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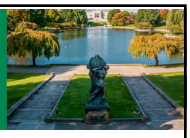
Cleveland Clinic Epilepsy Update and Review Course

Live-streamed September 21-24, 2020 | 5:00-9:30 PM (EDT)



# Classical Electroclinical Features of Partial (surgically amenable) Epilepsies

*Andreas V. Alexopoulos, MD, MPH*
*Cleveland Clinic Epilepsy Center*
*Wednesday, September 23rd 2020*

## Outline

1. Introduction & conceptual approach
2. Anatomical divisions and functions
3. Semiology and electroclinical correlations
4. Selected References
5. Pitfalls
6. Conclusions

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39% Frontal

76% Extra-temporal

25% Parietal

24% Temporal

13% Occipital

Left lobar surface area ($cm^2$) for automatic and manual parcellation and SI between 2 parcellated regions with 10 cortical surfaces

| Region | Automatic parcellation | Manual parcellation | SI |
|---|---|---|---|
| Frontal | 387.86 ± 27.12 | 384.73 ± 29.96 | 0.98 ± 0.003 |
| Temporal | 245.06 ± 20.63 | 238.50 ± 21.06 | 0.95 ± 0.015 |
| Parietal | 235.18 ± 18.58 | 245.12 ± 21.58 | 0.93 ± 0.018 |
| Occipital | 139.12 ± 8.66 | 127.59 ± 12.06 | 0.91 ± 0.047 |

Cerebral Cortex September 2008;18:2181-2191
doi:10.1093/cercor/bhm244
Advance Access publication January 29, 2008

**Brain Size and Cortical Structure in the Adult Human Brain**

Kiho Im[1], Jong-Min Lee[1], Oliver Lyttelton[2], Sun Hyung Kim[1], Alan C. Evans[2] and Sun I. Kim[1]

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# Selected Concepts

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## Conceptual Approach

- Concept 1: Limitations of terminology
- Concept 2: Chronology
- Concept 3: Locational descriptions of cortex
- Concept 4: The beginnings of the seizure

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## Concept 1: The terminology "trap"

**Possible Seizure Classifications Could be Based On:**

| Pathophysiology | Anatomy | Networks | Practical, by: | Modify Existing |
|---|---|---|---|---|
| But this is currently impossible with our limited understanding | Temporal<br>Frontal<br>Parietal<br>Occipital<br>Diencephalic<br>Brainstem | Neocortical<br>Limbic<br>Thalamo-Cortical<br>Brainstem | AED response<br>Surgical target<br>Disabling<br>EEG pattern<br>Many others | 1981 ILAE System<br>2010 ILAE update |

- In the absence of fundamental knowledge, ILAE chose to extend the existing classification
- The is an operational (practical) system, not a true scientific classification
- Others might devise special operational classifications for specific use, e.g., neonatal, ICU
- This classification is predominantly for clinicians

The 2017 ILAE Classification of Seizures

Robert S. Fisher, MD, PhD

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## ILAE 2017 Classification of Seizure Types Expanded Version

**Focal Onset**

Aware | Impaired Awareness

**Motor Onset**
- automatisms
- atonic[2]
- clonic
- epileptic spasms[2]
- hyperkinetic
- myoclonic
- tonic

**Non-Motor Onset**
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic-clonic

**Generalized Onset**

**Motor**
- tonic-clonic
- clonic
- tonic
- myoclonic
- myoclonic-tonic-clonic
- myoclonic-atonic
- atonic
- epileptic spasms[2]

**Non-Motor (absence)**
- typical
- atypical
- myoclonic
- eyelid myoclonia

**Unknown Onset**

**Motor**
- tonic-clonic
- epileptic spasms

**Non-Motor**
- behavior arrest

**Unclassified[3]**

[1] Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms.

[2] These could be focal or generalized, with or without alteration of awareness

[3] Due to inadequate information or inability to place in other categories

From *Fisher et al. Instruction manual for the ILAE 2017 operational classification of seizure types. Epilepsia doi: 10.1111/epi.13671*

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## *Wording changes*

| OLD TERM | NEW TERM |
|---|---|
| | |
| Unconscious (still used, not in name) | Impaired awareness (surrogate) |
| Partial | Focal |
| Simple partial | Focal aware |
| Complex partial | Focal impaired awareness |
| Dyscognitive (word discontinued) | Focal impaired awareness |
| Psychic | Cognitive |
| Secondarily generalized tonic-clonic | Focal to bilateral tonic-clonic |
| Arrest, freeze, pause, interruption | Behavior arrest |

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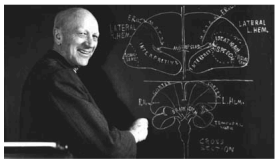
## Concept 2: Chronology & Syntaxis



'It is always necessary to consider the whole seizure pattern before reaching a conclusion as to the value of any element in that pattern' (Penfield 1954)

'Bancaud regarded the concept of order and sequence of semiological elements as crucial, and found analogies with the syntactic organization of words in a meaningful sentence' (Chauvel 2001)

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## *History & Video analysis*

1. Prior to seizure onset
2. Seizure onset
3. Seizure presentation & evolution
4. Postictal symptoms

*Loddenkemper & Kotagal, 2005*

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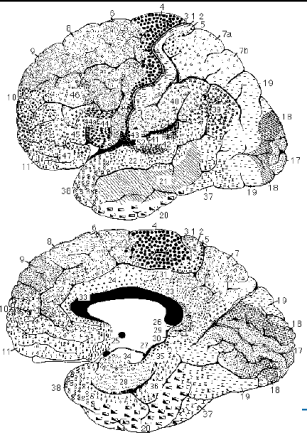
## Concept 3: *Organization of the cerebral cortex*



*Brodmann areas >100 years later*

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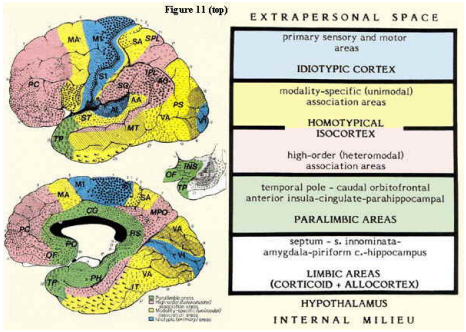


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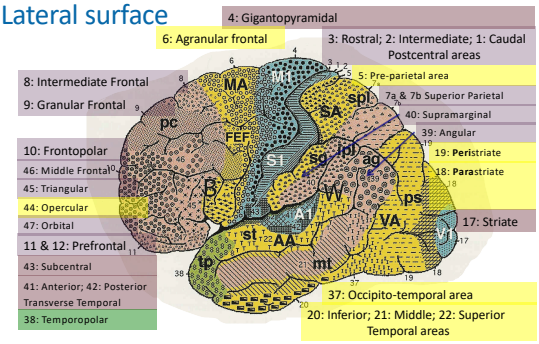
*M.-Marsel Mesulam 1985*

## Locational descriptions of human Brodmann areas
### Lateral surface

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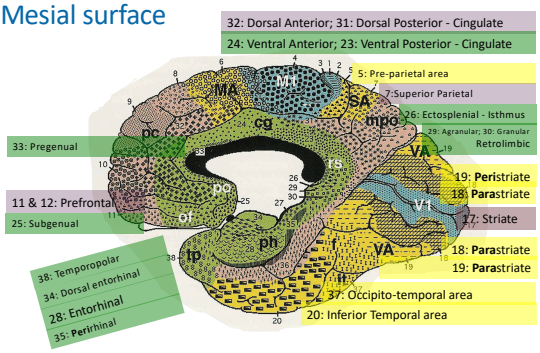
*M.-Marsel Mesulam 1985*

## Locational descriptions of human Brodmann areas
### Mesial surface

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*M.-Marsel Mesulam 1985*

# *Concept 4: Auras*

# *the starting point*

*"the beginning of the sentence"*

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## *Aura is a symptom, not a sign*

- Auras consist exclusively of **subjective sensations**
- No signs visible by the observer except perhaps the "voluntary" reactions the patient may have
- Auras are usually *of short duration* (seconds), occur *at the beginning* of a seizure, and are the result of epileptic activation of *a limited cortical region*

"***aura***" = *breath; wind* (*in Greek*)

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## *Epileptic Auras*

| | |
|---|---|
| • Somatosensory* | • Abdominal |
| • Visual* | • Olfactory |
| • Auditory* | • Gustatory |
| • Autonomic* | • Psychic |
| *Left/Right | No somatotopic distribution |

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## The localizing value of auras in partial seizures:
### A prospective and retrospective study

André Palmini, MD, and Pierre Gloor, MD, PhD

**Article abstract**—Doubts concerning the localizing significance of auras in partial seizures have recently been expressed. Prompted by this, we studied this issue by re-examining two groups of patients: the first, studied retrospectively, consisted of patients in whom the site of origin of the seizures was known beyond a reasonable doubt; the second, studied prospectively, comprised patients in whom specific auras were correlated with the localization of interictal epileptiform EEG abnormalities and the final diagnostic impression. The data from the retrospective series were suitable for rigorous statistical analysis. The two groups yielded similar results: the frequency of auras in partial seizures and the localizing significance of those for which large enough numbers could be collected was high. We conclude that the type of aura, when elicited by careful history-taking, provides as useful localizing, but often not lateralizing, information as the EEG and modern high-technology procedures such as CT, MRI, and PET.

