Semiological Differences between Adult and Pediatric Epilepsy Patients

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Outline

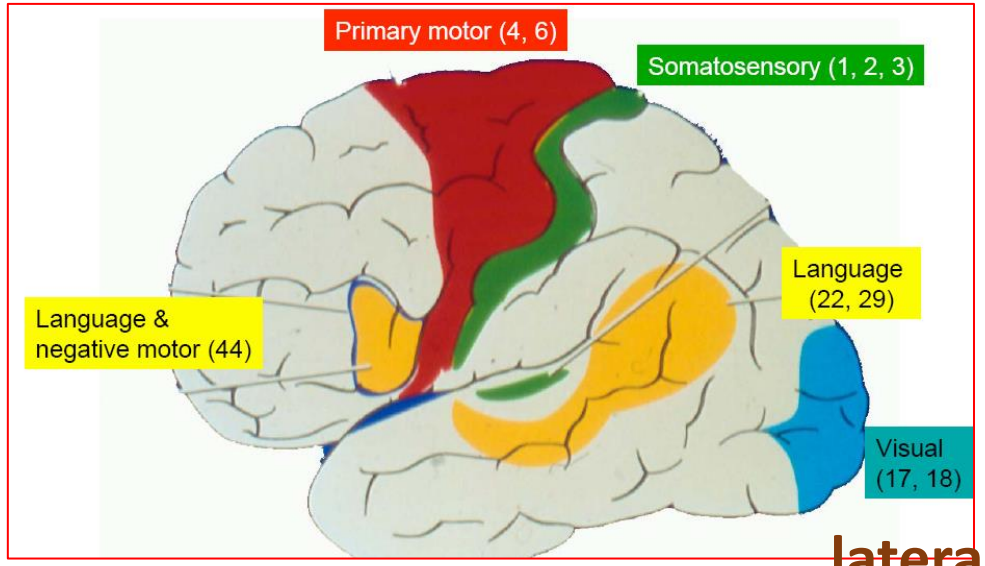
- Epileptic seizure semiology in different age groups
- Example of video semiology in Pediatric epilepsy patients

Seizure semiology

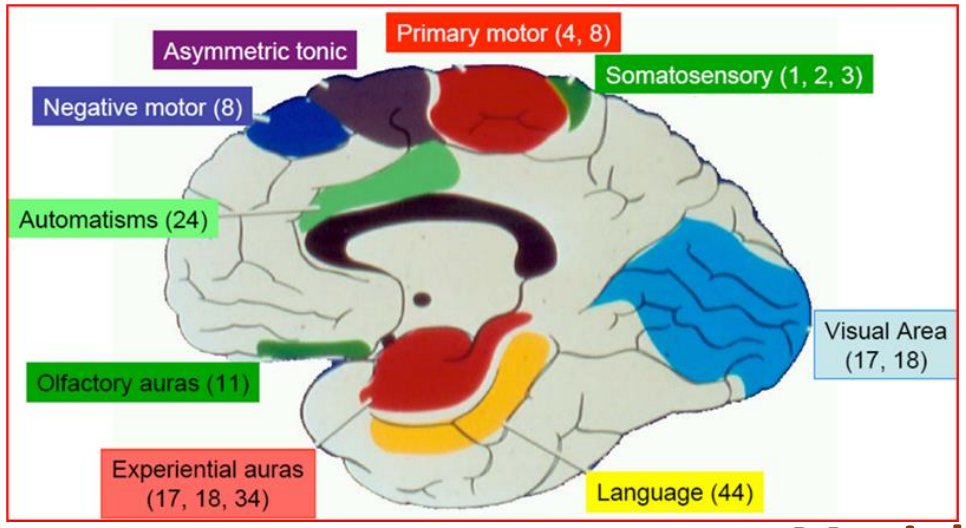
- Seizure semiology is the manifestation of the activation of the **symptomatogenic zone**
- *A simple and cost-effective tool* that allows localization of the symptomatogenic zone which either overlaps or close proximity of

  “the epileptogenic zone”

