Pattern EEG critici

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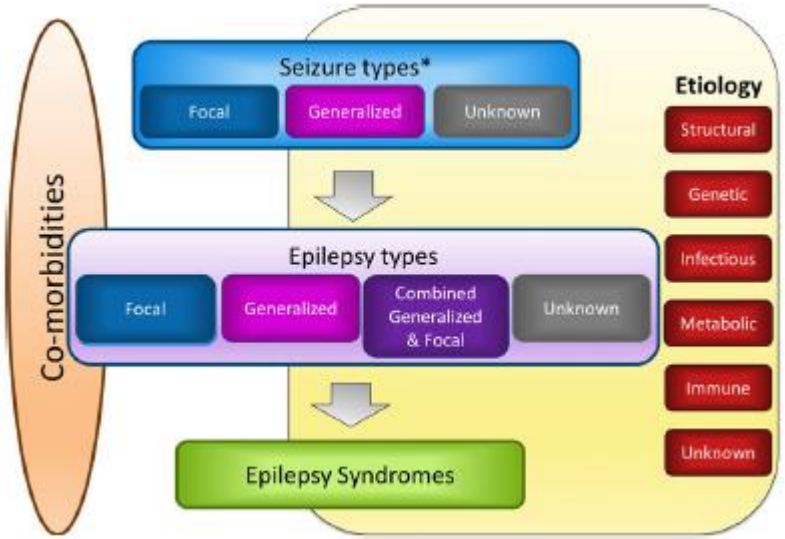
4° CORSO RESIDENZIALE
EEG e POTENZIALI EVOCATI
22 – 27 NOVEMBRE 2021



ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

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

Nonmotor Onset
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic–clonic

Generalized Onset

Motor
- tonic–clonic
- clonic
- tonic
- myoclonic
- myoclonic–tonic–clonic
- myoclonic–atonic
- atonic
- epileptic spasms

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

Unknown Onset

Motor
- tonic–clonic
- epileptic spasms

Nonmotor
- behavior arrest

Unclassified [3]

Scheffer 2017; Fisher 2017

Epilepsy Syndrome
Neonate Infant

Self-limited epilepsies

- Self-limited neonatal epilepsy
- Self limited infantile epilepsy
- Febrile seizures+ (part of GEFS+)
- Myoclonic epilepsy in infancy

Developmental and epileptic encephalopathies (DEEs)

- Early-infantile DEE
- Epilepsy of infancy with migrating focal seizures
- Infantile spasms syndrome
- Dravet syndrome

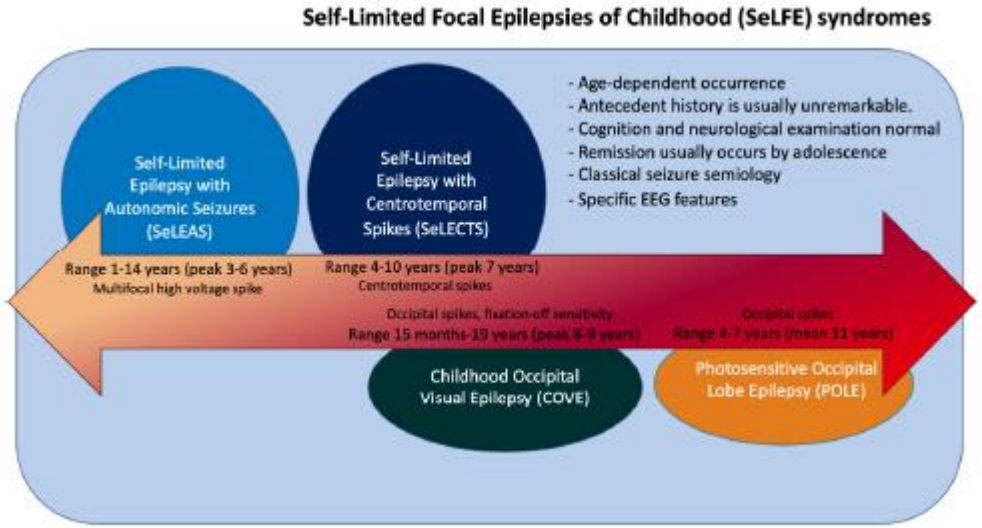
Etiology-specific epilepsies

- *KCNQ2*-DEE
- Pyridoxine-dependent (*ADH7A1*)-DEE
- Pyridox(am)ine-5'-phosphate-deficiency (*PNPO*)-DEE
- *CDKL5*-DEE
- *PCDH19* clustering epilepsy
- GLUT1DS-DEE
- Sturge-Weber Syndrome
- Gelastic seizures with hypothalamic hamartoma

ILAE Classification & Definition of Epilepsy Syndromes in the Neonate and Infant: Position Statement by the ILAE Task Force on Nosology and Definitions

