

CLASSIFYING SEIZURES & EPILEPSIES

ILAE, Semiological Classifications

Prakash Kotagal, M.D.

SEIZURE

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

SEIZURE TYPES

Focal seizure: originating within networks limited to one hemisphere

Generalized seizure: originating at some point within, and rapidly engaging bilaterally distributed networks

Unknown onset

NEW DEFINITION OF EPILEPSY

Disease of the brain defined by any of the following:

1. At least two unprovoked (or reflex) seizures occurring more than 24 hours apart
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome.

Epilepsy may be considered resolved when:

- individuals with age-dependent epilepsy syndrome are now past the applicable age
- seizure-free for 10 years, with no seizure medicines for last 5 years.

- **Epilepsy Syndrome:** refers to a cluster of features incorporating seizure types, EEG and imaging features that tend to occur together. Often has age-dependent features, seizure triggers, diurnal variation and sometimes prognosis
 - West Syndrome
 - Lennox-Gastaut Syndrome
 - Dravet Syndrome
- **Epileptic Encephalopathy:** epileptic activity itself may contribute to severe cognitive and behavioral impairments above and beyond what might be expected from the underlying pathology alone
 - Continuous Spikes in Slow Wave Sleep (CSWS/ESES)

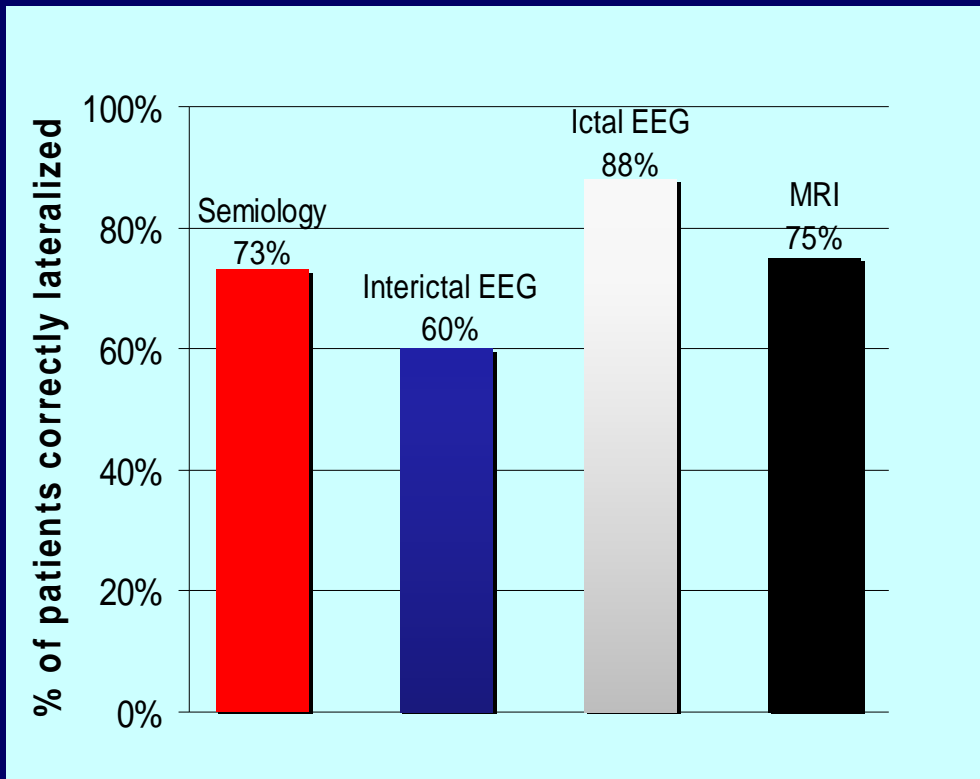
SEMIOLOGY

The branch of linguistics concerned with signs and symptoms

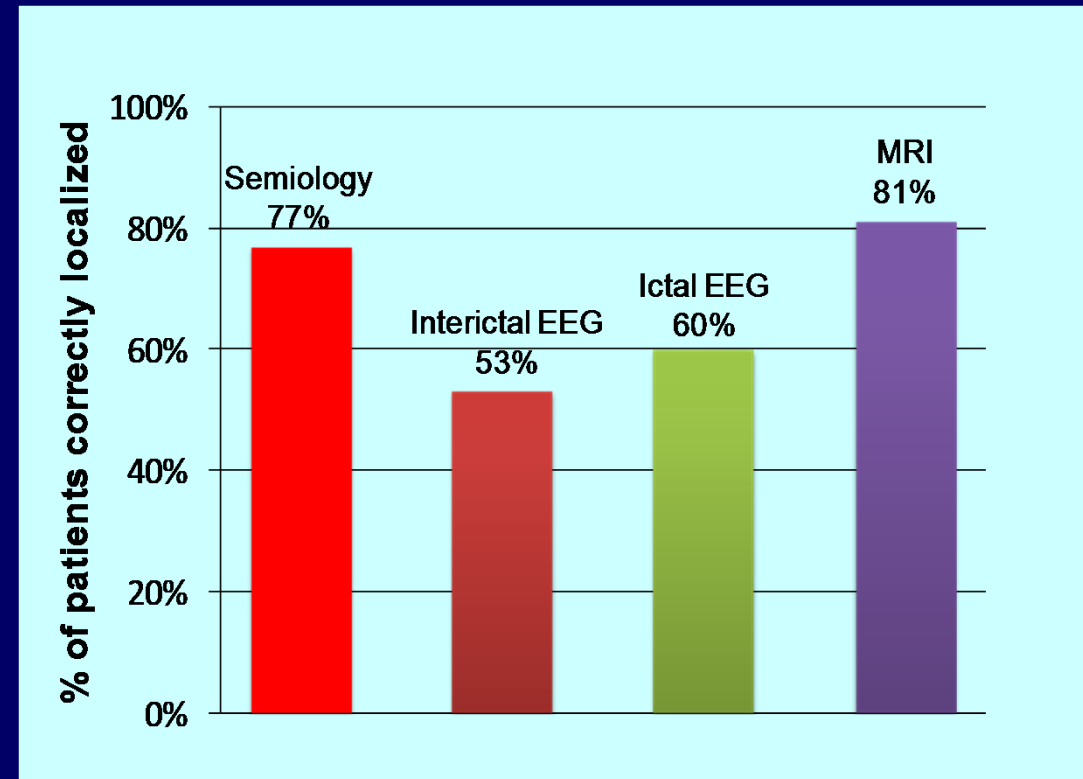
Blume WT, Lüders HO, Mizrahi E et al. Glossary of Descriptive Terminology for Ictal Semiology: Report of the ILAE Task Force on Classification and Terminology. Epilepsia 2001;42:1212-8

