SNOMED CT Web Series

Dr. Lynette Sadleir, Professor, University of Otago, Wellington (NZ)

SNOMED CT and Epilepsy: New International League Against Epilepsy seizure and epilepsy classifications

November 17, 2021 | 19:00 UTC

SNOMED CT CLINICAL WEBINAR

snomed.org/web-series linkedin.com/company/ihtsdo twitter.com/SnomedC

Julie – 2 years old

- Presents with multiple daily events
- Normal development
- Normal examination
- No family history

Julie – 8 years old

Julie – 8 years old

EPILEPSY and SNOMED

SNOMED International

1. Epilepsy
2. International League Against Epilepsy (ILAE)
3. ILAE classification history, process and progress
4. ILAE Position Papers: new concepts and terminology
5. Rationale for evolution of terms and concepts: absence seizures
6. SNOMED ILAE collaboration

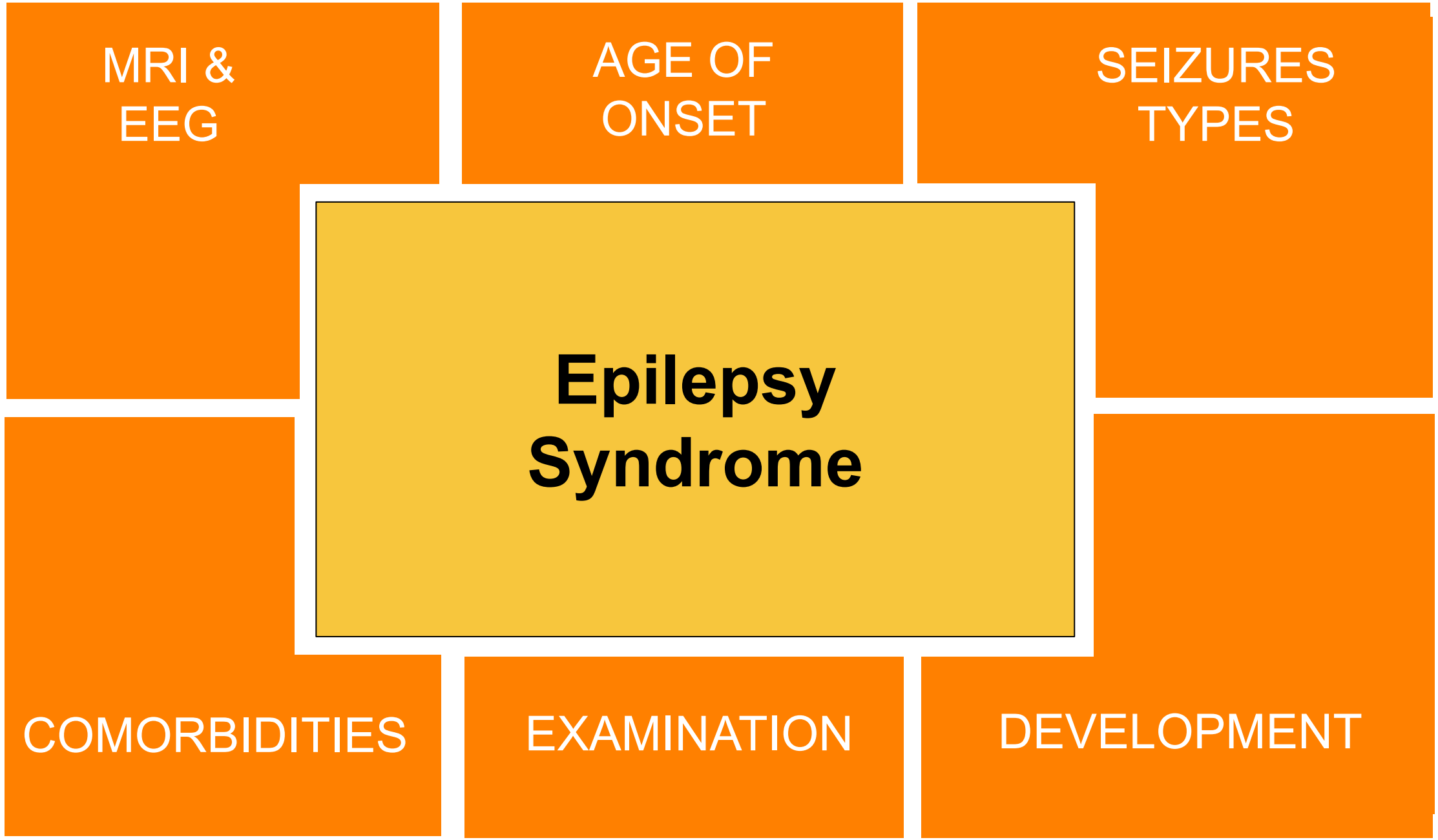
Epileptic Seizure

A transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.


Epilepsy
Two or more unprovoked epileptic seizures

Epilepsy Syndrome:

A characteristic cluster of clinical and EEG features, often supported by specific etiological findings.


MRI &
EEG
AGE OF
ONSET
SEIZURES
TYPES
Epilepsy
Syndrome
COMORBIDITIES
EXAMINATION
DEVELOPMENT

Why are epilepsy syndromes important?

Guide investigations

Provide prognosis: seizure and comorbitidies

Provide aetiological information

Guide management

Enable effective research

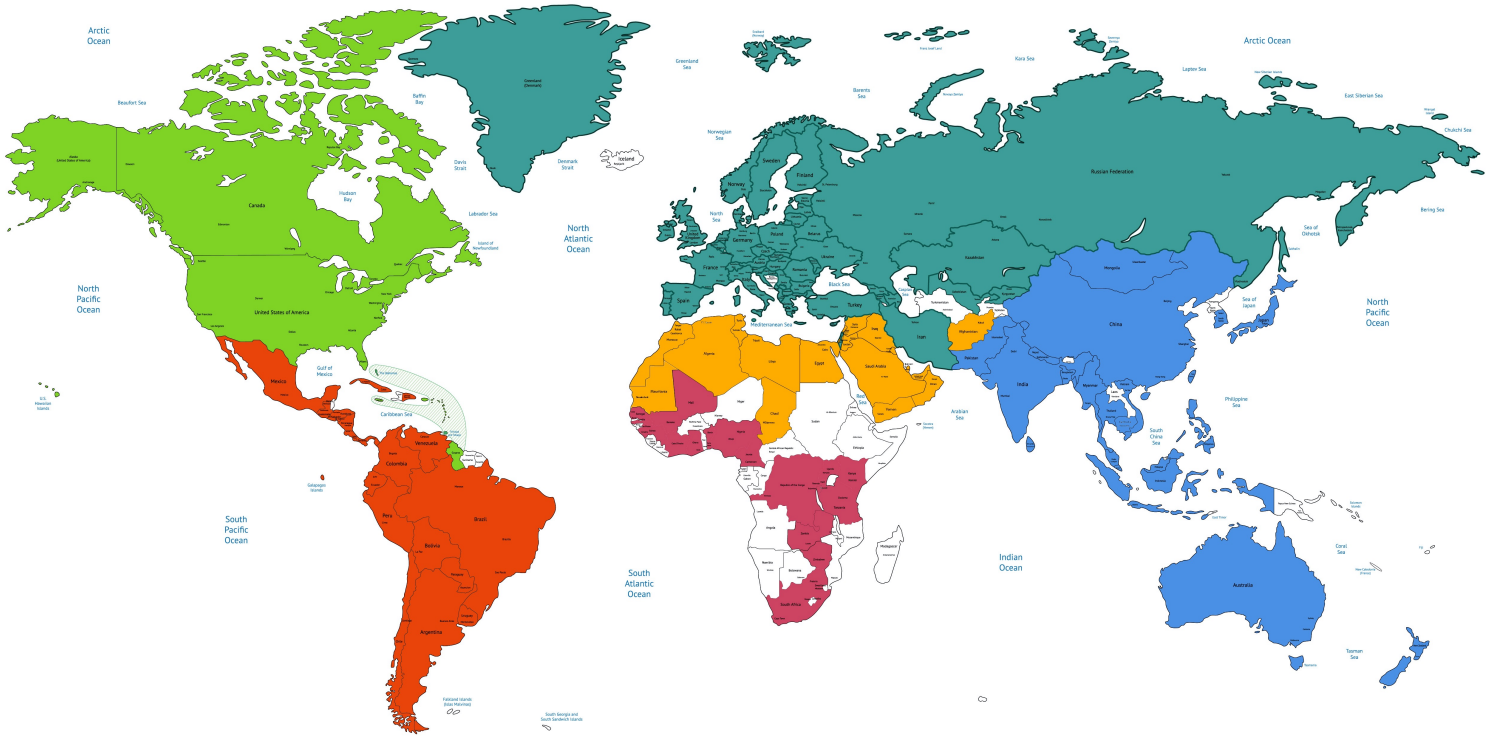
“Epilepsy is the most common serious brain disorder worldwide with no age, racial, social class, national, nor geographic boundaries.”