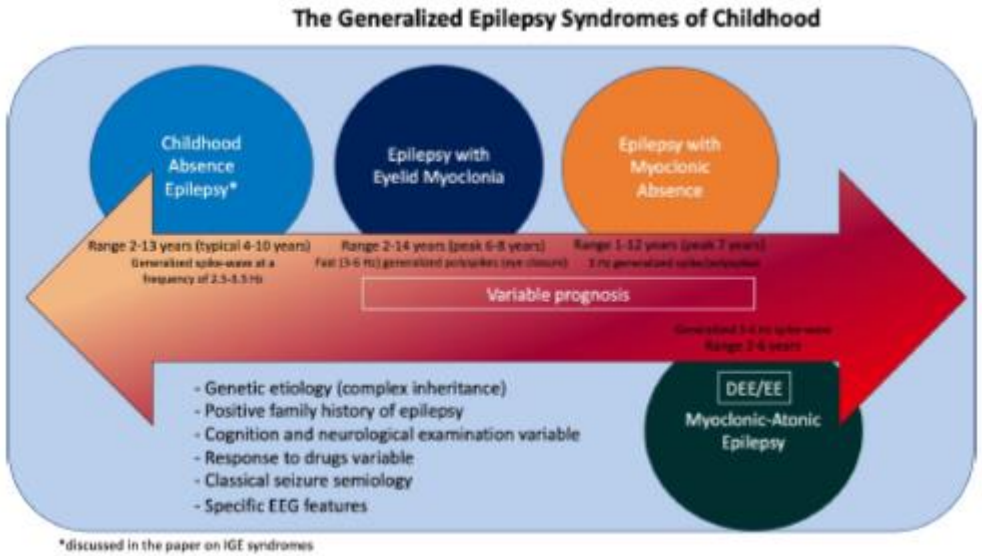
Sameer M Zuberi[1], Elaine Wirrell[2], Elissa Yozawitz[3], Jo M Wilmshurst[4], Nicola Specchio[5], Kate Riney[6], Ronit Pressler[7], Stephane Auvin[8], Pauline Samia[9], Edouard Hirsch[10], O Carter Snead[11], Samuel Wiebe[12], J Helen Cross[13], Paolo Tinuper[14,15], Ingrid E Scheffer[16], Rima Nabbout[17]

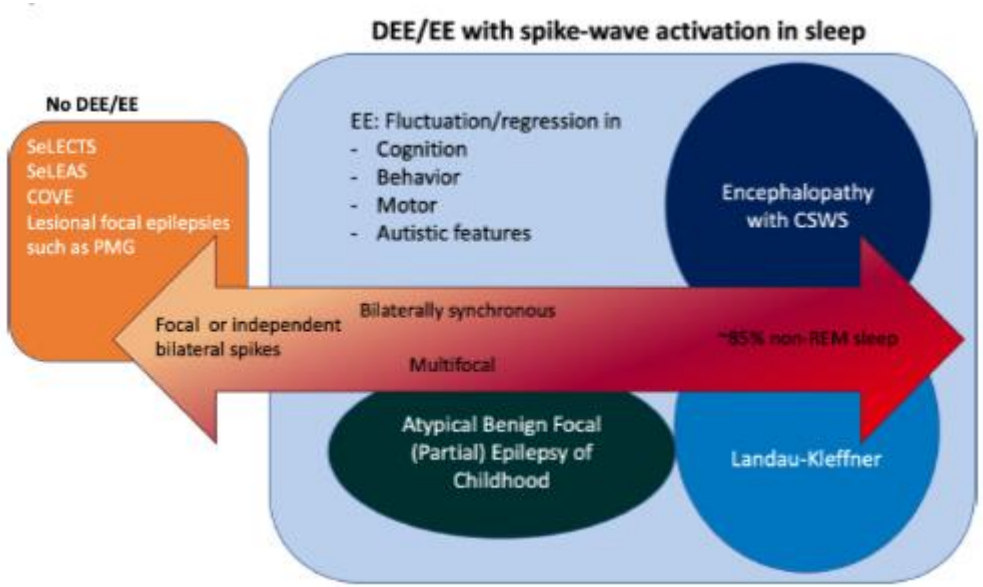
Epilepsy Syndrome Childhood



ILAE Classification and Definition of Epilepsy Syndromes with Onset in Childhood: Position Paper by the ILAE Task Force on Nosology and Definitions

N Specchio[1], EC Wirrell[2*], IE Scheffer[3], R Nabbout[4], K Riney[5], P Samia[6], SM Zuberi[7], JM Wilmshurst[8], E Yozawitz[9], R Pressler[10], E Hirsch[11], S Wiebe[12], JH Cross[13], P Tinuper[14], S Auvin[15]