NEUROLOGY 1992;42:0000-0000

…In this era of high-technology investigative procedures commonly used in the diagnostic work-up of patients with intractable seizures considered for surgical therapy, ***careful history-taking*** has lost none of its diagnostic power and yields reliable **localizing** (albeit, often not lateralizing) evidence

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## *Auras with high localizing value*

- **Elementary** Visual hallucination
- **Elementary** Auditory hallucination

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## Autonomic manifestations

- **Cardiac**: Tachy-, Brady-cardia / asystole, arrhythmia
- **Respiratory**: Tachy-, Brady-pnea / Hyper-pnea
- **Vasomotor**: flushing, pallor, cyanosis, piloerection
- **Pupillary**: mydriasis, miosis, hippus
- **Sexual arousal**: penile erection, orgasm
- **Glandular**: perspiration, lacrimation, bronchial secretion
- **Loss of bladder control**

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## Autonomic Auras: Overlap between Central Autonomic Networks & Epileptic regions

- **Insular cortex**
- **Amygdala**
- **Prefrontal / Cingulate cortex**
- **Hypothalamus**

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## Electrical stimulation of the brain

- **Experimentally**, heart rate (HR) changes, including bradycardia, tachycardia, and even asystole, have been repeatedly provoked by electrical **stimulation of the limbic system and insular cortex**.
- **Cortical limbic structures** are actually thought to be the principal mediators of autonomic functions, with candidate areas including the **cingulate gyrus, insula and orbitofrontal cortex**.

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## Epilepsy arising from the Temporal Lobe

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## *The Focal Aware Seizures*

## *Auras of Temporal Lobe origin*

NEUROLOGY PERSPECTIVE

**Tales from the Temporal Lobes**

Adam Zeman, D.M. *NEJM 2005*

***a.k.a. "Simple Partial"***

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## Mesial TLE: Anatomoclinical correlations

| Symptoms & Signs | Frequency |
|---|---|
| Auras | **1. Epigastric**; Visceral 79%<br>**2. Fear** 37%<br>**3. Déjà vu**, déjà vecu, dreamy state 29% |
| Loss of contact | Very frequent 71% |
| Gestural Automatisms | Frequent 67% |
| Oroalimentary Automatisms | Frequent 62.5% |
| Duration >1 min | Almost always 96% |
| Secondary Generalization | Exceptional or very rare in most patients |

*Maillard L et al. Epilepsia 2004*

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## Auras of TLE *(in order of prevalence)*

- ***Epigastric (visceral) aura***: rising abdominal sensation
- ***Other Autonomic auras***
- ***Experiential (psychic) auras***: fear, affective, dysmnesic etc.
- ***False feeling of familiarity dèjá vu*** (already seen) ***or unfamiliarity jamais vu*** (never seen) and their variations
- **Auditory** hallucinations & illusions
- **Olfactory & Gustatory** hallucinations
- **Other symptoms**

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*Epilepsia*, **45**(12):1590–1599, 2004
Blackwell Publishing, Inc.


Semiologic and Electrophysiologic Correlations in Temporal Lobe Seizure Subtypes

*†Louis Maillard, †Jean-Pierre Vignal, *Martine Gavaret, *Maxime Guye, ‡Arnaud Biraben, *Aileen McGonigal, *Patrick Chauvel, and *Fabrice Bartolomei

*Service de Neurophysiologie Clinique, Hôpital de la Timone, Inserm EMI 99-26, Université de la Méditerranée, Marseille; †Service de Neurologie, Centre Hospitalier Universitaire, Université Henri Poincaré, Nancy, and ‡Service de Neurologie, Centre Hospitalier Universitaire, Rennes, France

*Epilepsia*, **45**(6):695–714, 2004
Blackwell Publishing, Inc.


ILAE Commission Report

Mesial Temporal Lobe Epilepsy with Hippocampal Sclerosis

Compiled by Heinz-Gregor Wieser for the ILAE Commission on Neurosurgery of Epilepsy

*Department of Neurology, University Hospital Zurich, Zurich, Switzerland*

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## "Mesial temporal lobe epilepsy"

A specific syndrome of **TLE associated with hippocampal sclerosis** (mTLE/HS)

- a strong association with *antecedent of febrile seizures*
- a *progressive development* that leads frequently to *drug resistance*
- a clinical picture of *fairly homogeneous seizures*
- *a topographic distribution of interictal and ictal EEG* abnormalities, which tend to be focused around the anterior and basal TL regions.
- *Neuropsychological* and *functional neuroimaging* data that also point to the mesial temporal lobe structures

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## Scalp EEG in mesial TLE

- **Pure epileptic generators confined within the mesial temporal lobe have a poor contribution to scalp EEG and are not visible on routine visual analysis of EEG records**
- **There are no EEG changes at the very beginning of a seizure**
- Earliest change on surface EEG consists of **regional attenuation** (flattening) involving the anterior temporal electrodes, which has a strong lateralizing value and **corresponds to the initial subjective sensation prior to the emergence of automatisms**
- This is followed by **a regional temporal rhythmic theta** which reflects propagation to the basal & lateral temporal neocortex and is **usually (but not always) ipsilateral** to the side of onset

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# *The Focal Impaired Awareness seizures*

***a.k.a. "Complex Partial seizures"***

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## Objective ictal signs of TLE *(in order of prevalence)*

*usually, but not always, occur when consciousness is impaired*

- ***Automatisms***
- ***Autonomic changes***: tachycardia, mydriasis, salivation
- ***Speech disturbances***: speech arrest, ictal speech
- ***Dystonic postures***
- ***Motor arrest with staring***
- **Head & eye deviation**
- **Unilateral** eyeblinking
- **Other signs**

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## Automatism: definition

- Involuntary inappropriate performance of non-reflex act

***(a) Perseverative***: semi-purposeful continuation of ongoing activity

***(b) de novo***: Occurrence of a new activity

- May take the form of seemingly meaningful acts
- May sometimes be modified by external stimuli
- Usually patient cannot recall the event
- Can occur during ictal and post-ictal phases
- "***uncontrollable, physiological motor patterns occurring at the wrong place and/or time***"

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## Automatisms

- ***Oroalimentary***: swallowing, chewing, smacking/ licking lips
- ***Gestural***: fumbling, scratching, rubbing, picking
- **Phonation** (unarticulated sounds: grunting, moaning)
- **Vocalization** (articulated sounds)
- **Verbalization** (words or complete sentences)
- **Ambulatory** (walking about)

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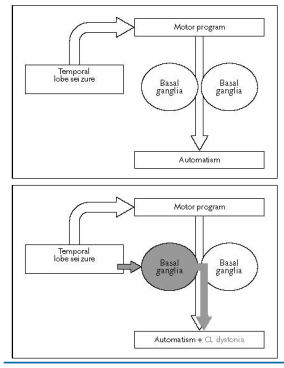
## *Hand Automatisms & Dystonia in TLE*

*(a theoretical view)*




*Vercueil; Epileptic Disord 2002*

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## Stereotyped motor behaviors *(ictal or peri-ictal)*

- **Dystonic limb posturing**
- Unilateral tonic posturing
- Unilateral immobile limb
- Eye motor manifestations
- Head turning & version
- Facial alterations
- Nosewiping or nose-rubbing

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Question 2

## Causes of Temporal Lobe surgery failures

- Incomplete resection **within** the Temporal lobe
- **Bilateral** Temporal lobe epilepsy
- Temporal-plus (**Multi-lobar**): incomplete resection in proximity to the TL "neighboring networks" i.e T-**Frontal**; T-**Sylvian-Insular**; T-Parieto-Occipital
- Pseudo-Temporal (**Extra-Temporal** epilepsies)

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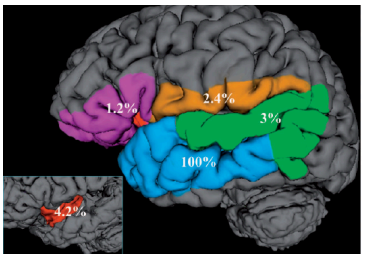
doi:10.1093/brain/aww372 BRAIN 2016: 139; 444–451 | 444

## Temporal plus epilepsy is a major determinant of temporal lobe surgery failures

Carmen Barba,[1] Sylvain Rheims,[2,3,4] Lorella Minotti,[5] Marc Guénot,[6] Dominique Hoffmann,[7] Stephan Chabardès,[7] Jean Isnard,[2] Philippe Kahane[5,8] and Philippe Ryvlin[3,4,9]

