Le Crisi e Le Epilessie del Lobo Frontale

VIDEO-SESSION

Giuseppe d'Orsi

Asbat BARLETTA-ANDRIA-TRANI

Centro per lo Studio e la Cura dell'Epilessia

U.O.C. Neurologia. Ospedale Bonomo, Andria

Catania 25 Ottobre 2021, Corso Video-EEG

♀, 17 anni

- Dall'età di 15 anni circa:
  - Episodi parossistici durante sonno (notturno)
  - Brevi e frequenti (più volte/die)
  - Movimenti ampi ai quattro arti, gemiti, deviazione del capo a destra e a sinistra
  - Rapida ripresa

Le Crisi e Le Epilessie del Lobo Frontale

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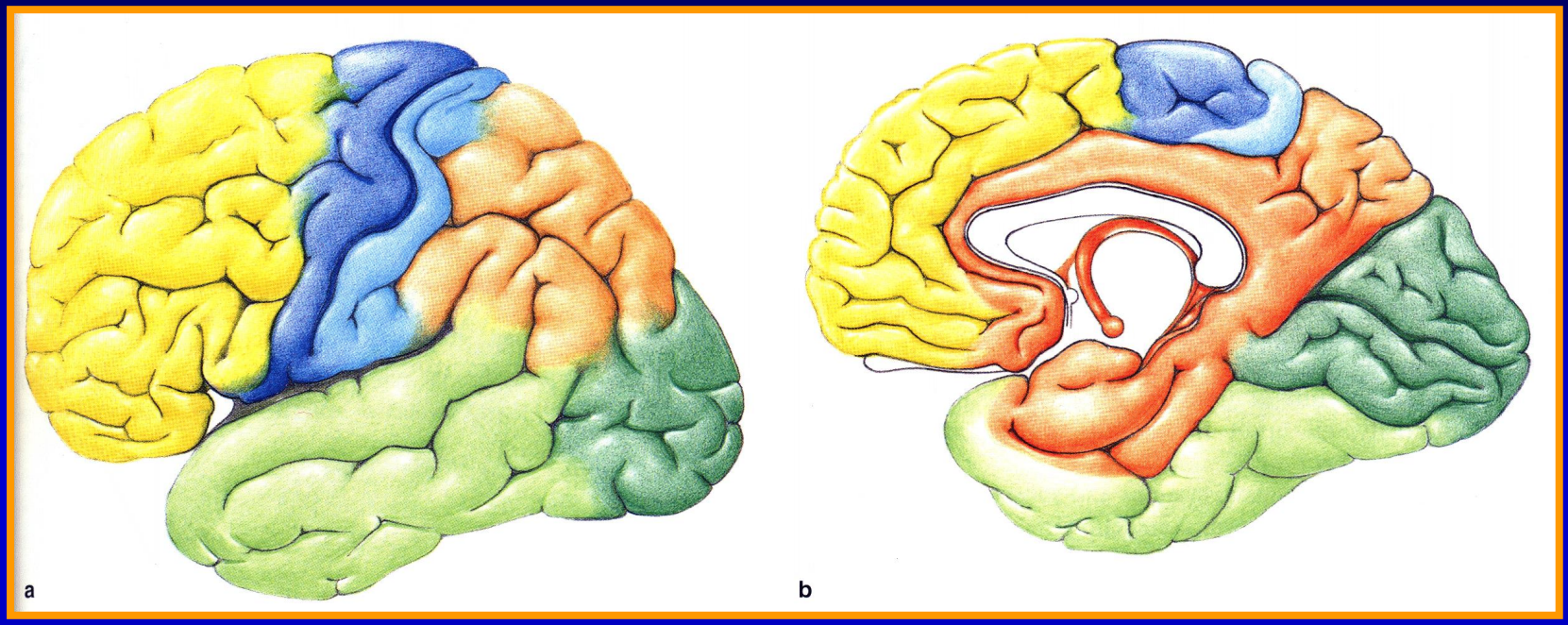
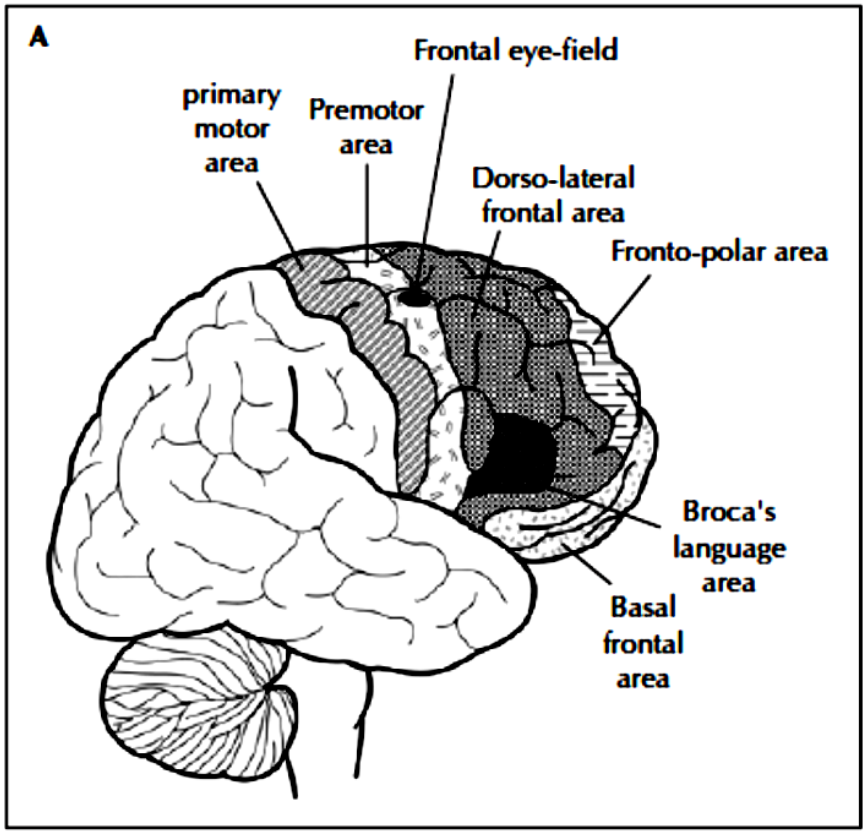


- Comprende 20-30% delle Epilessie Focali
- Non completamente studiata…
- Complessità del Lobo Frontale

HYPERMOTOR OR HYPERKINETIC SEIZURES

SEEG Studies



- Cortex cingulate cortex (82%)
- Orbitofrontal cortex (45%)
- Frontal polar cortex (27%)
- Mesial premotor cortex

(Rheims et al., 2008)

- Dorsolateral frontal cortex

(Tal et al, 2010)

HYPERMOTOR OR HYPERKINETIC SEIZURES

Extrafrontal Brain Regions

- Anterior Superior portion of insula (Ryvlin et al., 2006)
- Parietotemporal region (Nishibayashi et al., 2009)
- Temporal lobe epilepsy (.....)

HYPERMOTOR OR HYPERKINETIC SEIZURES IN TLE

SLEEP IN NEUROLOGICAL DISORDERS

Epileptic Nocturnal Wanderings with a Temporal Lobe Origin: A Stereo-Electroencephalographic Study

SLEEP, Vol. 25, No. 6, 2002

Lino Nobili, MD;[1,2] Stefano Francione, PhD;[1] Francesco Cardinale, MD;[1] and Giorgio Lo Russo, MD[1]

R. Mai • I. Sartori • S. Francione • L. Tassi • L. Castana • F. Cardinale • M. Cossu • A. Citterio N. Colombo • G. Lo Russo • L. Nobili

Sleep-related hyperkinetic seizures: always a frontal onset?

Neural networks underlying hyperkinetic seizures of ''temporal lobe'' origin

Epilepsy Research (2009) **86**, 200–208

L. Vaugier[a], S. Aubert[a], A. McGonigal[a,c], A. Trébuchon[c], M. Guye[a,b,c], M. Gavaret[a,b,c], J. Regis[b,c], P. Chauvel[a,b,c], F. Wendling[d,e], F. Bartolomei[a,b,c,*]

Hypermotor seizures in patients with temporal pole lesions

Lina Wang[a], Gregory C. Mathews[a,*], William O. Whetsell[b], Bassel Abou-Khalil[a]

Epilepsy Research (2008) **82**, 93–98

Temporal lobe origin is common in patients who have undergone epilepsy surgery for hypermotor seizures

Amir M. Arain [a,*], Nabil J. Azar [a], Andre H. Lagrange [a], Michael McLean [a], Pradumna Singh [a], Hasan Sonmezturk [a], Peter Konrad [b], Joseph Neimat [b], Bassel Abou-Khalil [a]

Epilepsy & Behavior 64 (2016) 57–61

Original Article

Sleep Medicine 12 (2011) S33–S38

Epileptic motor behaviors during sleep: Anatomo-electro-clinical features

P. Proserpio [a,b], M. Cossu [a], S. Francione [a], F. Gozzo [a], G. Lo Russo [a], R. Mai [a], A. Moscato [a], M. Schiariti [a], I. Sartori [a], L. Tassi [a], L. Nobili [a,b,*]

Hyperkinetic seizures in patients with temporal lobe epilepsy: Clinical features and outcome after temporal lobe resection

Epilepsia, 52(8):1439–1446, 2011

*Anke M. Staack, *Sofia Bilic, *Anne-Sophie Wendling, *Julia Scholly, *Uwe Kraus, *Karl Strobl, *Frédéric Bodin, †Josef Zentner, and *Bernhard J. Steinhoff

Sleep-related hyperkinetic seizures of temporal lobe origin

NEUROLOGY 2004;62:482–485

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

Complex motor behaviors in temporal lobe epilepsy

Abstract—Complex motor behaviors differing from typical automatisms were found in 12 of 502 patients with temporal lobe epilepsy. Movements involved proximal limb segments (6) or body axis (6) and were often preceded by auras and followed by automatisms. Seven of 12 patients are seizure free after surgery. The other 5 patients declined surgery.

NEUROLOGY 2005;65:1805–1807

M. Carreño, MD, PhD; A. Donaire, MD; M.A. Pérez Jiménez, MD, PhD; R. Agudo, MD; A. Quilez, MD; J. Rumià, MD; F. Villarejo, MD; N. Bargalló, MD, PhD; T. Boget, PsyD; T. Raspall, MD; L. Pintor, MD, PhD; and X. Setoain, MD, PhD

Disturbo del Movimento

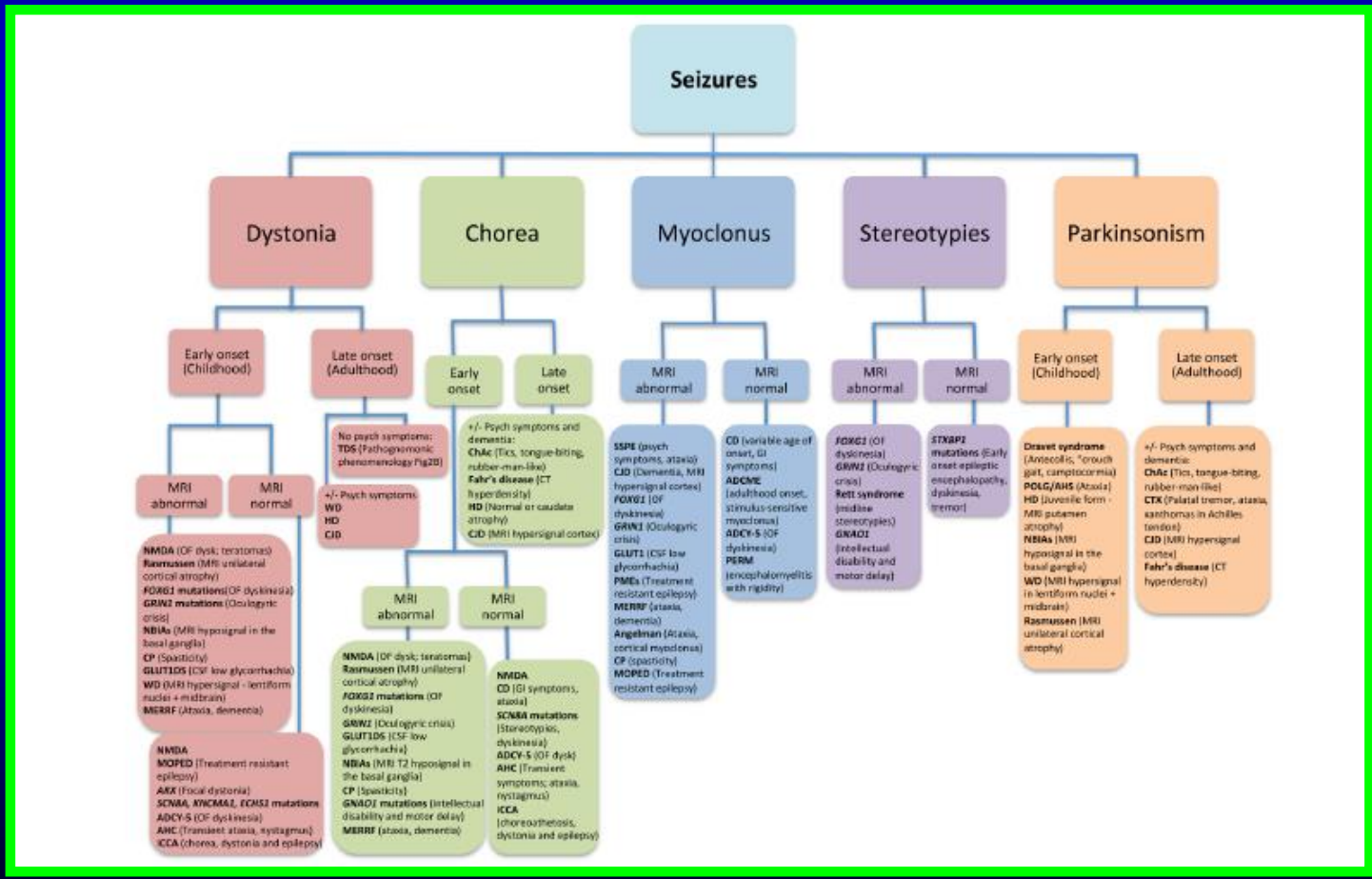
Altre Crisi Epilettiche ed Epilessie

Le Crisi e le Epilessie del Lobo Frontale

REVIEW *J Neurol Neurosurg Psychiatry* 2019;**90**:920–928. doi:10.1136/jnnp-2018-320039