Seizure Semiology may be as good as EEG, MRI

LATERALIZING VALUE



LOCALIZING VALUE



Elwan S, Alexopolous A, Silveria D, Kotagal P. Seizure 2018;61:203-8

INITIAL OFFICE VISIT

- Good **seizure history** is critical
 - Events surrounding seizure onset, precipitating factors, relationship to sleep / wakefulness
 - Auras
 - Lateralizing signs
 - Postictal phenomena
 - Any other seizure types?
 - Response to treatment
- Try to determine **etiology** (past history, family history)
- General physical **examination** (dysmorphic features, neurocutaneous stigmata, other systems)
- Neurological examination
- **EEG** is essential to the evaluation

LATERALIZING SIGNS IN SEIZURES

- Auras: somatosensory, visual
- Versive movements of the eyes and head
- Ipsilateral head turning
- Unilateral clonic movements
- Ictal vomiting
- Ictal speech and postictal dysphasia
- Unilateral automatisms
- Unilateral tonic posturing
- Unilateral dystonic posturing
- Lateralized ictal paresis
- Todd's palsy

LATERALIZING SIGNS IN SEIZURES

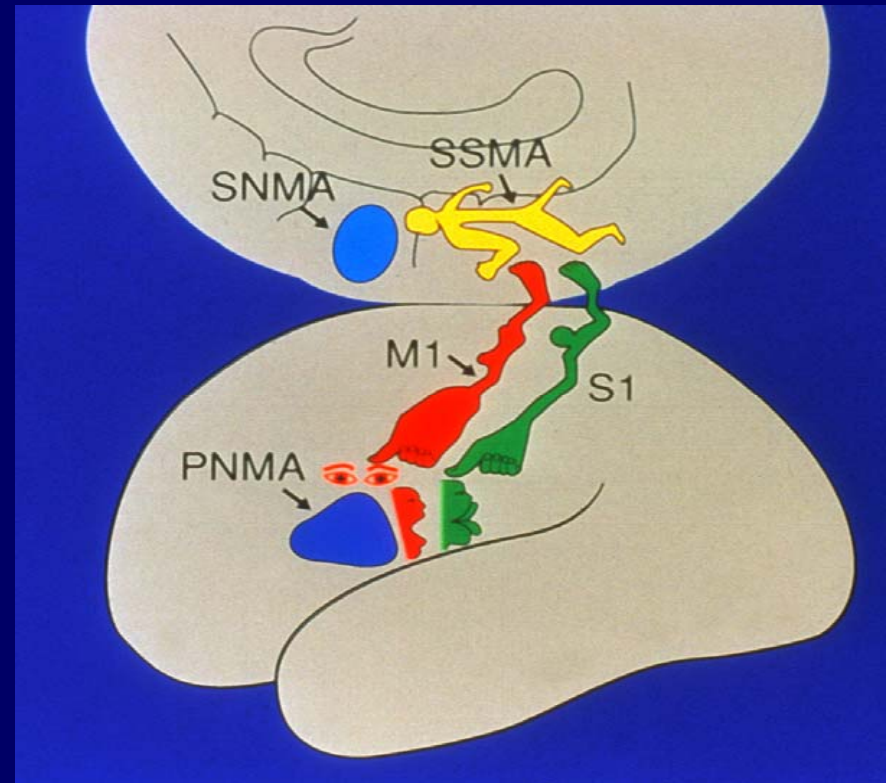
- Postictal nose wiping
- Automatism with preserved responsiveness
- Unilateral eye blinking
- Asymmetrical tonic limb posturing during GTC seizures (Figure 4 sign)
- Postictal headache
- Ictal crying
- Facial asymmetry
- Tongue biting
- Ictal spitting

AURAS OF LOCALIZING VALUE

- Somatosensory and elementary visual auras have localizing and lateralizing
- Epigastric sensation, psychical and olfactory auras are typical of temporal lobe epilepsy
- Cephalic and body sensations seen in frontal, temporal or parietal onset
- Auditory, vestibular and formed visual auras: posterior temporal, parietal
- Forced thinking - frontal lobe
- Gustatory, autonomic auras - perisylvian area

FOCAL CLONIC MOVEMENTS

- Indicate involvement of contralateral sensori-motor cortex
- Also seen when seizures spread from other regions
- Good reliability
- Epilepsia Partialis Continua: focal motor status involving a small portion of the sensorimotor cortex

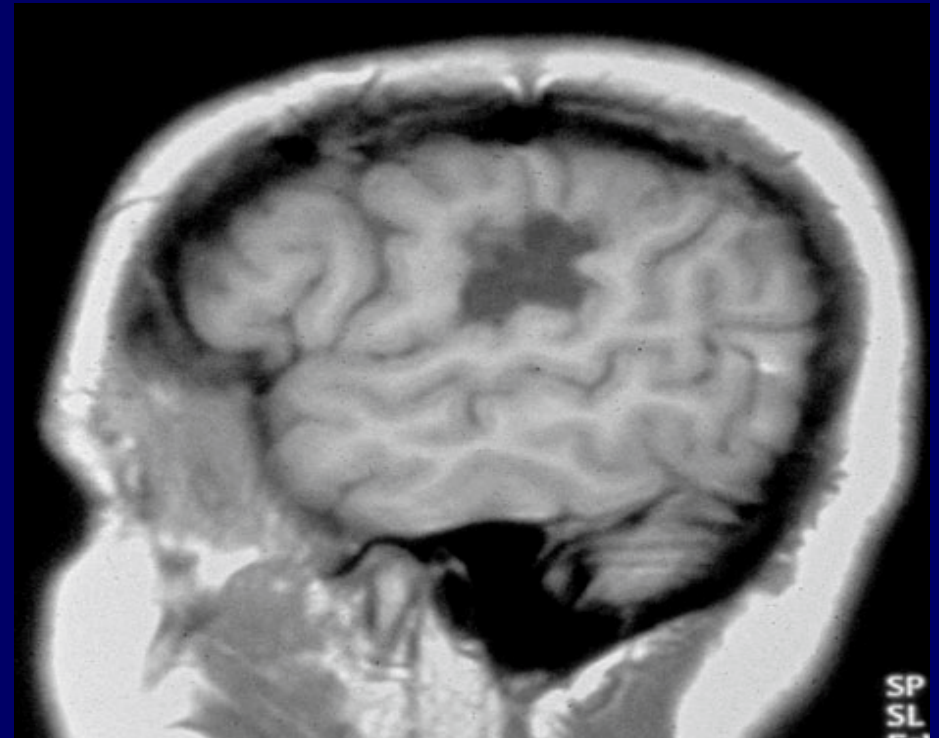
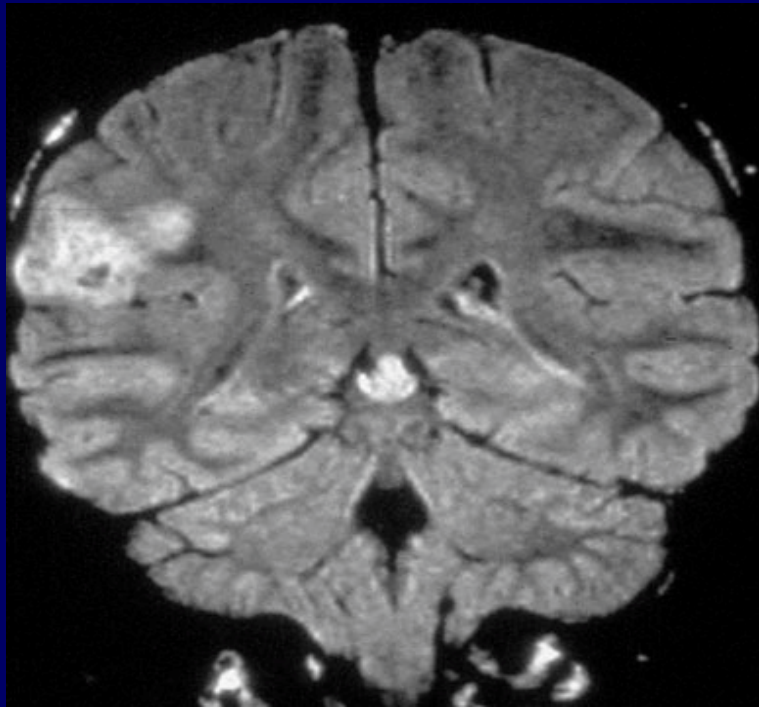