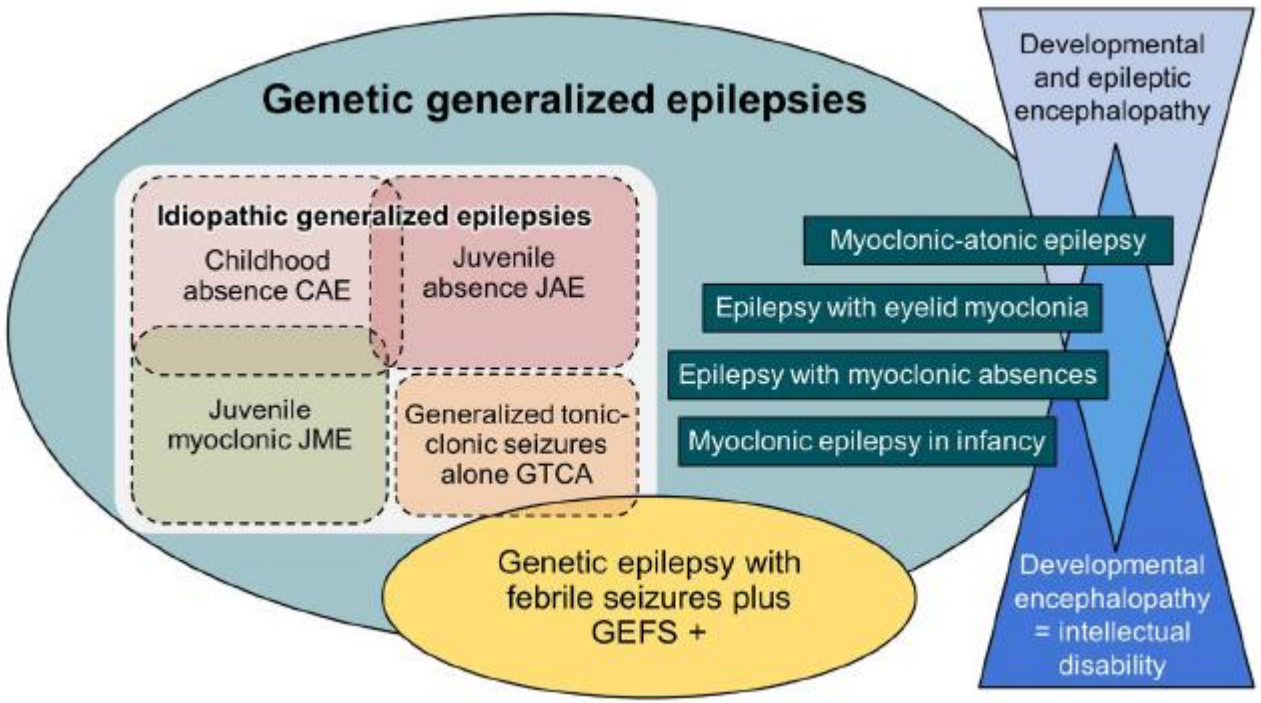




Epilepsy Syndrome:
Idiopathic Generalized

ILAE Definition of the Idiopathic Generalized Epilepsy Syndromes: Position Statement by the ILAE Task Force on Nosology and Definitions

E Hirsch[1], J French[2], IE Scheffer[3], SM Zuberi[4], E Trinka[5,6], N Specchio[7], E Somerville[8], P Samia[9], K Riney[10], R Nabbout[11], S Jain[12], A Bogacz[13], T Alsaadi[14], JM Wilmshurst[15], S Auvin[16], S Wiebe[17], P Tinuper[18,19]*, E Wirrell[20]*



ILAE Classification and Definition of Epilepsy Syndromes with Onset at a Variable Age: Position Statement by the ILAE Task Force on Nosology and Definitions

Authors: K Riney[1,2], A Bogacz[3], E Somerville[4,5], E Hirsch[6-8], R Nabbout[9-13], IE Scheffer[14], SM Zuberi[11,15,16], T Alsaadi[17], S Jain[18], J French[19], N Specchio[20,21], E Trinka[22-24], S Wiebe[25], S Auvin[10,13,26], EC Wirrell*[27], P Tinuper*[21,28]

Focal epilepsy syndromes

- Sleep related hypermotor epilepsy
- Familial focal epilepsy with variable foci
- Epilepsy with auditory features

Generalized epilepsy syndromes

- Idiopathic generalized epilepsies (juvenile myoclonic epilepsy, juvenile absence epilepsy, epilepsy with generalized tonic-clonic seizures alone)

Combined generalized and focal epilepsy syndrome

- Epilepsy with reading induced seizures

Developmental and epileptic encephalopathy

- Progressive myoclonus epilepsies

Etiology-specific epilepsy syndromes

- Mesial temporal lobe epilepsy with hippocampal sclerosis
- Rasmussen encephalitis

OUTLINE DELLA PRESENTAZIONE
- PATTERN CRITICI DI QUESTE SINDROMI

CHILDHOOD EPILEPSY SYNDROME
- CHILDHOOD ABSENCE EPILEPSY
- EPILEPSY WITH EYELIDS MYOCLONIA
- EPILEPSY WITH MYOCLONIC ABSENCES
- MYOCLONIC-ATONIC EPILEPSY