Seizures and movement disorders: phenomenology, diagnostic challenges and therapeutic approaches

Maria Eliza Freitas,[1] Marta Ruiz-Lopez,[2] Josep Dalmau,[3] Roberto Erro,[4] Michael Privitera,[5] Danielle Andrade,[6] Alfonso Fasano[7]



Disturbo del Movimento

Altre Crisi Epilettiche ed Epilessie

Le Crisi e le Epilessie del Lobo Frontale

Manifestazioni Parossistiche NON Epilettiche

Psicogene

Psycogenic Non-Epileptic Seizures

Neurol Clin 29 (2011) 149–162
doi:10.1016/j.ncl.2010.10.005

Differentiating Frontal Lobe Epilepsy from Psychogenic Nonepileptic Seizures

W. Curt LaFrance Jr, MD, MPH[a,b,*], Selim R. Benbadis, MD[c,d]

Table 1
Behaviors to distinguish between psychogenic nonepileptic and epileptic seizures

Observation	PNES	ES
Situational onset	Common	Rare
Gradual onset	Common	Rare
Precipitated by stimuli (noise, light)	Occasional	Rare
Purposeful movements	Occasional	Very rare
Opisthotonus (*arc de cercle*)	Occasional	Very rare
Tongue biting (tip)	Occasional	Rare
Tongue biting (side)	Very Rare	Common
Prolonged ictal atonia	Occasional	Very rare
Vocalization during tonic-clonic phase	Occasional	Very rare
Reactivity during unconsciousness	Occasional	Very rare
Rapid postictal reorientation	Common	Unusual
Undulating motor activity	Common	Very rare
Asynchronous limb movements	Common	Rare
Rhythmical pelvic movements	Occasional	Rare
Side-to-side head shaking	Common	Rare
Ictal crying	Occasional	Very rare
Ictal stuttering	Occasional	Rare
Postictal whispering	Occasional	Not present
Closed mouth in tonic phase	Occasional	Very rare
Closed eyelids during seizure onset	Very common	Rare
Convulsion >2 min	Common	Very rare
Resisted lid opening	Common	Very rare
Pupillary light reflex	Usually retained	Commonly absent
Cyanosis	Rare	Common
Ictal grasping	Rare	Occurs in FLE and TLE
Postictal nose rubbing	Not present	Can occur in TLE
Stertorous breathing postictally	Not present	Common
Self-injury	May be present (especially excoriations)	May be present (especially lacerations)
Incontinence	May be present	May be present

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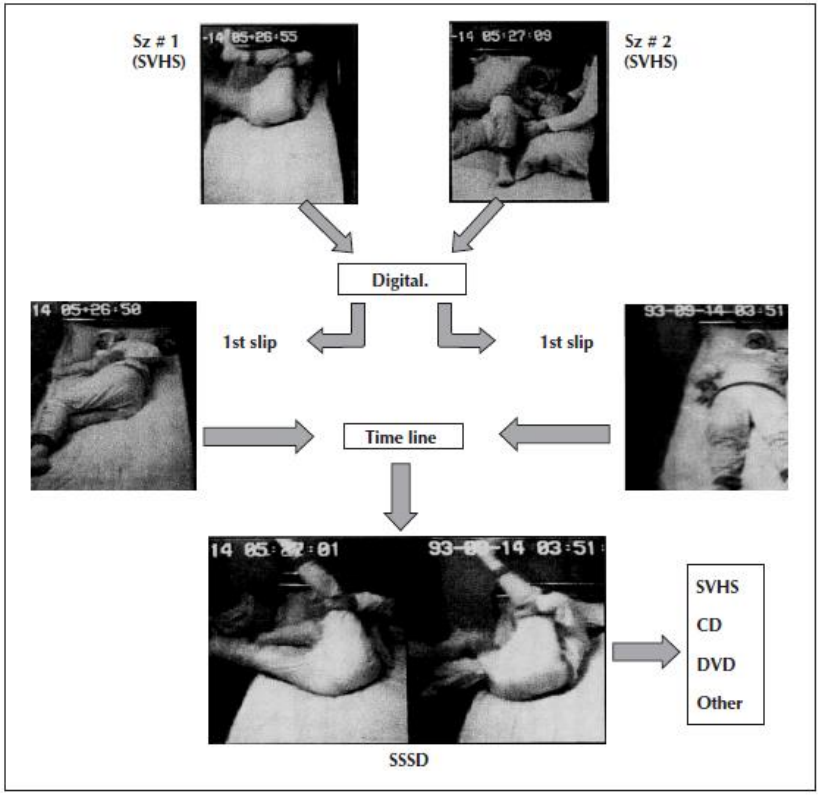
W. Curt LaFrance Jr, MD, MPH[a,b,*], Selim R. Benbadis, MD[c,d]

Original article

Epileptic Disord 2004; 6: 27-30

Split-screen synchronized display. A useful video-EEG technique for studying paroxysmal phenomena

Paolo Tinuper, Carlo Grassi, Francesca Bisulli, Federica Provini, Giuseppe Plazzi, Elena Zoni, Elio Lugaresi



Review article

Epileptic Disord 2019; 21 (1): 1-29

Classification of paroxysmal events and the four-dimensional epilepsy classification system

Hans Lüders [1], Guadalupe F
Naoki Akamatsu [3], Shahram
Christoph Baumgartner [6], Se
Andrew Bleasel [8], Adriana B
Mar Carreño [11], Michael De
Naiara García Losarcos [14], H
Shirin Jamal-Omidi [17], Giri K
Andrés M. Kanner [19], Susan
Samden Lhatoo [22], Shih Hui
Jayanti Mani [25], Riki Matsumoto
Soheyl Noachtar [28], André Palmini [29], Jun Park [30],
ele [33],
mde Boas [38],

Brain (2001), **124**, 1683–1700

Review article

Epileptic Disord 2004; 6: 223-39

Frontal lobe epilepsy

Christoph Kellinghaus, MD[1,2], Hans O. Lüders, MD, PhD[1]

[1] Dept. of Neurology, The Cleveland Clinic Foundation, Cleveland, Ohio, USA
[2] Dept. of Neurology, University of Münster, Germany

Received January 30, 2004; Accepted September 13, 2004

INVITED REVIEW

Presurgical evaluation of epilepsy

Felix Rosenow[1] and Hans Lüders[2]

SEMEIOLOGIA CRITICA

CRISI MOTORIE

- **SIMPLE:** Motor movements are relatively «simple», unnatural, and consist of movements similar to movements elicited by electrical stimulation of the primary motor areas (Brodmann areas 4 and 6).
- **COMPLEX:** Motor movements are relatively «complex» and simulate natural movement, except that they are inappropriate for the situation

Luders et al, 1998. Luders et al, 2019.

SEMEIOLOGIA CRITICA

SIMPLE MOTOR SEIZURES

- MYOCLONIC SEIZURES
- CLONIC SEIZURES
- TONIC SEIZURES
- TONIC-CLONIC SEIZURES
- EPILEPTIC SPASMS
- VERSIVE SEIZURES

Luders et al, Epilepsia 1998

Luders et al, Epileptic Disorders 2019

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COMPLEX MOTOR SEIZURES

- AUTOMOTOR SEIZURES
- HYPERMOTOR SEIZURES
- GELASTIC SEIZURES

Luders et al, Epilepsia 1998

Luders et al, Epileptic Disorders 2019

SEMEIOLOGIA
EPILESSIA DEL LOBO TEMPORALE

- Sensazione iniziale tipica (AURA)
- Graduale perdita di consapevolezza

  (con o senza automatismi)
- Ripresa graduale
- Confusione post-critica

SEMIOLOGIA CRITICA

SIMPLE MOTOR SEIZURES

- MYOCLONIC SEIZURES
- CLONIC SEIZURES
- TONIC SEIZURES
- TONIC-CLONIC SEIZURES
- EPILEPTIC SPASMS
- VERSIVE SEIZURES

COMPLEX MOTOR SEIZURES

- **AUTOMOTOR SEIZURES**
- HYPERMOTOR SEIZURES
- GELASTIC SEIZURES

Luders et al, Epilepsia 1998

Luders et al, Epileptic Disorders 2019

CRISI AUTOMOTORIE

- Repetitive, stereotyped, semipurpoself motor behaviors, involving primarily the distal limbs (fumbling, picking, gesticulating movements), mouth and tongue (mastication, swallowing, lip smacking, blowing, whistling, kissing).
- Awareness is generally impaired (except nondominant temporal lobe)
- Homogeneous perseverative automatisms, complex gestures, and upper limb automatisms prolonged in duration

Luders et al, 1998; Blume et al. 2001

AUTOMOTOR SEIZURES

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- Awareness is generally impaired (except nondominant temporal lobe)
- Homogeneous perseverative automatisms, complex gestures, and upper limb automatisms prolonged in duration

Luders et al, 1998; Blume et al. 2001

Ictal hyperperfusion patterns according to the progression of temporal lobe seizures

NEUROLOGY 2002;58:373–380

Won Chul Shin, MD; and S. Eun Kim, MD

Dystonia, and regional cerebral blood flow changes in temporal lobe seizures

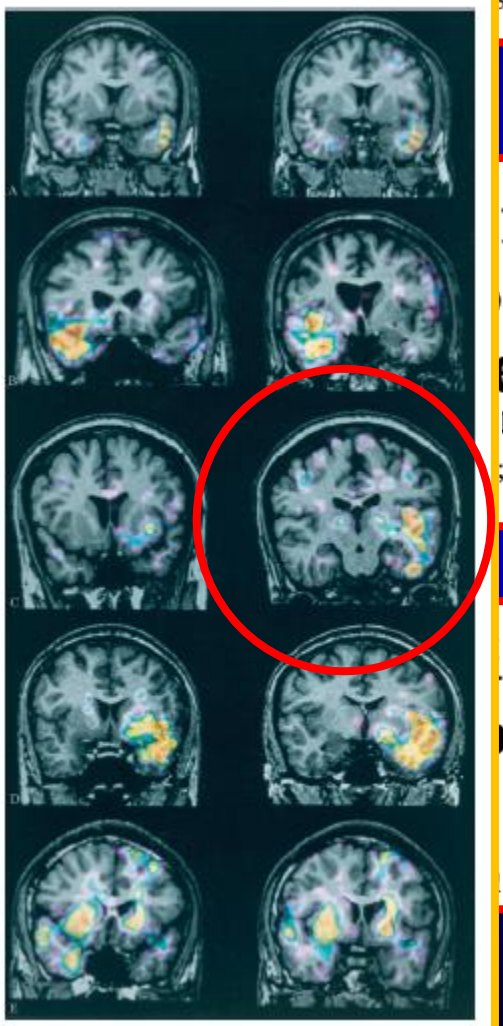
NEUROLOGY 1992;42:371-377

M.R. Newton, MB, ...; M.C. Austin, DipApplSci; D.C. Reutens, MB, ... P.F. Bladin, MD, FRACP

The underlying pathology of ictal dystonia in temporal lobe epilepsy

NEUROLOGY 1998;51:1289–1292

S. Dupont ... Y. Samson, MD



SEMEIOLOGIA
EPILESSIA DEL LOBO FRONTALE

- crisi frequenti (spesso pluri-quotidiane)
- soprattutto in sonno
- breve durata
- inizio e fine bruschi
- rare sensazioni soggettive e poco definite
- perdita di consapevolezza precoce
- confusione post-critica modesta
- frequenti secondarie generalizzazioni TC

SEMIOLOGIA CRITICA

SIMPLE MOTOR SEIZURES

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- TONIC SEIZURES
- TONIC-CLONIC SEIZURES
- EPILEPTIC SPASMS
- VERSIVE SEIZURES

