Semiology: Witness to a Seizure – What to Note and How to Report

James W. Jordan, M.D.

Neurological Institute
University Hospitals Case Western Medical Center
Cleveland, Ohio

ABSTRACT. This article reviews the Cleveland Epilepsy Classification (CEC) of seizure semiology. It defines the concept of semiology, and reviews its importance in epilepsy in defining the Symptomatogenic Zone, one of the five zones that make up the Epileptogenic Zone. It details the four broader spheres that contain all of the possible patient signs and symptoms (cognitive, autonomic, consciousness, and motor spheres), and reviews the specific seizure types that are classified within those spheres. Evidence-based information on usefulness of semiology for localization and lateralization is reviewed. A “user-friendly” reference table is provided.

KEY WORDS. Classification, Cleveland Epilepsy Classification (CEC), epilepsy, seizure, semiology, symptomatogenic zone.

INTRODUCTION

This article will review the systematic way many in the field of epilepsy classify seizure semiology. “Seizure” is defined as “a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain” (Engel 2006). “Semiology” – literally translated – means “the study of signs” and, thus in epilepsy, it refers to the study of the signs and symptoms of a seizure (i.e., what does the behavioral phenomenon that makes up a “seizure” reveal about a patient’s particular epilepsy?). The semiological classification method reviewed herein is part of a larger five-dimensional, patient-based classification system (Loddenkemper et al. 2005b), recently named the Cleveland Epilepsy Classification (CEC) by Dr. Hans O. Lüders (Lüders 2007), who is the leading proponent of this
system. The CEC has been used successfully by epileptologists throughout the world for nearly two decades, and can be used in pediatric and adult settings, as well as in cases of status epilepticus (Bautista and Lüders 2000, Bonelli et al. 2007, Hifanoglu et al. 2007, Kellinghaus et al. 2004b, Loddenkemper et al. 2005b, Lüders et al. 1998, Lüders et al. 1999, Lüders and Noachtar 2000, Lüders 2007, Rona et al. 2005).

Semiology has blossomed over the years, giving evidence-based guidance as to what should be noted during a seizure. The CEC gives those working in the field of epilepsy a way to report the most important details about a seizure succinctly and systematically. How often does a technologist get called by a physician reading a routine EEG, who asks, “what did the seizure look like exactly?” This classification allows massive amounts of information to be shared in a useful and universal manner. Those not in the field of epilepsy will likely find the information fascinating, since knowledge of semiology is essentially knowledge of functional neuroanatomy, or “where the brain does what.”

While reviewing seizure classifications, one can refer to the user-friendly summary table (Table 1) as a reference. Although the table might appear intimidating at first, it is meant to be a simple, comprehensive reference for those using the classification system. It includes colloquial examples that patients or their families might actually present to the healthcare worker, as well as the associated Symptomatogenic Zone (see below), and associated cartoons for any visual learners.

**WHY STUDY SEMIOLOGY?**

Studying semiology helps to localize the Symptomatogenic Zone in the brain, which ultimately aids in localizing the Epileptogenic Zone (EZ) – essential in cases of possible epilepsy surgery. Further, the EZ is, itself, closely related to the Epilepsy Syndrome in the CEC, and diagnosing the Syndrome is essential to clinical management (Lüders and Noachtar 2000, Lüders et al. 2006).

Briefly, the EZ is defined as the “minimal area of cortex that must be resected to produce seizure-freedom” (Lüders et al. 2006). Although this is a theoretical volume, its localization is thought to be qualitatively successful if a patient, post-surgically, has achieved seizure-freedom and has no new deficits. The strategy employed to localize the EZ is analogous to pinpointing one’s car with overlapping satellites using global positioning systems (GPS). The EZ is derived from five sub-zones: the Irritative Zone, the Seizure-Onset Zone, the Epileptic Lesion, the Functional-Deficit Zone, and the Symptomatogenic Zone. The EZ is then concluded to be the volume where the sub-zones are most logically related or overlapping with one another.

The Irritative Zone is the part of the brain that produces the interictal epileptiform activity on electroneurodiagnostic studies. The Seizure-Onset Zone is, then, that part of the cortex that seems to produce the actual seizure on electroneurodiagnostic studies. The Epileptogenic Lesion is a lesion found on neuroimaging that is known
Table 1. Cleveland Epilepsy Classification of Seizure Semiology with Examples of Patient Symptoms and Symptomatogenic Zone.