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## Features of TL vs. TL-plus epilepsies

| Symptoms main categories | Subcategories | Relative frequency of ictal events(%) TL group | T+ group | P |
|---|---|---|---|---|
| seizure duration (>1 min) | | 78 | 73.9 | 0.7 |
| consciousness impairement ability | | 94.9 | 100 | 0.27 |
| to warn at seizure onset aura # | | 78 | 43.5 | **0.003** |
| none | | 15.3 | 8.7 | 0.4 |
| somatosensory | (Ipsilateral/contralateral/bilateral) | 16.9 (11.9/15.3/11.9) | 26.1 (8.7/26.1/26.1) | 0.3 (ns/ns/ns) |
| visual | (Illusions/hallucinations) | 6.8 (5.1/1.7) | 13 (13/4.3) | 0.36 (0.02/ns) |
| auditory | (Illusions/hallucinations) | 3.4 (0/3.4) | 8.7 (8.7/4.3) | 0.32 (ns/ns) |
| olfactory | (Hallucinations) | 1.7 | 4.3 | 0.48 |
| gustatory | (Hallucinations) | 5.1 | 21.7 | **0.02** |
| vestibular | (Rotatory/not rotatory) | 1.7 (0/1.7) | 13 (8.7/4.3) | **0.03** (**0.02**/ns) |
| dysmnesic | (Familiarity illusion/memory flashback/dreamy state) | 5.1 (0/3.4/1.7) | 13 (0/8.7/4.3) | 0.2 (ns/ns/ns) |
| emotional | (Fear/anxiety/anger/pleasure) | 33.9 (22/11.9/3.4/0) | 39.1 (13/21.7/4.3/0) | 0.6 (ns/ns/ns/ns) |
| other psychic | (Forced thinking/distorsion of reality/urge to move) | 3.4 (0/3.4/0) | 4.3 (0/4.3/0) | 0.8 |
| cephalic | | 6.8 | 4.3 | 0.8 |
| digestive | (Throat/chest/abdomen) | 71.2 (16.9/54.2/6.8) | 43.5 (13/30.4/4.3) | **0.02** (ns/ns/0.05) |
| uro-genital | | 1.7 | 4.3 | 0.48 |

doi:10.1093/brain/awm108 *Brain* (2007), **130**, 1957–1967

lctal clinical and scalp-EEG findings differentiating temporal lobe epilepsies from temporal 'plus' epilepsies

C. Barba,[1,2] G. Barbati,[3] L. Minotti,[4] D. Hoffmann[5] and P. Kahane[4]

ORIGINAL CONTRIBUTION

ONLINE FIRST

## Cingulate Epilepsy

*Report of 3 Electroclinical Subtypes With Surgical Outcomes*

*Rafeed Alkawadri, MD; Norman K. So, MD; Paul C. Van Ness, MD; Andreas V. Alexopoulos, MD, MPH*

**Importance:** The literature on cingulate gyrus epilepsy in the magnetic resonance imaging era is limited to case reports and small case series. To our knowledge, this is the largest study of surgically confirmed epilepsy arising from the anterior or posterior cingulate region.

**Objective:** To characterize the clinical and electrophysiological findings of epilepsies arising from the anterior and posterior cingulate gyrus.

**Design, Setting, and Participants:** We studied consecutive cingulate gyrus epilepsy cases identified retrospectively from the Cleveland Clinic and University of Texas Southwestern Medical Center epilepsy databases from 1992 to 2009. Participants included 14 consecutive cases of cingulate gyrus epilepsies confirmed by restricted magnetic resonance image lesions and seizure freedom or marked improvement following lesionectomy.

**Main Outcomes and Measures:** The main outcome measure was improvement in seizure frequency following surgery. The clinical, video electroencephalography, neuroimaging, pathology, and surgical outcome data were reviewed.

**Results:** All 14 patients had cingulate epilepsy confirmed by restricted magnetic resonance image lesions and seizure freedom or marked improvement following lesionectomy. They were divided into 3 groups based on anatomical location of the lesion and corresponding seizure semiology. In the posterior cingulate group, all 4 patients had electroclinical findings suggestive of temporal origin of the epilepsy. The anterior cingulate cases were divided into a typical (Bancaud) group (6 cases with hypermotor seizures and infrequent generalization with the presence of fear, laughter, or severe interictal personality changes) and an atypical group (4 cases presenting with simple motor seizures and a tendency for more frequent generalization and less-favorable long-term surgical outcome). All atypical cases were associated with an underlying infiltrative astrocytoma.

**Conclusions and Relevance:** Posterior cingulate gyrus epilepsy may present with electroclinical findings that are suggestive of temporal lobe epilepsy and can be considered as another example of pseudotemporal epilepsies. The electroclinical presentation and surgical outcome of lesional anterior cingulate epilepsy is possibly influenced by the underlying pathology. This study highlights the difficulty in localizing seizures arising from the cingulate gyrus in the absence of a magnetic resonance image lesion.

*JAMA Neurol. Published online June 10, 2013. doi:10.1001/jamaneurol.2013.2940*

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## *Pseudotemporal epilepsy: another scenario*
## *Seizures arising from posterior cingulate*

| Patient | Sex | Handedness | Age, y | | Lesion | Generalization | Frequency/mo | Aura | Prominent Phase by Video EEG | Other Clinical Features | Resection | Pathology | Outcome (Engel) | Follow-up, y | Medication |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | Onset | Evaluation | | | | | | | | | | | |
| 11 | F | R | 13 | 18 |  | About 2 in 3 | 10 | Depersonalization | L hand automatisms > R version > generalized tonic-clonic | Early generalization (< 15 s) | Lesionectomy | Gliosis | I | 4 | No |
| 12 | M | R | 15 | 16 |  | About 2 in 3 | 6 | Falling, gustatory, abdominal | Bilateral asymmetric tonic | | Lesionectomy + adjacent frontal cortex | Astrocytoma vs cortical dysplasia | I | 11 | No |
| 13 | M | R | 9 | 13 |  | Rare | 60 | Déjà vu, jamais vu, abdominal | Auras with frightened look or dialeptic | | Lesionectomy | Low-grade astrocytoma | I | 8 | Yes |
| 14 | F | R | 19 | 26 |  | About 1 in 3 | 3 | Déjà vu | R version > generalized tonic-clonic or dialeptic | Early generalization (< 15 s) | Lesionectomy | Cortical dysplasia | I | 2 | Yes |

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# Epilepsy arising from the Frontal Lobe

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## FLE: the next frontier

Clin Electroencephalogr. 1998 Oct;29(4):163-9.

**Frontal lobe epilepsy: the next frontier.**

Niedermeyer E.

Department of Neurology, Johns Hopkins University School of Medicine and Hospital, Baltimore, Maryland, USA.

Abstract

Frontal lobe epilepsy, the great new epileptological challenge, presents enormous difficulties that still preclude a more profound understanding at the present time. The major subdivision of the frontal lobe into a prefrontal and premotor portion is the first step toward a better and yet limited comprehension of the frontal lobe epilepsies. Prefrontal implies higher mental functions (e.g., ictal forced thinking); rapid generalization to full grand mal evolves quite often from prefrontal foci. The frontal accentuation of classical generalized 3/sec spike-wave absences adds to the conceptual difficulties of the frontal lobe epilepsies. The unique type of disturbed consciousness in classical absences is presumed to be based upon ictal "suspension of the working memory." Limbic components (via orbitofrontal and cingulate mechanisms) also play an important role. Correlations between ictal semiologies and regional frontal lobe functions are still quite controversial.

*"The frontal lobe vexes the epileptologist with problems of differential diagnosis, exact localization, and in particular with questions of functional anatomy in correlation with seizure manifestations"*

*Niedermeyer E. Clin Electroencephalogr. 1998*

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## *Central challenges in Frontal lobe epilepsies*

- Deciding whether epilepsy is indeed focal or **generalized**
- Identifying **specific syndromes** and **age-dependent changes**
- Dealing with the **rapid propagation of seizures**
- Determining **localization** within (or outside) the frontal lobe
- Improving **outcome after surgery** which is less favorable compared to mesial temporal lobe epilepsy
- Approaching patients with intractable frontal lobe epilepsy **without MRI evidence of a structural lesion**

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Question 3

## Clinical Seizure Lateralization in Frontal Lobe Epilepsy

Silvia Beatrice Bonelli, Stefanie Lurger, Fritz Zimprich, Elisabeth Stogmann, Eva Assem-Hilger, and Christoph Baumgartner

*Department of Neurology, Medical University of Vienna, Vienna, Austria*

*Epilepsia*, 48(3):517–523, 2007
Blackwell Publishing, Inc.


**Summary:** *Purpose:* We systematically analyzed the lateralizing value of clinical seizure semiology in patients with frontal lobe epilepsy (FLE).

*Methods:* We studied the incidence, positive predictive value (PPV), and the lateralizing significance of various clinical symptoms in 228 seizures (s) of 31 patients (p) with medically refractory FLE (17 with left-sided and 14 with right-sided seizure onset). Seizures recorded during prolonged video-EEG monitoring were assessed by two independent reviewers blinded for the patient's clinical data. Analysis was performed both for patients and seizures.

*Results:* Version [16 p (52%); PPV, 94%; p = 0.001; 47 s (21%); PPV, 75%; p = 0.001], unilateral clonic movements [16 p (52%); PPV, 81%; p = 0.021; 32 s (14%); PPV, 81%; p = 0.001], unilateral dystonic posturing [eight p (26%); PPV, 75%; p = 0.289; 46 s (20%); PPV, 80%; p = 0.001], unilateral tonic posturing [10 p (32%); PPV, 80%; p = 0.109; 19 s (7.4%); PPV, 79%; p = 0.019], and unilateral grimacing [10 p (32%); PPV, 100%; p = 0.002; 19 s (8%); PPV, 100%; p = 0.001] were of lateralizing significance, indicating a contralateral seizure onset. Asymmetric ending [five p (16%); PPV, 80%; p = 0.375; nine s (4%); PPV, 89%; p = 0.039] after secondarily generalized tonic–clonic seizures was significantly associated with an ipsilateral seizure onset. Pure ictal vocalizations occurred significantly more frequently in seizures of right hemispheric onset [13 p (42%); PPV, 62%; p = 0.581; 63 s (28%); PPV, 73%; p = 0.001], whereas in individual patients, this symptom showed no lateralizing significance. The remaining clinical symptoms (figure 4 sign, unilateral hand automatisms, early head turning, postictal nose wiping, and unilateral eye blinking) were not of lateralizing significance in our patients. The results of clinical seizure lateralization corresponded with the final lateralization of the seizure-onset zone in 81% of our patients.