VIDEO 1

WHERE IS THIS SEIZURE ARISING FROM?

SENSORY AURA LEFT FACE -> LEFT FACE & ARM CLONIC

LESION IN POST-CENTRAL GYRUS



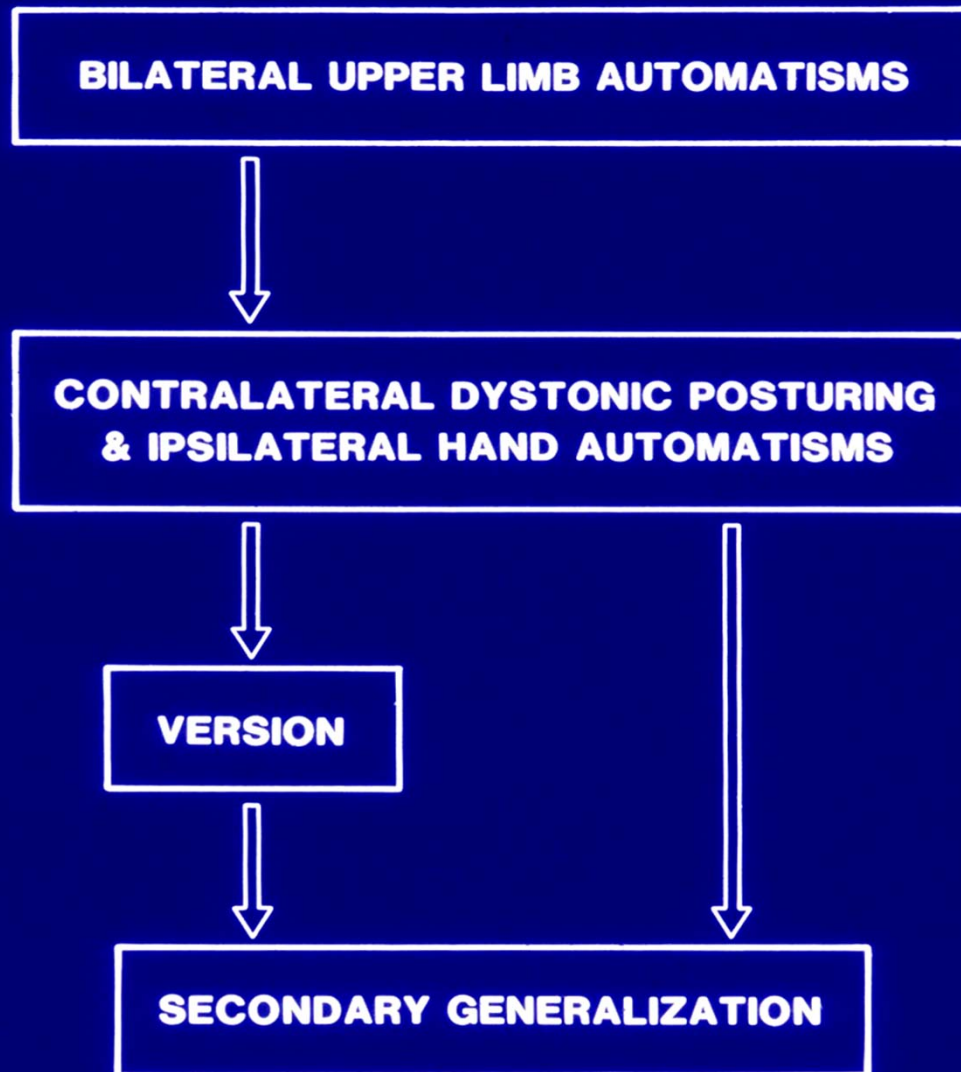
VIDEO 2

LOOK FOR ALL THE LOCALIZING AND LATERALIZING CLUES IN THIS SEIZURE

**Seizure begins
with abdominal
sensation**



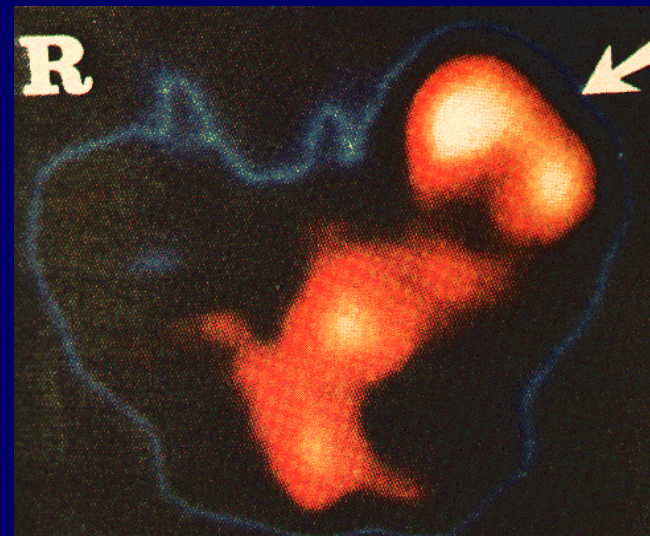
SEQUENCE OF LATERALIZING SIGNS IN MESIAL TEMPORAL LOBE SEIZURE



HYPERPERFUSION OF CONTRALATERAL BASAL GANGLIA DURING DYSTONIC POSTURING



LT ARM DYSTONIA



RIGHT ARM DYSTONIA

Newton 1992

FIGURE 4 SIGN: AYSMMETRIC LIMB POSTURING OF LEFT ARM



Kotagal P et al. Epilepsia 2000;41:457-62

VIDEO 3

**ASYMMETRIC LIMB POSTURING
(THE FIGURE 4 SIGN)**

INTER-OBSERVER AGREEMENT & PPV

SIGN	TLE n=34	XTLE n=20	p value
Dystonic Posturing			
Frequency	35%	20%	
Kappa	0.78	0.31	< 0.001
PPV	92%	100%	
Tonic Posturing			
Frequency	17.7%	15%	
Kappa	0.23	0.08	0.032
PPV	100%	-	
Immobile Limb			
Frequency	12%	-	
Kappa	0.23	0.06	0.030
PPV	100	-	

Bleasel A et al. Epilepsia 1997 ;38:168-74

ASYMMETRICAL LIMB POSTURING DURING SECONDARILY GENERALIZED SEIZURES

54 patients) seizure free > 1 year after resection
(34 temporal, 14 frontal, 3 parietal, 3 occipital)
238 seizure videos analyzed by 3 observers

Figure 4 sign	TLE	XTLE
Frequency	78.6%	87.5%
Predictive value	90.9%	87.5%
Version	TLE	XTLE
Frequency	64.3%	40%
Predictive value	100%	100%

Kotagal P et al Epilepsia 2000 ;41:457-62
Bleasel A et al. Epilepsia 1997 ;38:168-74

VALUE OF LATERALIZING SIGNS IN TLE

SIGN	FREQUENCY	PREDICTIVE VALUE
Focal clonic movements	11%	100% (p=0.05)
Version		
• < 10sec before gen	27%	100% (p<0.01)
• at any time during sz	45%	60% (NS)
Ipsilateral head tilt	9%	60% (NS)
Unilateral dystonia	18%	90% (p=0.05)
Unilateral tonic post.	13%	86% (NS)
Ipsilateral automatisms	9%	80% (NS)
Eye deviation	27%	57% (NS)
Face deviation	2%	100% (NS)
Postictal hemiparesis	2%	100% (NS)