<table>
<thead>
<tr>
<th>Classification</th>
<th>Example of Symptoms</th>
<th>Lateral/Localization</th>
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<tbody>
<tr>
<td>I. Auras</td>
<td></td>
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<tr>
<td>I.a. Somatosensory Aura</td>
<td>&quot;My right side is warm.&quot;</td>
<td>Contralateral hemisphere.  Primary Somatosensory area.</td>
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<tr>
<td></td>
<td>&quot;My shoulders feel warm.&quot;</td>
<td>Poorly lateralized. SSMA.</td>
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<td></td>
<td>&quot;Both of my hands tingle.&quot;</td>
<td>Poorly lateralized. Secondary Sensory Area (superior bank of Sylvian fissure).</td>
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<tr>
<td></td>
<td>&quot;I see birds on my right.&quot; (complex)</td>
<td>Contralateral hemisphere.  Association visual cortex.</td>
</tr>
<tr>
<td>Classification</td>
<td>Example of Symptoms</td>
<td>Lateral/Localization</td>
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<tr>
<td>I.e. Gustatory Aura</td>
<td>&quot;Yuck! I taste something bitter and awful.&quot;</td>
<td>Poorly lateralized. Insula or Secondary Sensory Area.</td>
</tr>
<tr>
<td>I.f. Autonomic Aura</td>
<td>&quot;I get goose bumps and my heart races.&quot; (No objective evidence of either autonomic activation nor a seizure.)</td>
<td>Poorly lateralized. Basal frontal region, ACC, and/or insula.</td>
</tr>
<tr>
<td>I.g. Abdominal Aura</td>
<td>&quot;I feel funny in my stomach, and it’s rising....&quot;</td>
<td>Poorly lateralized. Insula or superior bank of Sylvian fissure. Common in MTLE.</td>
</tr>
<tr>
<td>I.h. Psychic Aura</td>
<td>&quot;I get scared, like someone’s after me.&quot; (e.g., fear, déjà vu)</td>
<td>Fear -- amygdala. Déjà/jamais vu -- basal temporal lobe. Multisensory hallucinations -- temporal lobe.</td>
</tr>
<tr>
<td>II. Autonomic Seizures</td>
<td>&quot;I get goose bumps and my heart races.&quot; (Needs objective evidence of both autonomic activation and a seizure.)</td>
<td>(See Autonomic Aura.)</td>
</tr>
<tr>
<td>III. Dialectic Seizures (includes a “Typical Absence” subtype)</td>
<td>&quot;He just gets this blank stare -- it only lasts a short time.&quot;</td>
<td>Poorly localized (either diffuse cortex vs. both hippocampi).</td>
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<tr>
<td>Classification</td>
<td>Example of Symptoms</td>
<td>Lateral/Localization</td>
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<tr>
<td>IV. Motor Seizures</td>
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<td></td>
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<tr>
<td>a. Simple Motor</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IV.a.1. Myoclonic Seizure</td>
<td>&quot;I get this quick jerk, like before you go to sleep?&quot;</td>
<td>Likely primary motor; could be RAS.</td>
</tr>
<tr>
<td>IV.a.2. Clonic Seizure</td>
<td>&quot;I get this rhythmic tremor of my right hand.&quot;</td>
<td>Contralateral hemisphere. Likely primary motor.</td>
</tr>
<tr>
<td>IV.a.3. Tonic Seizure</td>
<td>&quot;Her right arm would just twist upwards, slowly.&quot;</td>
<td>Contralateral hemisphere. SSMA, RAS.</td>
</tr>
<tr>
<td>IV.a.4. Epileptic Spasms (&quot;salaam seizures&quot;)</td>
<td>&quot;He lurched forward with arms outstretched.&quot;</td>
<td>Variable.</td>
</tr>
<tr>
<td>IV.a.5. Tonic-Clonic Seizures</td>
<td>&quot;She had one of her Grand Mals....&quot;</td>
<td>Generalized by definition.</td>
</tr>
<tr>
<td>IV.a.6. Versive Seizures</td>
<td>&quot;His head and eyes turned slowly, like he was staring to the right and upwards....&quot;</td>
<td>Contralateral hemisphere. Primary motor area between the face and hand.</td>
</tr>
</tbody>
</table>
### Table 1. Continued.