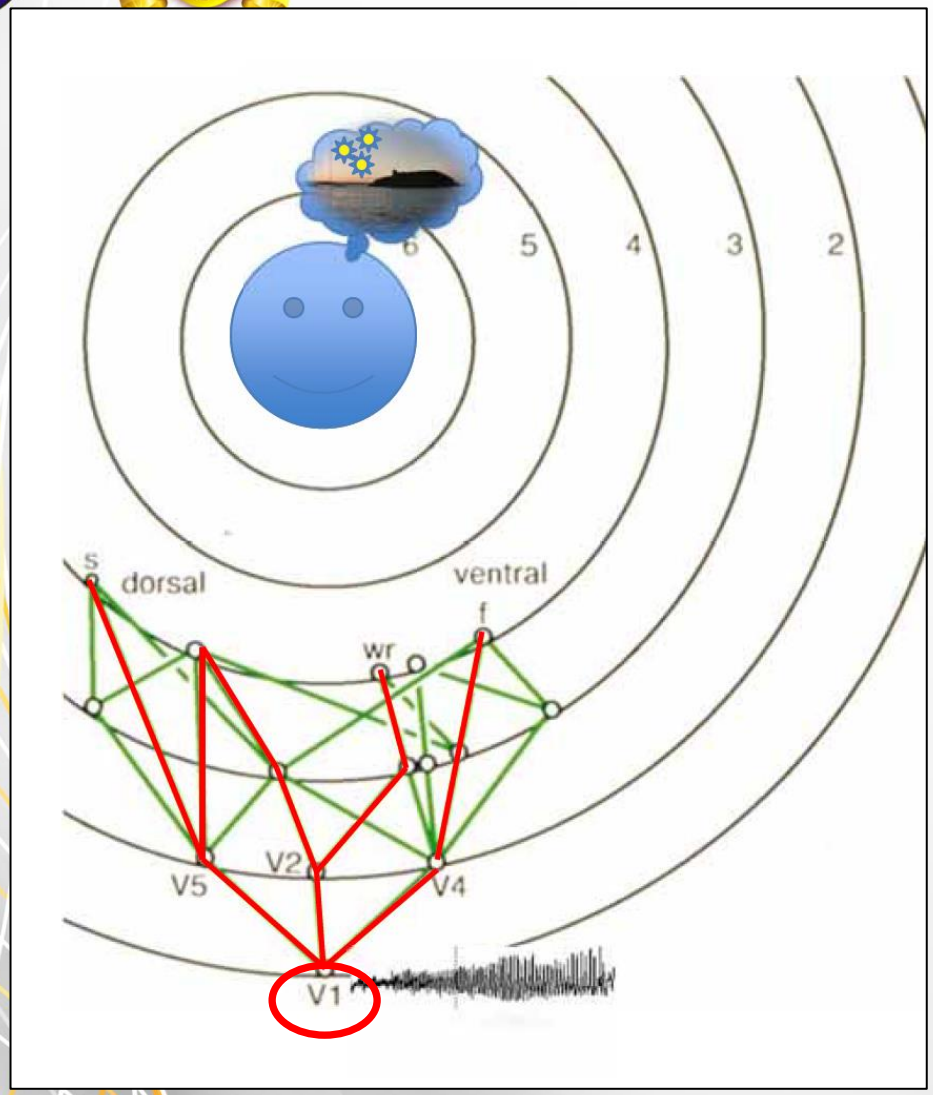
- Semiology is shaped by *cable wiring* of the brain and *hierarchical* organization of the cortex (Chauvel, 2014)



