EPILEPSY SOCIETY OF THAILAND

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Pediatric Epilepsy Center of Excellence
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Ictal semiology:
Localization of Seizure Onset and Propagation Networks

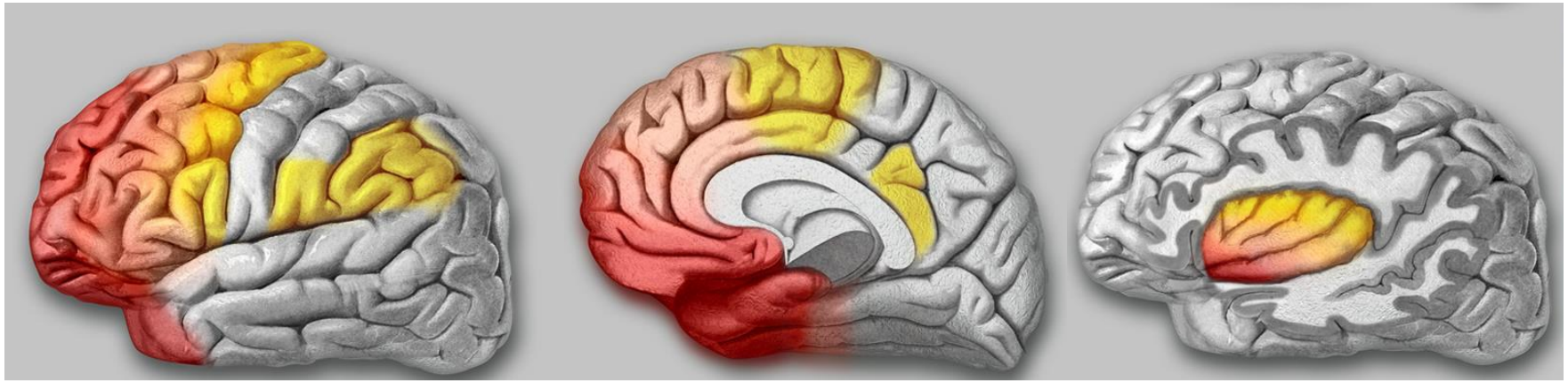
Piradee Suwanpakdee, MD
Head of Division of Neurology
Department of Pediatrics
Phramongkutklao Hospital and College of Medicine, Bangkok, Thailand

Ictal semiology

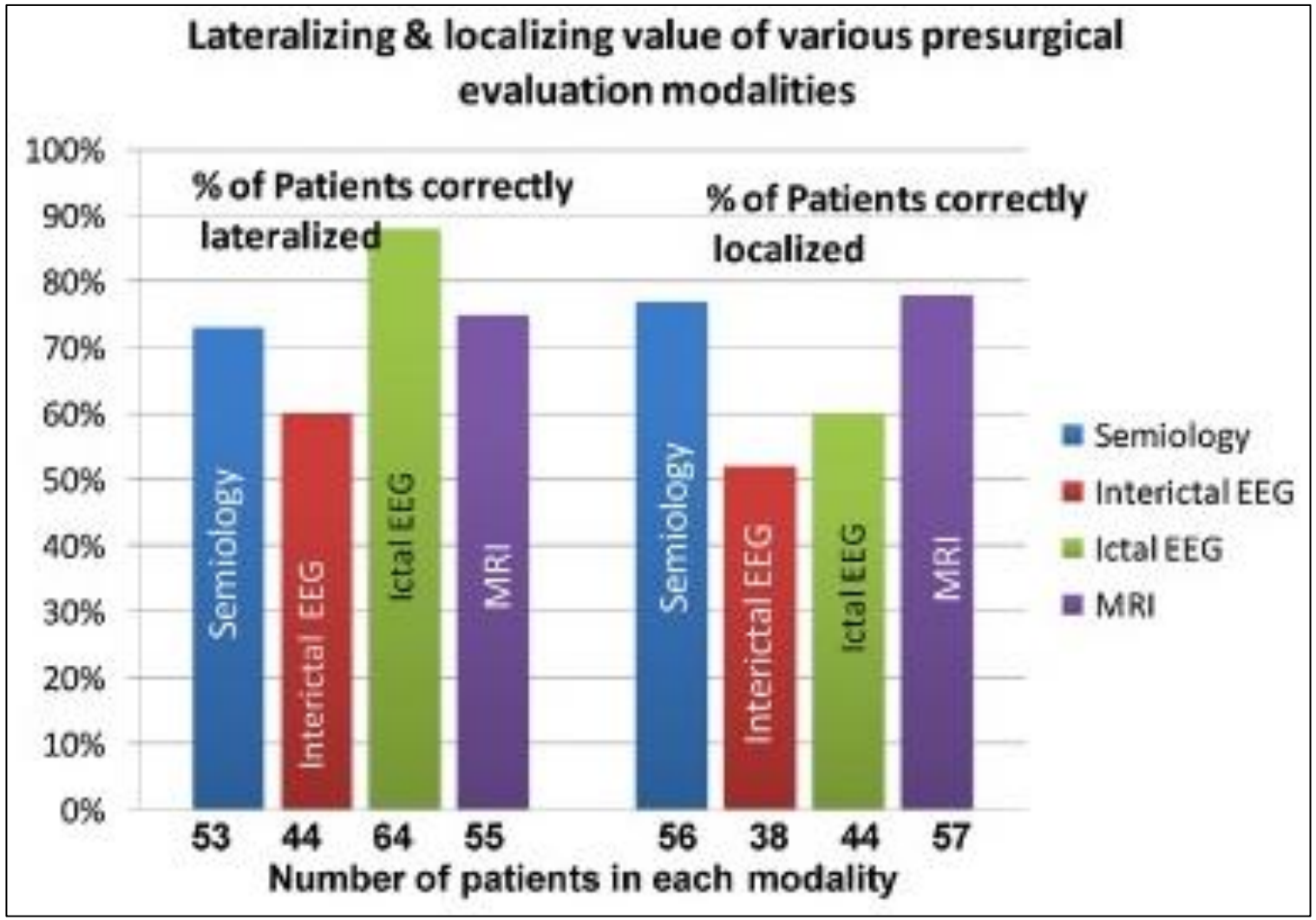
- ***Semiology or Semiotics : "The study of signs and sign-using behaviour"***

- The semiology of epileptic seizures reflects activation, or dysfunction, of areas of brain (often termed 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"**



The important of ictal semiology



Sherif Elwan, et al. Seizure 2018.

Ictal Semiology approach

Description and Terminology

Symptoms	Medical Term
automatic behaviors	automatisms
emotions or appearance of emotions	emotions
extension or flexion postures	tonic
flushing/sweating/piloerection	autonomic
jerking arrhythmically	myoclonus
jerking rhythmically	clonus
language or thinking problems, deja vu	cognitive
lid jerks	eyelid myoclonia
limp	atonic
numb/tingling, sounds, smells, tastes visions, vertigo	sensations
pausing, freezing, activity arrest	behavior arrest
thrashing/pedaling	hyperkinetic
trunk flexion	spasm

ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
- automatisms
- atonic [2]
- clonic
- epileptic spasms [2]
- hyperkinetic
- myoclonic
- tonic