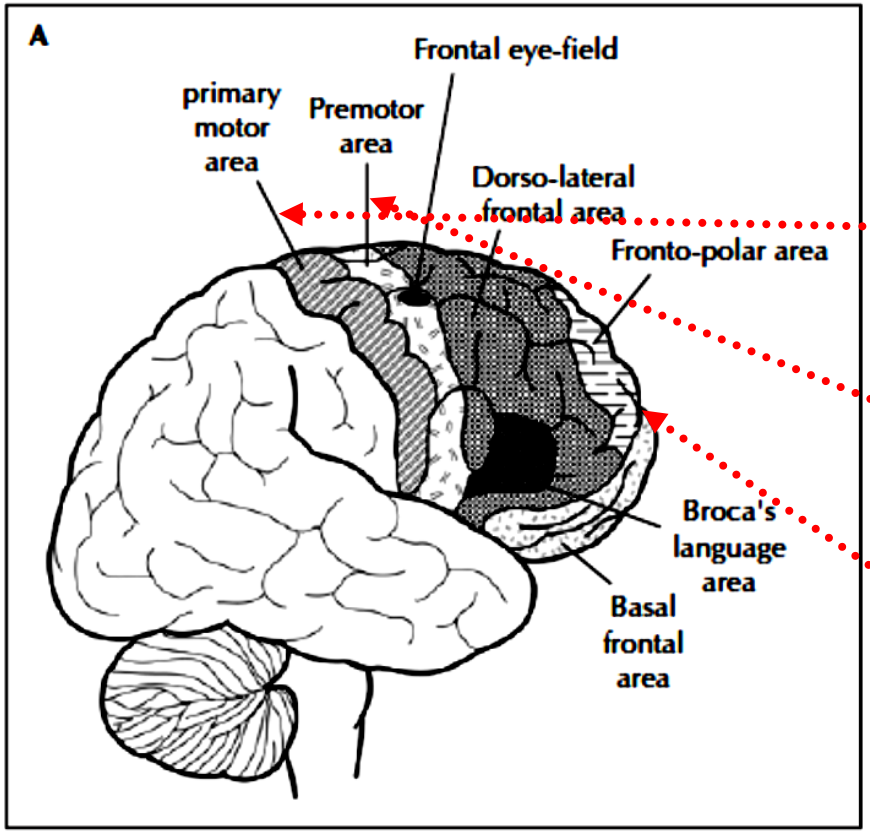
COMPLEX MOTOR SEIZURES

- AUTOMOTOR SEIZURES
- HYPERMOTOR SEIZURES
- GELASTIC SEIZURES

Luders et al, Epilepsia 1998

Luders et al, Epileptic Disorders 2019

LOBO FRONTALE



- Area Precentrale
- Area Premotoria
- Area Prefrontale

Modificato da Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239

CLASSIFICAZIONE ANATOMO-CLINICA EPILESSIA DEL LOBO FRONTALE

- Crisi focali motorie → Area Motoria Primaria

- Crisi toniche posturali → Area Motoria Supplementare

- Crisi psico-motorie (Crisi parziali complesse) →
  - Frontale anteriore (polare)
  - Orbito-frontale
  - Giro del cingolo
  - Dorso-laterale
  - Corteccia mesiale intermedia

ILAE 1985. Workshop on frontal lobe seizures 1988

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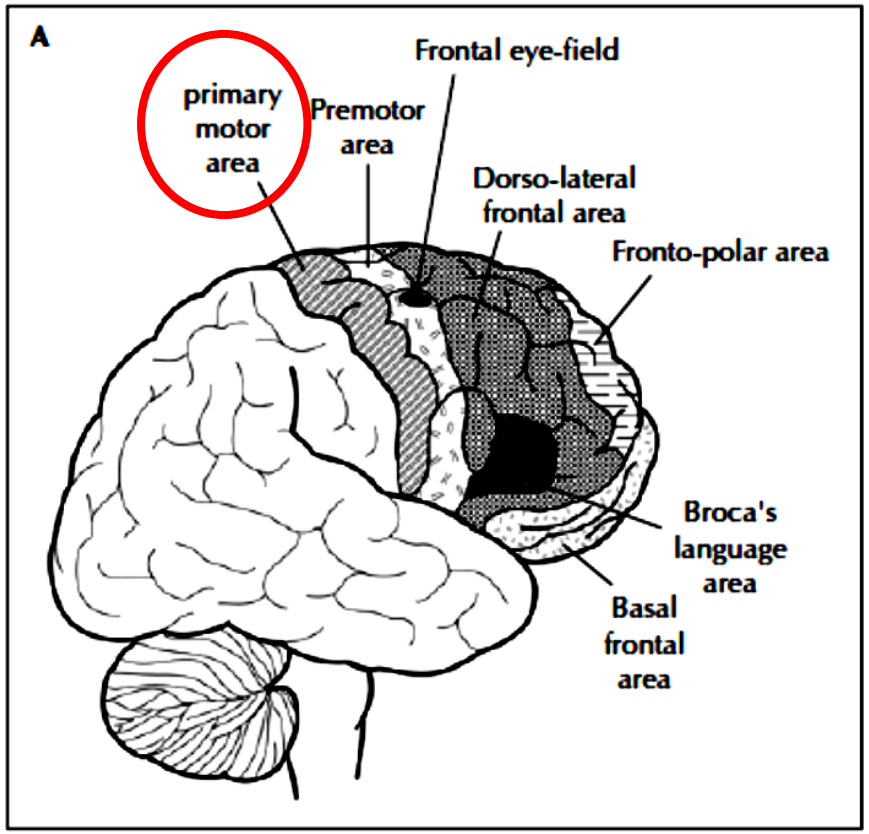
ILAE 1985. Workshop on frontal lobe seizures 1988

CRISI FOCALI MOTORIE

AREA MOTORIA PRIMARIA



Limiti anatomici: giro precentrale, parte posteriore dei giri superiore, mediale, frontale inferiore.

Afferenze: **regioni sensori-motorie**, aree visive, acustiche, talamiche, reticolari o cortico-corticali.

Efferenze: **cortico-spinali**, cortico-corticali, cortico-sottocorticali

Stimolazione elettrica:

- **risposte motorie controlaterali**.
- risposte sensitive-negative

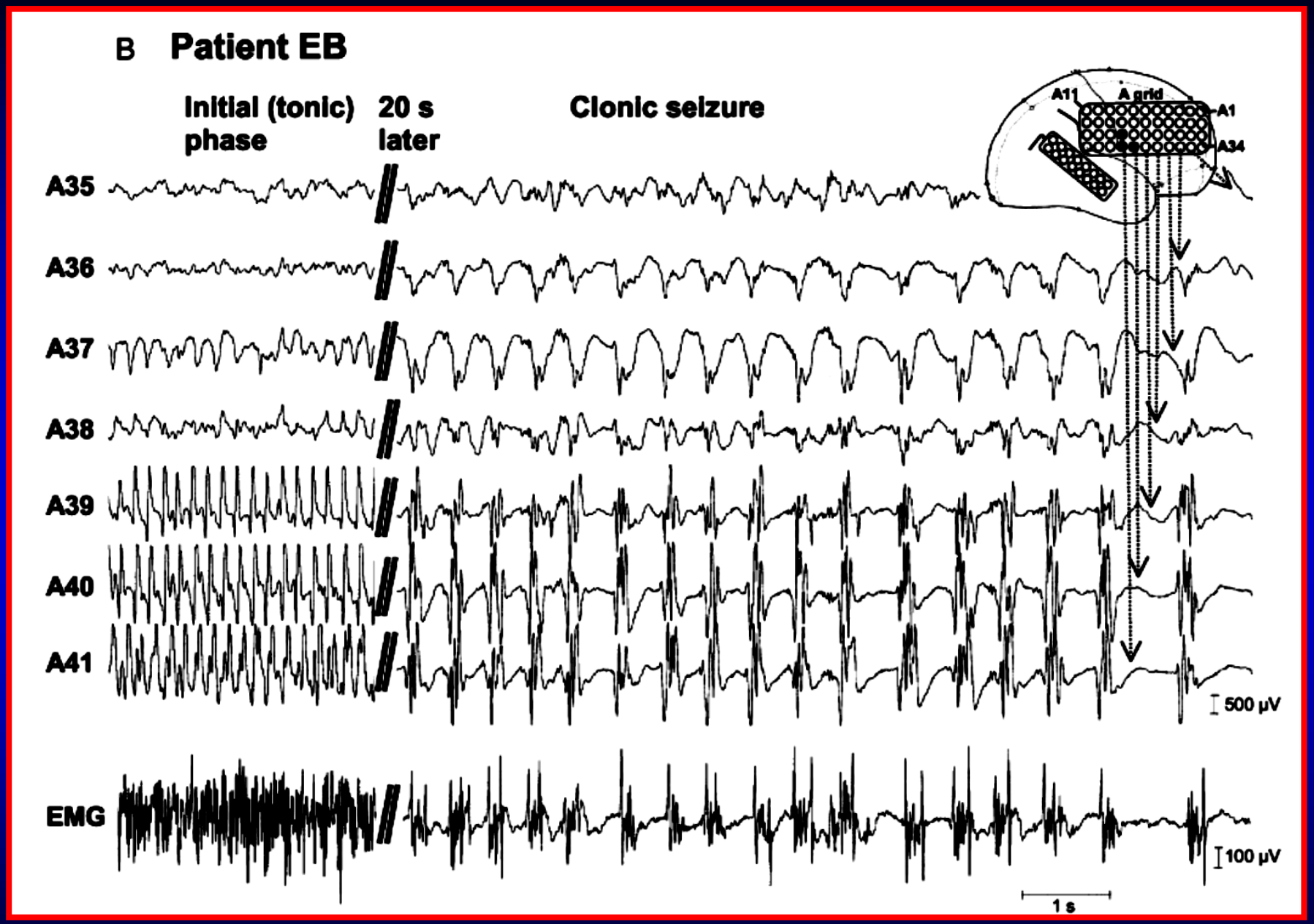
Modificato da Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239

CRISI FOCALI MOTORIE

AREA MOTORIA PRIMARIA

- **Clinicamente**: attività motoria controlaterale. Di solito, esordio unilaterale nella faccia, quindi coinvolgimento dell'arto superiore (*marcia jacksoniana*), seguiti da arresto della parola e clonie palpebrali. Preservazione del contatto. Talora, secondarie generalizzazioni.
- **Quadro poligrafico-clinico**: incremento nel tono muscolare associato a *punte rapide ripetitive* sul giro pre-centrale, seguito da un regolare pattern di breve e sincrone contrazioni dei muscoli agonisti e antagonisti alternate a "rilassamento" muscolare.

Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239



Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239

SEMIOLO

SIMPLE MOTOR SEIZURES

- MYOCLONIC SEIZURES

a. Myoclonic seizures. Myoclonic seizures consist of short muscle contractions lasting <400 ms.

- CLONIC SEIZURES

d. Clonic seizures. Clonic seizures are a series of myoclonic contractions that regularly recur at a rate of 0.2–5/s.

- TONIC-CLONIC SEIZURES
- EPILEPTIC SPASMS
- VERSIVE SEIZURES

) or by direct observation. Docu-
utonomic dysfunction may be clini-
episodes may only be detected by a
patient experiencing any unusual

is a new term coined to identify
predominant symptomatology con-
of consciousness (see discussion of
Alteration of consciousness is dif-
ever, for the purpose of identifying
dialeptic seizures, we classify altered consciousness as
does the ILAE classification of epileptic electroclinical
complexes, e.g., as episodes of unresponsiveness or de-
creased responsiveness that are not caused by motor al-
s. Dialeptic seizures are associated with complete
st partial amnesia for the episode; therefore, am-
the episode is necessary to establish the diagno-
aleptic seizure.
al dialeptic seizures consist of short episodes of
consciousness: <20 s. The alteration of con-
sciousness begins and ends abruptly and frequently is
associated with rhythmical eye blinking at a rate of ~3
Hz. Patients with generalized absence epilepsy often
pical dialeptic seizures. All seizures that consist
of an alteration of consciousness but that are not
l dialeptic seizures'' should be classified as just
seizures.

seizures

res in which the main symptomatology are motor
e identified as motor seizures. Two major sub-
can be differentiated:

1. Simple motor seizures in which the motor movements are relatively ''simple,'' unnatural, and consist of movements similar to movements elicited by electrical stimulation of the primary motor areas (Brodmann areas 4 and 6).
2. Complex motor seizures, in which the movements are relatively complex and simulate natural movements, except that they are inappropriate for the situation.

Simple motor seizures

Simple motor seizures can be subdivided into the following subgroups:

a. Myoclonic seizures. Myoclonic seizures consist of short muscle contractions lasting <400 ms.
b. Tonic seizures. Tonic seizures consist of sustained muscle contractions, usually lasting >3 s, that lead to ''positioning.''
c. Epileptic spasms. The term epileptic spasm is used to identify muscle contractions of variable duration which affect predominantly axial muscles. Epileptic spasms frequently occur in clusters in which the duration of the muscle contractions may vary from a short myoclonic jerk to a sustained tonic posturing. Usually the epileptic spasm consists of abduction of both arms in a ''salaam'' posture.
d. Clonic seizures. Clonic seizures are a series of myoclonic contractions that regularly recur at a rate of 0.2–5/s.
e. Tonic-clonic seizures. Generalized tonic-clonic seizures are characterized by an initial tonic posturing of all limbs. The sustained muscle contractions that determined the tonic phase then tend to slow, evolving into a clonic phase with contractions of progressively decreasing frequency until the contractions disappear completely. The muscles included in the tonic and clonic phase should be essentially the same. Focal motor seizures showing such a tonic-clonic evolution are infrequent.
f. Versive seizures. Versive seizures are seizures during which the patient either has a conjugate eye movement to one side or moves the head, and occasionally the whole body, to one side. Only conjugate eye movements or lateral head and body movements that are *sustained and extreme* should be classified as versive seizure. The lateral movement of the eyes frequently consists of a combination of a smooth tonic lateral movement on which are superimposed small saccades that progressively move the eye out to an extreme position. On other occasions, a smooth lateral movement without any saccades may be observed. The version of body parts has a similar character, but the saccades are replaced by small clonic lateral movements of the head or body. During these lateral movements, the chin frequently moves not only laterally but also upward, resulting in an *unnatural* position of the eyes and head. Occasionally, the patient's body will also turn and may complete one or more 360° turns.