World Health Organization

The World's preeminent association of health care professionals and scientists working toward a world where no person's life is limited by epilepsy

Established in 1909

International League Against Epilepsy



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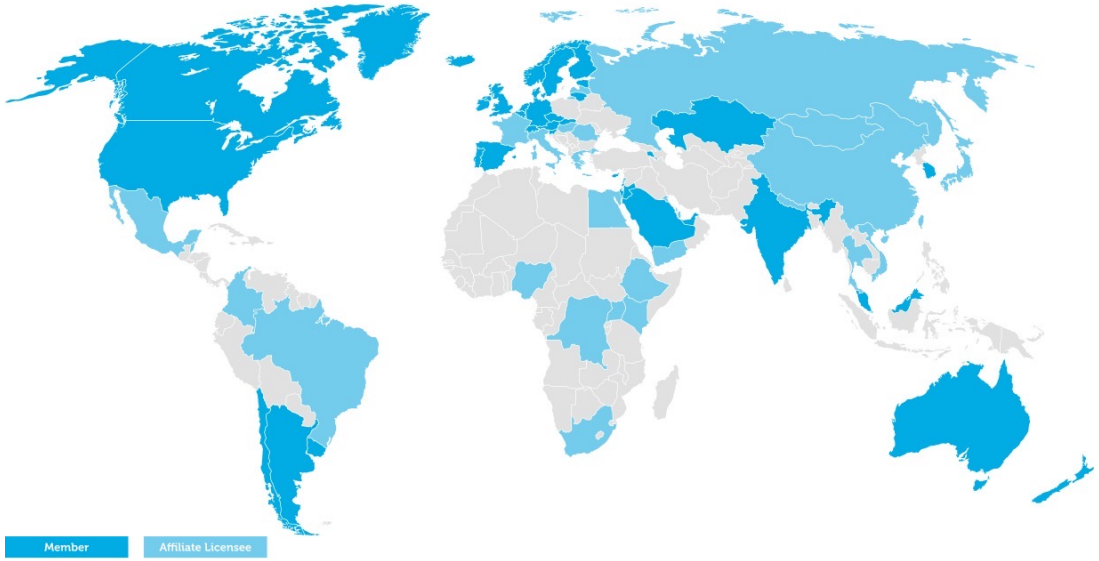
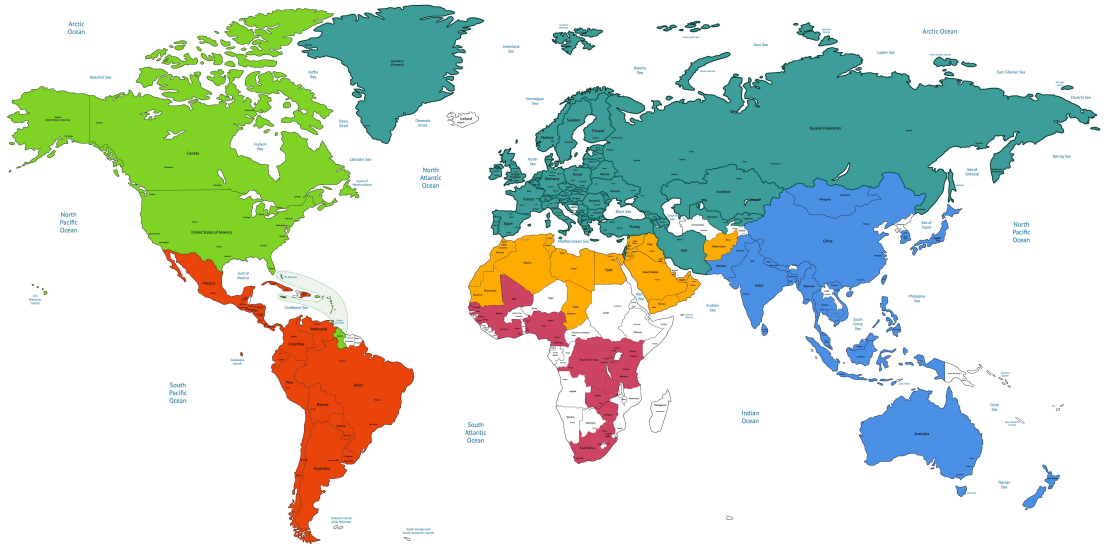
International League Against Epilepsy





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Goal is to enhance the content related to seizures and epilepsy in SNOMED CT to ensure clinicians have the latest evidence-based content available to them for point of care recording.

Terminology and Classification of the Epilepsies

Purpose: for clinical diagnosis

Transparent language: words that mean what they say

Previous ILAE Classification and Terminology Publications

1970: Clinical and EEG Classification of Epileptic Seizures

1981: Revised Clinical and EEG Classification of the Epileptic Seizures

1985: Classification of Epilepsies and Epileptic Syndromes

1989: Revised Classification of Epilepsies and Epileptic Syndromes

2010: Revised terminology and concepts for organization of seizures and epilepsies

ILAE International League Against Epilepsy

ILAE Position Paper Process

International League Against Epilepsy

Epilepsia
Official Journal of the International League Against Epilepsy

ILAE Position Papers: Commission for Classification and Terminology

2014: A practical and clinical definition of Epilepsy

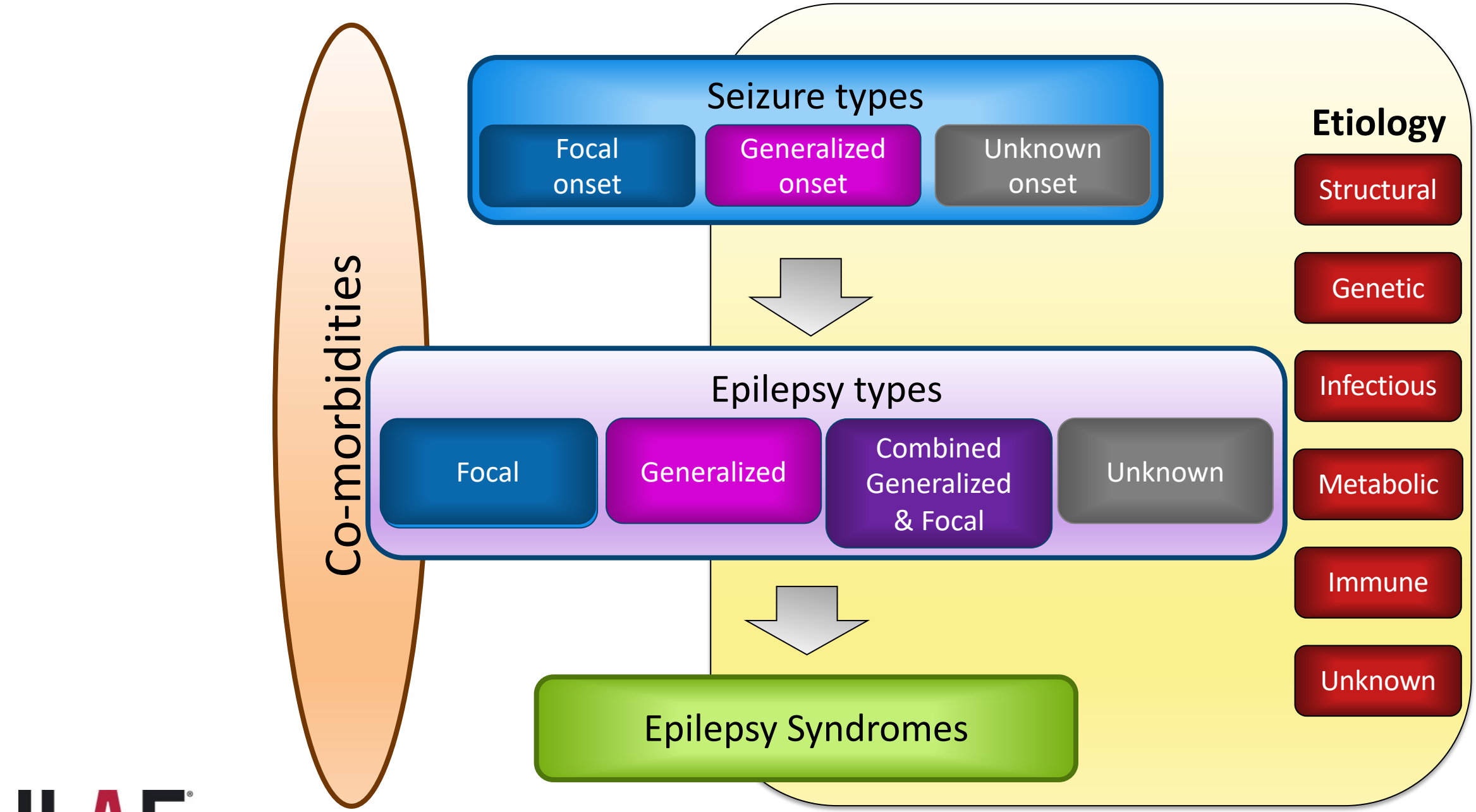
2017: **Operational classification of seizure types**

2017: **Classification of the epilepsies**

2020: **Modifications for seizures in the neonate**

2022: **Classification & definitions of epilepsy syndromes**

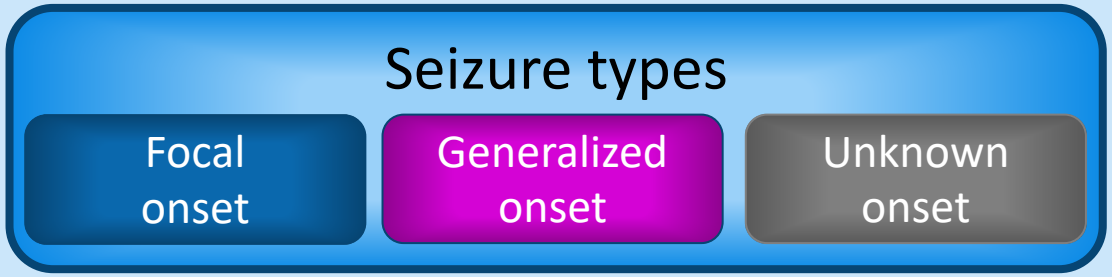
International League Against Epilepsy



ILAE International League Against Epilepsy

FINDING

SNOMED International



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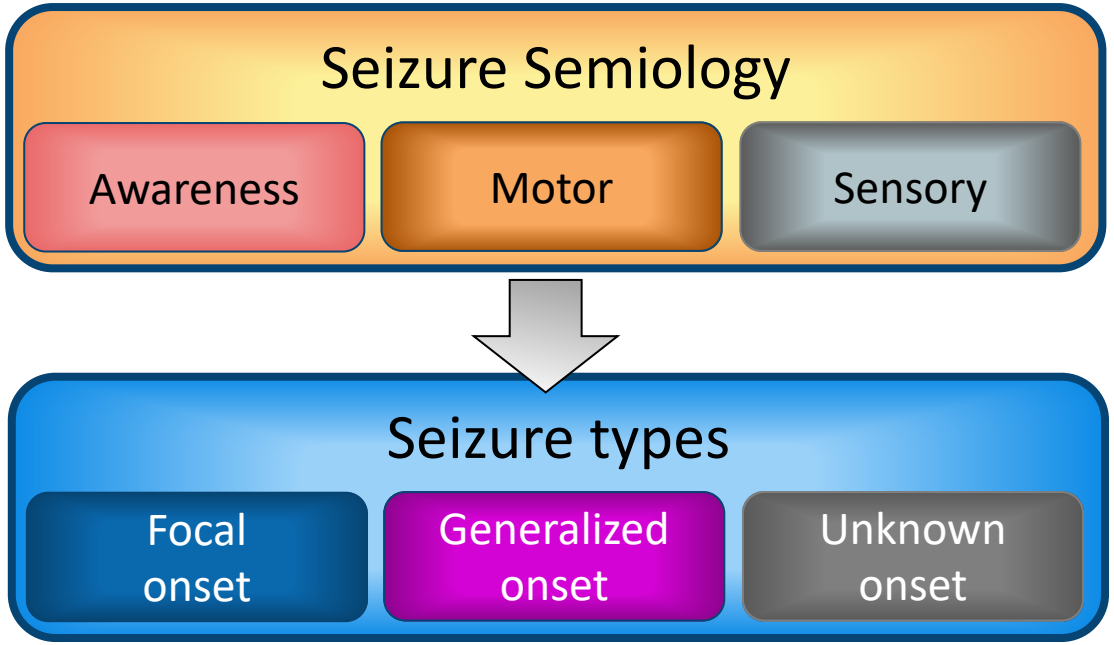