Nonmotor 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

Nonmotor (absence)
- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

Motor
- tonic–clonic
- epileptic spasms

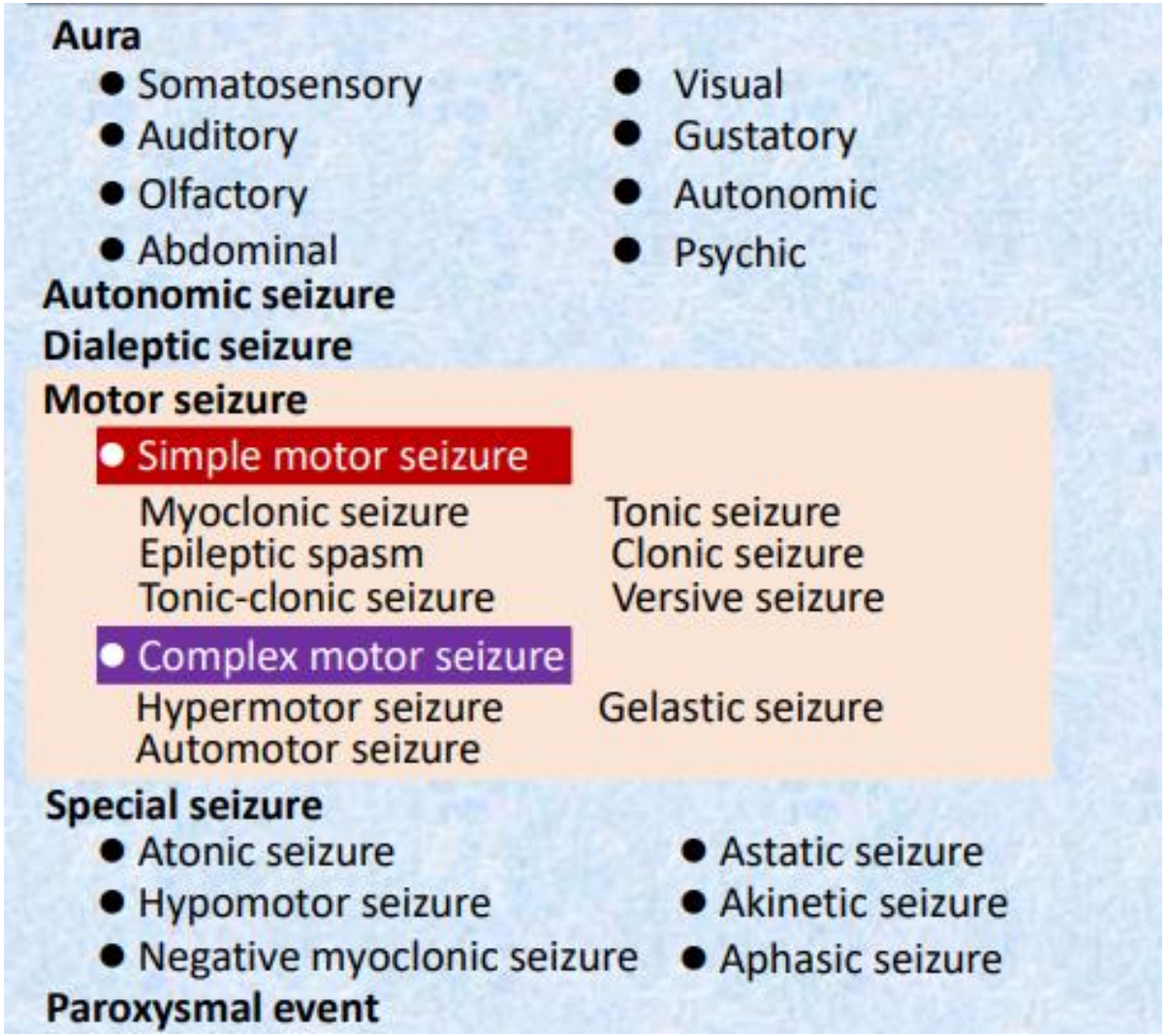
Nonmotor
- behavior arrest

Unclassified [3]

Ictal semiology: Less details

Localization and lateralization value

Seizure Semiological Classification



Hans Lüders, 1998

Motor seizure

- Simple Motor Seizure:
  - Unnatural but simple movements, usually involving only one articulation in one plane
  - Can be reproduced by electrical cortical stimulation of the motor areas

- Complex Motor Seizure:
  - Imitate natural movements involving several articulations in different planes
  - Tend to be repetitive
  - Cannot be elicited by electrical cortical stimulation unless a seizure discharge is triggered

K. Tufenkjian, HO Lüders, J Clin Neurol 2012

Motor seizure

- Simple Motor Seizure:
  - Unnatural but simple movements, usually involving only one articulation in one plane
  - Can be reproduced by electrical cortical stimulation of the motor areas

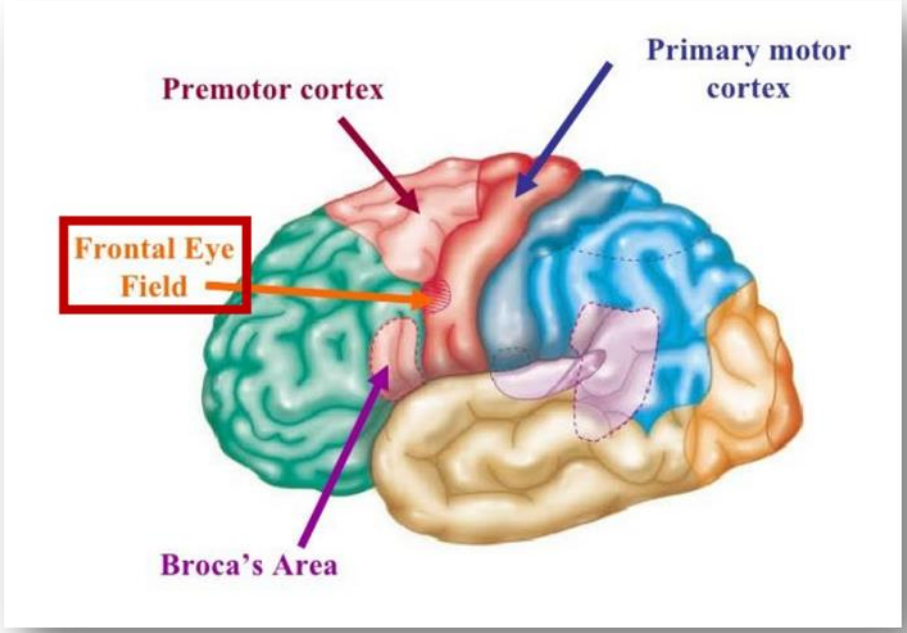
Semiology: Clonic seizure
Localization: primary motor cortex
Lateralization: contralateral

K. Tufenkjian, HO Lüders, J Clin Neurol 2012

Versive seizure

- Forced and involuntary turning of the head and eyes in one direction with an associated neck extension resulting in a sustained unnatural position

- Symptomatogenic zone-> Frontal eye fields, highly lateralizing to the contralateral hemisphere



Common lateralizing signs of focal seizures

Subtype	Symptomatogenic zone	Lateralization
Dystonic limb posturing	Activation of basal ganglia	Contralateral
Tonic posturing	Activation of SSMA, basal ganglia, cingulum, and primary motor cortex	Contralateral
Eye version	Frontal eye fields (area 8) and extrastriate cortex (area 19)	Contralateral
Head version	Premotor area and Frontal eye fields (areas 6 and 8)	Contralateral
Asymmetric tonic limb posturing	SSMA and precentral area	Contralateral

Modified from Foldvary-Schaefer N, Unnwongse K. *Epilepsy Behav*. 2011

Complex Motor Seizure

Hypermotor	• Involving the proximal segments of the limbs and trunk • Large movements that appear “ violent” when they occur at high speeds
Automotor	• Repetitive, stereotyped, semipurposeful motor behaviors, involving primarily distal limbs, mouth, and tongue
Gelastic seizure	• Main motor is “laughing”

Localization of hypermotor seizure

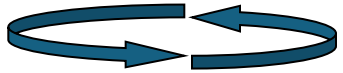
- HMS are reported in 15–27% of frontal lobe epilepsies (FLE) (Swartz, 1994; Manford et al., 1996)

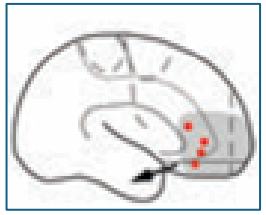
- Hypermotor seizures are generally considered to originate from the medial frontal gyrus, anterior cingulate cortex, or orbitofrontal and frontal polar regions (Williamson et al., 1985)

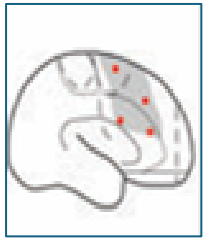
- Might also originate from temporal lobe, insula and parietal lobe