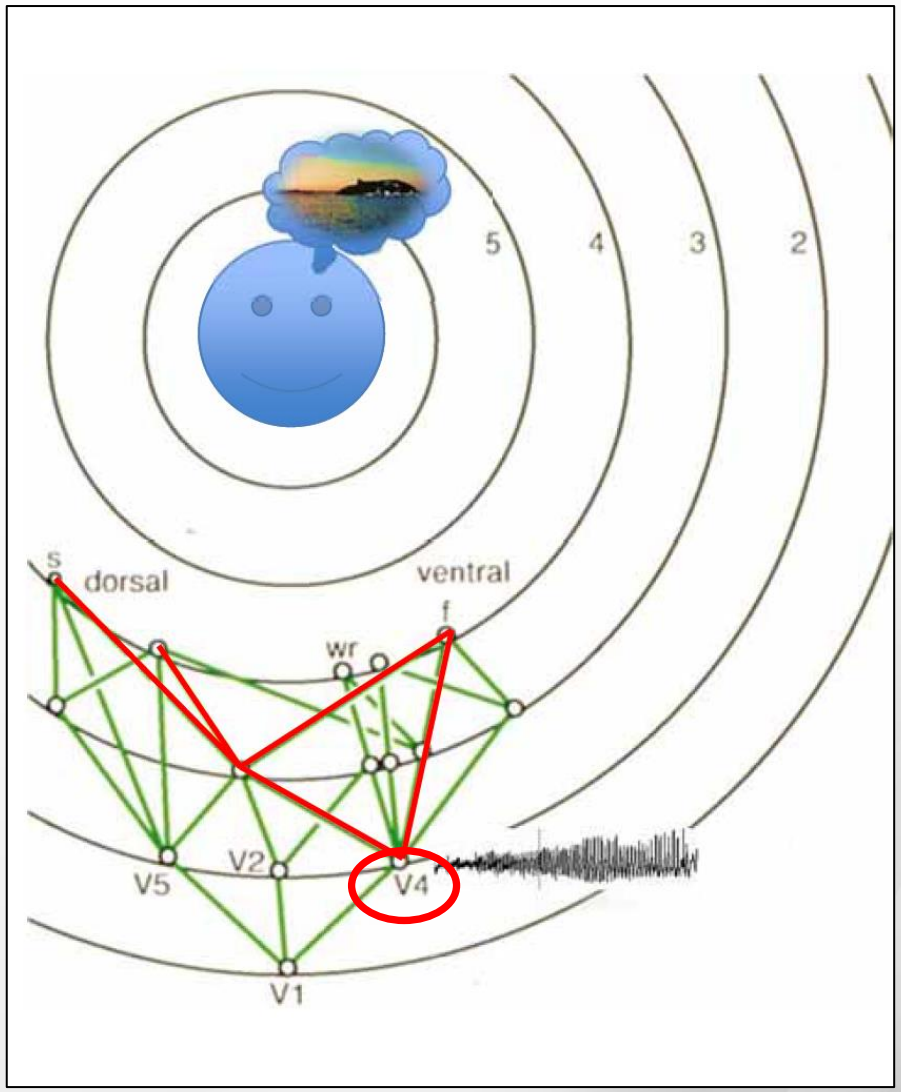
lateral



Mesial



Visual hallucination



Visual illusion

Halgren & Chauvel, 1993

Semiology in pediatric epilepsy patients

- The semiological analysis in children is often difficult
- Semiology is **related to age and cerebral maturation**
- Less detailed description of the seizure
- The younger children can not explain some semiology (such as aura, autonomic symptom, etc.)
- History taking and observation -> important!

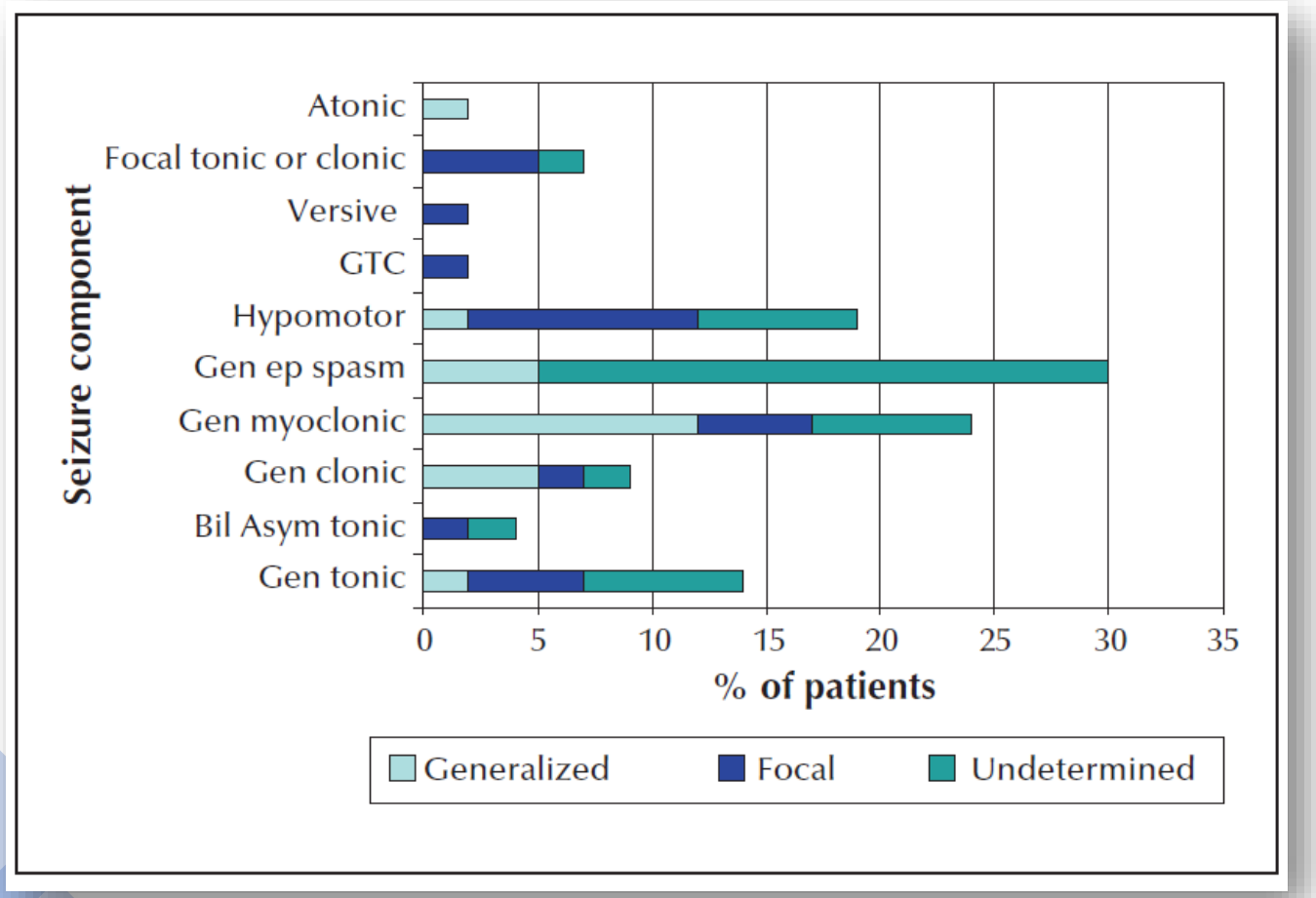
Epileptic seizure semiology in different age groups