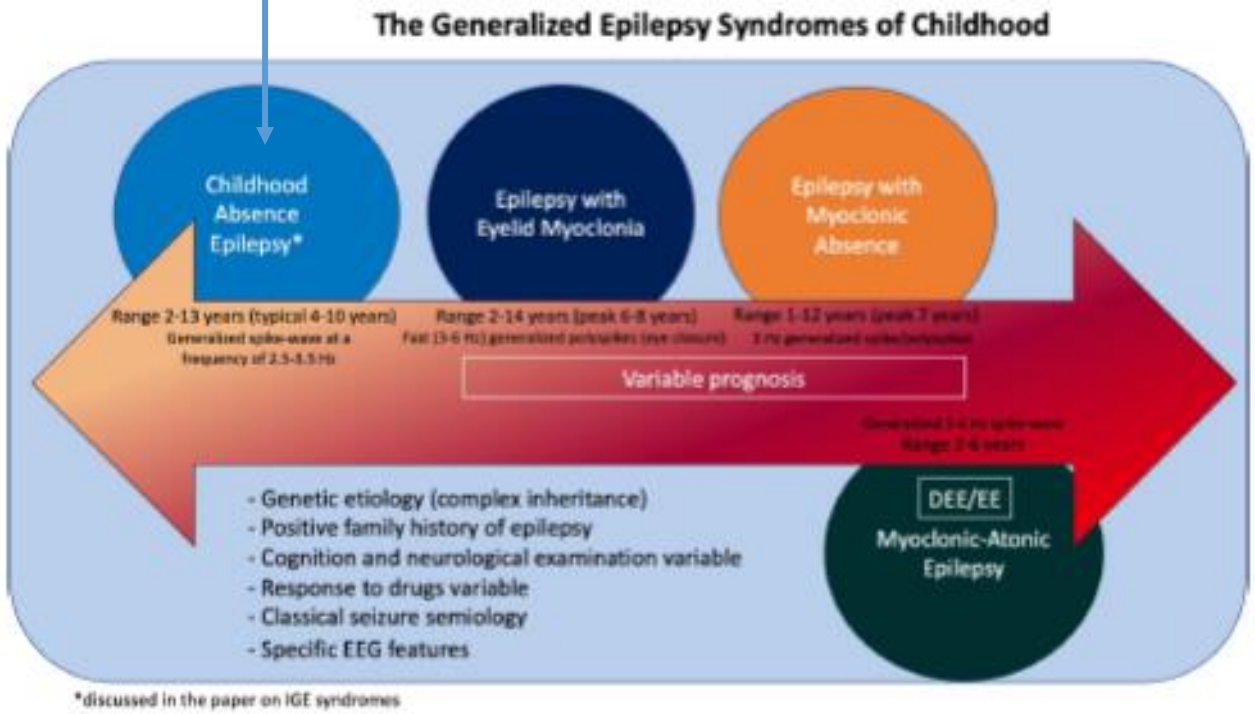
DEE/EE WITH SPIKE-AND-WAVE SLEEP ACTIVATION
- ENCEPHALOPATHY WITH CONTINUOUS SPIKE-AND-WAVE SLEEP (CSWS)