N=55

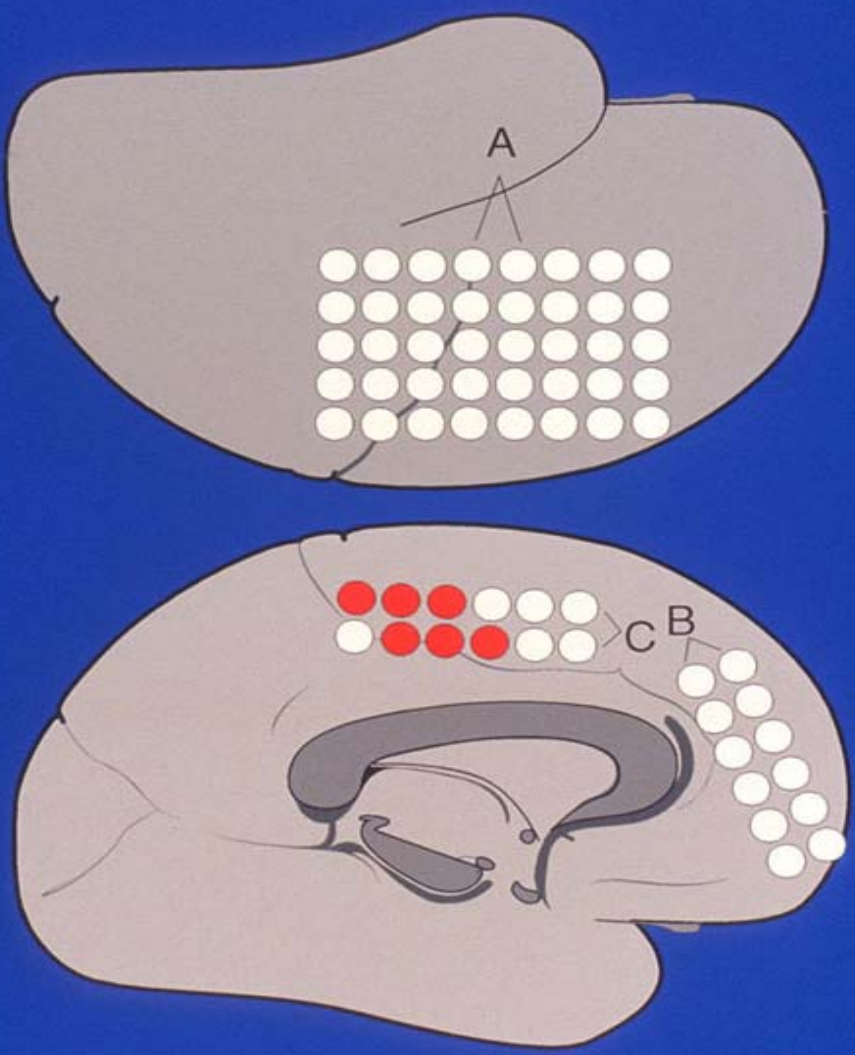
Marks WJ et al. Epilepsia 1998;39:721-6

VIDEO 4

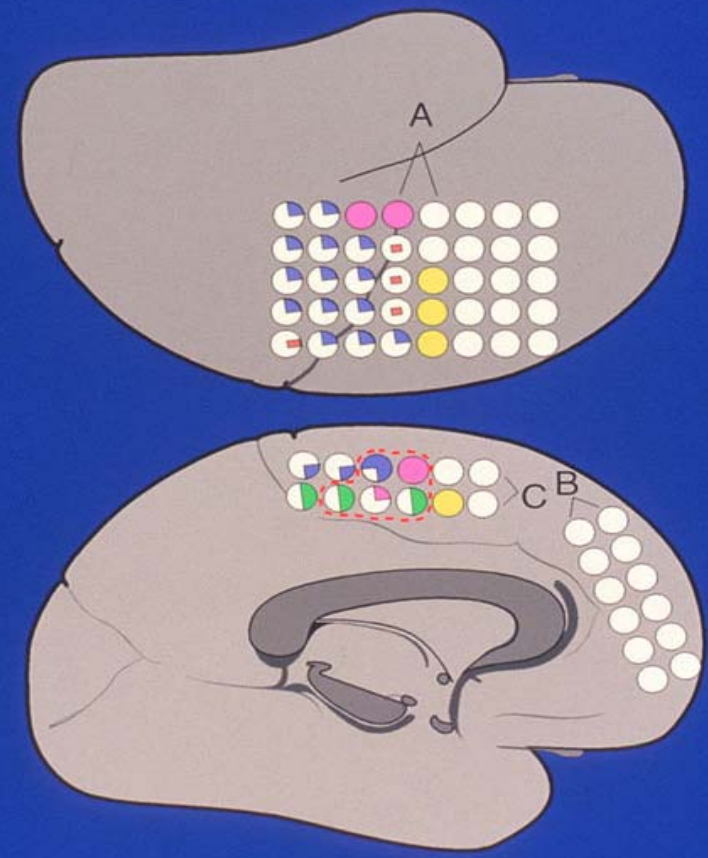
BILATERAL ASYMMETRIC TONIC SEIZURE
NOTICE RIGHT ARM EXTENSION

SUPPLEMENTARY MOTOR AREA EPILEPSY

ICTAL ONSET



CORTICAL STIMULATION



- Speech
- Negative Motor
- Face Motor
- Frontal Eye Field
- PMA
- SMA

CLASSIFICATION OF SEIZURES, EPILEPSIES & EPILEPTIC SYNDROMES



Henri Gastaut
(1915-1995)

- In 1965, **Henri Gastaut** started efforts to develop a seizure classification based on EEG and clinical features, principally consciousness
- 1971 ILAE adopted Gastaut's proposal for Classification of Epileptic Seizures, revised in 1981
- Followed by the ILAE Classification of Epilepsies and Epileptic Syndromes in 1989

1981 ILAE CLASSIFICATION OF EPILEPTIC SEIZURES

PARTIAL

Simple Partial

- motor signs
- somatosensory or special sensory symptoms
- autonomic symptoms/signs
- psychic symptoms

Complex Partial

- Simple Partial -> Complex Partial
- Complex Partial from onset

Partial -> GTC

SPS -> GTC

CPS -> GTC