- Retrospective studied seizure semiology in all age groups (1 month - 90 years old), n = 270 patients
- Group 1: >1 month to 3 years (n=36)
- Group 2: > 3 years to 6 years (n=22)
- Group 3: > 6 years to 10 years (n=33)

(> 10 years old, the seizure semiology closely resembled of the adult population)

Fernandez et al. Epileptic seizure semiology in different age groups. Epileptic Disord 2018

Group 1: >1 month to 3 years



- The most common seizure types
  - Epileptic spasm (30%)
  - Myoclonic seizure (24%)
  - Hypomotor seizure (19%)

Fernandez et al. Epileptic seizure semiology in different age groups. Epileptic Disord 2018

Case a 7-month-old female infant

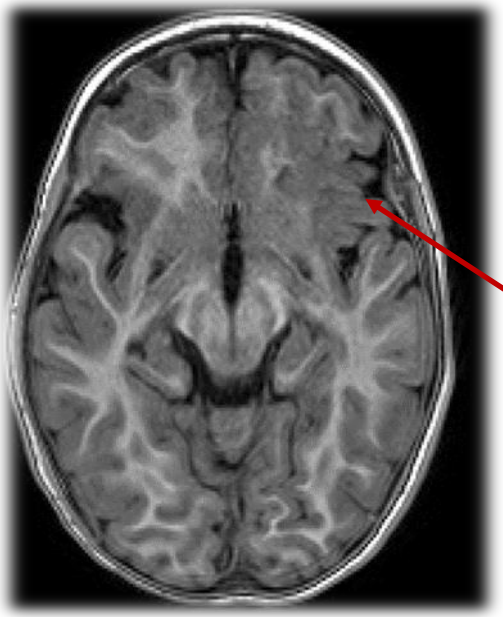
Epileptic spasms

- Sudden flexion, extension or mixed flexion-extension of proximal and truncal muscles, lasting 1-2 seconds
- Myoclonic jerk > Epileptic spasm > Tonic seizure
- Spasms typically **occur in a series**, usually on wakening
- Generalized epilepsies > focal epilepsy (parieto-occipital)

Tufenkjian K., Lüders H. J Clin Neurol. 2012

Generalized epileptic spasms in younger children with localization-related epilepsy may *falsely suggest generalized epilepsy*