*Conclusions:* Clinical seizure semiology can provide correct information on the lateralization of the seizure-onset zone in >80% of patients with medically refractory frontal lobe epilepsy. **Key Words:** Frontal lobe epilepsy—Clinical seizure semiology—Clinical seizure lateralization.

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## Lateralizing signs in FLE: Frequency & Predictive value

**TABLE 2.** *Relation between the lateralization of the seizure-onset zone and the unilateral clinical symptom (positive predictive value)*

| Total (31/228) Symptom (patients/seizures) | Patient no. (%) | | | | Seizure no. (%) | | |
|---|---|---|---|---|---|---|---|
| | Ipsilateral | Contralateral | No lateralization | Significance | Ipsilateral | Contralateral | Significance |
| Version (16/47) | 0 (0) | 15 (93.8) | 1 (5.9) | 0.001 | 12 (25.5) | 35 (74.5) | 0.001 |
| Unilateral clonic seizures (16/32) | 3 (18.8) | 13 (81.3) | 0 (0) | 0.021 | 6 (18.8) | 26 (81.3) | 0.001 |
| Unilateral dystonic posturing (8/46) | 2 (25) | 6 (75) | 0 (0) | 0.289 | 9 (19.6) | 37 (80.4) | 0.001 |
| Unilateral tonic posturing (10/19) | 2 (20) | 8 (80) | 0 (0) | 0.109 | 4 (21.1) | 15 (78.9) | 0.019 |
| Unilateral grimacing (10/19) | 0 (0) | 10 (100) | 0 (0) | 0.002 | 0 (0) | 19 (100) | 0.001 |
| Figure 4 sign (10/18) | 4 (40) | 6 (60) | 0 (0) | 0.754 | 6 (33.3) | 12 (66.7) | 0.238 |
| Asymmetric ending (5/9) | 4 (80) | 1 (20) | 0 (0) | 0.375 | 8 (88.9) | 1 (11.1) | 0.039 |
| Unilateral hand automatisms (6/28) | 2 (33.3) | 3 (50) | 1 (16.7) | 1.000 | 17 (60.7) | 11 (39.3) | 0.345 |
| Early head turning (5/21) | 2 (40) | 3 (60) | 0 (0) | 1.000 | 7 (33.3) | 14 (66.7) | 0.189 |
| Postictal nose wiping (1/4) | 1 (100) | 0 (0) | 0 (0) | / | 4 (100) | 0 (0) | 0.125 |
| Unilateral eye blinking (1/3) | 0 (0) | 1 (100) | 0 (0) | / | 0 (0) | 3 (100) | 0.250 |

Clinical Seizure Lateralization in Frontal Lobe Epilepsy

*Epilepsia*, 48(3):517–523, 2007
Blackwell Publishing, Inc.


Silvia Beatrice Bonelli, Stefanie Lurger, Fritz Zimprich, Elisabeth Stogmann, Eva Assem-Hilger, and Christoph Baumgartner

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# *The Transition to Secondary Generalization*

## *a.k.a. "Focal to Bilateral Tonic-Clonic"*

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NEUROLOGY 1986;36:606-611

## The lateralizing significance of versive head and eye movements during epileptic seizures

Elaine Wyllie, MD; Hans Lüders, MD, PhD; Harold H. Morris, MD; Ronald P. Lesser, MD; and Dudley S. Dinner, MD

Contro-versies: *adversion*, *contraversion* vs. *ipsiversion*

- **Versive** = ***unquestionably forced and involuntary, resulting in sustained unnatural positioning***
- **Nonversive** = mild, unsustained, wandering or seemingly involuntary

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## *"the figure 4 sign"*
## Asymmetric Tonic Limb Posturing

*Epilepsia*, 41(4):457–462, 2000
Lippincott Williams & Wilkins, Inc., Philadelphia


Clinical Research

Lateralizing Value of Asymmetric Tonic Limb Posturing Observed in Secondarily Generalized Tonic–Clonic Seizures

Prakash Kotagal, *Andrew Bleasel, *Eric Geller, Pongkiat Kankirawatana, Bhagwan I. Moorjani, and †Lisa Rybicki

*Sections of Pediatric Epilepsy, *Adult Epilepsy, and †Department of Biostatistics, Cleveland Clinic Foundation, Cleveland, Ohio, U.S.A.*

*Kotagal; Epilepsia 2000*

Cleveland Clinic

## *"figure 4 sign" early on*

*Kotagal; Epilepsia 2000*

Cleveland Clinic

## *"figure 4 sign" cont'd (same seizure)*

*Kotagal; Epilepsia 2000*

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# Frontal Lobe Epilepsies: What do we know from case series?

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## Frontal Lobe Epilepsy (FLE)
## Main ictal motor manifestations

- **Tonic posturing** (extension, flexion, dystonic)
- **Axial signs**
- **Complex motor** (rocking, pelvic thrusting, trunk flexing, rolling, rotating)
- **Hypermotor / Hyperkinetic** (intense motor activity, biking, attempts to run away, tearing off electrodes)
- **Secondary generalization** (quick sometimes so quick that the focal beginning is not apparent)
- **Variable** (eyelid flutter, unilateral jerks, massive myoclonic jerks)

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## Typical FLE seizure

- **Nonspecific** aura
- **Abrupt** beginning & end
- **Brief** duration (usually <60 sec)
- Frequently in **clusters** with tendency to status epilepticus
- **Rapid secondary** generalization (and **falling** with bilateral discharges)
- **Nocturnal** preponderance
- **Variable semiology** in individual patients
- **Complex motor** automatisms
- Complex **vocalization**
- **May be misdiagnosed as psychogenic events**

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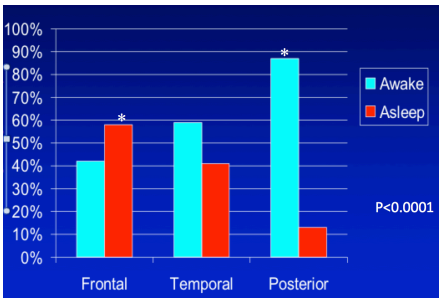
## Activation of seizures with sleep vs. awake




*Herman et al. Neurology, 2001*

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ORIGINAL CONTRIBUTION

Distinguishing Sleep Disorders From Seizures

*Diagnosing Bumps in the Night*

Christopher Paul Derry, MRCP; Margot Davey, FRACP; Murray Johns, FRACP; Katie Kron, BSc; Deborah Glencross, BSc; Carla Marini, PhD; Ingrid E. Scheffer, PhD; Samuel F. Berkovic, MD

*Arch Neurol.* 2006;63:705-709

**Table. The Frontal Lobe Epilepsy and Parasomnias (FLEP) Scale**

| Clinical Feature | | Score |
|---|---|---|
| Age at onset<br>At what age did the patient have their first clinical event? | <55 y<br>≥55 y | 0<br>-1 |
| Duration<br>What is the duration of a typical event? | <2 min<br>2-10 min<br>>10 min | +1<br>0<br>-2 |
| Clustering<br>What is the typical number of events to occur in a single night? | 1 or 2<br>3-5<br>>5 | 0<br>+1<br>+2 |
| Timing<br>At what time of night do the events most commonly occur? | Within 30 min of sleep onset<br>Other times (including if no clear pattern identified) | +1<br>0 |
| Symptoms<br>Are the events associated with a definite aura? | Yes<br>No | +2<br>0 |
| Does the patient ever wander outside the bedroom during the events? | Yes<br>No (or certain) | -2<br>0 |
| Does the patient perform complex, directed behaviors (eg, picking up objects, dressing) during events? | Yes<br>No (or uncertain) | -2<br>0 |
| Is there a clear history of prominent dystonic posturing, tonic limb extension, or cramping during events? | Yes<br>No (or uncertain) | +1<br>0 |
| Stereotypy<br>Are the events highly stereotyped or variable in nature? | Highly stereotyped<br>Some variability/uncertain<br>Highly variable | +1<br>0<br>-1 |
| Recall<br>Does the patient recall the events? | Yes, lucid recall<br>No or vague recollection only | +1<br>0 |
| Vocalization<br>Does the patient speak during the events and, if so, is there subsequent recollection of this speech? | No<br>Yes, sounds only or single words<br>Yes, coherent speech with incomplete or no recall<br>Yes, coherent speech with recall | 0<br>0<br>-2<br>+2 |

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## Normal EEG findings

*Across studies up to 21% of patients with Frontal Lobe Epilepsy do not have any interictal epileptiform discharges*

| Study | Patient numbers | % of no spikes |
|---|---|---|
| Quesney. Adv Epileptology, 1984 | 22 patients | 9% |
| Morris Neurology 1988 | 17 patients | 36% |
| Laskowitz Neurology 1995 | 16 patients | 19% |
| Bautista Neurology 1998 | 9 patients surgical well controlled | 33% |
| Gross. J Clin Neurophys, 2000 | 24 patient with frontocentral epilepsy | 26% |
| Vadlamudi Epileptic Disord 2004 | 53 patients (surgical well controlled) | 19% |

