



4° CORSO RESIDENZIALE EEG e POTENZIALI EVOCATI

22 – 27 NOVEMBRE 2021

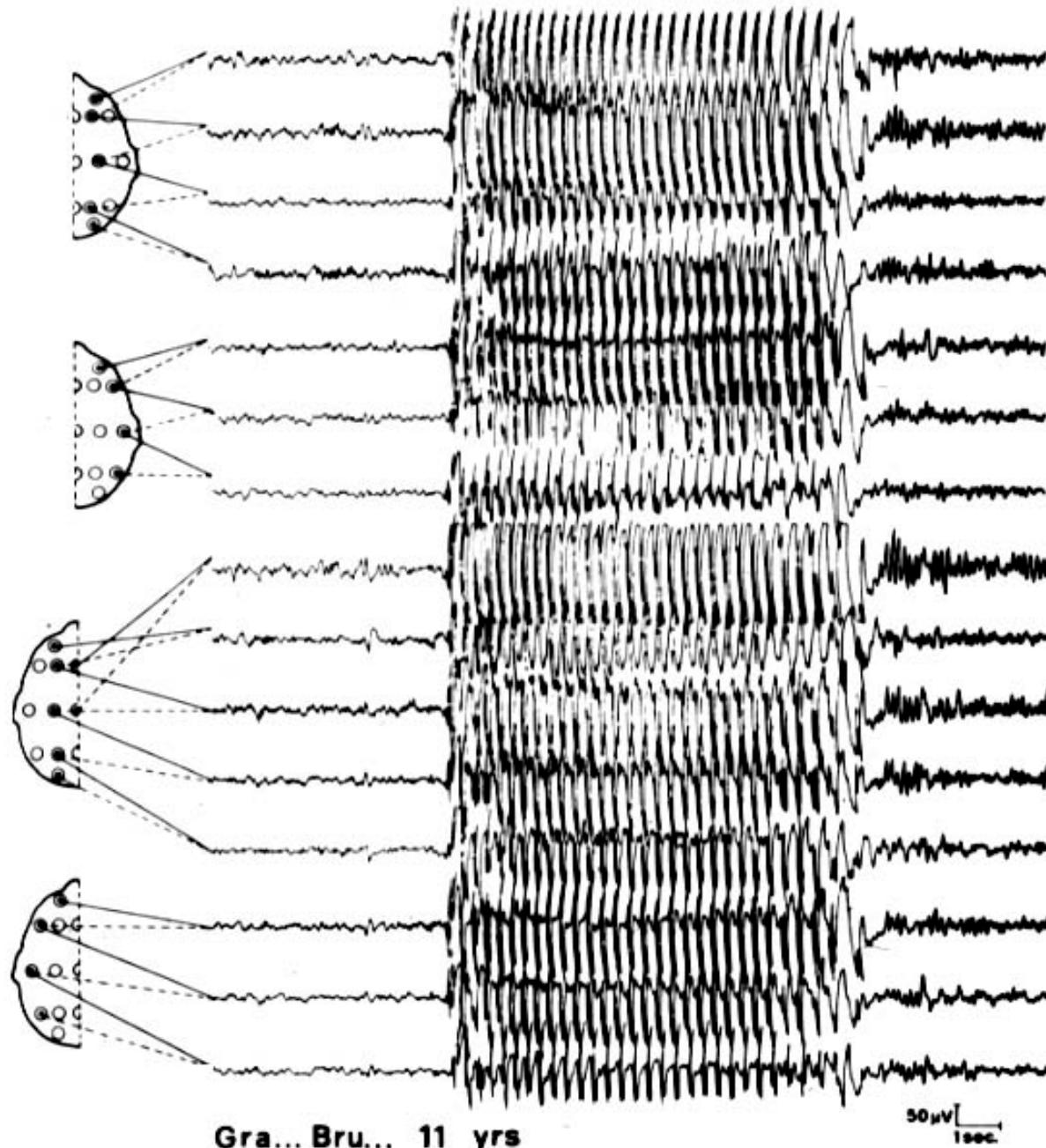
L'EEG nelle epilessie generalizzate



Paolo Tinuper



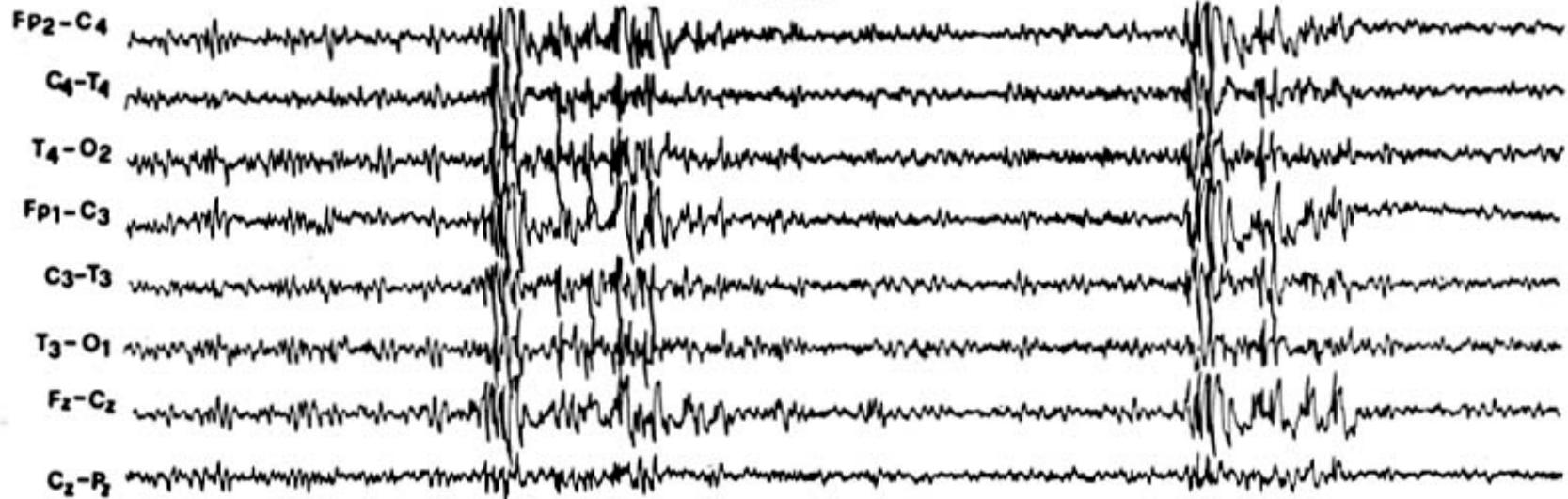
*IRCCS Istituto delle Scienze Neurologiche di Bologna.
Dipartimento di Scienze Biomediche e Neuromotorie. Università di
Bologna*



Gra... Bru... 11 yrs

50 μV
1 sec.

Awake



EOG

Mylo EMG

R.Delt.

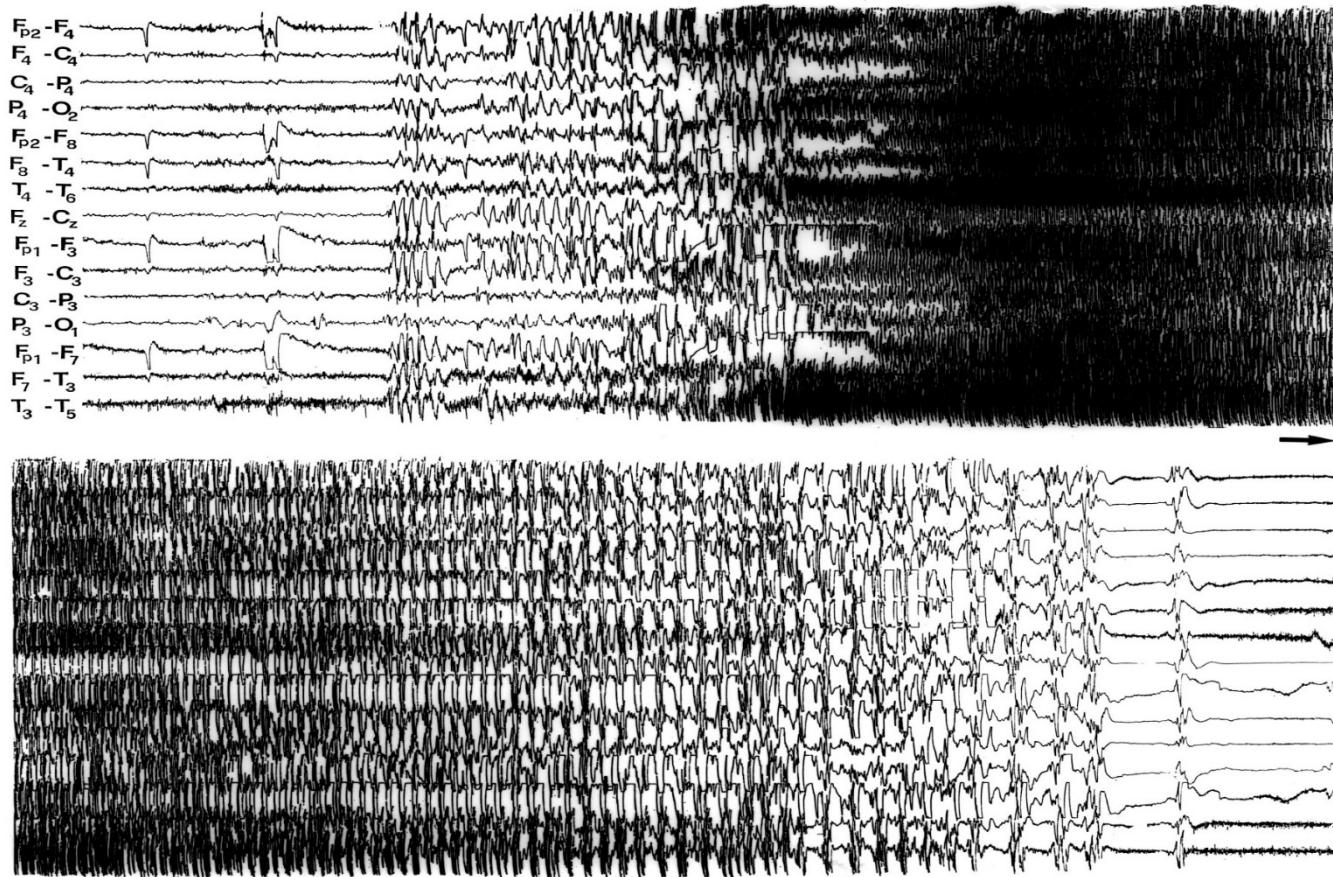
L.Delt.

ECG

Thor. Resp.

A.G. 14 yrs

50 μ V
1 sec.



Van.Ali. ♀ 19yrs

n°83661

50 μ V 1 sec.

ILAE Classification Task Force 2013-7



Torbjörn Tomson, Emilio Perucca, Ingrid Scheffer, Jackie French, Yue-Hua Zhang
Satish Jain, Gary Mathern, Sam Wiebe, Edouard Hirsch, Sameer Zuberi, Nico Moshe

Operational classification of seizure types by the International League Against Epilepsy: Position Paper of the ILAE Commission for Classification and Terminology

*Robert S. Fisher, †J. Helen Cross, ‡Jacqueline A. French, §Norimichi Higurashi, ¶Edouard Hirsch, #Floor E. Jansen, **Lieven Lagae, ††Solomon L. Moshé, ‡‡Jukka Peltola, §§Eliane Roulet Perez, ¶¶Ingrid E. Scheffer, and #**Sameer M. Zuberi

Epilepsia, **(*):1–9, 2017
doi: 10.1111/epi.13670

ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology

^{1,2,3}Ingrid E. Scheffer, ¹Samuel Berkovic, ⁴Giuseppe Capovilla, ⁵Mary B. Connolly, ⁶Jacqueline French, ⁷Laura Guilhoto, ^{8,9}Edouard Hirsch, ¹⁰Satish Jain, ¹¹Gary W. Mathern, ¹²Solomon L. Moshé, ¹³Douglas R. Nordli, ¹⁴Emilio Perucca, ¹⁵Torbjörn Tomson, ¹⁶Samuel Wiebe, ¹⁷Yue-Hua Zhang, and ^{18,19}Sameer M. Zuberi

Epilepsia, **(*):1–10, 2017
doi: 10.1111/epi.13709

Co-morbidities

Seizure types

Focal onset

Generalized onset

Unknown onset

Etiology

Structural

Genetic

Infectious

Metabolic

Immune

Unknown

Epilepsy types

Focal

Generalized

Combined
Generalized
& Focal

Unknown

Epilepsy Syndromes

Focal seizures:

- originating within networks limited to one hemisphere. These may be discretely localised or more widely distributed....

Generalised seizures

- originating at some point within, and rapidly engaging, bilaterally distributed networks. ...can include cortical and subcortical structures, but not necessarily include the entire cortex.

ILAE 2017 Classification of Seizure Types Expanded Version ¹

Focal Onset

Aware

Impaired Awareness

Motor Onset

automatisms

atonic²

clonic

epileptic spasms²

hyperkinetic

myoclonic

tonic

Nonmotor Onset

autonomic

behavior arrest

cognitive

emotional

sensory

focal to bilateral tonic-clonic

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

Generalized Onset

Motor

tonic-clonic
clonic
tonic
myoclonic
myoclonic-tonic-clonic
myoclonic-atonic
atonic
epileptic spasms

Nonmotor (absence)

typical
atypical
myoclonic
eyelid myoclonia

focal to bilateral tonic-clonic

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

Generalized Onset

Motor

tonic-clonic
clonic
tonic
myoclonic
myoclonic-tonic-clonic
myoclonic-ataxic
ataxic
epileptic spasms

Nonmotor (absence)

typical
atypical
myoclonic
eyelid myoclonia

Unknown Onset

Motor

tonic-clonic
epileptic spasms

Nonmotor

behavior arrest

Unclassified ³

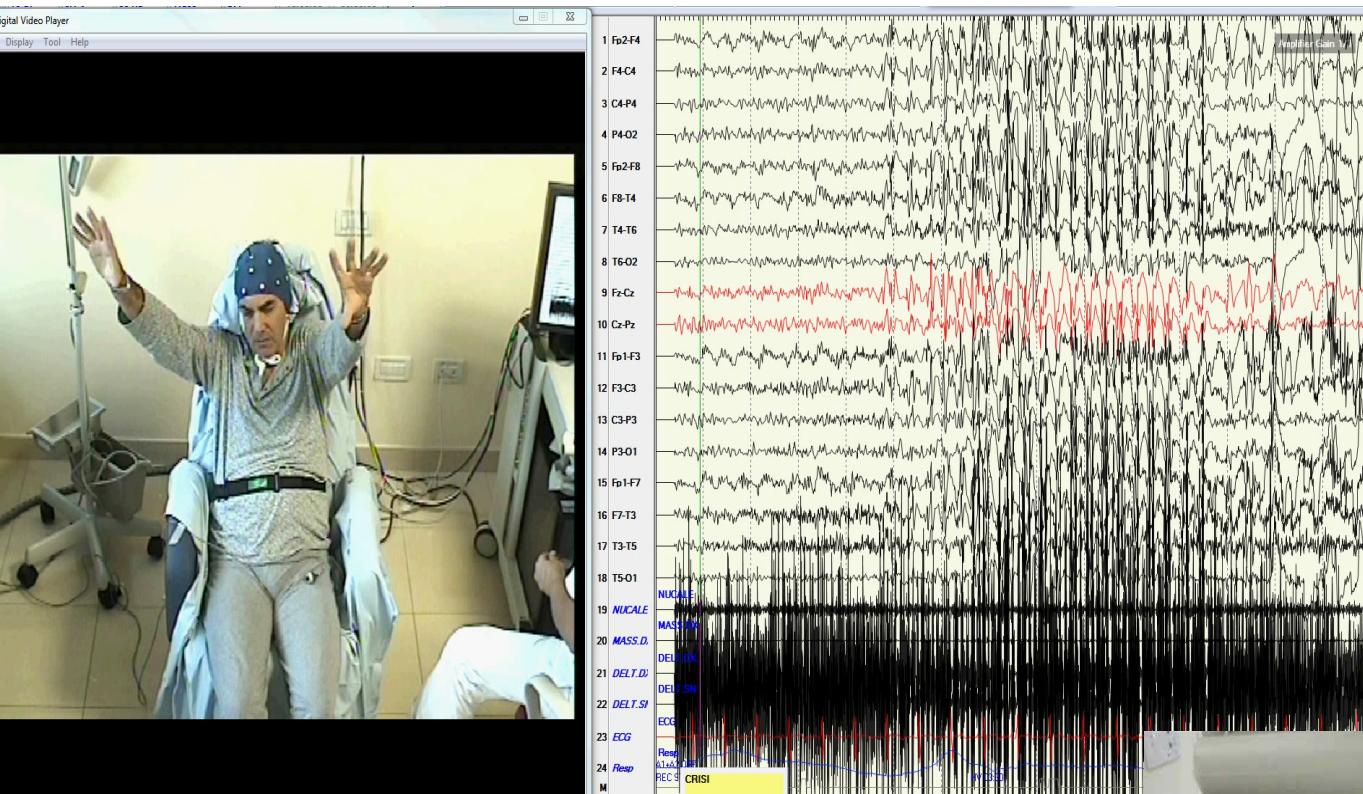
focal to bilateral tonic-clonic

Some seizures could be either focal or generalized onset

- **Clonic**
- **Myoclonic**
- **Tonic**
- **Atonic**
- **Spasms**
- **Tonic-clonic**
- **Absences ???**

Clonic seizure

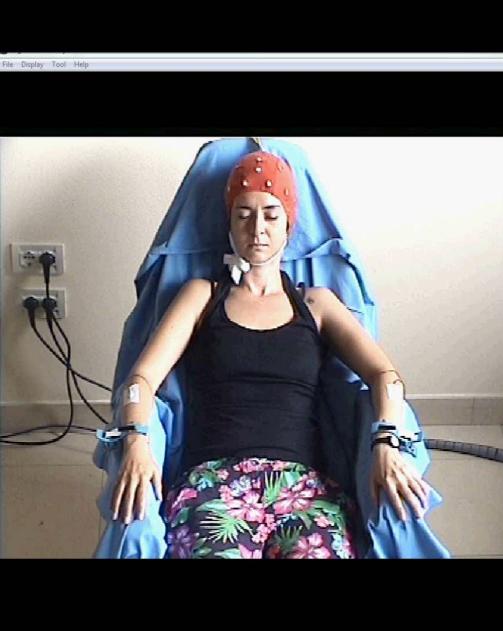
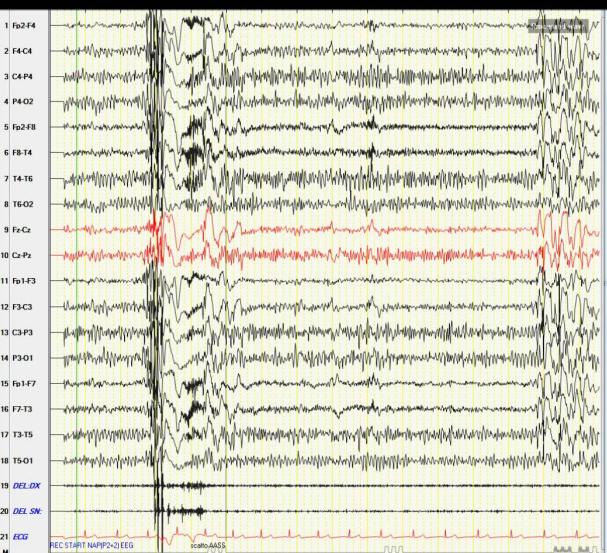
Generalized onset



Focal onset

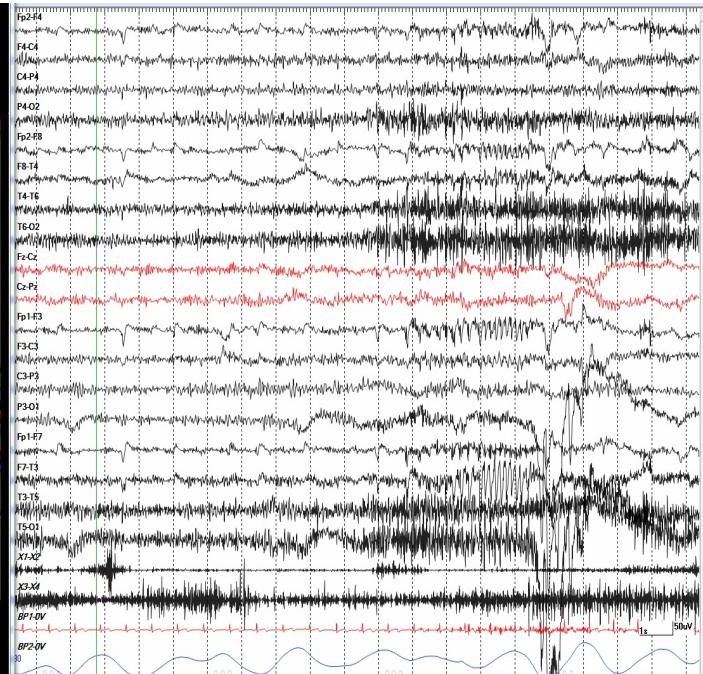


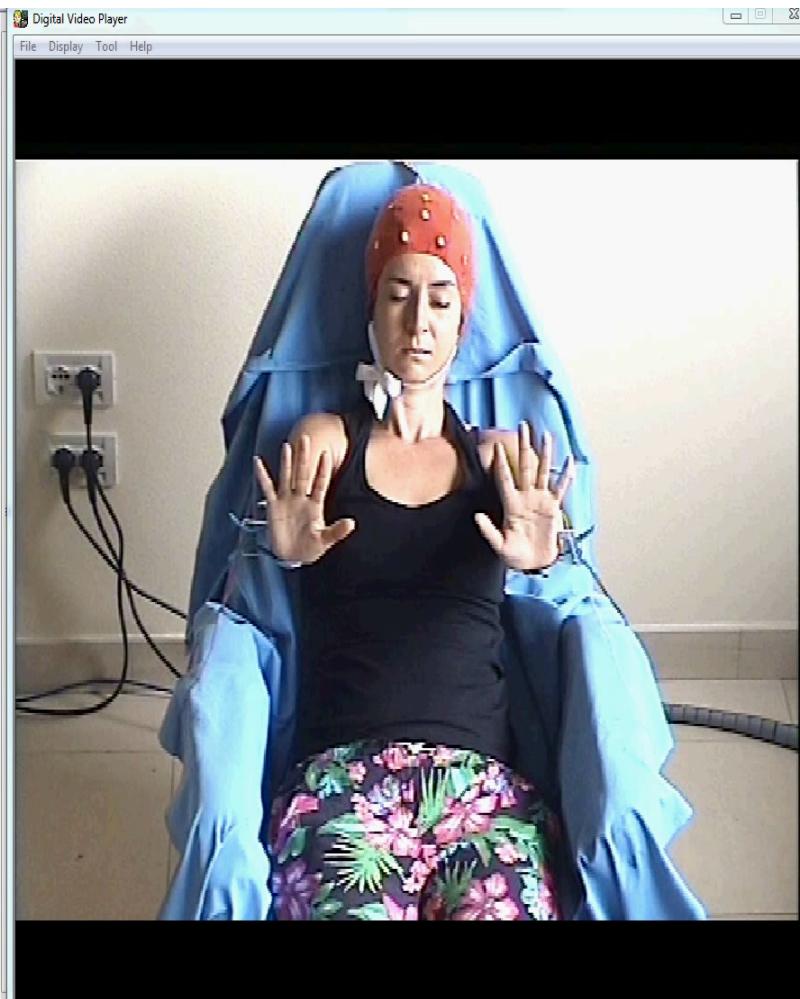
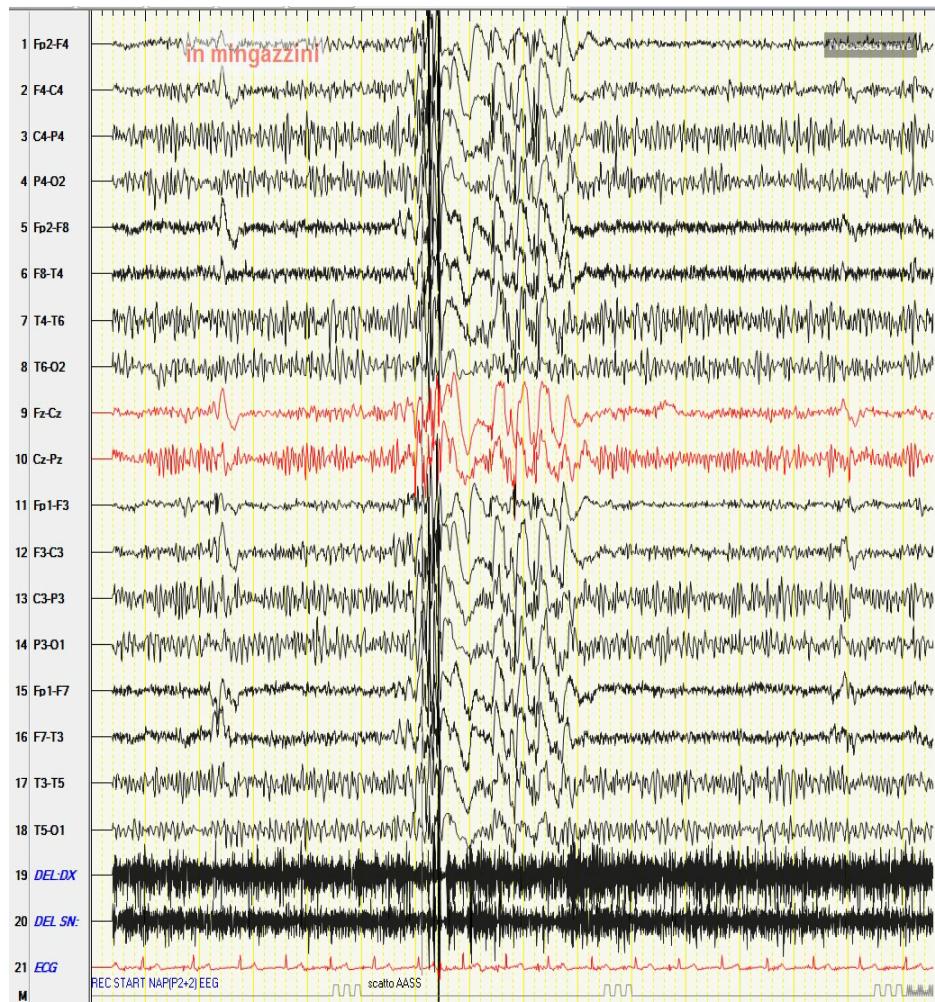
Generalized onset



Myoclonic seizure

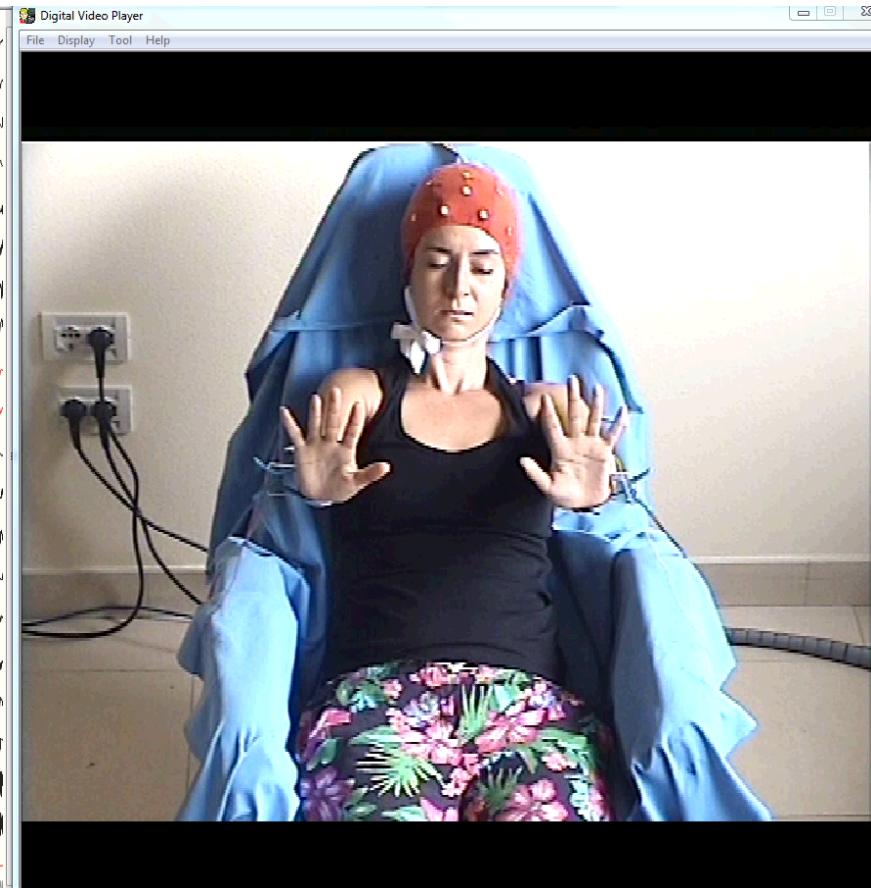
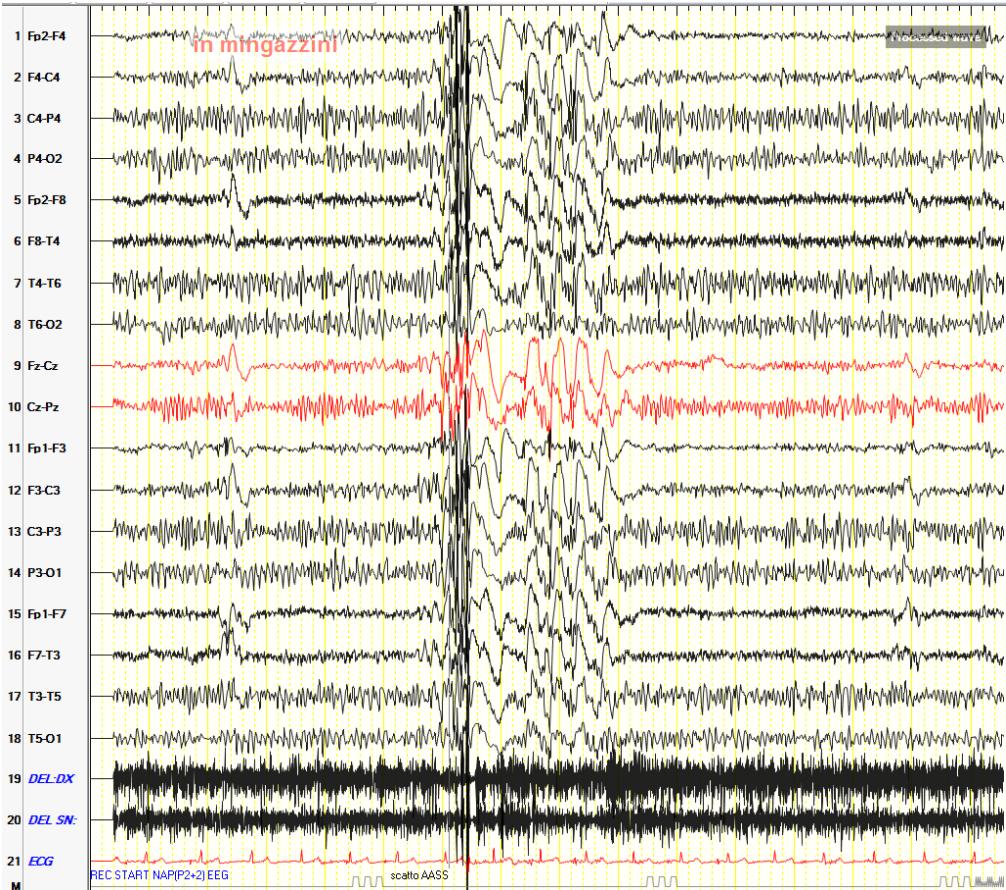
Focal onset



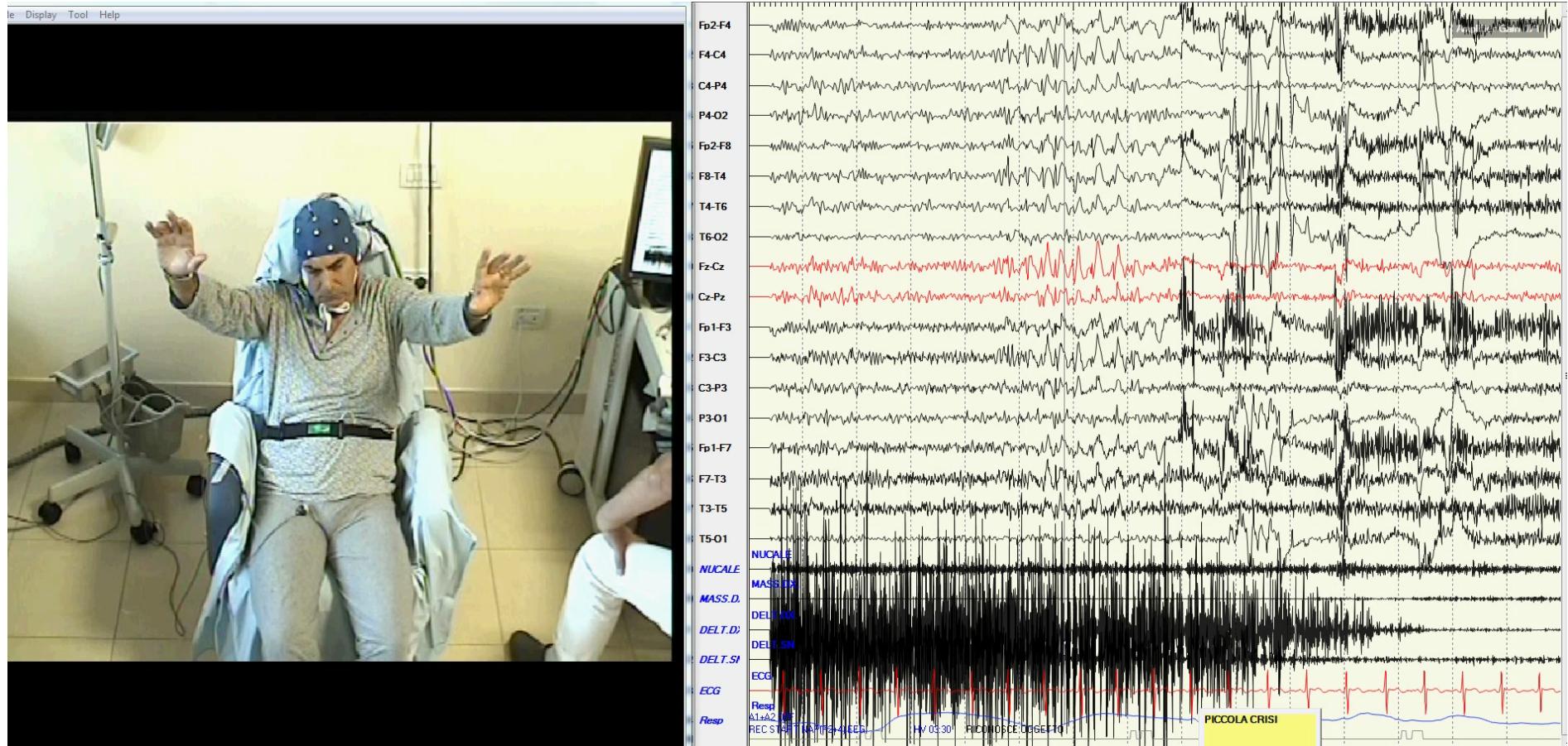


Le anomalie possono essere focali !!!!