CHARACTERISTICS OF HYPERMOTOR SEIZURES

	TYPE I	TYPE II
Agitation	Marked	Mild
Hypermotor	Sitting up Laying down Kicking/boxing	Rotation of trunk horizontally
Head deviation	Ipsilateral	+/- contra
Expression	Fear, anger	-
LOC	Yes	Variable
Amnesia	Yes	Variable
Vegetative	urine incontinence facial flushing	
Location of EZ	Ventromesial	mesial premotor

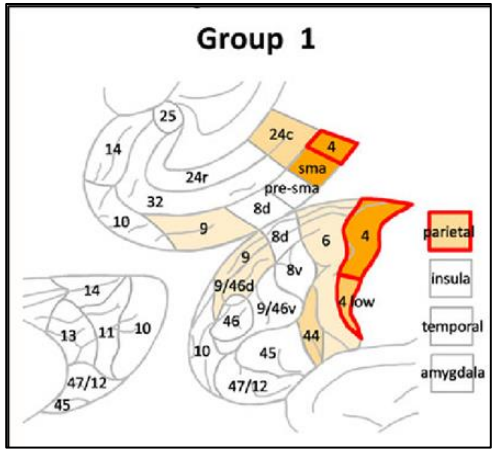




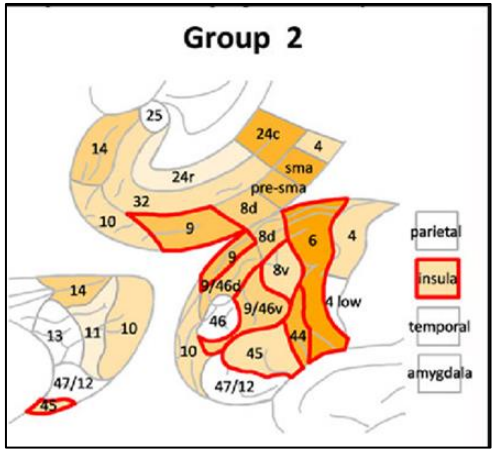


Rheims S et al. Epilepsia 2008

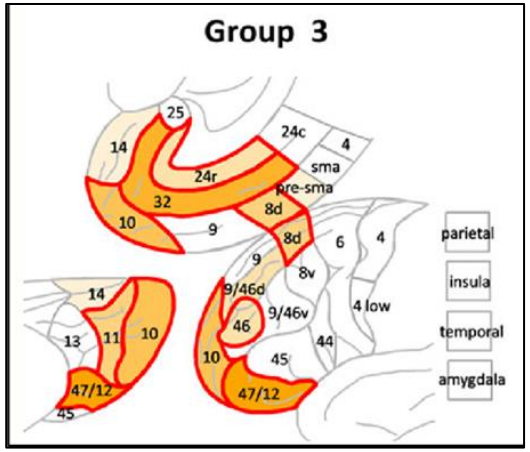
Frontal lobe seizures: From clinical semiology to localization



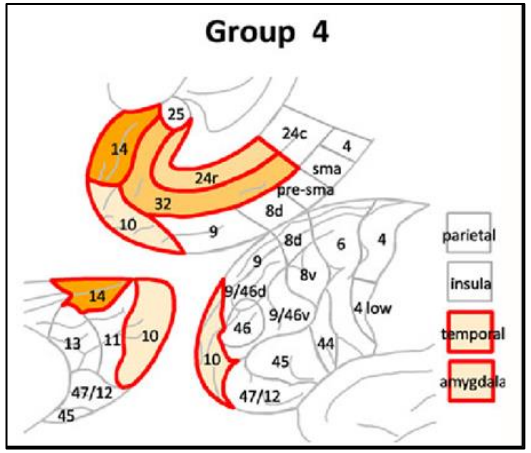
One or more motor signs
- Somesthetic localized aura and tonic vocalization
- Absence of gestural motor behavior and of emotional features
- Early spread network: significant involvement of rolandic cortex, rolandic operculum, parietal cortex; minor involvement of lateral and medial premotor cortices.
- Ictal discharge: medial and lateral premotor regions at onset



Co-occurrence of elementary motor signs (typically symmetric axial tonic posture and facial contraction such as “chapeau de gendarme”) and nonintegrated gestural motor behavior
- Nonlocalized aura and more complex nonverbal vocalization
- Absence of integrated gestural motor behavior, distal stereotypies, early clonic signs, and fixed facial expression
- Early spread network: both premotor and lateral prefrontal regions
- Ictal discharge: both medial and lateral aspect at onset



Integrated gestural motor behavior with distal stereotypies, fixed facial expression or positive emotional expression, proximal stereotypies and speech production
- Absence of any elementary motor signs
- Early spread network and ictal discharge: rostral prefrontal ventrolateral regions, rostral cingulate gyrus



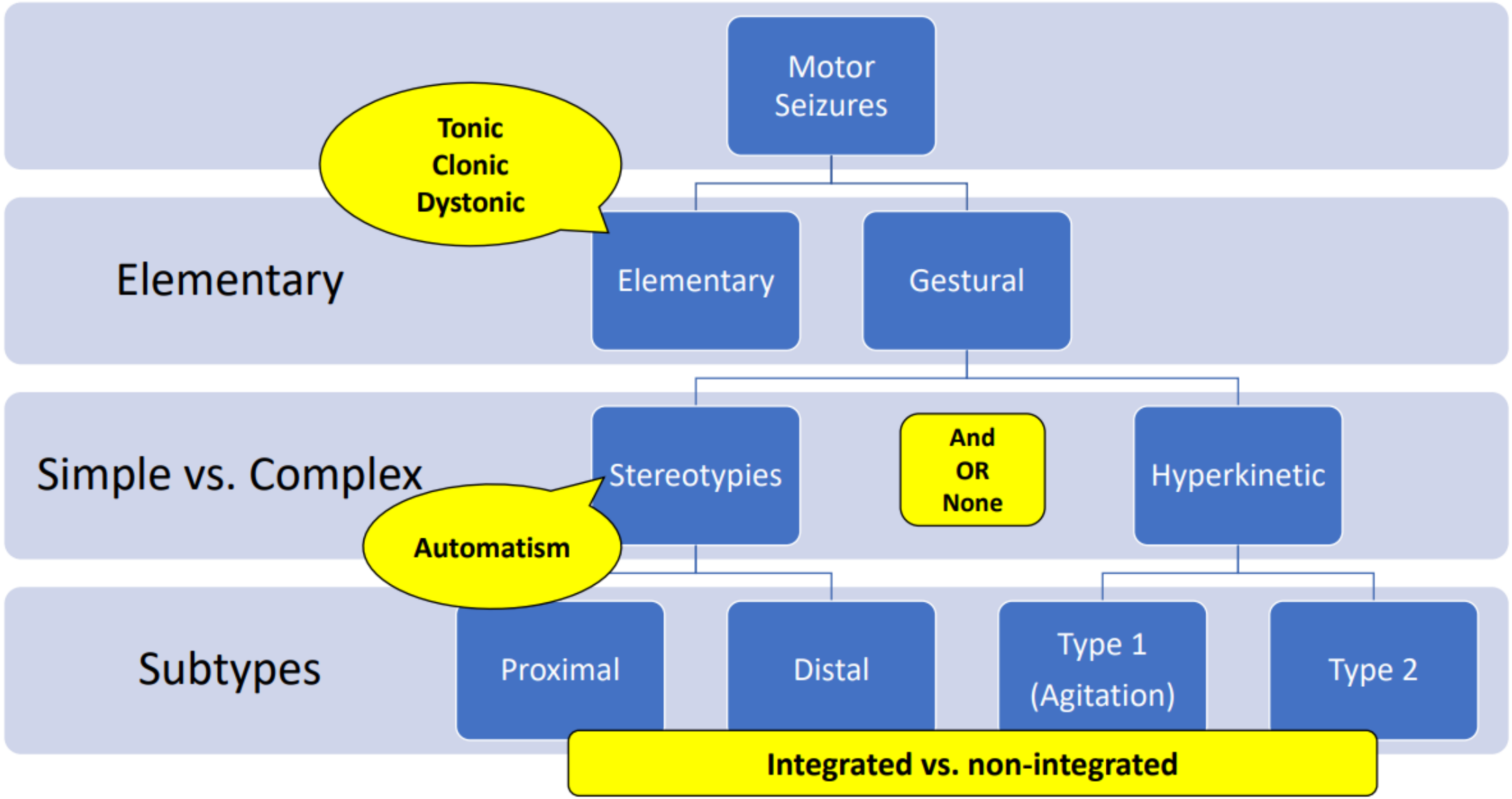
Integrated gestural behavior of fear, sometimes hyperkinetic, with attempt to fight or to escape,frightened facial expression,sometimes screaming or swearing,
and autonomic signs.
- Absence of elementary motor signs
- Early spread network and ictal discharge: orbital and medial prefrontal network with propagation to amygdala and anterior temporal regions

