



#### Martedì 26 ottobre

**Moderatori:** *Francesco Pisani* (Messina) – *Rosalia Silvestri* (Messina)

# Manifestazioni motorie

### Giancarlo Di Gennaro

### CORSO VIDEO EEG LICE 3° EDIZIONE CATANIA, 24-27 OTTOBRE 2021



#### LE CRISI E LE EPILESSIE DEL LOBO FRONTALE

# Outline

- Anatomy/Physiology
- From clinical semiology to localization
- Lateralizing value of motor signs in FLE
- Unilateral clonic seizures
- Epileptic negative myoclonus
- Unilateral tonic seizures



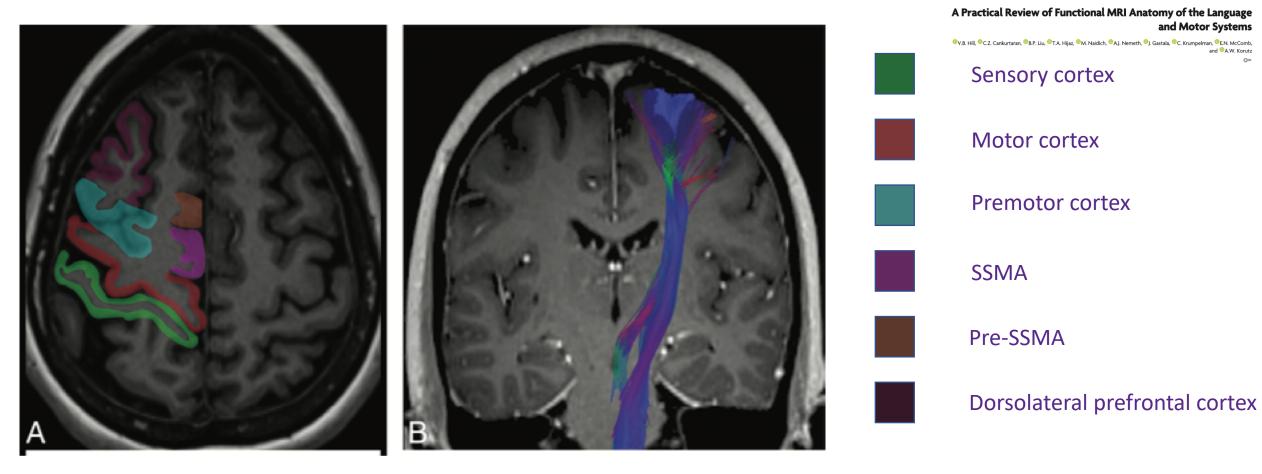
- Bilateral tonic seizures (SSMA, insula)
- Hand posture
- Eye and head deviation
- Rotatory seizures





# Anatomy and physiology (1)

#### Frontal lobe motor areas





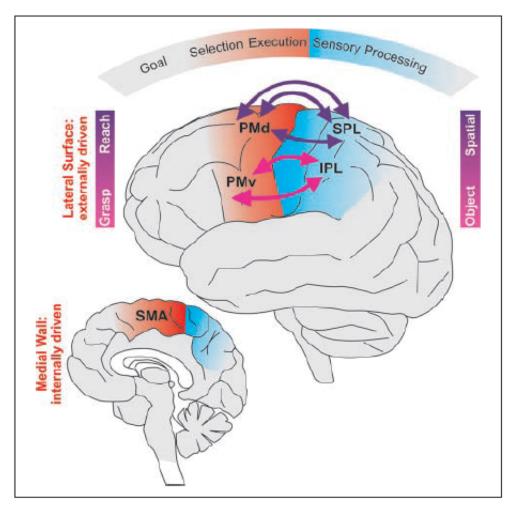


Published June 13, 2019 as 10.3174/ajnr.A6089

REVIEW ARTICLE

# Anatomy and physiology (2)

#### Parieto-premotor connections



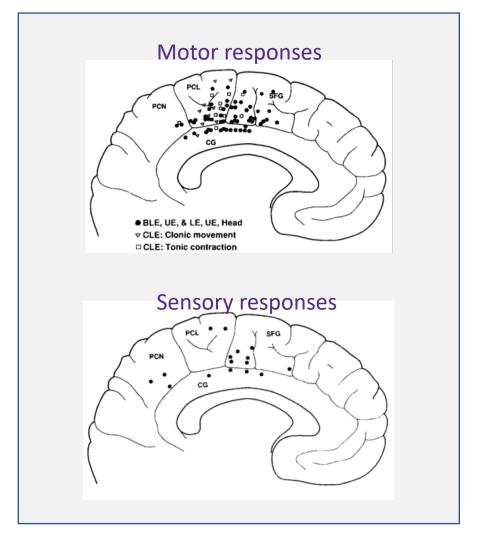
Neurological Principles and Rehabilitation of Action Disorders: Computation, Anatomy, and Physiology (CAP) Model Neurorehabilitation and Neural Repair Supplement to 25(5) 65-205 © The Author(s) 2011 Reprints and permission: http://www. apepub.com/journals/Permissions.nav DOI: 10.1177/154568311410940 http://nrs.agepub.com

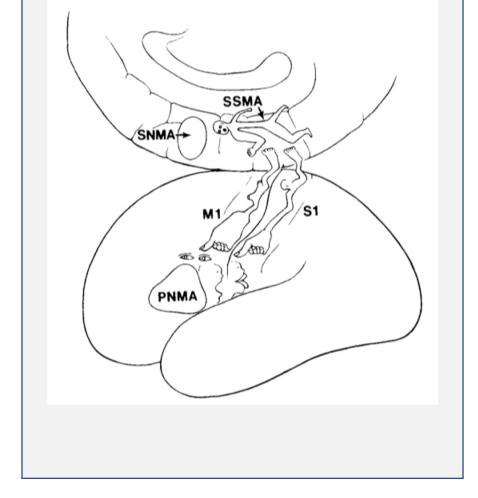
Scott H. Frey, PhD<sup>1</sup>, Leonardo Fogassi, PhD<sup>2</sup>, Scott Grafton, MD<sup>3</sup>, Nathalie Picard, PhD<sup>4</sup>, John C. Rothwell, PhD<sup>5</sup>, Nicolas Schweighofer, PhD<sup>6</sup>, Maurizio Corbetta, MD<sup>7</sup>, and Susan M. Fitzpatrick, PhD<sup>7,8</sup>