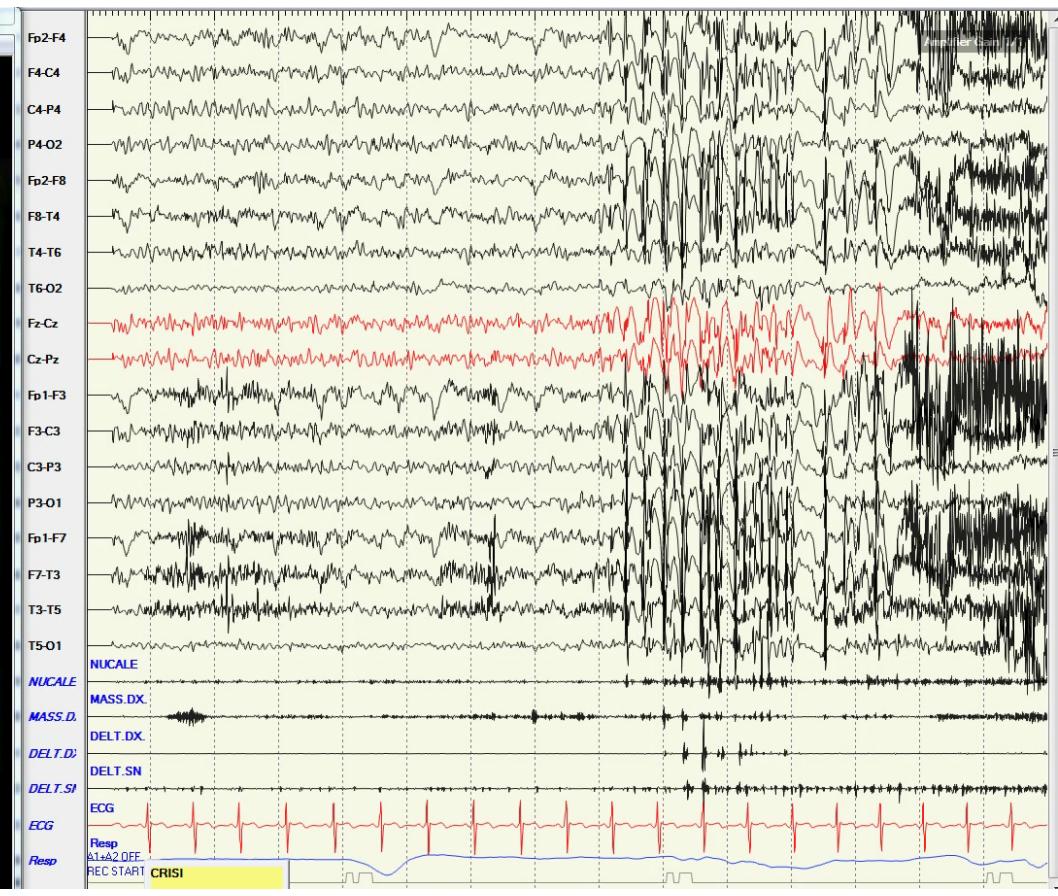




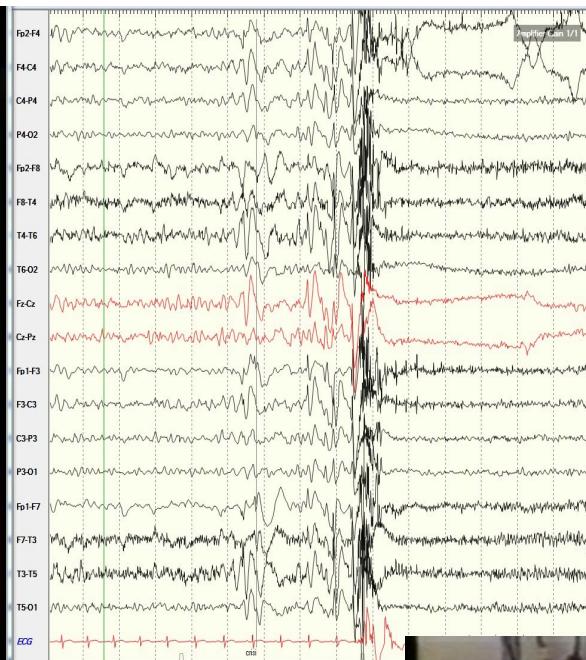


Digital Video Player

File Display Tool Help



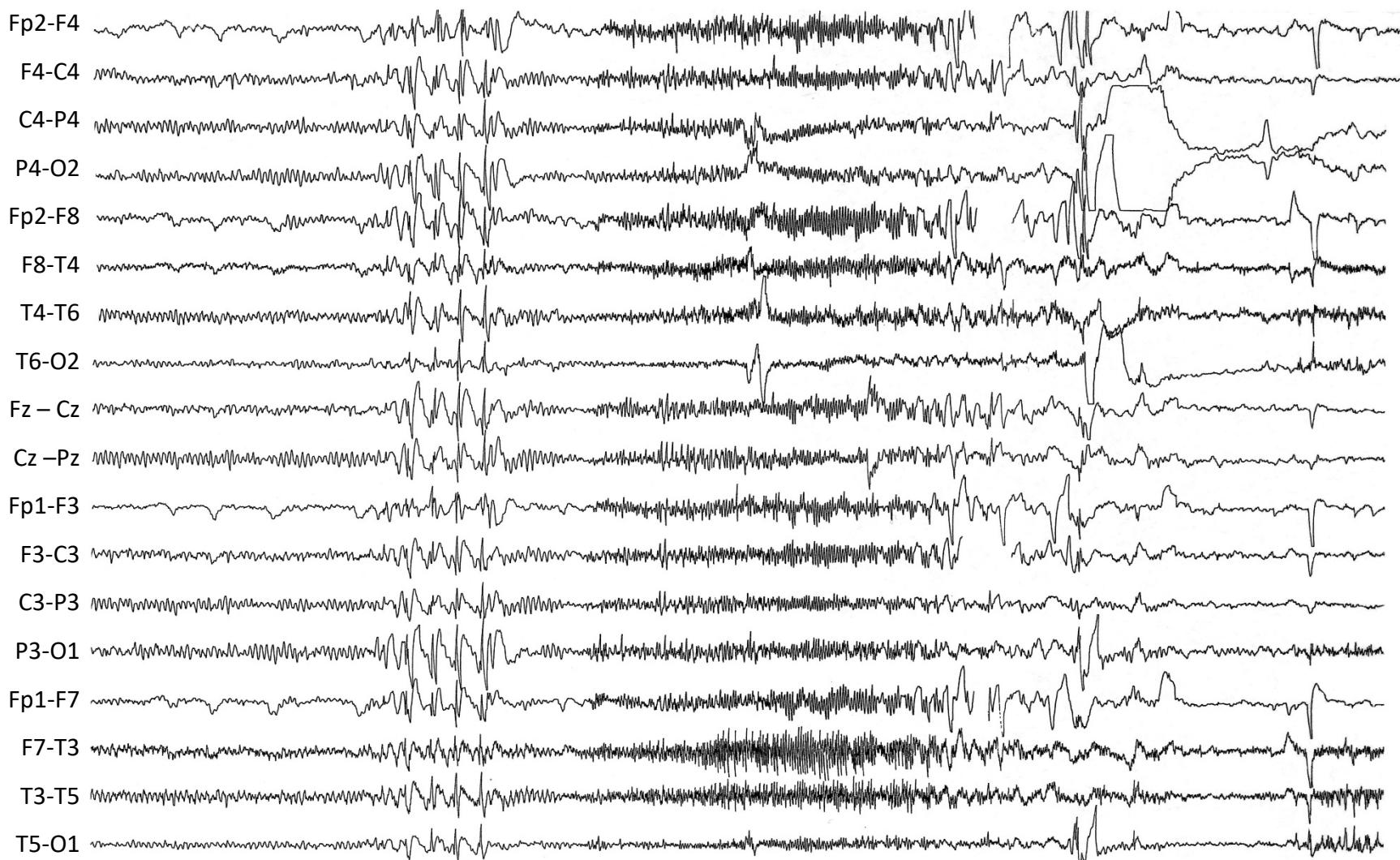
Generalized onset



Tonic seizure

Focal onset





13:56':05"
 00 supina AASS lungo i
 braccioli della poltrona
 capo verso dx

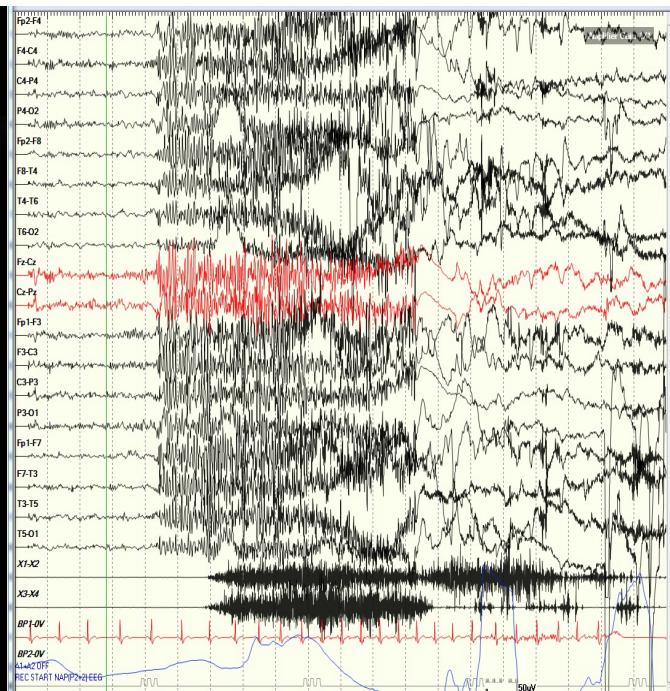
visibili mioclonie
 palpebrali

mostrate 3 dita non risponde
 stira angoli della bocca verso il
 basso allarga leggermente poi
 flette in avanti AASS solleva Al
 sn 00 capo leggermente in
 avanti

Mostrata penna non
 risponde riappoggia il capo
 alla poltrona e con le mani
 si sistema il pigiama

D. è passata ?
 R: sì
 tec: "apri gli occhi"
 pz: esegue

Generalized onset



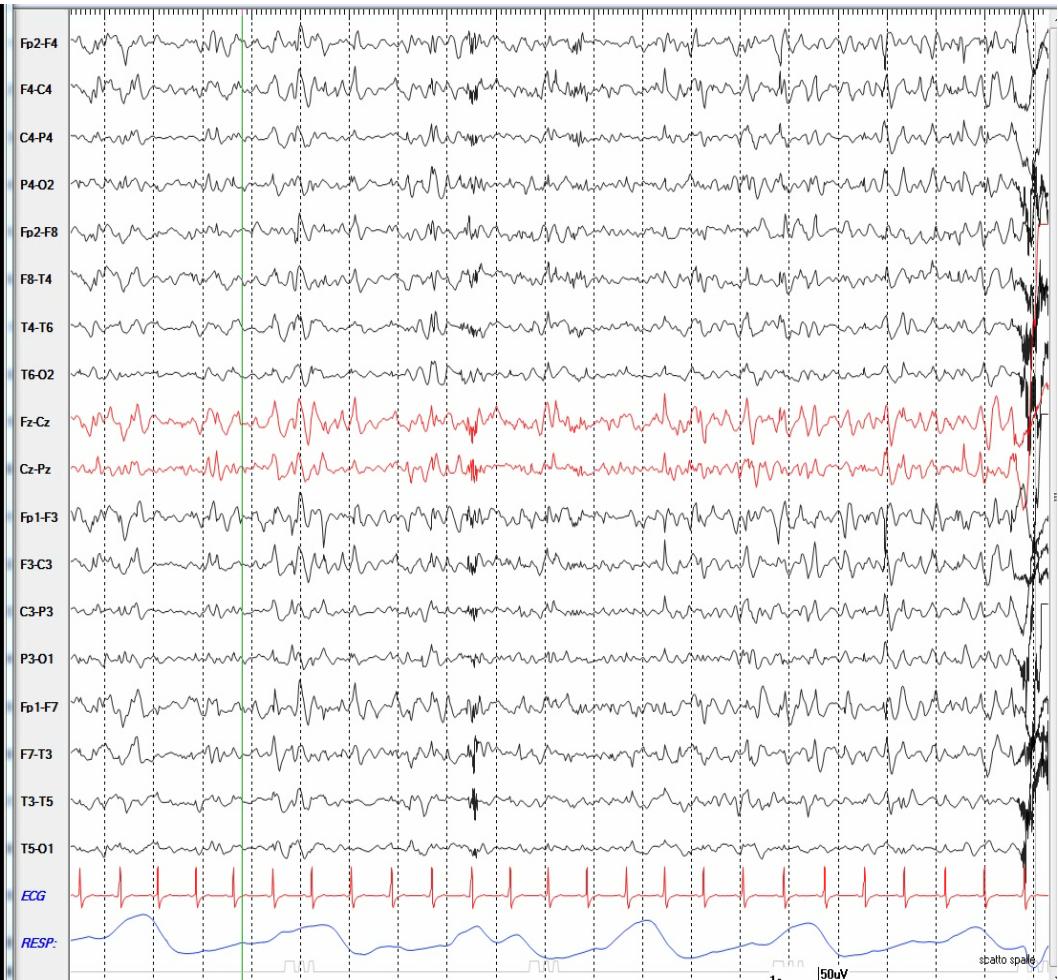
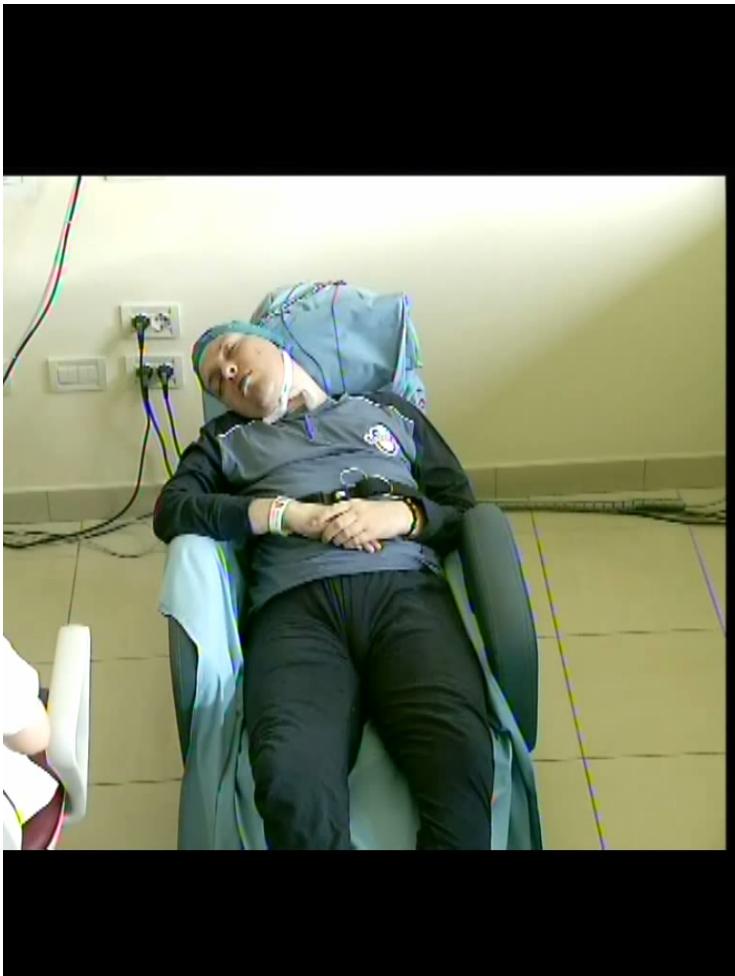
Tonic seizure



Focal onset

Tonic seizure

Generalized onset



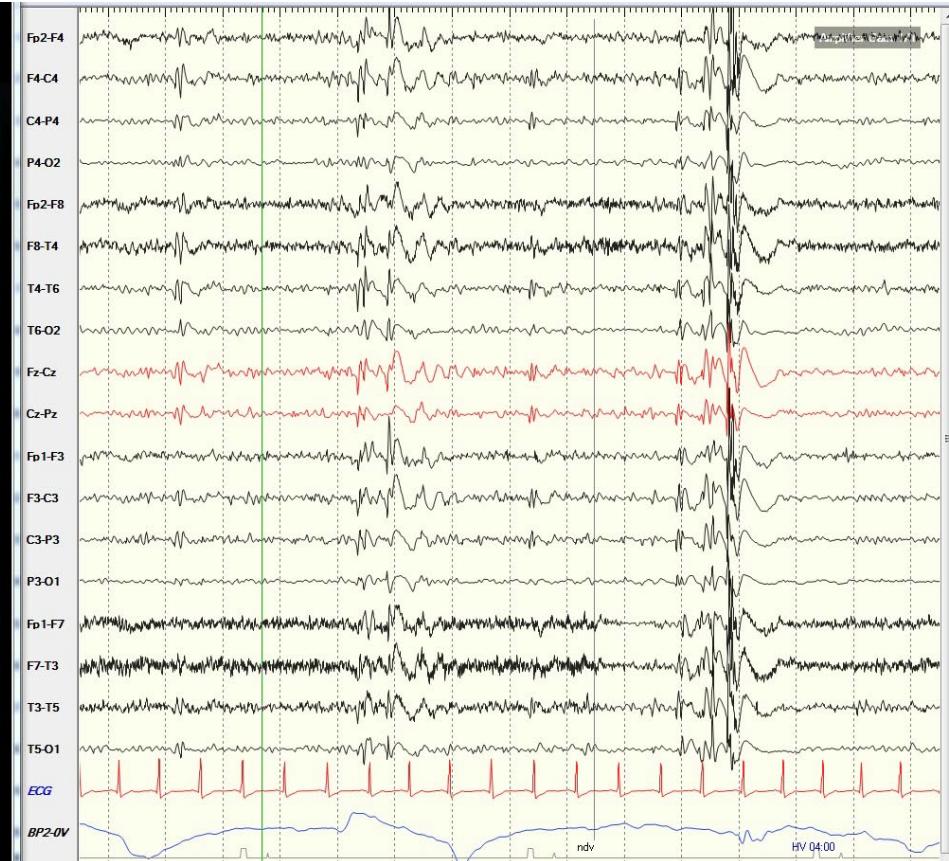
Tonic seizure

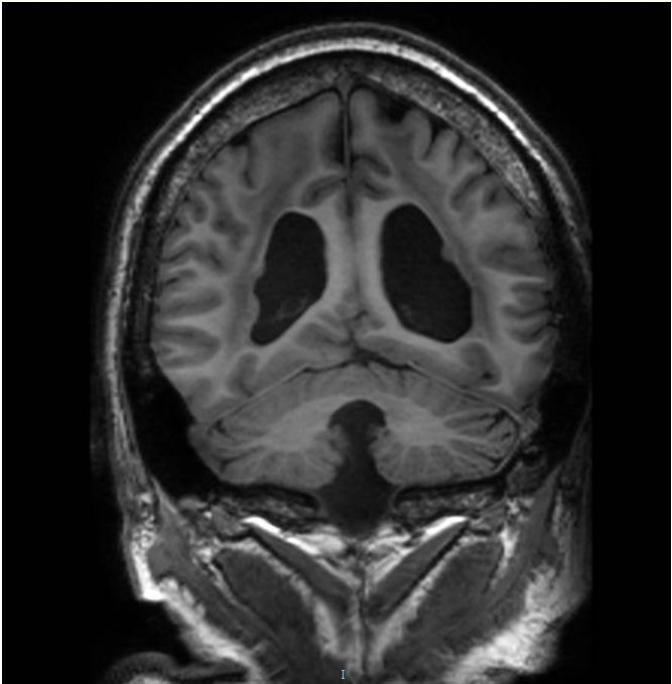
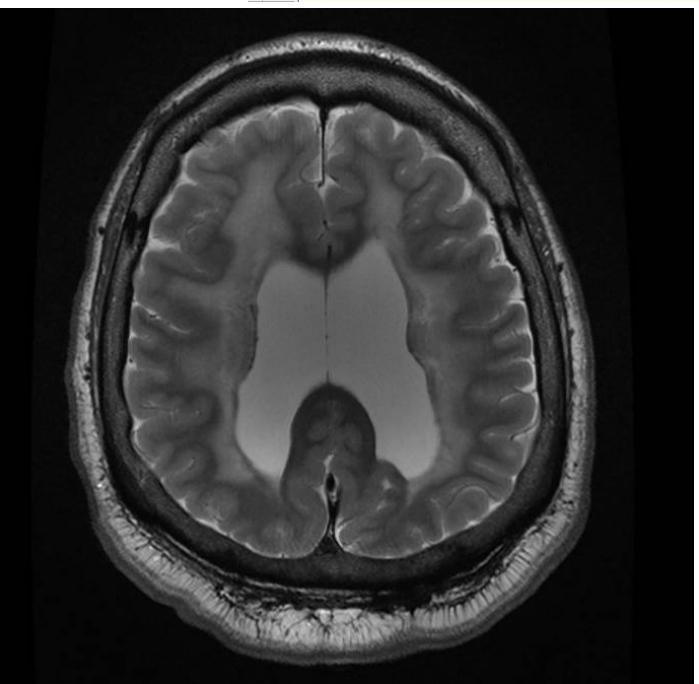
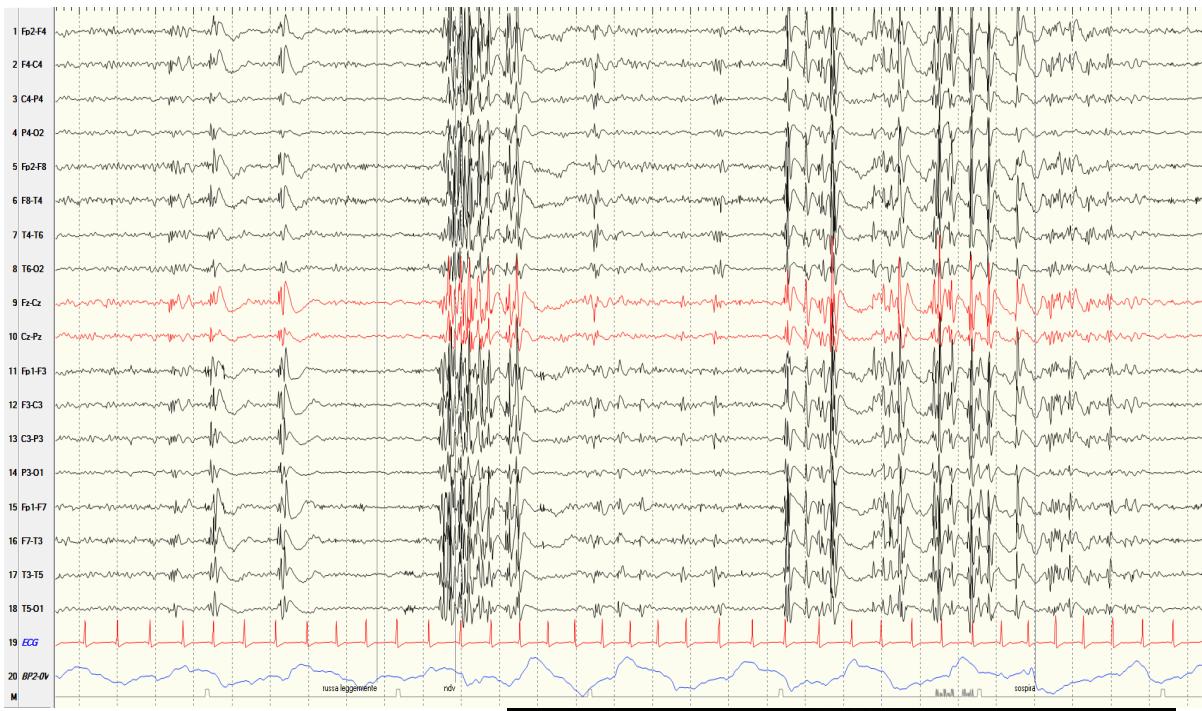
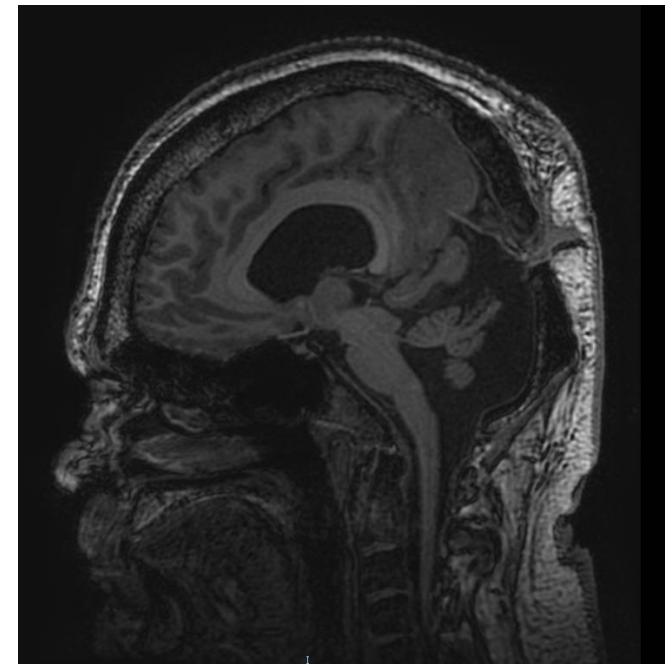
Focal onset