Case a 2-year-old girl with intractable epilepsy and developmental delay



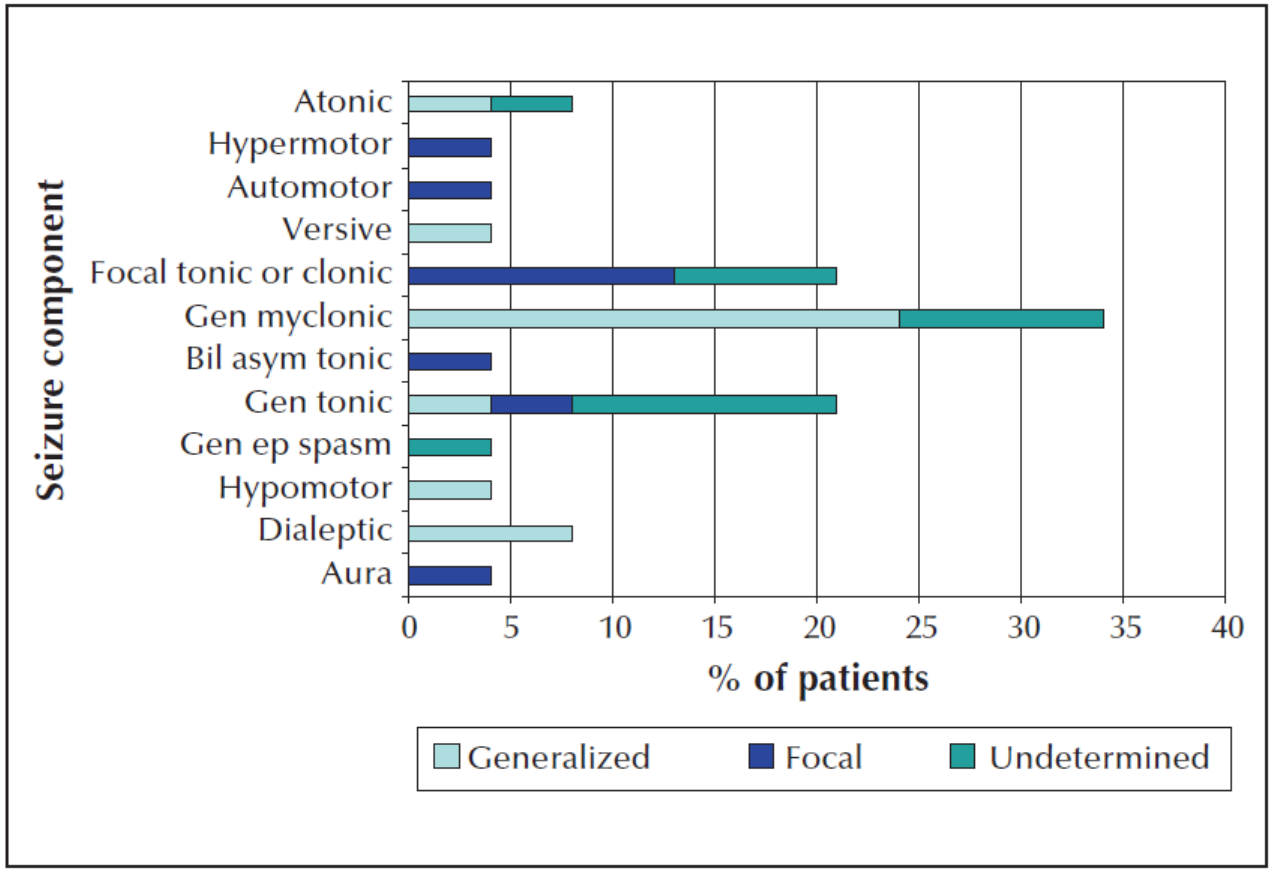
Focal cortical dysplasia

Hypomotor seizure

- The main manifestation of these seizures is a decrease or total absence of motor activity
- This expression is only used in patients in whom consciousness cannot be tested (newborns, infants and children under 3 years; mentally retarded patients)
- Hypomotor seizures may be either generalized or focal (frequently in temporal and parietal regions)

Källén K et al. Hypomotor seizures in infants and children. Epilepsia. 2002

Group 2: > 3 years to 6 years



- The most common seizure types
  - Generalized myoclonic seizure (34%)
  - Generalized tonic seizure (21%)
  - Focal tonic or clonic seizure (21%)

  Simple motor seizure
- This was the youngest age group in which we observed **auras and automotor seizures**
- Dialeptic and hypomotor seizures were not frequent, and these occurred only with a generalized EZ

Fernandez et al. Epileptic seizure semiology in different age groups. Epileptic Disord 2018

Myoclonic seizure

- Sudden muscle jerks of variable topography (distal, proximal, axial): uni- or bilateral, focal, multifocal or generalised
- Prominently affecting shoulders and proximal arms
- Consciousness likely preserved
- 100-400 msec in duration
- Lateralization: Unilateral myoclonic seizures -> contralateral primary motor area or premotor cortex

Epilepsia partialis continua (EPC)

- Spontaneous regular or irregular clonic muscular twitching affecting a limited part of the body, sometimes aggravated by action or sensory stimuli,occurring for a minimum of one hour, and recurring at intervals of no more than ten seconds[1]
- Localization: involving a small portion of the contralat. **sensorimotor cortex**
- The pathologies that underlie EPC are heterogeneous and may differ between adults and children
- The main diagnoses of EPC in children[2]:
  - **Rasmussen's encephalitis**
  - Mitochondrial disease
  - MRI lesion-positive focal epilepsy
  - MRI lesion-negative EPC (inflammatory, neurometabolic, genetic)

[1]Bien CG et al. Epileptic Disord. 2008
[2]Surana et al. Epilepsia 2020

Criteria for Rasmussen's encephalitis

Part A	Need 3/3
1. Clinical	Focal seizures (+/− EPC) and unilateral cortical deficit(s)
2. EEG	Unihemispheric slowing +/− epileptiform activity and unilateral seizure onset
3. MRI	Unihemispheric focal cortical atrophy and at least one of the following: Grey or white matter T_2/FLAIR hyperintense signal Hyperintense signal or atrophy of the ipsilateral caudate head
Part B	**Need 2/3**
1. Clinical	EPC or Progressive unilateral cortical deficit(s)
2. MRI	Progressive unihemispheric focal cortical atrophy
3. Histopathology	T-cell–dominated encephalitis with activated microglial cells (typically, but not necessarily forming nodules and reactive astrogliosis Numerous parenchymal macrophages, B cells, or plasma cells or viral inclusion bodies exclude the diagnosis of RE