Complex motor seizures

The following three types of complex motor seizures can be distinguished. Again, ''complex'' herein refers to the complex characteristics of the movement and does not mean that the patient loses awareness during the seizure.

a. Hypermotor seizures. Hypermotor seizures are seizures in which the main manifestations consist of complex movements involving the proximal segments of the limbs and trunk. This results in large movements that appear ''violent'' when they occur at high speeds. The ''complex motor manifestations'' imitate normal movements, but the movements are inappropriate for the situation and usually serve no purpose. Frequently, the movements

Luders et al., Epilepsia 1998

CLASSIFICAZIONE ANATOMO-CLINICA EPILESSIA DEL LOBO FRONTALE

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  - Frontale anteriore (polare)
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  - Dorso-laterale
  - Corteccia mesiale intermedia

ILAE 1985. Workshop on frontal lobe seizures 1988

CRISI TONICHE POSTURALI

AREA MOTORIA SUPPLEMENTARE

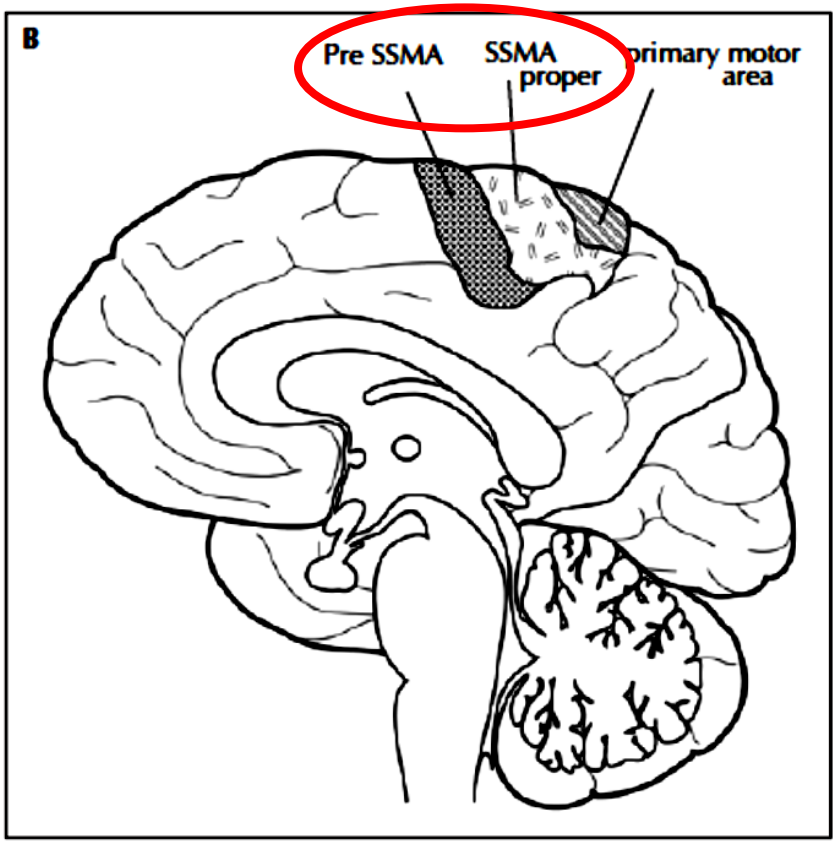
Limiti anatomici: parte posteriore del giro frontale superiore, lobulo paracentrale, area motoria primaria.

Afferenze: talamo, corteccia pre-motoria e post-centrale, area motoria primaria, proiezioni cortico-spinali (*SSMA proper*

Ruolo: *SSMA proper*: proprietà esecutive motorie; *pre-SSMA*: iniziare

Stimolazione elettrica:

- **risposte motorie toniche, prossimali, ipsi/contro/bilaterali**.
- risposte sensitive contro/bilaterali
- deviazioni oculari controlaterali
- . Risposte negative (pre-SSMA)



Modificato da Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239

CRISI TONICHE POSTURALI

AREA MOTORIA SUPPLEMENTARE

- **Clinicamente**: brevi (10-40 secondi), bilaterali, asimmetriche posture toniche con abduzione/elevazione degli arti, flessione degli arti superiori (gomiti), preservazione del contatto.
- **Quadro poligrafico-clinico**: 1) artefatti; 2) attività critica sul vertice e/o adiacente alla SMA; 3) attività tonica simmetrica o asimmetrica arti e tronco.

Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239

CRISI TONICHE POSTURALI

AREA MOTORIA SUPPLEMENTARE

SEMEIOLOGIA TIPICA

Modificazione posturale asimmetrica delle estremità (1? 2? 4?) -> arresto del linguaggio a coscienza conservata -> abduzione in elevazione dell'arto superiore controlaterale e deviazione capo e occhi verso l'arto superiore coinvolto.

Manifestazioni soggettive e vocalizzazione meno frequenti.

Kellinghaus et Luders, Frontal Lobe Epilepsy, Epileptic Disord 2004; 6:223-239

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Review article

Epileptic Disord 2019; 21 (1): 1-29

Classification of paroxysmal events and the four-dimensional epilepsy

Motor seizures*	Simple motor seizure*	Clonic seizure*	
		Epileptic spasm*	
		Myoclonic seizure*	
		Nystagmoid seizure*	
		Tonic seizure*	
		Tonic-clonic seizure*	
		Versive seizure*	
		Vocalization seizure	
	Complex motor seizure	Alien limb seizure	
		Automotor seizure	
		Dacrystic seizure	
		Gelastic seizure	
		Hypermotor seizure	Emotional hypermotor seizure
		Kissing seizure	
		Singing seizure	
		Spitting seizure	
		Verbalization seizure	

HYPERMOTOR OR HYPERKINETIC SEIZURES

- Complex movements involving the proximal segments of the trunk and limbs such as body rocking, pelvic thrusting, pedaling, boxing, and kiching.

(Luders et al, 1998; Blume et al. 2001)

- Additional symptoms: fear, facial grimacing, emotional and aggressive behavior, language and nonlanguage vocalization, dystonic posturing, head version

(Provini et al, 1999; Jobst et al, 2000; Kotagal et al. 2003)

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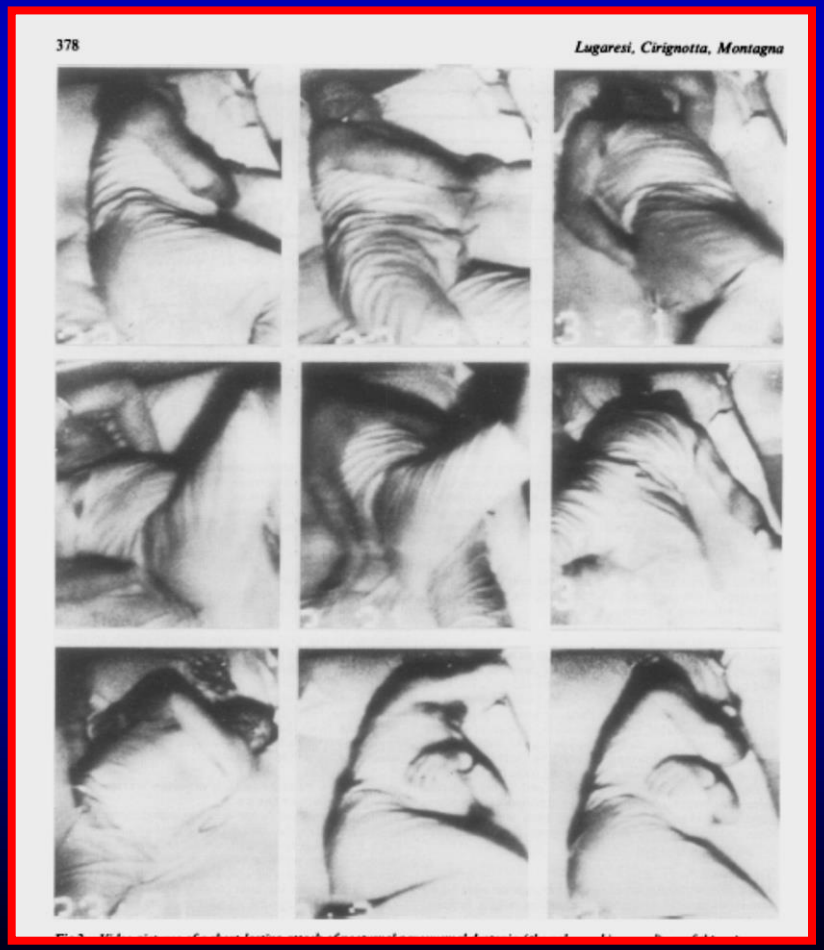
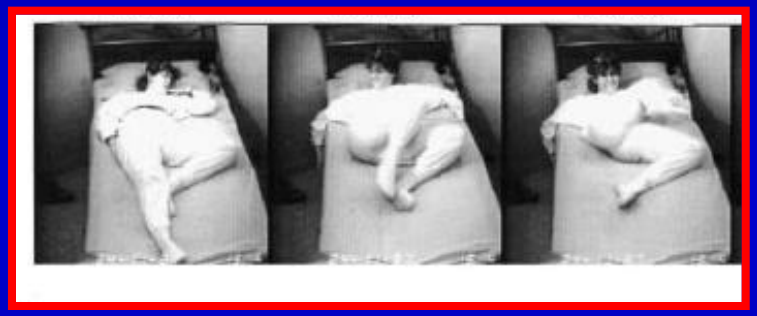
Journal of Neurology, Neurosurgery, and Psychiatry 1986;**49**:375–380

Nocturnal paroxysmal dystonia

E LUGARESI, F CIRIGNOTTA, P MONTAGNA

From the Institute of Neurology, University of Bologna, Italy





Brain (1999), 122, 1017–1031

Nocturnal frontal lobe epilepsy

A clinical and polygraphic overview of 100 consecutive cases

Federica Provini, Giuseppe Plazzi, Paolo Tinuper, Stefano Vandi, Elio Lugaresi and Pasquale Montagna

Summary

Nocturnal frontal lobe epilepsy (NFLE) has been delineated as a distinct syndrome in the heterogeneous group of paroxysmal sleep-related disturbances. The variable duration and intensity of the seizures distinguish three non-rapid eye movement-related subtypes: paroxysmal arousals, characterized by brief and sudden recurrent motor paroxysmal behaviour; nocturnal paroxysmal dystonia, motor attacks with complex dystonic–dyskinetic features; and episodic nocturnal wanderings, stereotyped, agitated somnambulism. We review the clinical and polysomnographic data related to 100 consecutive cases of NFLE in order to define the clinical and neurophysiological characteristics of the different seizure types that constitute NFLE. NFLE seizures predominate in males (7 : 3). Age at onset of the nocturnal seizures varies, but centres during infancy and adolescence. A familial recurrence of the epileptic attacks is found in 25% of the cases, while 39% of the patients present a family history of nocturnal paroxysmal episodes that fit the diagnostic criteria for parasomnias. A minority of cases (13%) have personal antecedents (such as birth anoxia, febrile convulsions) or brain CT or MRI abnormalities (14%). In many patients, ictal (44%) and interictal (51%) EEGs are uninformative. Marked autonomic activation is a common finding during the seizures. NFLE does not show a tendency to spontaneous remission. Carbamazepine completely abolishes the seizures in ~20% of the cases and gives remarkable relief (reduction of the seizures by at least 50%) in another 48%. VideoEEG recordings confirm that NFLE comprises a spectrum of distinct phenomena, different in intensity but representing a continuum of the same epileptic condition. We believe that the detailed clinical and videoEEG characterization of patients with NFLE represents the first step towards a better understanding of the pathogenic mechanisms and different clinical outcomes of the various seizure types that constitute the syndrome.

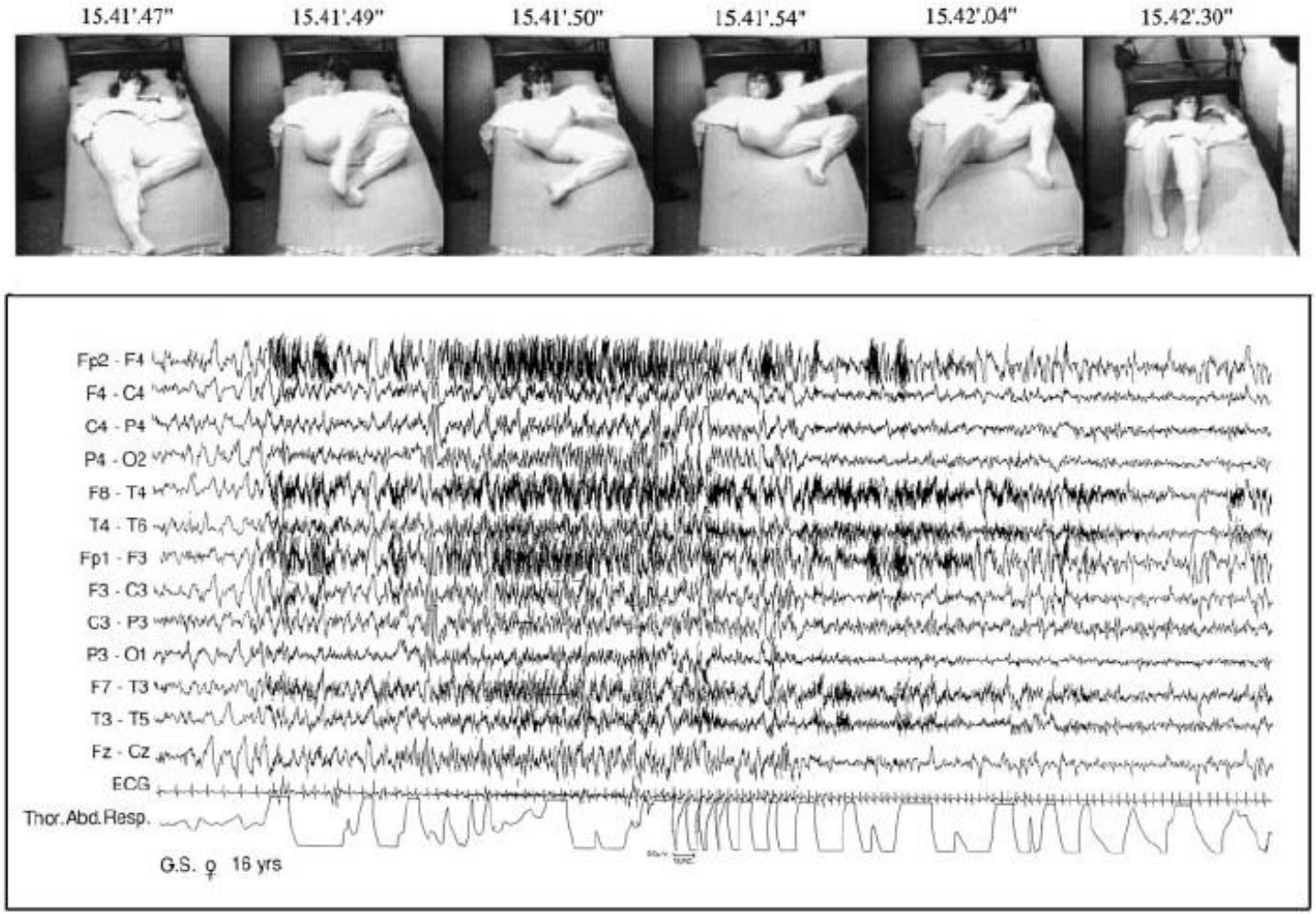
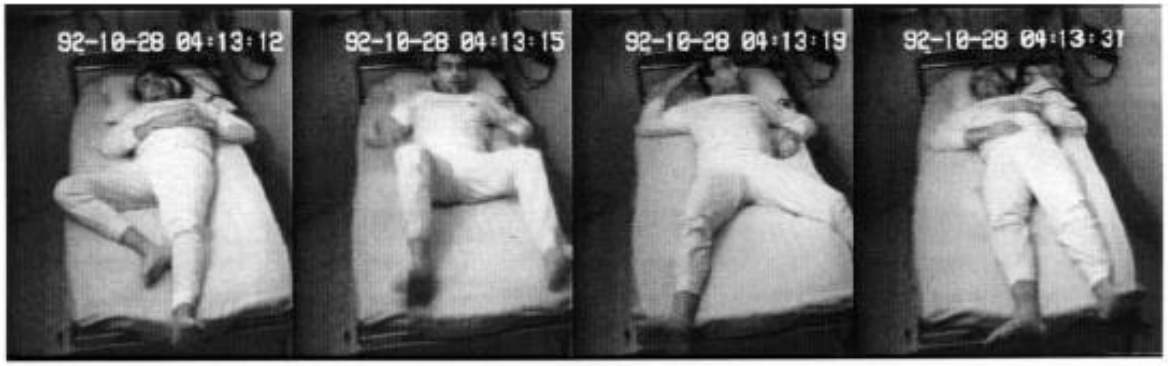