Epileptic spasm

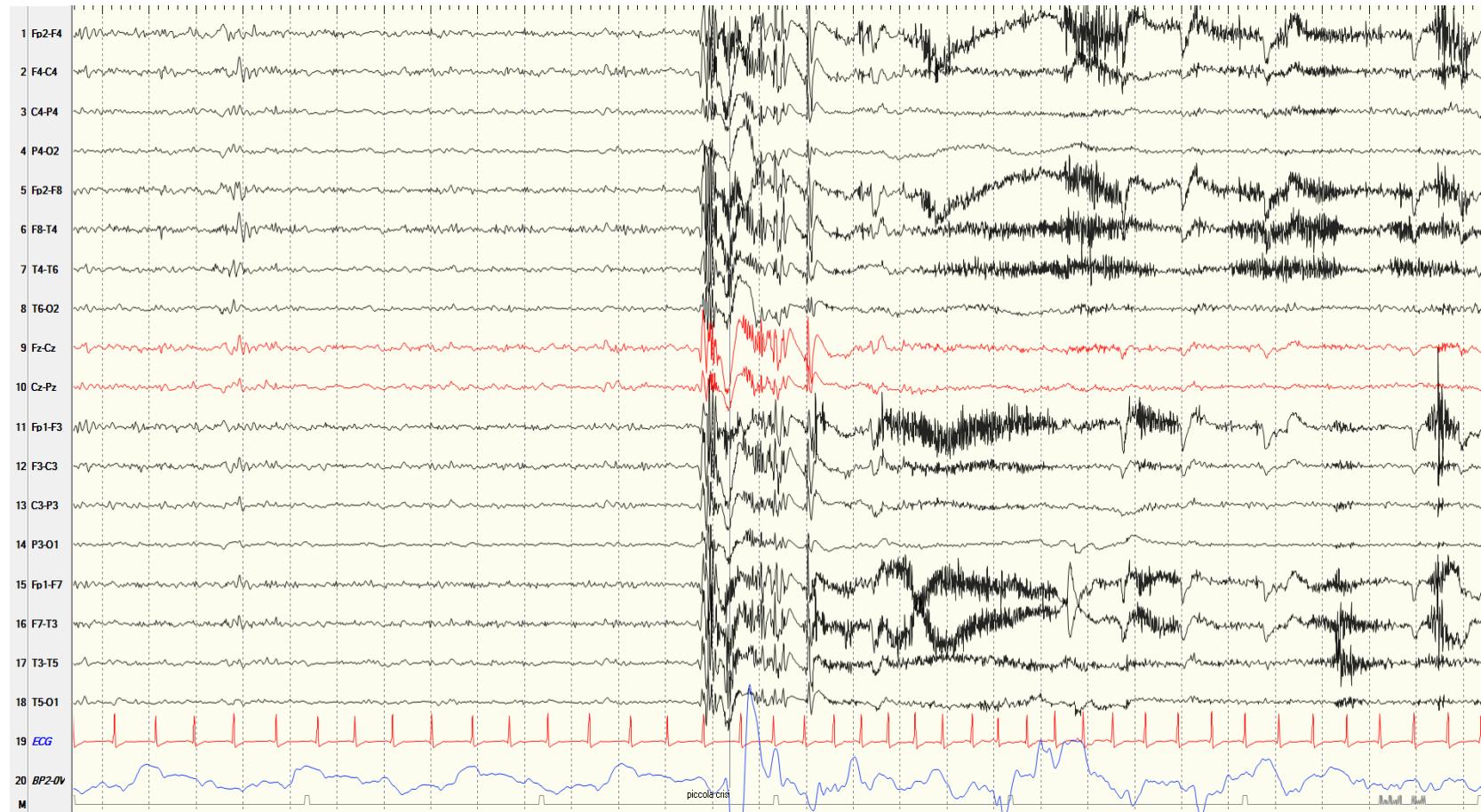
Generalized onset





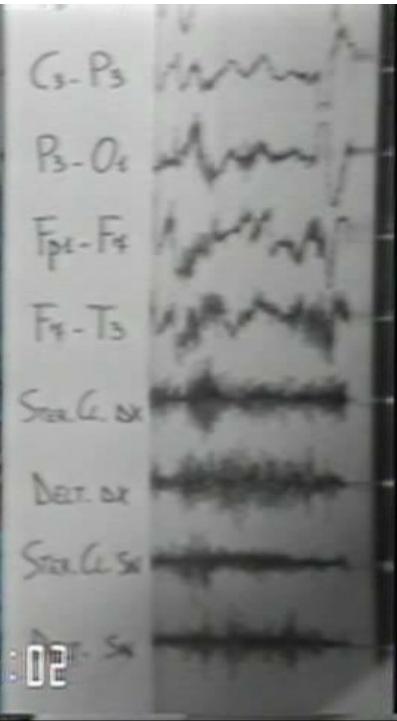
Picc. Gianni

Pic Gianni 2013

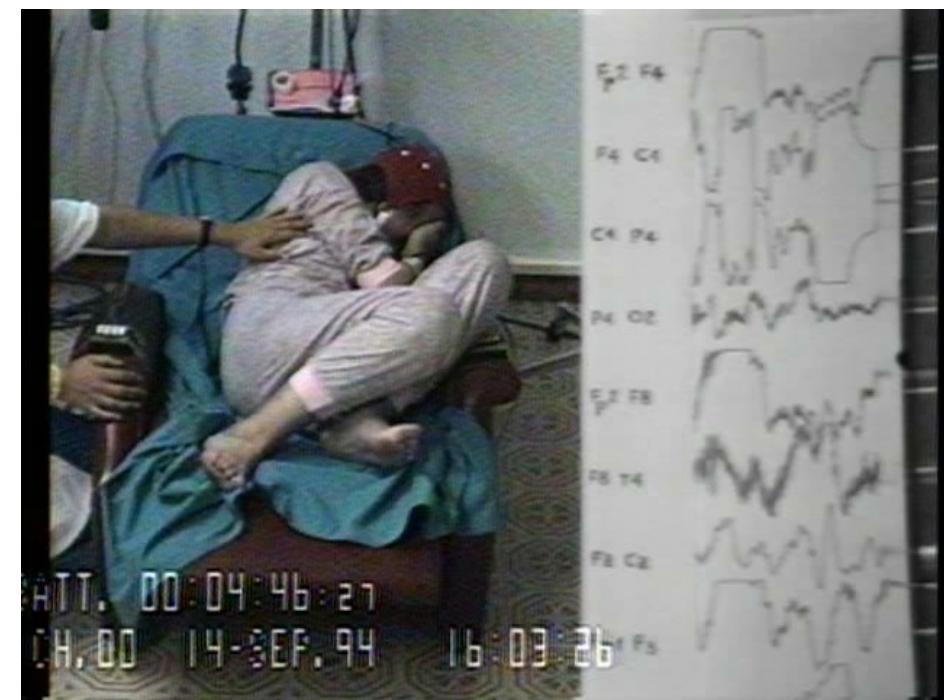


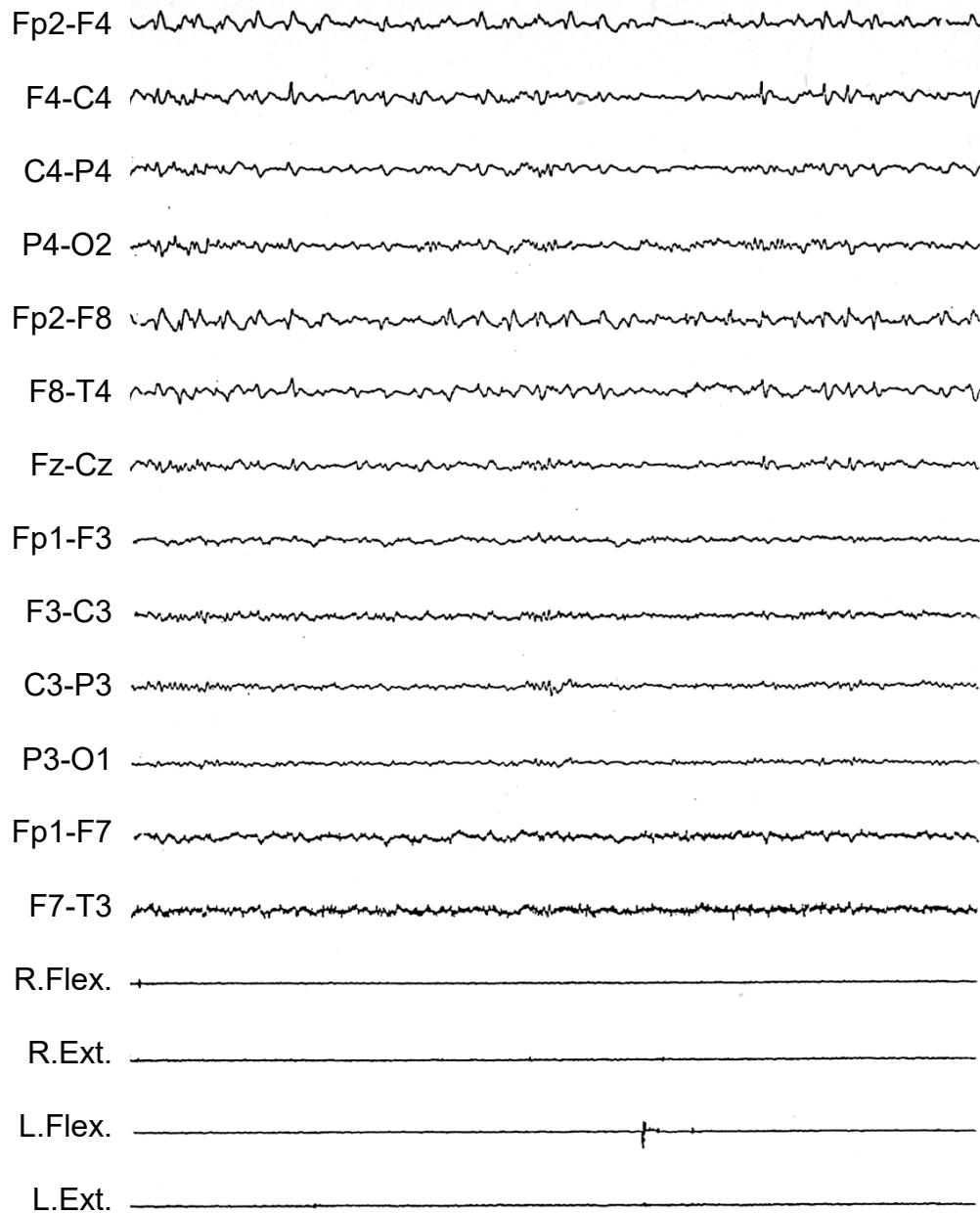
Epileptic spasm

Focal onset



M.P. 21 yrs

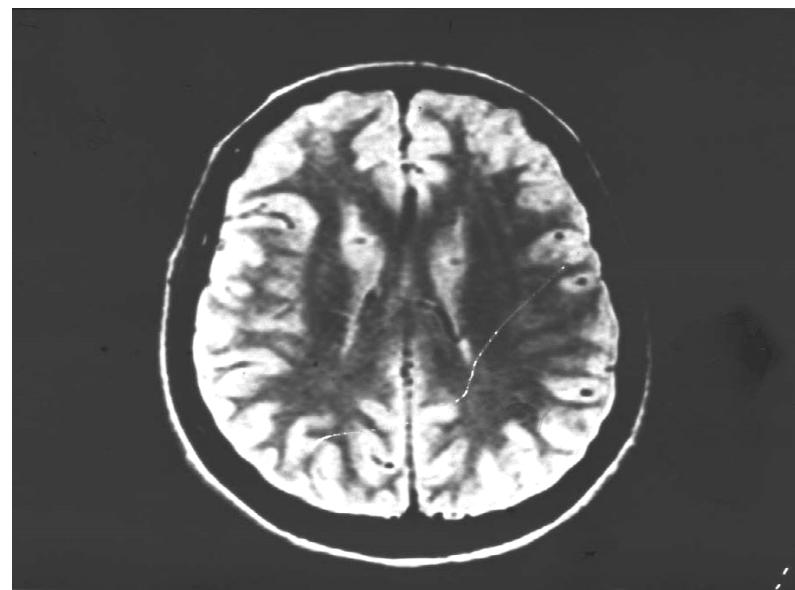
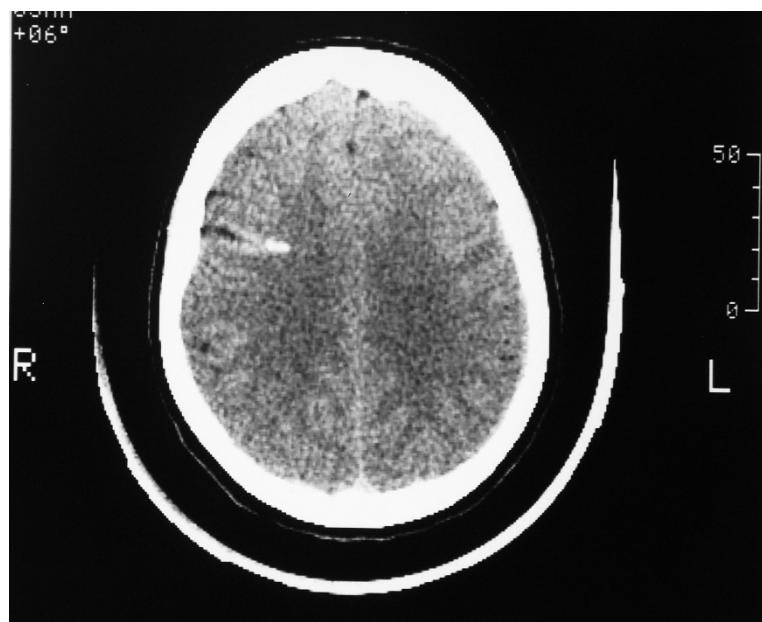
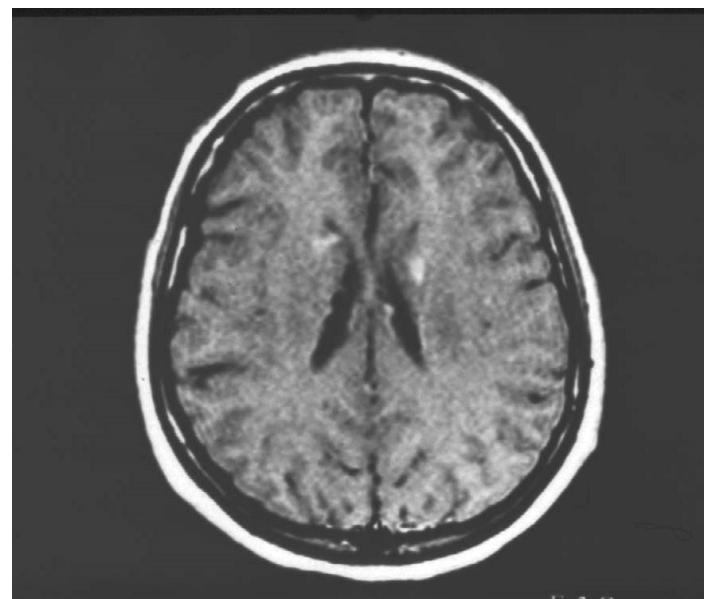
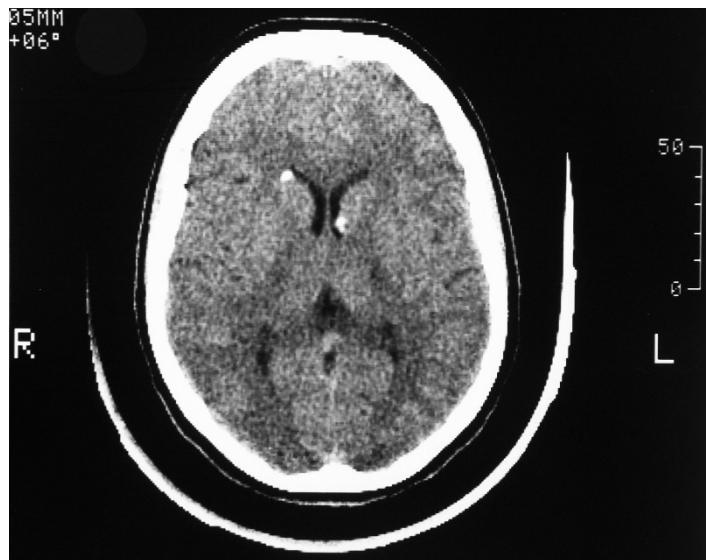




50 μ V T
1sec.

Mas . Pas. 21 aa





NMR: Cortical nodular formation in posterior R frontal, L cerebellar and sub ependymal regions. Focal R fronto-parietal eterotopia.

(M.P) female 21 yrs TUBEROUS SCLEROSIS (Bourneville' s disease)

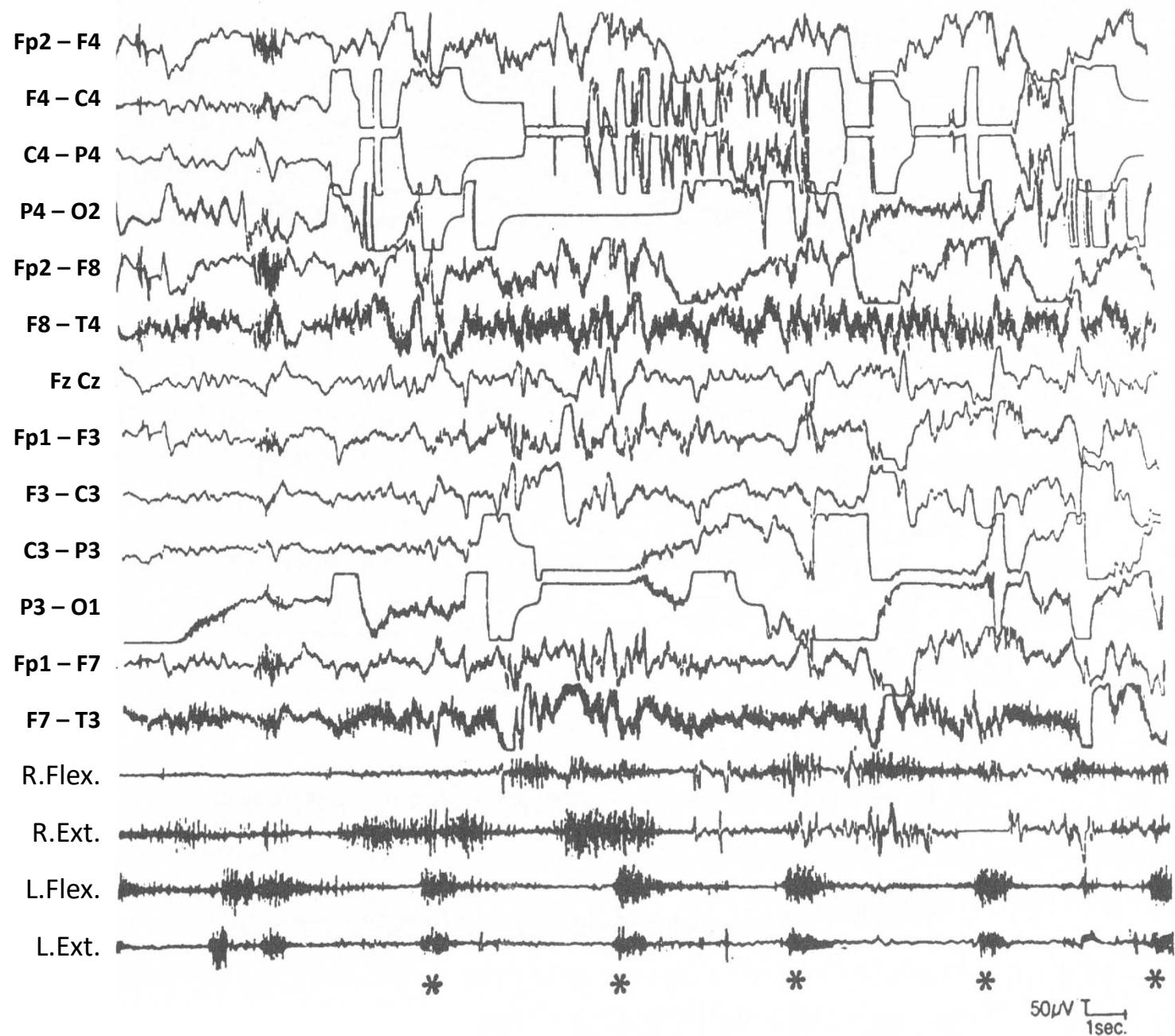
Since 1 months of life: upward deviation of eyes and “jerks” of the left limbs (ES?).

Since 14 yrs: asymmetrical spasms involving upper limbs, flexion of the head, facial grimace and LOC

Prolonged clusters every day, drug - resistant.



Severe mental deficit, left harm hypoplasia,
facial angiofibromas, shagreen patches,
cardiac rhabdomyoma, renal angiomyolipoma

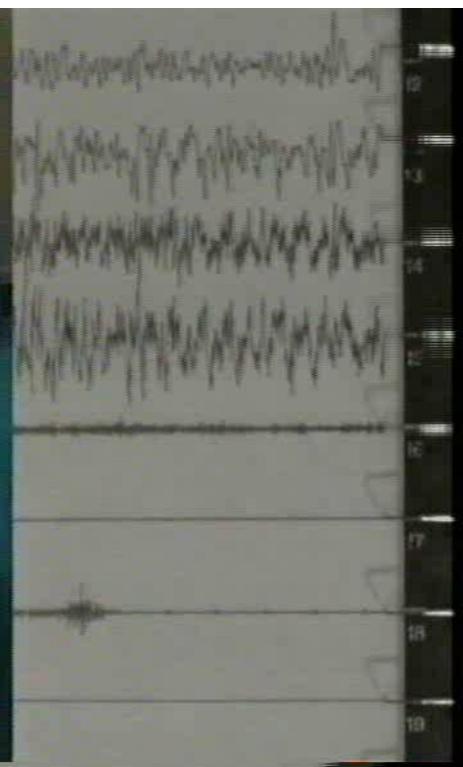


Mas . Pas. 21 yrs ♀

Tonic-clonic

Generalized onset

Focal onset





International League Against Epilepsy

Working toward a world where no person's life is limited by epilepsy



> Generalized seizure

CONVULSIVE

Clinical Overview

Videos

EEG

Differential diagnoses

Related syndromes

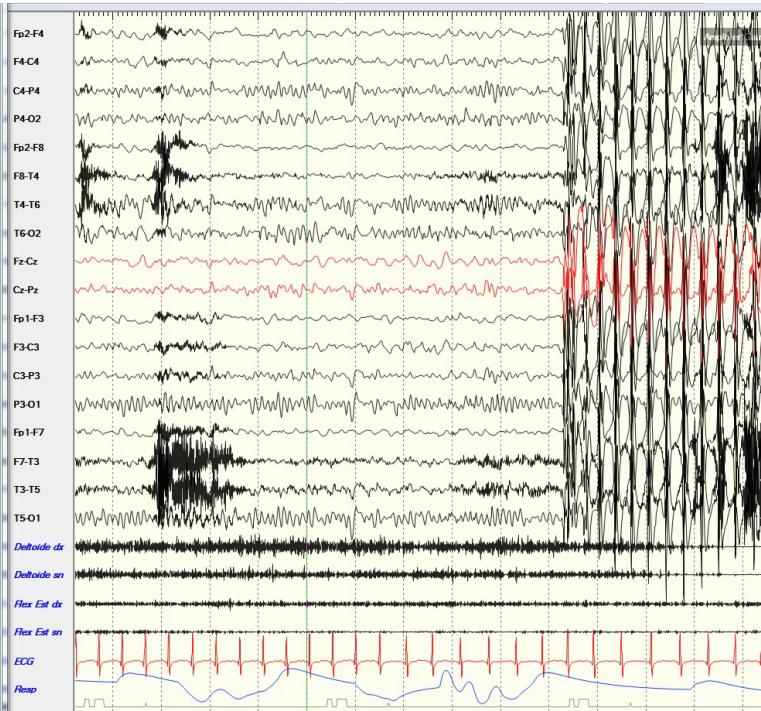
Generalized convulsive seizures are typically bilateral and symmetric although variants with asymmetry including head and eye deviation can be seen. A **tonic clonic** seizure is a seizure consisting of a tonic and a clonic phase, typically in this order, however variations such as **clonic-tonic-clonic** are also seen. A **clonic** seizure is a seizure involving bilaterally rhythmic jerking and may occur alone or in combination with **tonic** activity where there is bilaterally increased tone of the limbs typically lasting seconds to a minute. The jerking in a clonic seizure is more sustained and rhythmic than seen in a myoclonic seizure.

www.epilepsydiagnosis.org

Focal to bilateral tonic-clonic

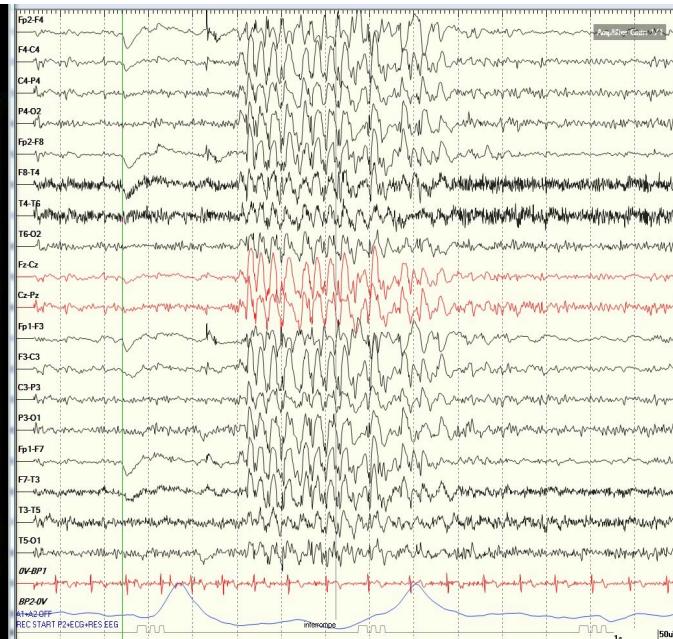


- **GENERALIZED**
- **FOCAL**
- **Clonic**
- **Myoclonic**
- **Tonic**
- **Atonic**
- **Spasms**
- **Tonic-clonic**
- **Absences ???**