*Patients need to meet either A or B criteria

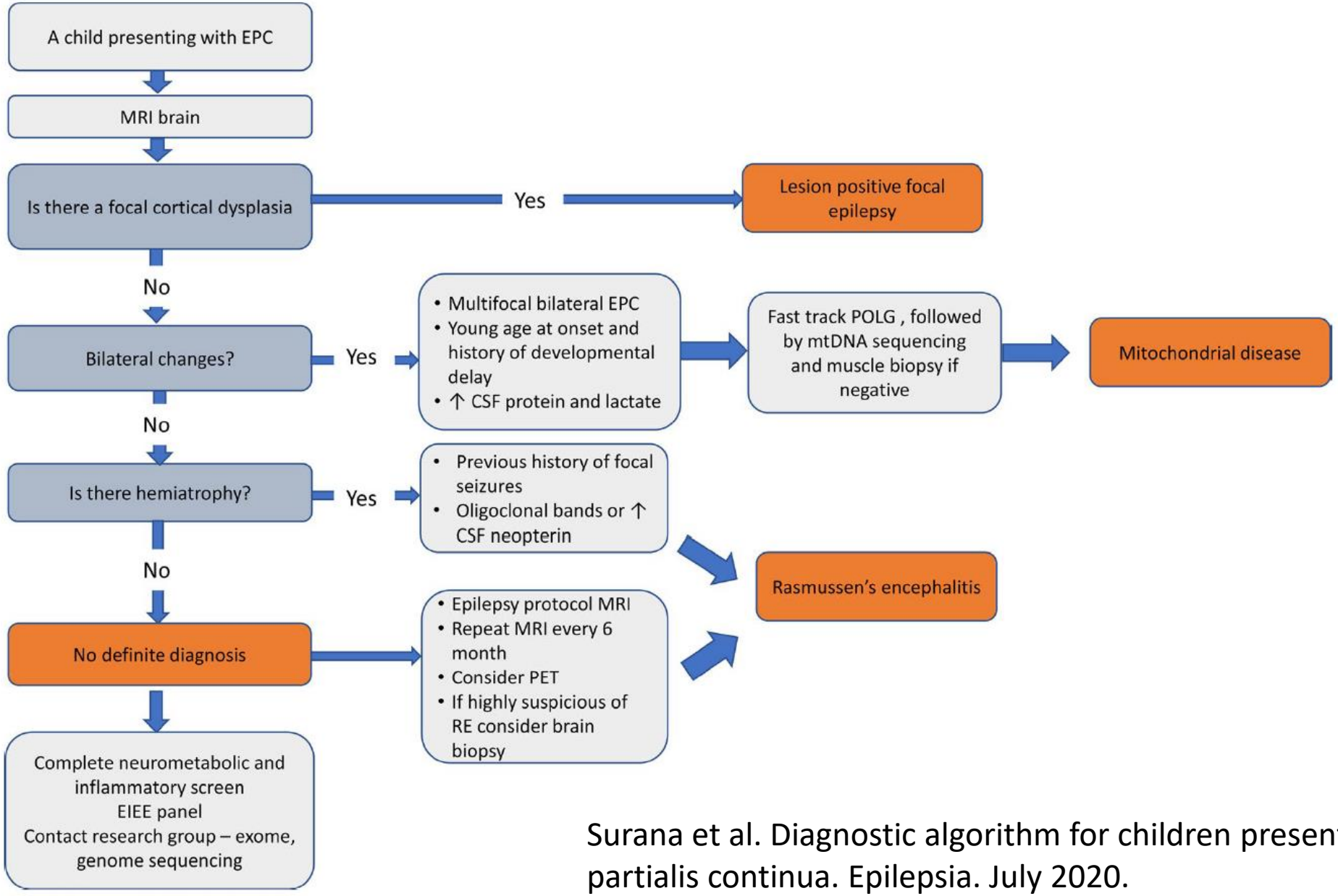
Bien CG et al. Pathogenesis, diagnosis and treatment of Rasmussen encephalitis: a European consensus statement. Brain 2005

Rasmussen's Encephalitis

- Age of onset usually 2-14 years
- Refractory focal epilepsy usually focal motor seizures
- Key features: Epilepsia Partialis Continua (EPC) in 75%
- Progressive hemiparesis, cognitive decline, and language impairment (dominant side)
- Progressive hemispheric atrophy on MRI

