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SEIZURE TYPES IN CHILDREN

From clinical observation to description and classification

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SEIZURE TYPES IN CHILDREN
From clinical observation to description and classification

EN AVANT PREMIERE

Operational Classification of Seizure Types by the International League Against Epilepsy

Robert S. Fisher[1], J. Helen Cross[2], Jacqueline A. French[3], Norimichi Higurashi[4], Edouard Hirsch[5], Floor E. Jansen[6], Lieven Lagae[7], Solomon L. Moshé[8], Jukka Peltola[9], Eliane Roulet Perez[10], Ingrid E. Scheffer[11], **Sameer M. Zuberi**[12]

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INTERNATIONAL LEAGUE AGAINST EPILEPSY
ILAE
ESTABLISHED 1909

But what is the definition of “seizure” ??

- An old debate …
- What will follow considers **semiology**, consequently we can only refer to “**clinical events** of relatively short duration (***paroxysmal*** in the sense that they are clearly different from the basal state of a person)
- However, the definition is also: an excessive and hypersynchronous neuronal discharge involving the cortex …

The Task Force avoided to provide a definition for “seizure” and preferred to define “operational seizure types” as : a useful grouping of seizure characteristics for purposes of communication in clinical care, teaching and research.

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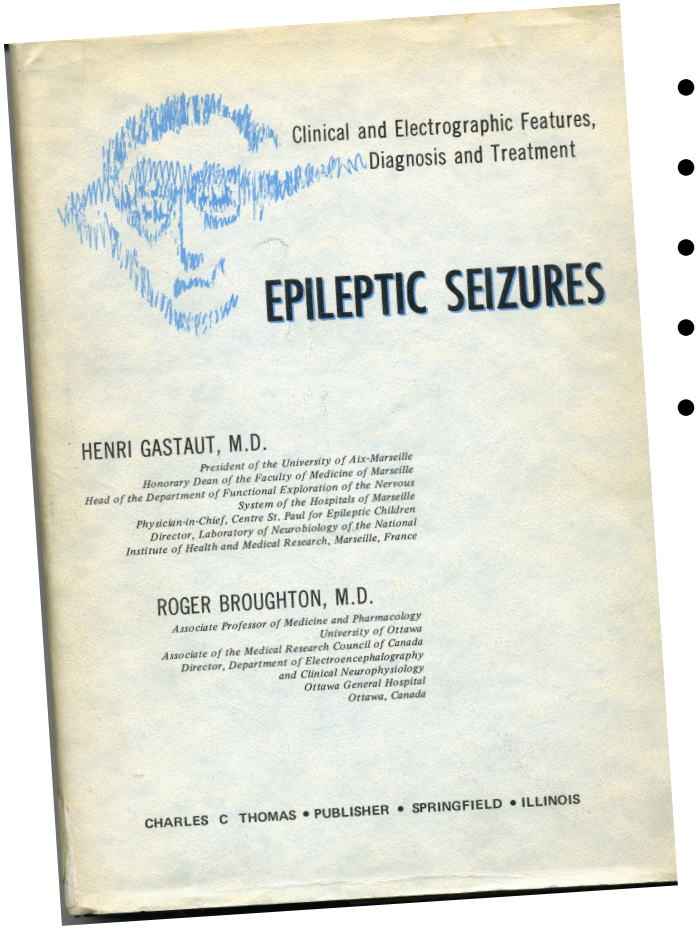
Historical Background - 1



- The 1969-1970 ILAE Classification
- The 1989 revision the 1969 classification
- The ILAE proposal (Engel 2001)
- The Lüders proposal (1998)
- The 2010 proposal (Berg et al)

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Historical Background - 2

Both the 1969 classification and the 1989 classifications
are dichotomous systems and utilize 2 criteria:

- the clinical seizure type
- the EEG seizure manifestations

The ILAE proposal (Engel 2001)
is termed a 'glossary of descriptive terminology' to help clinical description of seizure manifestations

The Cleveland proposal (Lüders et al 1998)
uses only one criterion: the clinical seizure type

Historical Background - 3

"Simple" versus "Complex" partial

In the 1969-70 Classification:

- **simple seizures** were those due to involvement of the primary cortical areas (motor, sensory , visual etc..
- **complex seizures** as those due to involvement of interpretative, association areas

In the 1989 Revision:

- **complex** seizures those with impaired consciousness
- **simple** seizures those without

Consciousness was operationally defined as impaired awareness and/or responsiveness, which does not reflect the intricacies of the concept

Problems with the 1969 and 1989 classifications

- Based on **simple dichotomies** such as ‘simple versus complex’ or ‘generalized versus localized’ that are of dubious significance or difficult to recognize clinically or both
- The use of an electroclinical classification supposes that there is a **consistent one to one relationship** between clinical and electrographic expression
- The 1969 **classification does not take into account the evolution of seizures over time**
- Both do not describe **many features of seizures**

Difficulties with seizure classifications

- Inclusion of **sufficiently precise clinical description** especially in seizures such as *automatisms* with resultant loss of information (denomination rather than description)