Bonini F, et al. Epilepsia 2014

Motor Semiology - Terminology

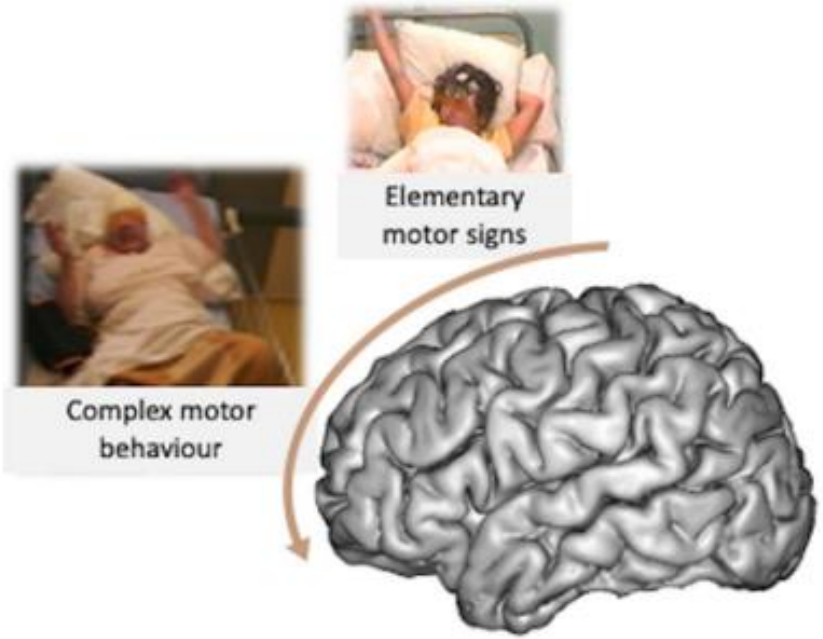
- Elementary: clonic movements and tonic or dystonic contraction
- Stereotypies: excessive production of one type of motor act, necessarily resulting in repetition.
  - proximal stereotypies: rhythmic, repetitive movements of trunk and limbs
  - distal stereotypies: hands/feet
  - non-purposeful appearance (e.g., whole body rocking) or a semi-purposeful one (e.g., manipulating an object)
- Hyperkinetic: an excessive amount of movement (hyperactivity) and/or excessive amplitude, speed, and acceleration
- Integrated: sequence of movements appeared to follow a recognisable, ordered sequence of movement (naturalistic) within the seizure, such as reaching, grasping, pedalling, kicking, tapping, rocking

