Seizure semiology in children

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Seizure Semiology

- Description of ictal events
- Using standardized glossary of descriptive terminology

Table 2. Glossary of terms

Word	Definition	Source
Absence, typical	A sudden onset, interruption of ongoing activities, a blank stare, possibly a brief upward deviation of the eyes. Usually the patient will be unresponsive when spoken to. Duration is a few seconds to half a minute with very rapid recovery. Although not always available, an EEG would show generalized epileptiform discharges during the event. An absence seizure is by definition a seizure of generalized onset. The word is not synonymous with a blank stare, which also can be encountered with focal onset seizures	Adapted from Ref. 12
Absence, atypical	An absence seizure with changes in tone that are more pronounced than in typical absence or the onset and/or cessation is not abrupt, often associated with slow, irregular, generalized spike-wave activity	Adapted from Ref. 11
Arrest	See behavior arrest	New
Atonic	Sudden loss or diminution of muscle tone without apparent preceding myoclonic or tonic event lasting ~1–2 s, involving head, trunk, jaw, or limb musculature	12
Automatism	A more or less coordinated motor activity usually occurring when cognition is impaired and for which the subject is usually (but not always) amnesic afterward. This often resembles a voluntary movement and may consist of an inappropriate continuation of preictal motor activity	12
Autonomic seizure	A distinct alteration of autonomic nervous system function involving cardiovascular, pupillary, gastrointestinal, sudomotor, vasomotor, and thermoregulatory functions	Adapted from Ref. 12
Aura	A subjective ictal phenomenon that, in a given patient, may precede an observable seizure (popular usage)	12

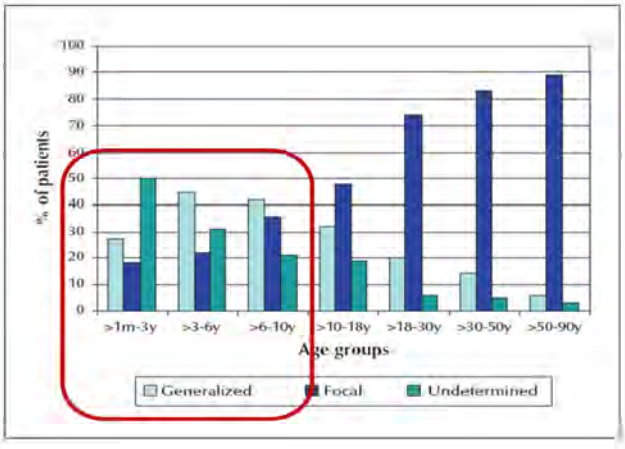
Fisher RS, et al. Epilepsia 2017;58(4):531-42

Seizure Semiology

- Produced by interactions between seizure onset and propagation
- Help to localize the epileptogenic zone (EZ)
  important especially in epilepsy surgery
- Seizure types changes with age:
  probably due to the maturation of the cortex

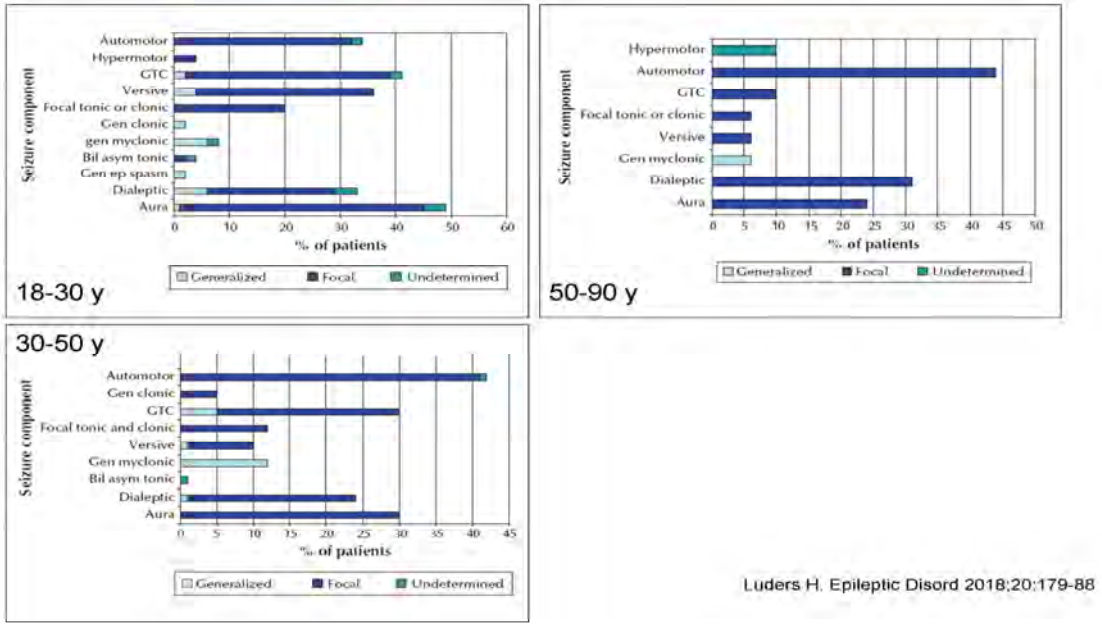
McGonigal A. Neurosurg Clin N Am 2020;31:373-85