# Anatomy and physiology (SSMA homunculus) (3)



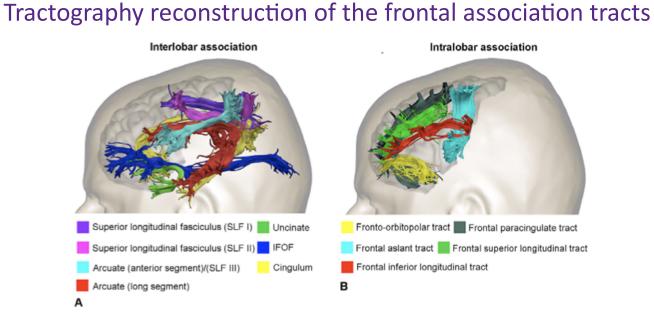




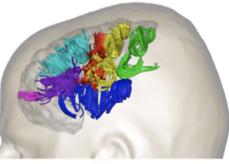




# **Anatomy** and physiology (4)



#### Short U-shaped fibers



Perirolandic U-fibers Precentral sulcus U-fibers Fronto-insular tracts Inferior frontal sulcus U-fibers

C

Fronto-marginal tract 📃 Superior frontal sulcus U-fibers



Handbook of Clinical Neurology Volume 163, 2019, Pages 95-122

Chapter 6 - The anatomy of the human frontal lobe

#### Marco Catani 온 🛛

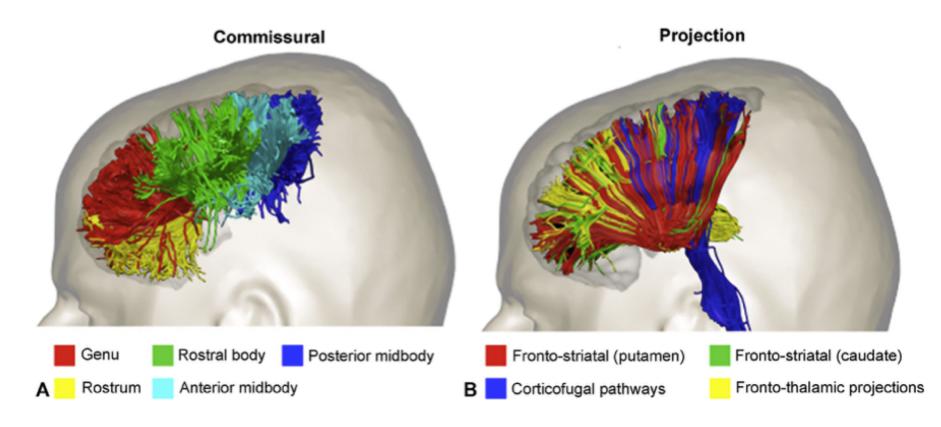
NatBrainLab, Department of Forensic and Neurodevelopmental Sciences, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, United Kingdom





# **Anatomy** and physiology (5)

Diagram of the main frontal lobe regions and their association and projection pathways.





Handbook of Clinical Neurology Volume 163, 2019, Pages 95-122

Chapter 6 - The anatomy of the human frontal lobe

Marco Catani 🙁 🖾

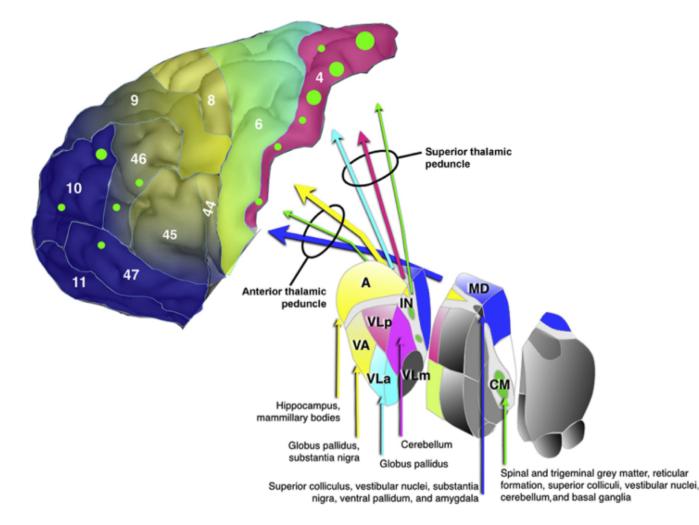
NatBrainLab, Department of Forensic and Neurodevelopmental Sciences, Institute of Psychiatry, Psychology and Neuroscience, King's College London, London, United Kingdom





# Anatomy and physiology (6)

Diagram of the principal thalamic nuclei, their afferent fibres, and their projections to the frontal cortex