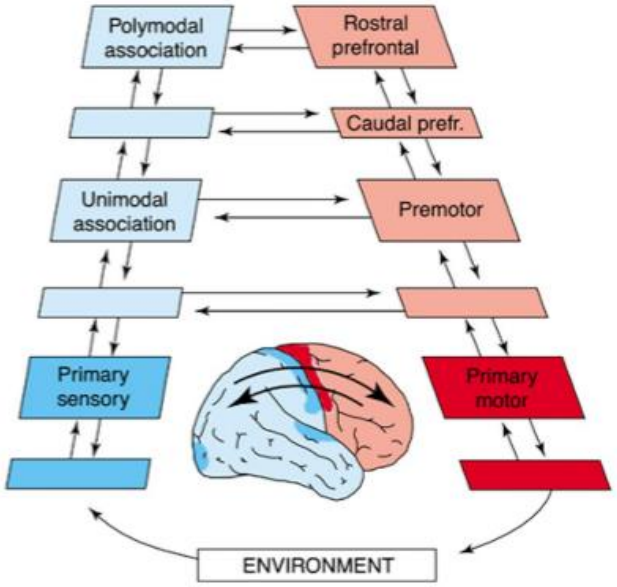
Motor Semiology - Classification

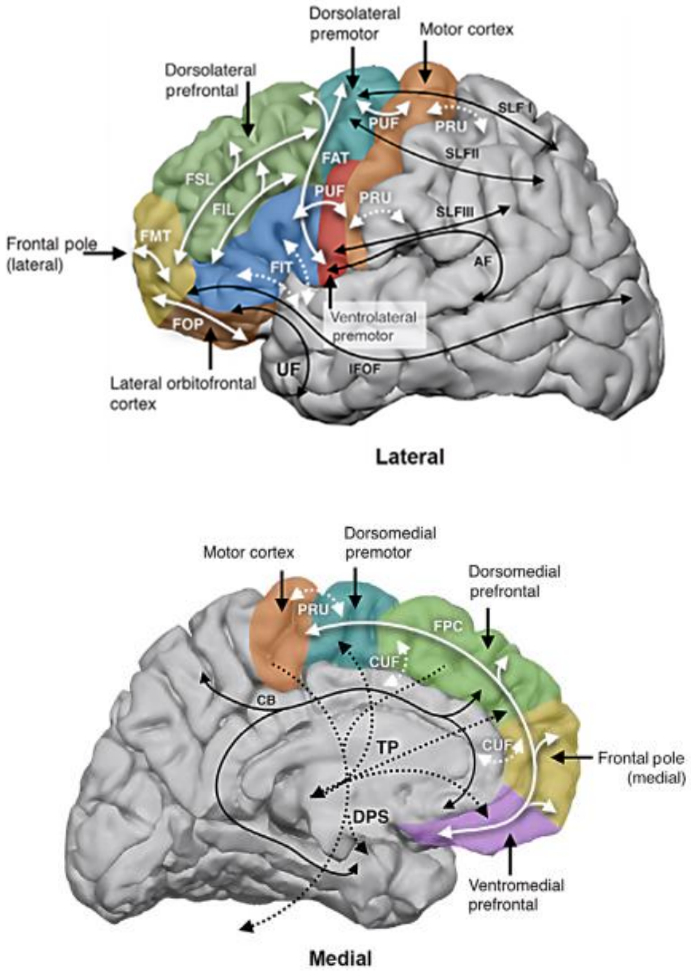


Courtesy: Prof. Lim Kheng Seang

Frontal lobe seizure semiology and neural correlates





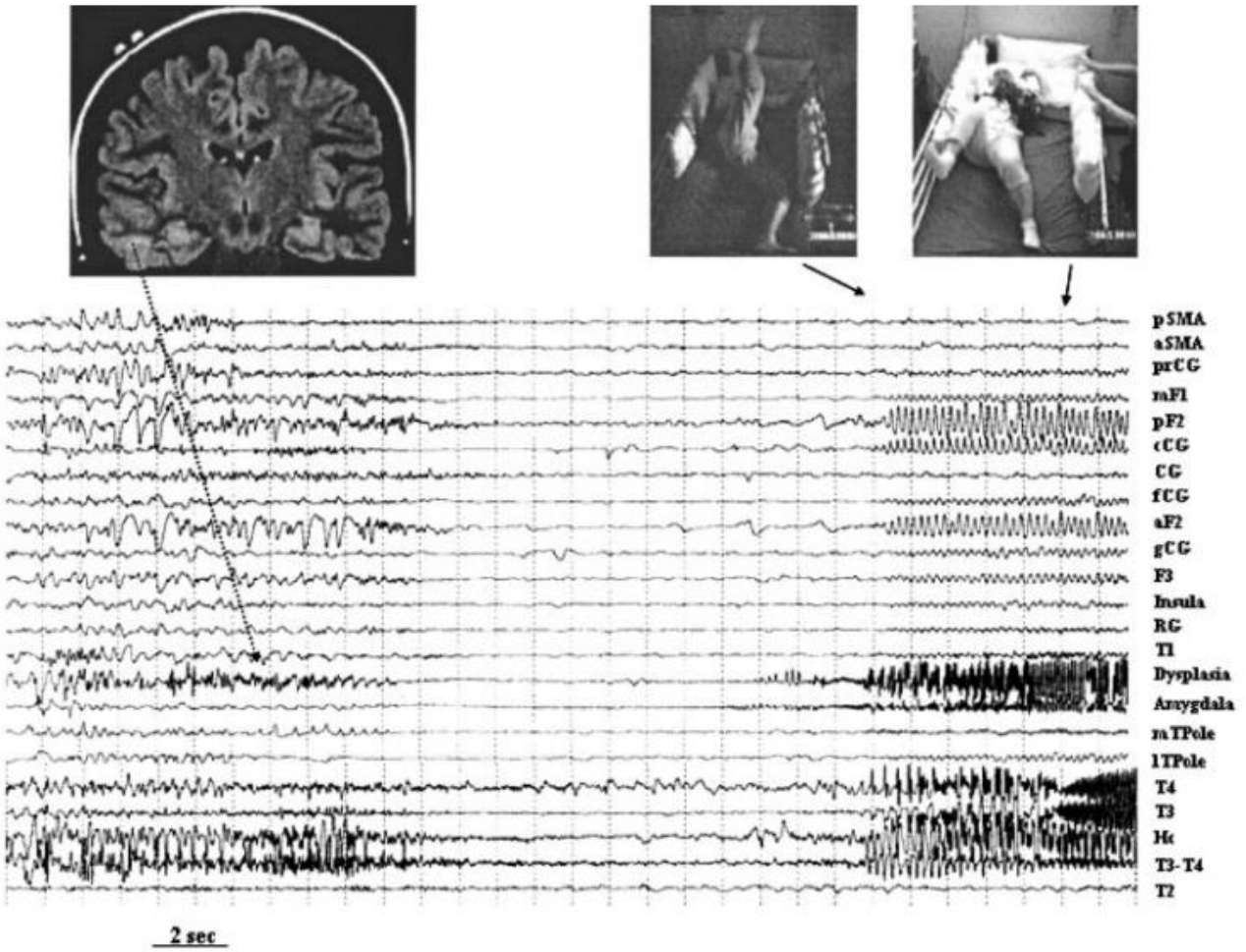


- A spatial hierarchy along a rostro-caudal (antero-posterior) axis
- elementary motor signs are associated with primary (and supplementary) motor regions
- complex motor behaviors occur with more anterior prefrontal seizure organization

McGonigal A, Journal of Neurology 2022

Sleep-related hyperkinetic seizures of temporal lobe origin

L. Nobili, MD, PhD; M. Cossu, MD; R. Mai, MD; L. Tassi, MD; F. Cardinale, MD; L. Castana, MD; A. Citterio, MD; I. Sartori, MD; G. Lo Russo, MD; and S. Francione, MD, PhD

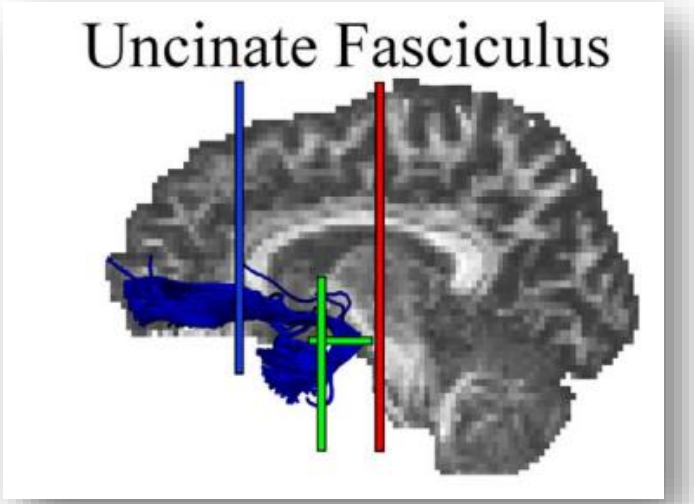


Nobili L, et al. Neurology 2004

Temporal lobe origin

- Hypermotor seizures are more common in patients with neocortical temporal lobe epilepsy (6%) than with mesial temporal lobe epilepsy (1%) (Pfander et al, 2002)

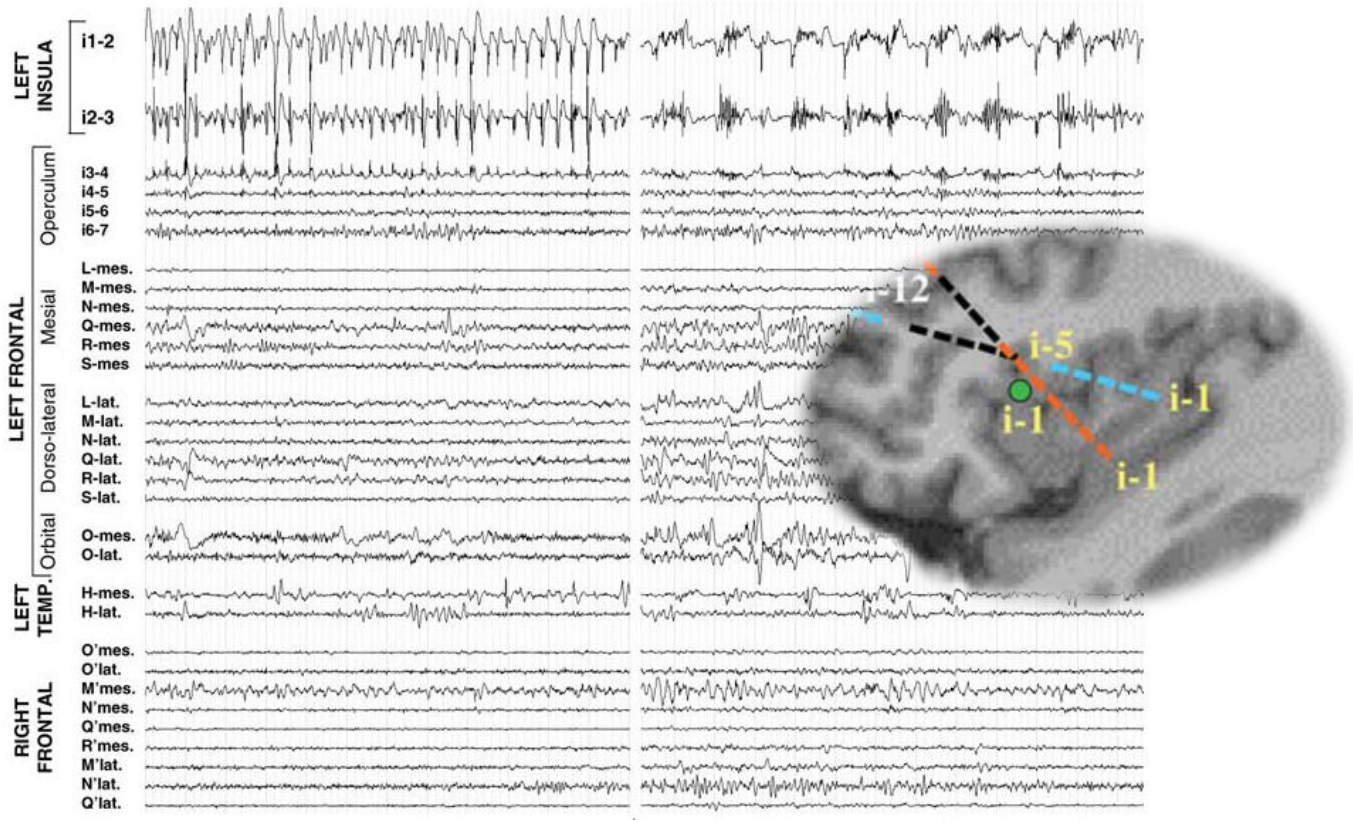
- The temporal pole is extensively connected with the orbitofrontal and medial prefrontal cortex in primate studies (Carmichael and Price, 1995; Kondo et al., 2003; Kendrick and Gibbs, 1958), and this connection has been demonstrated physiologically in humans (Kendrick and Gibbs, 1958).