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## Focal Onset Ictal Patterns

*seen more frequently with **Lateral FLE** than with **Mesial FLE***

| Study | Seizures | % of focal onsets |
|---|---|---|
| Quesney. Can J Neurol Sci 1991 | 302 seizures in frontal lobe epilepsy | 22% |
| Swartz Can J Neurol Sci 1991 | 172 seizures of frontal lobe epilepsy | 12% |
| Blume. Adv Neurol 1996 | 16 seizures of SMA patients | 6% |
| Bautista. Neurology, 1998 | 9 patients (5 with MFLE and 4 LFLE)-surgical well controlled | 44% all in LFLE |
| Lee. Epilepsia 2000 | 26 cases of frontal lobe epilepsy | 23% |
| Foldvary. Neurology 2001 | 8 cases of MFLE<br>15 cases of LFLE | 25%<br>60% |

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## *Functional anatomy of the Frontal Lobe*

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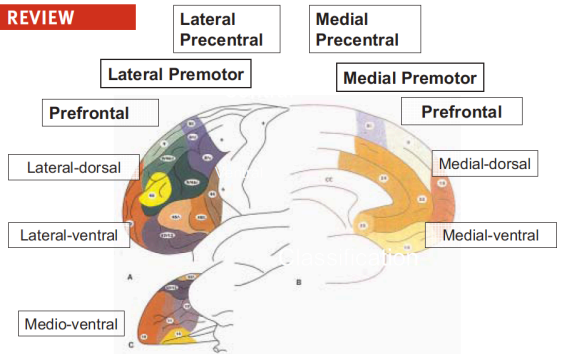
REVIEW

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*McGonigal A & Chauvel P, Practical Neurol 2004*

## Functional frontal lobe anatomy

- **Motor** cortex
  - —Primary
  - —Premotor
  - —Supplementary
  - —Frontal eye field
  - —Broca's speech area
- **Prefrontal** cortex
  - – Dorsolateral
  - – Medial
  - – Orbitofrontal

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## *Seizures arising from the Motor areas*

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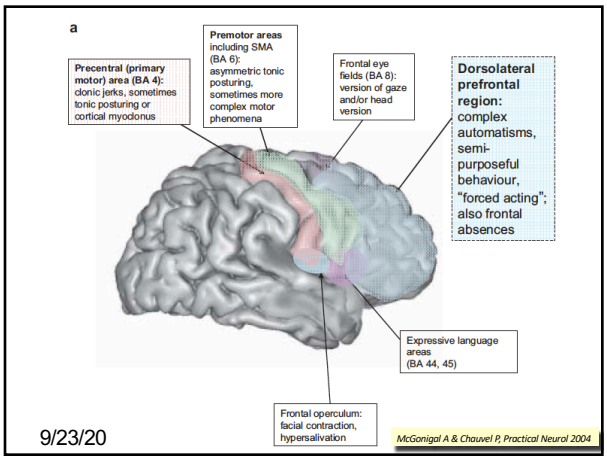


**TABLE 2.** *Seizures originating from areas 4 and 6 (154 seizures in 43 patients)*

Area 4 (39 patients, 126 seizures)
- Isolated myoclonic jerks
- Partial motor seizures with Jacksonian march
- Partial complex motor seizures
- (Kojewnikoff's syndrome, Rasmussen's syndrome, startle epilepsy)

Supplementary motor area (4 patients, 28 seizures)
- Speech arrest
- Vocalization
- Palilalia (dominant hemisphere)
- Abduction and lifting contralateral upper limb
- Adversive movement of head and eyes
- $M^2E$

**Jean Bancaud and Jean Talairach 1992**

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# *Frontal Eye Fields*

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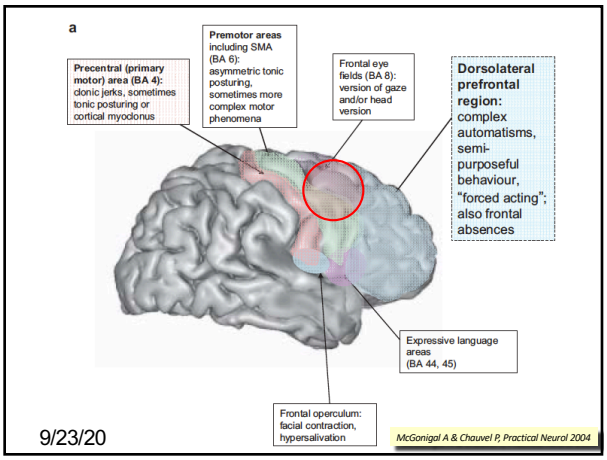


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CME

## The predictive localizing value of tonic limb posturing in supplementary sensorimotor seizures

Y. Aghakhani, MD; A. Rosati, MD; A. Olivier, MD, FRCS, PhD; J. Gotman, PhD; F. Andermann, MD, FRCP; and F. Dubeau, MD, FRCP

**Abstract**—*Objective:* To determine whether early tonic limb posturing is reliable in lateralizing or localizing of the seizure generator in 14 patients with pharmacoresistent supplementary sensorimotor area (SSMA) seizures. *Methods:* All patients underwent high-quality MRI scans and stereo-EEG recordings. *Results:* The SSMA seizure semiology predicted focal or regional ictal onset in the SSMA in six (43%) patients: Three had a focal SSMA seizure onset, and three had a regional seizure onset with involvement of one SSMA plus adjacent neocortex. The eight remaining patients had diffuse uni- or bilateral seizure onset. Eight of 14 patients underwent a frontal or central cortical resection, but a good outcome was seen in only 3: 2 with no SSMA resection and 1 with an extensive central removal. *Conclusions:* SSMA semiology is suggestive of early involvement of this region but is by no means a reliable indicator that the primary SSMA contains the seizure focus.

NEUROLOGY 2004;62:2256–2261

## *SSMA seizures vs. SSMA epilepsy*

…SSMA semiology is suggestive of ***early involvement of this region***, but is by no means a reliable indicator that the primary SSMA contains the seizure focus

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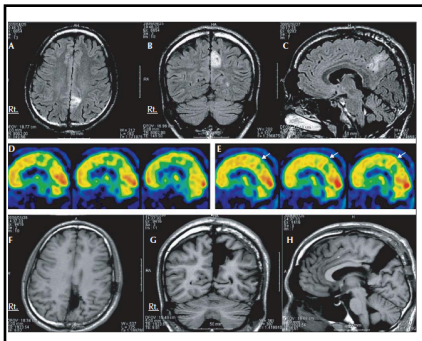

Clinical commentary with video sequences

Bilateral symmetric tonic posturing suggesting propagation to the supplementary motor area in a patient with precuneate cortical dysplasia

Shuichi Umeoka[1], Koichi Baba[1], Kiyohito Terada[2], Kazumi Matsuda[1], Takayasu Yoton[3], Naotaka Usui[1], Keiko Usui[1], Fumihiro Nakamura[3], Yushi Inoue[1], Taiki Fujiwara[4], Tadahiro Mihara[1]

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# Ventromesial Frontal

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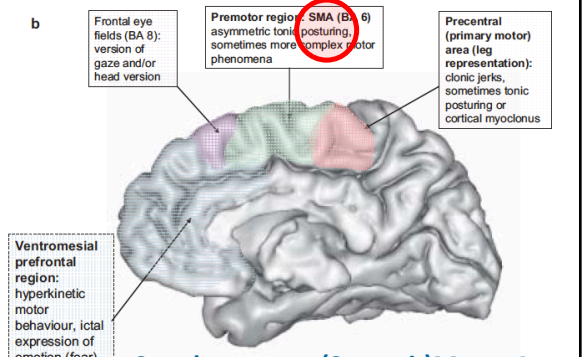


Supplementary (Sensori-)Motor Area

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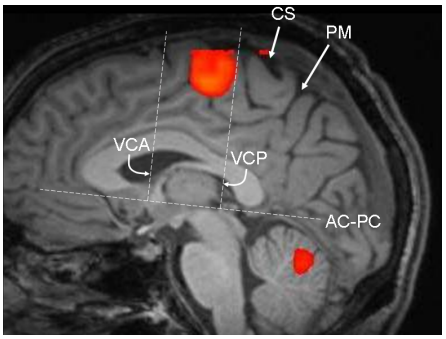
*McGonigal A & Chauvel P, Practical Neurol 2004*

## Anatomical localization & fMRI activation of SMA




*Alexopoulos AV & Jones SE - in Wyllie E. Treatment of Epilepsy 2010*

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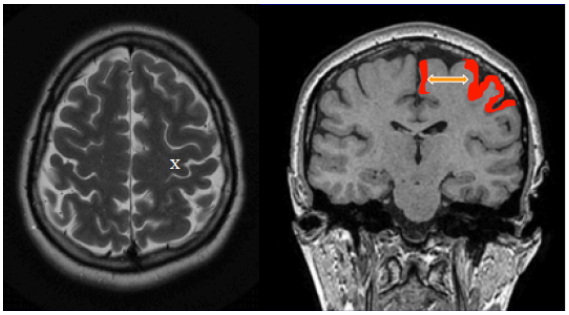
## Somatotopically organized monosynaptic pathways between SSMA and M1