<table>
<thead>
<tr>
<th>Classification</th>
<th>Example of Symptoms</th>
<th>Lateral/Localization</th>
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<tbody>
<tr>
<td>IV. Motor Seizures</td>
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<tr>
<td>b. Complex Motor</td>
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<tr>
<td>IV.b.1. Hypermotor</td>
<td>&quot;She was rolling her shoulders up and down.&quot;</td>
<td>Probably frontal or ACC.</td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IV.b.2. Automotor</td>
<td>&quot;He was picking at things and smacking his lips.&quot;</td>
<td>Temporal lobe epilepsies. Some evidence ACC.</td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td></td>
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<tr>
<td>IV.b.3. Gelastic</td>
<td>&quot;She laughs in this weird, rhythmic way. Always the same laugh.&quot;</td>
<td>Hypothalamic hamartoma. Also neocortical frontal and temporal.</td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
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<tr>
<td>V. Special Seizures</td>
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<tr>
<td>V.a. Atonic</td>
<td>&quot;He just goes limp. He's never gotten hurt doing it.&quot;</td>
<td>Possibly RAS with generalized epilepsy; can also occur in focal epilepsy.</td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>V.b. Astatic</td>
<td>&quot;He jerked back and fell flat on his face. He's been badly hurt several times.&quot;</td>
<td>Variable localization.</td>
</tr>
<tr>
<td>Classification</td>
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<td>------------------------</td>
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<tr>
<td>V.c. Hypomotor Seizures</td>
<td>&quot;My baby -- sometimes she just stops moving suddenly, staring off....&quot;</td>
<td>Variable localization.</td>
</tr>
<tr>
<td>V.d. Akinetic Seizures</td>
<td>&quot;My left hand will just stop working, and sometimes there's a left-facial twitch with it.&quot;</td>
<td>Contralateral hemisphere. Primary and supplementary negative motor areas.</td>
</tr>
<tr>
<td>V.e. Negative Myoclonic Seizures</td>
<td>&quot;When he's holding his hands up, sometimes they just go limp all of a sudden.&quot;</td>
<td>Poorly localizing.</td>
</tr>
<tr>
<td>V.f. Aphasic Seizures</td>
<td>&quot;He suddenly stops talking and looks really confused.&quot;</td>
<td>Dominant hemisphere, usually left. Cortical language areas.</td>
</tr>
</tbody>
</table>

SSMA - supplementary sensorimotor area; ACC - anterior cingulate cortex; MTLE - mesial temporal lobe epilepsy; RAS - reticular activating system
to be potentially epileptogenic and that is associated with the other zones. The Functional Deficit Zone is the volume of brain that shows functional deficits based on neuropsychological testing and/or functional neuroimaging. Finally, the Symptomatogenic Zone is the anatomical correlate of a seizure’s initial semiology. It is the part of the brain that an epileptic discharge stimulates to produce the patient’s initial symptoms (i.e., it is by definition an “eloquent” area of the brain, and never a “silent” area). We know much about “where the brain does what” thanks to a long history of carefully performed electrical cortical stimulation studies. While the Symptomatogenic Zone is usually not the EZ, it is usually nearby (Lüders and Noachtar 2000, Lüders et al. 2006).

Sometimes, semiology and its anatomically correlating Symptomatogenic Zone can be the main clue in deducing the likely area of the EZ, for example, in cases where a focus is too deep or small to be detected electrophysiologically. Or in another example, suppose one has an occipital lobe focal epilepsy, and because of the inferior longitudinal fasciculus (a white matter tract that rapidly connects the visual cortex with the temporal lobe), the EEG might show temporal spikes. The Irritative Zone, here, would be in the temporal lobe, yet the patient’s history of initially “seeing flashing colors” would better localize the EZ to the occipital lobe with almost 100% certainty (Loddenkemper and Kotagal 2005a).