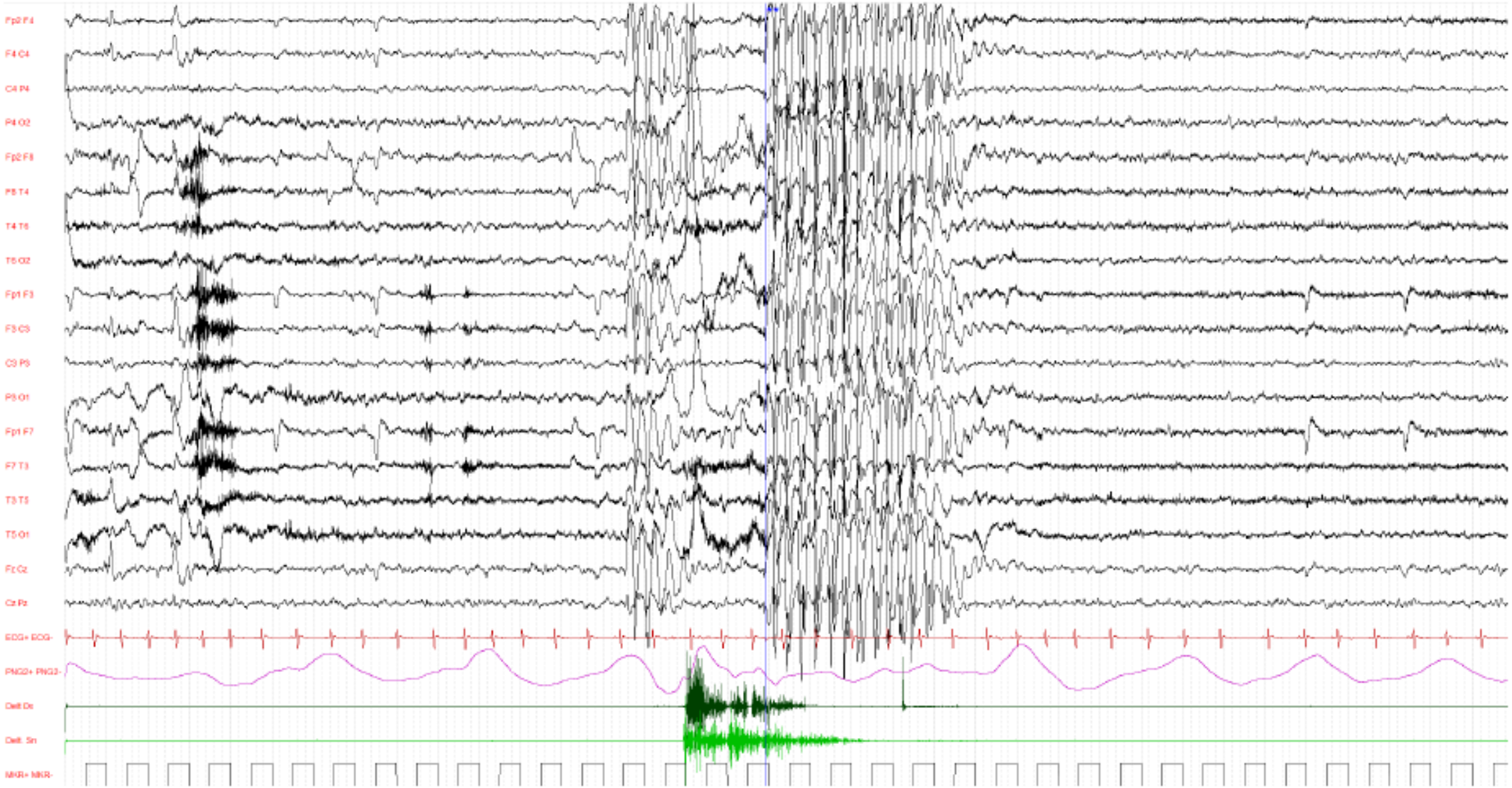
ATYPICAL BENIGN FOCAL EPILEPSY
- NEGATIVE MYOCLONUS
- CSWS

SELF-LIMITED FOCAL EPILEPSY OF CHILDHOOD
- WITH AUTONOMIC SEIZURES
- WITH CENTRO-TEMPORAL SPIKES

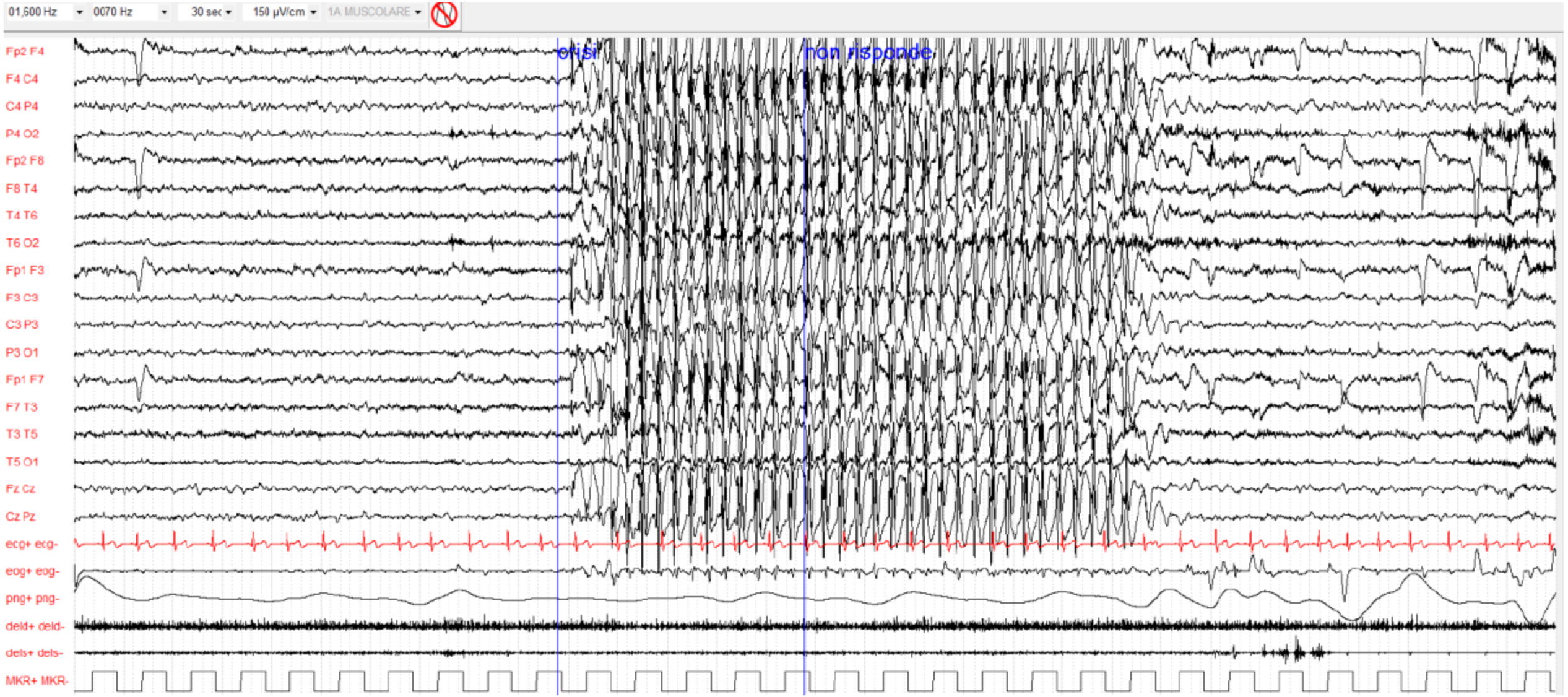
INFANTS NEONATE
- SELF LIMITED (NON) FAMILIAL EPILEPSY
- MYOCLONIC EPILEPSY OF INFANCY
- DEE
  - EARLY ONSET DEE
  - WEST SYNDROME
  - MIGRATING FOCAL SEIZURE