SPS -> CPS -> GTC

GENERALIZED

Absence

Atypical absence

Myoclonic

Clonic

Tonic

Tonic-Clonic

Atonic

UNCLASSIFIED

(Neonatal)

ADDENDUM

Precipitating Factors

Status Epilepticus

1989 ILAE CLASSIFICATION OF EPILEPSIES & EPILEPTIC SYNDROMES

LOCALIZATION-RELATED

Idiopathic

- Benign childhood epilepsy w C-T spikes
- Childhood epilepsy w occipital paroxysms

Symptomatic

- Chronic EPC
- Epilepsies w specific modes of precipitation
- **Epilepsies by lobe of origin**
- Temporal (mesial, lateral)
- Frontal (SMA, cingulate, frontopolar, orbitofrontal, dorsolateral, opercular, motor cortex)
- Parietal
- Occipital

Cryptogenic

GENERALIZED

Idiopathic

- BFNC, BNC
- Childhood Absence
- Juvenile Absence
- Juvenile Myoclonic
- Epilepsy w GTCs on awakening, etc.

Symptomatic or Cryptogenic

- West Syndrome
- Lennox-Gastaut Syndrome
- Epilepsy w Myoclonic-Astatic seizures
- Epilepsy w Myoclonic Absences

Symptomatic

SPECIAL SYNDROMES

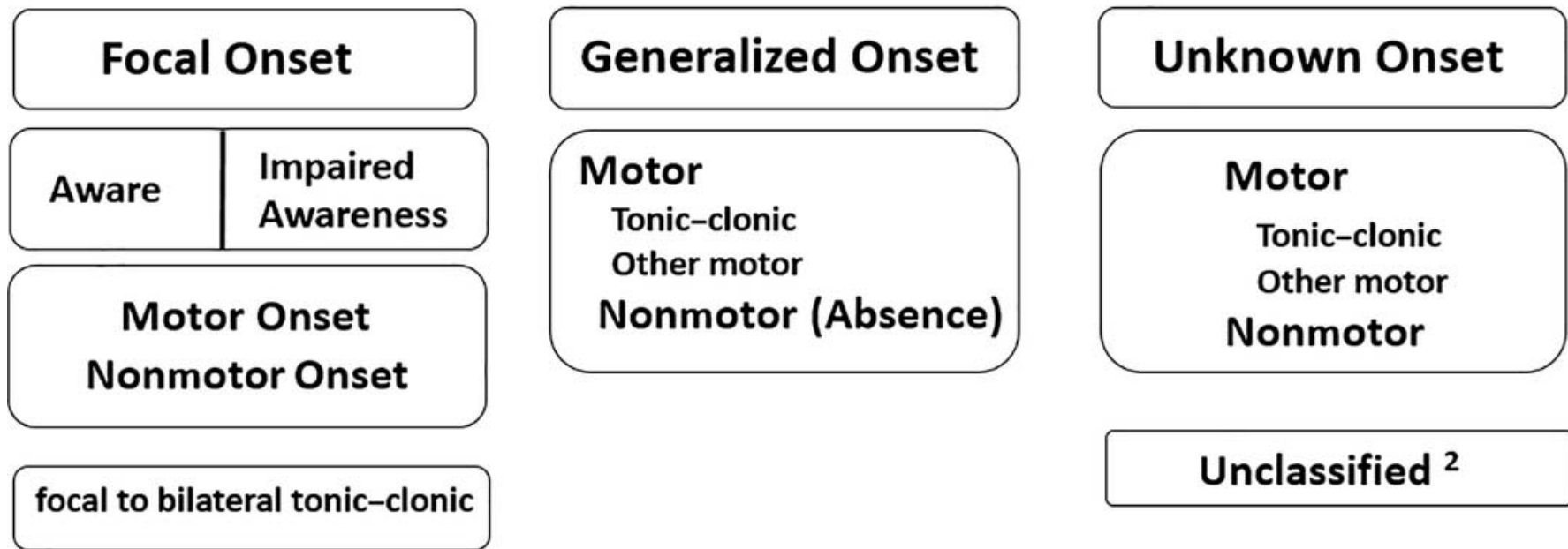
- Situation related
- Isolated seizures/status
- Metabolic / toxic