Unfortunately, those working within the field of epilepsy today are confronted by two completely separate classification systems. This article covers the CEC’s classification of seizure semiology which is most analogous to the International League against Epilepsy’s (ILAE’s) current “Axis I: Ictal Phenomenology” (Engel and ILAE 2001). However, Axis I allows for subjectivity and no clear guidance about how to describe a seizure, merely referring readers to a long glossary (Blume et al. 2001), while the CEC provides a systematic way to organize one’s thoughts when witnessing and reporting a seizure. Overall, the differences between the two systems are many, and too detailed to review completely here. Suffice it to say that while the ILAE’s old system (ILAE 1985, ILAE 1989) contained too many ambiguities (e.g., the terms “simple,” “complex,” “cryptogenic,” and “partial” are no longer recommended), the ILAE’s new system (Engel and ILAE 2001, Engel 2006) is arguably too complex and not practical (Kellinghaus et al. 2004b, Loddenkemper et al. 2005b). The alternate CEC system presented here is arguably more intuitive and practical for epileptologists (Bautista and Lüders 2000, Bonelli et al. 2007, Hifranoglu et al. 2007, Kellinghaus et al. 2004b, Loddenkemper et al. 2005b, Lüders et al. 1998, Lüders et al. 1999, Lüders and Noachtar 2000, Lüders 2007, Rona et al. 2005).

WITNESS TO A SEIZURE – WHAT TO NOTE

Suppose one is a technologist recording an EEG, and a patient produces a sign that might be a seizure. What should one be aware of? EEG technologists know
already to look for and test responsiveness, awareness, language, memory, and motor function (Verdoorn-Markhorst et al. 2004). Doing this should give one all the information one needs to classify the seizure. An important point: although more than one symptom can manifest simultaneously, the classification is always based on the symptom that predominates over the others (Lüders and Noachtar 2000).

Generally, semiological phenomena have been divided into four possible “spheres”: the cognitive (subjective/sensory/experiential) sphere, the autonomic sphere, the consciousness sphere, and the motor sphere. In the CEC, there is a broadly defined seizure-type for each sphere. The cognitive sphere contains the auras, which are sensory or experiential seizures (e.g., visual flashes or déjà vu, respectively). The autonomic sphere contains the rare Autonomic Seizures, where objectively-determined autonomic signs are the predominant manifestation of the seizure. The consciousness sphere contains the Dialectic Seizures, where altered consciousness is the predominant manifestation of the seizure. The motor sphere contains the Motor Seizures, where motor signs are the predominate manifestation of the seizure. Motor Seizures are further subdivided into simple motor seizures and complex motor seizures – although here, the word “simple” means “simple” (that is, simple movements in one plane that do not seem like normal, natural movements), while “complex” means “complex” (that is, complex movements that seem coordinated and natural somehow). Finally, while there is one seizure-type for each of the four “spheres,” there is lastly a fifth seizure-type, designated “Special Seizures.” These include negative signs or symptoms as the predominant manifestation of a seizure (e.g., atonia or aphasia), as well as seizure phenomena that are impossible to classify due to a patient’s inability to communicate (e.g., infants or people with severe mental retardation).

If one is witnessing a seizure first-hand and suspects an aura, one asks the patient to verbalize what he or she is experiencing, possibly with leading questions from the behavior (e.g., if the patient is holding the stomach, is it an abdominal aura?; if the patient is cringing and holding an ear, is it auditory aura?, etc.). If one suspects an autonomic seizure, one could quickly assess for tachycardia, diaphoresis, piloerection (“goose bumps”), and flushing. If one suspects altered awareness, one confirms this by asking the patient to remember words, by asking the patient to speak, by determining if they are oriented and can follow simple commands, and by asking them to repeat and name. If one is witness to a motor seizure, one classifies the various motor patterns as detailed below.

WITNESS TO A SEIZURE – HOW TO REPORT

Now how would one report the seizure?

In classifying a seizure, one starts by deciding whether or not one believes the patient actually had a seizure, rather than something nonepileptic (psychogenic
nonepileptic seizure, a tremor or other movement disorders, narcolepsy or other sleep disorders, a transient ischemic attack, etc.). If one is not sure that a sign or symptom is a seizure, one classifies it in the broadest possible category as a “Paroxysmal Event” (see Figure 1). These two words simply and immediately communicate that one has witnessed a behavior, but that one is not confident that the behavior was a seizure. One should then qualify this and explain what features of the event led one to believe that it could have been one of the other possibilities on the differential diagnosis of epilepsy (Lüders and Noachtar 2000).