- Temporal evolution and changes in seizure

- Necessity of special description for infants and young children

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The Lüders contribution - 1

Topographic modifiers

- Right/left
- Somatotopy: hand, arm, leg etc..
- Bilateral, symmetric/asymmetric, axial, generalized

  ex clonic seizure right hand

  somatosensory aura throat

  tonic seizure right leg
- Hemispheric markers: Aphasia, upper limb dystonia

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The Lüders contribution - 2

Seizure Sequence. Timing

- **Visual aura left**

➢ clonic seizure left hand

➢ tonic clonic seizure generalized

(CPS evolving to secondary generalization)

- **Abdominal aura**

➢ automotor seizure with R hand dystonia (Lhemisphere)

(CPS evolving to CPC + automatisms)

- **Olfactory aura**

➢ automotor seizure

➢ Versive seizure left

➢ tonic clonic

(SPS evolving to CPC + automatisms, evolving to secondary generalization)

2001 ILAE Task Force: ‘a diagnostic scheme rather than a fixed classification’

- Axis 1: Ictal phenomenology
  (glossary to describe ictal events)
- Axis 2: Seizure types
- Axis 3: Syndromes
- Axis 4: Aetiology (list of diseases)
- Axis 5: Impairment resulting from disease

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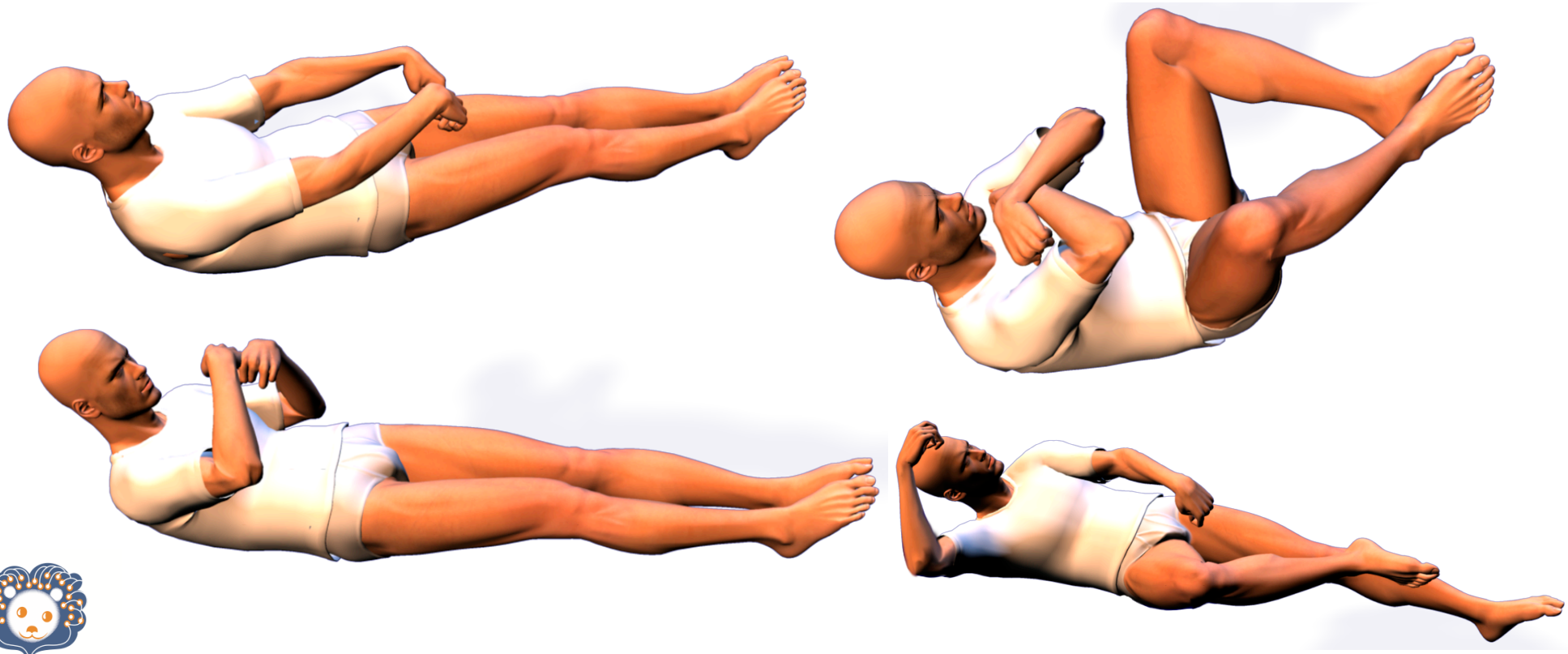
Message 1:

A detailed **description** of a clinical (paroxysmal) event experienced by your patient is far more important than all classification systems.