Childhood Absence Epilepsy

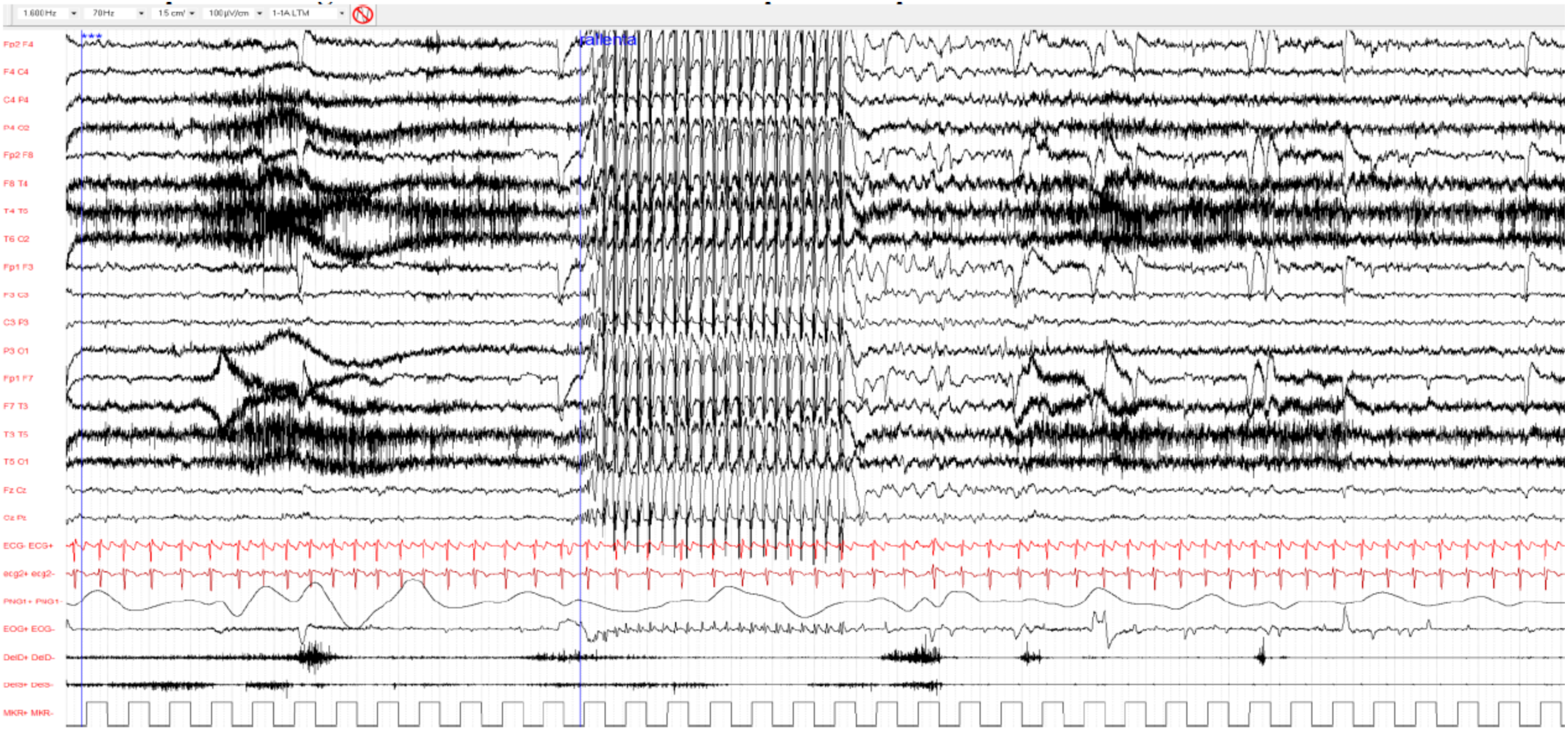




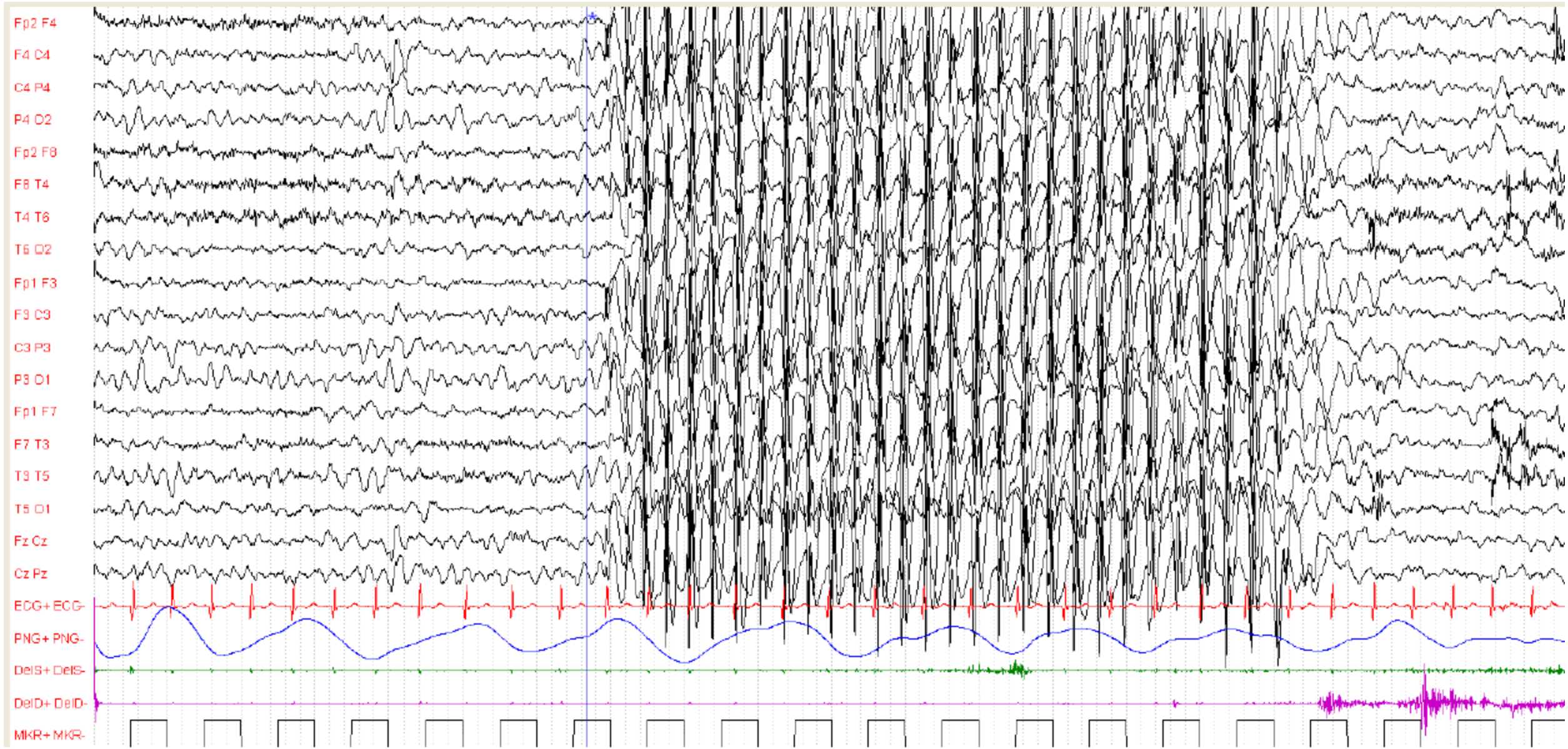
6 - assenza con arresto psicomotorio


01,600 Hz
0070 Hz
30 sec
150 µV/cm