UNDETERMINED

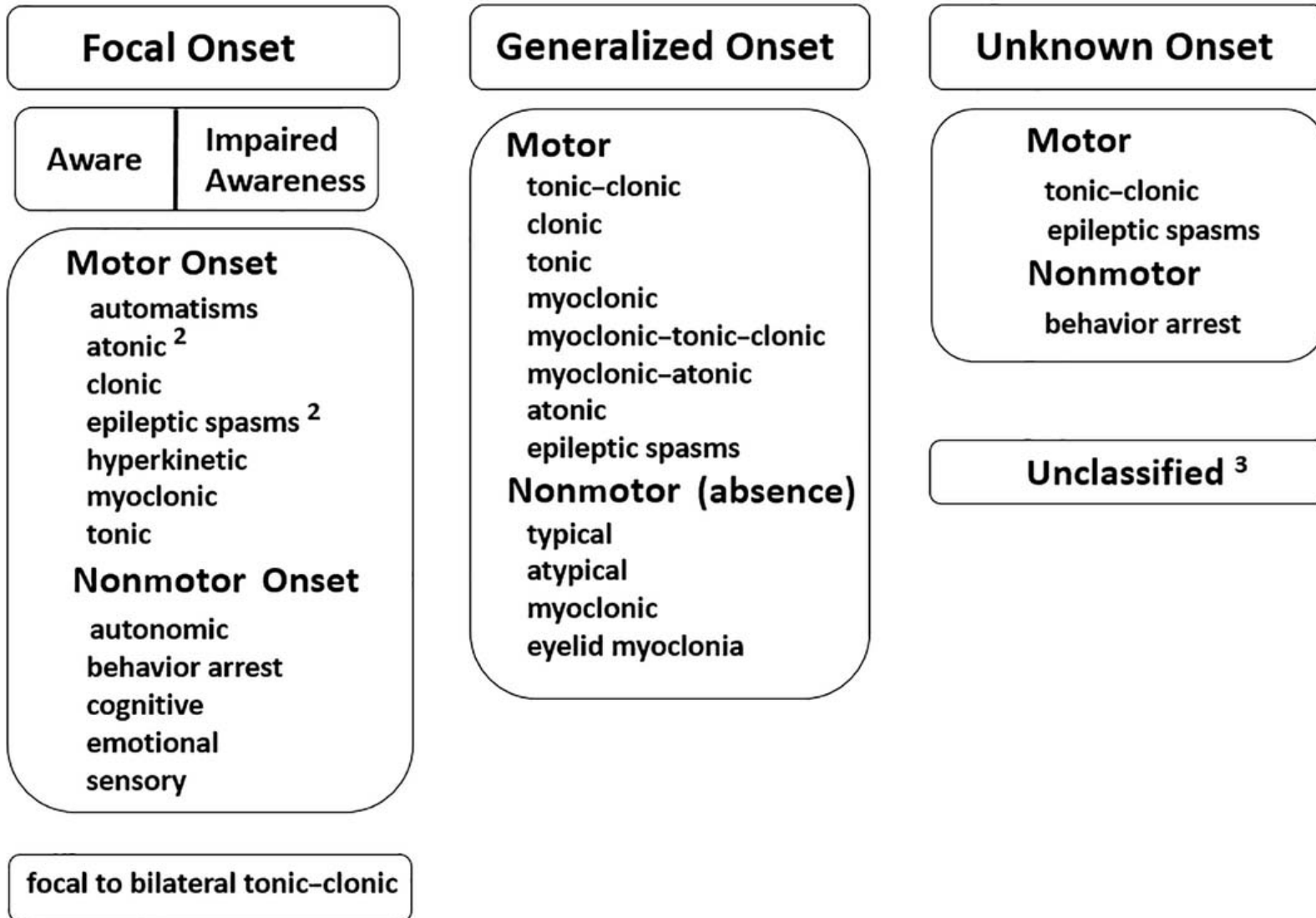
- Neonatal
- Severe Myoclonic Epilepsy of Infancy
- CSWS

Epilepsia 1989;30:389-99

ILAE 2017 Classification of Seizure Types Basic Version ¹



ILAE 2017 Classification of Seizure Types Expanded Version ¹



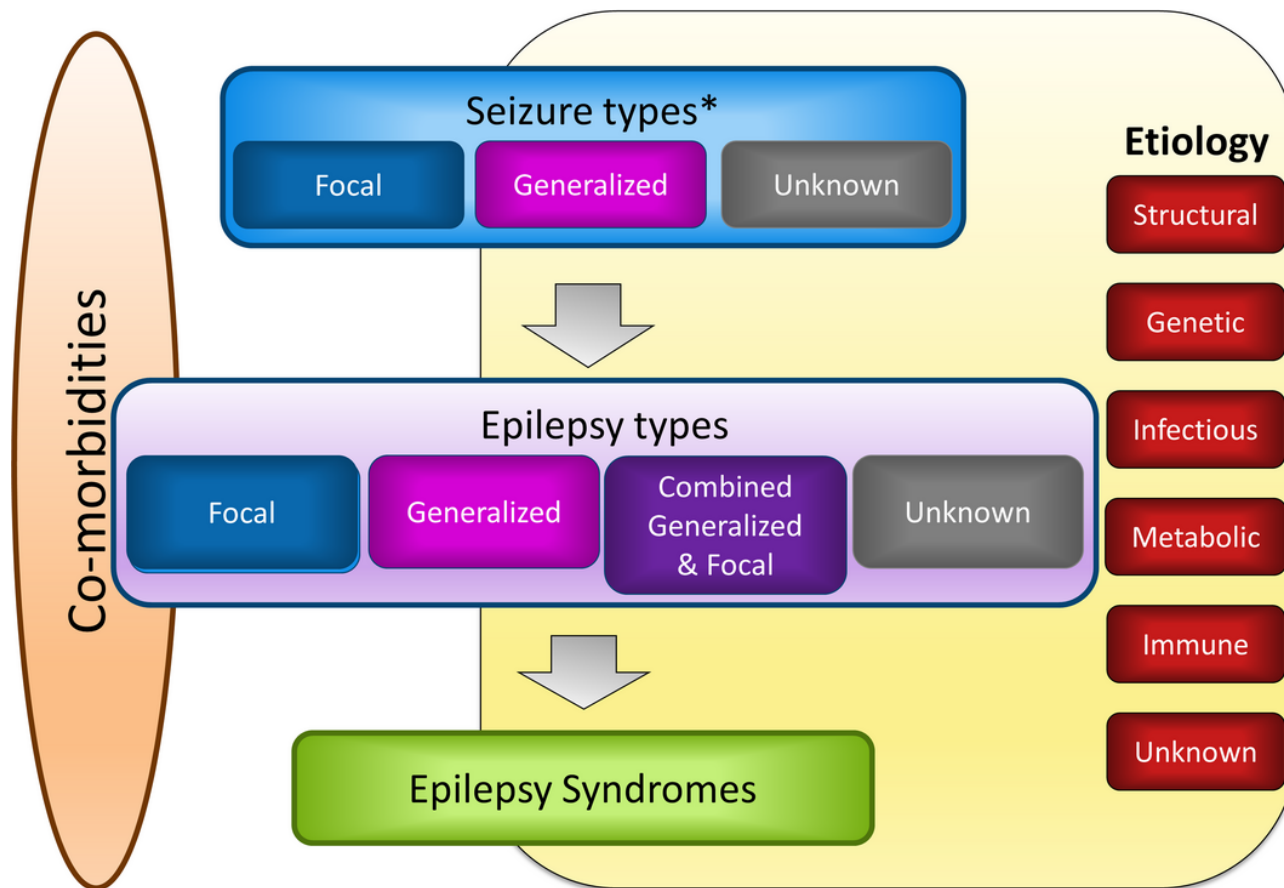
CHANGES TO SEIZURE CLASSIFICATION: FROM 1981 TO 2017