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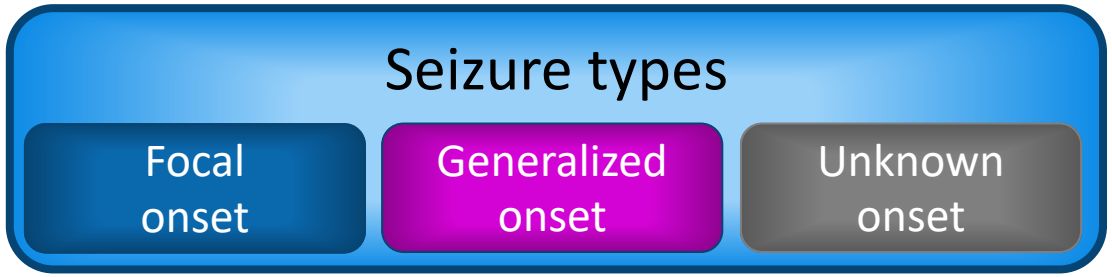
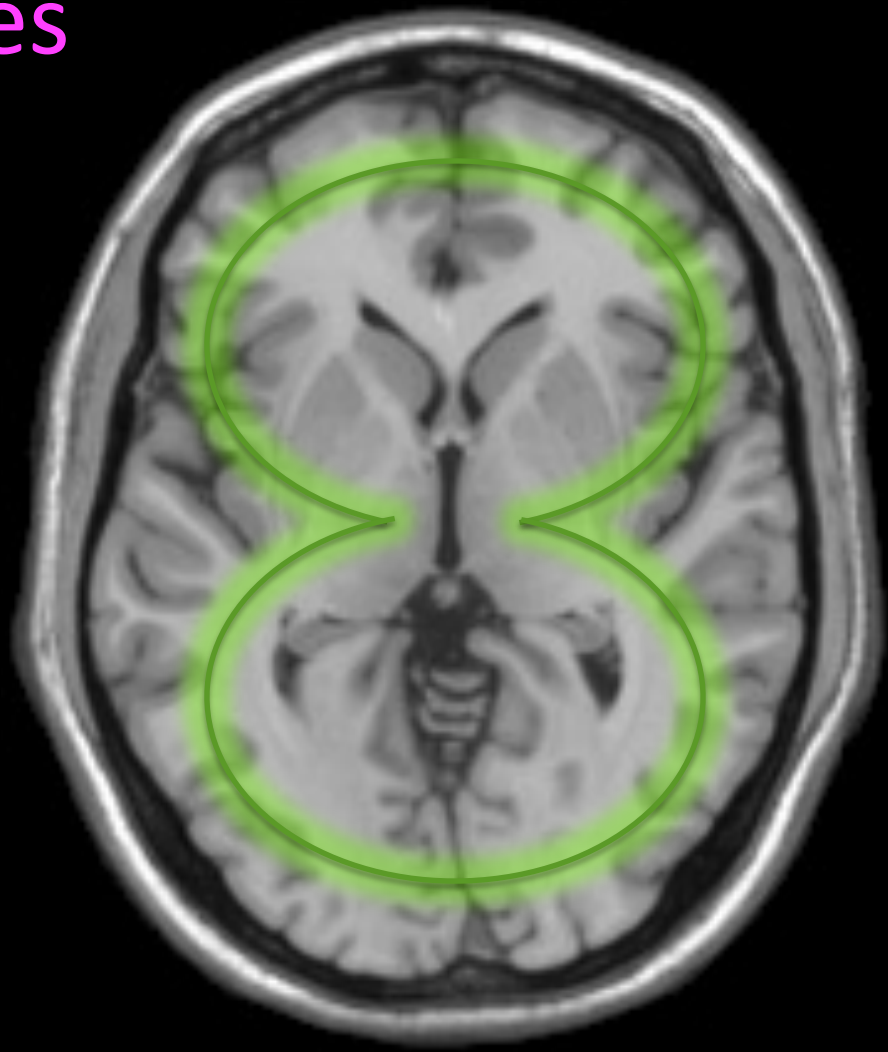


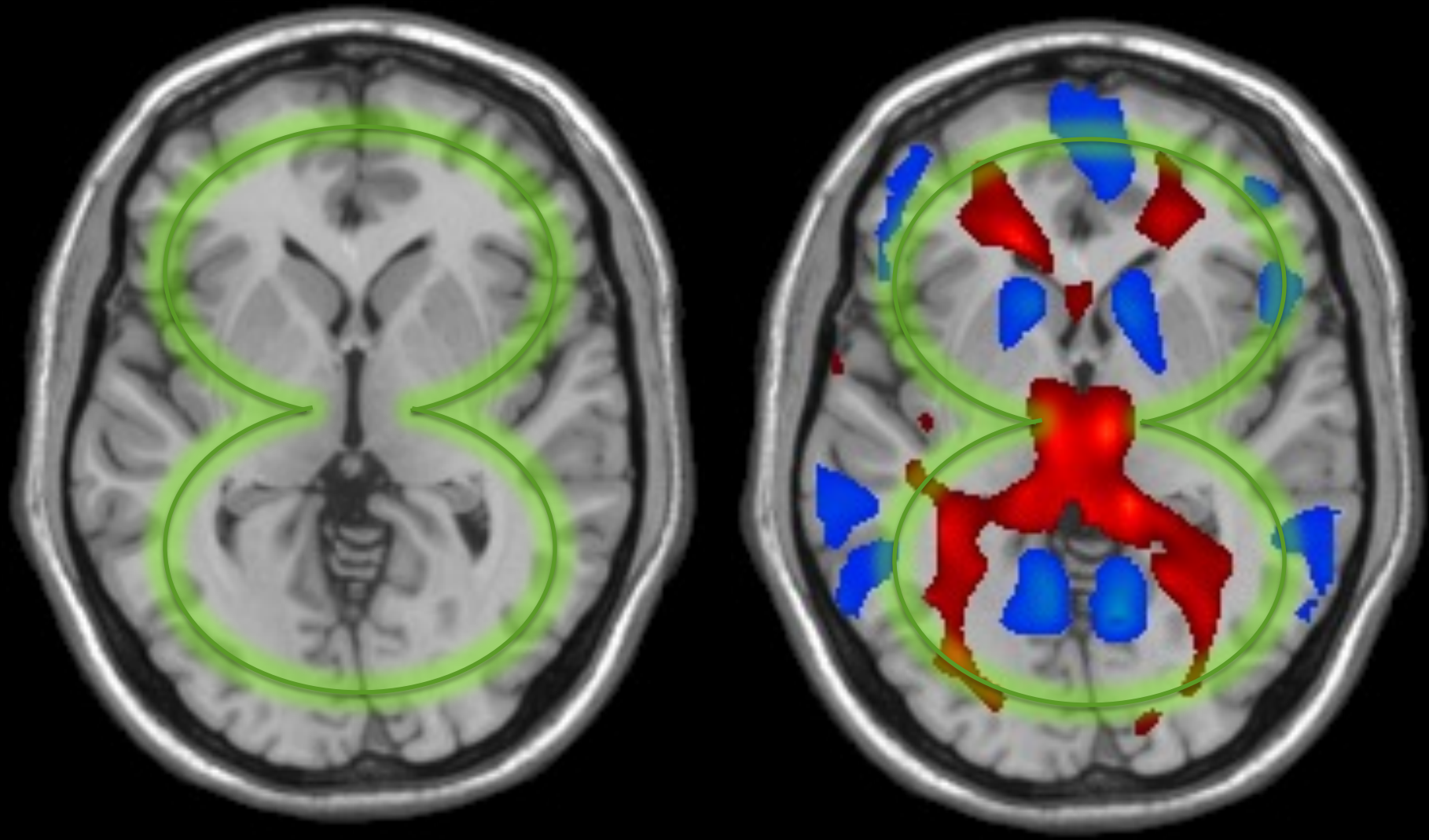
International League Against Epilepsy

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

Generalized seizures

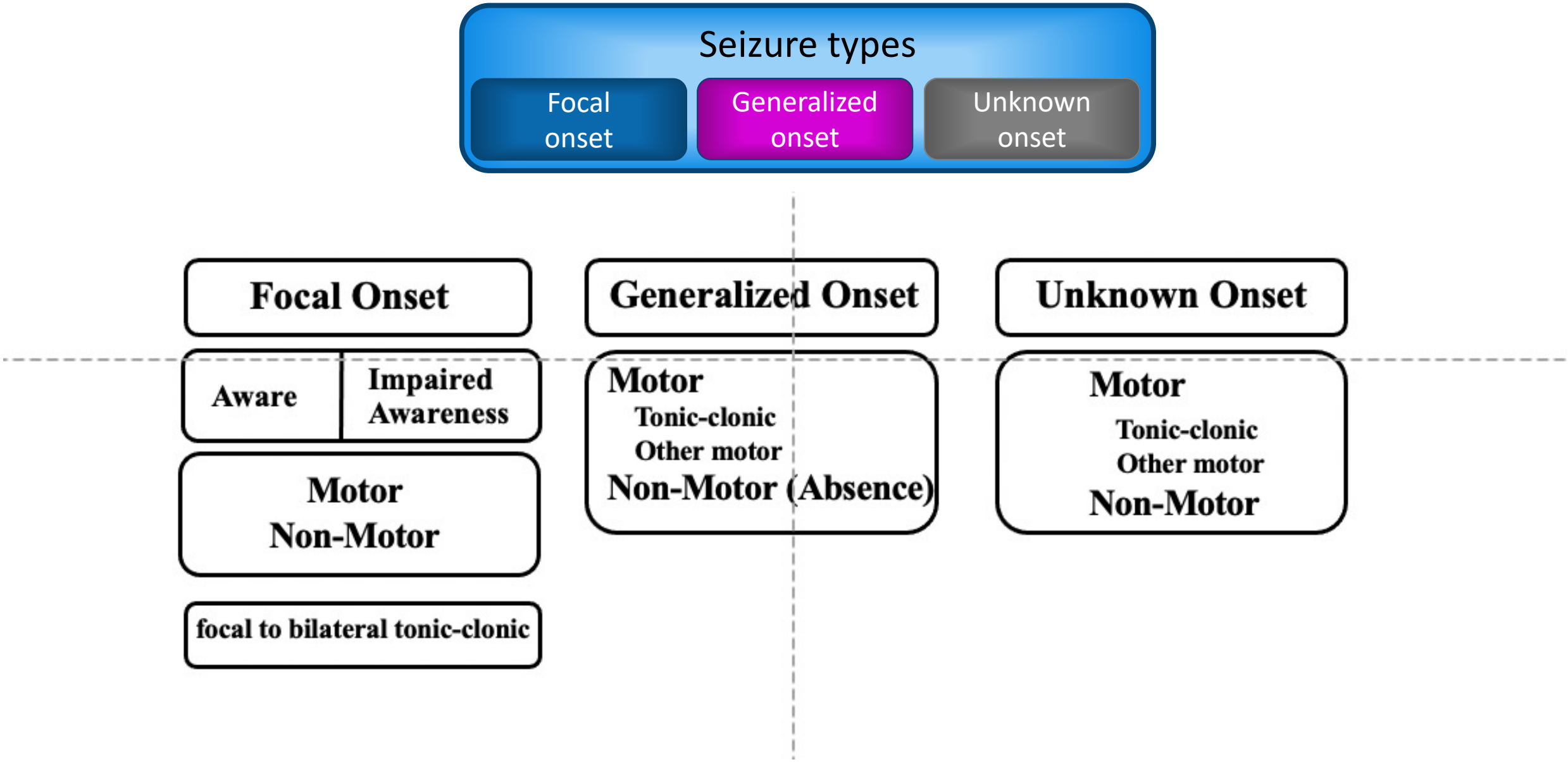
- Originate at some point within and rapidly engage bilaterally distributed networks