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Regards to Dr Lhatoo

Ictal phenomena:

Be **descriptive**, seek for information in a methodical way

(or just carefully review 10 times the videos from your patient's smartphone)

Teach your patient and his family how to furnish you with valuable information on semeiology;

Reconstruct (several times if needed), with the patient and / or his family, the temporal sequence of events

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The new ILAE proposal clearly supports such an approach

Operational Classification of Seizure Types by the International League Against Epilepsy (2016)

Cognitive
- acalculia
- aphasia
- attention impairment
- déjà vu
- dysphasia
- hallucinations
- illusions
- jamais vu
- memory impairment
- neglect
- forced thinking
- responsiveness impairment

Emotional or affective
- agitation
- anger
- anxiety
- crying (dacrystic)
- fear
- laughing (gelastic)
- paranoia
- pleasure

Autonomic
- asystole
- bradycardia
- cold
- erection
- flushing
- gastrointestinal
- heat
- hyperventilation
- hypoventilation
- nausea or vomiting
- pallor
- palpitations
- piloerection
- tachycardia

Common Descriptors of Behaviors During Focal Seizures (alphabetically in English)

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The new ILAE proposal clearly supports such an approach

Operational Classification of Seizure Types by the International League Against Epilepsy (2016)

Sensory
- auditory
- gustatory
- olfactory
- somatosensory
- vestibular
- visual

Automatisms
- aggression
- manual
- oral-facial
- perseveration
- sexual
- undressing
- vocalization
- walking/running

Motor
- arrest
- astatic
- dysarthria
- dystonic
- fencer's posture
- figure-4
- hypomotor
- hypokinetic
- hypermotor
- incoordination
- Jacksonian
- paralysis
- paresis
- pedaling
- pelvic thrusting
- versive

Common Descriptors of Behaviors During Focal Seizures (alphabetically in English)

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Message 2:

Become familiar with:

- Various types of seizures
- Main features and characteristics
- Patterns of appearance

Infantile spasms

usually occur in clusters…

Tonic seizures

are almost exclusively encountered in NON-idiopathic forms of epilepsy…

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Atypical absences

- are rarely the sole type of seizure…

Typical absences

- usually occur several times a day…

Myoclonic seizures

- are usually associated with other seizure types……
- and these associations provide valuable clues for a syndromic diagnosis

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Become familiar with …

Lennox-Gastaut Syndrome, Doose Syndrome, Syndromes with Predominantly Myoclonic seizures

LGS	LGS + myocl.	MAE	BMyoclEpil

ATYPICAL ABSENCES

TONIC SEIZURES

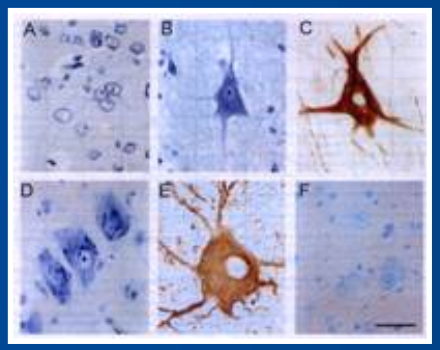
MYOCLONIC SEIZURES

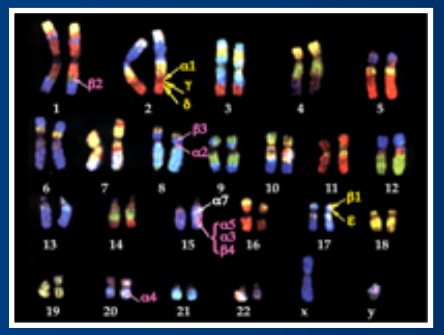
ATONIC (ASTATIC) SEIZURES [mechanism and EEG may differ]

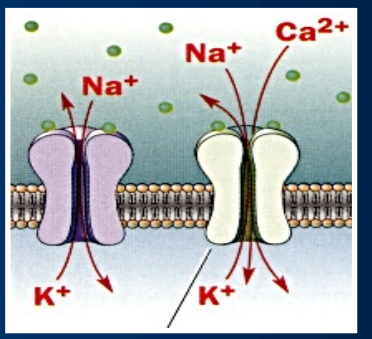
NONCONVULSIVE SE

10 Hz Slow Spike-Waves

Fast Spike-Waves







Arzimanoglou and Aicardi 2004

Last but not least

Almost 60% of seizures are focal while only 23% are generalised TCL…

- **…therefore, secondarily generalised seizures are much more frequent than is usually stated in emergency departments**