Handbook Neur VIER Volume 163, 20

Handbook of Clinical Neurology Volume 163, 2019, Pages 95-122

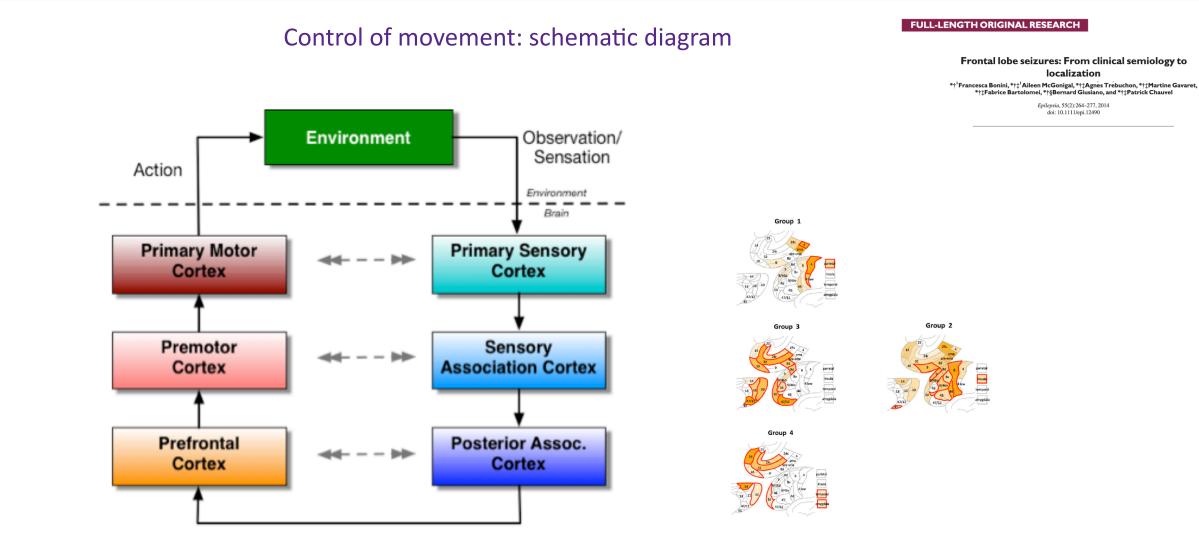
Chapter 6 - The anatomy of the human frontal lobe

Marco Catani 옷 🛛

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# **Anatomy and physiology** (7)







# The Epileptogenic Zone and AEC correlations (1)

frontiers in Human Neuroscience



High-frequency oscillations in epilepsy and surgical outcome. A meta-analysis

Yvonne Höller<sup>1+</sup>, Raoul Kutil<sup>2</sup>, Lukas Klaffenböck<sup>2</sup>, Aljoscha Thomschewski<sup>1</sup>, Peter M. Höller<sup>1</sup>, Arne C. Bathke<sup>2</sup>, Julia Jacobs<sup>2</sup>, Alexandra C. Taylor<sup>1</sup>, Raffaele Nardone<sup>1,4</sup> and Eugen Trinka<sup>1</sup>

• Ictal motor signs often do not necessarely coincide with the function

Motor signs are frequent in seizures involving frontal cortical areas

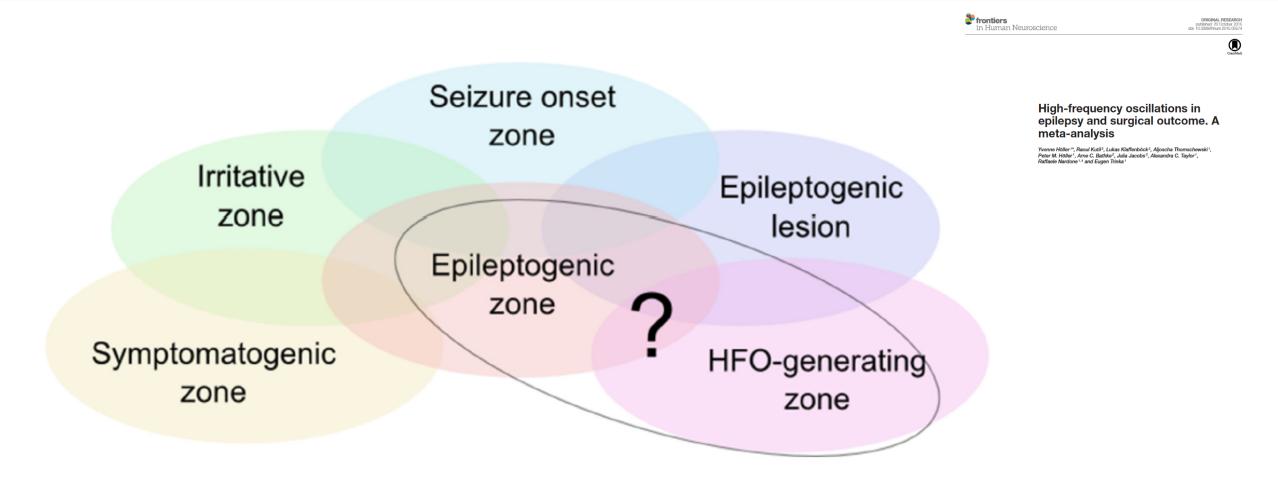
- Ictal semiology reflects the origin and 3D propagation of the discharge to areas both close to and remote from its origin
- Seizure onset zone often do not coincide with symptomatogenic zone
- Do not think in terms of a «naval battle» but in terms of AEC correlations



ullet



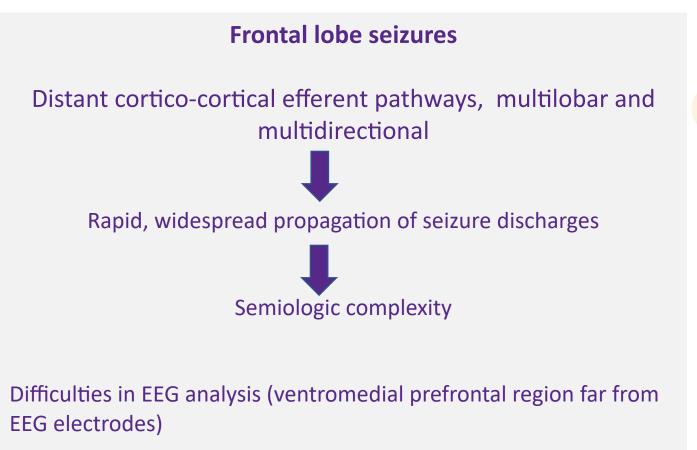
# The Epileptogenic Zone and AEC correlations (1)