1. **Changed “partial” to “focal”**
2. Seizure onset is either **focal, generalized or unknown**
3. Seizures of unknown onset may have features that can still be classified
4. **Awareness** is used as a classifier of focal seizures
5. The terms **dyscognitive, simple partial, complex partial, psychic and secondarily generalized** are eliminated
6. **New focal seizure types** include automatisms, autonomic, behavior arrest, cognitive, emotional, hyperkinetic, sensory and focal to bilateral tonic–clonic seizures.
7. Atonic, clonic, epileptic spasms, myoclonic, and tonic seizures can be either focal or generalized
8. **New generalized seizure types** include absence with eyelid myoclonia, myoclonic absence, myoclonic–tonic–clonic, myoclonic–atonic, and epileptic spasms

CHANGES TO SEIZURE CLASSIFICATION TERMS

OLD TERM	NEW TERM
Absence, atypical absence	(Generalized) absence, atypical absence
Akinetic	Focal behavioral arrest Generalized absence
Atonic	(Focal/generalized) atonic
Complex partial	Focal unaware
Grand mal Generalized tonic-clonic	Bilateral tonic-clonic Focal to bilateral tonic-clonic Unknown onset tonic-clonic
Infantile spasms	(focal, generalized, unknown) epileptic spasms
Myoclonic	(focal/generalized) myoclonic
Petit mal	Absence
Psychomotor	Focal unaware automatism
Simple partial	Focal aware
Temporal lobe	Focal aware/unaware automatism
Tonic	(focal/generalized) tonic

ILAE CLASSIFICATION OF THE EPILEPSIES



SEMIOLOGICAL SEIZURE CLASSIFICATION



Hans Lüders

Recommended classifying seizures by semiology alone (independent of EEG, MRI)

Introduced new seizure terms: hypermotor, hypomotor, automotor, complex motor, dialeptic, and B/L asymmetric tonic

Showed how to describe the seizure evolution:

Abdominal aura -> automotor seizure -> right versive seizure -> GTC seizure

Semiology of Epileptic Seizures

- Symptoms occurring during epileptic seizures can affect the following spheres:
 - Cognitive sphere → Aura
 - Autonomic sphere → Autonomic seizure
 - Consciousness sphere → Dialeptic seizure
 - Motor sphere → Motor seizure

Dialeptic Seizures

Dialeptic seizures are characterized primarily by **alterations of consciousness or relative unresponsiveness**. Subtle motor manifestations may be observed but the alterations of consciousness is always the predominant symptom.

VIDEO 5

HYPERKINETIC (Hypermotor) SEIZURE

VIDEO 6

seizure with behavioral arrest (hypomotor)



Contents lists available at ScienceDirect

Seizure: European Journal of Epilepsy

journal homepage: www.elsevier.com/locate/seizure



Review

Could the 2017 ILAE and the four-dimensional epilepsy classifications be merged to a new “Integrated Epilepsy Classification”?



Felix Rosenow^{a,*}, Naoki Akamatsu^b, Thomas Bast^{c,V}, Sebastian Bauer^a, Christoph Baumgartner^d, Selim Benbadis^e, Adriana Bermeo-Ovalle^f, Stefan Beyenburg^g, Andrew Bleasel^h, Alireza Bozorgiⁱ, Milan Brázdil^j, Mar Carreño^k, Norman Delanty^l, Michael Devereaux^m, John Duncanⁿ, Guadalupe Fernandez-Baca Vaca^m, Stefano Francione^o, Naiara García Losarcos^m, Lauren Ghanma^m, Antonio Gil-Nagel^p, Hajo Hamer^q, Hans Holthausen^r, Shirin Jamal Omid^s, Philippe Kahane^t, Giri Kalamangalam^u, Andrés Kanner^v, Susanne Knake^w, Stjepana Kovac^x, Karsten Krakow^y, Günter Krämer^z, Gerhard Kurlmann^A, Nuria Lacuey^B, Patrick Landazuri^C, Shi Hui Lim^D, Luisa V. Londoño^E, Giorgio LoRusso^o, Hans Luders^m, Jayanti Mani^F, Riki Matsumoto^G, Jonathan Miller^H, Soheyl Noachtar^I, Rebecca O’Dwyer^J, André Palmi^K, Jun Park^L, Philipp S. Reif^a, Jan Remi^I, Americo C. Sakamoto^M, Bettina Schmitz^N, Susanne Schubert-Bast^O, Stephan Schuele^P, Asim Shahid^L, Bernhard Steinhoff^{c,V}, Adam Strzelczyk^a, C. Akos Szabo^Q, Nitin Tandon^R, Kiyohito Terada^S, Manuel Toledo^T, Walter van Emde Boas^U, Matthew Walkerⁿ, Peter Widdess-Walsh^I

Example of Classification

- ILAE classification
 - Focal aware seizure motor seizure
- Semiological classification
 - left face somatosensory aura -> left head and arm clonic seizure
 - Indicative of seizure from Right peri-Rolandic region (near face sensory area)

Example of Classification

ILAE classification

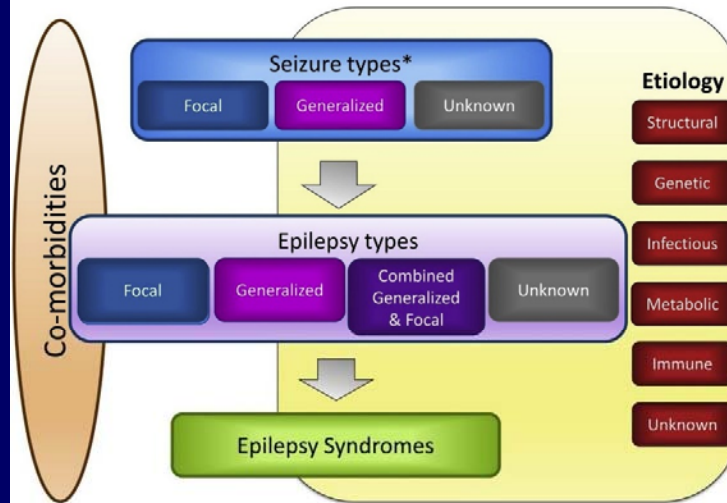
- Focal unaware automatism to bilateral tonic-clonic seizure