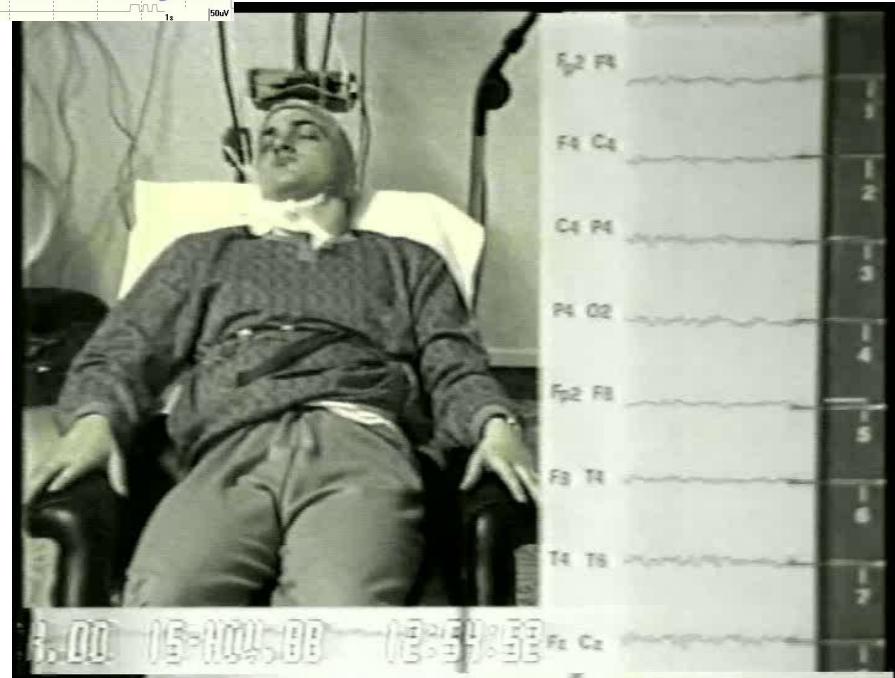


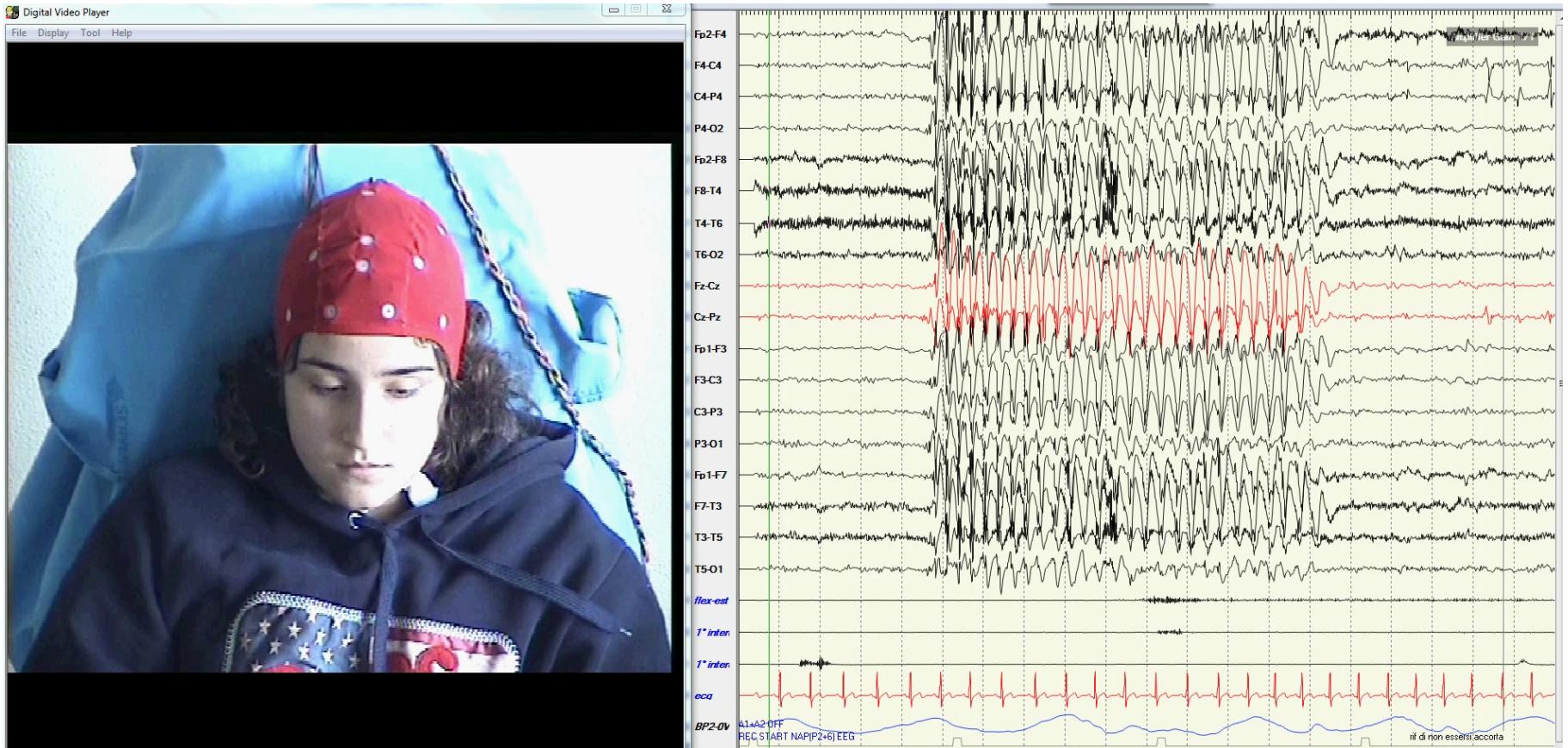
«Absence» seizure

Generalized onset

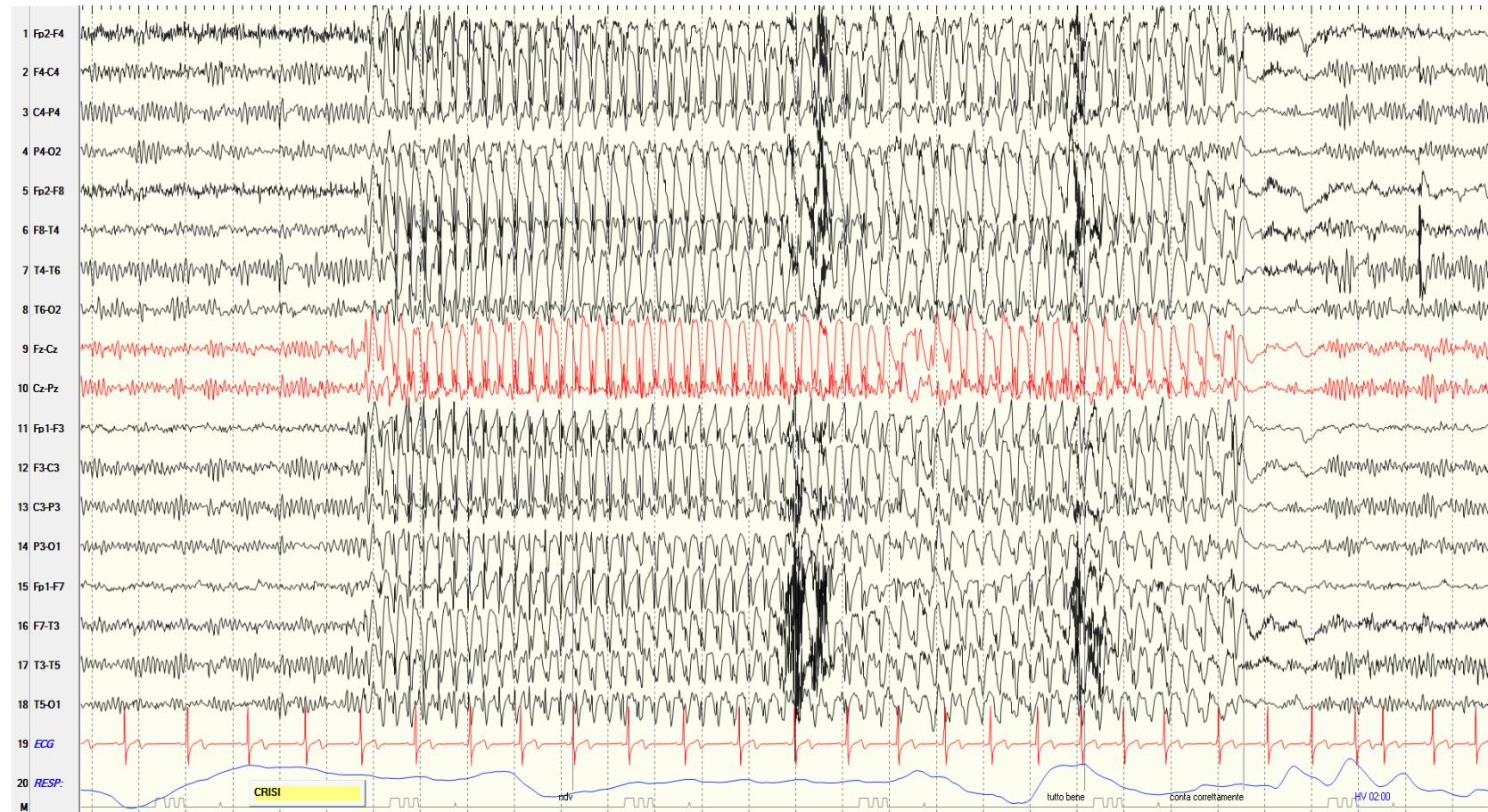


Focal onset

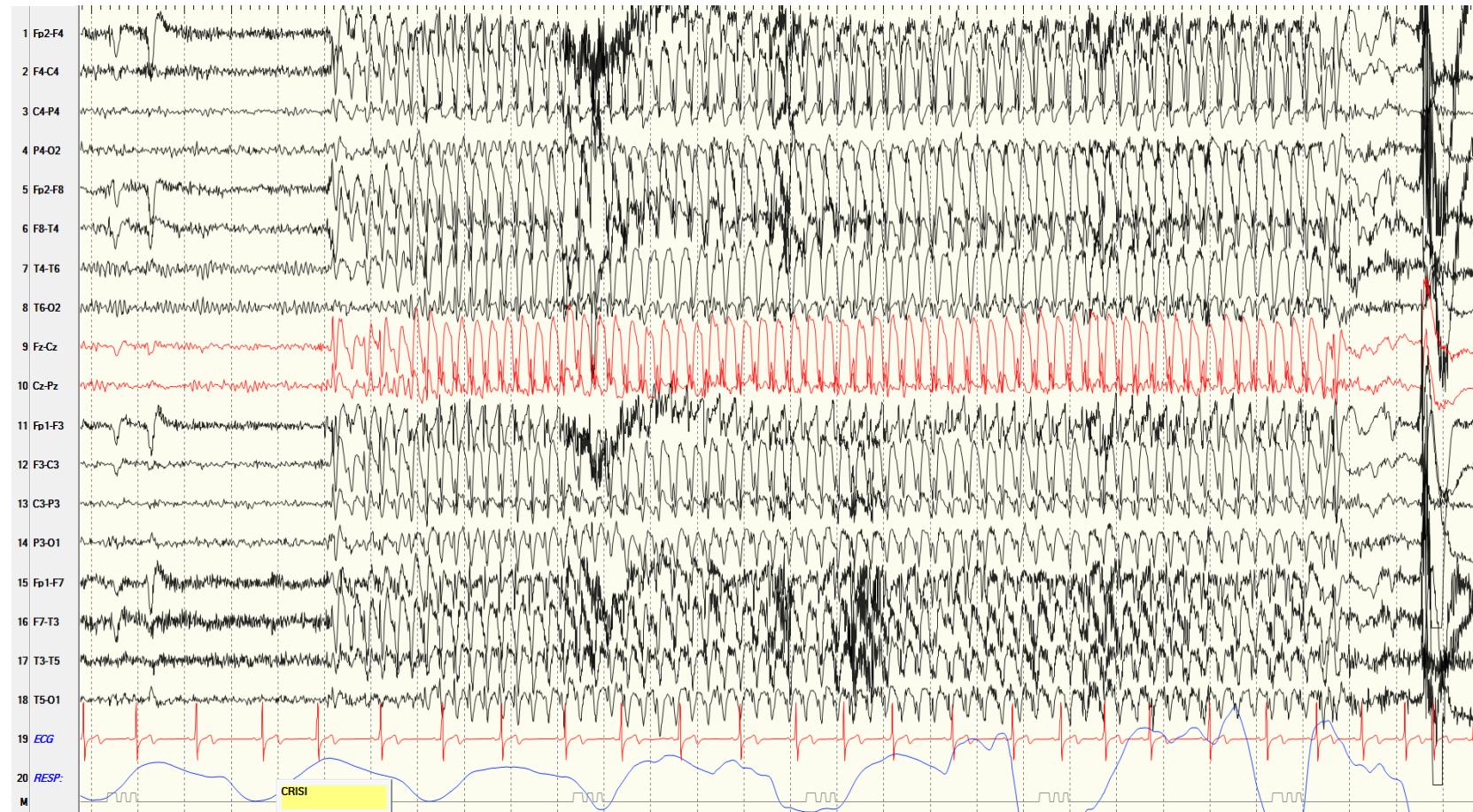




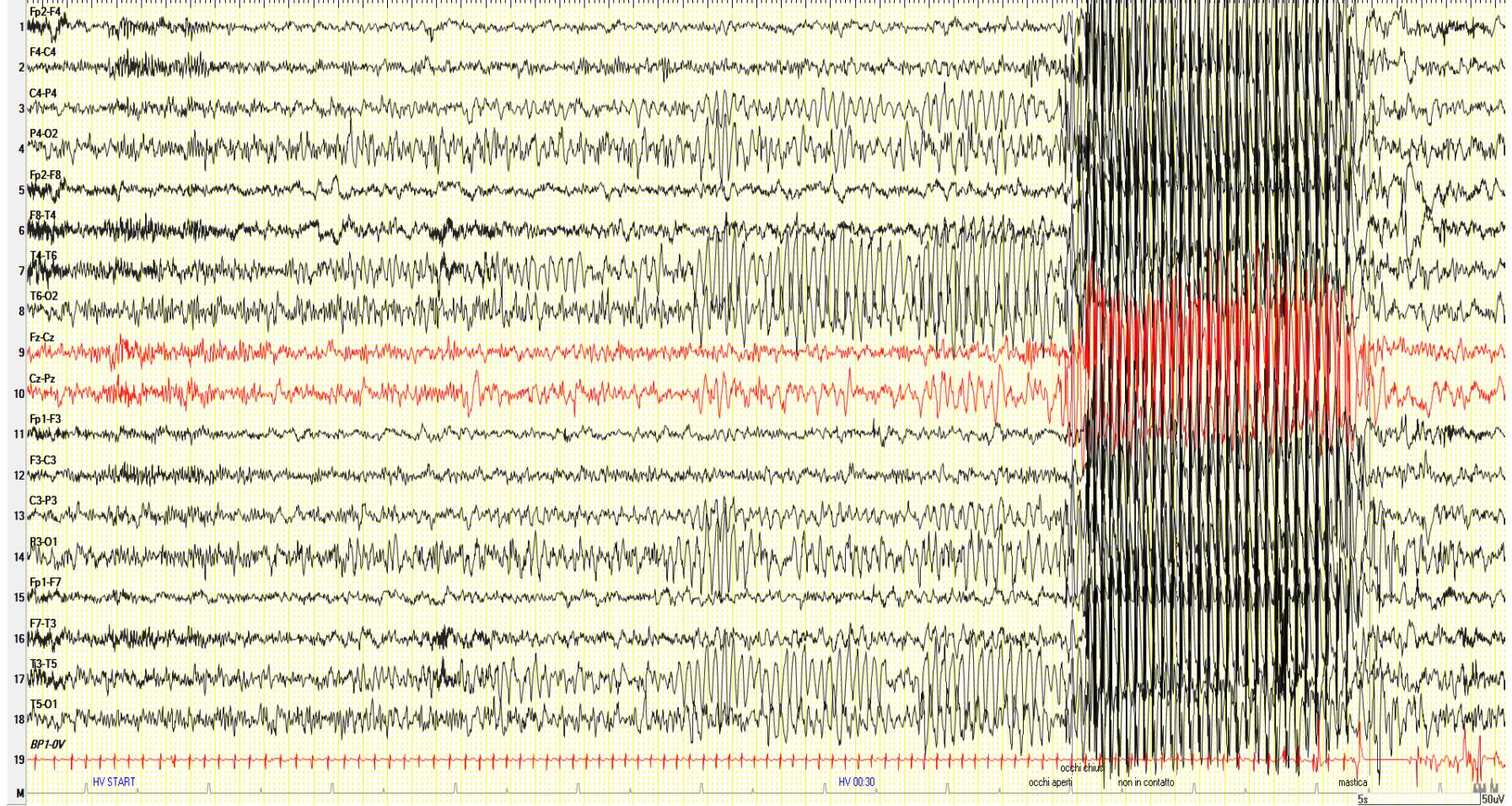
Spag. HP



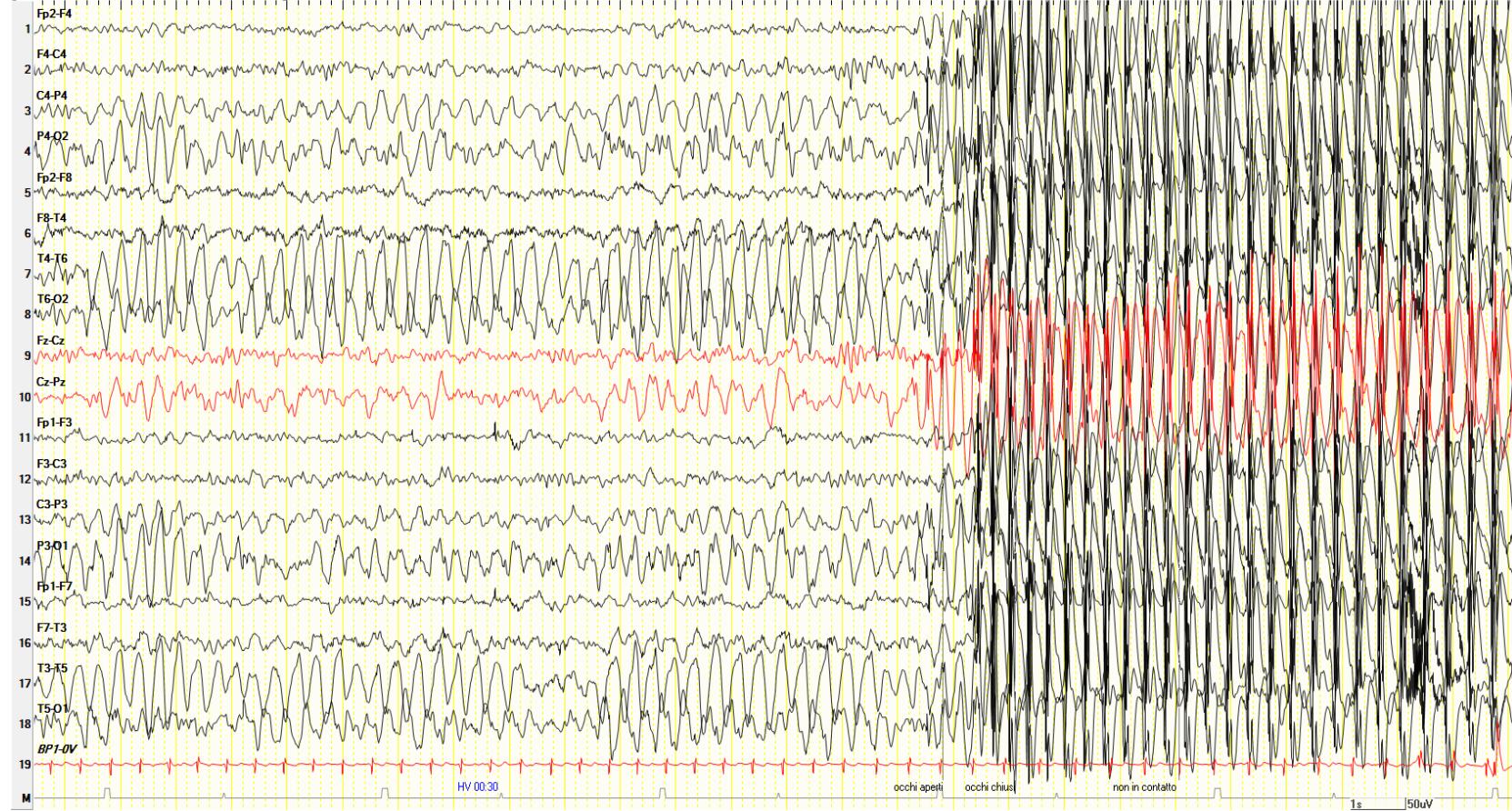
Spag.

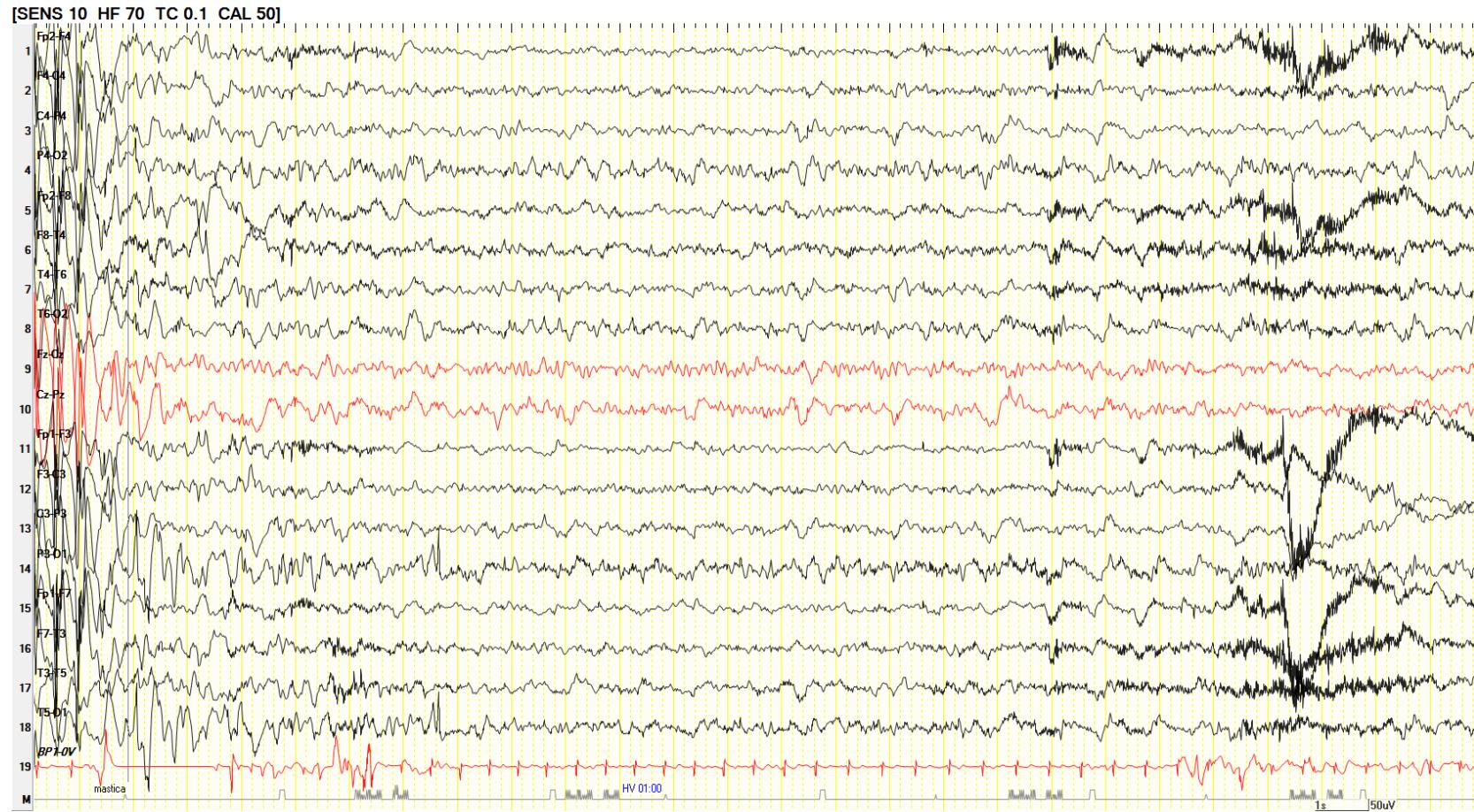


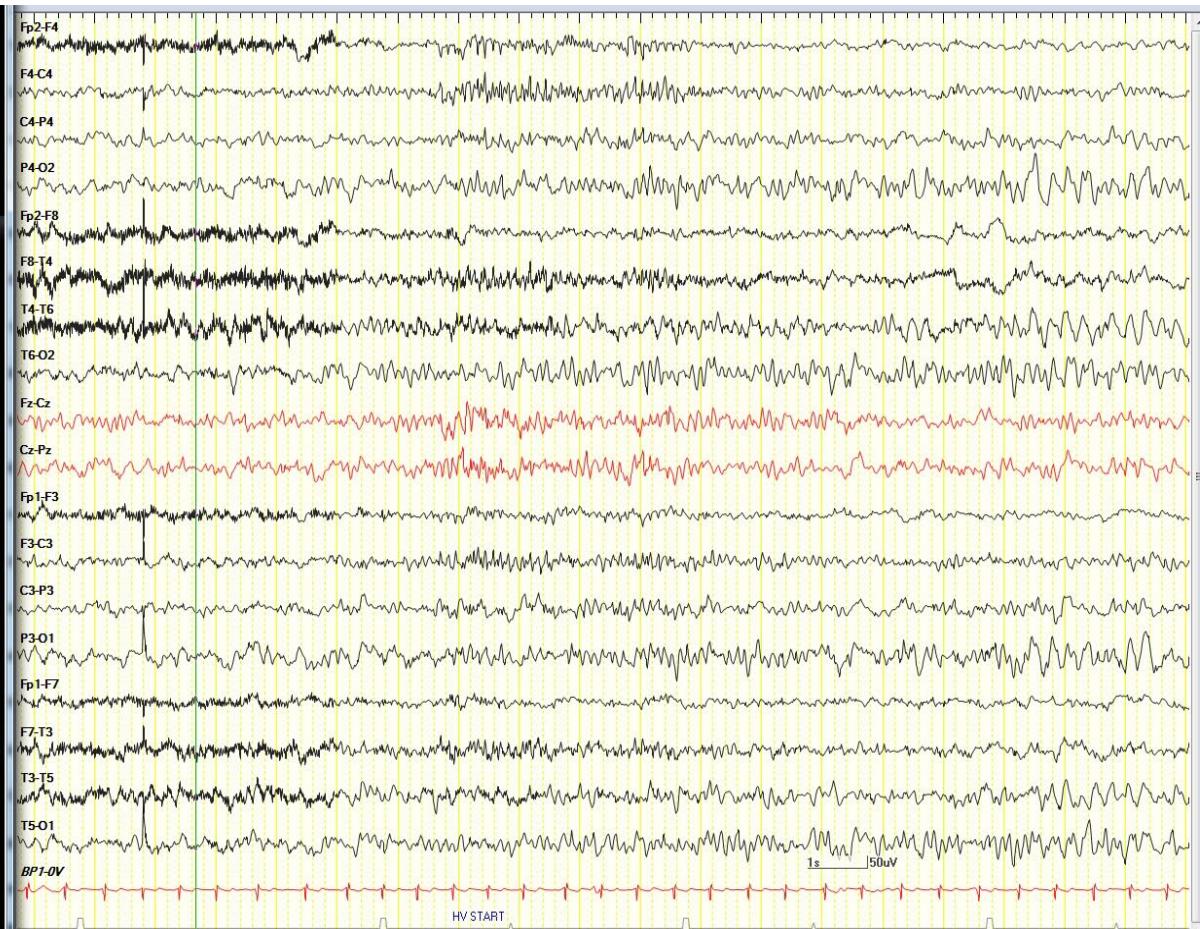
[SENS 10 HF 70 TC 0.1 CAL 50]



[SENS 10 HF 70 TC 0.1 CAL 50]

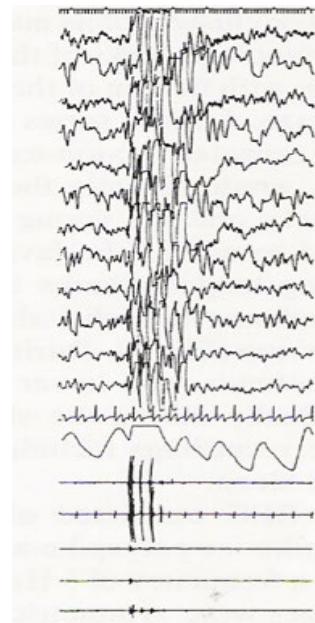
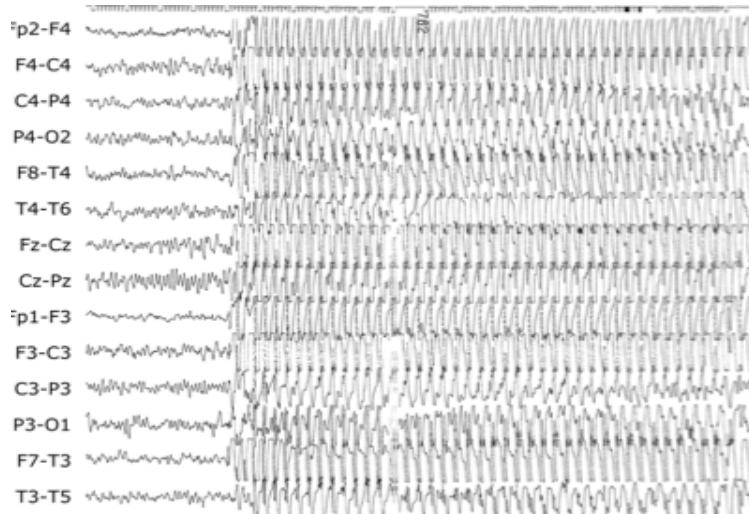


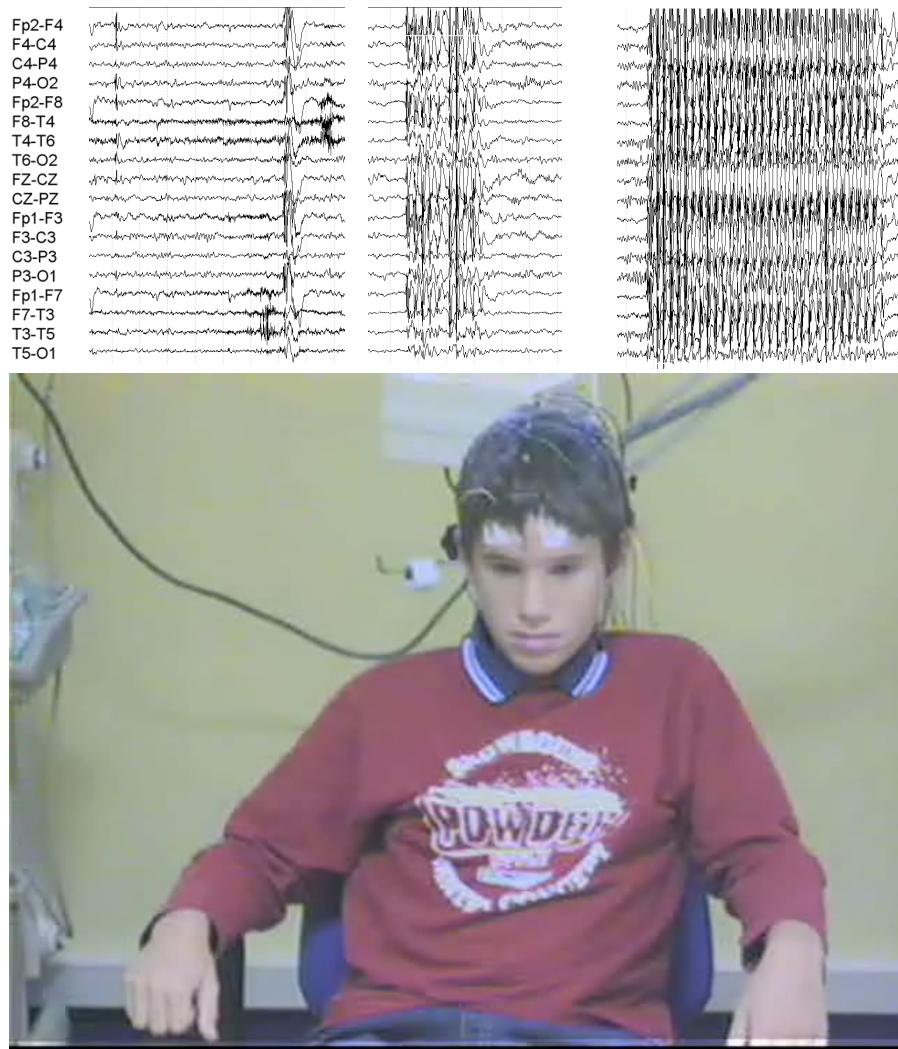


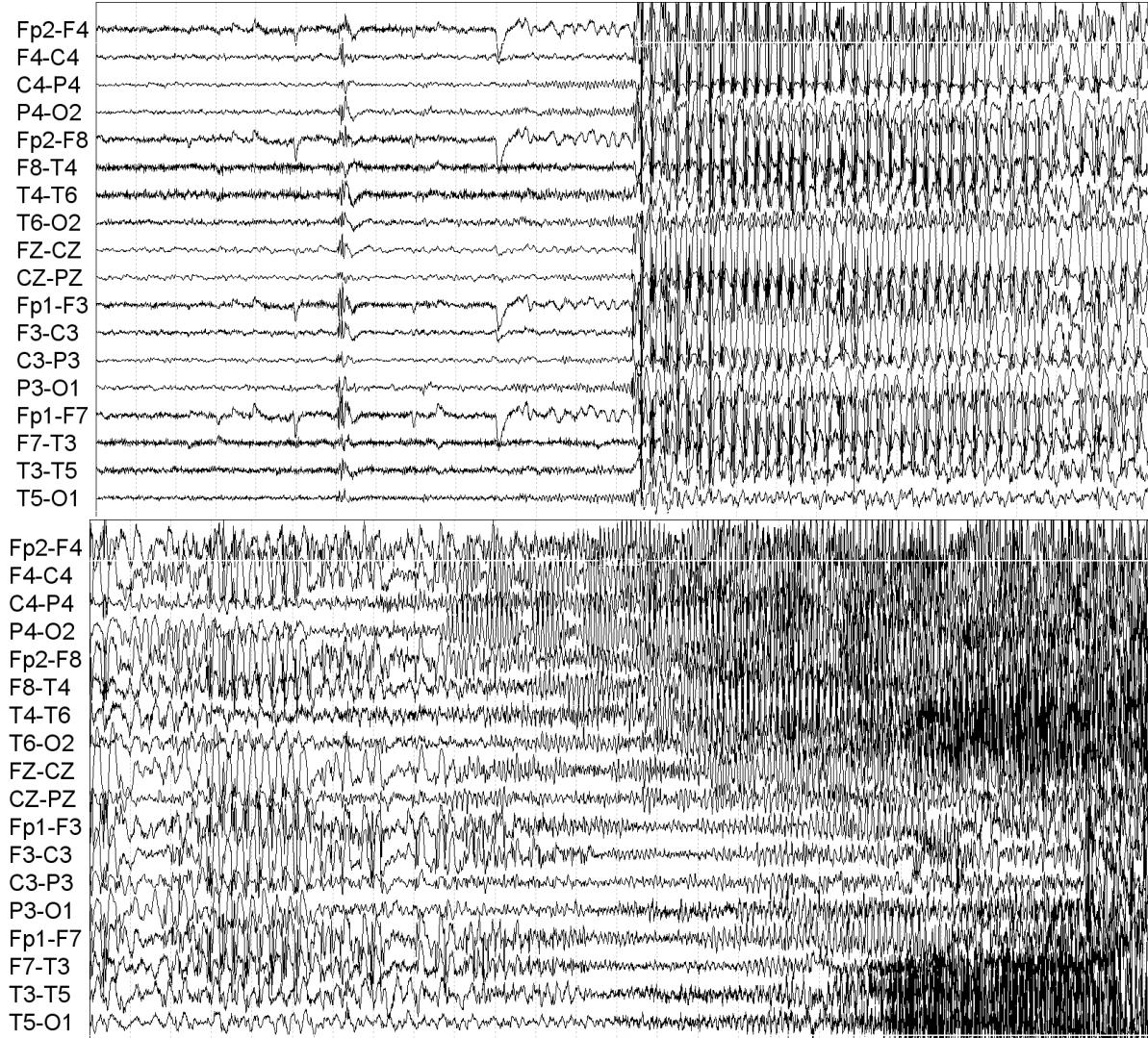


“Generalized Seizures”

- Myoclonic jerks
- Absence seizures Typical, Atypical:
 - absence with eyelid myoclonia;
 - myoclonic absence.
- Generalized Tonic-Clonic; Tonic,
- Clonic, Atonic, Myoclonic
- Myoclonic-Tonic-Clonic
- Epileptic spasms







BRIEF COMMUNICATION

Generalized-onset seizures with secondary focal evolution

Randy Williamson, Samrina Hanif, Gregory C. Mathews,
Andre H. Lagrange, and Bassel Abou-Khalil

Department of Neurology, Vanderbilt University, Nashville, Tennessee, U.S.A.

Epilepsy & Behavior 54 (2016) 20–29



Contents lists available at ScienceDirect

Epilepsy & Behavior

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



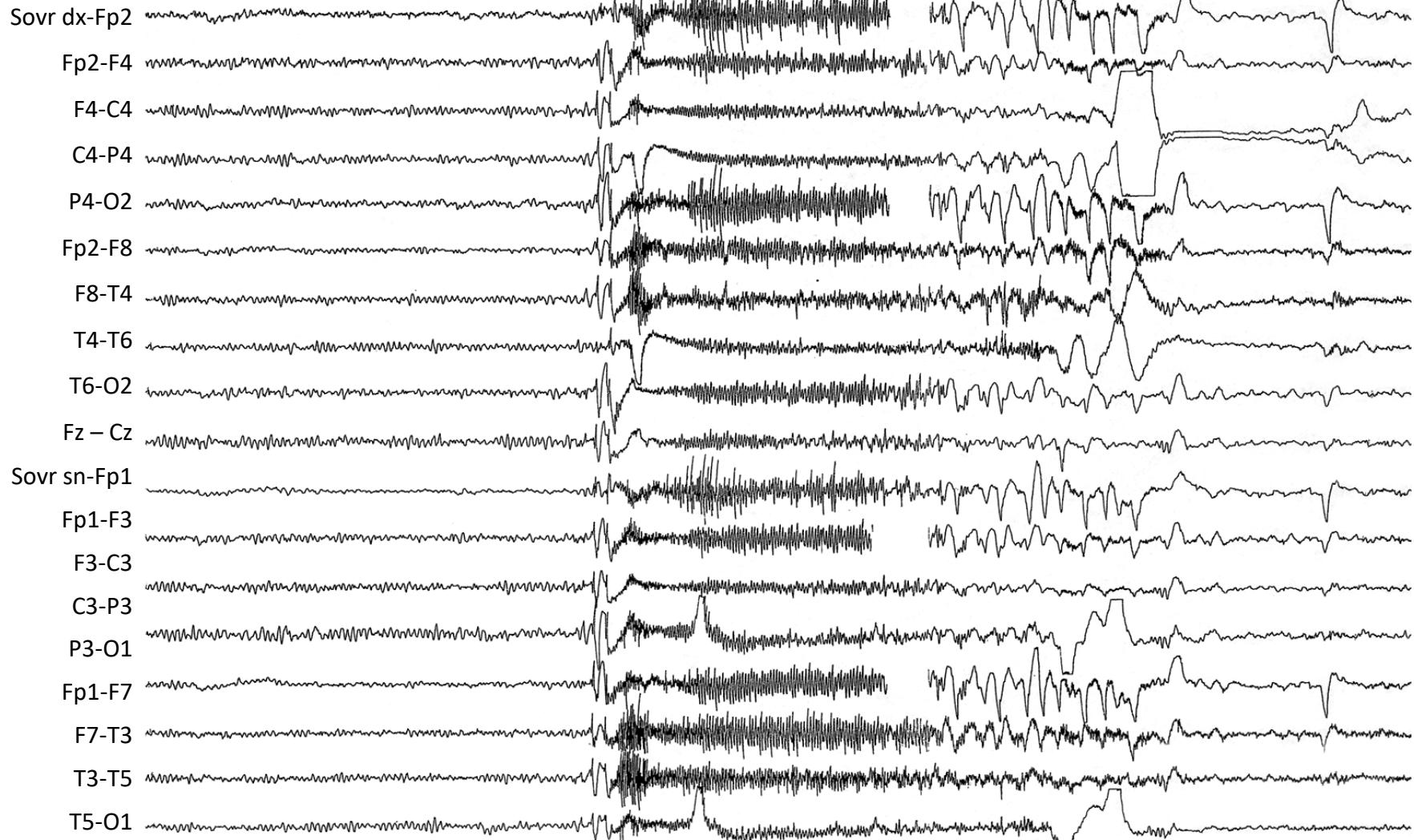
Generalized onset seizures with focal evolution (GOFE) – A unique seizure type in the setting of generalized epilepsy

Avriel Linane, Andre H. Lagrange, Cary Fu, Bassel Abou-Khalil *

Department of Neurology, Vanderbilt University, Nashville, TN, USA







ECG

Resp.

pz. sdraiata ad 00
mano dx sul bracciolo
mano sn sull'inguine

15:53':57"
Rimanendo a 00 e poi leggermente a 00 verso dx, deviazione verso il basso angoli della bocca, allarga e solleva AASS (dx e sn) allarga leggermente AAII chiamata e mostrato oggetto non risponde

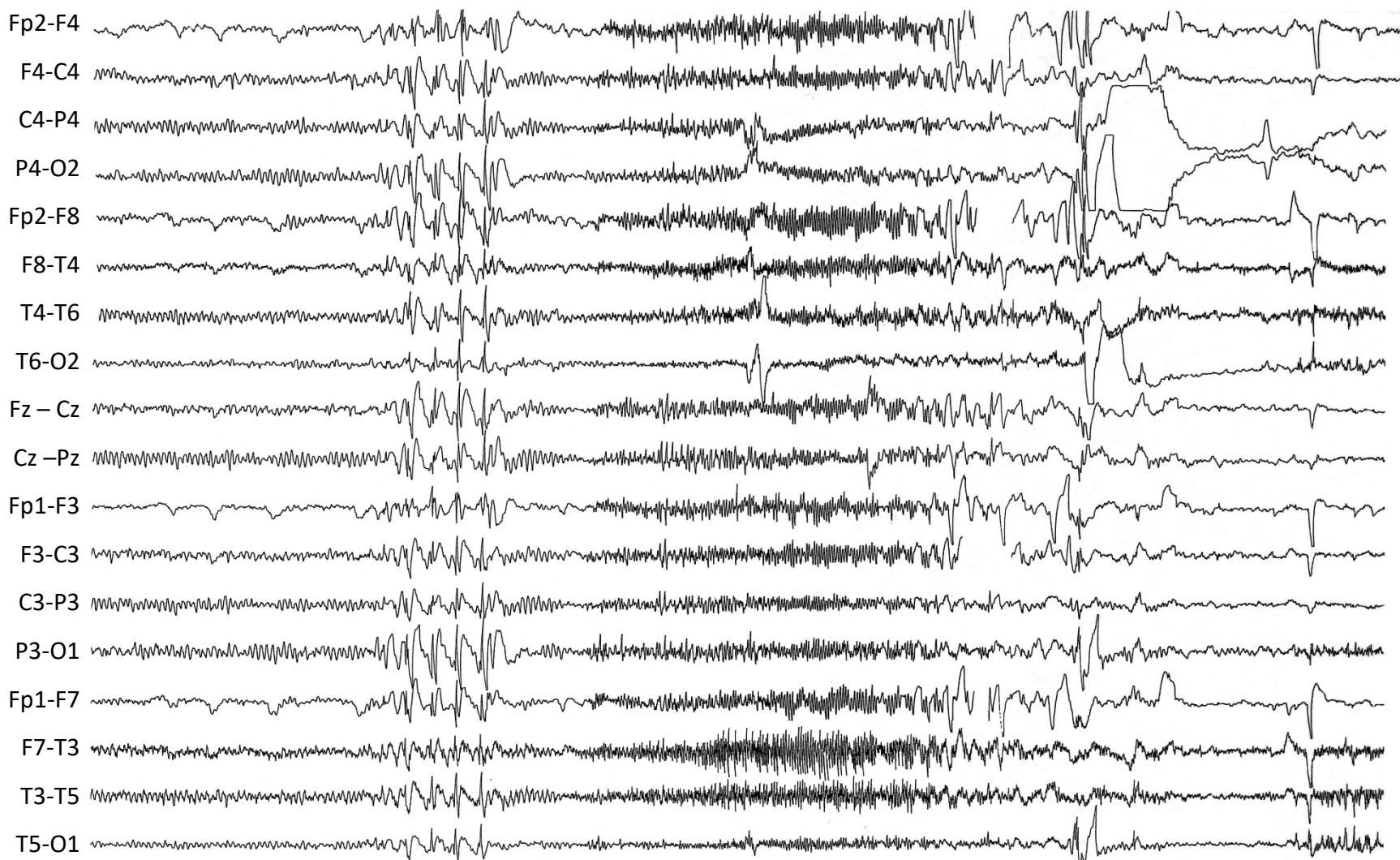
rossa in volto

torna nella posizione iniziale e si aggiusta un po' i vestiti

00 su comando

D. cosa è successo?
R: mi sono sentita male
D che cosa hai sentito?
R: un formicolio alle gambe

50 μ V
1 sec.