Focal seizures

- Originate within networks limited to one hemisphere



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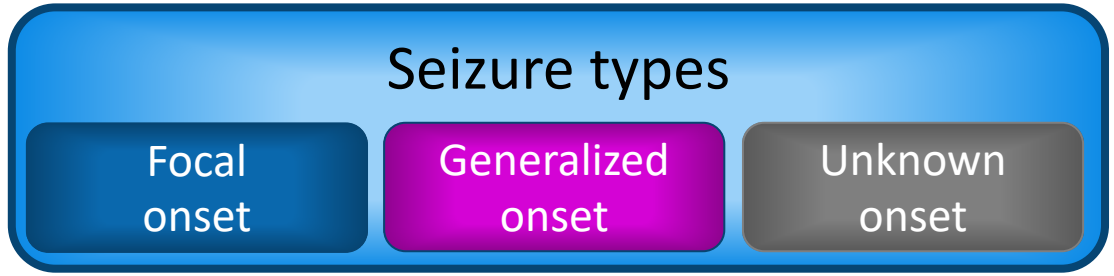


Focal Onset

Aware	Impaired Awareness

Motor Onset
- automatisms
- atonic
- clonic
- epileptic spasms
- 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

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

Unknown Onset

Motor
- tonic-clonic
- epileptic spasms

Non-Motor
- behavior arrest

ILAE International League Against Epilepsy

Terms no longer in use

- Complex partial
- Simple partial
- Partial
- Psychic
- Dyscognitive
- Secondarily generalized tonic-clonic

International League Against Epilepsy

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

FINDING

SNOMED International

Seizure types

- Focal onset
- Generalized onset
- Unknown onset

DISORDER

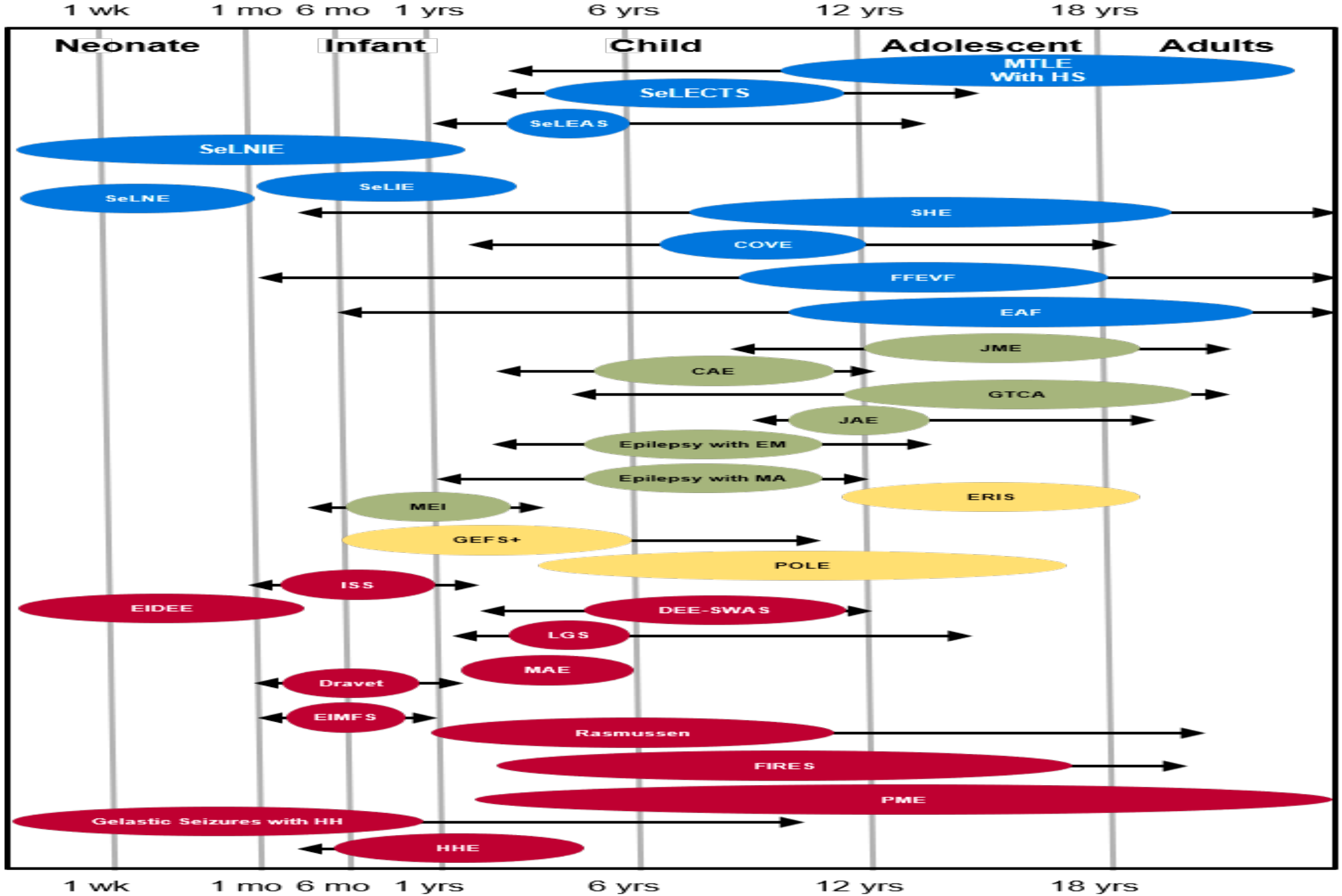
SNOMED International

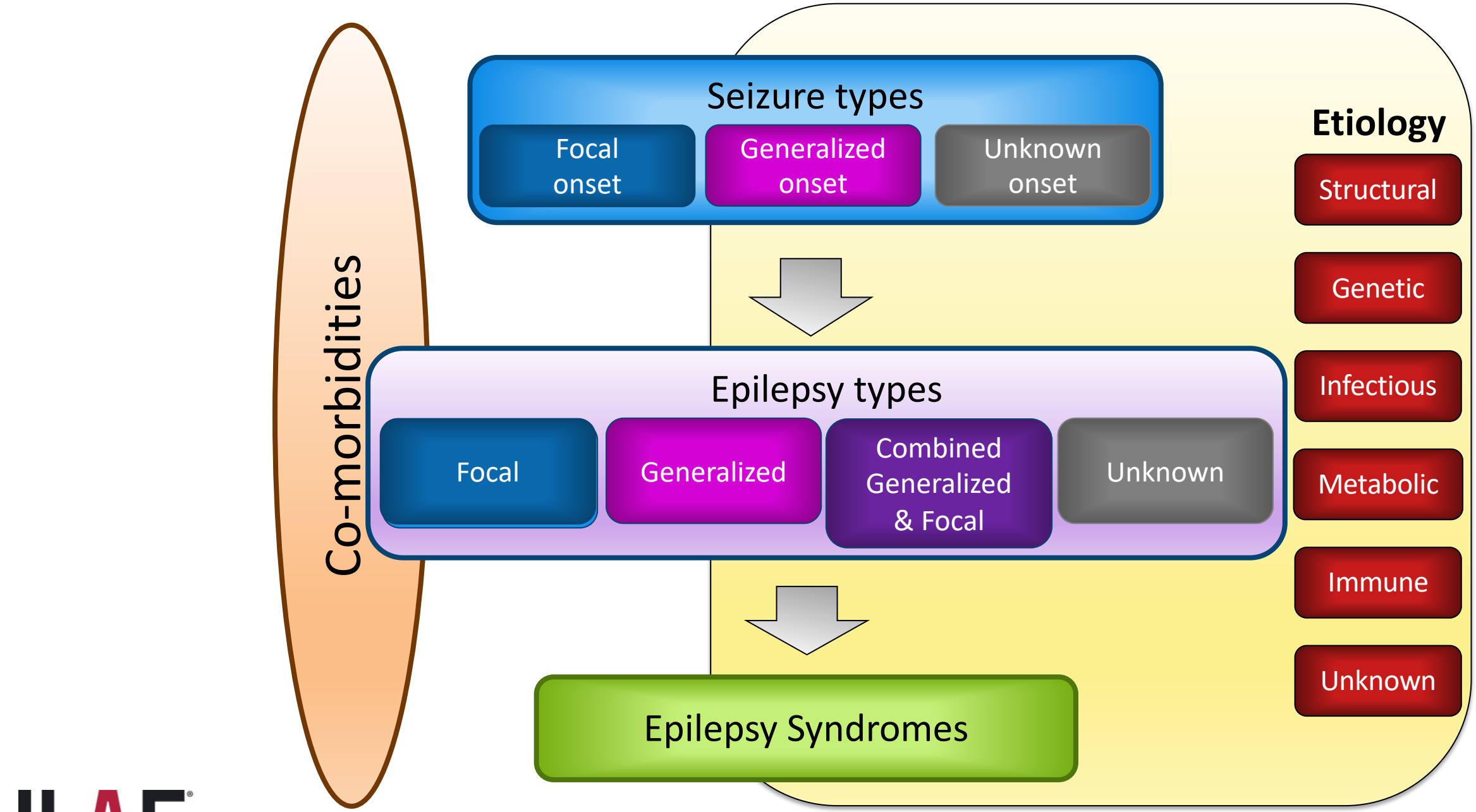
Epilepsy types

- Focal
- Generalized
- Combined Generalized & Focal
- Unknown

Epilepsy Syndromes

ILAE International League Against Epilepsy


1 wk
1 mo
6 mo
1 yrs
6 yrs
12 yrs
18 yrs
Neonate
Infant
Child
Adolescent
Adults
MTLE With HS
SeLECTS
SeLEAS
SeLNIE
SeLIE
SeLNE
SHE
COVE
FFEVF
EAF
JME
CAE
GTCA
JAE
Epilepsy with EM
Epilepsy with MA
ERIS
MEI
GEFS+
POLE
ISS
EIDEE
DEE-SWAS
LGS
MAE
Dravet
EIMFS
Rasmussen
FIRES
PME
Gelastic Seizures with HH
HHE