Semiological classification

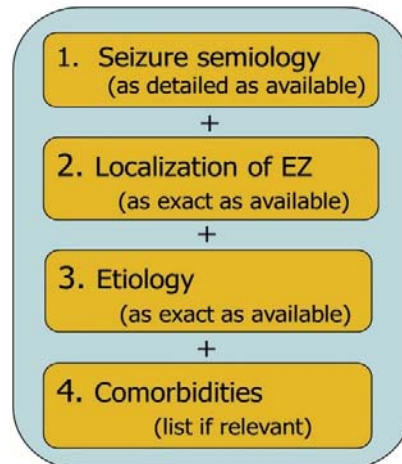
- Abdominal aura → Automotor seizure -> Right versive -> GTC

INTEGRATED EPILEPSY CLASSIFICATION

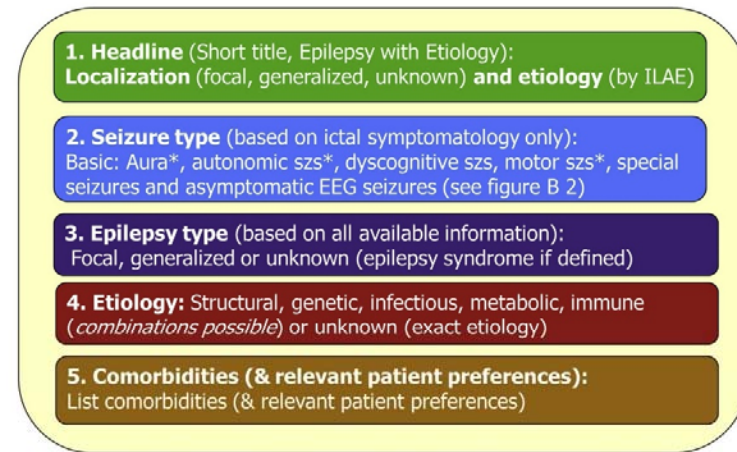
A) 2017 ILAE Epilepsy Classification (from reference 1)



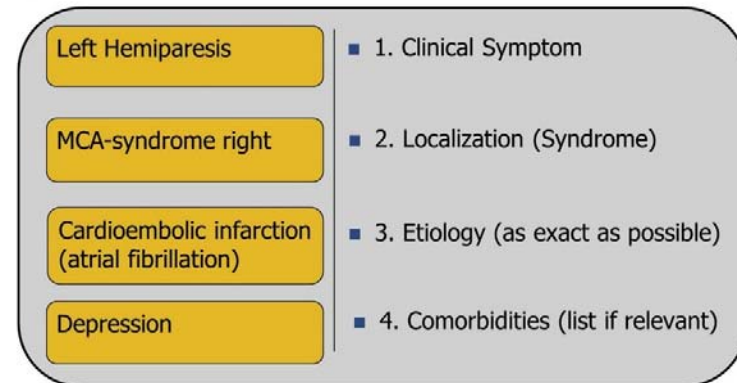
B) 4-Dimensional Epilepsy Classification (19)



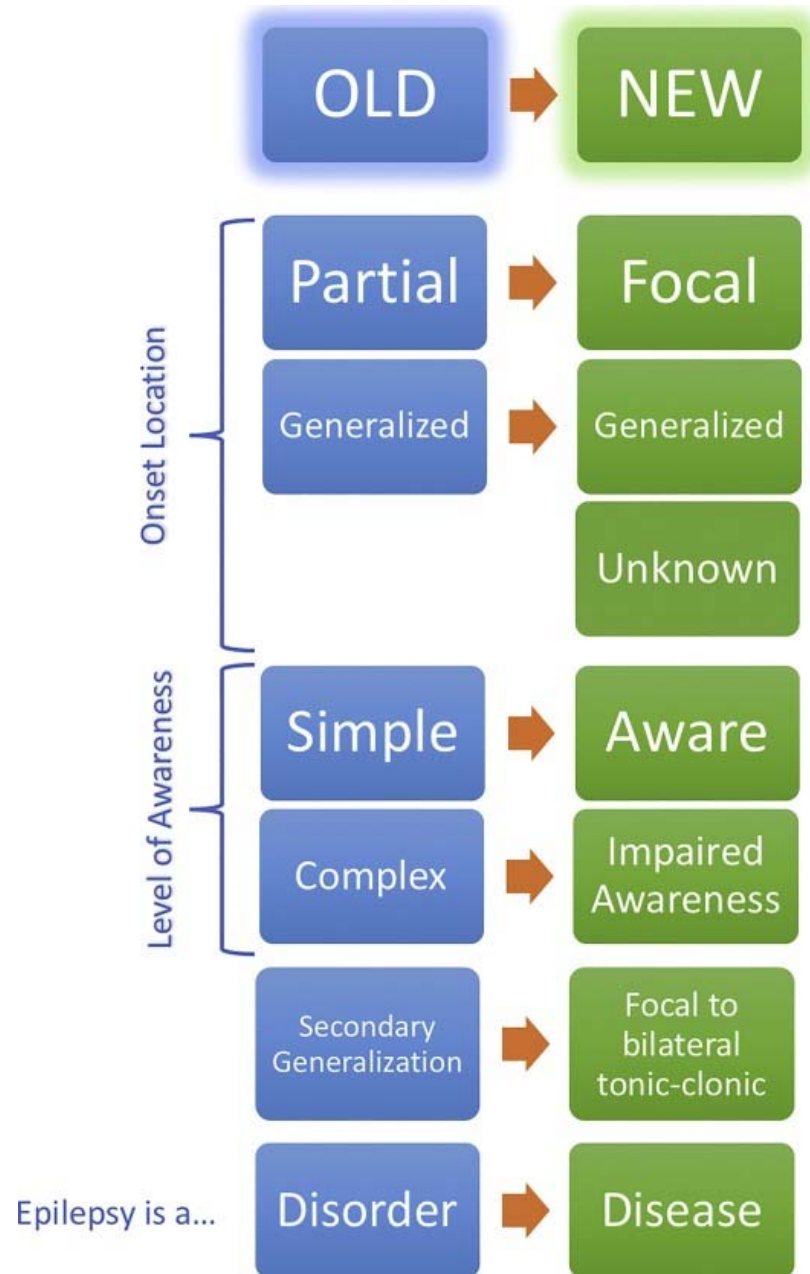
C) Integrated Epilepsy Classification



D) Classification of a Stroke Patient in General Neurology



THE NEW ILAE CLASSIFICATION SIMPLIFIED!



*Falco-Walter JJ, Scheffer IE, Fisher RS.
Epilepsy Res 2018;139:73-9*