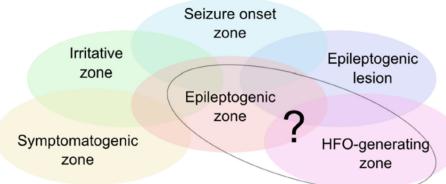






# The Epileptogenic Zone and AEC correlations (2)

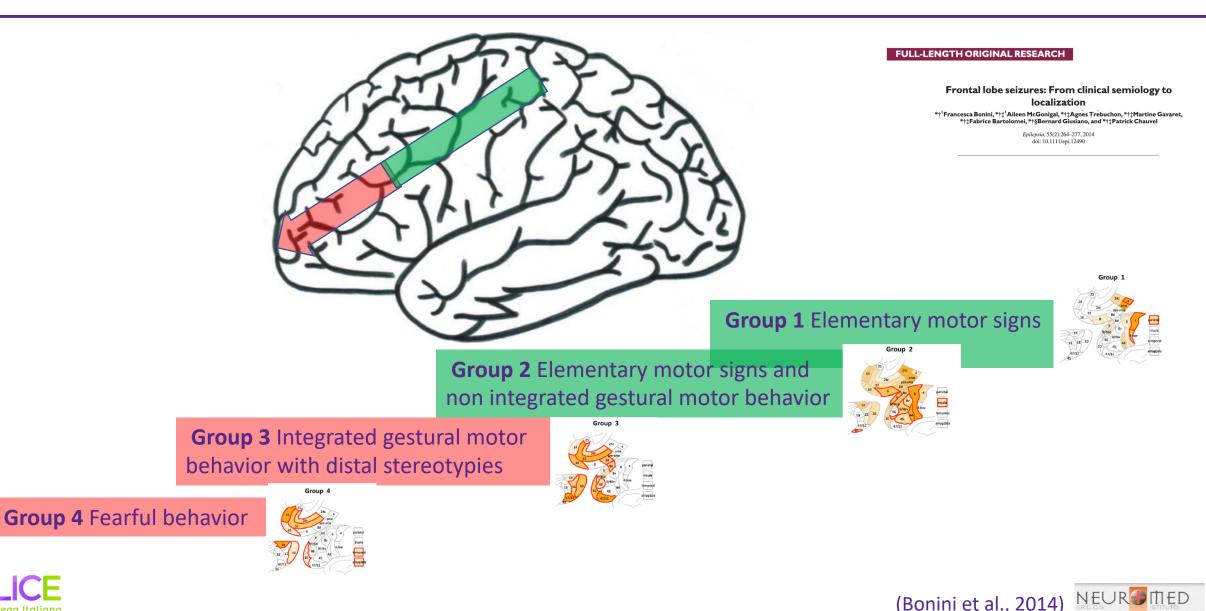






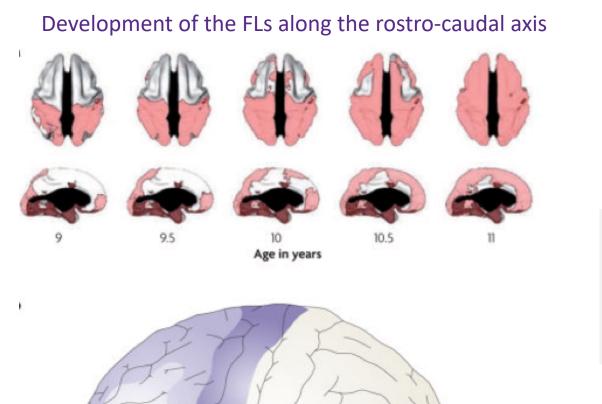


### Frontal lobe seizures: From clinical semiology to localization (1)





### Frontal lobe seizures: From clinical semiology to localization (2)



Nat Rev Neurosci. 2009 September ; 10(9): 659-669. doi:10.1038/nrn2667.

#### Is the rostro-caudal axis of the frontal lobe hierarchical?

David Badre\*.\* and Mark D'Esposito<sup>||.§</sup> 'Department of Cognitive & Linguistic Sciences, Brown University, Providence, Rhode Island 02912, USA <sup>‡</sup>Department of Psychology, Brown University, Providence, Rhode Island 02912, USA <sup>§</sup>Helen Wills Neuroscience Institute, University of California, Berkeley 94720, USA <sup>II</sup>Department of Psychology, University of California, Berkeley 94720, USA

Temporal differentiation in maturation Functional differences along the rostrocaudal axis.





# Lateralization value of ictal signs

Brain (2001), 124, 1683-1700

Table 2 Lateralizing ictal and postictal symptoms in patients with temporal lobe or extratemporal epilepsy					_ INVITED REVIEW	
Symptom	Location of the	Specificity	Frequency	References	Presurgical evaluation of epilepsy	
	epileptogenic zone				Felix Rosenow <sup>1</sup> and Hans Lüders <sup>2</sup>	
Forced head-version (<10 s) before	Contralateral	>90%	TLE 35%	Wyllie et al., 1986		
secondary generalization			ETE 40%	Kernan et al., 1993		
				Bleasel et al., 1997		
	~			Chee et al., 1993		
Unilateral ictal dystonia	Contralateral	90–100%	TLE 35 %	Kotagal et al., 1989		
			ETE 20%	Steinhoff et al., 1998 Bleasel et al., 1997		
Ictal speech	Non-dominant	>80%	10-20%	Chee et al., $1997$		
Preserved consciousness during	Non-dominant	100%	Rare, 5%	Ebner <i>et al.</i> , 1995		
ictal automatisms		100%	10a0, 570	Loner er un, 1990		
Postictal dysphasia	Dominant	>80%	20% (depends on the	Steinhoff et al., 1998		
			testing)	-		
				Chee et al., 1993		
Postictal nosewiping	Ipsilateral	80–90%	TLE 40–50%	Geyer et al., 1999		
			FLE 10%	Hirsch et al., 1998		
** **	* ··· ·	000	D 150	Leutmezer et al., 1998		
Unilateral eyeblinking	Ipsilateral	80%	Rare, 1.5%	Benbadis et al., 1996		
Ictal vomiting	Non-dominant	>90%	Rare	Kramer et al., 1988		
Figure of four	Contralateral	90%	65% of patients with sGTCSs	Kotagal et al., 2000		

ETE = extratemporal epilepsy; FLE = frontal lobe epilepsy; sGTCSs = secondary generalized tonic-clonic seizures; TLE = temporal lobe epilepsy.