If, however, one feels confident that a patient has had a seizure, one then moves to a less broad, more specific seizure classification. See Figure 1 for a concise table of possible classifications: as one gets more specific in classifying a seizure, one keeps moving to the right of the figure. Table 1 is merely a more detailed look at the same figure, with examples and correlating Symptomatogenic Zone.

A caveat to reporting and classifying seizures: if one is reporting semiology based on history, rather than on direct observation, one must corroborate the patient’s history with a witness – especially in instances where there is loss of consciousness (when, by definition, the patient cannot tell first-hand what happened) – and note specifically who reported which detail.

CEC OF SEIZURE SEMIOLOGY

Auras

In the CEC, the cognitive sphere contains the auras, where a sensory or experiential symptom (e.g., visual or déjà vu, respectively) is the predominate manifestation of the seizure. Auras include somatosensory auras, visual auras, auditory auras, olfactory auras, gustatory auras, autonomic auras, abdominal auras, and psychic auras (Lüders and Noachtar 2000). Auras have been shown to be as useful as EEG in terms of their localizing value (Palmini and Gloor 1992, Fried et al. 1995).

Somatosensory auras include paresthesias (and rarely, pain) that are either: 1) limited to a strict dermatomal area, 2) bilateral and proximal, or 3) bilateral and distal. The Symptomatogenic Zone for these auras is felt to be either the primary somatosensory area (post-central gyrus, Brodmann’s areas 3, 1, and 2), the supplementary sensorimotor area, or the secondary sensory area, respectively (Lüders and Noachtar 2000). A unilateral aura is a reliable lateralizing sign to the contralateral hemisphere (Loddenkemper and Kotagal 2005a).

Visual auras usually consist of flashing colors, dark dots, or even blindness, and can be contained within one hemi-field. If so contained, they are very lateralizing, as well as localizing, to the contralateral primary visual cortex (Loddenkemper and Kotagal 2005a). If one sees faces or other complex visual hallucinations, the Symptomatogenic Zone is more likely to be a visual association area (Salanova et al. 1992).
FIG. 1. Cleveland Epilepsy Classification of Seizure Semiology. In classifying a seizure, one first determines if an event is likely a seizure of not, and then moves as far to the right of the figure as possible for increasing specificity. Evolution to a different seizure is connoted with an arrow, loss of consciousness by including ("LOC"), and any pertinent somatotopic qualifiers should be included (left, right, arm, face, etc.). See text for details.

Auditory auras are rare, usually consist of a buzzing noise, and usually correlate with Heschl's gyrus and the planum temporale (Liegoois-Chauvel et al. 1991). They are often hard to lateralize, as they are often bilateral thanks to the multiple bilateral pathways of the auditory system. More complex auditory hallucinations like voices are extremely rare, and should raise the suspicion of schizophrenia or some other etiology (Lüders and Noachtar 2000).
While olfactory auras (usually offensive smells, but not always) are popularly thought of as common in temporal lobe epilepsy, they are actually fairly rare. The amygdala is the likely Symptomatogenic Zone for these seizures (Acharya et al. 1998, Lüders and Noachtar 2000). However, gustatory auras – which are usually disagreeable tastes – probably signify excitation of the insula and its surrounding opercula (Hauser-Hauw and Bancaud 1987).

If there is no objective evidence of autonomic excitation (tachycardia, piloerection, flushing, diaphoresis, etc.), and a patient claims that these signs are associated with his seizure, then the event is called an autonomic aura. The basal frontal, anterior cingulate, and insular cortices have been implicated as possible Symptomatogenic Zones for this aura (Wannamaker 1985, Lüders and Noachtar 2000). An abdominal aura can be thought of as a sub-set of autonomic auras, but given its own status due to its high prevalence in mesial temporal lobe epilepsy. It is usually a feeling of nausea or unpleasantness in the epigastric area, and tends to rise up into the chest (Lüders and Noachtar 2000). An abdominal aura localizes to temporal lobe epilepsy (TLE) in nearly three-fourths of cases and if it evolves into an Automotor Seizure (see below), the patient is 98.3% certain to have a TLE (Henkel et al. 2002).