Become familiar with …

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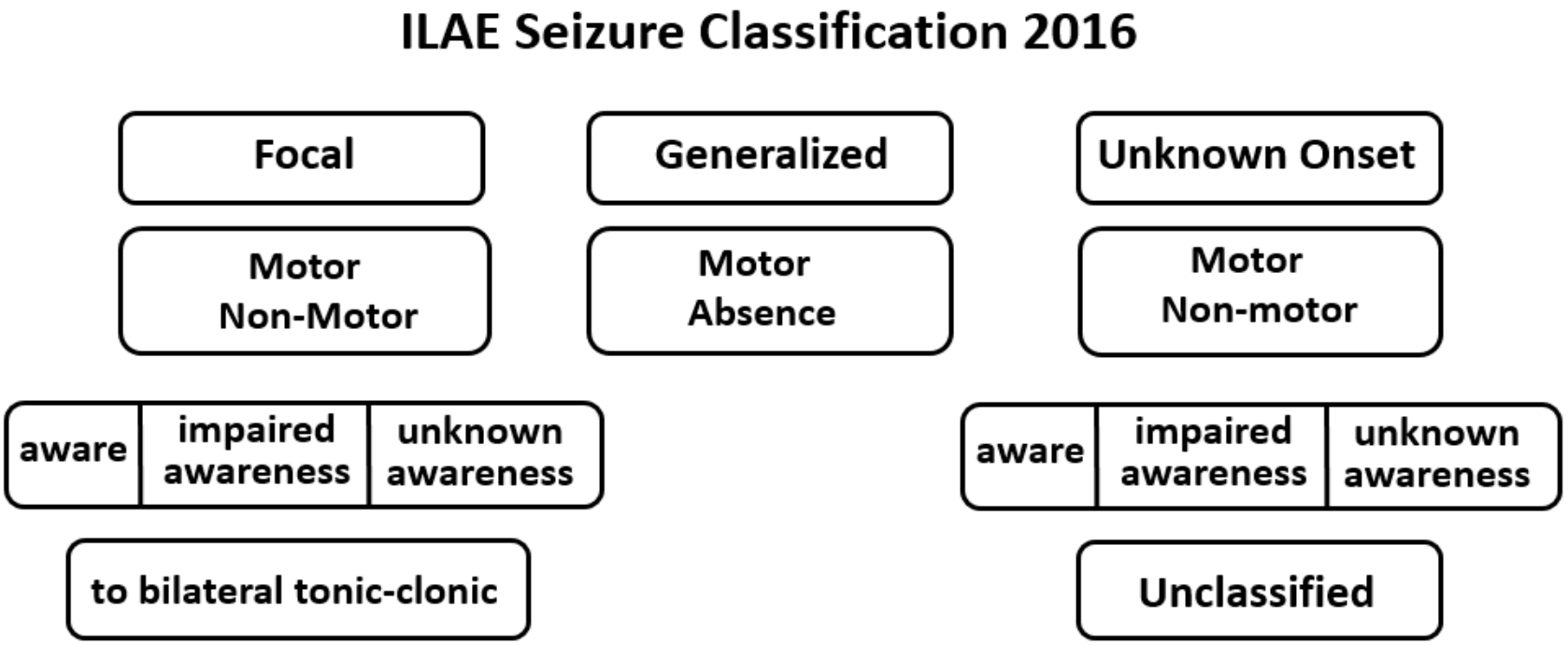

Modern research changed our view of the pathophysiological mechanisms involved and has shown **epilepsy to be a network disease** and not only a symptom of local brain abnormalities. **From a network perspective, seizures could arise** in neocortical, thalamo-cortical, limbic, and brainstem networks.

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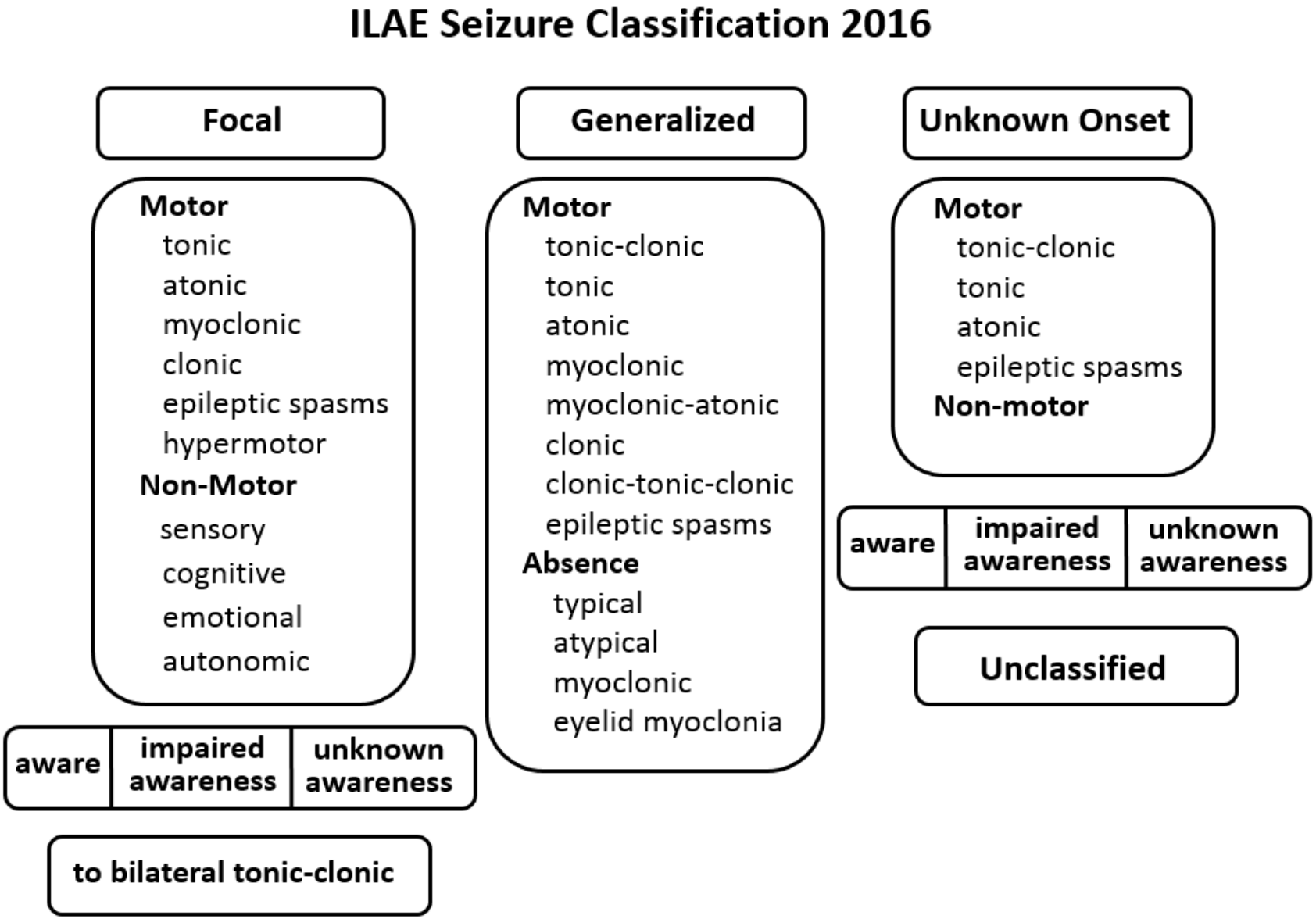