Fig. 2 An NPD seizure. The patient violently rocks her legs, presents a dystonic posture of the arms and an asymmetric grimace. The seizure lasts 40 s. Polysomnograph: the seizure arises during slow wave sleep. A slow wave bouffee with superimposed fast rhythms appears over the right frontal region. Subsequently, a rhythmic fast activity spreads to the right central and parietal regions and to the left frontal regions. Tachycardia and irregular breathing are evident.



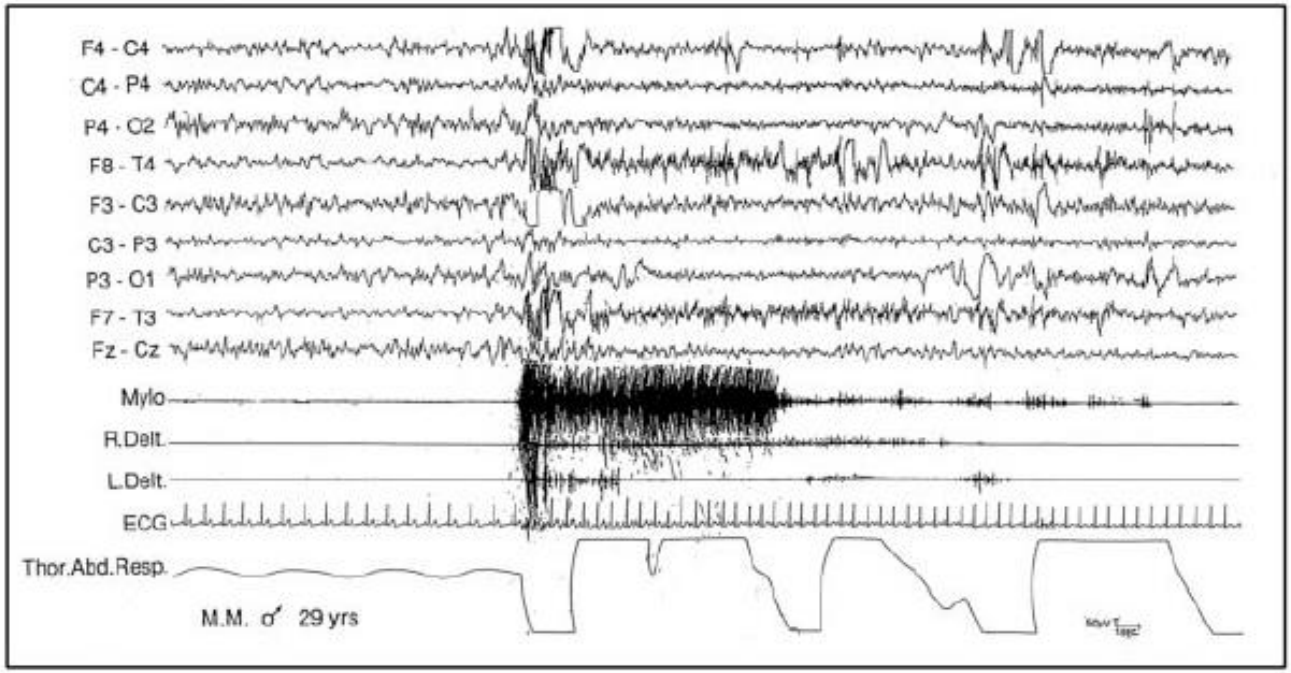


Fig. 1 A typical PA seizure. An abrupt arousal during which the patient opens his eyes and raises his head, trunk and limbs with a fearful expression. The episode lasts 19 s. Polysomnograph: a K-complex precedes the seizure onset. Marked tachycardia and irregular breathing also occur.


A
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23:38
23. 4.1991

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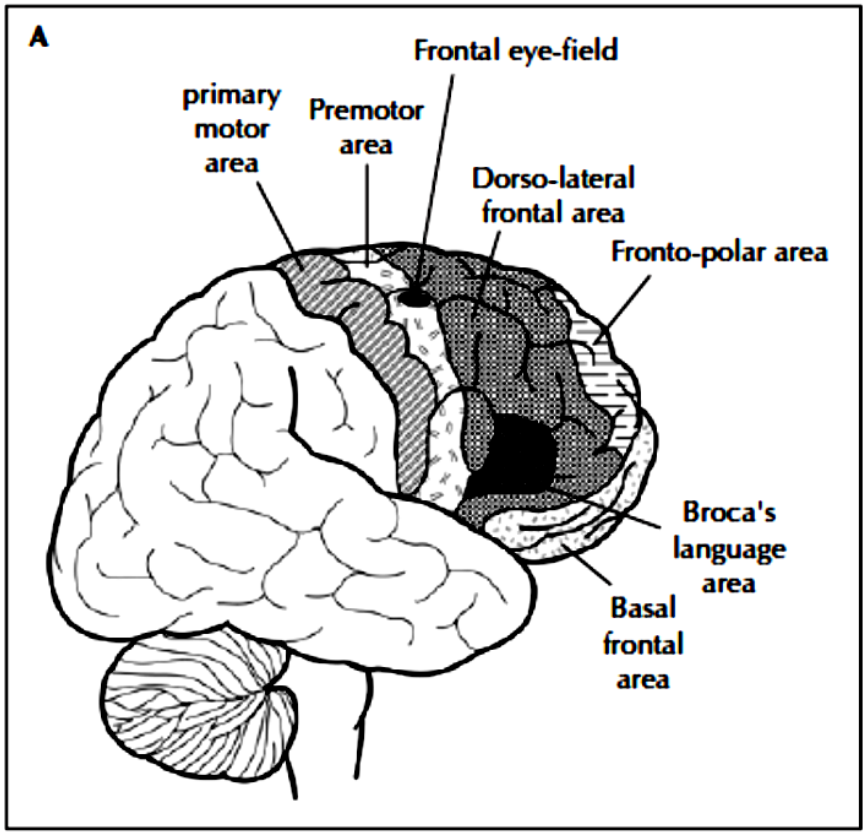
CRISI IPERMOTORIE

Studi SEEG



- Cortex cingulate cortex (82%)
- Orbitofrontal cortex (45%)
- Frontal polar cortex (27%)
- Mesial premotor cortex (18%)

(Rheims et al., 2008)

- Dorsolateral frontal cortex

(Tal et al, 2010)

CRISI IPERMOTORIE

Zone Epilettogena

- **Tonic or Dystonic Posturing** → Posterior Mesial Frontal Cortex (frequent involvement of the supplementary motor area and the posterior mesial frontal cortex)

(Nobili et al, 2007; Morris et al, 1988; Baumagartner et al., 1996; Kellinghaus et al., 2004; Rheims et al, 2008; Proserpio et, 2011)

- **Hyperkinetic behavior** → Dorsolateral Frontal Cortex, Anterior Frontal Cingulate Gyrus, Orbito-polar Region

(Nobili et al, 2007; Rheims et al, 2008; Proserpio et, 2011)

HYPERMOTOR OR HYPERKINETIC SEIZURES

Extrafrontal Brain Regions

- Anterior Superior portion of insula (Ryvlin et al., 2006)
- Parietotemporal region (Nishibayashi et al., 2009)
- Temporal lobe epilepsy (.....).

CRISI IPERMOTORIE IN TLE

SLEEP IN NEUROLOGICAL DISORDERS

Epileptic Nocturnal Wanderings with a Temporal Lobe Origin: A Stereo-Electroencephalographic Study

SLEEP, Vol. 25, No. 6, 2002

Lino Nobili, MD;[1,2] Stefano Francione, PhD;[1] Francesco Cardinale, MD;[1] and Giorgio Lo Russo, MD[1]

R. Mai • I. Sartori • S. Francione • L. Tassi • L. Castana • F. Cardinale • M. Cossu • A. Citterio N. Colombo • G. Lo Russo • L. Nobili

Sleep-related hyperkinetic seizures: always a frontal onset?

Neural networks underlying hyperkinetic seizures of ''temporal lobe'' origin

Epilepsy Research (2009) **86**, 200—208

L. Vaugier[a], S. Aubert[a], A. McGonigal[a,c], A. Trébuchon[c], M. Guye[a,b,c], M. Gavaret[a,b,c], J. Regis[b,c], P. Chauvel[a,b,c], F. Wendling[d,e], F. Bartolomei[a,b,c,*]

Hypermotor seizures in patients with temporal pole lesions

Lina Wang[a], Gregory C. Mathews[a,*], William O. Whetsell[b], Bassel Abou-Khalil[a]

Epilepsy Research (2008) **82**, 93—98

Temporal lobe origin is common in patients who have undergone epilepsy surgery for hypermotor seizures

Amir M. Arain [a,*], Nabil J. Azar [a], Andre H. Lagrange [a], Michael McLean [a], Pradumna Singh [a], Hasan Sonmezturk [a], Peter Konrad [b], Joseph Neimat [b], Bassel Abou-Khalil [a]

Epilepsy & Behavior 64 (2016) 57–61

Original Article

Sleep Medicine 12 (2011) S33–S38

Epileptic motor behaviors during sleep: Anatomo-electro-clinical features

P. Proserpio [a,b], M. Cossu [a], S. Francione [a], F. Gozzo [a], G. Lo Russo [a], R. Mai [a], A. Moscato [a], M. Schiariti [a], I. Sartori [a], L. Tassi [a], L. Nobili [a,b,*]

Hyperkinetic seizures in patients with temporal lobe epilepsy: Clinical features and outcome after temporal lobe resection

Epilepsia, 52(8):1439–1446, 2011

*Anke M. Staack, *Sofia Bilic, *Anne-Sophie Wendling, *Julia Scholly, *Uwe Kraus, *Karl Strobl, *Frédéric Bodin, †Josef Zentner, and *Bernhard J. Steinhoff

Sleep-related hyperkinetic seizures of temporal lobe origin

NEUROLOGY 2004;62:482–485

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

Complex motor behaviors in temporal lobe epilepsy

Abstract—Complex motor behaviors differing from typical automatisms were found in 12 of 502 patients with temporal lobe epilepsy. Movements involved proximal limb segments (6) or body axis (6) and were often preceded by auras and followed by automatisms. Seven of 12 patients are seizure free after surgery. The other 5 patients declined surgery.