*Baumgartner et al. Neurology 1996*

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## Features of SSMA seizures

1. Frequent, Brief, occurring in Clusters
2. Sudden Onset & Offset w/out postictal confusion
3. Prominent Tonic Posturing
4. Nocturnal preponderance
5. Preserved Consciousness (commonly)

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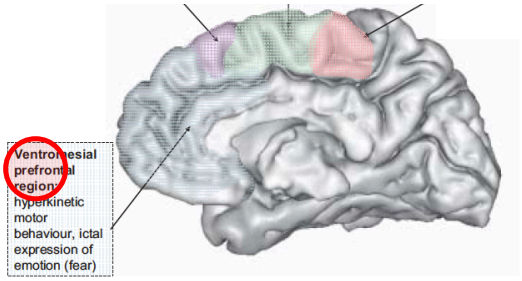
## Ventromesial prefrontal region

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*McGonigal A & Chauvel P, Practical Neurol 2004*

## *Types of hyperkinetic seizures*


*Rheims S et al. Epilepsia 2008*

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## *Characteristics of hyperkinetic seizures*

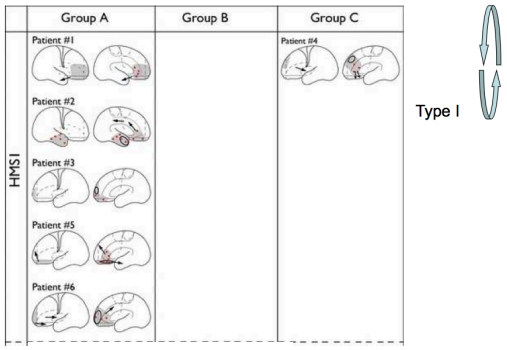
| | Type I | Type II |
|---|---|---|
| **Agitation** | Marked | Mild |
| **Hyperkinetic** Behavior | Sitting up, laying down, kicking, boxing | Rotation of trunk **horizontally** |
| **Expression** | Fear, Anger | - |
| **Head** deviation | Ipsilateral | +/- Contralateral |
| **Autonomic** | Incontinence, **Flushing** | Variable |
| **Loss of Contact** | Yes | Variable |
| **Amnesia** | Yes | Variable |

*Rheims S et al. Epilepsia 2008*

Cleveland Clinic

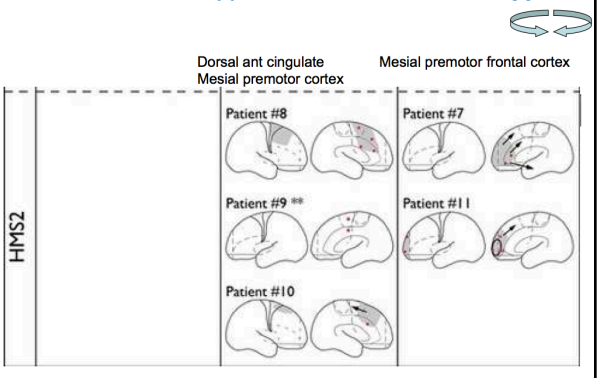
## **Ictal onset** of hyperkinetic seizures: **type I**

Rostral ant cingulate — Mesial premotor frontal cortex




*Rheims S et al. Epilepsia 2008*

## **Ictal onset** of hyperkinetic seizures: **type II**




*Rheims S et al. Epilepsia 2008*

## Pitfalls


Hypermotor seizures in lateral and mesial parietal epilepsy

Alexandra Montavont [a,b,c,*], Philippe Kahane [d], Hélène Catenoix [c], Karine Ostrowsky-Coste [a,b], Jean Isnard [c], Marc Guénot [e], Sylvain Rheims [b,c], Philippe Ryvlin [a,b,c]

*Montavont A et al. Epilepsy & Behavior 2013*

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## Orbitofrontal Epilepsy

Semiology

- **Absent**
- Hypermotor (Hyperkinetic)
- Dialeptic (Absence-like)
- Automotor (Distal Automatisms)
- SSMA

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## Psychogenic versus FLE Seizures

- PNES *general impression*
  - A. Hx of psychiatric disorder
  - B. Pelvic thrusting, body rocking
  - C. Side-to-side head movement
  - D. Rapid postictal recovery

***But there was no difference between the two groups in any of the above categories***

- **FLE** Seizures*
  1. **Turning** to **prone** position
  2. **Nocturnal** occurrence
  3. **Short** duration
  4. **Younger** age of **onset**
  5. **Stereotyped** movements

*Saygi S et al. Neurology 1992*

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## Epilepsy arising from the Parietal Lobe

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## Parietal Lobe Epilepsy

- **Perirolandic cortex**: onset with ***somatosensory sensations***
  - Tingling > vibrations, numbness, unpleasant pressure, rarely pain
  - ***Body parts with largest representation*** *are most frequently affected*
  - Hand, face, mouth, tongue
  - Tendency to *evolve into a "Jacksonian March"*
- Involvement of **inferior parietal cortex**
  - **Vertigo** or **spatial disorientation**
- Involvement of the **parietal operculum**
  - Whole contralateral body and/or ipsilateral **body sensations**
- ***Other Auras***: rare often too brief to be remembered
  - Distortion of visual perception (metamorphopsias), **alteration of body perception** (dysmorphopsia/gnosia), loss of awareness of part of half of the body (asomatognosia)
- ***Spread of ictal activity can be to frontal or temporal lobe***
  - Seizure manifestations then representative of these symptomatogenic zones

Cleveland Clinic

doi:10.1093/brain/bev187 BRAIN 2015: 138, 2596–2607 | 2596

BRAIN
A JOURNAL OF NEUROLOGY

**Multimodal responses induced by cortical stimulation of the parietal lobe: a stereo-electroencephalography study**

Simona Balestrini,[1] Stefano Francione,[2] Roberto Mai,[2] Laura Castana,[2] Giuseppe Casaceli,[2] Daniela Marino,[2] Leandro Provinciali,[1] Francesco Cardinale[2] and Laura Tassi[2]

The functional complexity of the parietal lobe still represents a challenge for neurophysiological and functional neuroimaging studies. While the somatosensory functions of the anterior parietal cortex are well established, the posterior parietal cortex has a relevant role in processing the sensory information, including visuo-spatial perception, visual attention, visuo-motor transformations and other complex and not completely understood functions. We retrospectively analysed all the clinical manifestations induced by intracerebral bipolar electrical stimulation in 172 patients suffering from drug-resistant focal epilepsy (mean age 25.6, standard deviation 11.6; 44% females and 56% males) with at least one electrode stereotactically implanted in the parietal cortex. A total of 1186 electrical stimulations were included in the analysis, of which 88 were subsequently excluded because of eliciting stimulation responses were classified into the following semiological categories: (i) somatosensory sensation (including anaesthesia, paraesthesia or thermic dysperception); (ii) visual, auditory, gustatory or multisensory illusion/hallucination; (iii) vertigo; (iv) body scheme alteration (including altered subjective perception of body image or movement); (v) dysarthria; (vi) speech arrest; (vii) neurovegetative symptom (including all types of autonomic symptoms); (viii) pain; (ix) eyeball/eyelid movement or sensation; (x) motor symptom (including clonic or tonic movements, atonia); (xi) psychic phenomenon (including derealization); (xii) multimodal response (including complex symptom belonging to more than one of the above categories); and (xiii) unclassified response (when the effect was not possible to include in any of the above categories).

Examples of symptoms classification are reported in Supplementary Table 1.

The parietal lobes were divided in the following subtractures (Duvernoy, 1999; Nieuwenhuys *et al.*, 2008) (Fig. 2): (i) postcentral gyrus, posterior part of paracentral lobule and parietal operculum; (ii) posterior cingulum; (iii) precuneus; (iv) intraparietal sulcus; (v) superior parietal lobule; and (vi) inferior parietal lobule.