Finally, if a patient ever has a “multi-modal sensory hallucination” – that is, one that includes more than one sensation, or one that is more like an “experience” than a sensation (like fear, déjà vu, jamais vu, etc.) – then the seizure is termed a psychic aura, and the Symptomatogenic Zone is not well-defined although fear has been associated with the amygdala and déjà vu with the basal temporal lobe (Bancaud et al. 1994).

Autonomic Seizures

The autonomic sphere contains the rare Autonomic Seizures. Unlike the autonomic aura, an Autonomic Seizure is classified if there is objective evidence of autonomic excitation (tachycardia, piloerection, flushing, diaphoresis, etc.) concurrent with objective evidence of a seizure. As with the aura, the basal frontal, anterior cingulate, and insular cortices have been implicated as possible Symptomatogenic Zones for this seizure (Wannamaker 1985, Lüders and Noachtar 2000).

Some have critiqued the CEC for recommending “appropriate monitoring” to diagnose Autonomic Seizures, something one should not need in a purely semiologic classification (Arzimanoglou and Aicardi 2000). It should be noted that one is not forbidden from using an EEG to help decide if a paroxysmal event is epilepsy or not. But if one objectively documents tachycardia or piloerection, for example, well before an objective loss of consciousness and then a generalized seizure, one would be right to classify this as an Autonomic Seizure regardless of an EEG (Lüders 2007). But being so rare, a prudent diagnostician would likely classify
these phenomena as “autonomic auras” unless there is the hardest of objective evidence of both autonomic activity and a corresponding seizure.

Dialeptic Seizures

The consciousness sphere contains the Dialeptic Seizures, where altered consciousness is the primary manifestation of the seizure. “Dialeptic” from the Greek “dialeipein” meaning to interrupt, stand still, or pass out was coined (Lüders and Noachtar 2000) to avoid confusion about whether one was dealing with “absence” or “complex partial seizures.” In other words, one can describe the seizure semiology regardless of whether or not one has an EEG or knows that a seizure is generalized or focal, respectively. “Loss of consciousness” (LOC) is a tricky subject in neurology, but in epilepsy, it generally means a state of unresponsiveness during which memories are unattainable. One should be sure to check that one is not actually dealing with an aphasia (if the patient is not speaking), with an apraxia or atonia (if the patient is not obeying commands), or with a patient who is being distracted by a hallucination (Gloor 1986).

There is an even more specific seizure-type that can be described in this classification, known as the Typical Dialeptic Seizure. These are short in duration (about ten seconds), start and stop abruptly, and might have associated 3 Hz blinking. Some have critiqued this classification for hinting at pathophysiology in a supposedly pure semiologial classification system (Arzimanoglou and Aicardi 2000). But the Typical Dialeptic Seizure classification is still based purely on a semiologic description – a Dialeptic Seizure with a more specific length of time and manner of onset and offset. While it is true that these seizures are usually associated with absence epilepsy and likely a faulty thalamocortical circuitry (Snead 1995), they do not have to be. Typical Dialeptic Seizures have been described in focal epilepsies (Grosso et al. 2005). Nonetheless, in determining the diagnosis (i.e., the determination of the Epilepsy Syndrome), the semiology of a short, abrupt Dialeptic Seizure – as opposed to a long Dialeptic Seizure that gradually evolves into an Automotor Seizure, for example – would be considered very valuable information to most epileptologists, and thus deserving of a semiological subclassification.

Motor Seizures – Simple Motor Seizures

The motor sphere includes any seizure that has a predominate motor manifestation, and includes the Motor Seizures, which in the CEC are subdivided into Simple Motor Seizures and Complex Motor Seizures. Simple Motor Seizures, as their name implies, consist of simple, stereotyped movements that are unnatural, and contained within one plane when there are repetitive movements. The rhythm,
duration of movement, and involved muscles determine the more-specific seizure-type. Several Simple Motor Seizures have excellent lateralizing value to the contralateral hemisphere, including versive seizures, and unilateral clonic or tonic seizures (Loddenkemper and Kotagal 2005a). These common seizures occur less frequently in the elderly (Kellinghaus et al. 2004a).