Epilepsia, **47**(4):755–765, 2006
Blackwell Publishing, Inc.

Nocturnal Hypermotor Seizures, Suggesting Frontal Lobe Epilepsy, Can Originate in the Insula

*Philippe Ryvlin, ‡Lorella Minotti, *Geneviève Demarquay, §Edouard Hirsch, ‖Alexis Arzimanoglou, ‡Dominique Hoffman, †Marc Guénot, ¶Fabienne Picard, *Sylvain Rheims, and ‡Philippe Kahane



Patients <u>did not</u> demonstrate clinical data that would clearly distinguish their phenotype from typical NFLE

The anterosuperior portion of the insula might play a pivotal role in generating nocturnal hypermotor seizures

Automatism - Subtypes

- Oral automatism
- Limbs and body – non-purposeful or semi-purposeful, stereotyped, and repetitive behaviours
    - Gestural motor behaviour
        - Stereotypies – distal or proximal
        - Integrated vs Nonintegrated
- Verbal automatism – production of single or repetitive words, phrases, or brief sentences
- Automatism in Absence Seizures

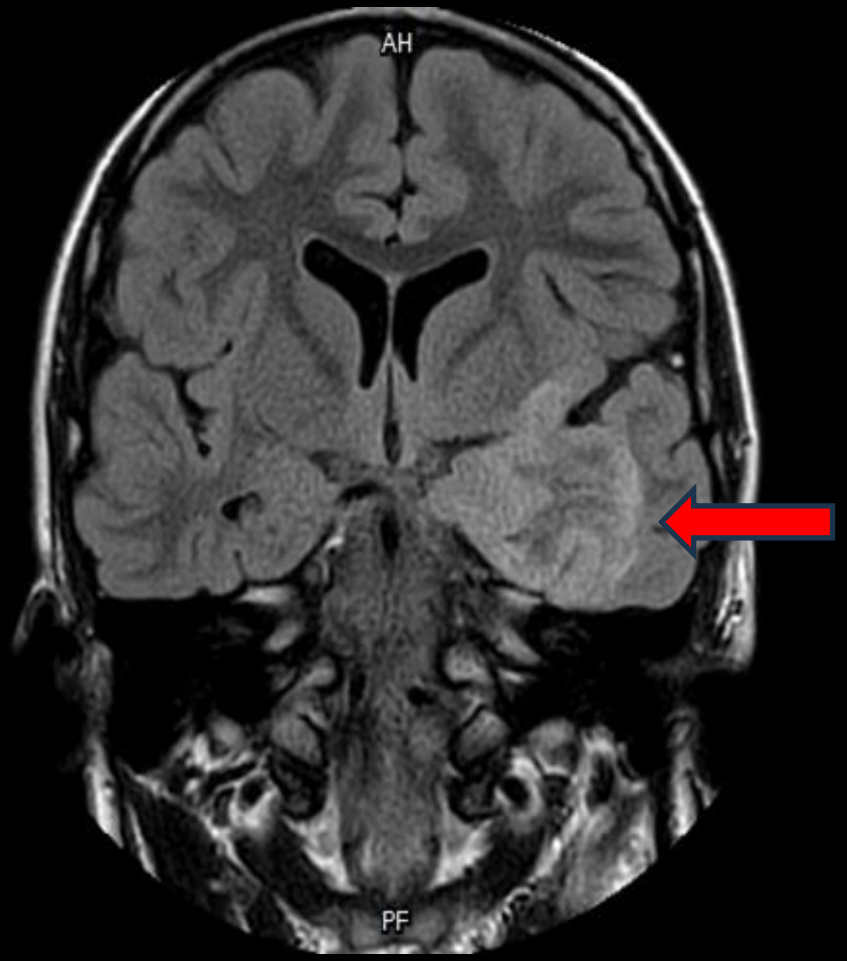
Case A 6 years old boy with drug-resistant epilepsy, RH

- Description:
  - Facial expression (some feeling?)
  - Proximal body movement
  - Bilateral hand tapping
  - Mouth chewing
  - Nonresponsive

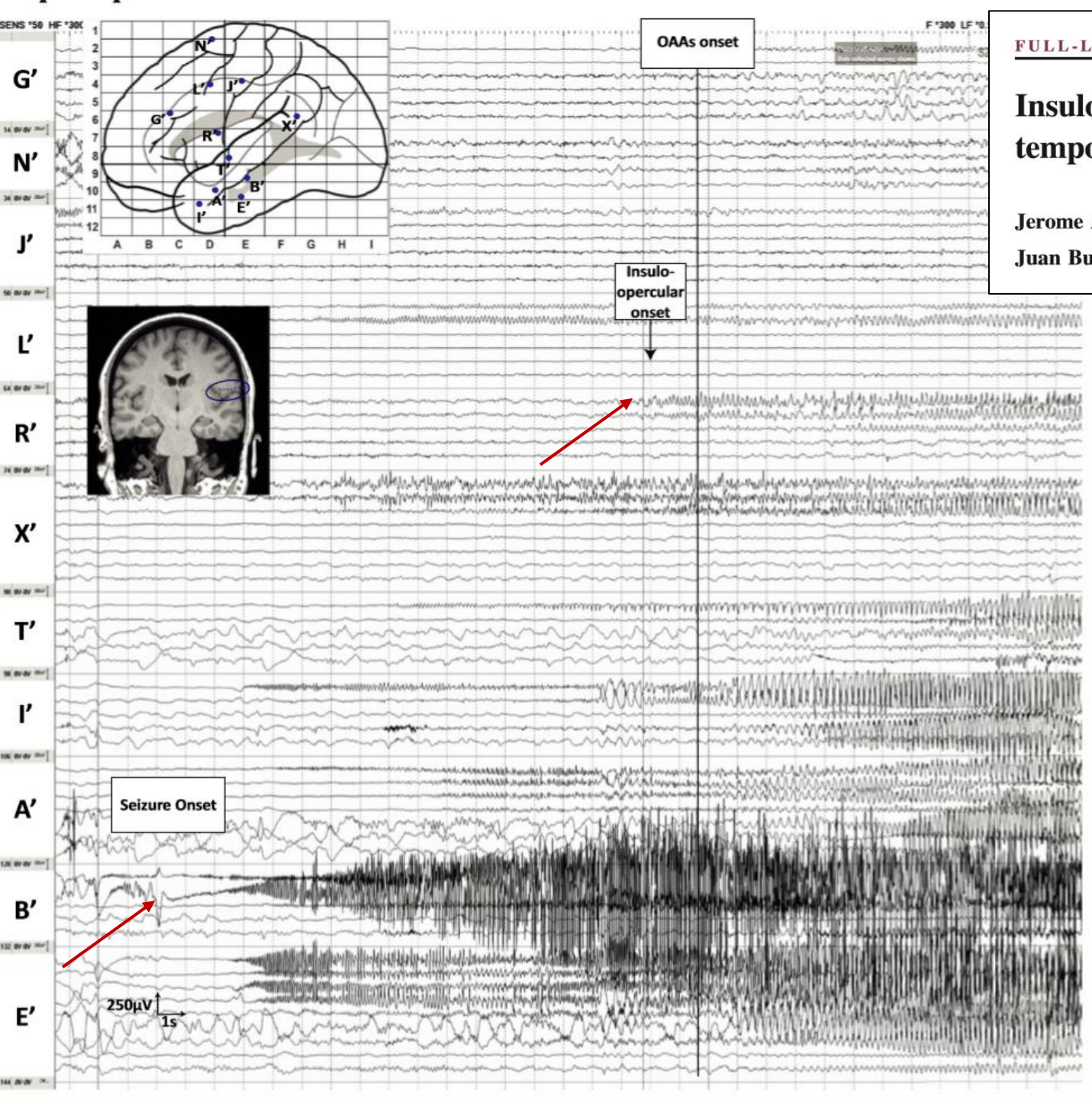
- Specific terminology:
  - Aura (fear)
  - Proximal stereotypies/complex motor
  - Bimanual automatism
  - Oroalimentary automatism
  - Impaired awareness