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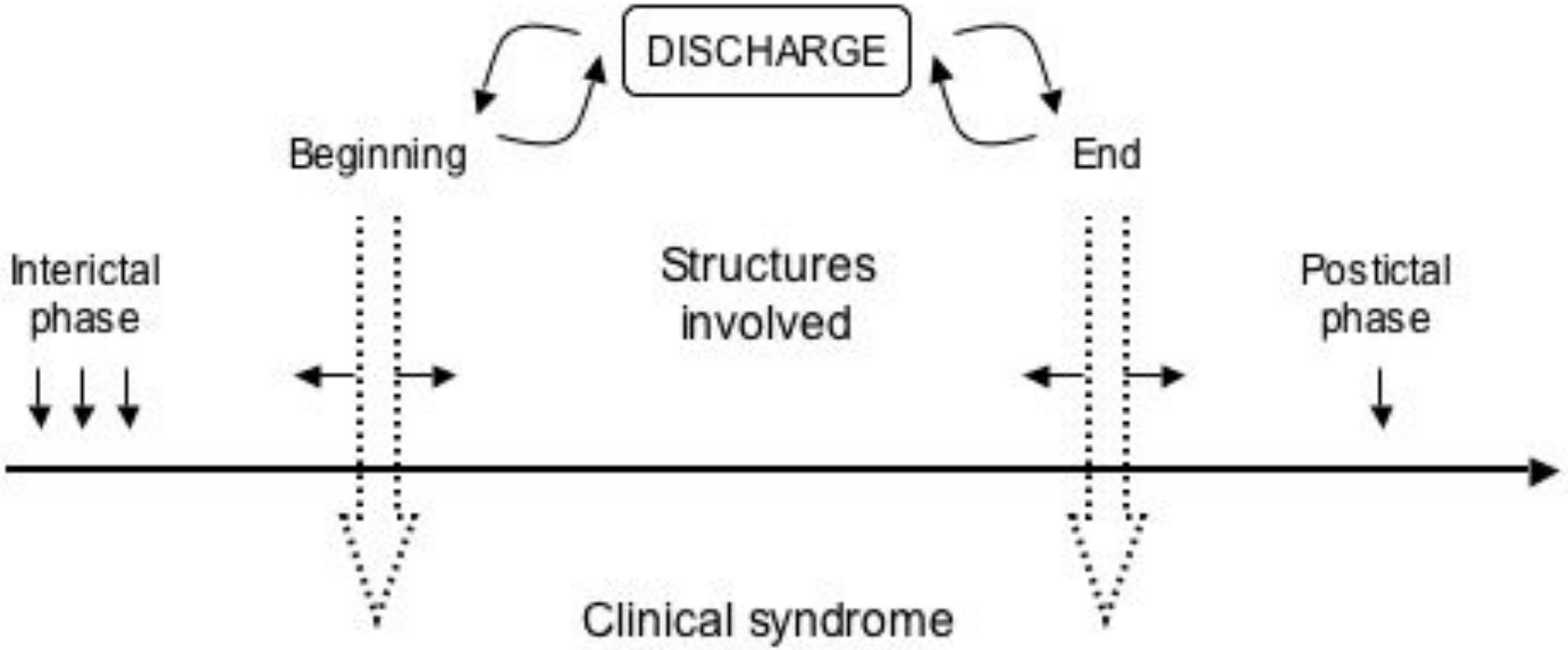
Operational Classification of Seizure Types by the International League Against Epilepsy (2016)

1. Change of “partial” to “focal”
2. Certain seizure types can be either focal, generalized or onset unknown
3. Seizures of unknown onset can still be classified
4. Awareness is used as a classifier of focal seizures
5. The terms dyscognitive, simple partial, complex partial, psychic, secondarily generalized were eliminated.
6. Focal (unihemispheric) tonic, clonic, atonic, myoclonic and epileptic spasms seizure types are recognized, along with bilateral corresponding seizure types.
7. Addition of new generalized seizure types: absence with eyelid myoclonia, myoclonic absence, clonic-tonic-clonic, myoclonic-atonic, epileptic spasms.

The best illustration: description of focal seizures



Children are not small adults

- *lack of aura*
- *epileptic spasms*
- *subtle symptoms*
- *rapid spreading*
- *diffuse abnormalities*
- *benign spikes*

Courtesy Philippe Kahane

Complex partial seizures of temporal lobe origin in children of different age groups.