Motor Seizures include the short myoclonic seizures (less than 200 milliseconds) which are shock-like “jerks” similar to the physiologic hypnagogic jerk people sometimes do while falling asleep. There are also clonic seizures which are stereotyped myoclonic jerks that repeat every one to two seconds. Both of these seizures likely correspond to a Symptomatogenic Zone within the primary motor cortex (Lüders and Noachtar 2000).

If the movement and muscle contraction is sustained for over five seconds, it is then called a tonic seizure. The Symptomatogenic Zone for these seizures is likely the supplementary sensorimotor area (SSMA), and if it is a unilateral tonic seizure, it has excellent lateralizing value to the contralateral SSMA (Werhahn et al. 2000). Epileptic spasms are also known as “salaam seizures” due to the posture of a flexed trunk with abducted and elevated arms. These are no longer referred to as “infantile spasms,” since infants are not the only age group to have these seizures. The Symptomatogenic Zone is variable, although there is some evidence that these seizures have a subcortically-mediated mechanism, with input from an abnormally organized cortex (Pranzatelli 2002).

Tonic-clonic seizures are by definition generalized, with arms and legs outstretched for a short period of tonicity (five to ten seconds) before evolving into the clonic phase; the whole seizure usually lasting about a minute. They can be seen in grand mal epilepsy and can also secondarily generalize from a focal epilepsy (Theodore et al. 1994). Lateral tongue biting is highly specific (99%) for a generalized tonic-clonic seizure (Benbadis et al. 1995).

Finally, there is the versive seizure, which is used to describe a forced, sustained, unnatural deviation of the head and eyes to one side, usually causing the patient to look up and back in a very unusual, unnatural manner. This seizure also likely corresponds to the primary motor area, and is a highly valuable lateralizing sign (Wyllie et al. 1986, Loddenkemper and Kotagal 2005a, Lüders and Noachtar 2000).

Motor Seizures – Complex Motor Seizures

There are three specific types of Complex Motor Seizures, which are seizures that involve complex, natural movements. If the movements are proximal and bilateral, like pedaling a bike or shrugging one’s shoulders, they are called hypermotor seizures. Because these seizures involve large muscle groups, they can appear violent. Because patients can be conscious with these seizures, they are often mistaken for a psychiatric disorder. The Symptomatogenic Zone is likely the frontal...
lobes or cingulate cortex, with possible insular involvement in nocturnal hypermotor seizures (Ryvlin et al. 2006).

If the motor activity is distal or oral, such that the patient is performing manual “automatisms” (e.g., rubbing fingers, picking, licking one’s lips), then the seizure is called an automotor seizure. These might localize to anterior cingulate activity. If there is preserved consciousness with the Automotor Seizure, it is a good lateralizing sign to the nondominant hemisphere (Ebner et al. 1995, Loddenkemper and Kotagal 2005a). The unilateral dystonic posturing sometimes seen in an automotor seizure is also an excellent lateralizing sign, to the contralateral hemisphere (it is better to note this than the hand that conducts the automatisms) (Loddenkemper and Kotagal 2005). Finally, there are the gelastic seizures, which consist of inappropriate and unusual laughter. These are frequently associated with hypothalamic hamartomas, although the anterior cingulate or basal temporal lobe could also be implicated (Arroyo et al. 1993, Lüders and Noachtar 2000).

Special Seizures

If one has “negative symptoms” (e.g., atonia, aphasia, etc.), the CEC classifies them as Special Seizures. Atonic seizures manifest as loss of tone, and patients fall, but usually do not hurt themselves. The Symptomatogenic Zone is possibly the reticular formation in the brainstem. Conversely, astatic seizures (“drop attacks”) usually are associated with injury, and these patients often need to wear a protective helmet. The astatic seizure is often a brief myoclonic jerk that creates instability, followed by an astatic or tonic seizure that knocks the patient down (Lüders and Noachtar 2000). Akinetic seizures are possibly apraxic seizures with inability to perform learned motions of the distal muscles, and a likely Symptomatogenic Zone of the negative motor areas (Noachtar and Lüders 1999). Negative myoclonic seizures are brief episodes of atonia. These seizures are poorly localizing, though some evidence shows involvement of the motor areas, the primary sensory area, as well as subcortical mechanisms (Rubboli and Tassinari 2006).