NEUROLOGY 2005;65:1805–1807

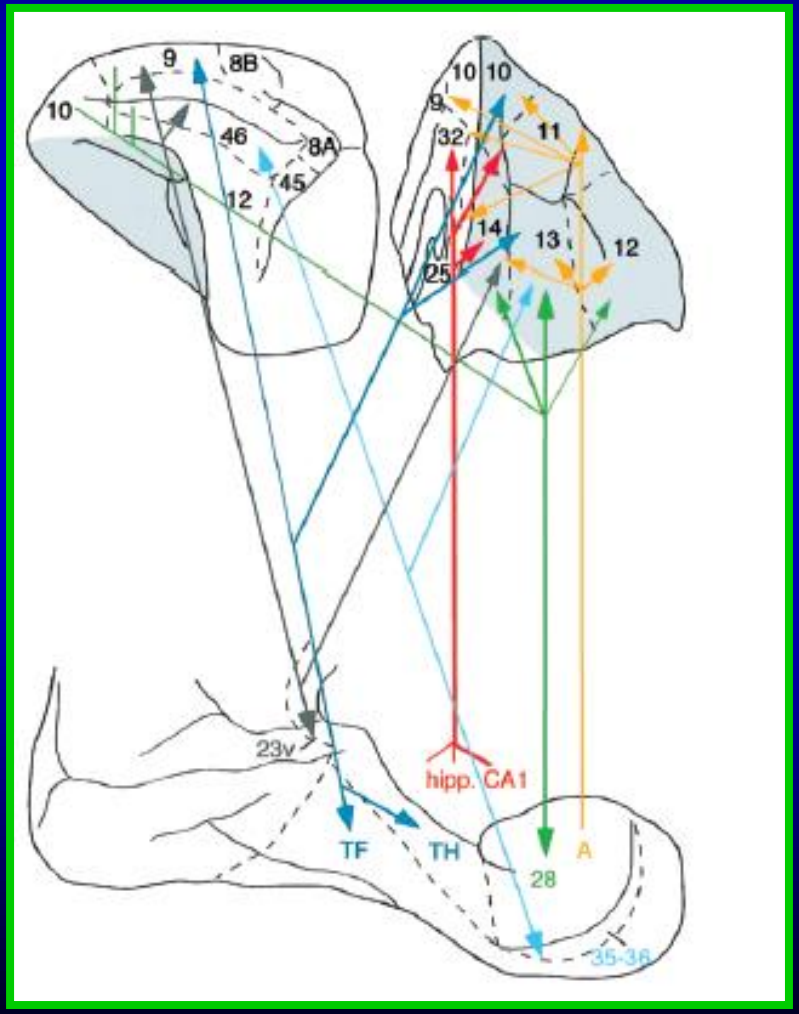
M. Carreño, MD, PhD; A. Donaire, MD; M.A. Pérez Jiménez, MD, PhD; R. Agudo, MD; A. Quilez, MD; J. Rumià, MD; F. Villarejo, MD; N. Bargalló, MD, PhD; T. Boget, PsyD; T. Raspall, MD; L. Pintor, MD, PhD; and X. Setoain, MD, PhD

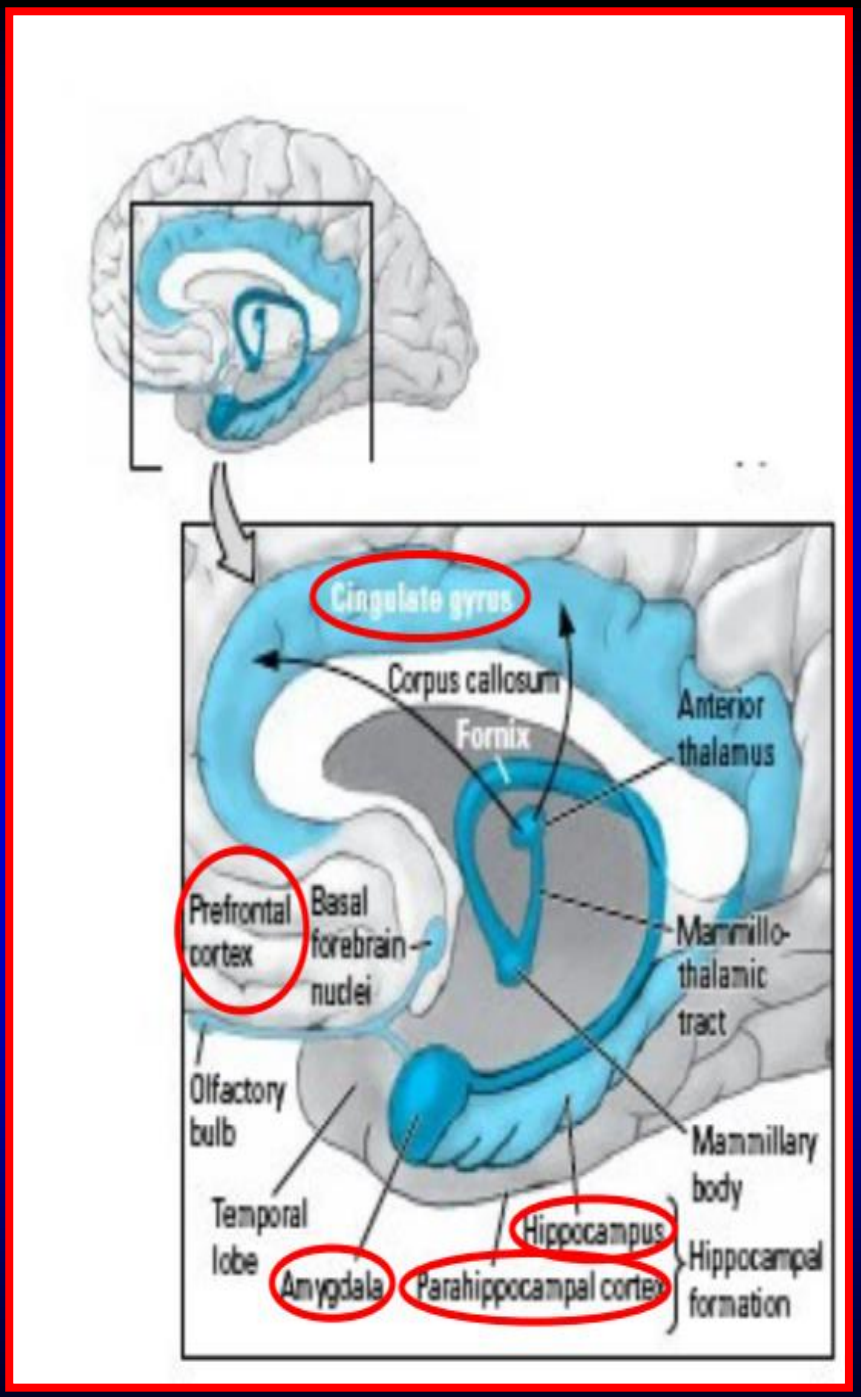
The Anatomical Connections of the Macaque Monkey Orbitofrontal Cortex. A Review

Cerebral Cortex Mar 2000;10:220–242; 1047–3211/00/$4.00

Carmen Cavada, Teresa Compañy, Jaime Tejedor, Roelf J. Cruz-Rizzolo[1] and Fernando Reinoso-Suárez

Departamento de Morfología, Facultad de Medicina, Universidad Autónoma de Madrid, 28029 Madrid, Spain
[1]Current address: Department of Basic Sciences, State University 'Júlio de Mesquita Filho', Campus of Araçatuba, São Paulo, Brazil




Cingulate gyrus
Corpus callosum
Fornix
Anterior thalamus
Prefrontal cortex
Basal forebrain nuclei
Mammillo-thalamic tract
Olfactory bulb
Mammillary body
Temporal lobe
Hippocampus
Amygdala
Parahippocampal cortex
Hippocampal formation

Journal of Neurology, Neurosurgery, and Psychiatry 1986;**49**:375–380

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VIEWS & REVIEWS

Definition and diagnostic criteria of sleep-related hypermotor epilepsy

OPEN

Paolo Tinuper, MD

Neurology 86 May 10, 2016

CRISI IPERMOTORIE IN TLE

Caratteristiche Critiche

- **Semiology**: bidirectional rotation, ballistic movements, frenetic festural automatisms. Aggressive and compulsive behavior, marked agitation. No aura.
- **Clinical Onset**: after seizure onset (3-10 s.; 8-15 s.).
- **Sleep** > Awake.
- **Duration**: 20-40 sec.
- **Frequency**: 1-5 seizures/24 h

(Nobili et al., 2004; Mai et al., 2005; O'Brien et al., 2008; Staack et al., 2011)

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CRISI IPERMOTORIE

TLE vs FLE

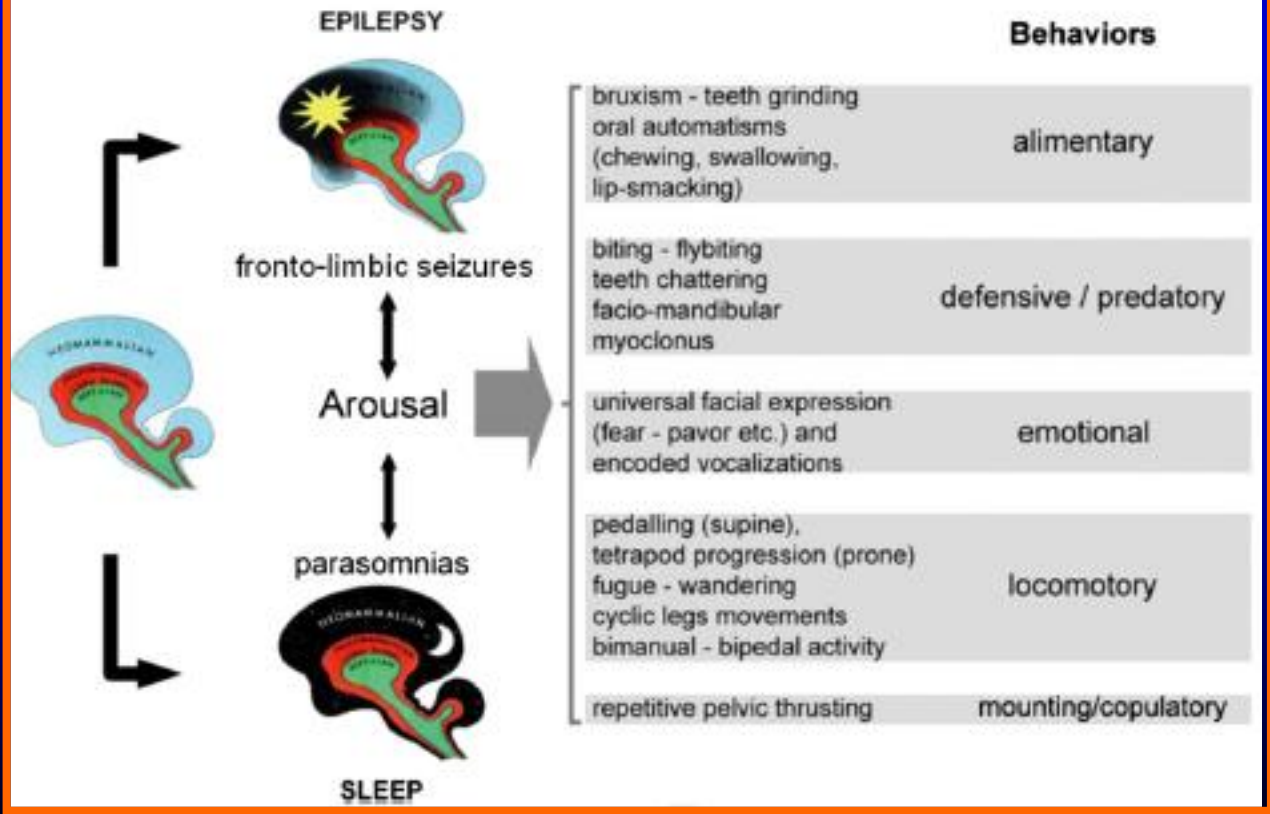
- Complex motor automatisms are more asymmetric.
- Ipsilaterally to the onset of the epileptic discharges.
- Warning sensation (fear, discomfort epigastric, déjà vu).
- An accurate analysis of all the clinical, electrophysiological, and neuroradiological data is essential in order to identify clues suggestive of a specific site of origin.

(Nobili et al., 2004; Mai et al., 2005)

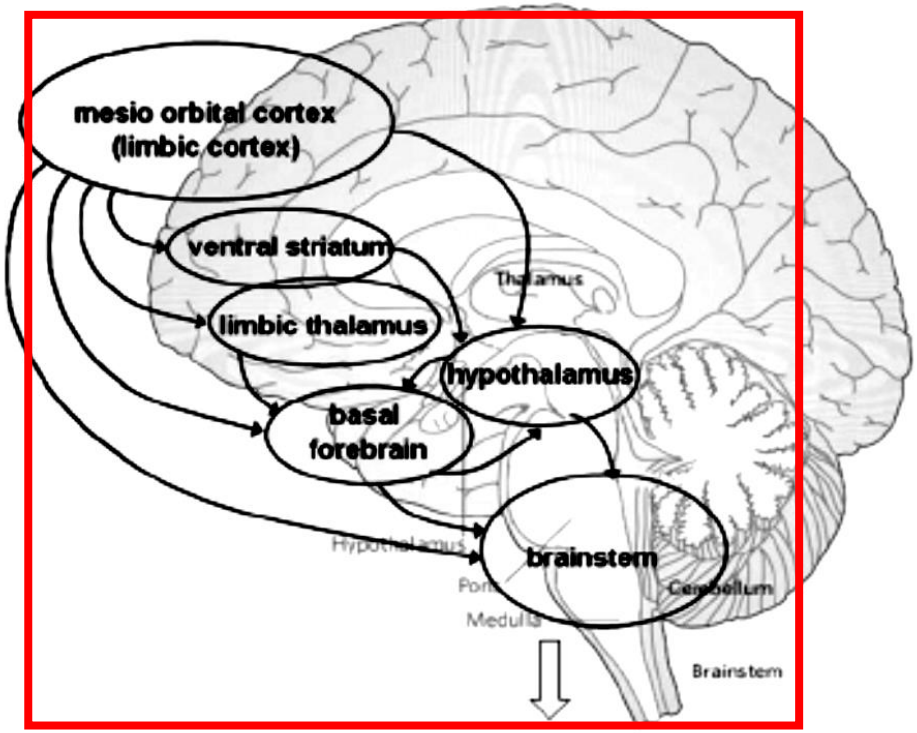
Relationship of Central Pattern Generators with Parasomnias and Sleep-Related Epileptic Seizures

Carlo Alberto Tassinari[a], Elena Gardella[b,c], Gaetano Cantalupo[d], Guido Rubboli, MD[c,e,*]

Sleep Med Clin ■ (2012)