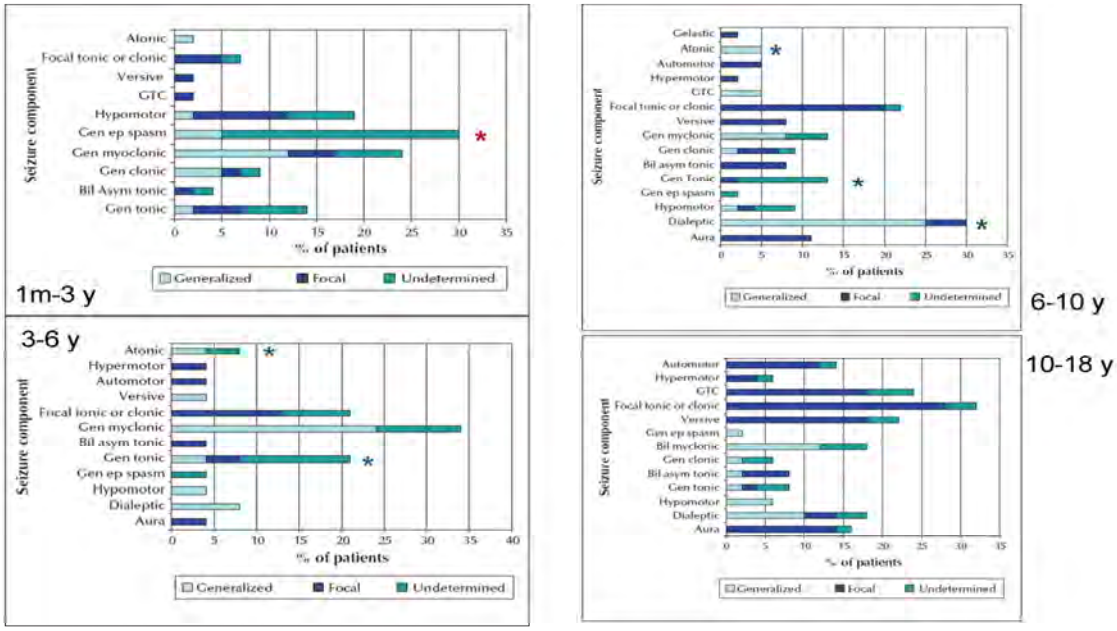
Types of EZ according to the Age Groups



- Focal EZ dominantly in adults (age 10+)
- Generalized EZ dominantly in pediatrics
- Undetermined EZ under 3y group

Luders H. Epileptic Disord 2018;20:179-88


18-30 y
Automotor
Hypermotor
GTC
Versive
Focal tonic or clonic
Gen clonic
gen myclonic
Bil asym tonic
Gen ep spasm
Dialeptic
Aura
Seizure component
% of patients
Generalized
Focal
Undetermined
50-90 y
Hypermotor
Automotor
GTC
Focal tonic or clonic
Versive
Gen myclonic
Dialeptic
Aura
30-50 y
Automotor
Gen clonic
GTC
Focal tonic and clonic
Versive
Gen myclonic
Bil asym tonic
Dialeptic
Aura
Luders H. Epileptic Disord 2018;20:179-88

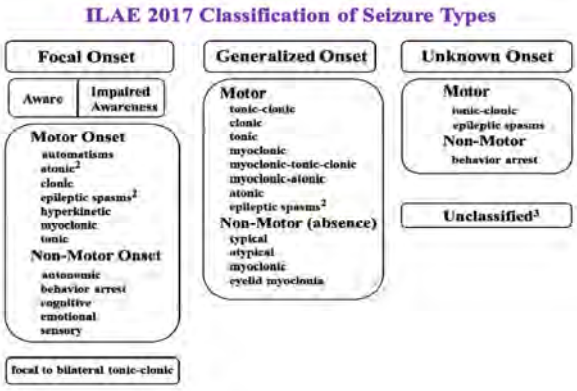

1m-3 y
Atonic
Focal tonic or clonic
Versive
GTC
Hypomotor
Gen ep spasm
Gen myoclonic
Gen clonic
Bil Asym tonic
Gen tonic
Seizure component
% of patients
Generalized
Focal
Undetermined
3-6 y
Atonic
Hypermotor
Automotor
Versive
Focal tonic or clonic
Gen myclonic
Bil asym tonic
Gen tonic
Gen ep spasm
Hypomotor
Dialeptic
Aura
6-10 y
Gelastic
Atonic
Automotor
Hypermotor
GTC
Focal tonic or clonic
Versive
Gen myclonic
Gen clonic
Bil asym tonic
Gen Tonic
Gen ep spasm
Hypomotor
Dialeptic
Aura
10-18 y
Automotor
Hypermotor
GTC
Focal tonic or clonic
Versive
Gen ep spasm
Bil myclonic
Gen clonic
Bil asym tonic
Gen tonic
Hypomotor
Dialeptic
Aura