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## Scalp EEG: Interictal

Original article

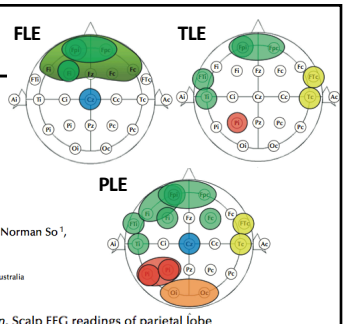
*Epileptic Disord 2012; 14 (1): 22-31*

**Parietal lobe epilepsy: the great imitator among focal epilepsies**

Aleksandar J Ristić [1,2], Andreas V Alexopoulos [1], Norman So [1], Chong Wong [1,3], Imad M Najm [1]

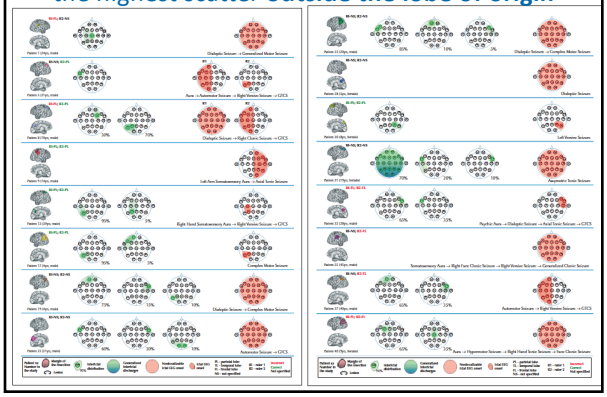
[1] Epilepsy Center, Cleveland Clinic, Cleveland, OH, USA
[2] Epilepsy Center, Clinic of Neurology CCS, Belgrade, Serbia
[3] Department of Neurology, Westmead Hospital, Westmead, NSW, Australia

Received October 23, 2011; Accepted February 14, 2012

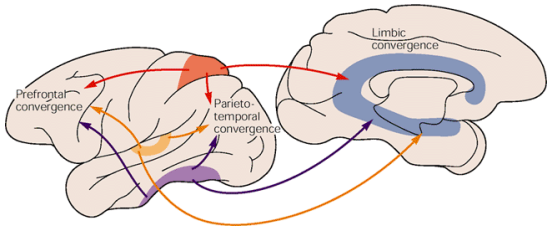



*Conclusion.* Scalp EEG readings of parietal lobe epilepsy patients showed a more variable scatter of interictal discharges and a lower localisation value of ictal recordings compared to temporal and frontal lobe epilepsy subjects, suggesting an increased likelihood of misidentification and mislocalisation of parietal lobe epilepsy. Combining seizure semiology with scalp interictal and ictal EEG readings facilitates a more accurate lobar classification in patients with temporal and frontal, but not parietal, lobe epilepsy.

## **Interictal discharges in Parietal lobe epilepsy** had the highest scatter **outside the lobe of origin**



## Parietal Lobe: the prototypical **Multimodal Association Area**




***Spread of ictal activity can be to frontal or temporal lobe***

– Seizure manifestations then representative of these symptomatogenic zones

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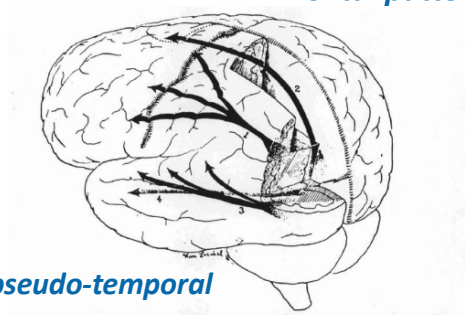
*Kandel et al. Principles of Neural Science 2000*

## Posterior epilepsy

***"Frontal pattern"***



***pseudo-temporal***

Patterns of spread from occipital lobe seizure origin. (From Ajmone-Marsan and Ralston, 1957, with permission.)

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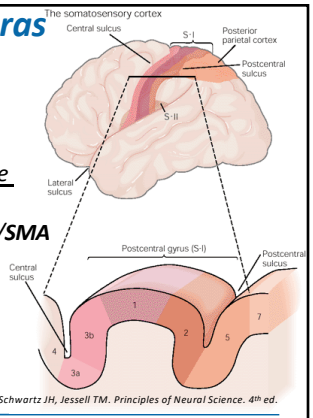
## *Somatosensory auras*

- *Primary **SI** area*
  - *Contralateral focal*
  - *Tingling, numbness*
- *Second **SII** area*
  - *Contra or Ipsi or B/L wide*
  - *Coldness, pain*
- *Supplementary Sensory **SSA/SMA***
  - *Contra or B/L extensive*
  - *Pulling sensation*
- *Posterior Insula*
  - *Contra localized or wide*
  - *Heat, pain*




*Kandel ER, Schwartz JH, Jessell TM. Principles of Neural Science. 4th ed.*

Cleveland Clinic

*Kandel et al. Principles of Neural Science 2000*

# Opercular-Insular perisylvian networks

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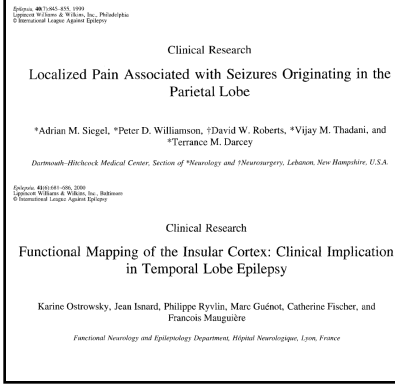
## *Insular-Perisylvian Networks*



Clinical Research

Localized Pain Associated with Seizures Originating in the Parietal Lobe

*Adrian M. Siegel, *Peter D. Williamson, †David W. Roberts, *Vijay M. Thadani, and *Terrance M. Darcey

*Dartmouth–Hitchcock Medical Center, Section of *Neurology and †Neurosurgery, Lebanon, New Hampshire, U.S.A.*

Clinical Research

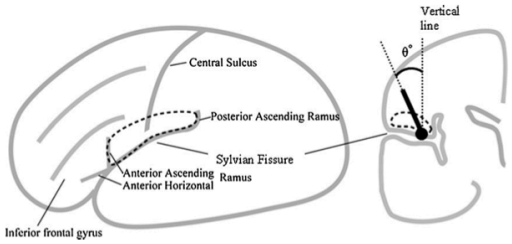
Functional Mapping of the Insular Cortex: Clinical Implication in Temporal Lobe Epilepsy

Karine Ostrowsky, Jean Isnard, Philippe Ryvlin, Marc Guénot, Catherine Fischer, and Francois Mauguière

*Functional Neurology and Epileptology Department, Hôpital Neurologique, Lyon, France*

The topographic organization of the induced responses within the insular cortex suggests that 2 different cortical networks are disturbed with stimulation of the **anterior** or the **posterior** insula, a **visceral** network extending to the *temporomesial structures* and a **somesthetic** network reaching the *opercular cortex*, respectively

## Frontoparietal operculum: schematic




*Kakisaka Y, Alexopoulos AV, Burgess RC, et al. Epilepsy Research 2012*
*Magnetoencephalography in fronto-parietal opercular epilepsy*

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## Opercular Epilepsy (Parietal)

- **Posterior (parietal) operculum**
  - –Somato-sensory symptoms affecting large cutaneous territories
  - –Somatotopic fields are much larger in the insula than in SII
  - –**Whole contralateral** body and/or **ipsilateral** body sensations

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## Opercular Epilepsy (Frontal)

- **Anterior (frontal) operculum**
  - –Difficulties in **speech** and **swallowing** due to the loss of voluntary control of the *oro-facio-linguo-pharyngo-masticatory muscles*, while automatic-emotional and reflexive activities remain intact
  - –Contraction of the contralateral facial musculature, particularly of **one corner of the mouth**
  - –Unilateral or bilateral contractions of the mouth, tongue or eyelids, positive or negative subtle **perioral** or other myoclonus, dysarthria, speech arrest, difficulties in swallowing, **buccofacial apraxia**, **hypersalivation** and ?gustatory hallucinations

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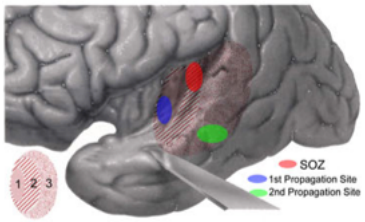
## Temporoparietal operculum: schematic




*Wang ZI, Alexopoulos AV, et al. Epilepsy Research 2012*
*Interconnections in superior temporal cortex revealed by musicogenic seizure propagation*

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## Opercular Epilepsy (Temporal)

- **Infrasylvian (temporal) operculum**
  - – **Auditory** hallucinations or early receptive aphasia
  - – Contraction of the contralateral facial musculature, particularly of one corner of the mouth

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## Insular Epilepsy

### *(but cannot exclude opercular)*

- Unpleasant, sometimes painful **laryngeal constriction**
- **Perioral dysesthesias**, dysarthria followed by clonic activity in face, arm without LOC
- **Unpleasant taste**

– May resemble temporal, frontal or parietal lobe epilepsy

– Thought to account for some of the patients who fail temporal lobectomy

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Clinical Manifestations of Insular Lobe Seizures:
A Stereo-electroencephalographic Study

*Jean Isnard, †Marc Guénot, †Marc Sindou, and *François Mauguière

*Functional Neurology and Epileptology Department and †Functional Neurosurgery Department, Neurological Hospital, Lyon, France.

Epilepsia, 45(9):1079–1090, 2004
Blackwell Publishing, Inc.