LIMBIC LOOPS ACTIVATED BY PATHOLOGICAL AROUSALS



CLINICAL MANIFESTATIONS :
SUDDEN AROUSAL, COMPLEX MOTOR AND AUTONOMIC BEHAVIOUR

Tinuper et al, Sleep Medicine 12 (2011) S39-S42

Disturbo del Movimento

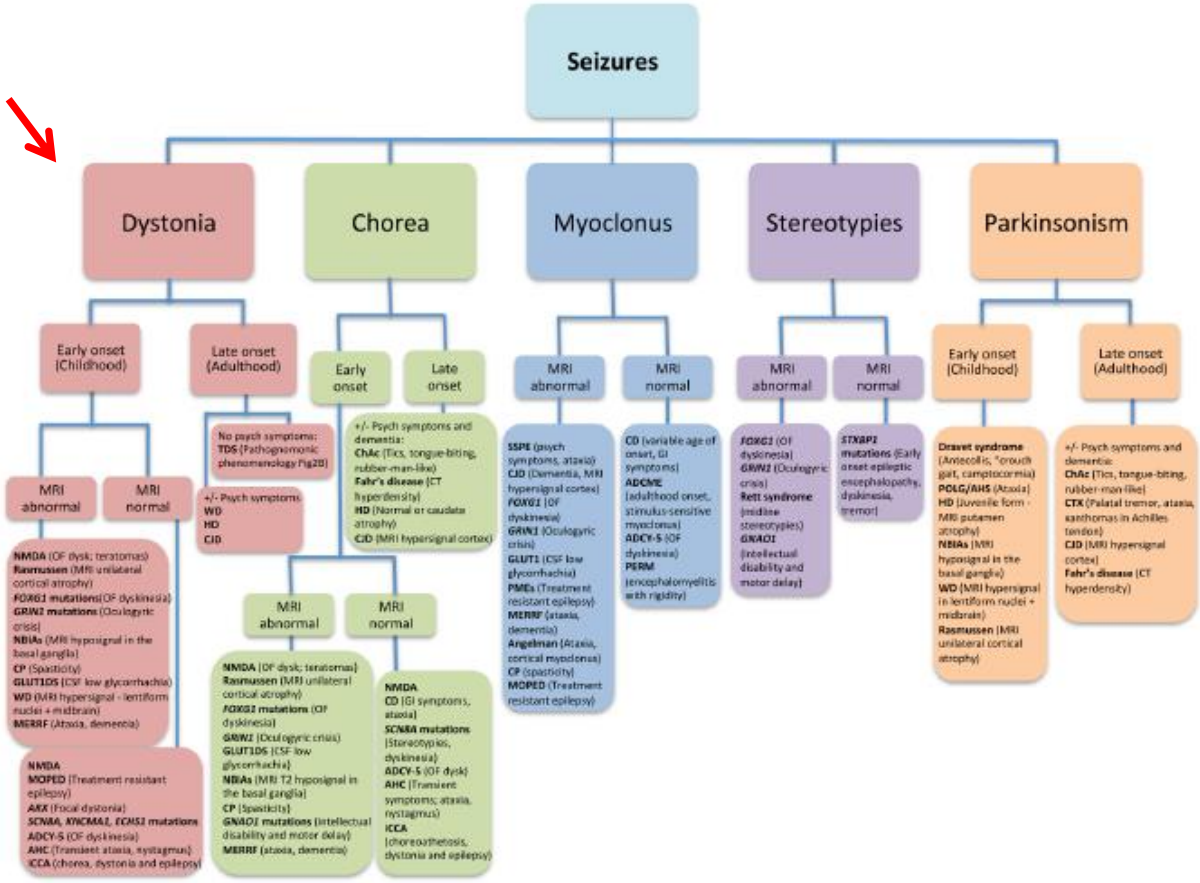
Altre Crisi Epilettiche ed Epilessie

Le Crisi e le Epilessie del Lobo Frontale

REVIEW *J Neurol Neurosurg Psychiatry* 2019;**90**:920–928. doi:10.1136/jnnp-2018-320039

Seizures and movement disorders: phenomenology, diagnostic challenges and therapeutic approaches

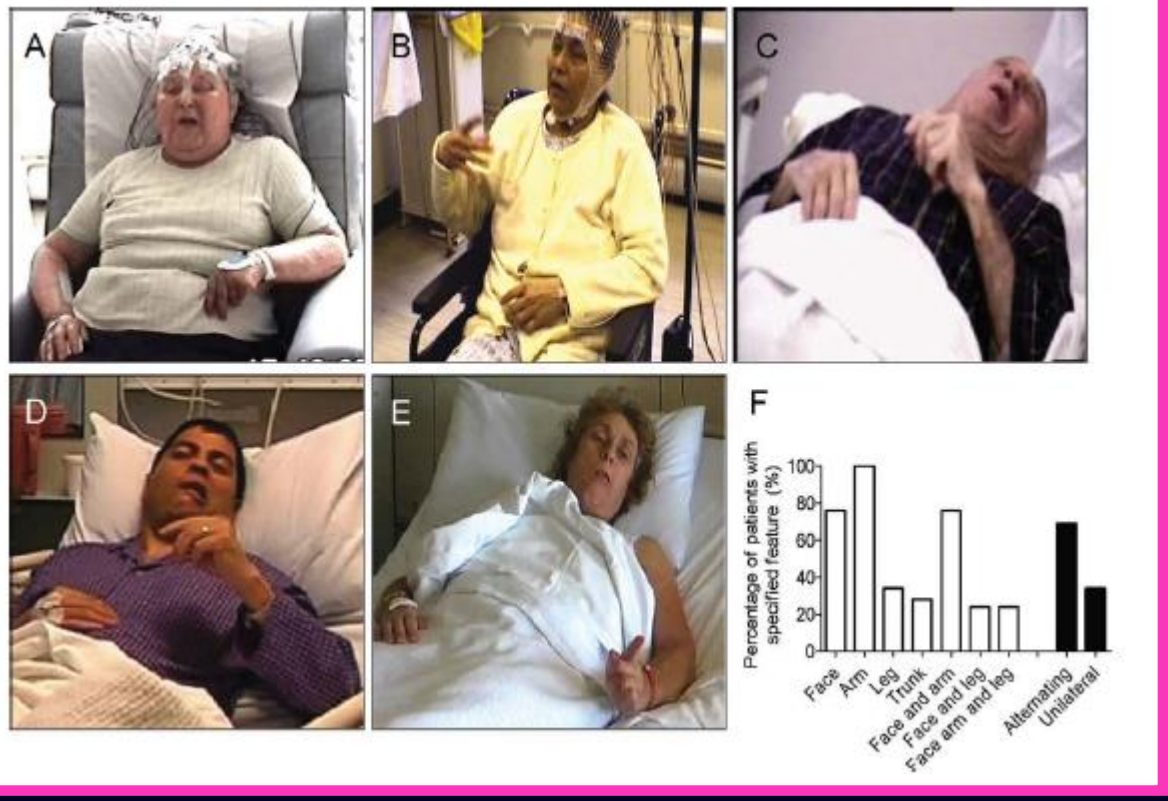
Maria Eliza Freitas,[1] Marta Ruiz-Lopez,[2] Josep Dalmau,[3] Roberto Erro,[4] Michael Privitera,[5] Danielle Andrade,[6] Alfonso Fasano[7]



Faciobrachial Dystonic Seizures Precede Lgi1 Antibody Limbic Encephalitis

ANN NEUROL 2010

Sarosh R. Irani, DPhil,[1] Andrew W. Michell, PhD,[2] Bethan Lang, PhD,[1] Philippa Pettingill, BSc,[1] Patrick Waters, PhD,[1] Michael R. Johnson, PhD,[3] Jonathan M. Schott, MD,[4] Richard J. E. Armstrong, PhD,[1,4] Alessandro S. Zagami, MD,[5] Andrew Bleasel, PhD,[6] Ernest R. Somerville, FRCAP,[5,7] Shelagh M. J. Smith, FRCP,[8] and Angela Vincent, FRCPath[1,9]

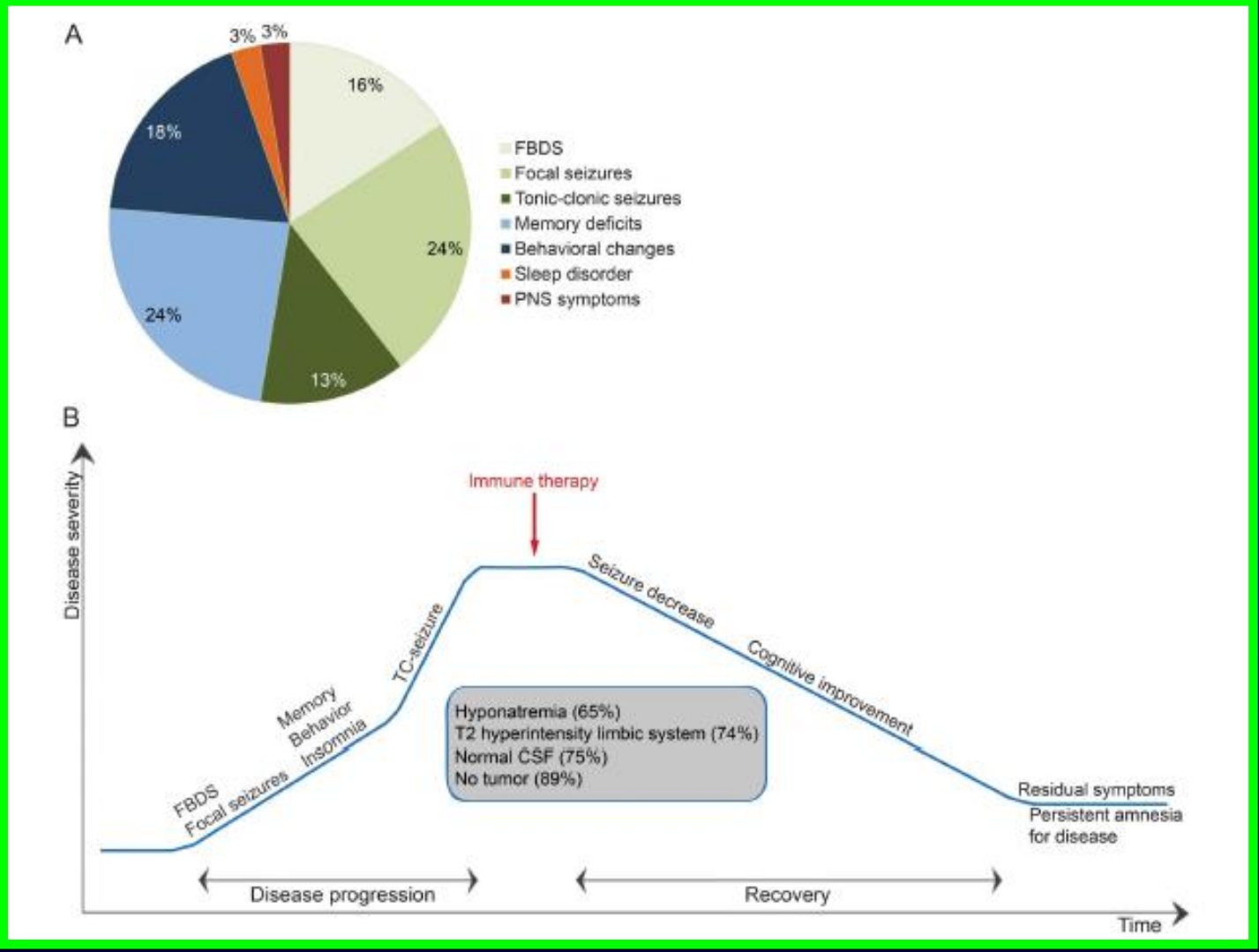


Anti-LGI1 encephalitis

Neurology® 2016;87:1449-1456

Clinical syndrome and long-term follow-up

Agnes van Sonderen, MD
Roland D. Thijs, MD, PhD
Elias C. Coenders, BSc
Lize C. Jiskoot, MSc
Esther Sanchez, MD
Marienke A.A.M. de Bruijn, MD
Marleen H. van Coevorden-Hameete, MD
Paul W. Wirtz, MD, PhD
Marco W.J. Schreurs, PhD
Peter A.E. Sillevis Smitt, MD, PhD
Maarten J. Titulaer, MD, PhD



ONLINE FIRST

Kinesigenic Dyskinesia in a Case of Voltage-Gated Potassium Channel–Complex Protein Antibody Encephalitis

Enrique Aradillas, MD; Robert J. Schwartzman, MD

Arch Neurol. 2011;68(4):529-532. Published online December 13, 2010. doi:10.1001/archneurol.2010.317

Faciobrachial Dystonic Attacks: Seizures or Movement Disorder?

Pasquale Striano, MD, PhD

ANNALS *of Neurology* July 2011

Images in Neurology

Rapidly Cycling Auras and Episodic Focal Dystonia in Anti-LGI1 Autoimmune Encephalitis

JAMA Neurology September 2016

Tadeu A. Fantaneanu, MD; Shamik Bhattacharyya, MD; Tracey A. Milligan, MD; Page B. Pennell, MD

Creutzfeldt-Jakob disease versus anti-LGI1 limbic encephalitis in a patient with progressive cognitive dysfunction, psychiatric symptoms, involuntary facio-brachio-crural movement, and an abnormal electroencephalogram: a case report

Neuropsychiatric Disease and Treatment 2015:11 1427–1430

Faciobrachial Dystonic Seizures Precede Lgi1 Antibody Limbic Encephalitis

ANN NEUROL 2010

Sarosh R. Irani, DPhil,[1] Andrew W. Michell, PhD,[2] Bethan Lang, PhD,[1] Philippa Pettingill, BSc,[1] Patrick Waters, PhD,[1] Michael R. Johnson, PhD,[3] Jonathan M. Schott, MD,[4] Richard J. E. Armstrong, PhD,[1,4] Alessandro S. Zagami, MD,[5] Andrew Bleasel, PhD,[6] Ernest R. Somerville, FRCAP,[5,7] Shelagh M. J. Smith, FRCP,[8] and Angela Vincent, FRCPath[1,9]

Neurology 76 April 12, 2011

Danielle M. Andrade, MD
Peter Tai, MD
Josep Dalmau, MD, PhD
Richard Wennberg, MD

TONIC SEIZURES: A DIAGNOSTIC CLUE OF ANTI-LGI1 ENCEPHALITIS?