ILAE International League Against Epilepsy

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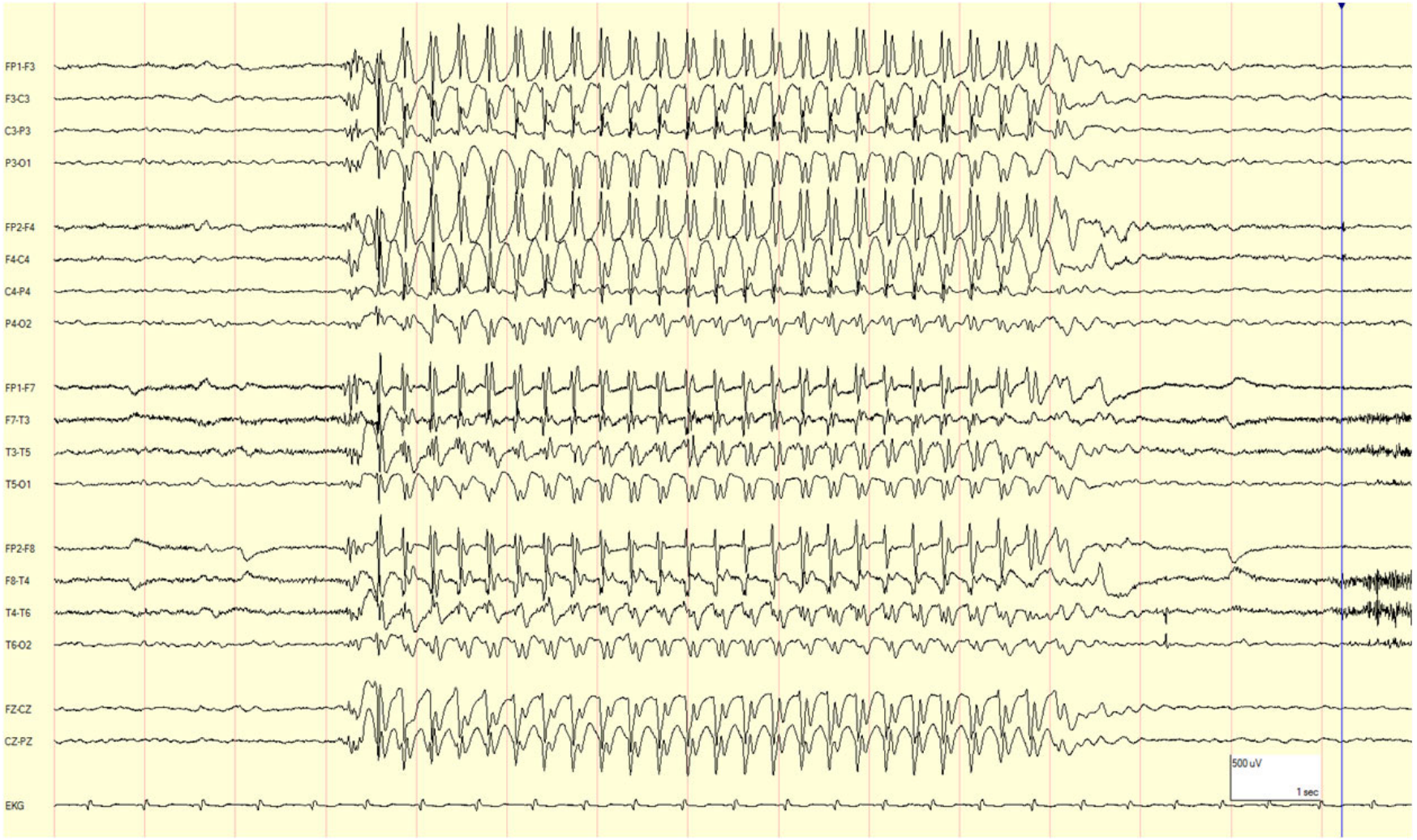

Julie – 8 years old

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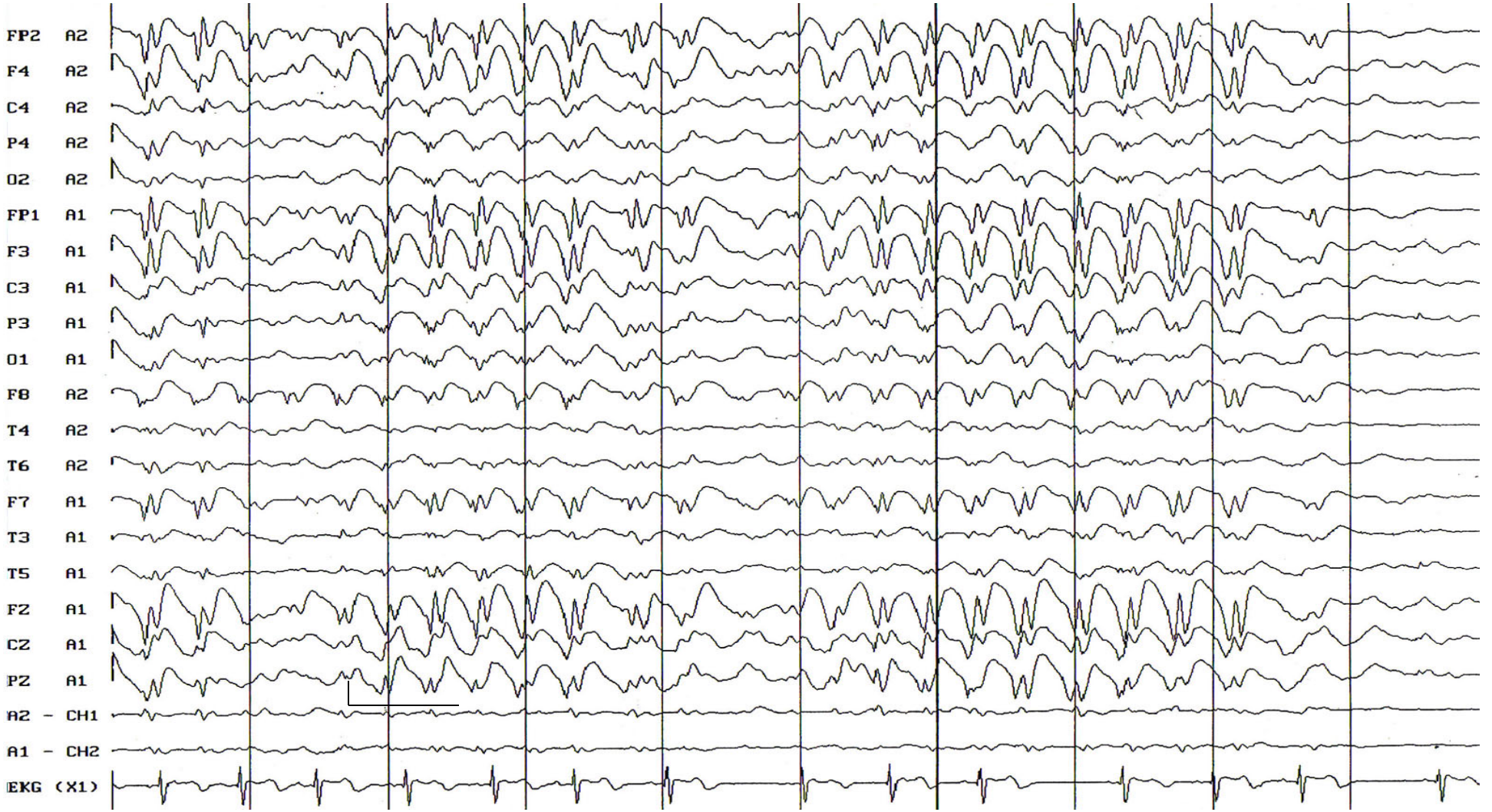