1A MUSCOLARE
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ecg+ ecg-
eog+ eog-
png+ png-
deld+ deld-
dels+ dels-
MKR+ MKR-
crisi
non risponde

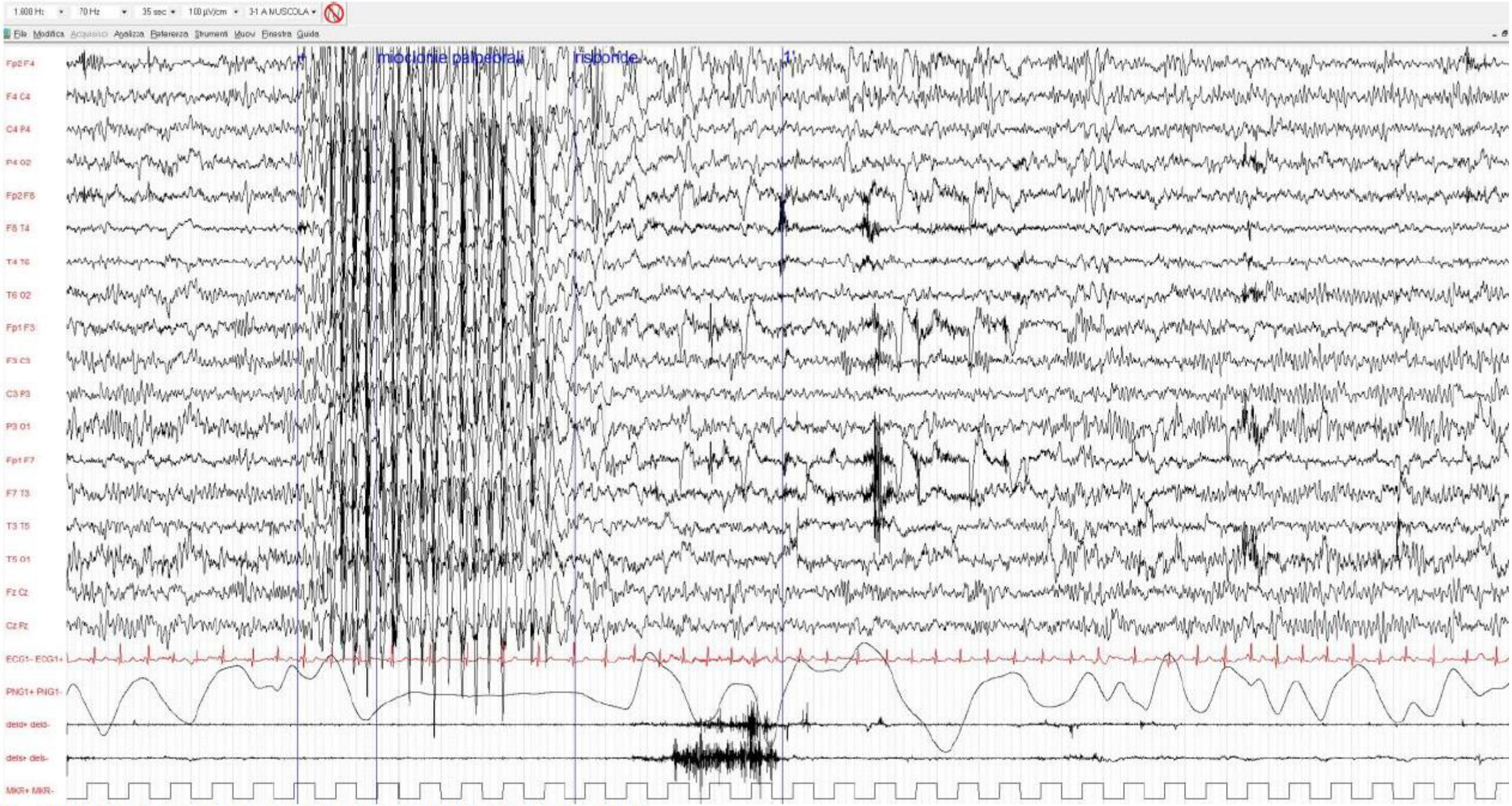
Crisi.