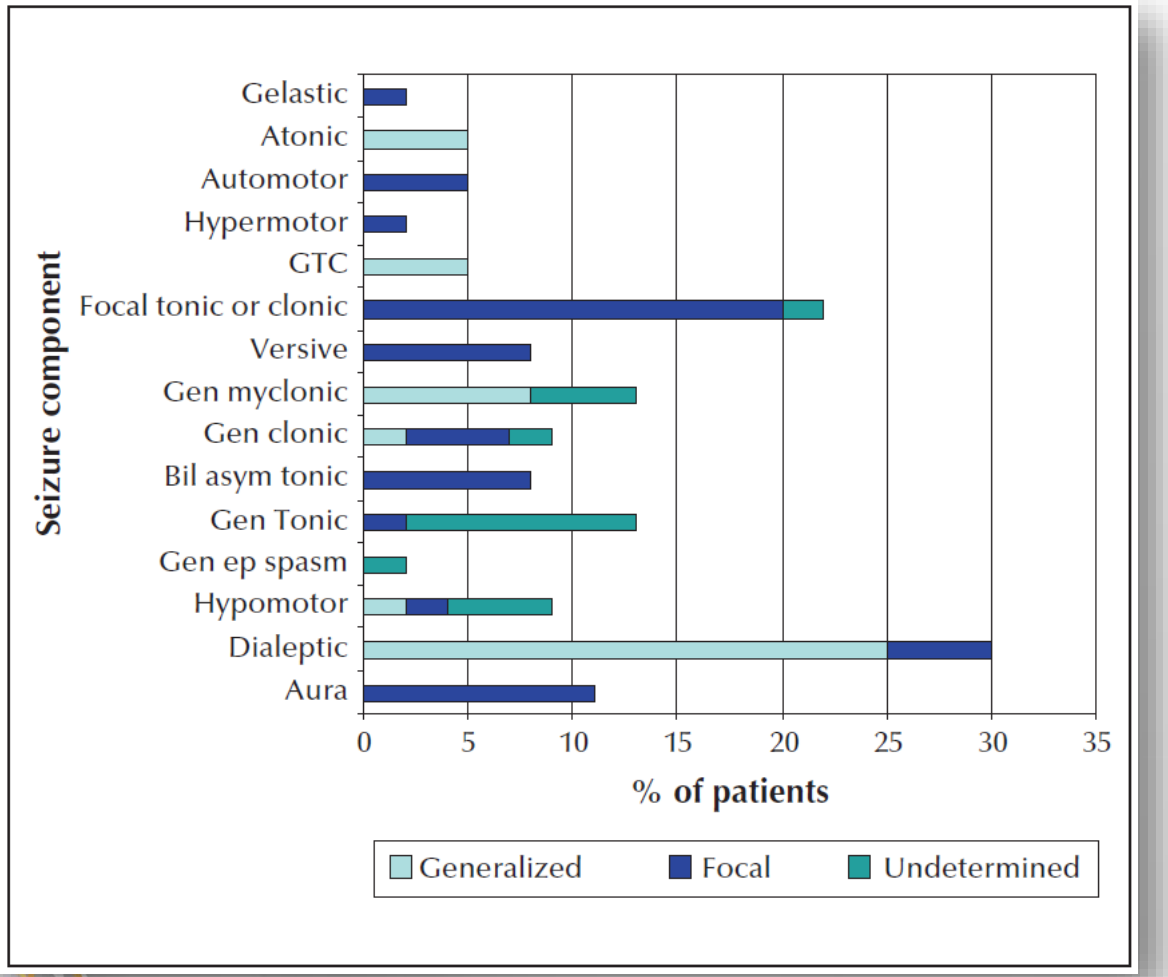
Bien CG et al. Brain 2005

A diagnostic algorithm for a child presenting with epilepsia partialis continua (EPC)



Surana et al. Diagnostic algorithm for children presenting with epilepsia partialis continua. Epilepsia. July 2020.

Group 3: 6 years to 10 years



- The most common seizure types
  - Dialeptic seizure (30 %)
  - Focal tonic or clonic (21 %),
  - Myoclonic (13 %)
  - Tonic (13 %)
- Less common seizure types
  - Gelastic seizure
  - Hypermotor

Fernandez et al. Epileptic seizure semiology in different age groups. Epileptic Disord 2018

Focal Onset Impaired Awareness Seizures (Dialeptic seizures)

- An alteration of consciousness consisting of unresponsiveness during the seizure and amnesia of the episode post-ictally
- Typically last 1 to 2 minutes.
- These seizures include automatisms (such as lip smacking, picking at clothes), becoming unaware of surroundings, and wandering.
- Not localized or lateralized
- Duration of seizures has a localizing value
  - Mesial temporal seizure -> longer duration than frontal lobe seizure

Wyllie E. Neurology 1986

Semiology and Epileptic Networks

Aileen McGonigal, MD, PhD[a,b]

Table 2
Examples of studies examining semiology in conjunction with signal analysis of stereoelectroencephalography