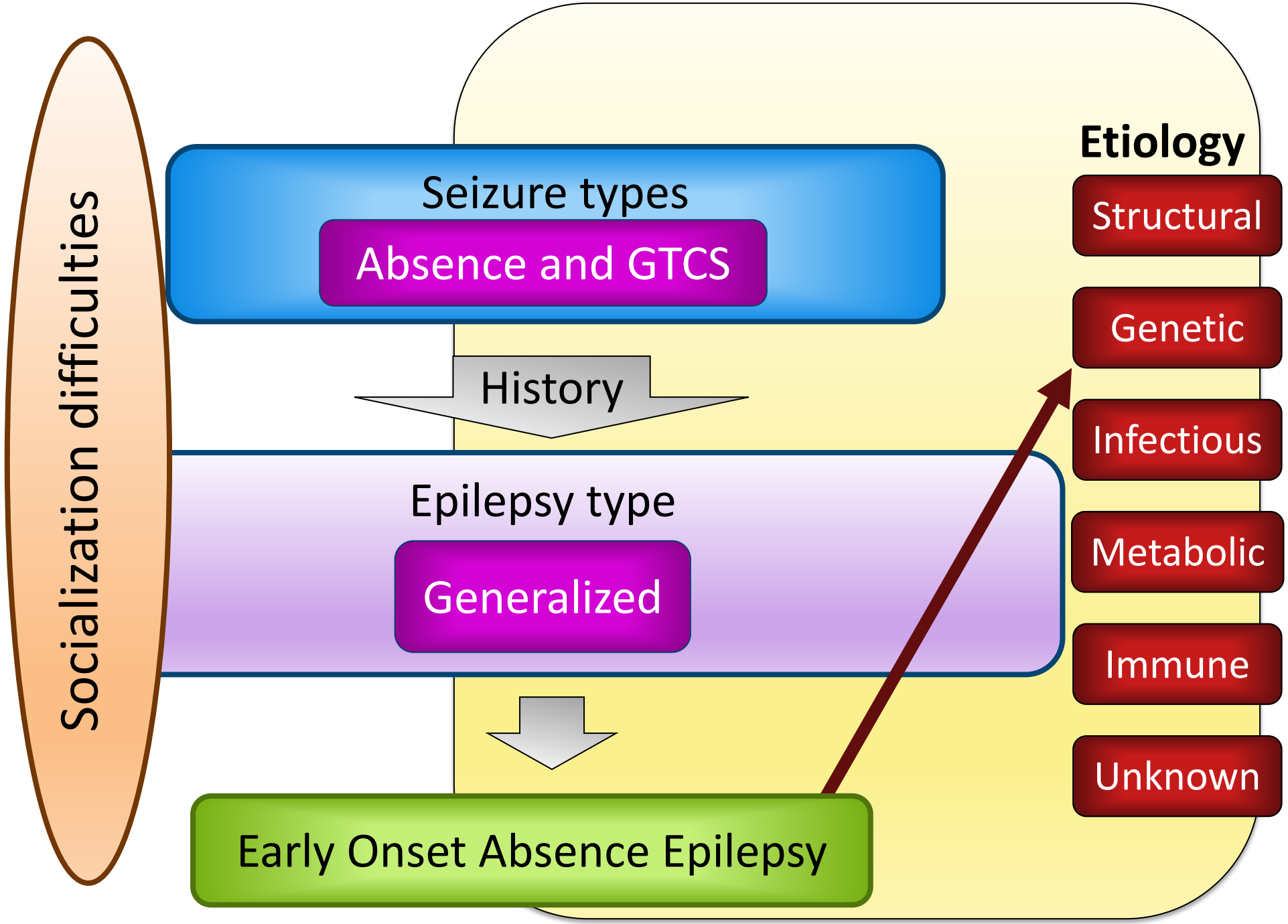
Julie – 8 years old

Seizure type

Typical Absence and GTCS

Julie – 12 years old

- Seizures
  - 2 years: infrequent typical brief absence seizures
  - 8 years: Absence status and GTCS
- Normal development and examination
- No family history
- EEG – generalised spike wave and polyspike wave
- MRI - normal

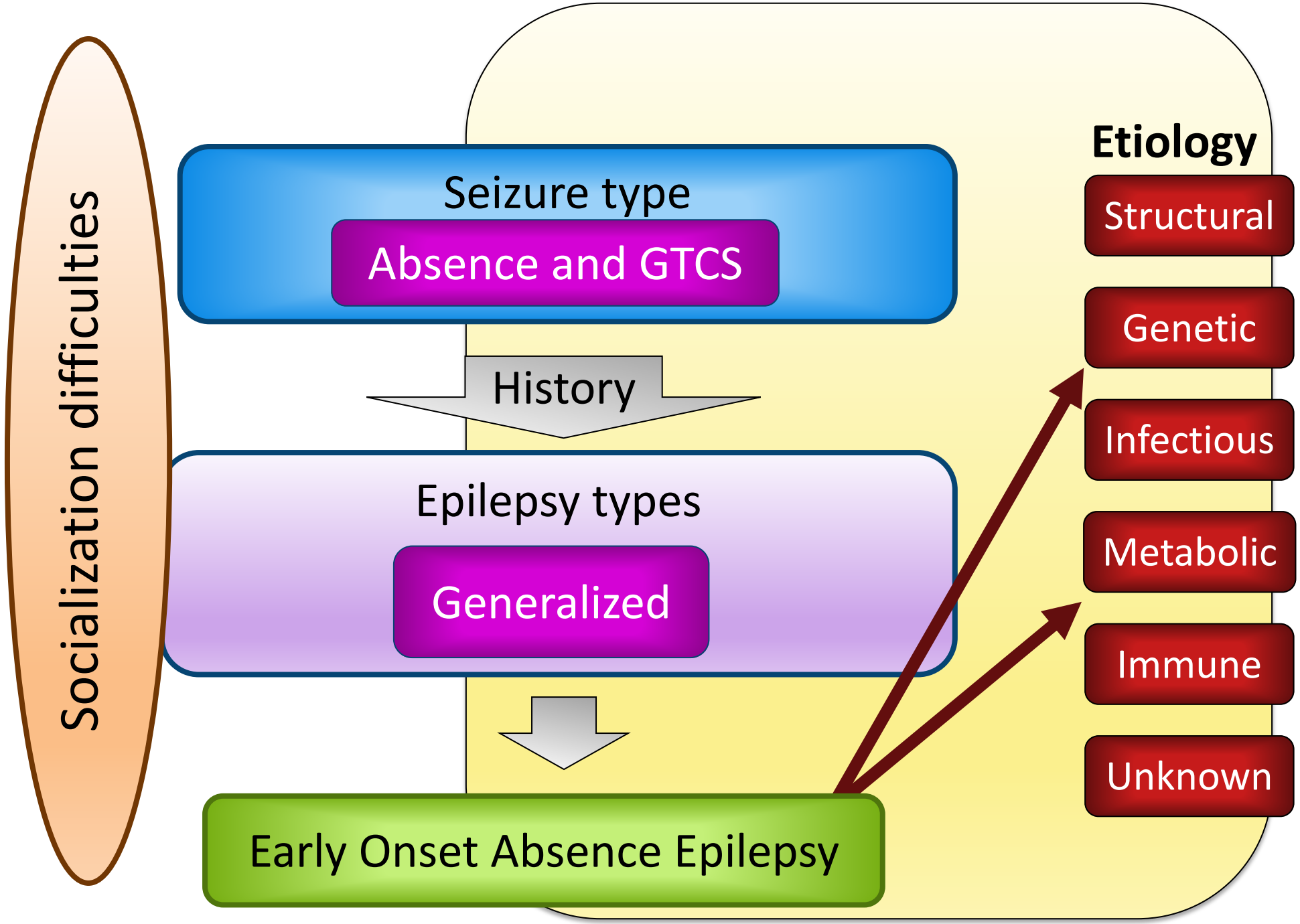

Socialization difficulties
Seizure types
Absence and GTCS
History
Epilepsy type
Generalized
Early Onset Absence Epilepsy
Etiology
Structural
Genetic
Infectious
Metabolic
Immune
Unknown

Julie – 8 years old

SLC2A1 pathogenic variant

GLUT1 deficiency

Precision therapy: Ketogenic diet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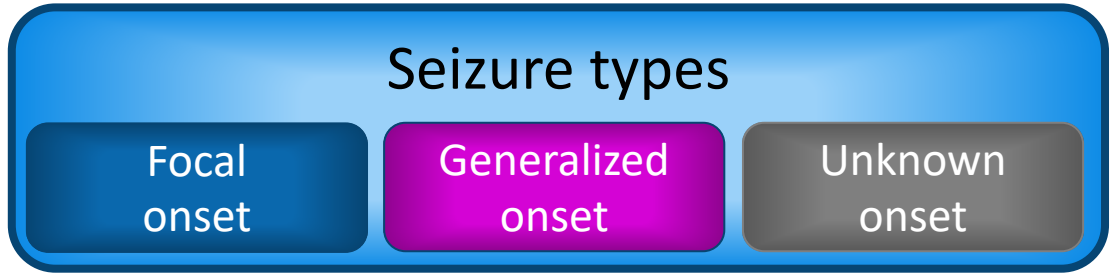
Rationale for evolution of terminology and concepts

ABSENCE SEIZURES



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Unknown Onset

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Non-Motor
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ABSENCE SEIZURES

Observation of events

1705: Poupart published first description
1815: Esquirol introduced “Petit Mal”
1824: Calmeil used “absence”

EEG

1933: Berger described 3 hz spike and wave

ABSENCE SEIZURES

Prior to 1963

Petit Mal

Petit Mal Variant

ABSENCE SEIZURES

1964

Petit Mal (typical absence)

Petit Mal Variant (atypical absence)

ABSENCE SEIZURES

Observation of events	**1705:** Poupart published first description **1815:** Esquirol introduced “Petit Mal” **1824:** Calmeil used “absence”
EEG	**1933:** Berger described 3 hz spike and wave
Video-telemetry monitoring	**1969, 1975** Penry; **1974** Browne; **1981** Stefan: described semiology in detail

ABSENCE SEIZURES

1970 Classification

SIMPLE ABSENCES

COMPLEX ABSENCES

+ mild clonic components

+ increased tone

+ decreased tone

+ automatisms

+ autonomic phenomena

ABSENCE SEIZURES

1970 Classification

- SIMPLE ABSENCES
- COMPLEX ABSENCES
  - + mild clonic components
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  - + automatisms
  - + autonomic phenomena

1981 Classification

- ABSENCE SEIZURES
  - + mild clonic movements
  - + tonic components
  - + atonic components
  - + automatisms
  - + autonomic components
- ATYPICAL ABSENCE SEIZURES

ABSENCE SEIZURES

Observation of events	**1705:** Poupart published first description **1815:** Esquirol introduced "Petit Mal" **1824:** Calmeil used "absence"
EEG	**1933:** Berger described 3 hz spike and wave
Video-telemetry monitoring	**1969, 1975** Penry; **1974** Browne; **1981** Stefan: described semiology in detail
Video-EEG	**1987** Holmes; **1989** Panayiotopoulos; **1994** Hirsch; **2001** Capovilla; **2006** Sadleir: described relevance of semiological features