Semiology of Mesial Temporal lobe epilepsy

- Motor manifestations are more common in children before 6 years of age (Fogarasi et al., 2002)
- Focal seizures are often hypomotor or automotor with limited motor movements and a fixed motionless stare. Additionally, oral and manual automatisms with impaired consciousness for 30-60 seconds frequently accompany the the symptoms
- Pupillary dilation, hyperventilation, piloerection, and tachycardia are common autonomic features
- Dystonic posturing contralateral to the hemisphere of seizure origin with ipsilateral automatisms during the seizure and lateralizing signs (So et al, 2006)



Operation: Tumor resection (23/05/2023)
Pathology : Low-grade glioma
Seizure outcome: Seizure free since surgery



FULL-LENGTH ORIGINAL RESEARCH

Epilepsia

Insulo-opercular cortex generates oroalimentary automatisms in temporal seizures

Jerome Aupy[1,2,3] | Ika Noviawaty[1,4] | Balu Krishnan[1] | Piradee Suwankpakdee[1,5] | Juan Bulacio[1] | Jorge Gonzalez-Martinez[1] | Imad Najm[1] | Patrick Chauvel[1]

- In seizures with medial temporal onset, oroalimentary automatism occurrence depends on ictal discharge propagation to ***operculo-insular areas***

Spatial features

- Rhythmically synchronized activity at ***theta frequency*** between amygdala-hippocampus and operculo-insular cortex underlies the emergence of oroalimentary automatisms in temporal seizures

Temporal features

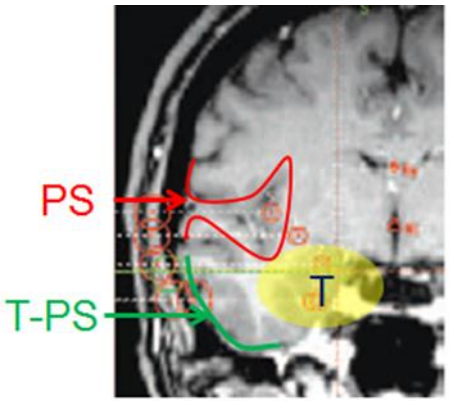
Consideration of automotor seizure

- More common are generally considered to originate from the temporal lobe
- Might also originate from frontal lobe, insula and operculum area or absence seizure
- Can occur early , mid or late of the whole seizure

TABLE 5. *Distribution of the late ictal signs according to the electrophysiologic subtypes*

Late ictal features	Medial 24 (%)	Medial–Lateral 18 (%)	Lateral 13 (%)	Degree of significance
Late oroalimentary automatisms	14 (58.3)	4 (22.2)	2 (15.4)	$p = 0.012^a$
Late upper-limb elementary automatisms	14 (58.3)	7 (38.9)	2 (15.4)	$p = 0.039^a$
Late vocalization	5 (20.8)	0	2 (15.4)	$p = 0.12$
Late verbal automatisms	3 (12.5)	2 (11.1)	0	$p = 0.40$
Late upper-limb tonic posturing	6 (25)	5 (27.8)	1 (7.7)	$p = 0.41$
Late head and/or eyes deviation	8 (33.3)	6 (33.3)	3 (23.1)	$p = 0.86$
Late dysphasia	6 (25)	3 (16.7)	0	$p = 0.14$

[a]Significant.