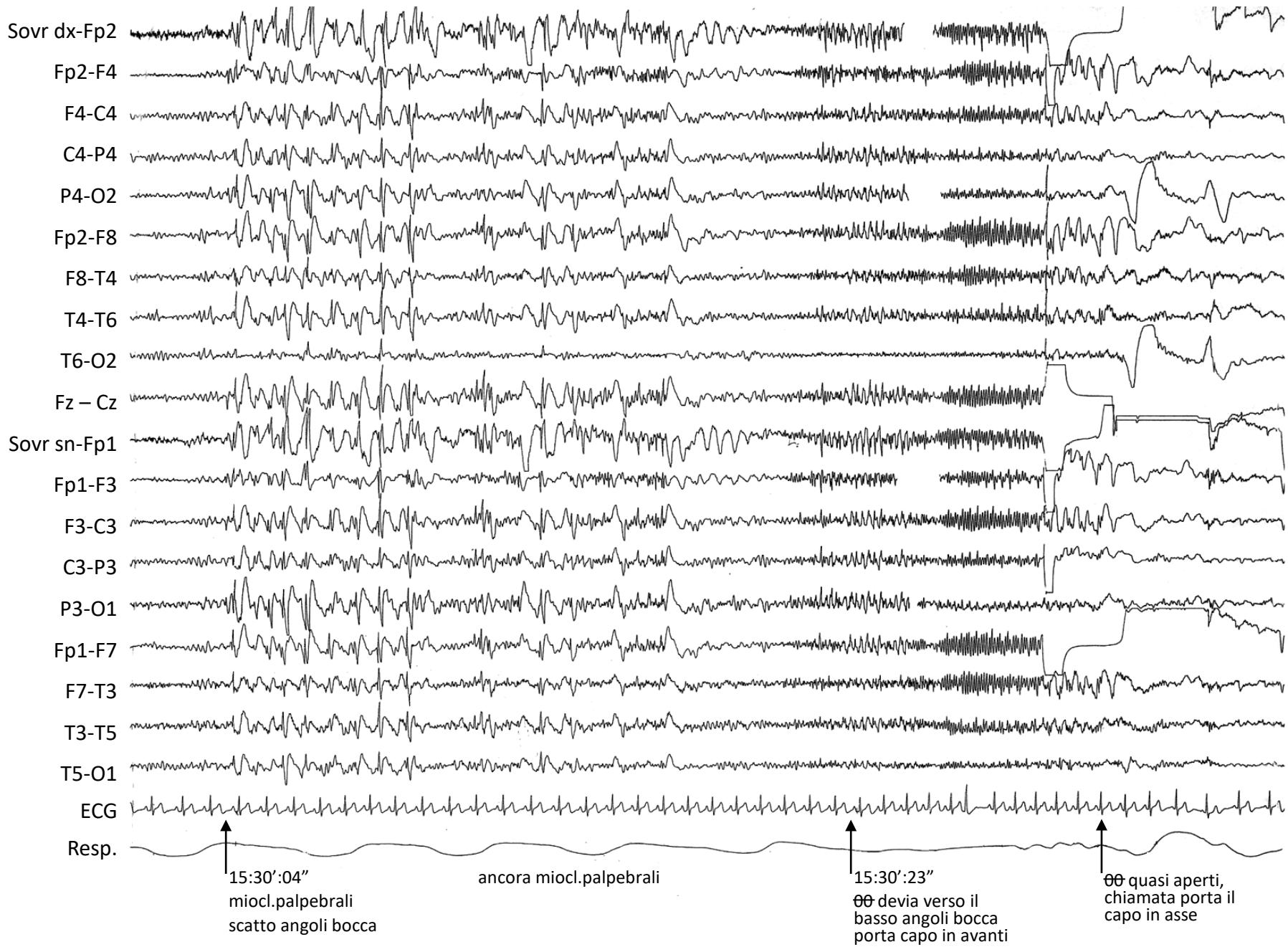
13:56':05"
 00 supina AASS lungo i
 braccioli della poltrona
 capo verso dx

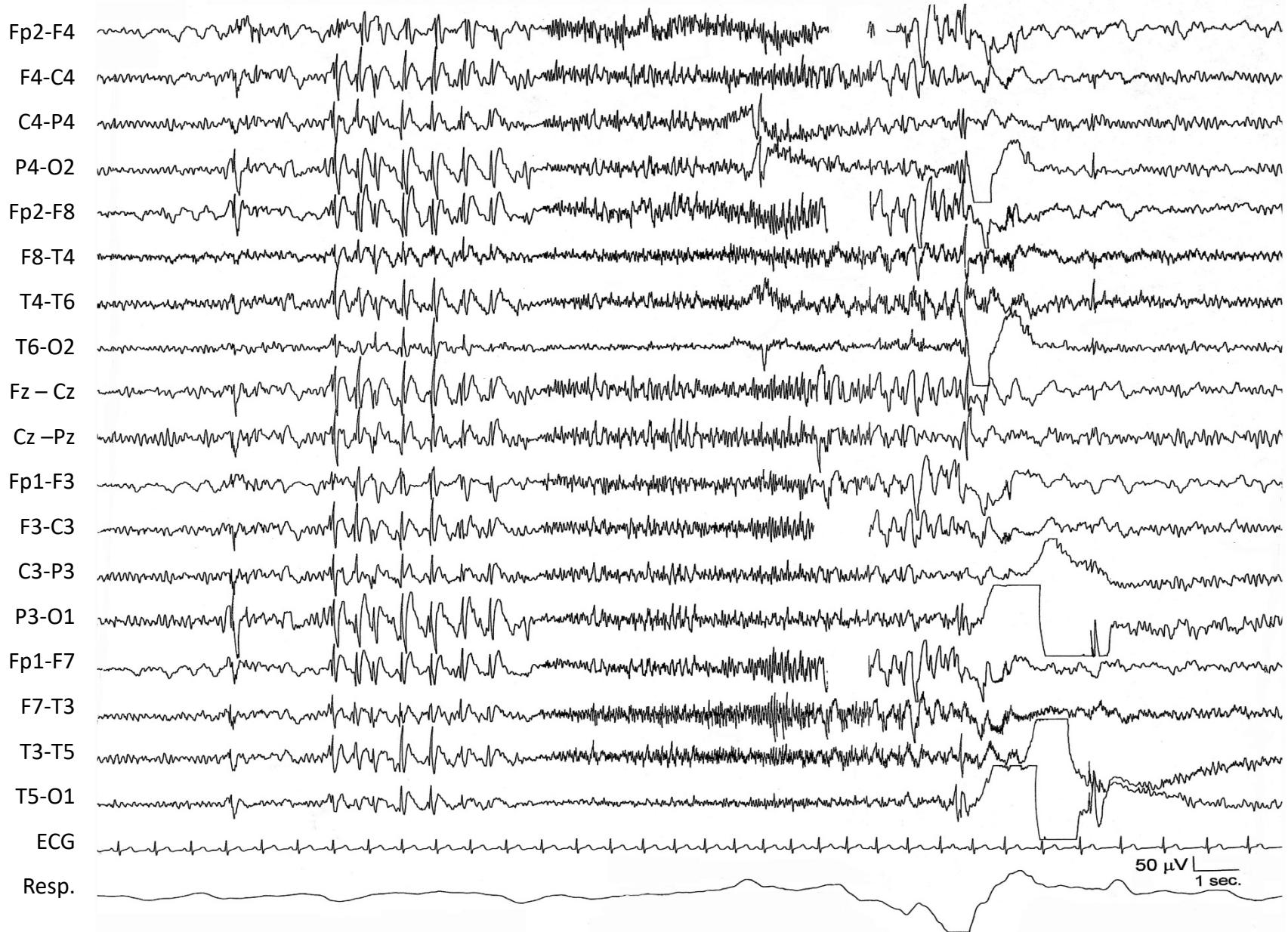
visibili mioclonie
 palpebrali

mostrate 3 dita non risponde
 stira angoli della bocca verso il
 basso allarga leggermente poi
 flette in avanti AASS solleva Al
 sn 00 capo leggermente in
 avanti

Mostrata penna non
 risponde riappoggia il capo
 alla poltrona e con le mani
 si sistema il pigiama

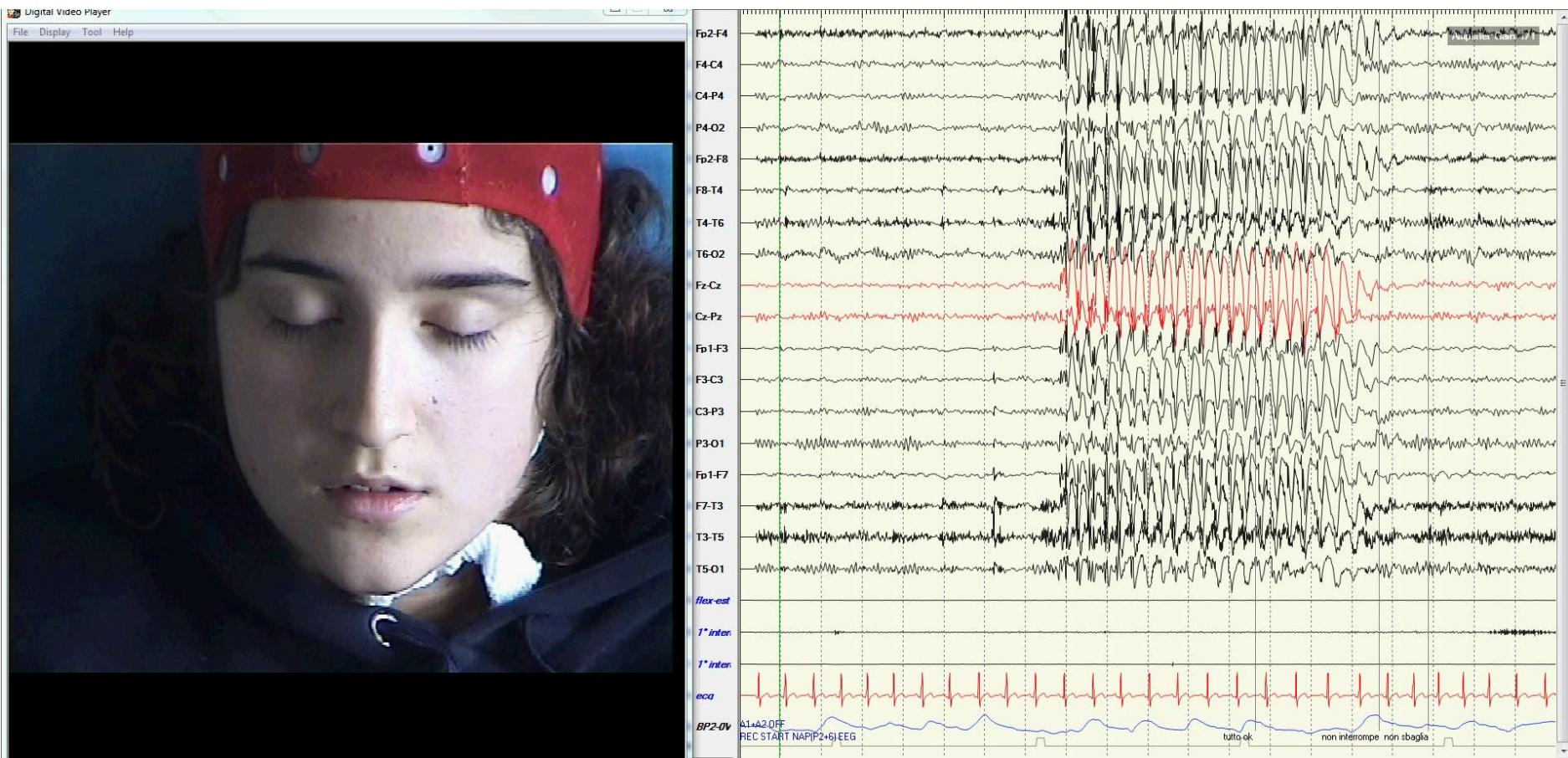
D. è passata ?
 R: sì
 tec: "apri gli occhi"
 pz: esegue

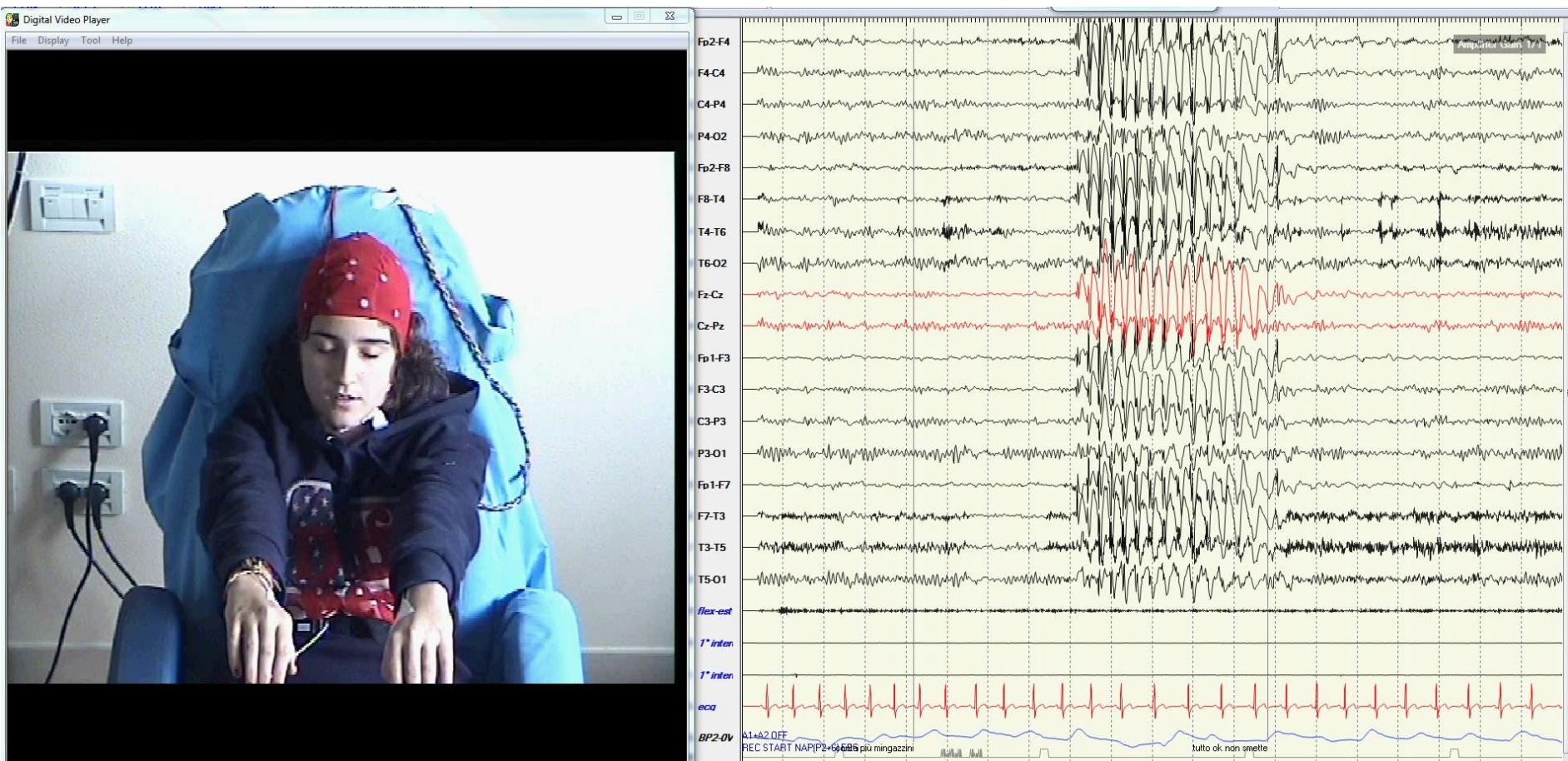




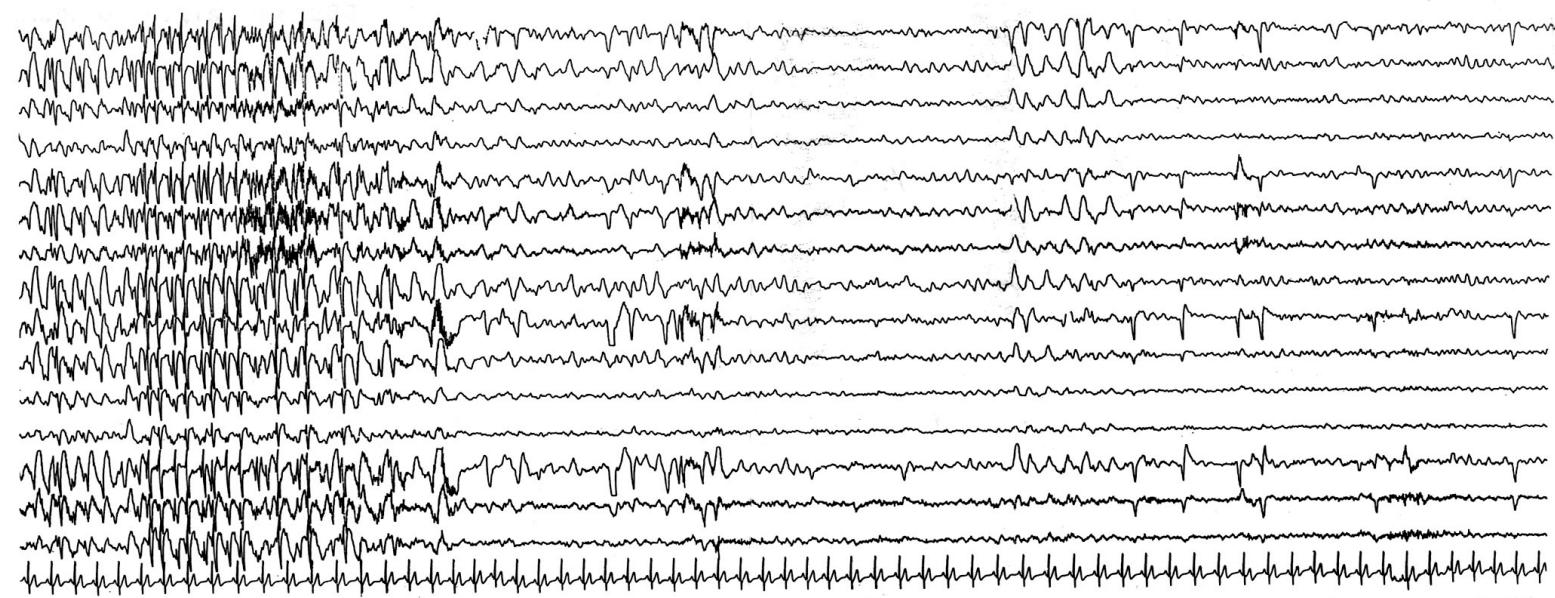
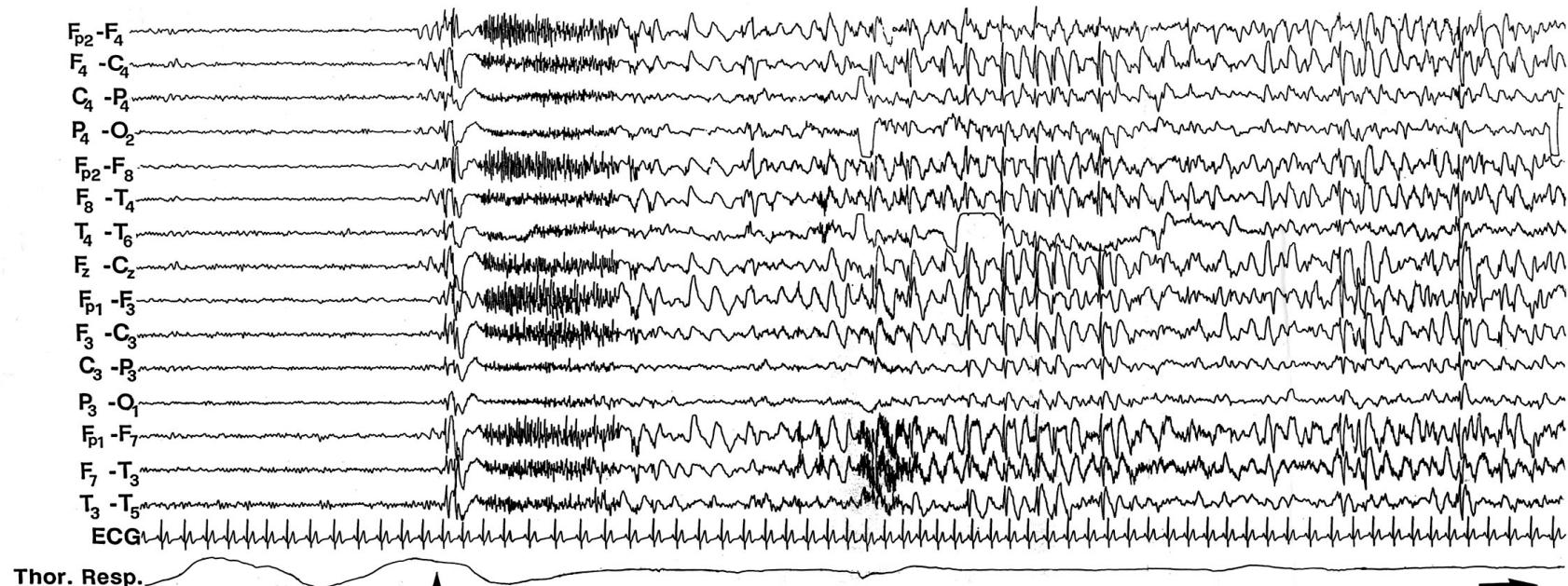
Stessa semeiologia della precedente
visibile rosore al volto

13:58':05"









Ber. Rob. ♂ 17yrs n° 81283

50 μ V
1 sec.

Genetic Generalized Epilepsies

Idiopathic Generalized Epilepsies

Childhood
Absence CAE

Juvenile
Myoclonic
JME

Juvenile
Absence JAE

Generalized Tonic-
Clonic Seizures
Alone GTCA

Genetic Epilepsy with
Febrile Seizures Plus
GEFS +

Myoclonic-Atonic Epilepsy

Epilepsy with Eyelid Myoclonia

Epilepsy with Myoclonic Absence

Myoclonic Epilepsy in
Infancy

Developmental
Encephalopathy
y
= Intellectual
Disability

ILAE Task Force on Nosology and Definitions 2017-2021



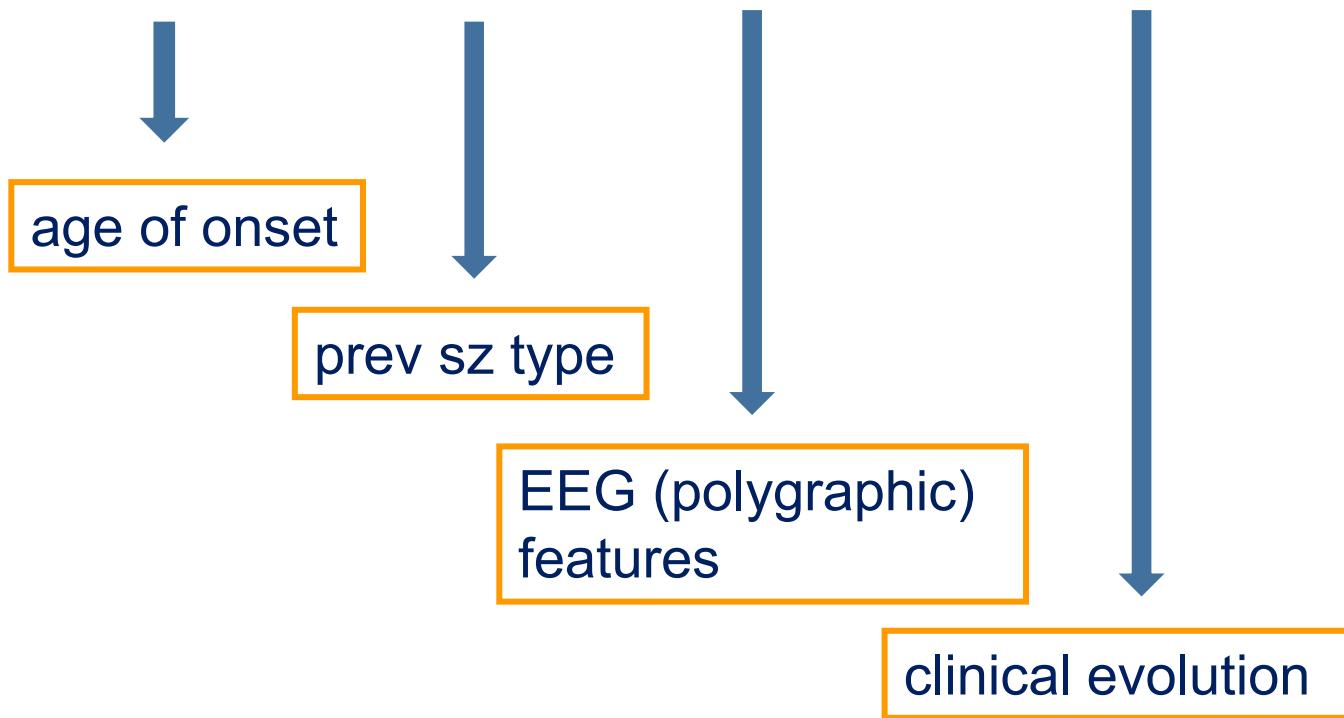
EC Wirrell, P Tinuper (Co-Chair), E Trinka, S Wiebe, JH Cross, E Hirsch, SM Zuberi, A Bogacz, E Somerville, K Riney, JA French, IE Scheffer, R Nababout, N Specchio, S Kaneko, S Jain, P Samia, T Alsaadi, OC Snead, S Balestrini

IGE: common features

- ❖ **Onset** : around infancy
- ❖ **Types of seiz** : absences
 - myoclonic seizures
 - tonic-clonic seizures
- ❖ **EEG** : bss S-W and PP-SW complexes
 - normal BG activity
- ❖ **Clin ctx** : normal neurological ex and intell
- ❖ **MRI find** : normal
- ❖ **Outcome** : usually good

not all IGEs are equal

specific syndromes are defined according to:



IGE syndromes

- ❖ Benign myoclonic epilepsy of infancy (BMEI)
 - ❖ Childhood absence epilepsy (CAE)
 - ❖ Juvenile absence epilepsy (JAE)
 - ❖ Juvenile myoclonic epilepsy (JME)
 - ❖ IGE with tonic-clonic seizures alone (IGE-TCS)
-
- ❖ Epilepsy with myoclonic-astatic seizures (EMAS)
 - ❖ Epilepsy with myoclonic absences (EMA)

Different IGE sub-types seem to present as distinctive entities with different age of onset, electroclinical patterns, duration of illness and response to therapy.

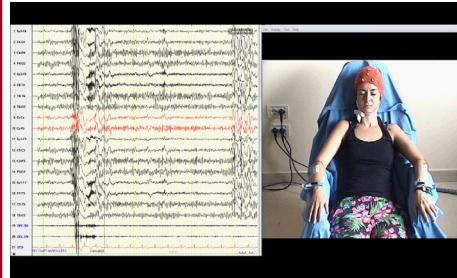
CAE

Onset 4-10 yrs
Typical absence S
Rare TCS
Remission at puberty



JME

Onset 10-18 yrs
Myocl S, photosensit
Freq TCS
Late remission



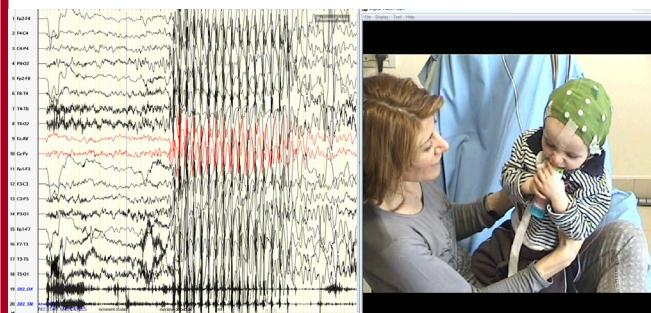
IGE-TCS

Onset chil to ad
TCS, Myocl S,
Normal intellect
Good outcome



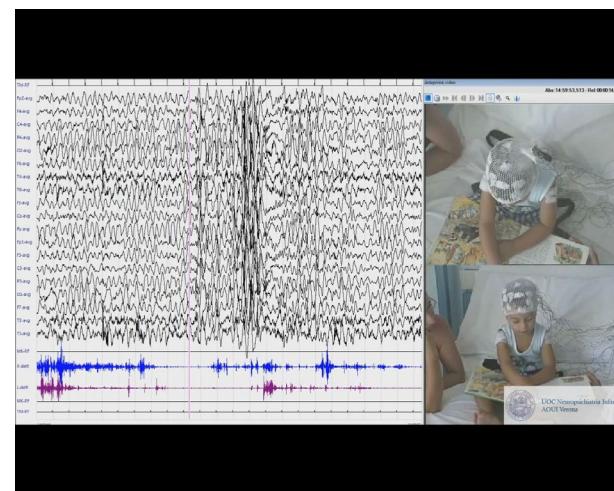
EMA

Onset 5-10 yrs
Myocl abs, intellect impairm
Variable outcome



MAE

Onset 2-6 yrs
Myocl ,atonic, GTCS, NCSE
long lasting intellect impairm
Variable outcome
Possible intractability

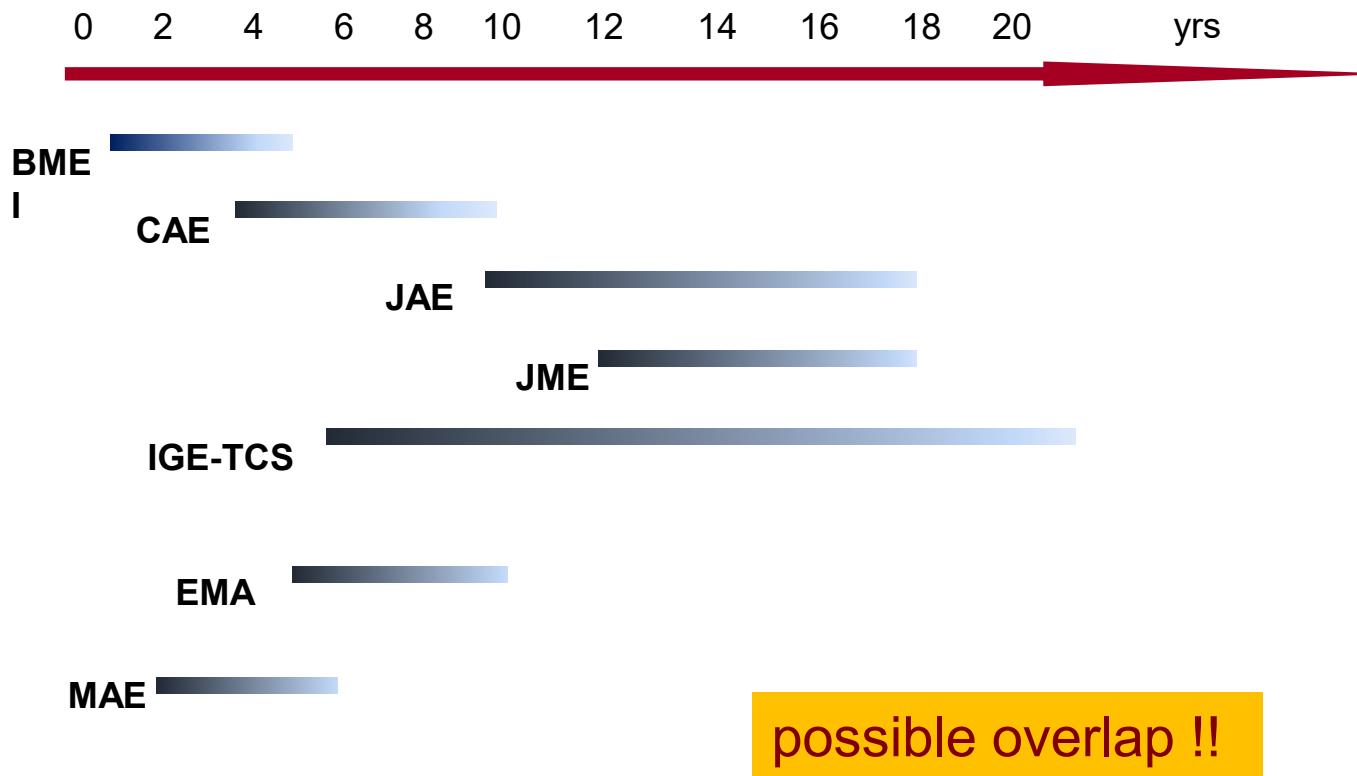


Summary of the clinical features of the IGE syndromes

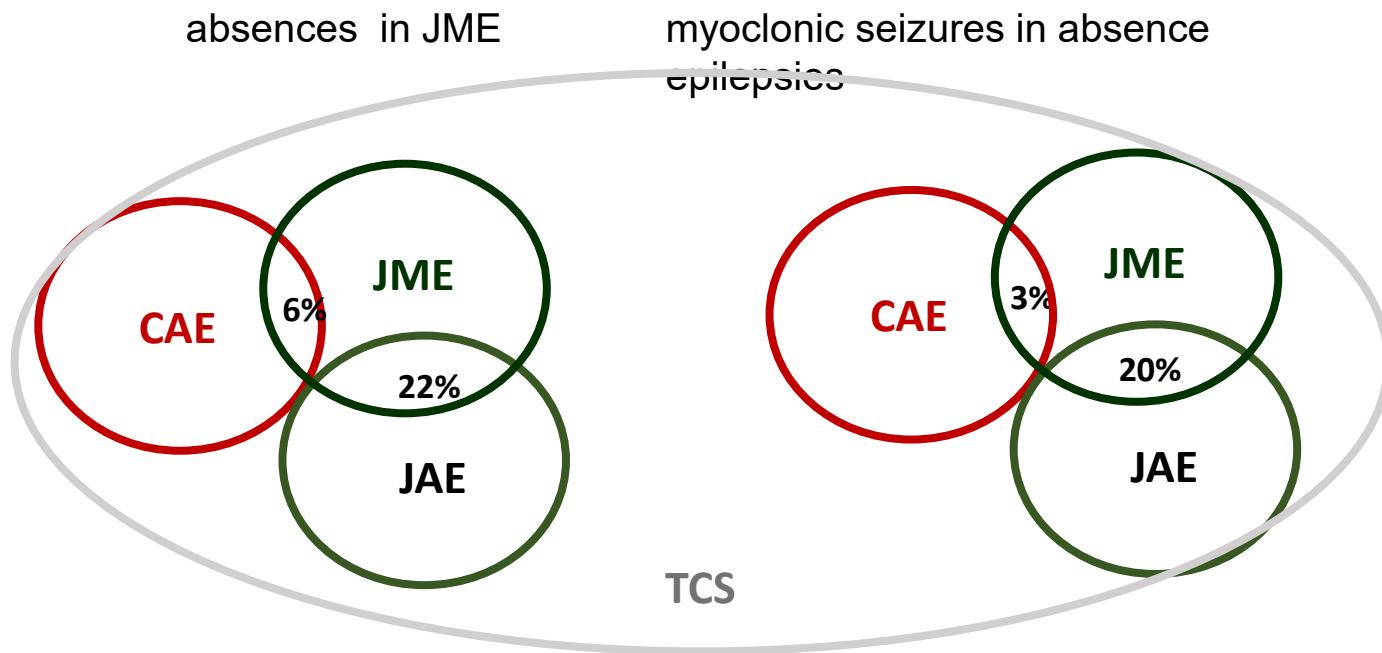
IGE subtype	Age of onset	1st seizure type	Other seizure types	EEG	Photosensitivity
Included in ILAE classification					
BMEI	0.4-5 yrs	My	FS	GSW	Rare
CAE	4-10 yrs	Ab	TCS, My	3-Hz GSW	Rare
JAE	10-18 yrs	Ab	TCS, My	3-4Hz GSW	Rare
JME	12-18 yrs	My	TCS, Ab	4-6Hz GSW	Frequent
IGE-TCS	Childhood to early adulthood	TCS	My, Ab	GSW	Rare
EMA	5-10 yrs	MA	TCS	3-Hz GSW	Rare
MAE	2-6 yrs	My-ast	Ab, TCS, non convulsive SE	GSW	Rare