Brockhaus A, Elger CE. Epilepsia 1995; 36(12): 1173-1181.

The semiology of complex partial seizures (CPS) of temporal lobe origin in adults is well known and is important in establishing seizure localization in patients considered for epilepsy surgery. In contrast, the behavioral features of temporal lobe seizures (TLS) in children described in the literature have not been consistent. In the present study, we investigated children with TLS to compare their attacks to TLS occurring in adults. The study was based on video recordings of 29 children with TLS aged 18 months to 16 years. Children were included, if they became seizure-free after temporal lobectomy (except 4 children with a marked reduction in seizure frequency and 1 with isolated auras), and if clear unitemporal seizure onset in ictal EEG-recordings, unilateral radiological lesions, and corresponding histopathological findings were detected. Children aged > 6 years had TLS with features similar to those of adults. **In younger children, typical semiology included symmetric motor phenomena of the limbs, postures similar to frontal lobe seizures in adults, and head nodding as in infantile spasms.** We concluded that the clinical features of TLS in younger children can be misleading and should therefore be considered with caution in selecting patients for surgical procedures on the temporal lobe.

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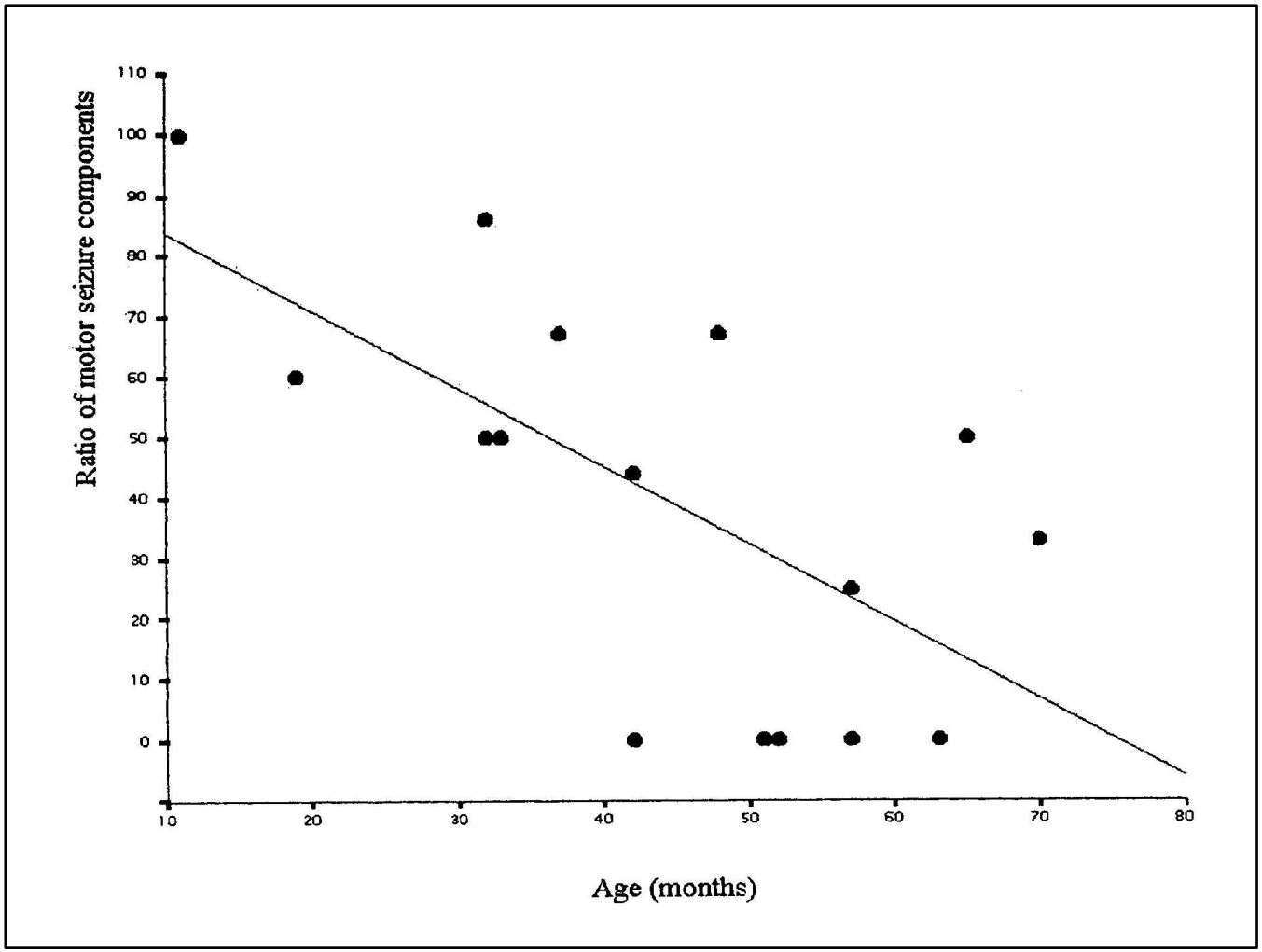
We concluded that the clinical features of TLS in younger children can be misleading and should therefore be considered with caution in selecting patients for surgical procedures on the temporal lobe.

