple oral medication. While this medical problem is not something one would wish to have, discovering that they are not psychologically out of balance or affected with a disordered personality or "crazy" is often a considerable relief. On medication they return to being "themselves" and then repair their family relationships and get along with what they want to do in life without the disruption the bioelectric mood disorder has forced upon them before treatment.

[The concepts contained in this introductory paper have been brought together from a number of sources and are derived from the views and publications of several researchers and clinicians interested in this area of medicine. In some cases a phrase or descriptive word from one source or another has been used if it is particularly apt or evocative of what is being described. This conceptual model is the work of many, only pulled together in this paper to provide a relatively concise introduction.]

--Some of the sources and best written reviews are listed on the accompanying reading list.  
--Acknowledgement: The author is indebted to Dr. Orin Devinsky, Dr. Irwin Greenberg, and Dr. Deitrich Blumer for their review of the manuscript and for their helpful comments and suggestions.