# Lateralization value of ictal motor signs

Table 4: Important se lateralizating and/or loca	miologic features and their dizing value	Seizure Semiology: Value in Identifying Seizure Origin
Semiologic features	Lateralization and/or localization	Mohammed M.S. Jan, John P. Girvin
	IOCALIZA UOLI	Can. J. Neurol. Sci. 2008; 35: 22-30
3- Motor abnormalities		
Early nonforced head turn	Ipsilateral to seizure origin	
Late forced head turn	Contralateral to seizure origin	
Eye deviation	Contralateral to seizure origin	
Focal clonic jerking	Contralateral to seizure origin,	not always
	peri-rolandic	not always
Asymmetric clonic ending	Ipsilateral to seizure origin	contralateral
Dystonic limb posturing	Contralateral to seizure origin	contratateral
Tonic limb posturing	Contralateral to seizure origin	
Fencing posture	Contralateral frontal lobe (supplementary motor) seizures	
Figure of 4 sign	Contralateral to the extended limb, usually temporal lobe	
Unilateral ictal paresis	Contralateral to seizure origin	
Postictal Todd's paresis	Contralateral to seizure origin	





# Automatisms Unilateral clonic seizure

More or less regular, repeated, **short contractions of various muscle groups** (0.2-5 Hz).

Distal part of the extremities or face and most probably originate from the primary motor or premotor area. (Noachtar and Arnold, 2000)

In contrast, **in TLE**, **clonic seizures typically occur after automatisms** (spread of epileptic activity from the temporal to the frontal lobe).

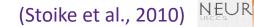
Positive predictive value of 92% for seizure onset in the **contralateral hemisphere** (Janszky et al., 2001).

Automotor seizure  $\rightarrow$  Clonic seizure of the left side of the face



A 44-year-old woman with right neocortical TLE of unknown origin. EEG seizure onsets were localised to the right posterior temporal region.





### Dx unilateral gestural automatisms







### Epileptic negative myoclonus/Epilepsia partialis continua

- Short (ca. 30-400 ms) phases of muscle atonia (clinically observed only during muscle contraction) (Tassinari and Gastaut, 1969).
- Focal negative motor phenomena frequently preceded by epileptiform discharges in the contralateral central region (20-30 ms before atonia).
- Origin in primary somatosensorimotor cortex (Ikeda et al., 2000), the premotor cortex (Baumgartner et al., 1996, Meletti et al., 2000) and the postcentral cortex (Noachtar et al., 1997)

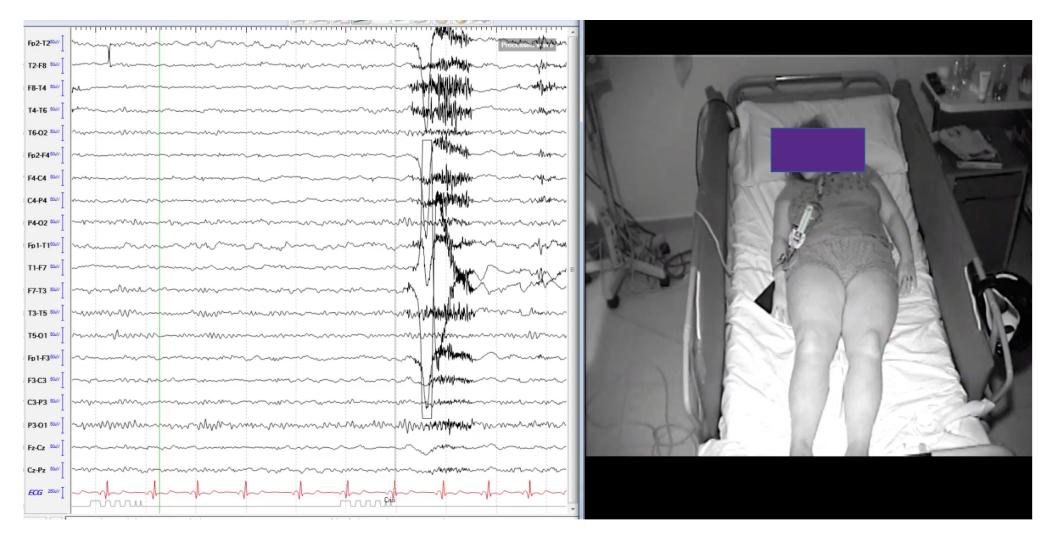








### Unilateral tonic seizure+ head/body rotation (2)







### Bilateral tonic facial contraction (ictal pouting or «chapeau du gendarme») (1)

- Symmetrical and sustained (> 5 s) lowering of labial commissures with contraction of chin, mimicking an expression of fear, disgust, or menace
- Early symptom in frontal seizures