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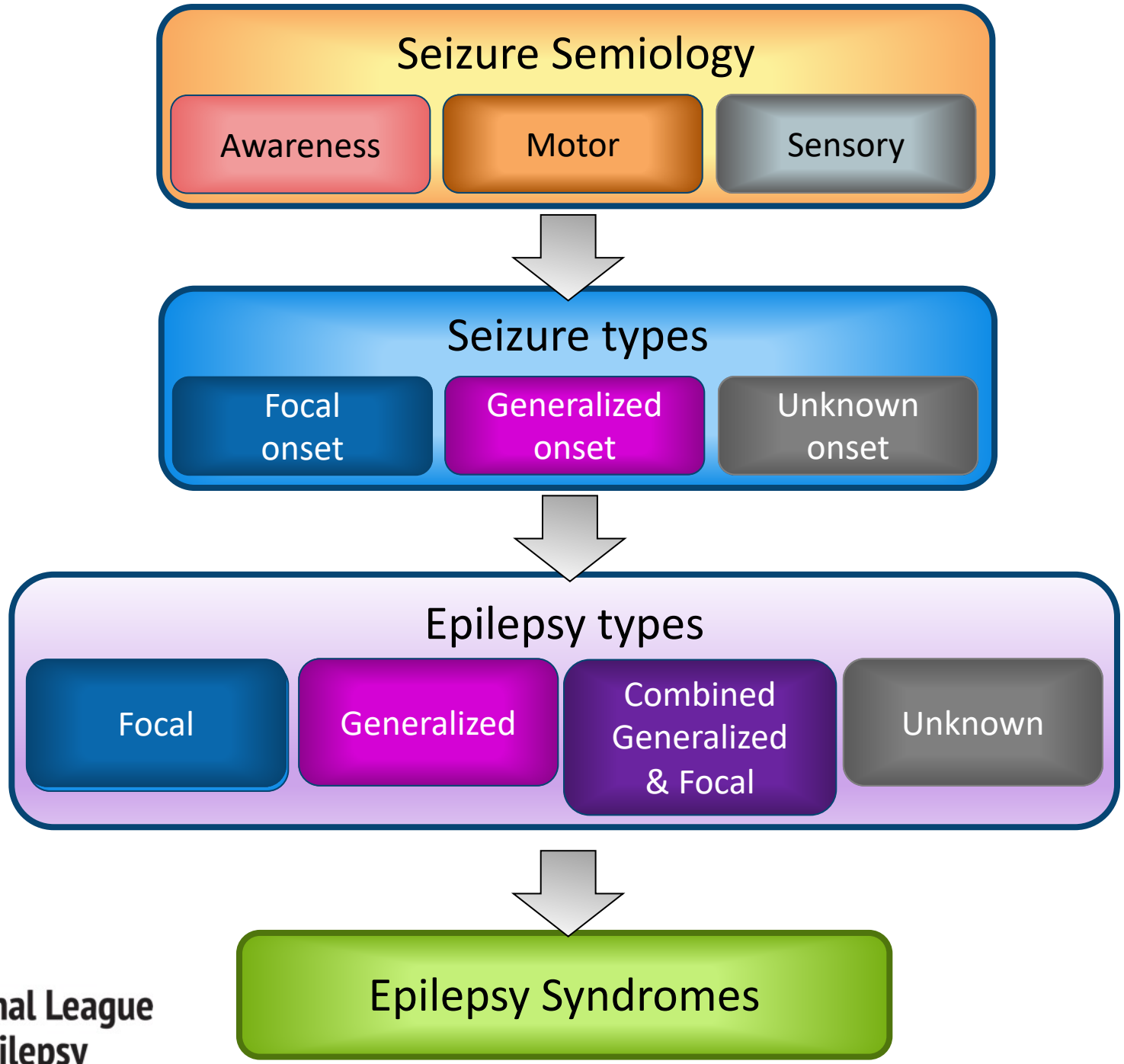
2017 Classification

- TYPICAL ABSENCE SEIZURES
- MYOCLONIC ABSENCE
- ABSENCE WITH EYELID MYOCLONIA
- ATYPICAL ABSENCE SEIZURES



International League Against Epilepsy

Automatisms

"A more or less coordinated repetitive motor activity usually occurring when cognition is impaired and for which the subject is amnesic afterwards"

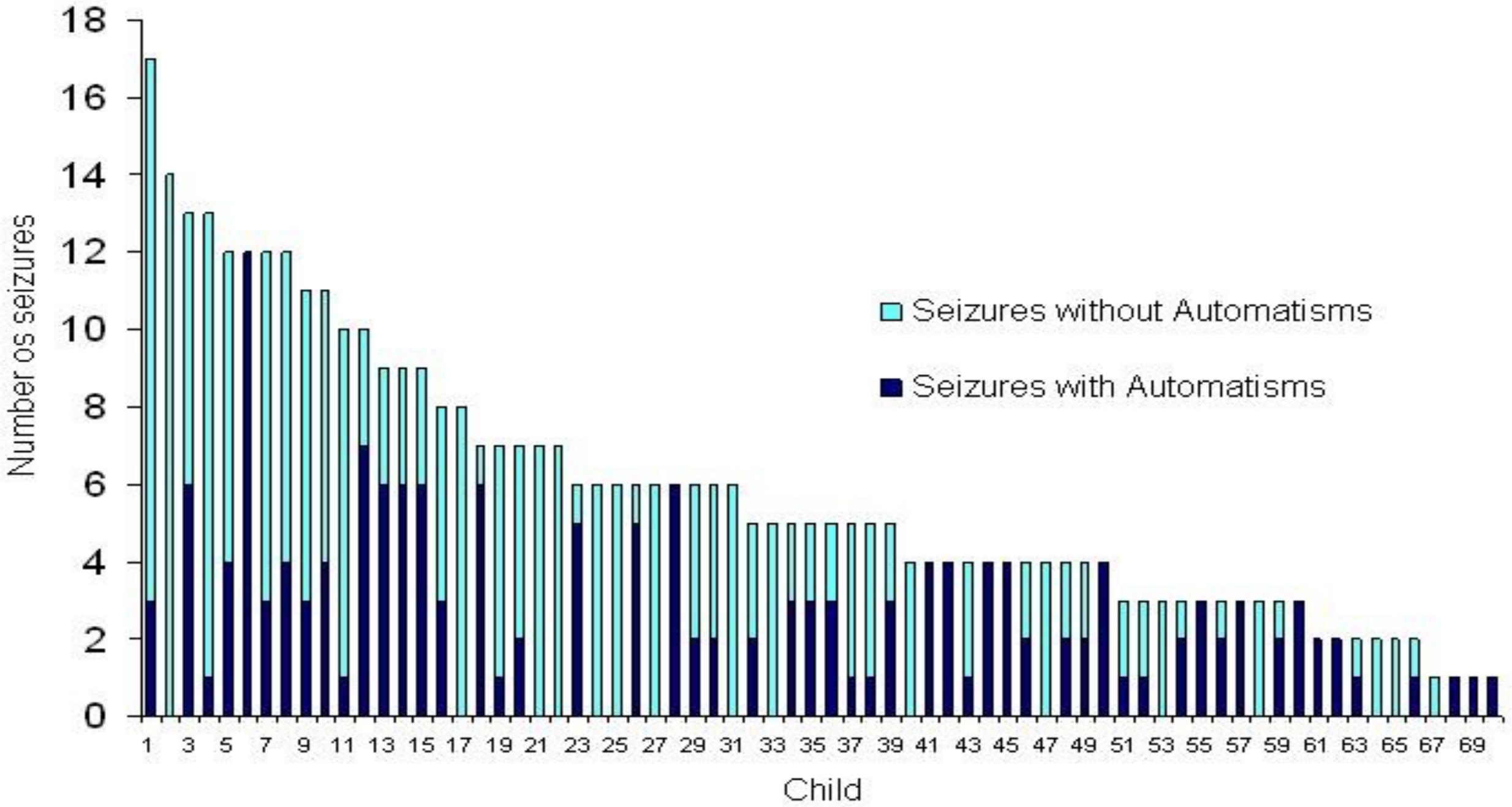
Common

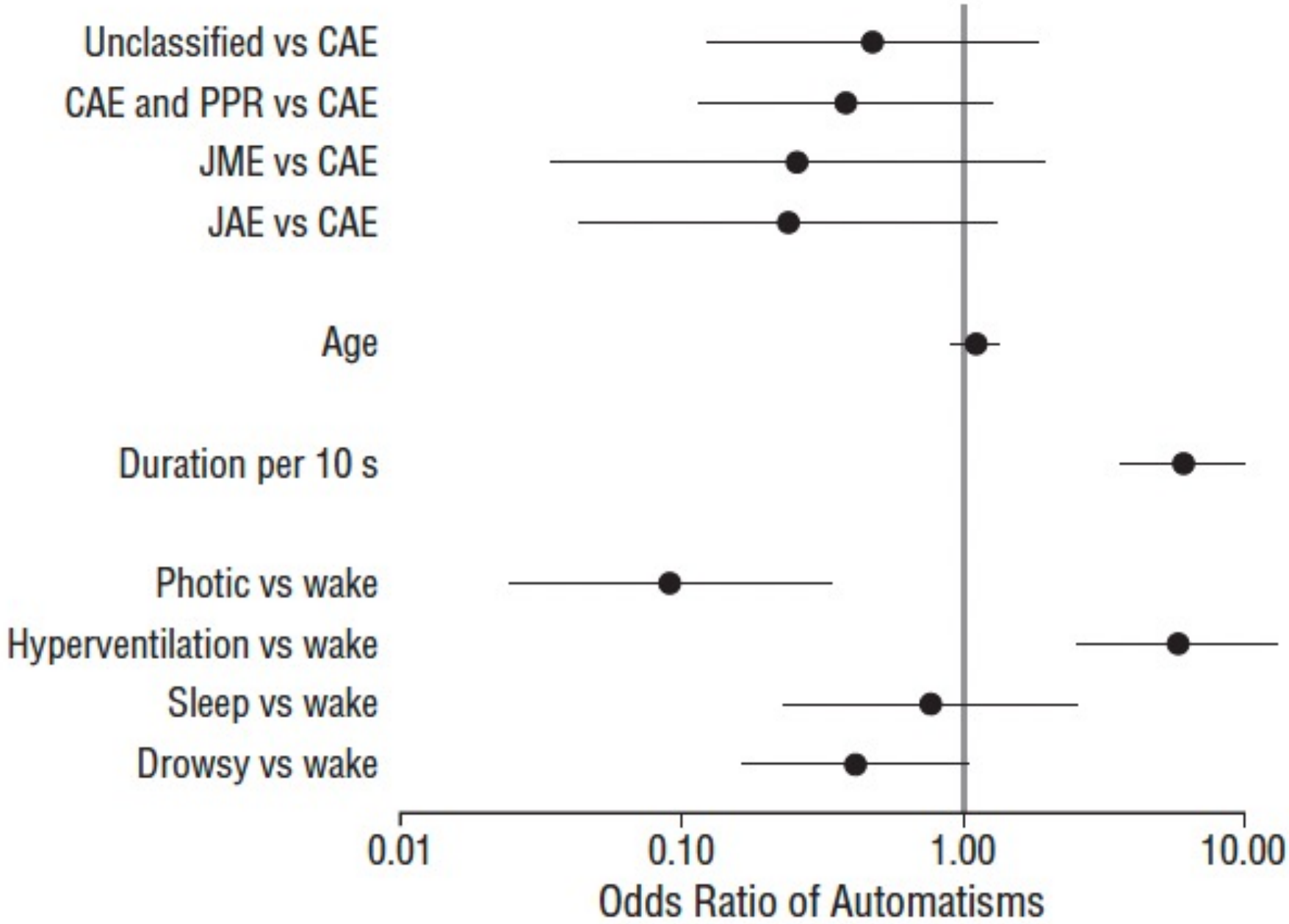
- Up to 63% of absence seizures
- Up to 100% of children.
- Up to 67% of children have automatism in every seizure