While the previous have been negative motor seizures, the aphasic seizures are negative seizures of language (possibly motor, if primarily Broca’s expressive area is involved). Aphasic seizures can consist of a global, expressive, or receptive aphasia, and the patient has to be awake in order to be classified. The Symptomatogenic Zone is likely to be one of the three language areas: Broca’s, Wernicke’s, and/or the basal temporal language area, which are highly interconnected (Matsumoto et al. 2004). Postictal aphasia is an extremely reliable lateralizing sign to the dominant hemisphere, and conversely, ictal speech is a decent lateralizing sign to the nondominant hemisphere (Gabr et al. 1989, Loddenkemper and Kotagal 2005a). Finally, there is the hypomotor seizure, which consists of an arrest of movement in a person who is not able to communicate why the movement stopped (e.g., an infant
or a person with severe mental retardation). These seizures can be focal or generalized (Källén et al. 2002). This seizure-type is useful in the pediatric setting, helping to able the CEC to classify nearly all infantile seizures (Acharya et al. 1997).

ADDITIONAL “QUALIFIERS” WHEN REPORTING SEIZURES

Evolving Seizure Signified with Arrows

Seizures often evolve into other predominant seizure-types. In the CEC, this is signified simply by using an arrow to separate the previous from the current seizure. Example:
Abdominal Aura → Clonic Seizure → Generalized Clonic Seizure

Loss of Consciousness (LOC)

Loss of consciousness, being extremely important in patients’ quality of life, should be included if applicable in any classification. It is noted with “(LOC)” immediately after the seizure during which it occurred. The following seizures are by definition associated with a loss of consciousness and thus do not need “(LOC)” due to redundancy: dialeptic seizures and generalized tonic-clonic seizures. Using this qualifier, the above seizure example might be better classified as:
Example:
Abdominal Aura → Clonic Seizure (LOC) → Generalized Clonic Seizure

Somatotopic Attributes

When classifying a seizure, one prefaces the seizure-type with as many somatotopic attributes as one can, for maximum specificity. Attributes one can use include the following:

1. Left, right, or bilateral;
2. Face, eyes, mouth, head, arm, hand, leg, or foot;

The following seizures have no somatotopic attribute by definition: olfactory, gustatory, abdominal, or psychic auras; dialeptic seizures; and complex motor seizures. Finally, then, in the above seizure example, the most specific and best classification could be:
Example:
Abdominal Aura → Left Arm Clonic Seizure (LOC) → Generalized Clonic Seizure
SUMMARY

This article has reviewed the systematic way in which many now classify seizure semiology. Like everything in medicine and science, the CEC's classification of seizure semiology might change over time as new information develops. However, the whole system shouldn't need to be overhauled very frequently, since its primary focus is on semiology, which is universal and is a fairly "closed set" when grouped into broad classifications. This is opposed to the ILAE's system of classifying seizure-types, where the healthcare worker does not classify purely on semiology, but needs electroneurodiagnostic and/or pathophysiologic test results. This leads to a theoretically "infinite set" of seizure-types and could change rapidly, thus needing frequent updating. It should be noted that the CEC's semiological classification has been useful and nearly constant for almost twenty years, while the ILAE's system has been frequently changed, and arguably has changed into a system that is more burdensome and impractical (Kellinghaus et al. 2004b, Loddenkemper et al. 2005b, Lüders 2007).

As mentioned above, the CEC of seizure semiology is part of a larger five-dimensional, patient-based classification system (not reviewed here) that includes Syndrome, Semiology (including localizing and lateralizing information), Etiology, Seizure Frequency, and Related Medical Conditions (Loddenkemper et al. 2005b). The semiology classification has been successfully employed by epileptologists throughout the world for nearly two decades. It can be used in pediatric and adult settings, as well as in cases of status epilepticus (Bautista and Lüders 2000, Bonelli et al. 2007, Hırfanoglu et al. 2007, Kellinghaus et al. 2004b, Loddenkemper et al. 2005b, Lüders et al. 1998, Lüders et al. 1999, Lüders and Noachtar 2000, Lüders 2007, Rona et al. 2005). Those not in the field of epilepsy will likely find semiology fascinating nonetheless, since it is essentially the study of functional neuroanatomy.

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