Investigators, Year	Semiological Pattern	Epilepsy Localization	Main Anatomic Structures	Signal Analysis	Change in Network Synchrony
Bartolomei et al,[51] 2002	Humming	Temporal lobe	STG, prefrontal cortex	Rhythmic discharge over STG (6 or 15 Hz). Increased coherence between STG and prefrontal cortex	Increased
Bartolomei et al,[52] 2005	Fear behavior	Prefrontal cortex	Ventromesial orbitofrontal cortex, anterior cingulate, amygdala (limbic system)	Sudden loss of synchrony between orbitofrontal cortex and amygdala at seizure onset/clinical onset	Decreased
Arthuis et al,[53] 2009	Impaired consciousness	Temporal lobe	Temporal structures, parietal lobe, thalamus	Excessive synchrony; ie, functional coupling, between temporal and extratemporal structures, notably parietal cortex and thalamus	Increased
Bartolomei et al,[54] 2012	Déjà vu	Mesial temporal lobe	Rhinal cortices, hippocampus	Increased high-frequency EEG signal correlation between mesial temporal structures in seizures producing déjà vu	Increased
Lambert et al,[55] 2012	Impaired consciousness	Parietal lobe	Superior and inferior parietal lobules, precuneus, parietal operculum, supplementary motor area	Increased synchrony was associated with progressively greater degrees of altered responsiveness. A statistically significant nonlinear relationship was found between h2 values and degree of alteration of consciousness, suggesting a threshold effect	Increased
Aupy et al,[56] 2018	Oroalimentary automatisms	Temporal lobe	Medial basal temporal lobe, opercular cortex	Increased coherence occurred between mediobasal temporal structures and insulo-opercular cortex before onset of rhythmic chewing movements	Increased

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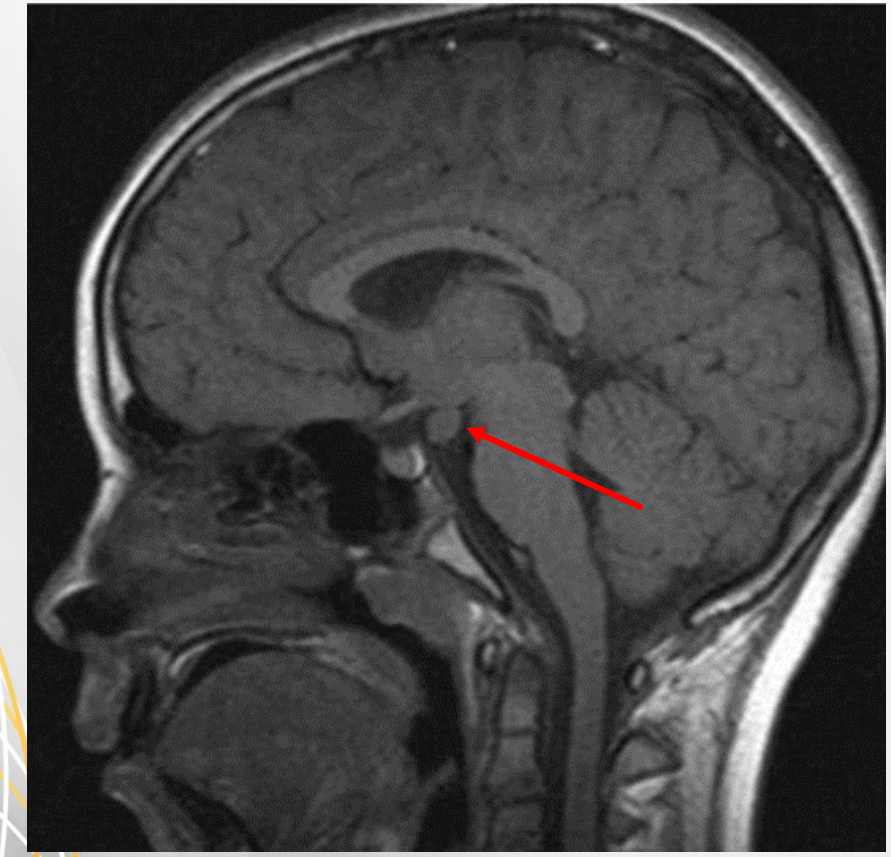
Gelastic seizure



- Characterized by unnatural forceful laughter with or without mirth
- This seizure type is characteristic of seizures arising in the hypothalamus (*Hypothalamic hamartoma*)
- But can occur in seizures arising in the *frontal* or *temporal* lobes.

Gelastic seizure

HH	Frontal lobe origin	Temporal lobe origin
• Mirthless • Usually begin in infancy or early childhood • Preserved consciousness • Autonomic symptoms	• Mirthless • Motor component	• Pleasurable aura or mirthful

- **Clouding of consciousness and impairment of memory**
- **Later age of onset**

Jeremy L Freeman, Orvar Eeg-Olofsson. Epilepsy: A comprehensive textbook 2nd edition

Summary

- The semiological analysis in children is different from adult
  - Less detailed description
  - Semiological expression is related to age and cerebral maturation
    (The patients < 3 years exhibited mostly generalized simple motor seizures and hypomotor seizures)
  - Does not always permit differentiation of focal or generalized epilepsies

It is necessary to

- Record sufficient number of seizures
- Look for consistency between seizures, look for specific signs
- Identifying features in common is the key to categorization
- May need to interpret with other tools (EEG, MRI, etc.)

Thank you for your attention