Absence video-EEG study

- New onset absence seizures
- 70 children (1 to 16 years)
- Syndromes: Childhood Absence Epilepsy, Juvenile Absence Epilepsy, Juvenile Myoclonic Epilepsy
- 509 absence seizures
- States: Awake, drowsy and sleep
- Provocation: Photic stimulation, Hyperventilation

Sadleir et al.
Archives of Neurology 2009


Number os seizures
18
16
14
12
10
8
6
4
2
0
Seizures without Automatisms
Seizures with Automatisms
1 3 5 7 9 11 13 15 17 19 21 23 25 27 29 31 33 35 37 39 41 43 45 47 49 51 53 55 57 59 61 63 65 67 69
Child


Unclassified vs CAE
CAE and PPR vs CAE
JME vs CAE
JAE vs CAE
Age
Duration per 10 s
Photic vs wake
Hyperventilation vs wake
Sleep vs wake
Drowsy vs wake
0.01
0.10
1.00
10.00
Odds Ratio of Automatisms

ABSENCE SEIZURES

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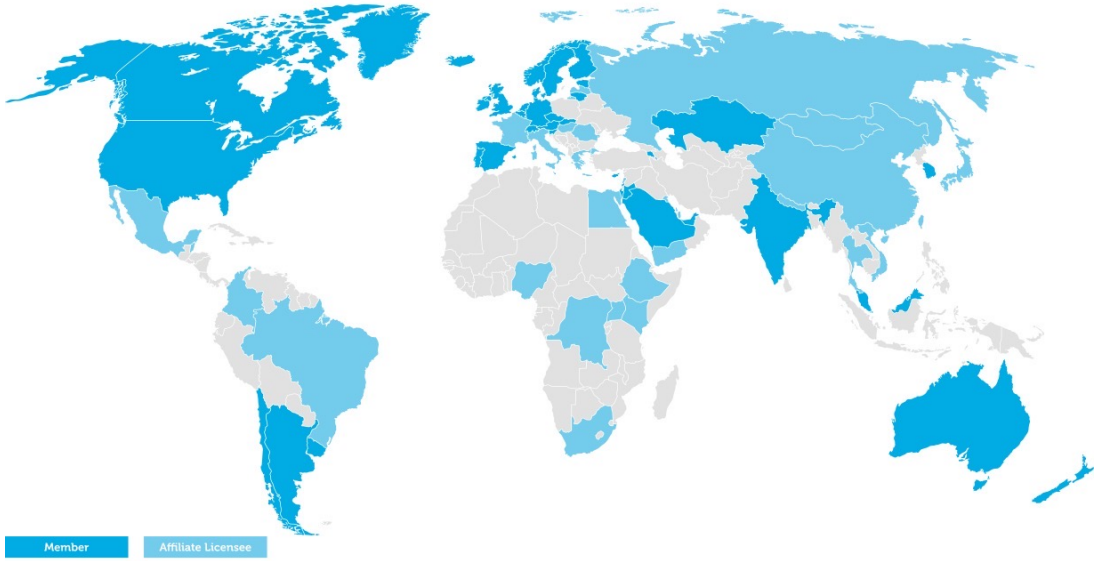

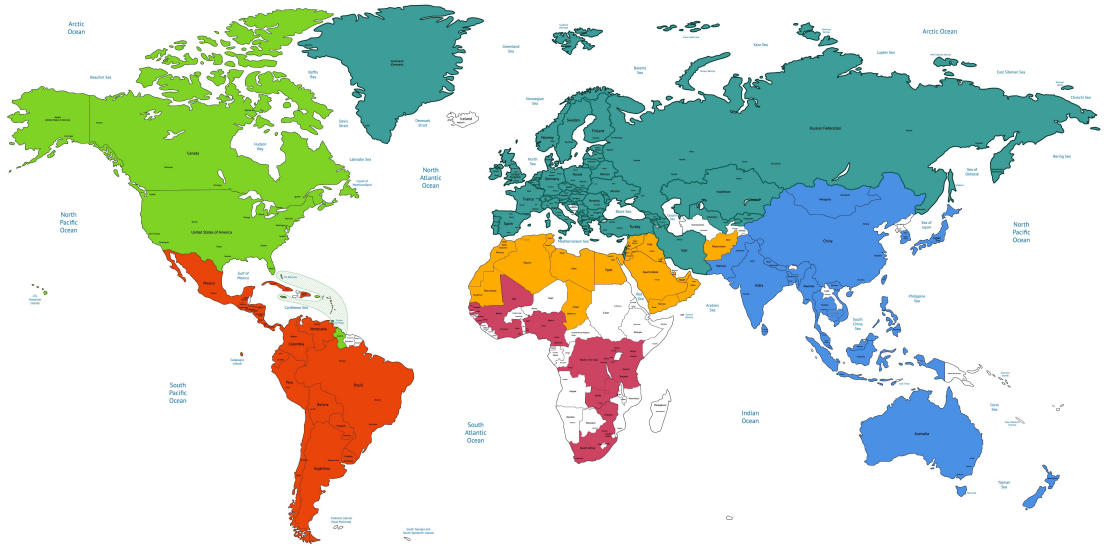

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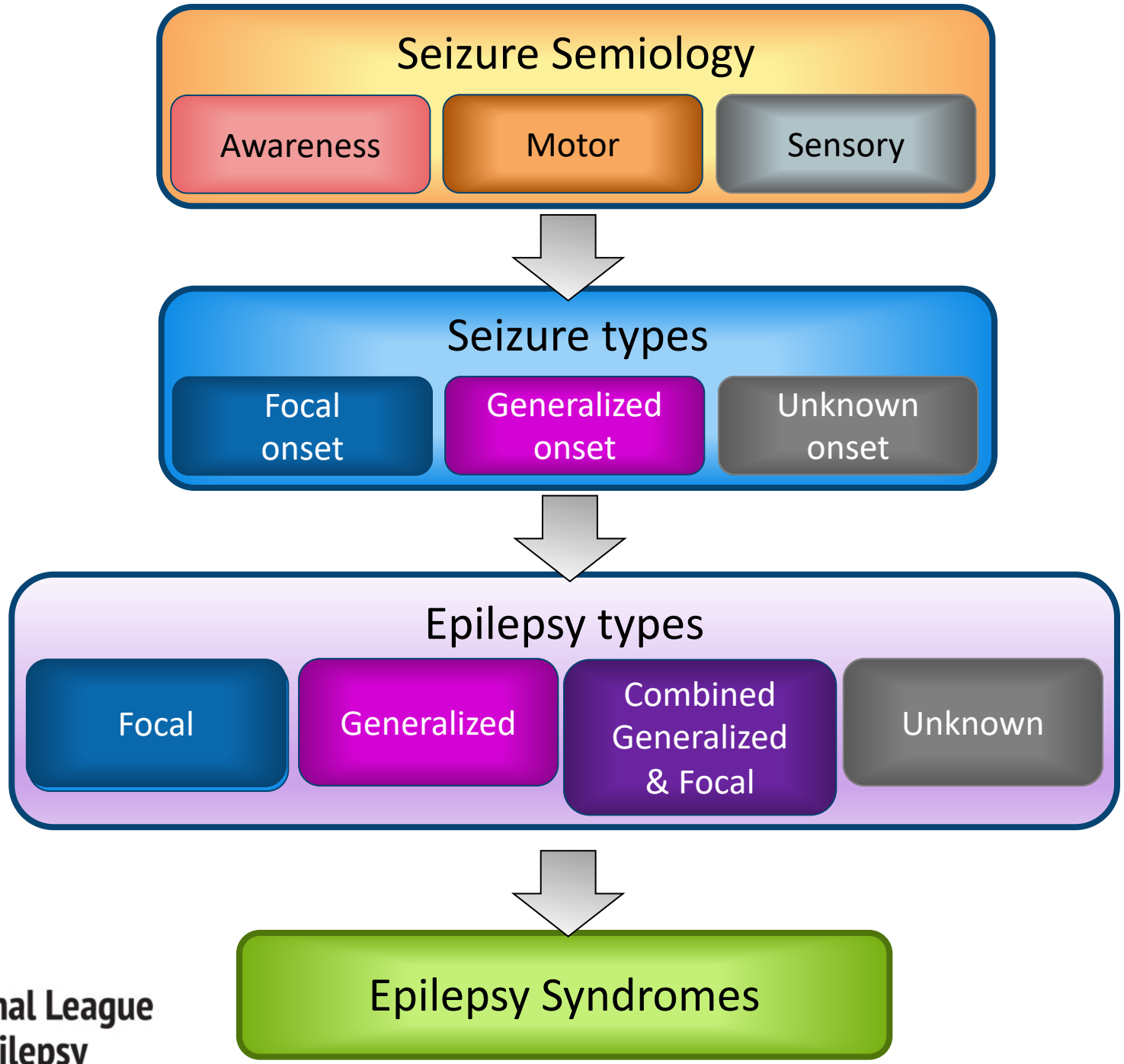
Plan:

- Add new seizure type and epilepsy syndromes terminology
- Inactivate seizure and epilepsy terminology which is outdated and not helpful
- Organise seizure and epilepsy terminology relationships in a conceptual way which makes sense with our present understanding of seizures and epilepsy

EPILEPSY and SNOMED

SNOMED International

- Epilepsy is a group of disorders (syndromes) defined predominantly (but not only) by seizure types
- 2017, 2020, 2022: ILAE introduced new concepts and terminology
- Changes reflect advances in understanding and use understandable words
- SNOMED ILAE collaboration will add new terminology, inactivate unhelpful terminology, realign terminology in SNOMED



International League Against Epilepsy