mod. from Guerrini and Marini, 2011

age of onset of IGE syndromes



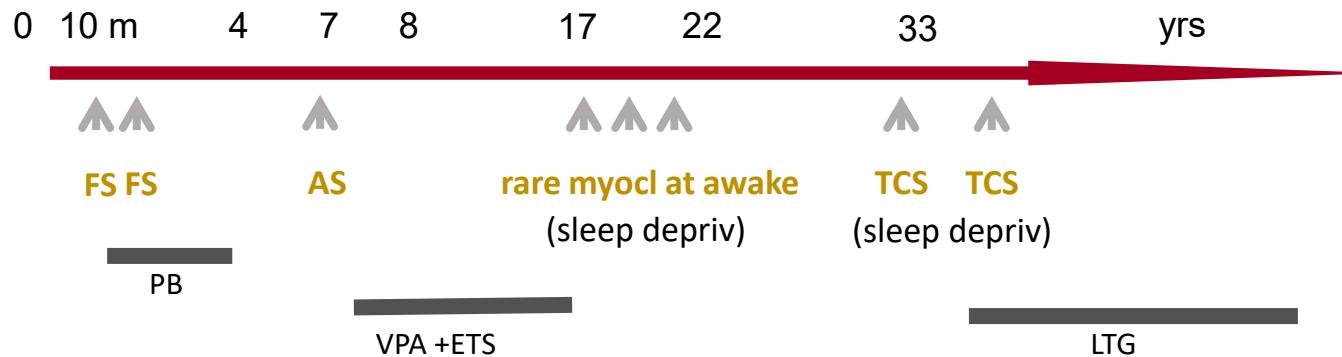
from Janz, 1990



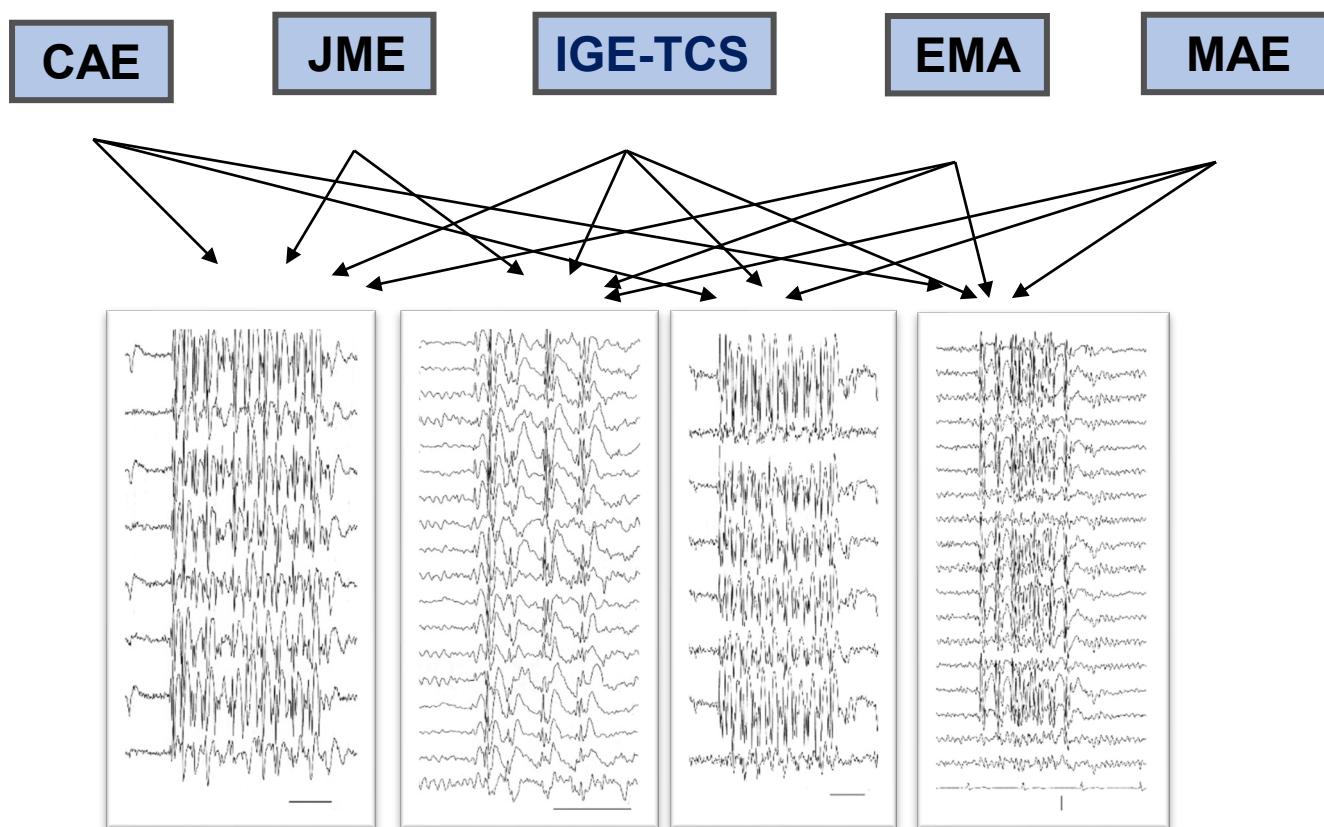
frequent clinical overlap !!

possible intraindividual overlap !!

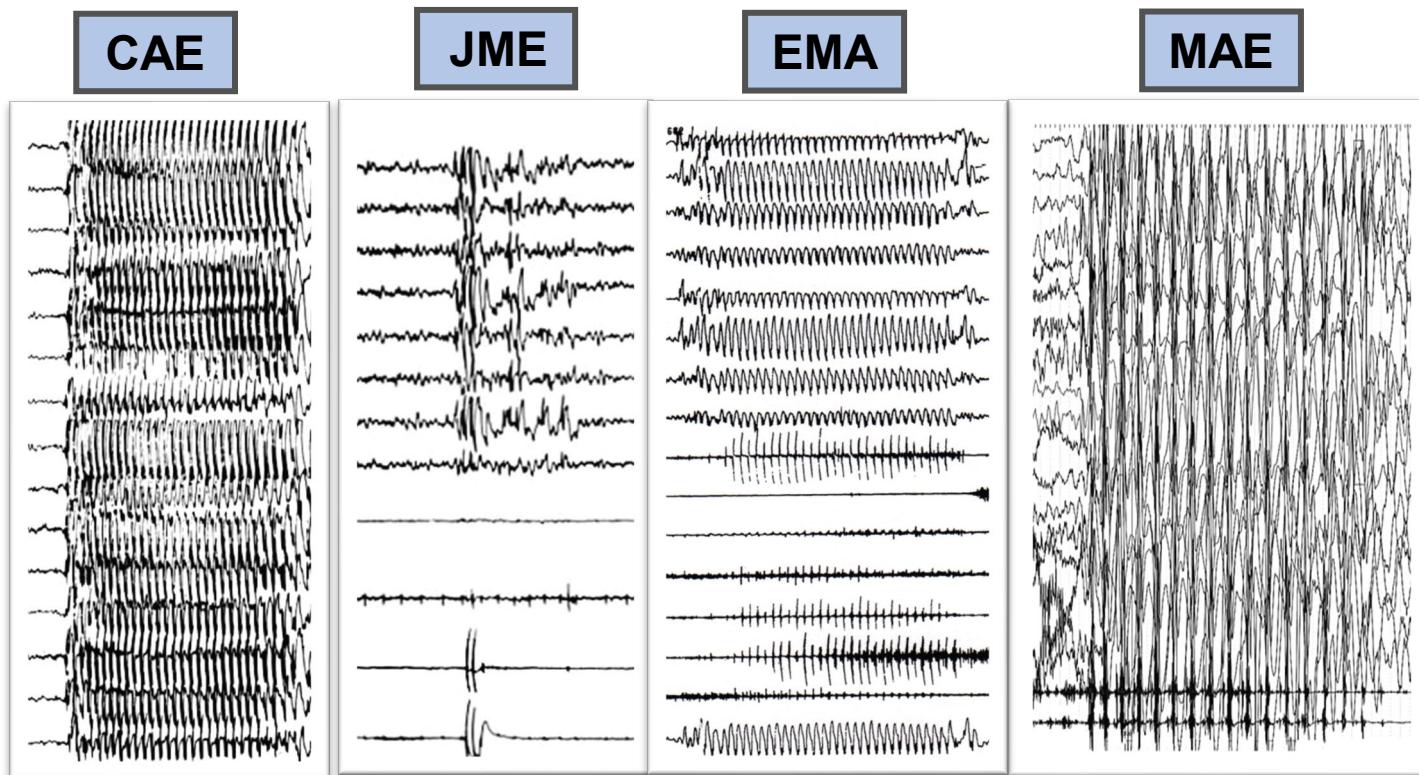
A.A. 35 yrs lady's hairdresser
Fam + for FS (the son of a daughter)



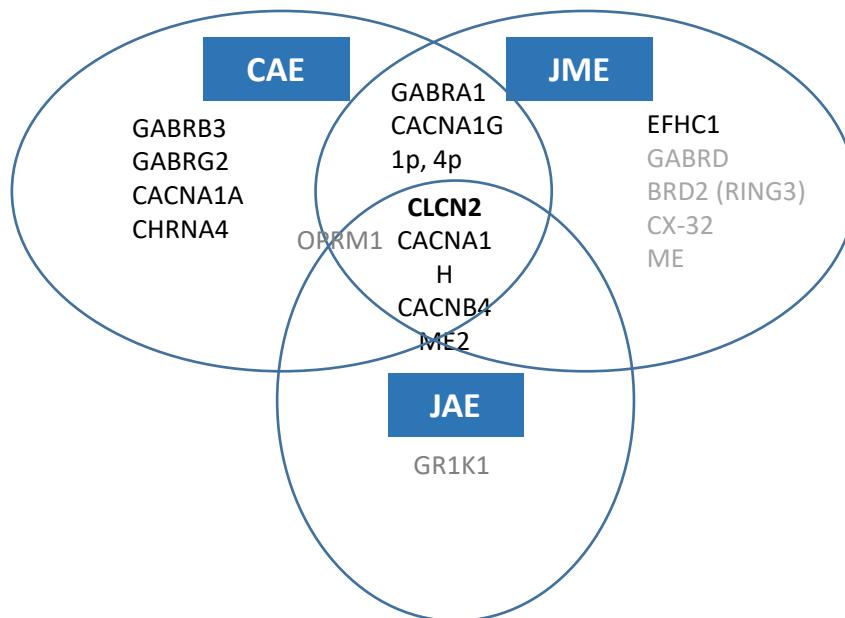
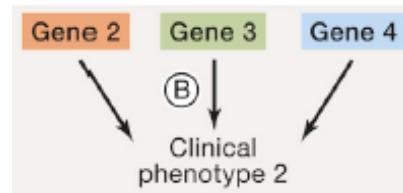
neurophysiological overlap !!



It is very important to describe EEG- polygraphic features to differentiate seizures and syndromes



genetic overlap !!



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FRACP

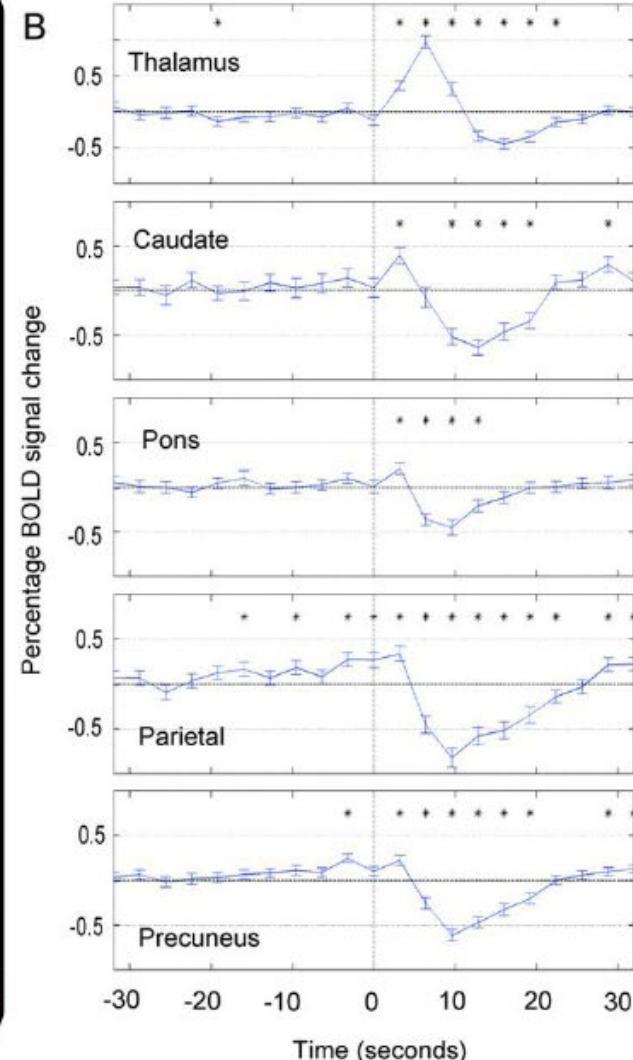
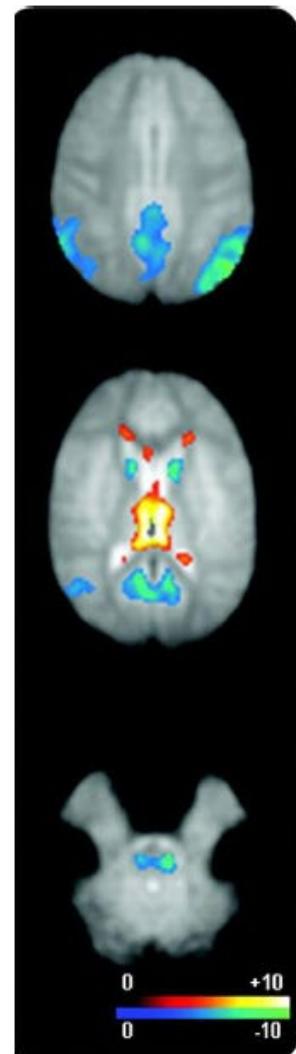
The core network in absence epilepsy

Differences in cortical and thalamic BOLD response



Neurology® 2010;75:904-911

11 children with typical AS, untreated

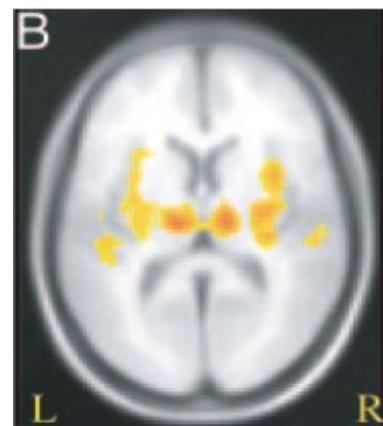


pathophysiological overlap !!

IGE

critical neuronal network

- neocortical pyramidal neurons
- thalamic relay neurons
- reticular thalamic neurons



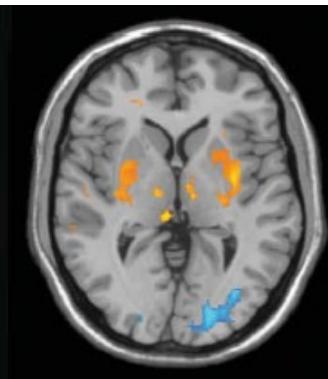
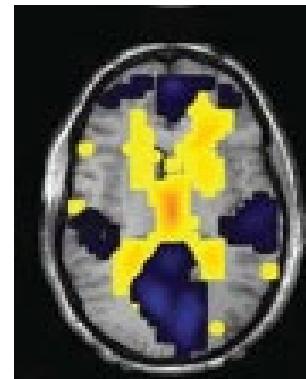
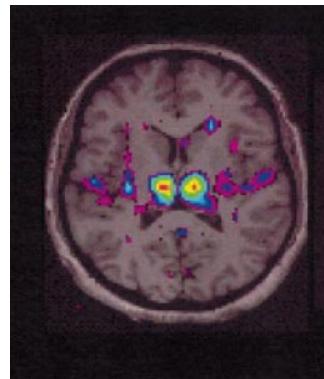
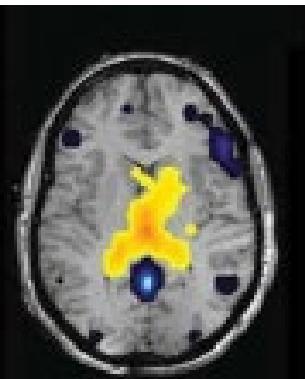
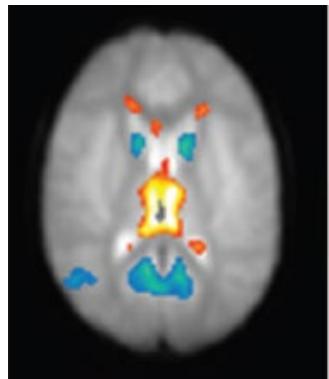
CAE

JAE

IGE-TCS

JME

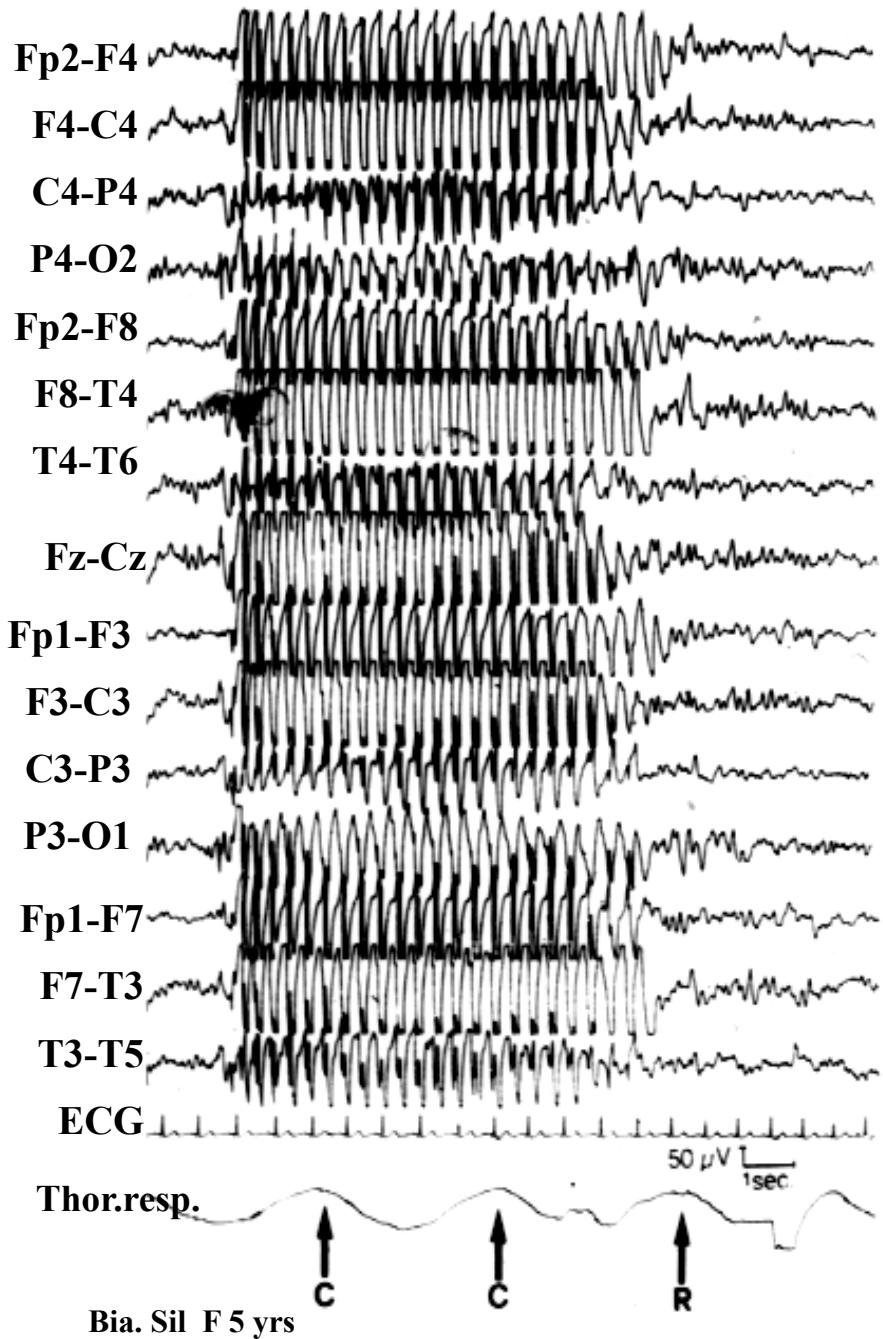
MAE



ASSENZA “PURA” (3 c/s)



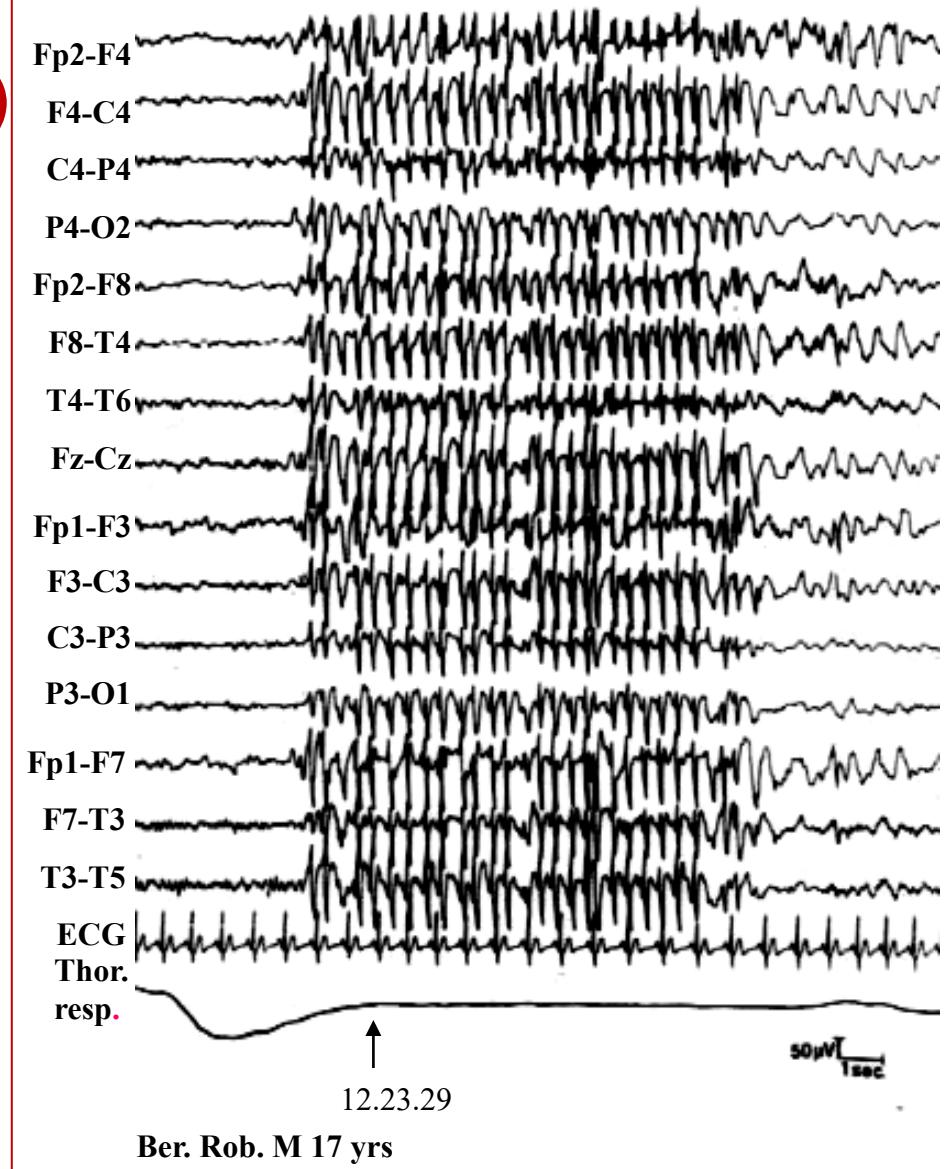
Breve sospensione della working memory



ASSENZA “NON PURA” (3c/s)



Disturbo dell'iniziativa e
dell'intenzione





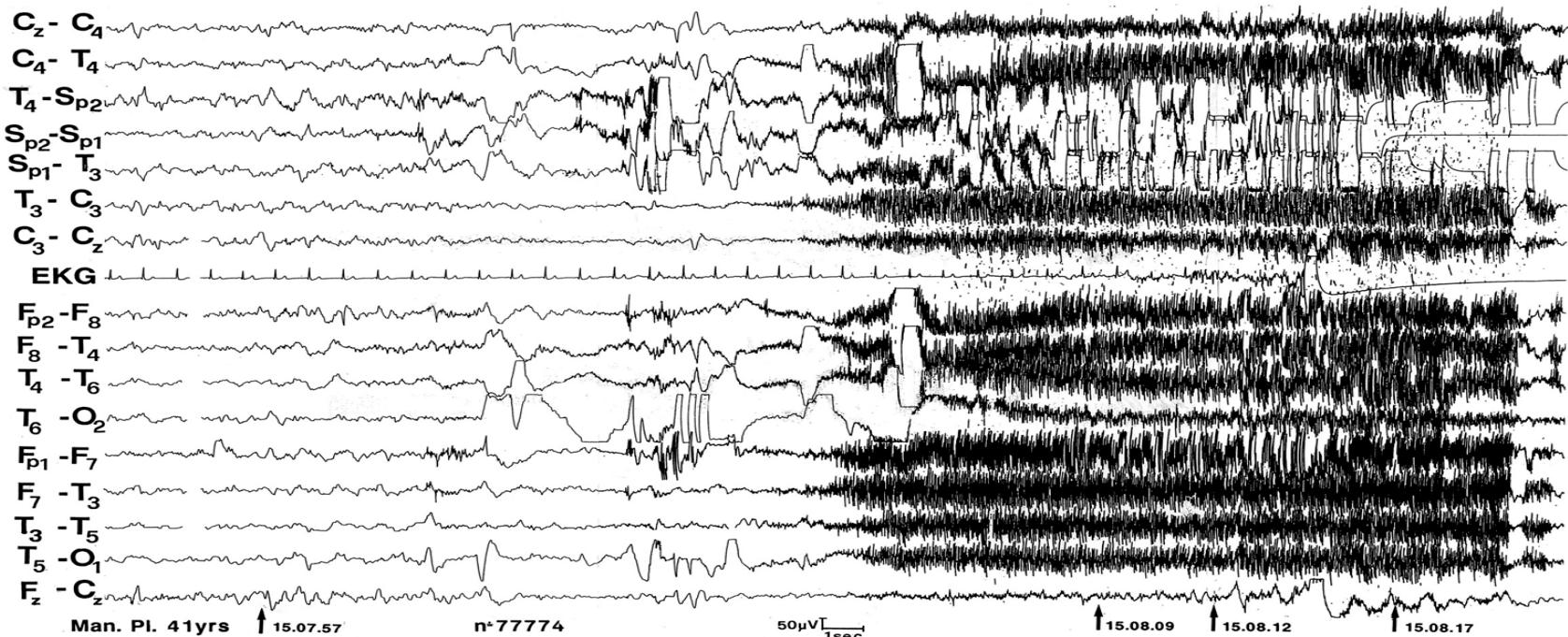
50 μ V/ cm
1 sec.

Thor. Resp.