In this Session,

1 Epileptic spasms:
from infantile spasms to epileptic spasms in adult

2 Absence seizures
in childhood absence epilepsy

3 Generalized tonic and atonic seizures
in Lennox-Gastaut Syndrome



1 Epileptic spasms

- Sudden flexion, extension, or mixed extension-flexion of proximal and truncal muscles
- Brief (< 2 sec), but more than myoclonic jerk (< 100 msec) and less than tonic seizure (2-10 sec)
- Frequently occurring in clusters
- Most often during infancy (Infantile spasms), but spasms can occur at all ages.

Epileptic spasms

- Bilateral, symmetric, flexor or extensor spasms
- Asymmetric spasms
- Subtle spasms

- Ictal phenomenon associated with spasms
  - Autonomic changes: heart rate alterations, cyanosis, pallor, sweating, and flushing
  - Respiratory rate changes
  - Vocalizations: crying, laughter, and grunting sounds
  - Hiccups
  - Grimacing
  - Ocular movements: eye deviation, nystagmus

Spasms in Lennox-Gastaut Syndrome

Male / 15 years
- Seizure since 2 mo.
- Developmental delay and arrest
- Previous TPM, VPA, Pb, KD
- But, follow up loss for few years

- Recently increased seizure frequency, x freq/day
- GTC, atonic head drop, tonic seizures
- Lennox-Gastaut Syndrome on VPA LEV

- "요즘 많이 놀래요"

Epileptic Spasms in Adult Epilepsy patient

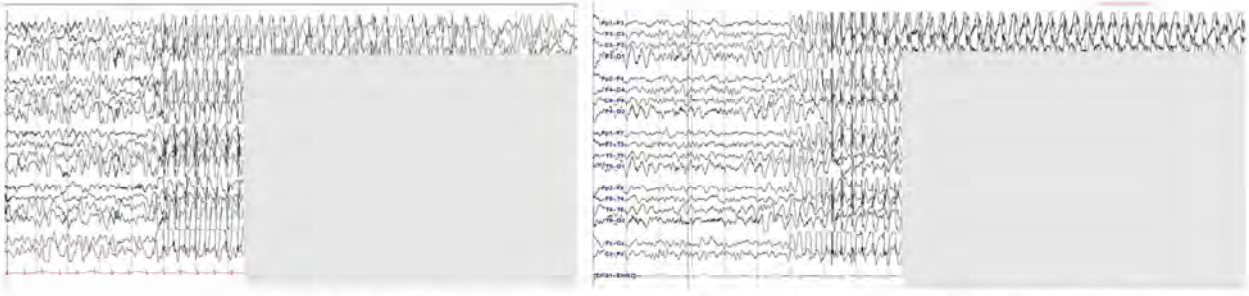
Female / 23 years

- Seizure since 13 years
- Developmental delay & Intellectual disability

- MRI: normal
- Interictal EEG: Right occipital spikes

- "머리와 목을 움찔거려요. 한번 할 때 10번 정도 몰아서 놀래요"

2 Typical absence seizure in Childhood absence epilepsy



- Sudden onset, a blank stare, possibly a brief upward deviation of the eyes
- Unresponsiveness and cessation of on-going activities
- Duration: usually lasting 8-20 seconds, very rapid recovery
- Abrupt onset of rhythmic 3-Hz generalized spike-wave activity in EEG

3 Lennox-Gastaut Syndrome (LGS)

- 1-10% of childhood epilepsies (1-2 % of all epilepsy)
- Severe form of epilepsy with onset in childhood
- Age at the onset of LGS: usually before 8 years

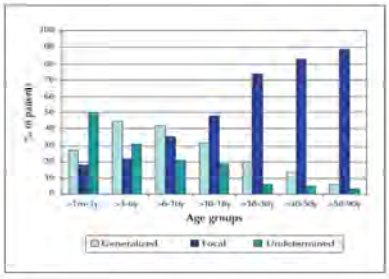
- **Symptomatic triad:**
  (1) Multiple drug-resistant seizure types:
  axial tonic, atonic and atypical absence
  (2) EEG abnormalities: bursts of diffuse slow spike-waves
  (3) Intellectual disability

Lennox-Gastaut Syndrome (LGS)

- Seizure types:

Tonic seizures (m/c), atypical absences, epileptic drop attack (~50%)

Conclusion



- Predominant seizure semiology *changes with age*.
  - Focal onset predominantly in adult
  - Generalized onset predominantly in children
    1. **Epileptic spasms**
    2. **Absence seizures in childhood absence epilepsy**
    3. **Generalized tonic and atonic seizures in Lennox-Gastaut Syndrome**

- Seizure semiology provides clue for the epileptogenic zone.
- It is important to describe detailed seizure semiology using standardized terminology.

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Thank you for your listening