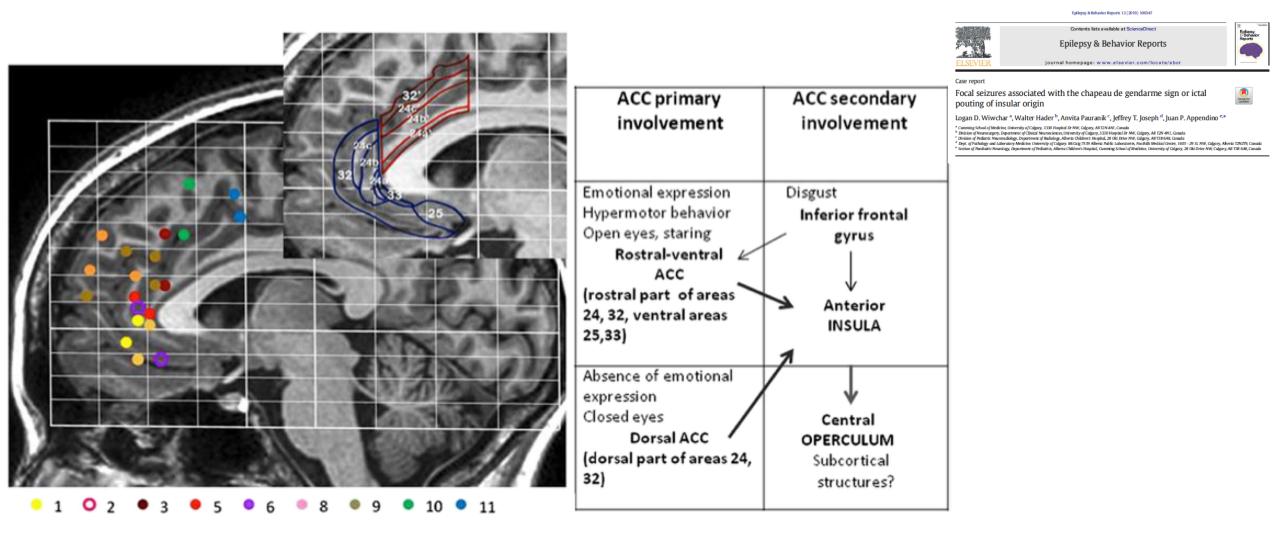
- PET-FDG hypometabolism include both the ٠ anterior cingulate cortex and lateral cortex including the anterior insula in all cases.







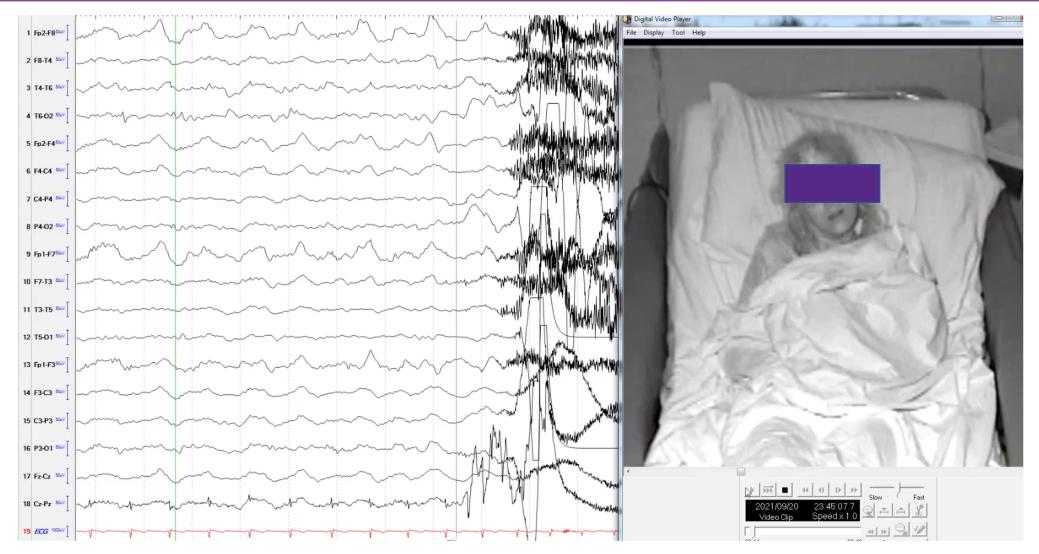
### Bilateral tonic facial contraction (ictal pouting or «chapeau du gendarme») (2)







### Dx vs paroxysmal hypnogenic/non kinesigenic dyskinesia (1)







### Dx vs paroxysmal hypnogenic/non kinesigenic dyskinesia (2)

Feature	PKD	PNKD	PED	PHD
Age at onset (years)	5–15 (range: 0.5–33)	8 (range: 2.5–79)	5 (range: 2–30)	Adolescence (range: 2–47)
Sex (male:female ratio)	4:1 (even up to 8:1)	2:1	2:3	1:1
Predominant movement	Dystonia >> chorea, ballism or a combination	Dystonia and chorea	Dystonia	Dystonia, chorea or ballism
Duration	<1 minute (seconds–5 min)	<1 hour (10 min–12 h)	2 min–2 h	30–45 seconds
Frequency	1–20 attacks per day	1 per week (3 per day)	1 per day–2 per month	5 per night–5 per year
Aura	70%	80% familial cases	No	-
Distribution	${\sf Limbs} >> {\sf trunk} \text{ and face}$	Face, trunk and limbs	Feet >> hemidystonia and hands	Limbs >> trunk and face
Triggers	Abrupt movement, an increase of speed, amplitude or strength. Light, sound and vestibular stimulation, startle, hyperventilation and stress	At rest; alcohol, coffee, tea, cola, tobacco, emotional excitement, hunger, fever, concentration and fatigue	Prolonged or maintained exercise; vibration, passive movements, electrical stimulation of the nerves, stress, hunger, sleep deprivation, cold exposure	Non-REM sleep
Exacerbating factors	Puberty, depression and anxiety	Menstruation, ovulation and postpartum	-	-
Response to antiepileptic drugs	+++	+/-	++ ('runner's dystonia')	+++
Evolution	Remission: pregnancy and adulthood	Variable; remission: pregnancy, sleep, cold water, the elderly, menopause	Variable; remission: sensory or motor 'tricks' ('runner's dystonia')	-





### Bilateral tonic seizure (SSMA vs Extra-SSMA)

#### SSMA group

- Unilateral/ bilateral asymmetrical tonic limb posturing (extended upper extremity was contralateral and flexed upper extremity was ipsilateral)
- Awareness during seizures was preserved
- Often aura and more than half of the auras were somatosensory (seven patients). Side of somatosensory aura was contralateral
- Bilateral tonic facial contraction (BTFC) and speech arrest are frequent