Seizures and memory loss are common features of anti-LGI1 limbic encephalitis,[1] a recently described au-

tonic seizures. Identical movements of shorter duration were not associated with definite EEG changes.

Interestingly, all 3 patients also developed frequent, repeated subclinical electrographic seizures of an entirely different sort, not associated with their

Tonic seizures: A diagnostic clue of anti-LGI1 encephalitis?

Pasquale Striano, Sarosh R. Irani, Danielle Andrade, et al.

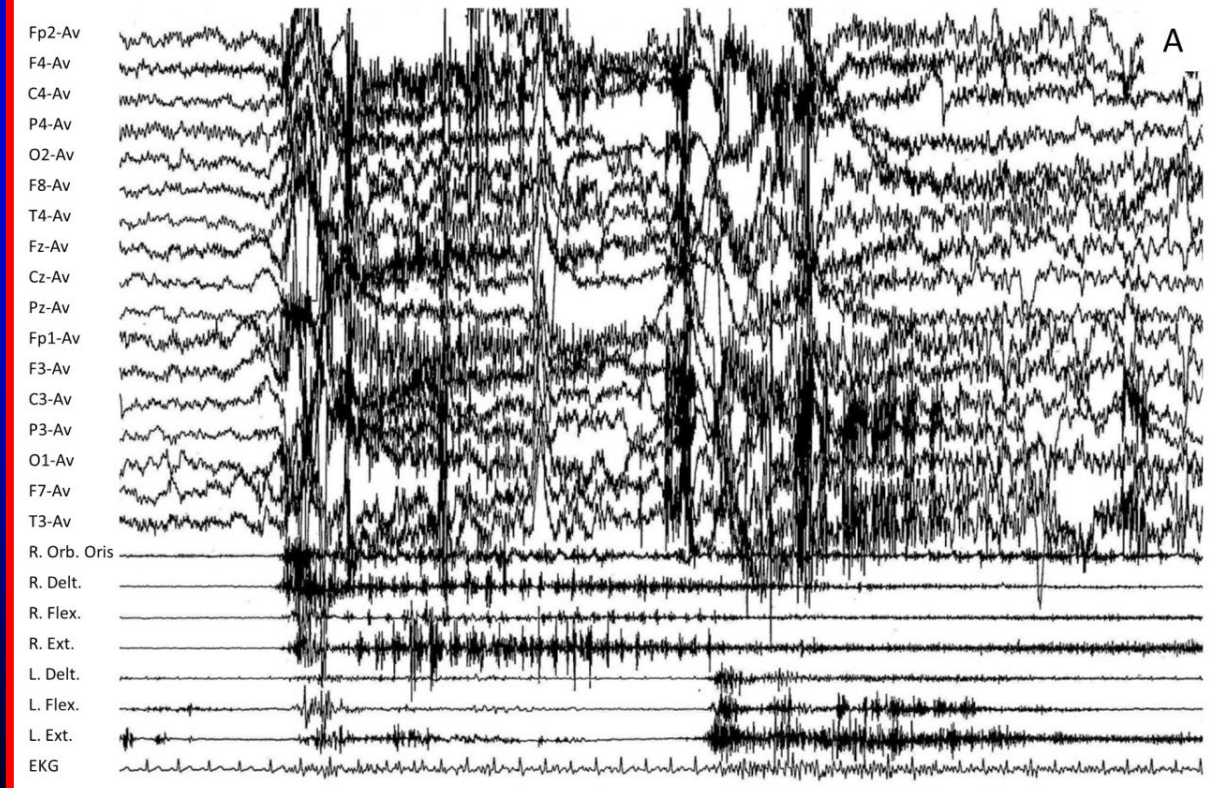
Neurology 2011;77;2140
DOI 10.1212/WNL.0b013e318239c3d7

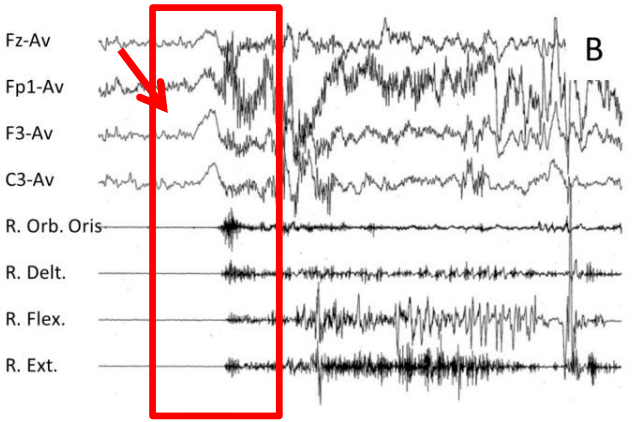
Faciobrachial dystonic seizures expressed as epileptic spasms, followed by focal seizures in anti-LGI1 encephalitis: a video-polygraphic study

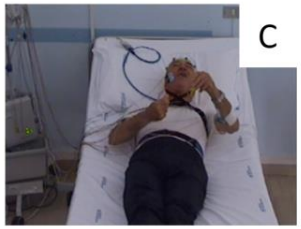
Giuseppe d'Orsi, Tommaso Martino, Alessandra Lalla, Maria Teresa Di Claudio, Elena Carapelle, Carlo Avolio

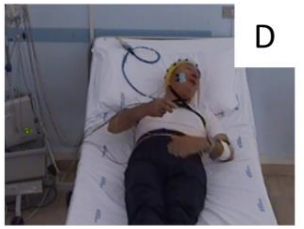
Epilepsy Centre - Clinic of Nervous System Diseases, Riuniti Hospital, Foggia, Italy

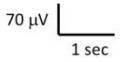
Epileptic Disord 2018; 20 (6): 525-9







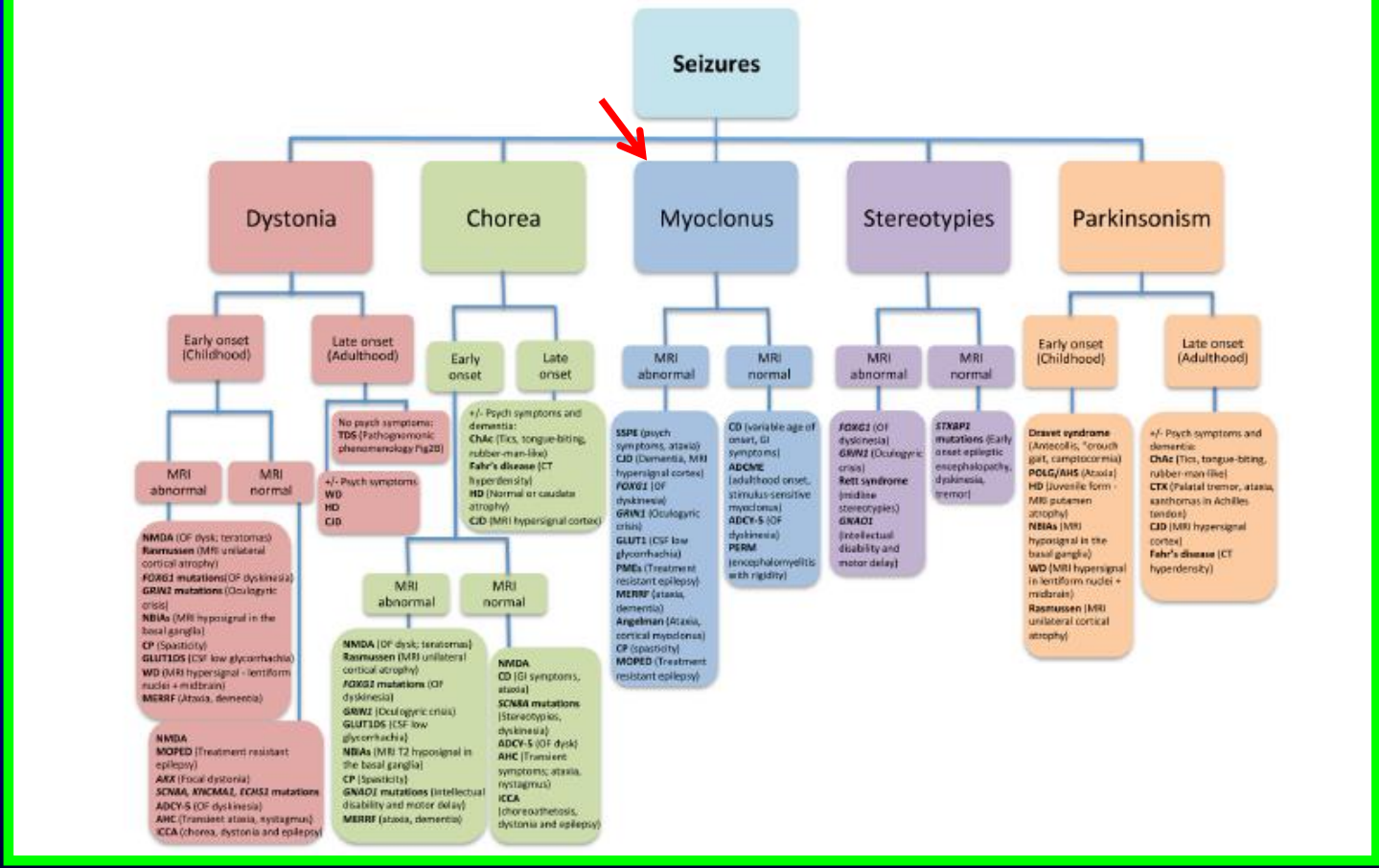




REVIEW *J Neurol Neurosurg Psychiatry* 2019;**90**:920–928. doi:10.1136/jnnp-2018-320039

Seizures and movement disorders: phenomenology, diagnostic challenges and therapeutic approaches

Maria Eliza Freitas,[1] Marta Ruiz-Lopez,[2] Josep Dalmau,[3] Roberto Erro,[4] Michael Privitera,[5] Danielle Andrade,[6] Alfonso Fasano[7]



CLINICA
EPILESSIA MIOCLONICA PROGRESSIVA

- Mioclono
- Crisi epilettiche
- Deterioramento neurologico progressivo

CLASSIFICAZIONE
EPILESSIA MIOCLONICA PROGRESSIVA

- Malattia di Unverricht-Lundborg
- Malattia di Lafora
- MERRF
- Ceroido lipofuscinosi neuronale
- Sialidosi

Disturbo del Movimento

Altre Crisi Epilettiche ed Epilessie

Le Crisi e le Epilessie del Lobo Frontale

Manifestazioni Parossistiche NON Epilettiche

Psicogene

Psycogenic Non-Epileptic Seizures

Psycogenic Non-Epileptic Seizures

- Ritardo di diagnosi di PNES di 7 anni (Reuber et al, Neurology 2002: 58: 493-495)
- 80% di pazienti con conferma diagnostica attraverso Video-EEG assumevano almeno un farmaco anti-epilettico al momento della diagnosi di PNES (Benbadis, Eur Neurol 1999: 41:114-115)
- Circa 25% dei pazienti inviati ad un monitoraggio Video-EEG per epilessia farmacoresistente hanno PNES (Smith et al, QJM 1999: 92:15-23)
- Prima di una diagnosi di PNES, costi sanitari di ~20,995 euro (Magee et al, Epilepsy Behav 2014; 33: 45-48)

Psycogenic Non-Epileptic Seizures

Presentazione - Andamento clinico

- **Durata media**: PNES (<1-150 min) > ES (≤ 2 min)

  Sensibilità 67%, Specificità 48%

- **Esordio**: Graduale

- **Decorso**: fluttuante (waxing-waning) →

  Sensibilità 69%, Specificità 96%

A.Avbersek and S.Sisodiya , JNNP, 2010; 81: 719-725

Epileptic Seizures

FENOMENI ACUSTICI

- **Crisi Focali**
  - Vocalizzazioni
  - Aumentata intensità all'inizio della vocalizzazione nelle crisi «frontali»
- **Crisi Tonico-Cloniche Generalizzate**
  - Urlo, Suono Laringeo (Sensibilità 85%, Specificità 100%).
  - Respiro Stertoroso (Sensibilità 22-93%, Specificità 50-100%).

Elzawahry et al., Epilepsy & Behavior 2010; Hartl et al., Seizure 2018

Psycogenic Non-Epileptic Seizures

FENOMENI ACUSTICI

- 3,7-8,9% PNES
- Pianti (Sensibilità 5-32%, Specificità 91-100%), gemiti, tosse, urla prolungate…
- Andamento fluttuante.
- Persistenza, aumento in intensità.

Chen et al, Curr Neurol Neurosci Rep 17: 71, 2017

A.Avbersek and S.Sisodiya , JNNP, 2010; 81: 719-725

Psycogenic Non-Epileptic Seizures

«Pattern Motori»

- **Scuotimento del capo** (side-to-side head shaking).

Andamento fluttuante.

- **Movimenti lato a lato** (side-to-side body movement)

Sensibilità 25-63%, Specificità 96-100%

- **Movimenti Asincroni** (asynchronous limb movements),

Sensibilità 44-96%, Specificità 93-96%

Chen et al, Curr Neurol Neurosci Rep 17: 71, 2017

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PNES - TRAPPOLE DIAGNOSTICHE

- PNES misdiagnosticate come ES in soggetti portatori di patologie cerebrali organiche
- PNES in presenza di lesioni cerebrali "potenzialmente epilettogene" (ad es. *TLMS*)
- PNES + ES
- ES misdiagnosticate come PNES….

Fenomeni Motori Parossistici…Video-EEG!