Maillard L, et al. Epilepsia 2004

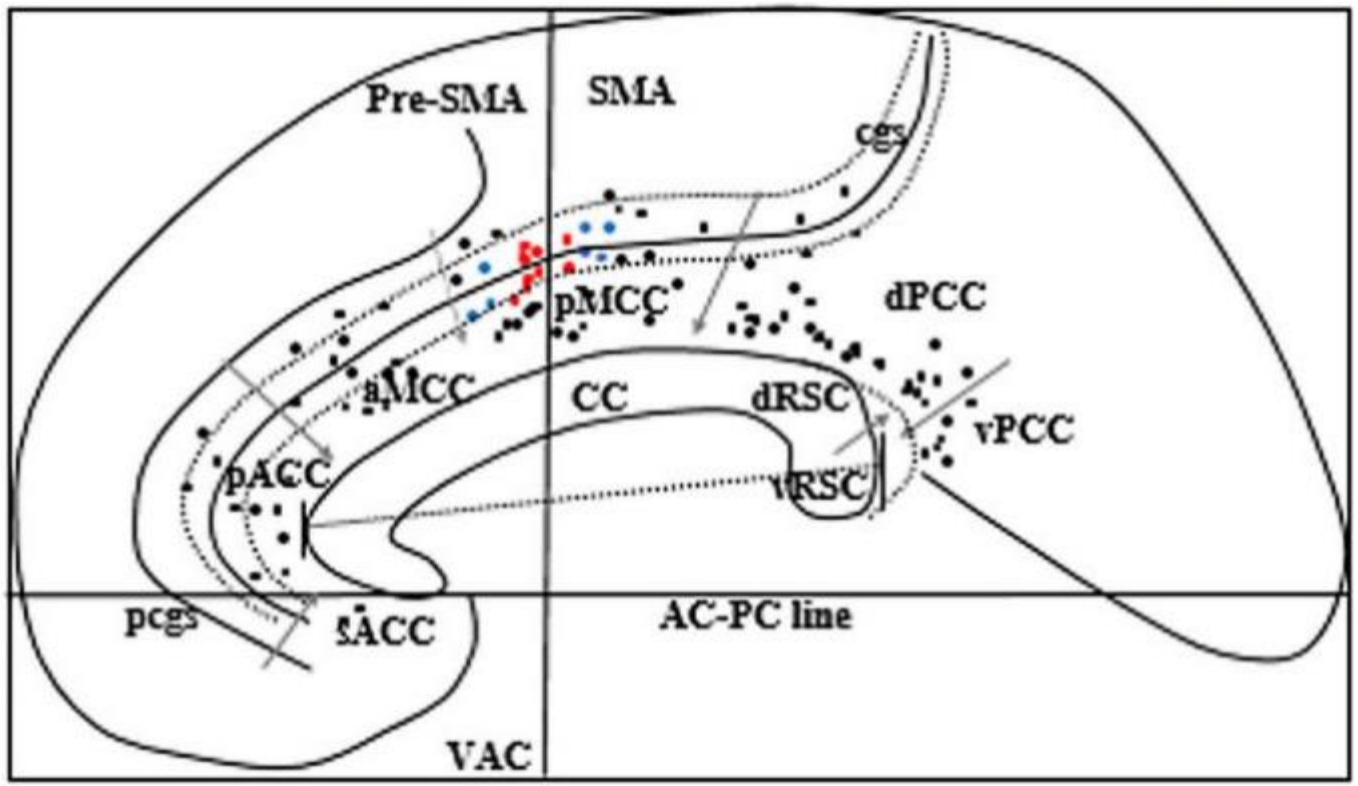
frontiers in Human Neuroscience

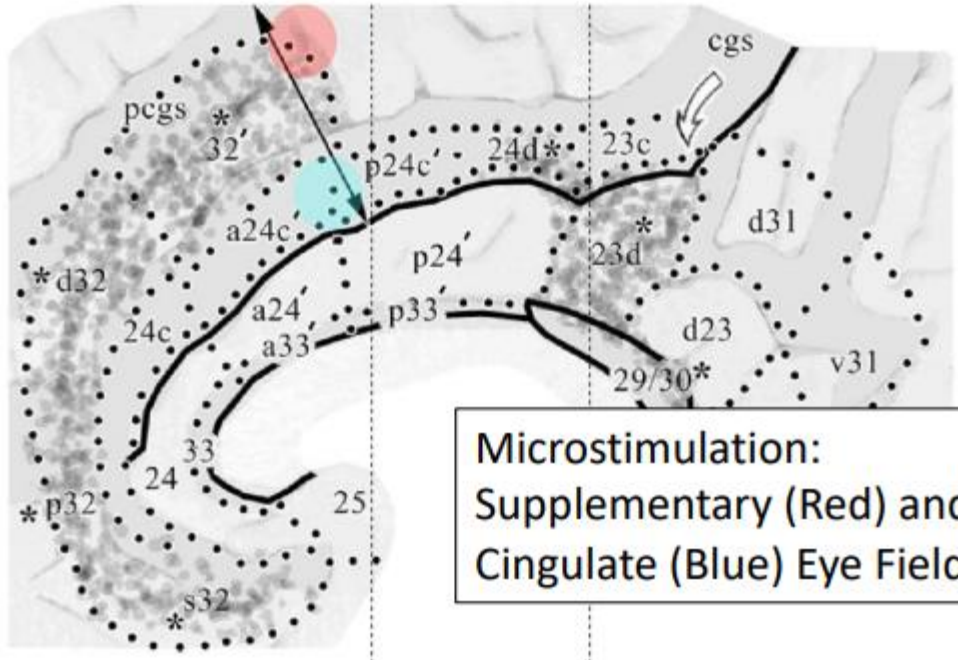
Front Hum Neurosci. 2022; 16: 815749.
Published online 2022 Feb 24. doi: 10.3389/fnhum.2022.815749

PMCID: PMC8909638
PMID: 35280209

Investigating the Precise Localization of the Grasping Action in the Mid-Cingulate Cortex and Future Directions

Zebunnessa Rahman,[1,2,*] Nicholas W. G. Murray,[1] Jacint Sala-Padró,[1] Melissa Bartley,[1] Mark Dexter,[1,2] Victor S. C. Fung,[1,2] Neil Mahant,[1] Andrew Fabian Bleasel,[1,2] and Chong H. Wong[1,2]

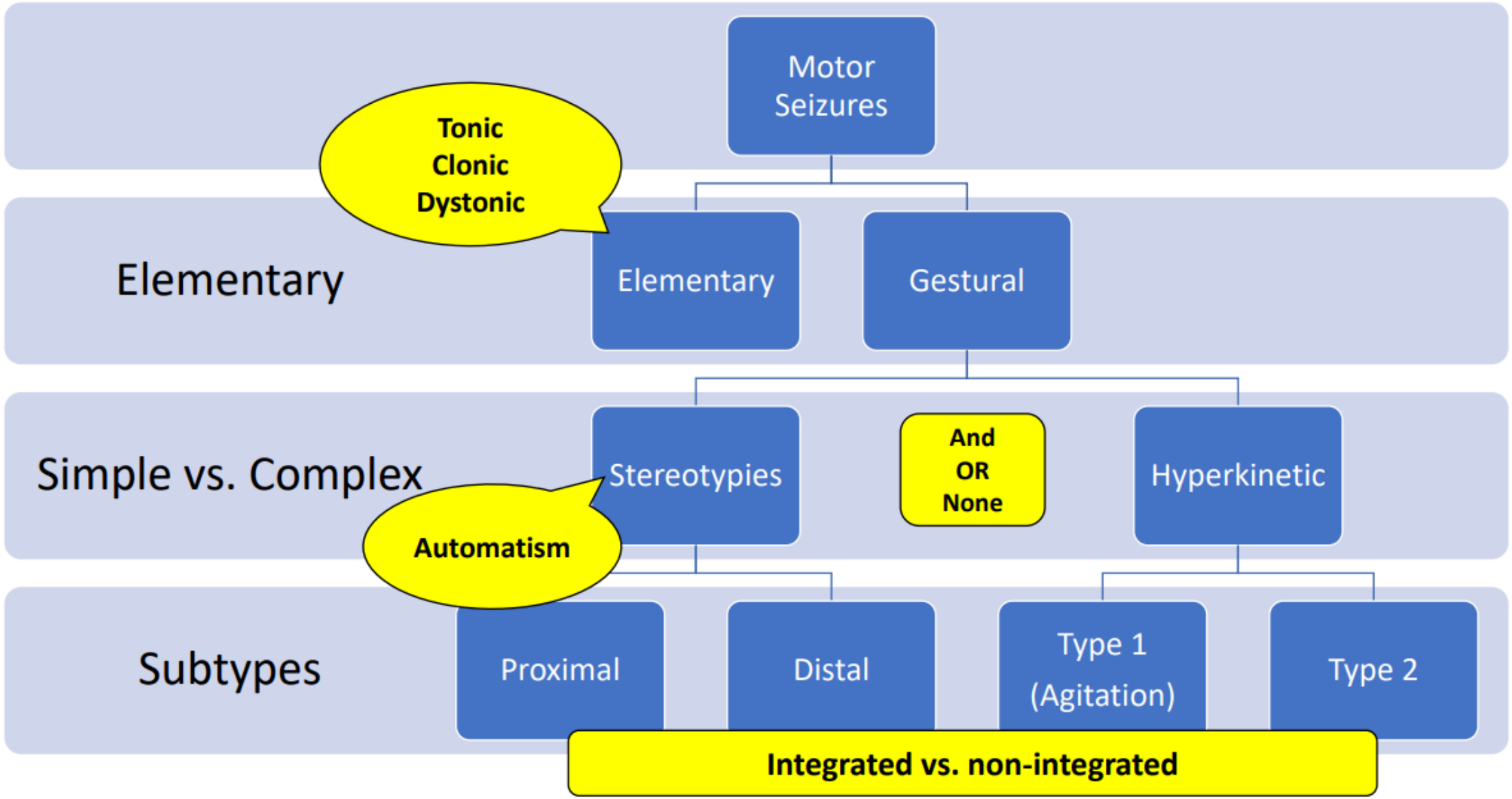




Microstimulation: Supplementary (Red) and Cingulate (Blue) Eye Fields

Brent A Vogt, Human Nervous System, 3rd Edition, Jurgen Mai, George Paxinos, p923, 2012

Motor Semiology - Classification



Courtesy: Prof. Lim Kheng Seang

Summary

- The elements of ictal semiology strongly suggests the seizure onset and propagation
- Carefully analyze semiology step by step from the initial symptom/sign until the end
  - Describe the event
  - Terminology
  - Localization/ Lateralization
  - Correlate with other investigations (EEG, MRI)
- Then you will understand epilepsy the underlying epileptic network

EPILEPSY SOCIETY
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Asian Epilepsy Academy
(ASEPA)
Asian and Oceanian Commission
International League Against Epilepsy

DIXI medical

ASEPA SEEG Workshop & DIXI SEEG Course

30th Oct – 3rd Nov 2024

80 Participants

Precongress DIXI SEEG Course

30th Oct – 1st Nov 2024

"The Fundamentals of SEEG "

Course Director: Prof. Patrick Chauvel

ASEPA SEEG Workshop

2nd – 3rd Nov 2024

"Workshop focusing on temporal and insuloperisylvian epilepsy "

Course Director: Prof. Lim Kheng Seang, Prof. Andrew Bleasel, Prof. Chong Wong and ASEPA faculties

Local organizing committees: Assoc.Prof. Chusak Limotai, Assist.Prof. Piradee Suwanpakdee and EST faculties

E-mail: Epilepsy09@gmail.com

Montien Hotel, Bangkok Thailand

Registration fee,
4.5 days: 600 USD