1 2 3 4 5 6

A
B
C
D

## Clinical features of Insular Lobe seizures

- Frequently retain awareness
- Unpleasant or painful **laryngeal** constriction and/or chest or abdominal heaviness
- **Paresthesiae** either limited to peri/**intraoral** region or contralateral (rarely bilateral) face/arms/legs/trunk without jacksonian march
- Dysphonic/**dysarthric** speech or speech arrest
- Progression to **contralateral tonic**/clonic face, limb involvement occasionally bilateral asymmetric
- Early onset of these symptoms should alert one to possibility of insular onset

*Isnard J et al. Epilepsia 2004;45:1079-90*

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## Other symptoms related to spread to the Insula

- Ictal Vomiting
- Spitting
- Piloerection
- Apnea
- Bradycardia or Asystole

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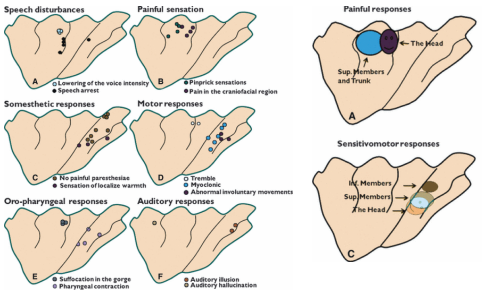
## Functional organization of the Insular Cortex




*Afif A et al. Epilepsia 2010*

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# Epilepsy arising from the Occipital Lobe

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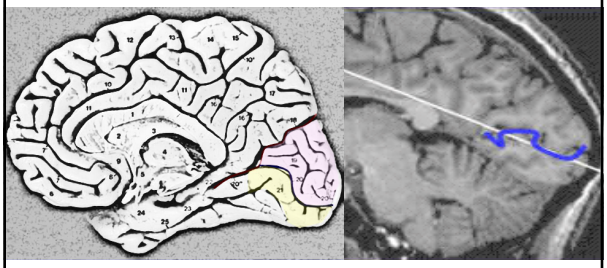
## Mesial Surface
## Parieto-occipital & Calcarine fissures



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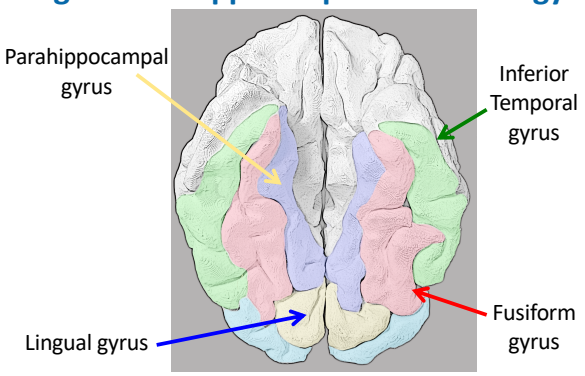
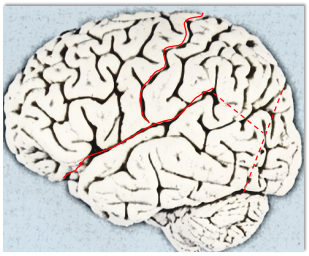
## Basal Surface
## Lingual-Parahippocampal & Fusiform gyri




## Lateral Surface
## The artificial "TPO" borders



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## Occipital Epilepsy

- 4-13 % of all partial epilepsies; 11% of symptomatic partial epilepsies in children
- 5-6% of surgical series

"…discharges arising from the visual region may possess the greatest potential for complexity of seizure formation"
*Ajmone-Marsan and Ralston 1957*

**Only 10-15% stay localized** to occipital lobe

- Infrasylvian spread – temporal lobe involvement (44-88%)
- Suprasylvian spread - frontal lobe seizure type (12%)
- Spread to contralateral occipital lobe
- Combination of spread patterns - multiple seizure types (44%)

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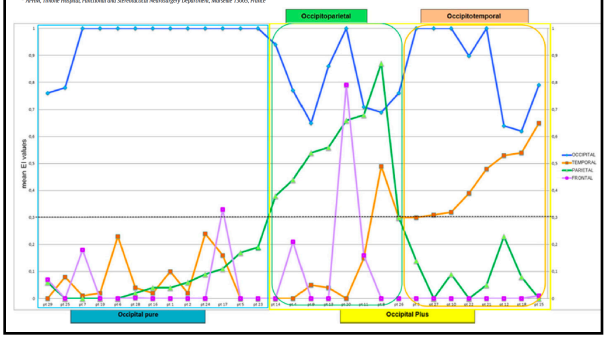
Occipital and occipital "plus" epilepsies: A study of involved epileptogenic networks through SEEG quantification

Angela Marchi [a,b], Francesca Bonini [a,b], Stanislas Lagarde [a,b], Aileen McGonigal [a,b], Martine Gavaret [a,b], Didier Scavarda [c], Romain Carron [d], Sandrine Aubert [a], Nathalie Villeneuve [a], Samuel Médina Villalon [a], Christian Bénar [b], Agnes Trebuchon [a,b], Fabrice Bartolomei [a,b,*]

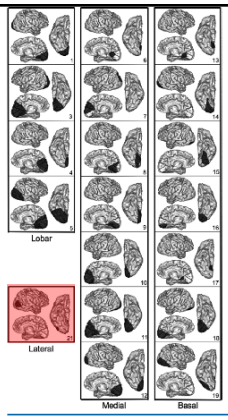
[a] APHM, Timone Hospital, Clinical Neurophysiology and Epileptology Department, Marseille 13005, France
[b] Aix-Marseille Université, Institut de Neurosciences des Systèmes, UMR_S 1106, Marseille 13005, France
[c] APHM, Timone Hospital, Paediatric Neurosurgery Department, Marseille 13005, France
[d] APHM, Timone Hospital, Functional and Stereotactical Neurosurgery Department, Marseille 13005, France

Epilepsy & Behavior 62 (2016) 104–114




## Seizures with visual symptomatology

- **Elementary visual hallucinations** are most common: crude sensations of light or colors - may take various shapes, be continuous, steady, moving, or interrupted
- Transient amaurosis either partial or complete may also occur (**ictally** or **postictally**) and is especially common in children
- **Formed visual hallucinations** are fairly often such as pictures of people, animals or scenes, either static or moving.
- Visual illusions of **spatial** interpretation, illumination, **coloring** of vision, or movement in space. Lines may be defective or fragmented, stationary objects seen as moving, or motion appears too slow or too fast.
- Micro- or **macropsia**: objects may appear diminished or enlarged
- **Teleopsia**: objects appear both small and at a distance,
- **Palinopsia** or visual perseveration, in which visual images recur or persist




Occipital epilepsy: spatial categorization and surgical management

Clinical article
J Neurosurg 110:306–318, 2009

Nitin Tandon, M.D.,[1] Andreas V. Alexopoulos, M.D., M.P.H.,[2] Ann Warbel, R.N.,[2] Iyad M. Najm, M.D.,[2] and William E. Bingaman, M.D.[2]

[1]Department of Neurosurgery, The University of Texas Medical School at Houston, Texas; and [2]Departments of Neurology and Neurosurgery, Cleveland Clinic Epilepsy Center, Cleveland, Ohio

- Resection between 1992 and 2004.

Included:

Resections extending to temporo-occipital junction, inferior precuneus, posterior parietal lobe

Excluded

multi-lobar resections

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## Semiology - Scalp EEG

- **Visual aura** in 11 **(52%)**
  - Elementary visual phenomena
    - Positive
    - Negative
    - Lateralizing value
  - Elementary & complex phenomena
    - One saw animals
    - One had micropsia
- Seizure types
  - Dialeptic 33%
  - Early frontal involvement 29%
  - Automotor 38%
- Interictal discharges:
  - occipital/temporo-occipital 38%
  - mid to posterior temporal 14%
  - anterior temporal lobe 5%
- Ictal onset - focal origin:
  - temporo-occipital /occipital 62%
  - **ipsilateral temporal lobe** 10%

*Tandon N, Alexopoulos AV et al. J Neurosurg 2009*

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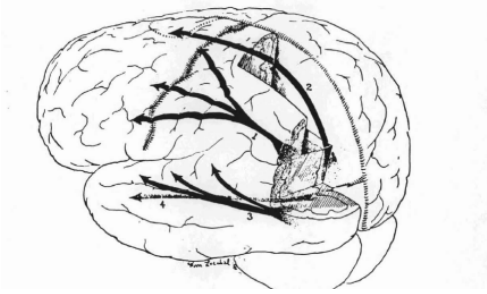

*Spread of ictal activity can be to frontal or temporal lobe*

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## One more thing…

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## *Age-dependent semiology of focal seizures*

| Manifestation | *Infants* | *Adults* |
|---|---|---|
| **Aura** | **Absent** | Sometimes present |
| ***Behavioral arrest**** | Prominent, often isolated | Present, often w/other signs |
| ***Limb clonus**** | Present | Present |
| **Perioral cyanosis** | Prominent w/ TL szs | Sometimes present |
| **Dystonic posture** | **Absent** | May be present |
| **Hand automatisms** | **Absent** | May be present |
| **Loss of consciousness** | Difficult to ascertain | Can be determined |
| **Secondary generaliz.** | Rare | Common |
| **Diffuse myoclonus** | At start or end of focal sz | Rare |
| ***Symmetric tonic**** | Frequent | Unusual |
| ***Spasms**** | Concurrent w/ focal szs | Absent |

*Adapted from Wyllie 2005*

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## Take home points

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## Important points regarding semiology

- Observe **sequence of appearance of signs** - early signs more reliable
- Expect **concordance between signs** in a seizure
- Record **sufficient number of seizures**
- Look for **consistency between seizures**
- **Show recorded seizures to family** or friends
- **Always correlate** with history, EEG, & imaging results

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## Take-home points

- There are tight anatomical connections and cytoarchitectonic commonalities (continuum) between the **fronto-parietal**, **occipito-parietal**, **occipito-temporal**, **perisylvian**, **parieto-insular** (as well as fronto-temporal, temporo-insular etc.) regions
- Focal epilepsies do not conform to anatomical lobar boundaries. Yet most case series and studies are based on lobar divisions.
- Take into account the importance of structural connectivity sub-serving normal brain functions along with the pathways of seizure spread
- Consider **which of the above networks could be involved** to account for each and every **electroclinical** scenario

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## *Thank you for your attention!*

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