↑ 12.23.29

Ber. Rob. ♂ 17 yrs n° 81283

Il problema dell'EEG nelle crisi frontali

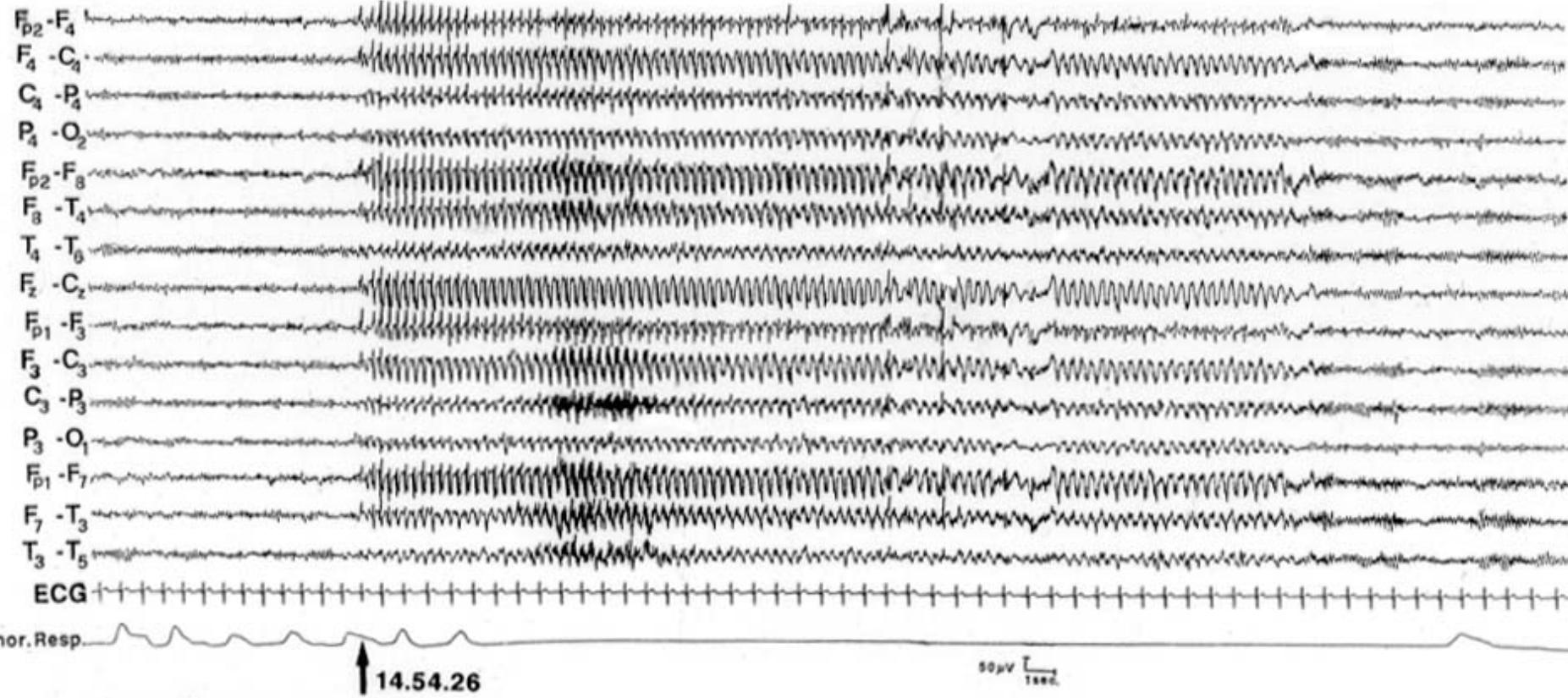


“... interictal and ictal scalp electroencephalograms are often not helpful and sometimes misleading.” P.D. Williamson, 1985

the electrographic diagnosis of orbital seizures depends exclusively on the findings from chronically implanted electrodes. C. Ajmone – Marsan, 1988

Tukel and Jasper. **The electroencephalogram in parasagittal lesions**
Electroenceph Clin Neurophysiol, 1952

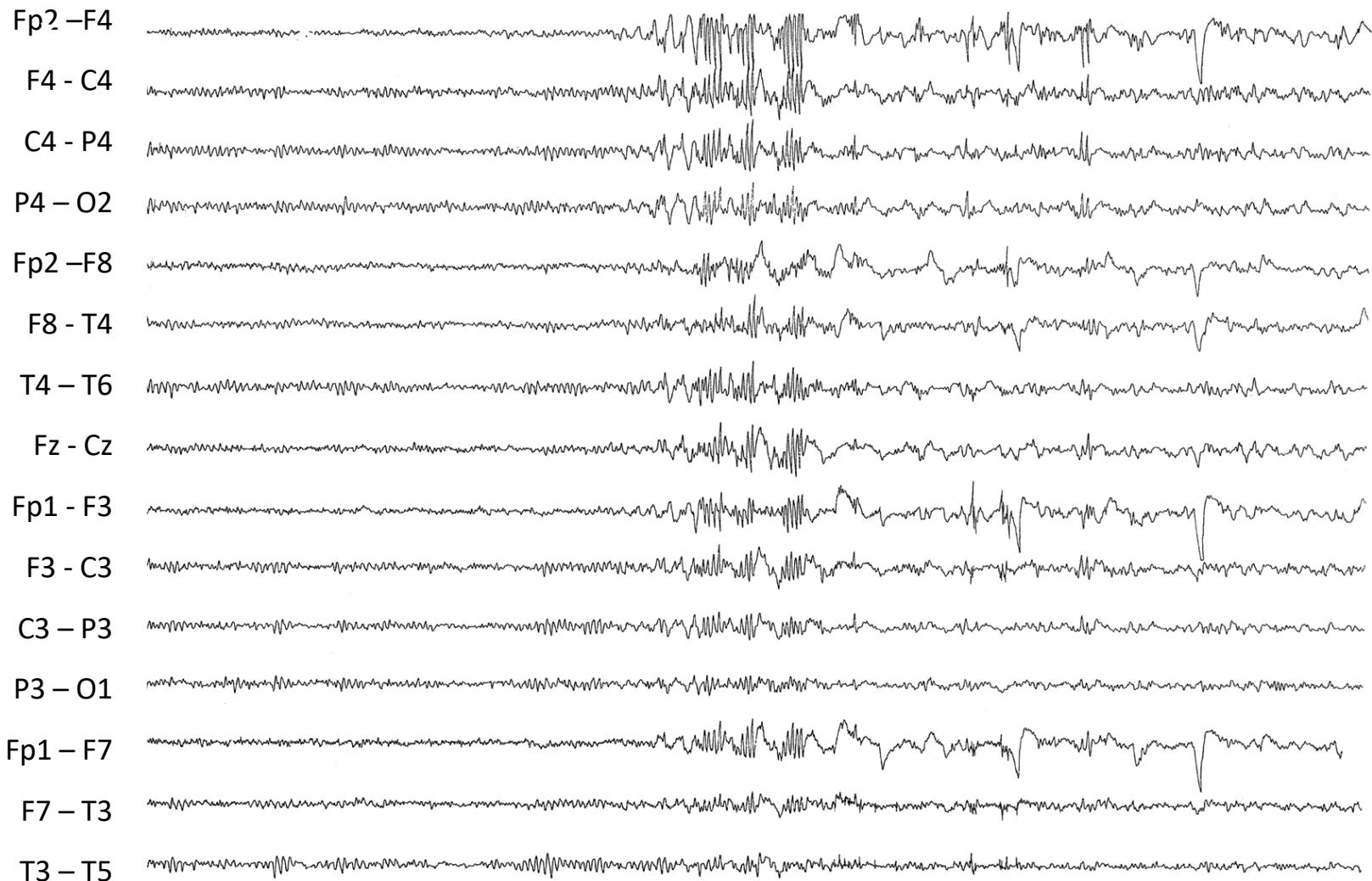
HPV 1'



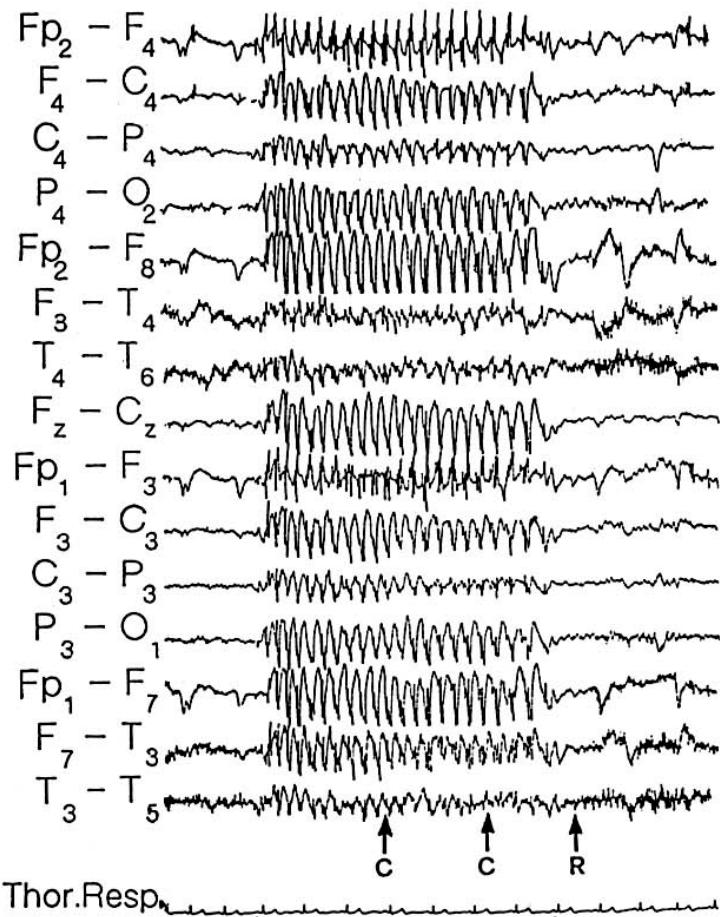
Mor. Sav. ♂ 38yrs n° 85650

....these eegs may resemble closely those of petit mal...epilepsy"

La bisincronia secondaria (SBS)

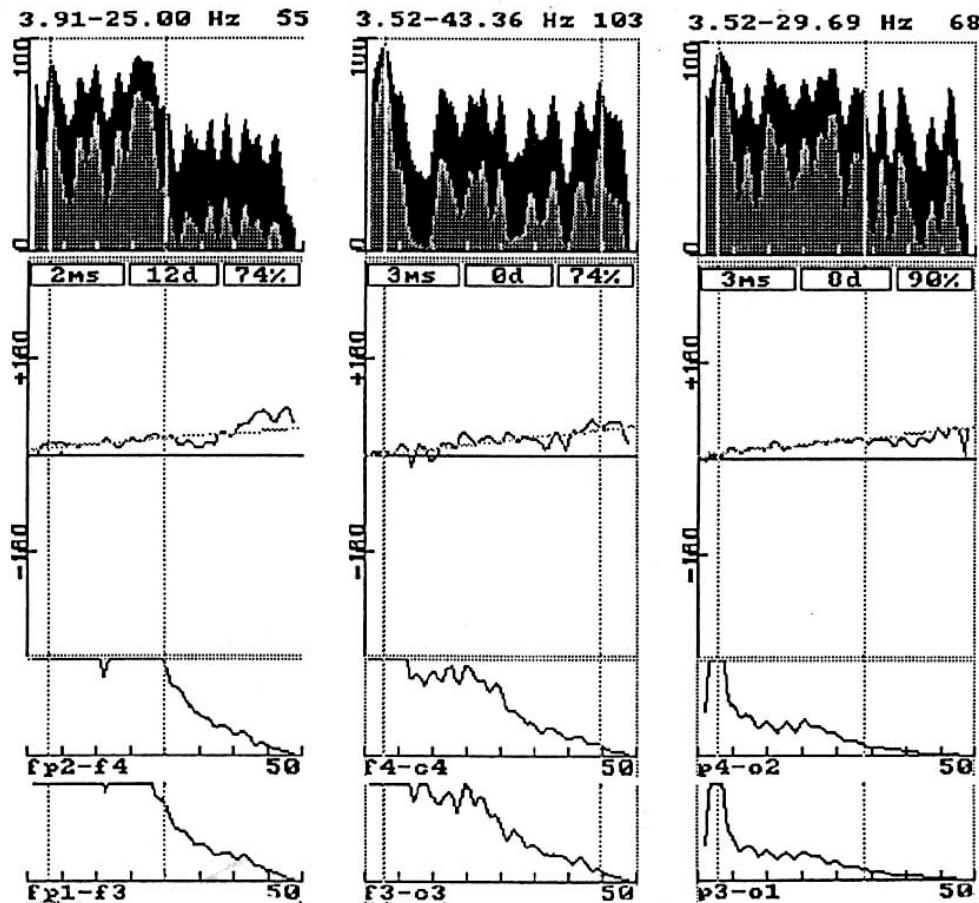


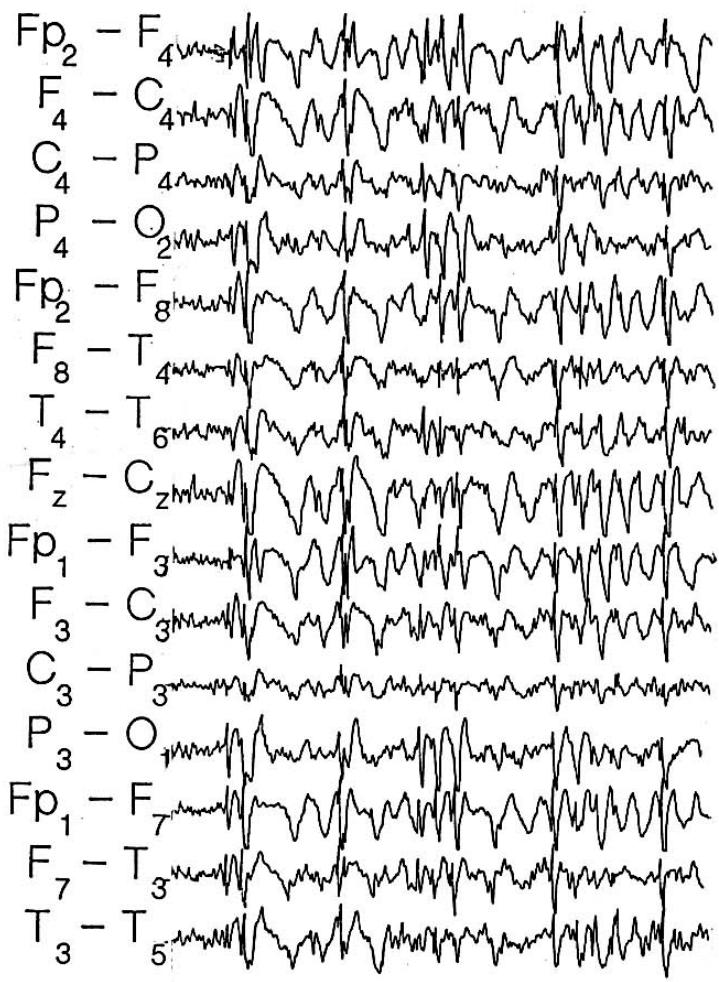
50 μ V
1 sec.



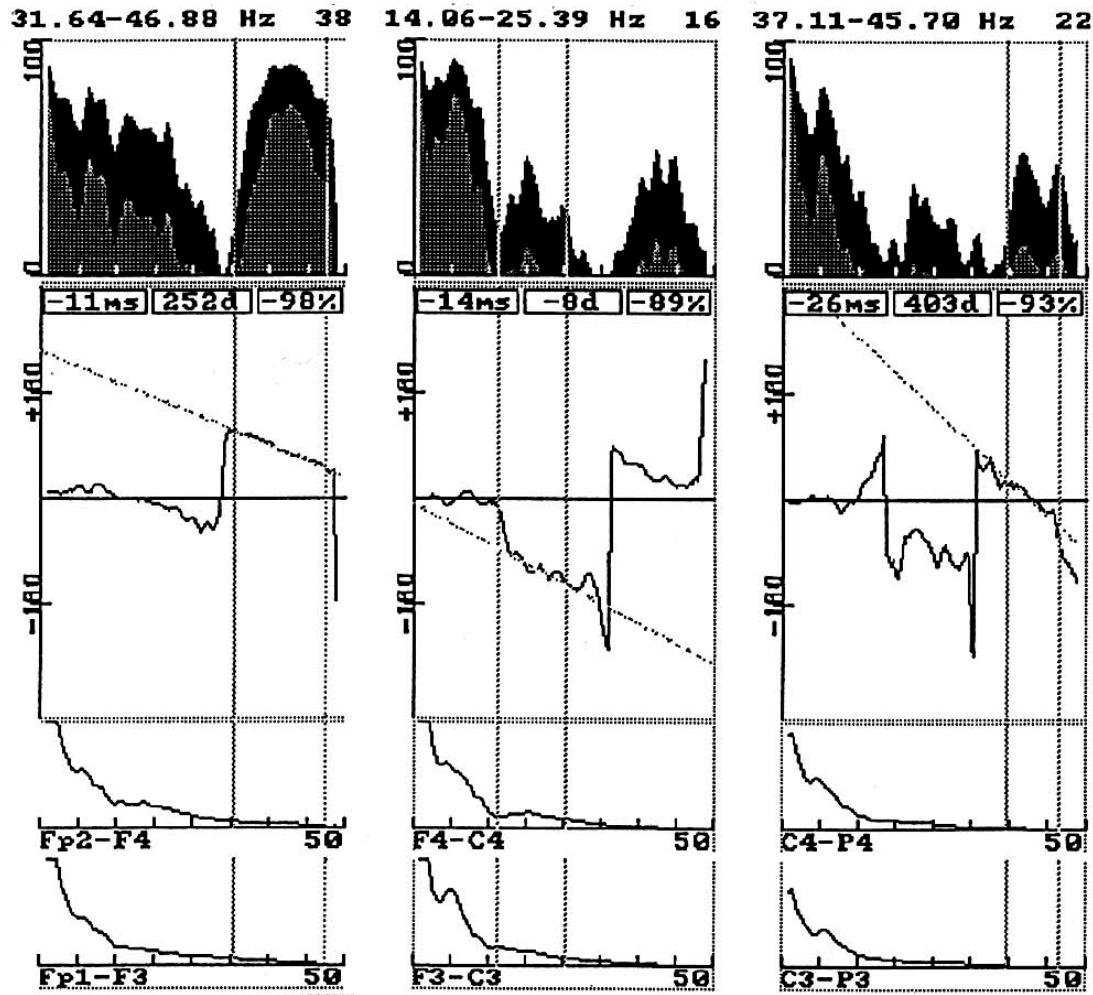
Cas.Pat. ♀ 34 yrs

50 μ V 1sec.

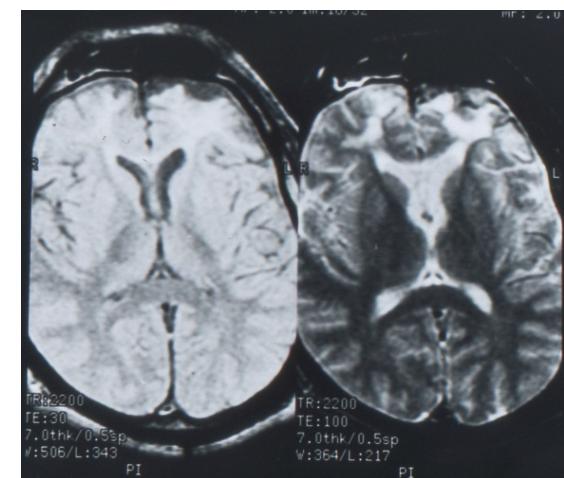
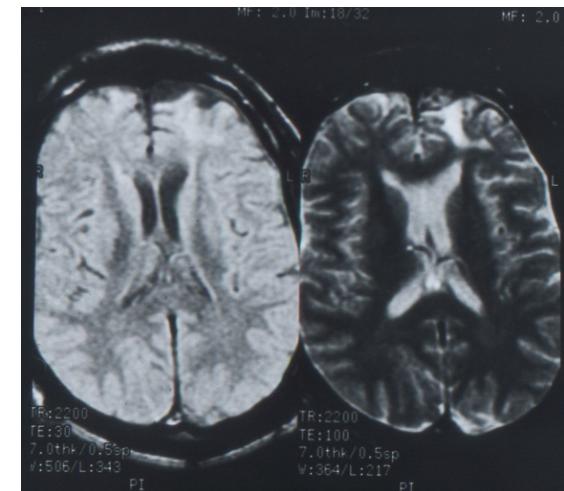
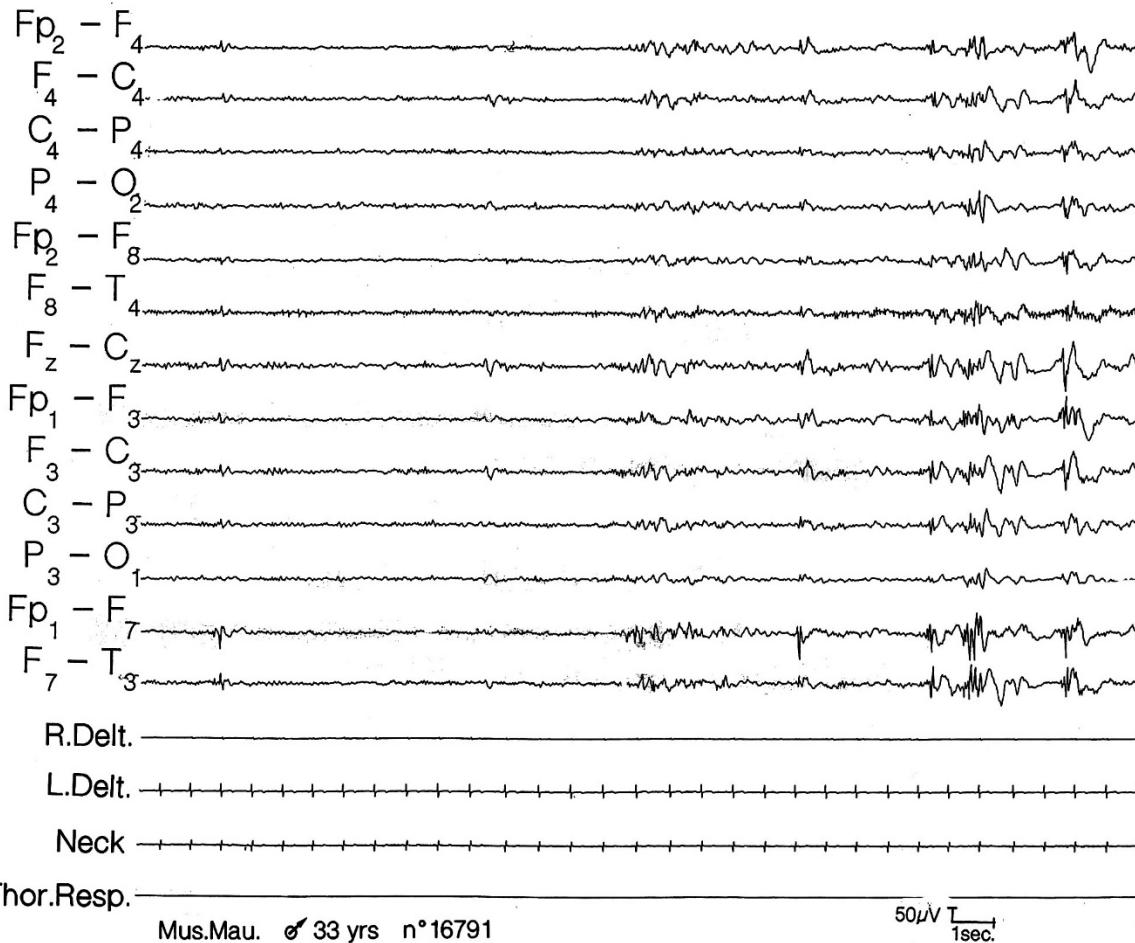




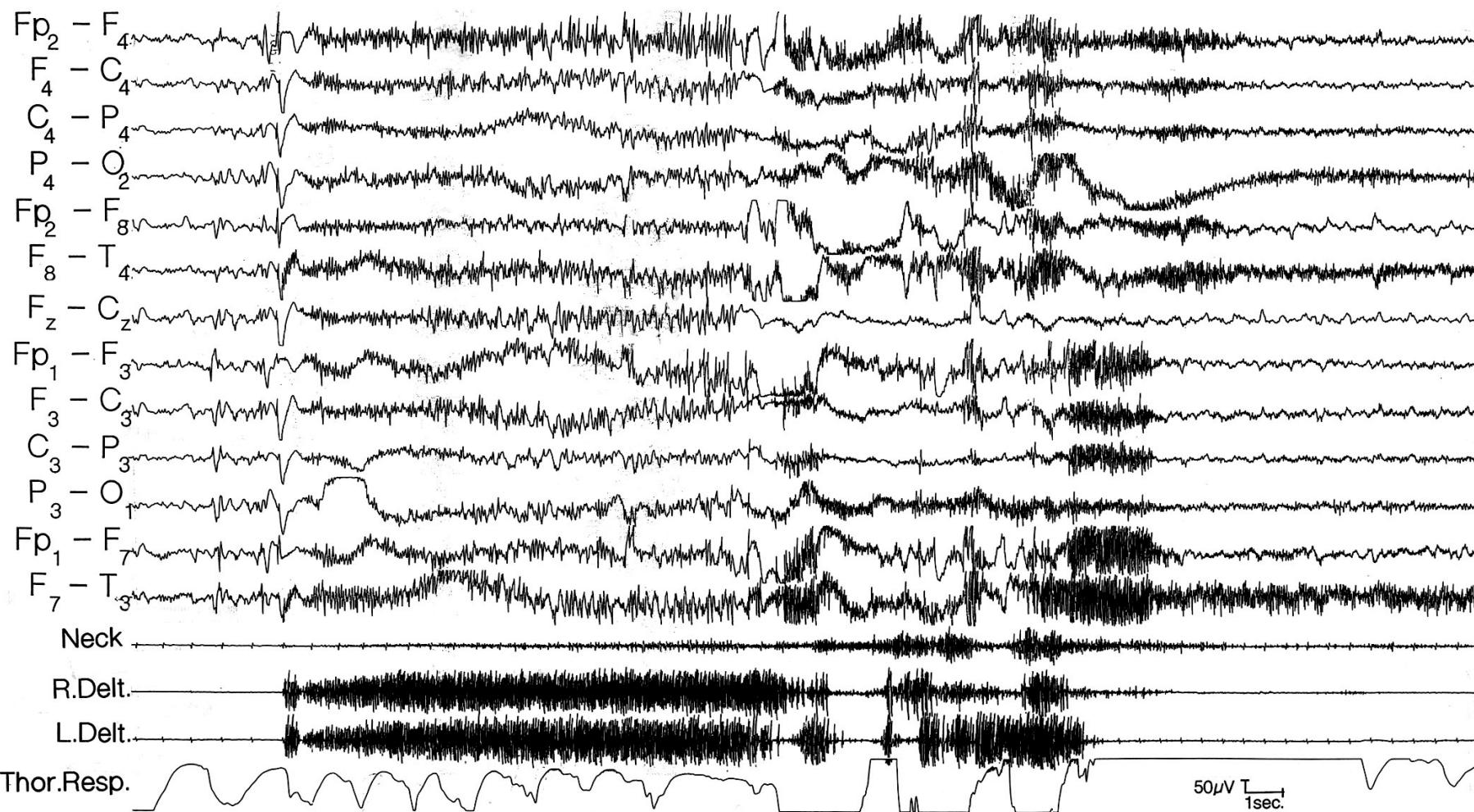
Mat.Sil. ♀ 53 yrs



SBS è un indice di prognosi sfavorevole nelle ep. focali



Clinical and EEG features of partial epilepsy with secondary bilateral synchrony. P.Tinuper 1995



M
C
D
R

<https://www.epilepsydiagnosis.org>

INTERNATIONAL LEAGUE AGAINST EPILEPSY  **International League Against Epilepsy**
Working toward a world where no person's life is limited by epilepsy
Established 1909

 International Reach

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Overview

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- Seizure Classification**
 - [Generalized seizures](#)
 - [Focal seizures](#)
 - [Focal/Generalized](#)
- Epilepsy syndromes**
 - [Neonatal/Infantile](#)
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 - [Structural](#)
 - [Metabolic](#)
 - [Immune](#)
 - [Infectious](#)
 - [Unknown](#)

EpilepsyDiagnosis.org

The ILAE Commission on Classification and Terminology welcomes you to EpilepsyDiagnosis.org, a cutting edge online diagnostic manual of the epilepsies.

Goal

The goal of [epilepsydiagnosis.org](https://www.epilepsydiagnosis.org) is to make available, in an easy to understand form, latest concepts relating to seizures and the epilepsies. The principle goal is to assist clinicians who look after people with epilepsy anywhere in the world to diagnose seizure type(s), classify epilepsy, diagnose epilepsy syndromes and define the etiology of the epilepsy. The site is principally designed for clinicians in primary and secondary care settings caring for people with epilepsy and we hope will also serve as a useful teaching aid.

Structure

The structure of this site reflects the importance of seizure type, syndrome, and etiology in clinical practice. On this website, you will find current classification concepts for seizures, with their clinical features, video examples, EEG correlate, differential diagnosis and related epilepsy syndromes. Epilepsy syndromes are detailed by their clinical features, seizure types, EEG, imaging and genetic correlates and differential diagnoses. The site includes sections on etiologies of epilepsies and epilepsy imitators with cross-referencing between these sections and seizure and syndrome sections.

Definition of epilepsy

Epilepsy is a disease of the brain defined by any of the following conditions:

- At least two unprovoked (or reflex) seizures occurring more than 24 hours apart

Francesca Bisulli

Barbara Mostacci

Laura Licchetta

Lidia Di Vito

Carlotta Stipa

Veronica Menghi

Lorenzo Ferri

Lorenzo Muccioli

Tecnici EEG



Atonic seizure

Generalized onset



Focal onset

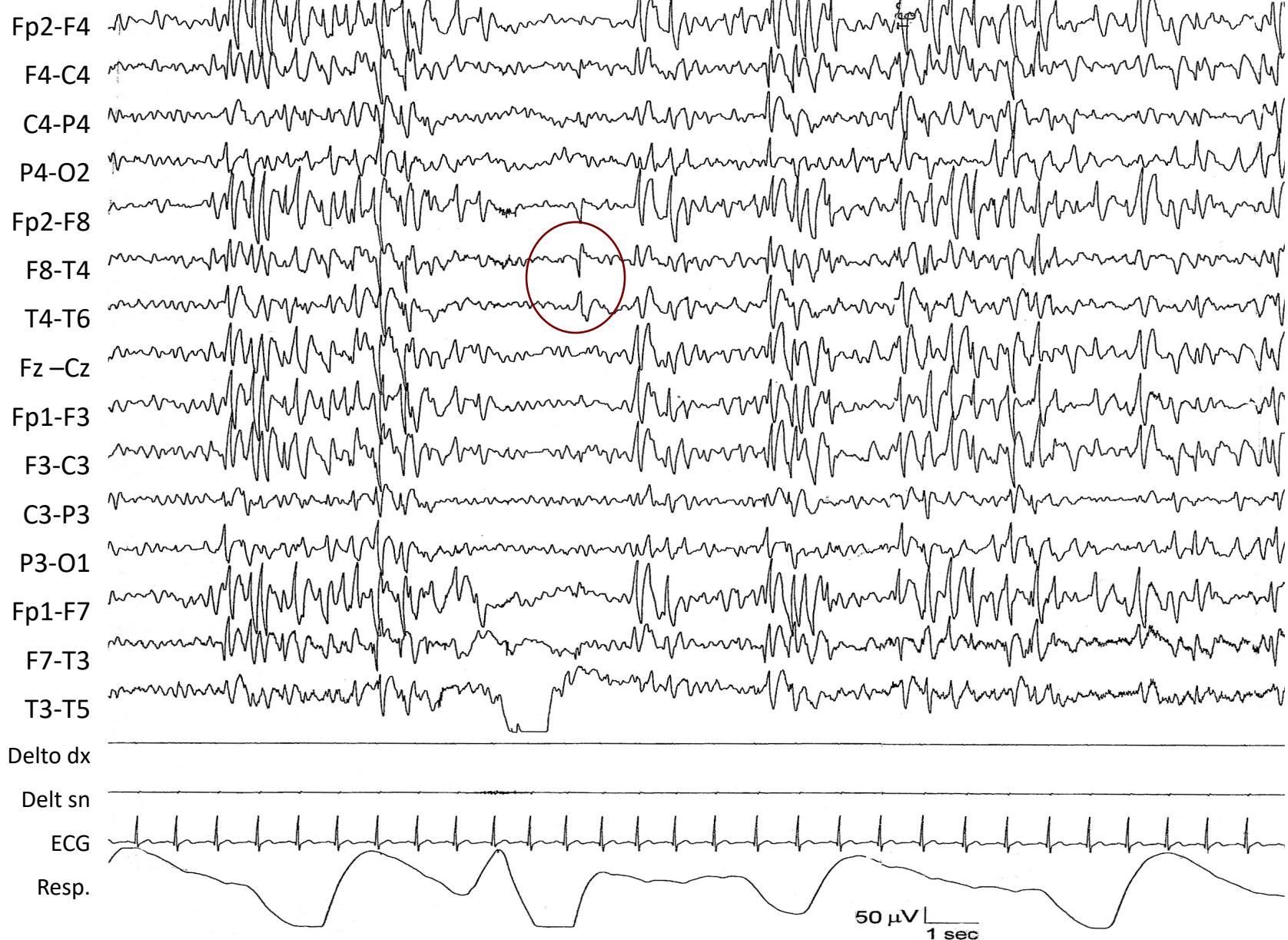


Epileptic spasm

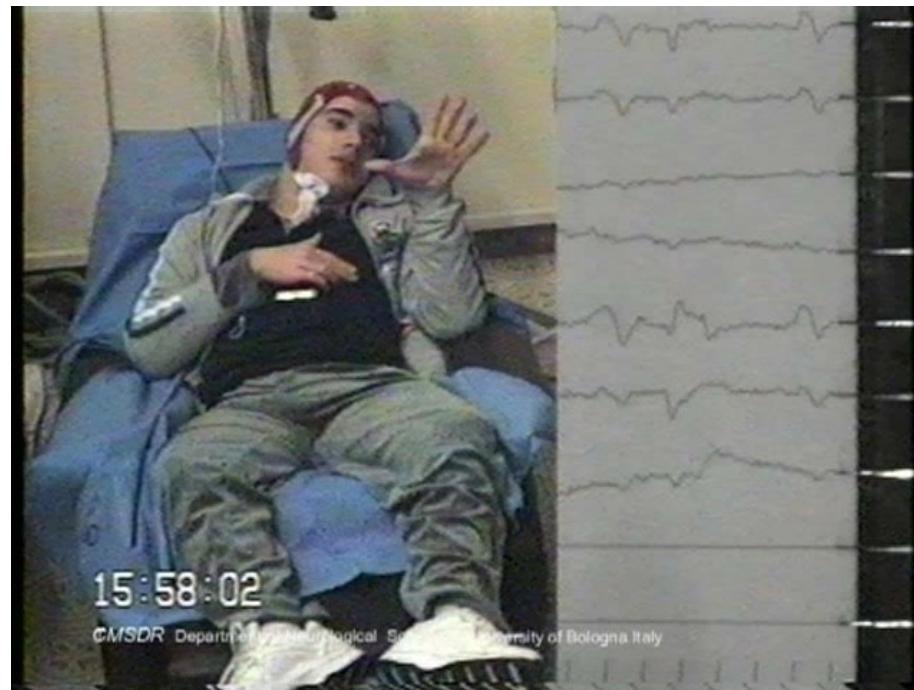
Generalized onset

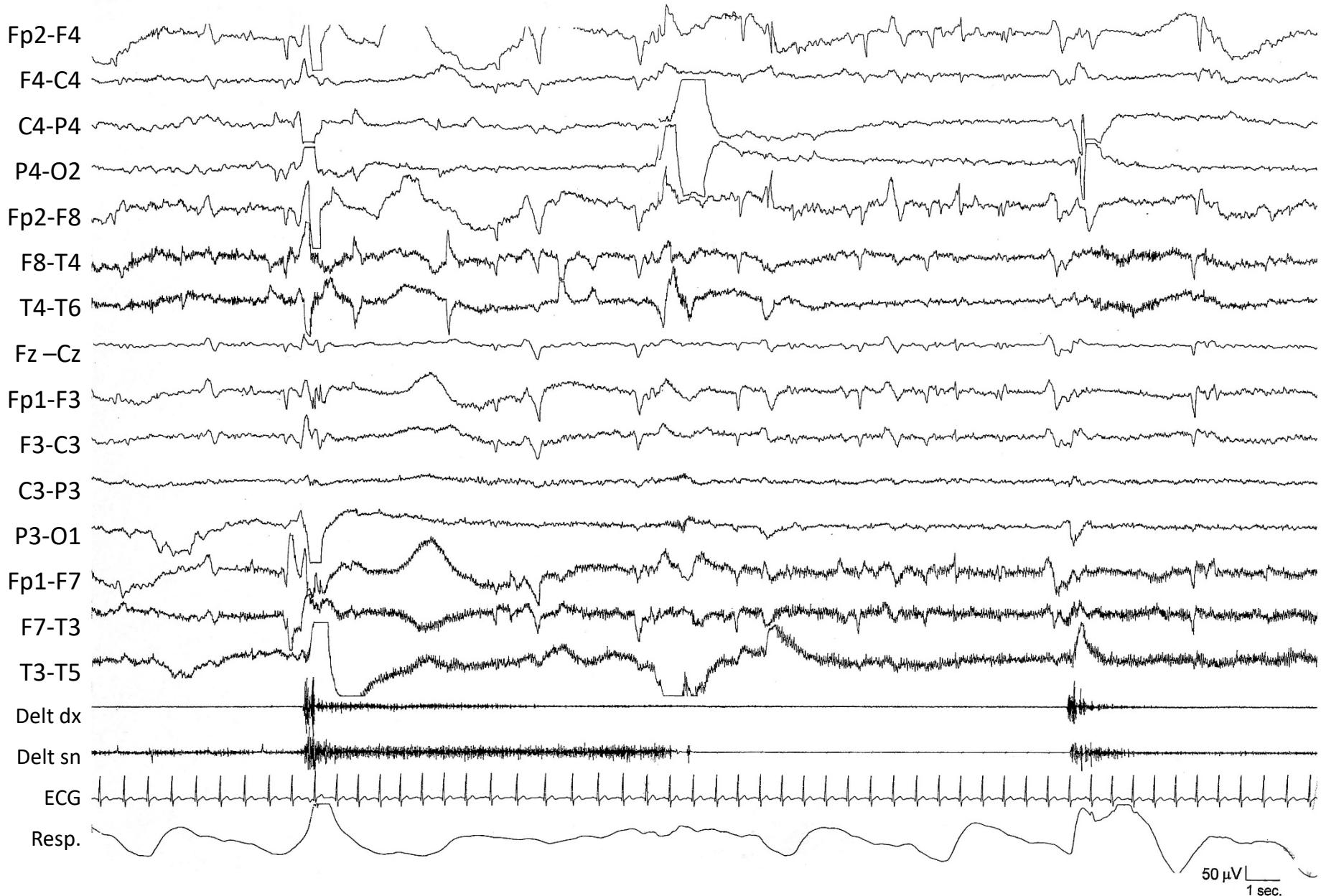
Focal onset

Sleep 1



14:51':33"