#### Extra SSMA group

- Bilateral symmetrical tonic limb posturing
- Impaired awareness during seizures

# SSMA group 22 ptsExtra-SSMA group 10 pts4 dlF or preF

2 insula

3 P



SSMASSMAExtra-SSMABilateral asymmetricalBilateral tonic facial Bilateral symmetrical)<br/>(extension<br/>contralateral)contraction





# Bilateral tonic/dystonic seizure (insula $\rightarrow$ frontal lobe) (1)

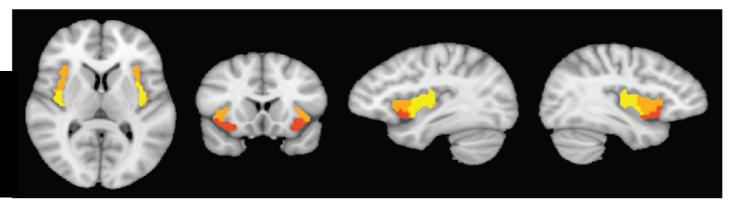
- Full awareness
- Laryngeal constriction
- Paresthesiae, often unpleasant, affecting large cutaneous territories
- 1. Evolution in hyperkinetic seizure
- 2. Evolution in Bilateral tonic/dystonic seizure

Isnard et al., 2004

Posterior antero-inferior  $\rightarrow$  TLE semiology Antero-superior  $\rightarrow$  FLE semiology Bilateral tonic/dystonic seizure R>L



Involvement of frontal cingulate gyrus and the posterior part of the mesial frontal cortex, in particular the SSMA (Proserpio et al., 2011)





Ryvlin et al., 2006



### Hand posture (FLE vs TLE) (1)

)	fist	cup	politician's fist	pincer	extended hand	pointing
	The second					Atta
humb MP	Flexed	Flexed	Flexed	Flexed	Extended	Extended
Thumb IP	Flexed	Extended	Extended	Extended	Extended	Extended
Index MP	Flexed	Flexed	Flexed	Flexed	Extended	Extended
Index IP	Flexed	Flexed	Flexed	Flexed	Extended	Extended
3-5 MP	Flexed	Flexed	Flexed	Ext/Flex	Extended	Flexed
3-5 IP	Flexed	Flexed	Flexed	Ext/Flex	Extended	Flexed

#### Hand Posture as Localizing Sign in Adult Focal Epileptic Seizures

RESEARCH ARTICLE

Isabella Ferando, MD, PhD <sup>0</sup>,<sup>1</sup> Jason R. Soss, MD,<sup>1</sup> Christopher Elder, MD,<sup>1,2</sup> Vishal Shah, MD,<sup>1,2</sup> Giorgio Lo Russo, MD,<sup>3</sup> Laura Tassi, MD,<sup>3</sup> Carlo Alberto Tassinari, MD,<sup>4,5</sup> and Jerome Engel Jr MD, PhD<sup>1,2,6,7,8</sup>

#### 79 pts, 489 seizures

- Seizure Disorder Center
   UCLA, Los Angeles (USA)
- C. Munari Epilepsy Surgery Center Niguarda Hospital, Milan, Italy



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### Hand posture (FLE vs TLE) (2)

#### 100% specificity and PPV in FLE



#### pointing



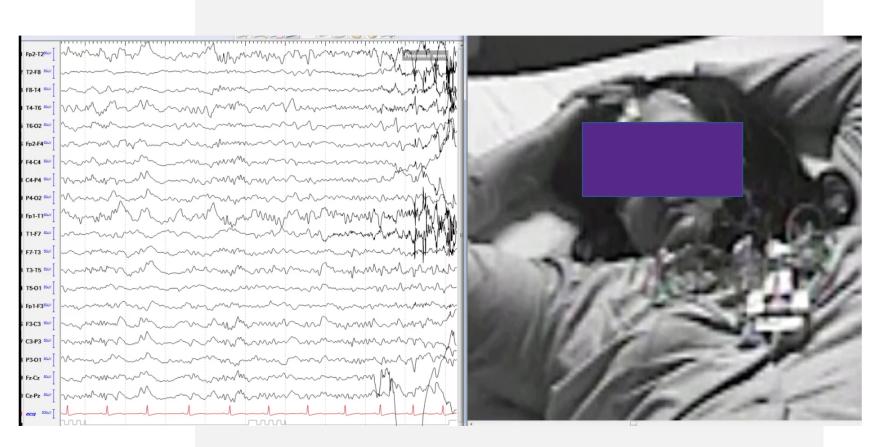






### Head and eye deviation (1)

- Versive seizures consist of a forced and involuntary head movement resulting in sustained unnatural positioning (Wyllie et al., 1986a), High specificity (>90%) of lateralisation for a contralateral seizure onset zone (Chee et al., 1993; Steinhoff et al., 1998), particularly when occurring immediately prior to generalisation (Kernan et al., 1993).
- IEpileptic activation of the frontal eye field contralateral to the side to which the eyes or head turn (Penfield and Jasper, 1954).