The Effect of Age on Seizure Semiology in Childhood Temporal Lobe Epilepsy

Epilepsia 2002; 43(6): 638-643

*†András Fogarasi, *Hennric Jokeit, *Eduardo Faveret, *‡József Janszky, and *Ingrid Tuxhorn



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Temporal Lobe Seizures: classical features

- daytime, a few days a month
- SGTCS & status epilepticus : rare
- Non-motor manifestation (auras) : frequent

(vegetative, psychic, experiential)

- consciousness impairment : frequent
- automatisms : frequent (oral, gestural, verbal)
- motor signs : rare (except dystonic posturing)
- post-ictal confusion

Frontal lobe epilepsy in childhood (1995-2000 period)

Sinclair et al. Pediatr Neurol 2004 ; 30 : 169-176

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21 children studied by VEEG (mean age 11.3 yrs, range 6-16 years)

✓ Age of onset : mean 7.5 yrs (10 months - 16 years)

✓ Family history of epilepsy in 9/21 (suggestive of ADNFLE in 2)

✓ **Referring dg : sleep disturb. (10/21), psychiatric problems (6/21)**

✓ **Szrs : stereotypic, brief (30s-2'), nocturnal (17/21), frequent (3-22/d)**

✓ Interictal EEG & MRI normal in most cases (N EEG: 18; N MRI: 18)

✓ Seizure control difficult in 10/21 (3 went on to epilepsy surgery)

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Differentiation between pediatric FLE and MTLE

Lawson et al. Neurology 2002 ; 58 : 723-729

Characteristics	FLE	MTLE
No. of seizures/d, mean	6.9	1*
Telemetry, d, mean	2.2	4.9*
Event duration, s, mean	36	75*
Sleep/wake ratio, mean	0.66	0.24*
Aura, n	8 total†	12 total‡
Oral and gestural automatisms, n	1	15*
Nonclonic, motor, n	0	7*
Focal clonic, n	7	0*
Motor agitation, n	7	0*
Paroxysmal arousal, n	2	0
Staring only, n	2	0
Vocalization, n	3	0
Secondary generalization, n	3	0
Symmetric bilateral tonic, n	2	0
Asymmetric bilateral tonic, n	21	0*

56 children (9-14.5 yrs)

39 FLE and 17 MTLE

FL szrs were ($p<0.01$) :

- ✓ more frequent
- ✓ briefer duration
- ✓ sleep predominance
- ✓ focal clonic
- ✓ motor agitation
- ✓ asymmetric tonic

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FL seizure semiology in children under 7 yrs

Fogarasi et al. Epilepsia 2001 ; 42 : 80-85

Seizure type	Frequency among patients	Frequency among seizure onsets
Tonic	9 (64%)	39 (35%)
Clonic	5 (36%)	27 (24%)
Epileptic spasm	5 (36%)	18 (16%)
Myoclonic	1 (7%)	7 (6%)
Psychomotor	2 (14%)	8 (7%)
Hypomotor	2 (14%)	6 (5%)
Unclassified motor	1 (7%)	4 (4%)
Isolated aura	1 (7%)	2 (2%)
Total	14 patients (100%)[a]	111 seizures (100%)

[a] Ten of the 14 patients had more than one seizure type.

Frontal Lobe Seizures : classical features

- sleep, in clusters (relative frequency of SE)
- inconsistency of Secondarily Generalized TCS
- brief attacks (20-50 sec)
- sudden onset - possible auras (polymorphic)
- early - and often predominant - motor signs
- consciousness impairement variable
- rapide postictal recovery

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Posterior Cortex Sseizures : semiology is diverse

- Visual auras
- Somatosensory auras
- Vestibular auras
- Gustatory auras
- Adversive and oculoclonic seizures
- Eyelid flutter, rapid blinking
- Anosognosia, apraxia, acalculia, alexia, aphemia

Sveinbjornsdottir & Duncan, Epilepsia 1993, 343: 493-521.

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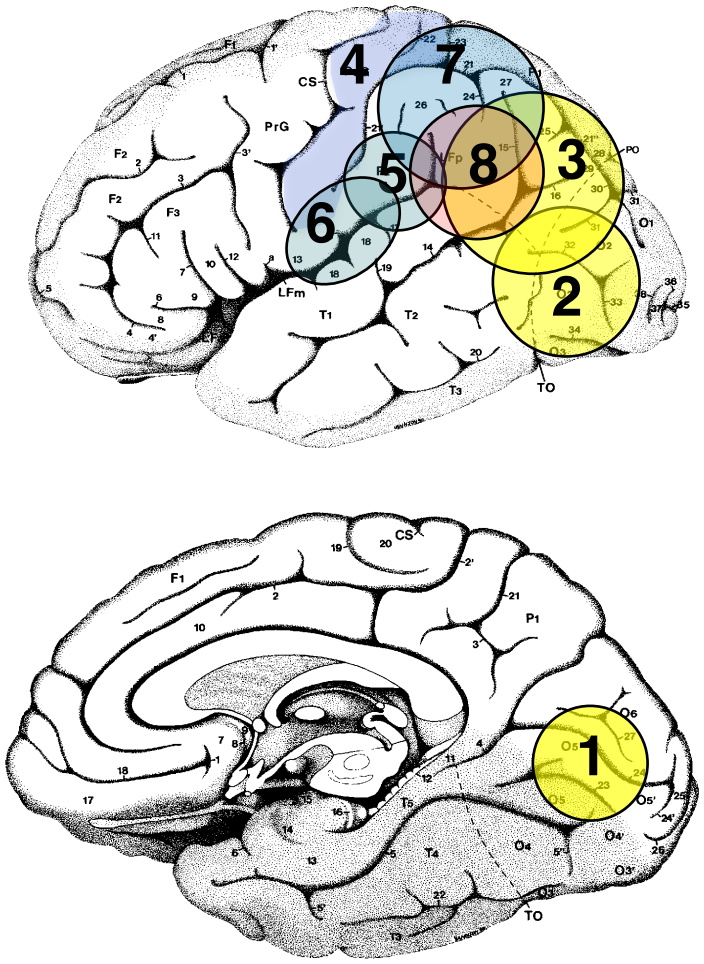