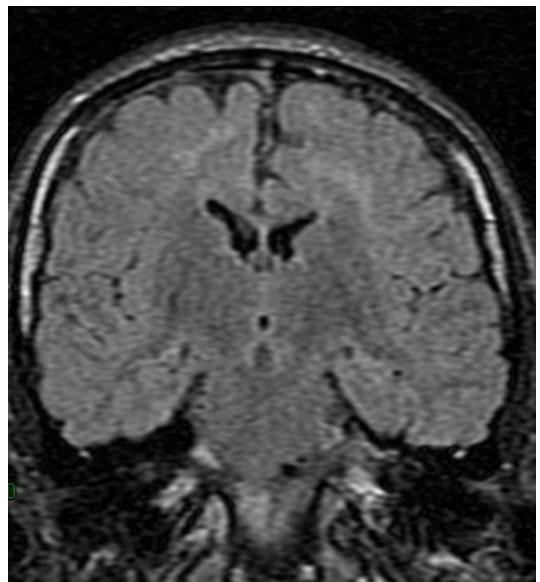
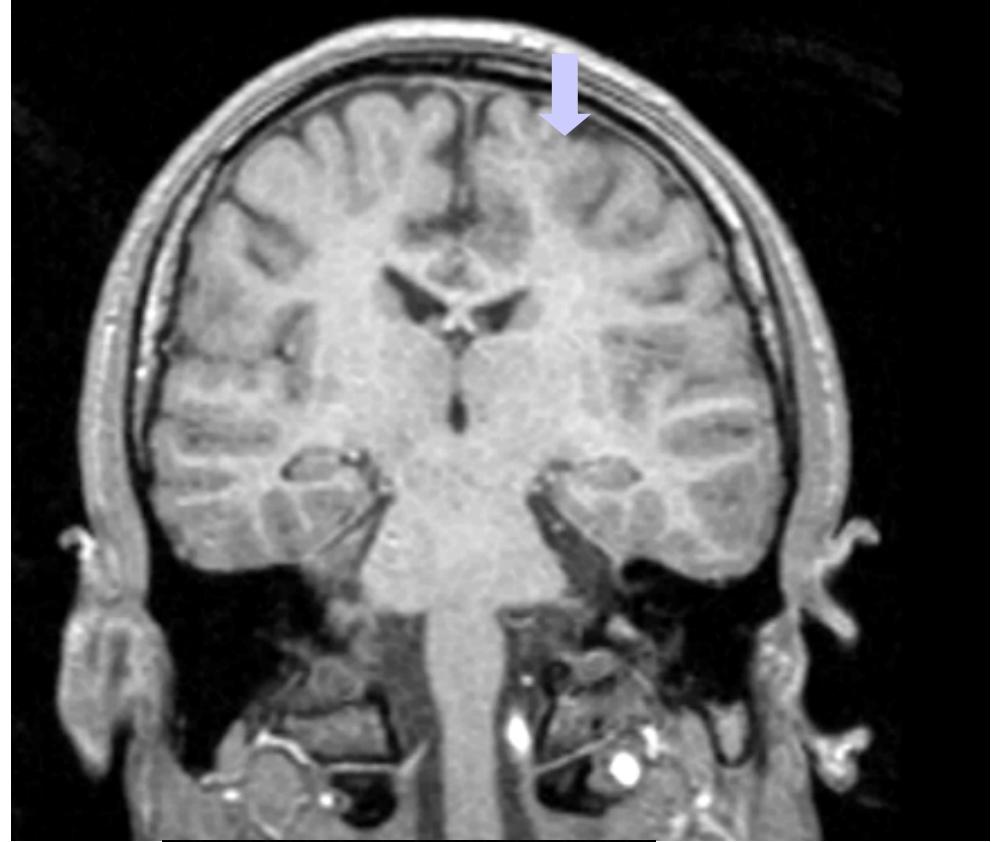
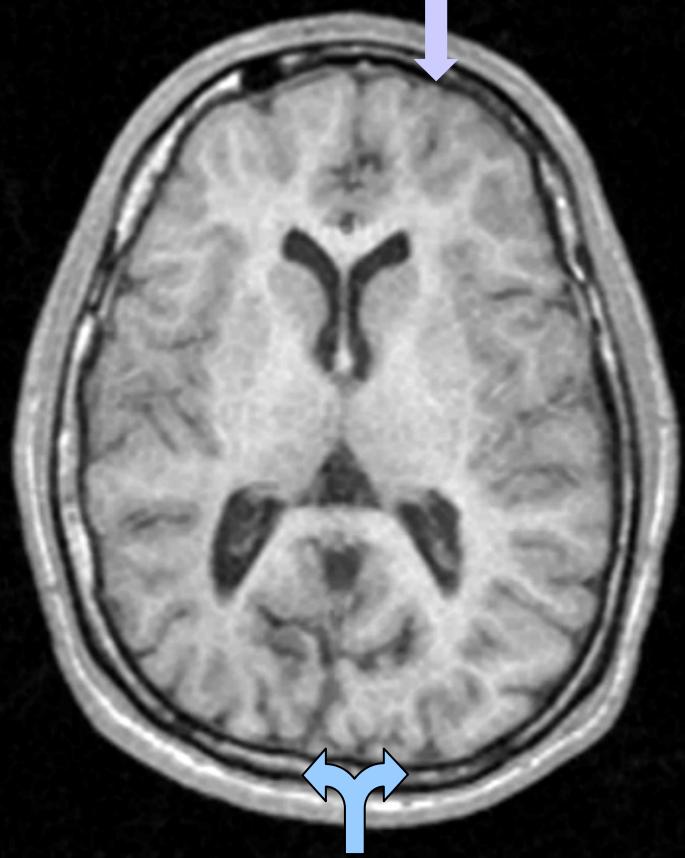




15:57':28" Spasmo in flessione + solleva il capo

Piccolo spasmo

Gat.Max. ♂ 20 aa 16-02-2011



Ulegiria bilaterale e
verosimile
polimicrogiria
frontale sinistra





(Z.G) female 57 yrs

Age 6 to 10 yrs: rare minor seizures with staring and tonic posturing

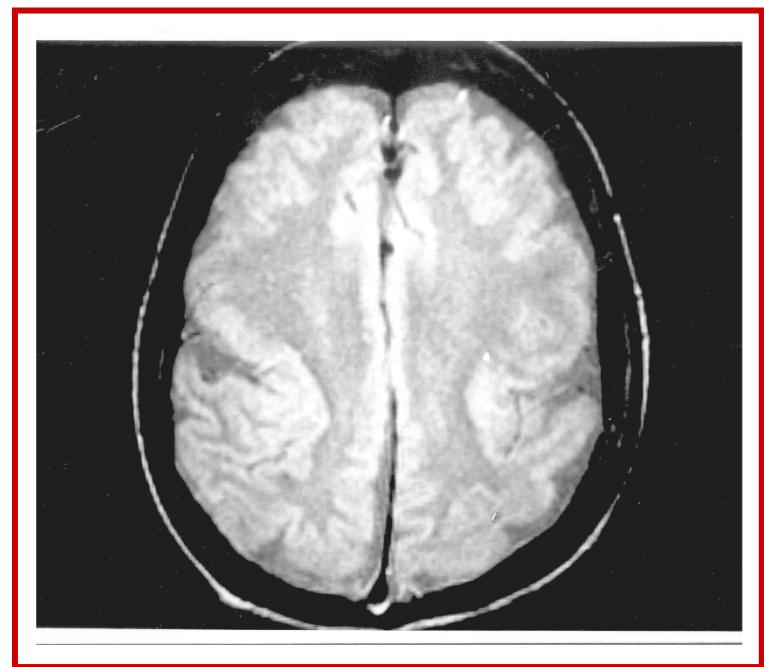
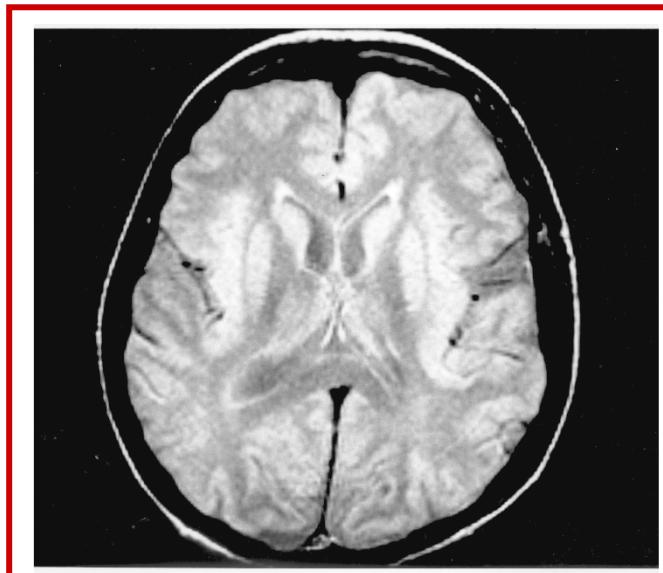
Age 11 to 27 yrs: TC s sometime preceded by L harm paresthesia

Since age 27: bilateral tonic contractions of peribuccal muscles (more on the left) with anteflexion of the head and trunk.

In cluster 3-4 / week

Partially responding to therapy (CBZ, VPA)

NMR: bilateral perisylvian polymicrogyria



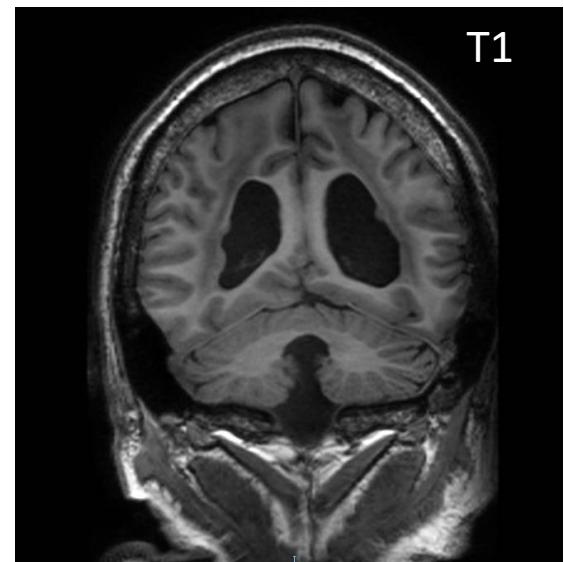
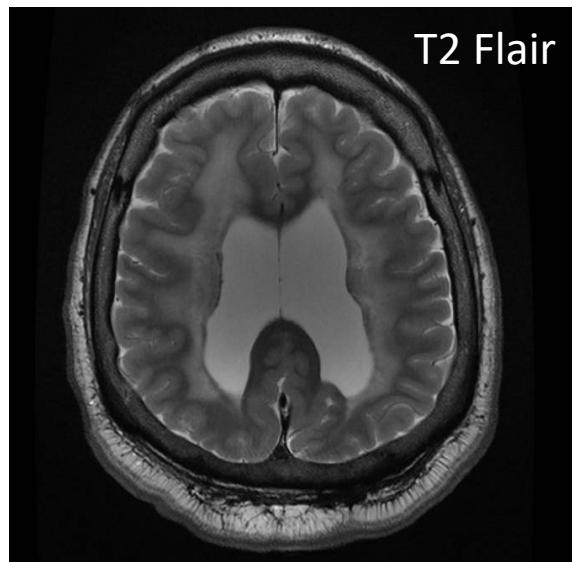
Pi. Gianni (49 aa)

Malattia di Whipple con interessamento linfonodale e surrenali

EON: strabismo divergente, ristagno rapido nello sguardo laterale verso sinistra, sindattilia del 2-3 dito del piede bilateralmente, atteggiamento distonico in estensione degli alluci bilateralmente ($dx > sn$), lieve ipotestenia a tricipite dx e bicipite dx, tremore postulare AS dx, marcia atassica con tendenza alla lateropulsione verso destra alla chiusura degli occhi.

CGH- Array: nn

Malformazione congenita complessa con encefalocele, complesso di Dandy- Walker, anomalie della migrazione neuronale e leucoencefalopatia.



Pi. Gianni (12 aa)

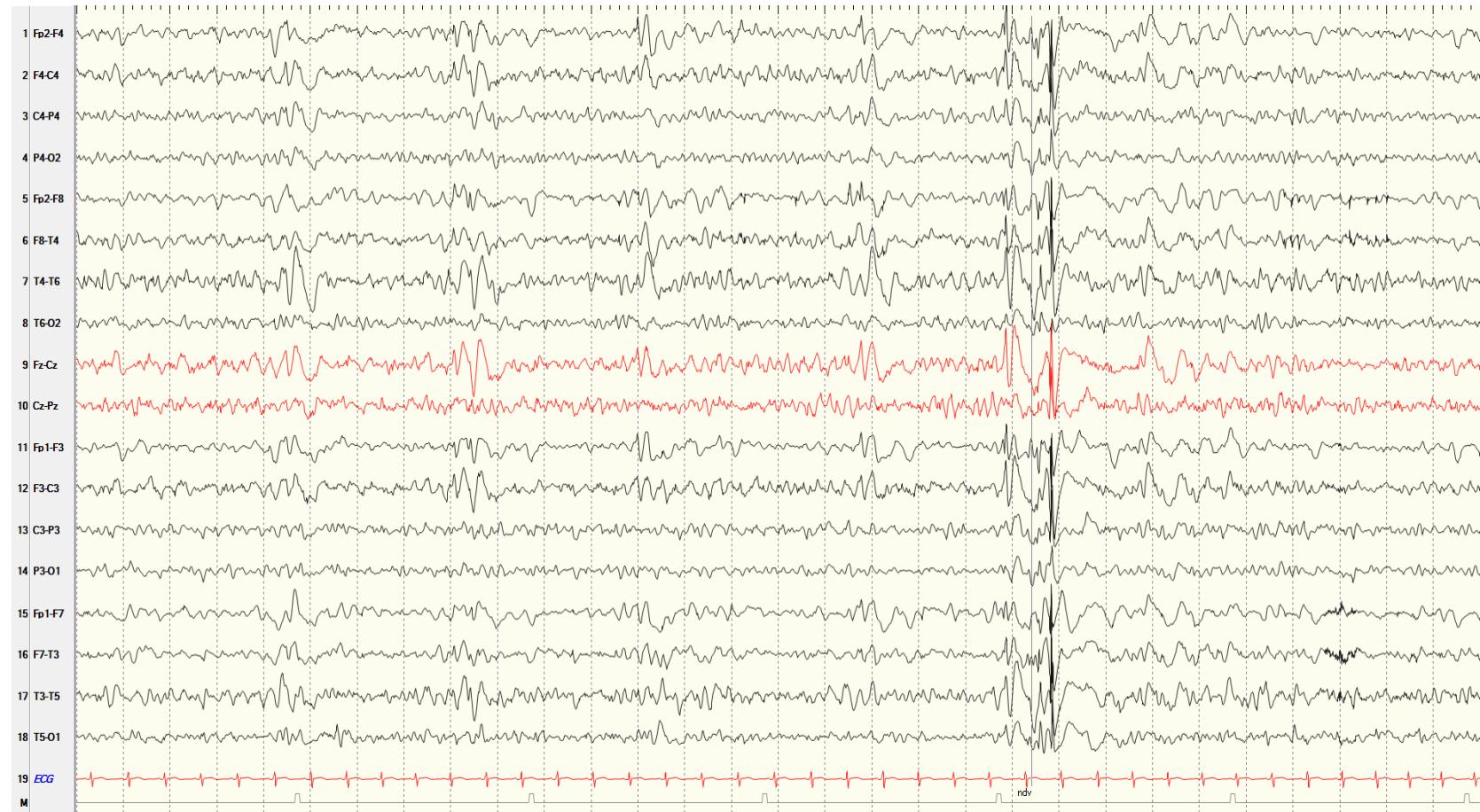


Pi. Gianni (12 aa)



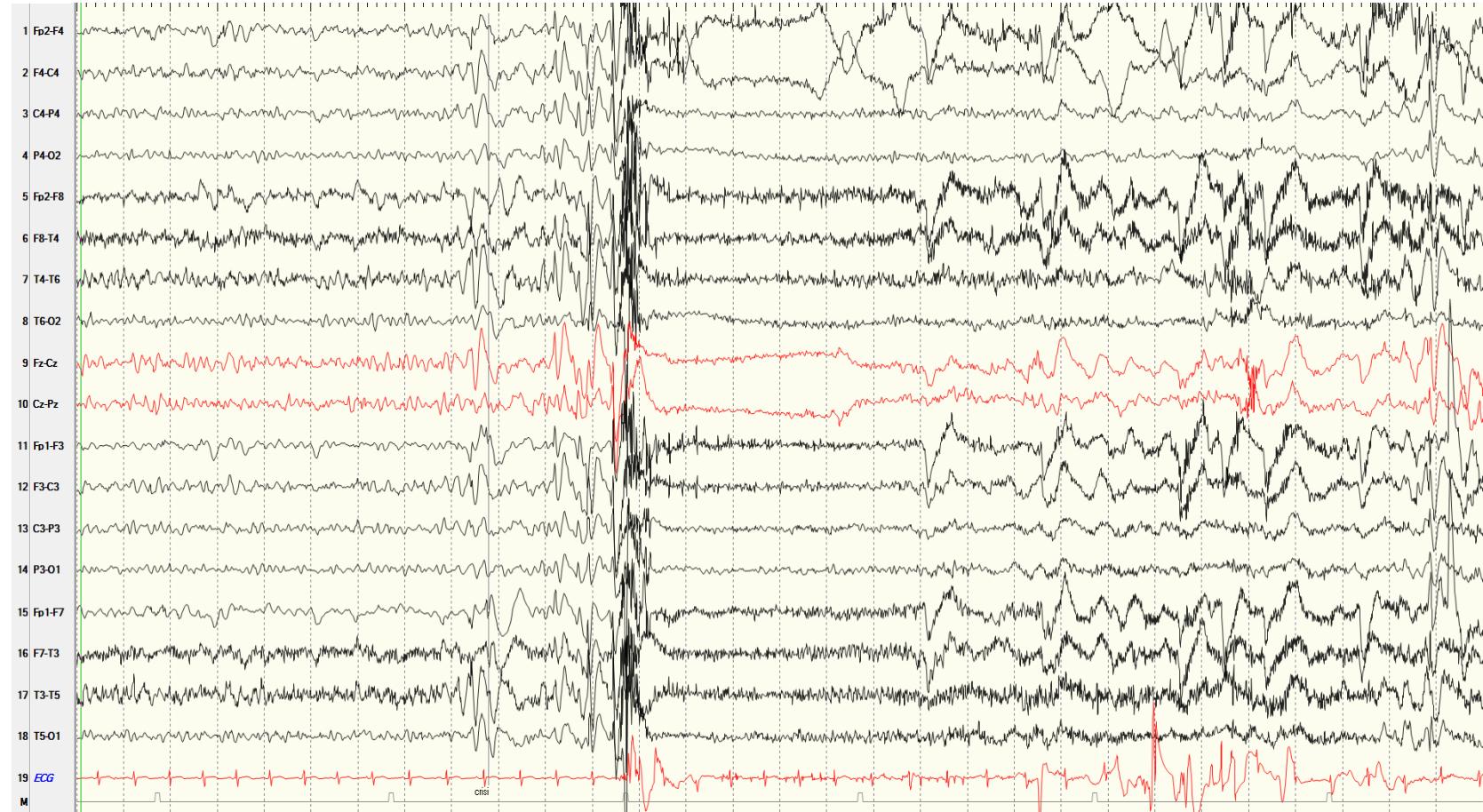
02-11-2012

Tonin luca

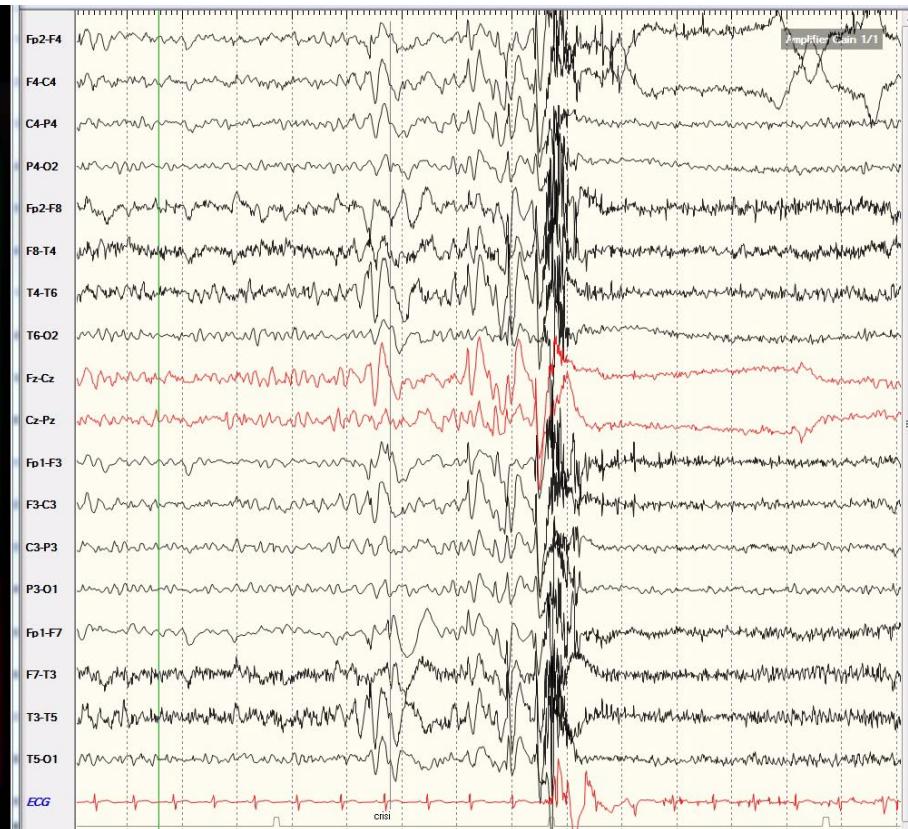


crisi

Tonin luca



Tonin luca



Francesca Bisulli

Barbara Mostacci

Laura Licchetta

Patrizia Avoni

Lidia Di Vito

Carlotta Stipa

Veronica Menghi

Lorenzo Ferri

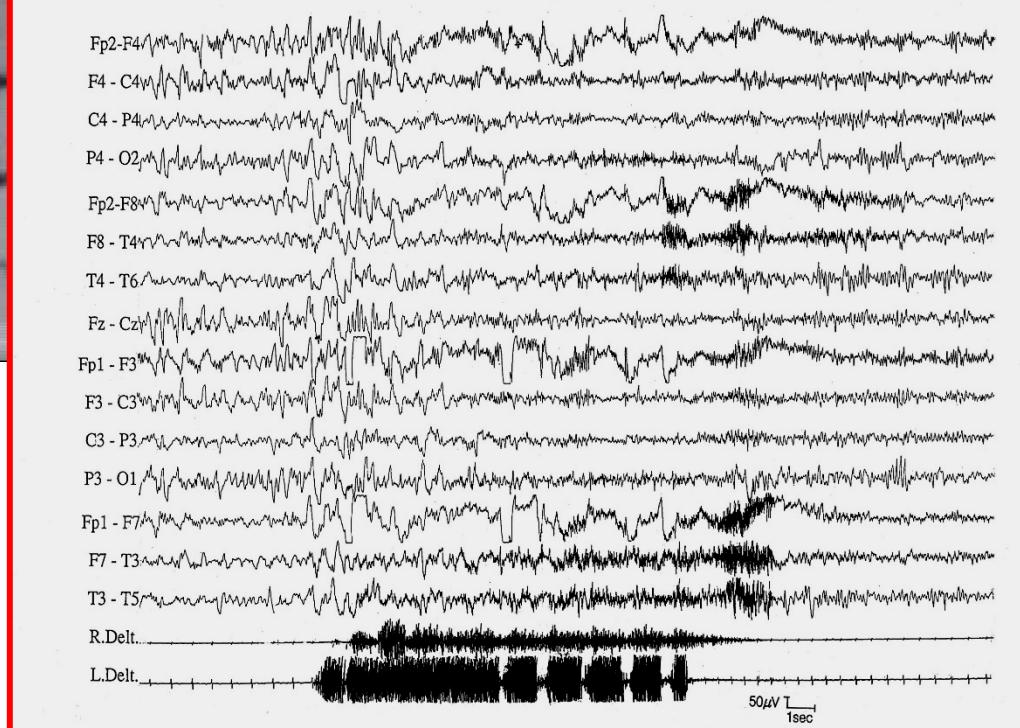
Tecnici EEG

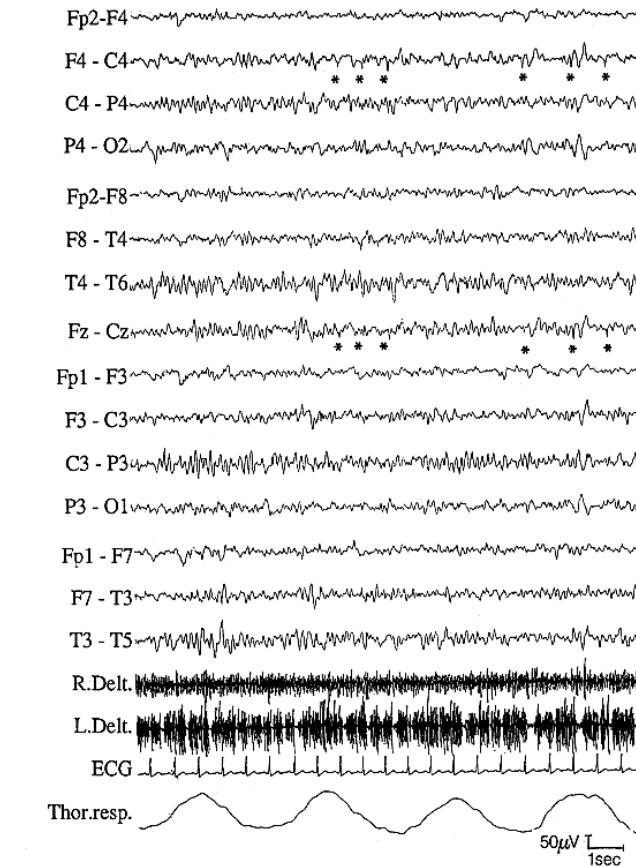
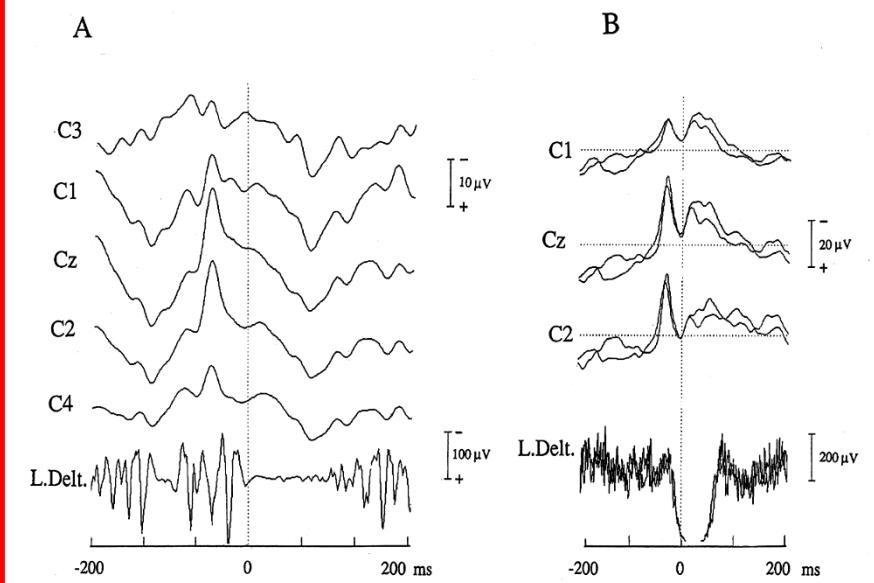


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tonic and atonic seizure

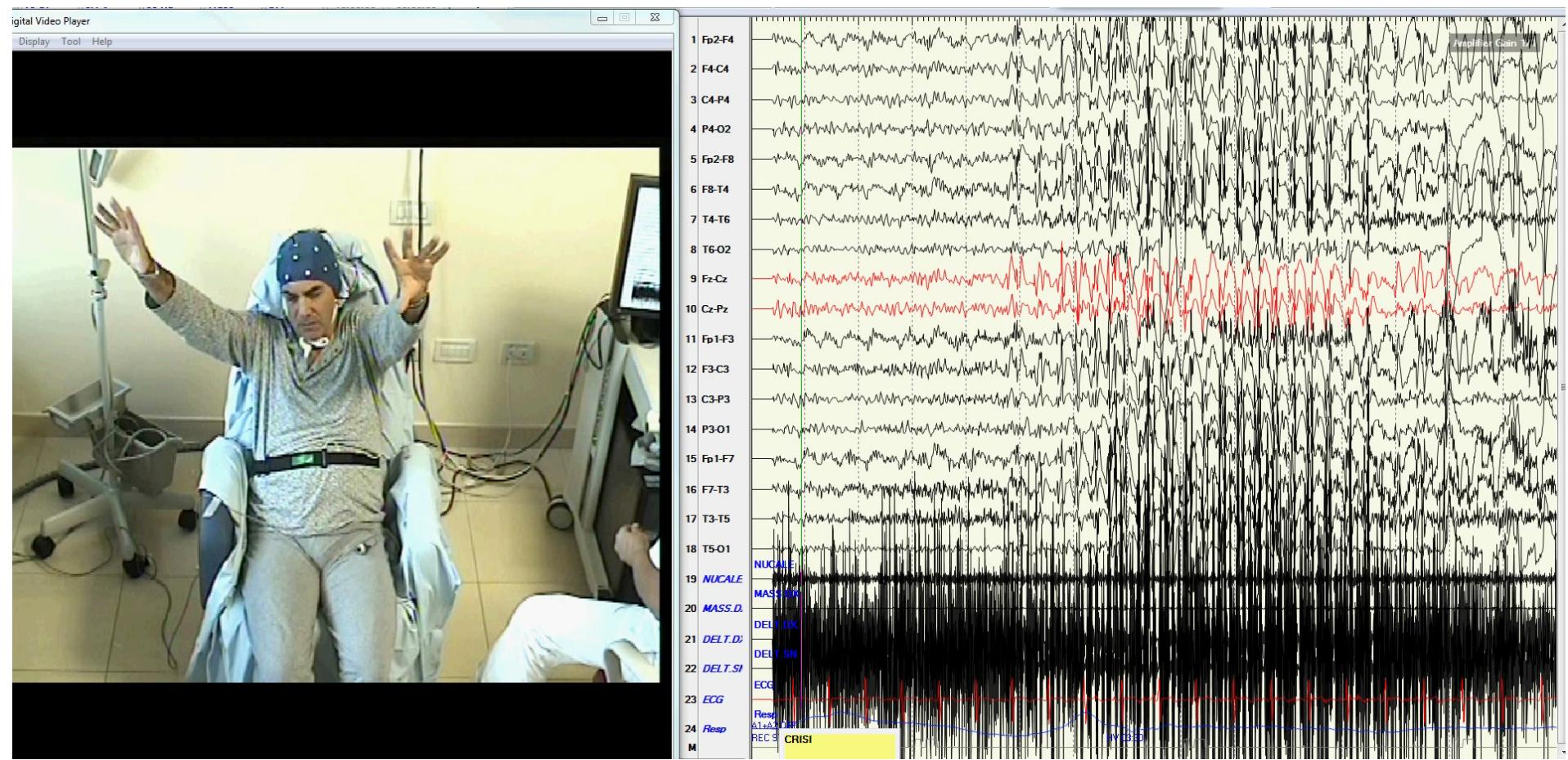




clonic+tonic+atonic







Clinical Research

Distinct Behavioral and EEG Topographic Correlates of Loss of Consciousness in Absences

*†P. Vuilleumier, *F. Assal, *O. Blanke, and *P. Jallon