### Head and eye deviation (ipsilateral/contralateral) (2)

#### In Frontal Lobe Seizures



#### Ipsilateral

- early
- no associated clonic movements
- (frontopolar/orbitofrontal)

#### Contralateral

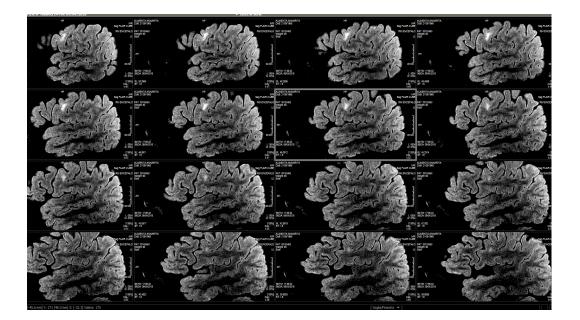
- Tardive
- Tonic unnatural
- Associated clonic moviments (FEF)

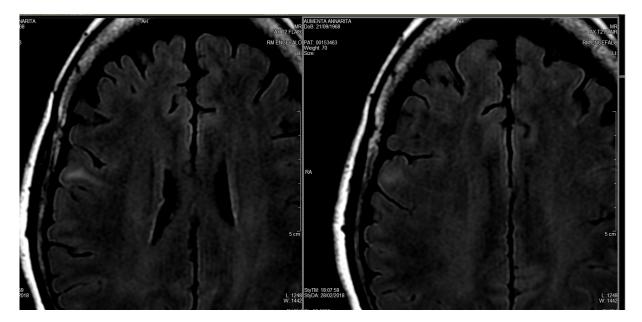
(Rheims, 2005)

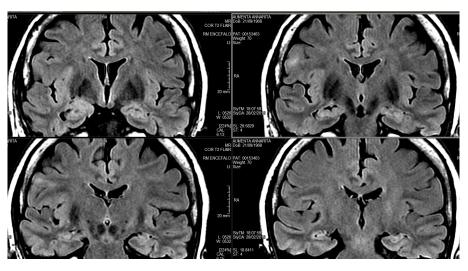




# Head and eye deviation (ipsilateral/contralateral) (2)



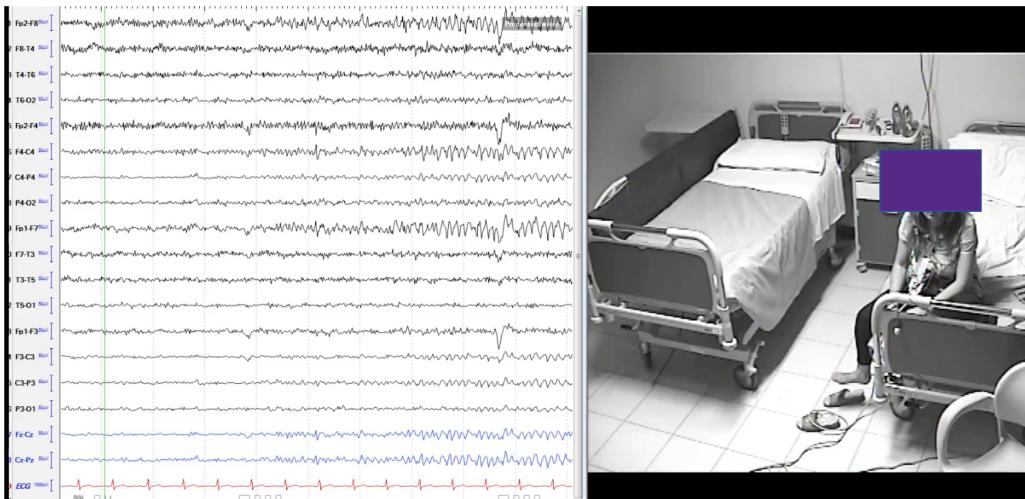








# Rotatory seizures (1)

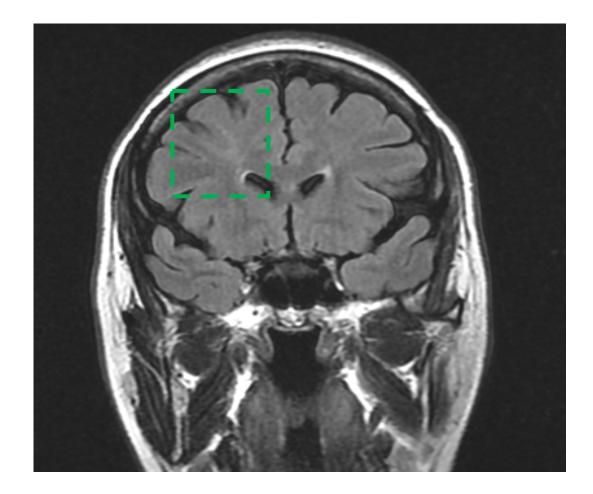


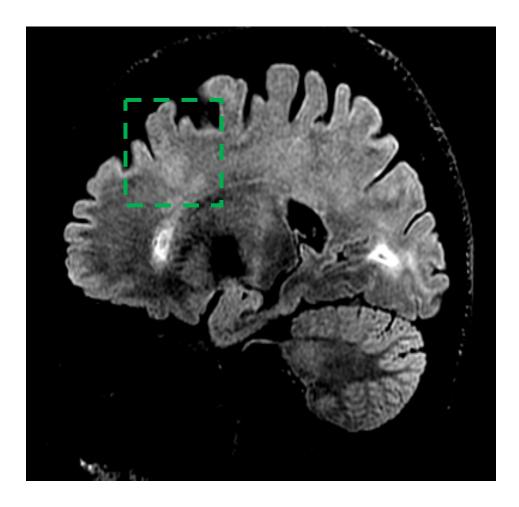
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5 F4-C4 50.V	
7 C4-P4 50.V	
8 P4-O2 500V	man man man and a second and as second and a
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8 Cz-Pz 50JV	~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~
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# Rotatory seizures (2)











Epilessia: Mattiana in crisi

#### Grazie per l'attenzione

Neurologi -Giancarlo Di Gennaro -Pier Paolo Quarato -Addolorata Mascia -Sara Casciato

**Neuropsichiatra infantile** -Alfredo D'Aniello

Neuropsicologa Psicoterapeuta -Liliana Grammaldo

Ingegnere biomedico -Luigi Pavone **Neurochirurghi** -Vincenzo Esposito -Roberta Morace

Neuroradiologo -Claudio Colonnese -Giovanni Grillea

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- Simona Fratini
- Maria Tedesco
- Valeria Bruno
- Giorgia Malinconico

Segreteria -Enrica Salvucci

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