Courtesy Philippe Kahane

VISUAL AURAS

1. elementary visual hallucinations amaurosis
2. visual illusions
3. complex visual hallucinations

SOMATO-SENSORY AURAS

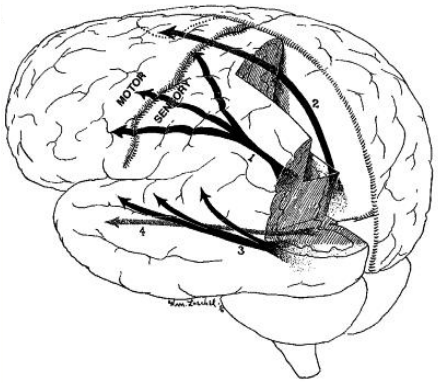
4. paresthesiae
5. pains
6. sensations of warmth and cold
7. somatognostic illusions

8. **VESTIBULAR AURAS**

70 seizures recorded in 32 children (30 aged less than 2 years) (Rathgeb et al. 1998)

1st ictal clinical sign	F (n=10)	T (n=13)	O (n=27)	C (n=10)	H (n=10)
Oculoclonia	0	0	3	0	0
Head +/- eyelid myocl.	1	0	1	1	0
Head or eye adversion	1	3	16	0	0
Uni/bilateral hypertonia	1	0	0	3/2	3/3
Unilateral myoclonias	0	0	0	1	1
Body movements	1	0	0	2	0
Staring or eye opening	6	10	7	1	1

Frontal Pattern



Temporal Pattern

The most common seizure components were motor manifestations (with myoclonic and tonic seizure), but psychomotor (automotor), hypomotor attacks, and isolated auras also were frequently observed…

Compared with adults, children with PCE have shorter but more frequent seizures, they rarely report auras or visual sensory signs, only sporadically develop hypermotor and 2d generalized tonic-clonic seizures, whereas ictal smile, flush, head node and behavioral change are typical features at this age.

Fogarasi et al. Epilepsia 2003; 44: 89-96.

Differentiation between childhood Frontal LE and Posterior Cortex Epilepsy

Fogarasi et al. Epilepsia 2005 ; 46 : 1280-1285

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35 children (11 mths - 12 years) - 20 FLE and 15 CPE

- ✓ Higher nocturnal dominance of seizures in FLE ($p<0.05$)
- ✓ Tonic seizures more frequent in FLE ($p<0.01$)
- ✓ Somatosensory aura and hypermotor seizures only in FLE
- ✓ Visual aura, nystagmus and versive seizures only in PCE

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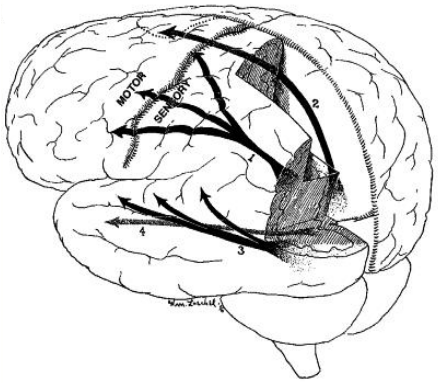
Frontal Pattern



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Epileptic Disorders

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VOLUME 16
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WITH VIDEOS

WWW.EPILEPTICDISORDERS.COM

John Libbey EUROTEXT

Original article

Epileptic Disord 2014; 16 (2): 141-64

Paediatric epilepsy surgery in the posterior cortex: a study of 62 cases*

Alexandra Liava [1,2], Roberto Mai [1], Laura Tassi [1], Massimo Cossu [1], Ivana Sartori [1], Lino Nobili [1], Giorgio Lo Russo [1], Stefano Francione [1]

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[2] Neuroscience Department, University of Milan-Bicocca, Milan, Italy

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CLASSIFICATION IS NOT A DIAGNOSTIC PROCESS *per se*

"As part of the **diagnostic process**, a clinician will commonly use supportive evidence to help to classify a seizure, even though that evidence is not part of the classification scheme.

Such evidence may include: EEG patterns, neuroimaging, laboratory results such as detection of anti-neuronal antibodies, gene mutations, family history or presence of an epilepsy syndrome known to be associated with either focal or generalized seizures.

The seizures usually can be classified on the basis of behavior, **provided that a good subjective and objective description is available**. However, where supportive information is available, it should be used to secure the most accurate classification."