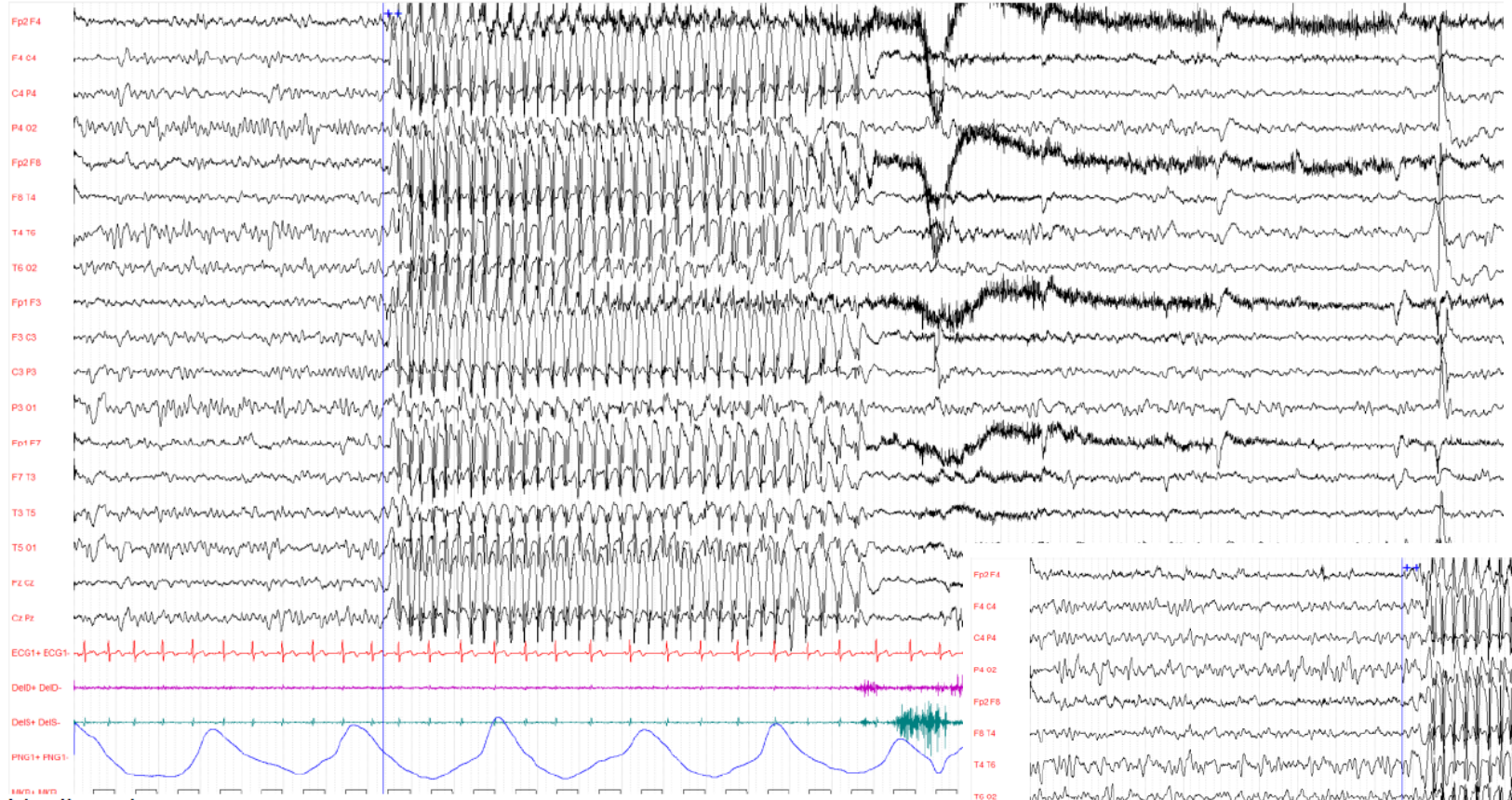
4 - ore 18:47 del 07.04 - sta parlando e rallenta


Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
PNG+ PNG-
DelS+ DelS-
DelD+ DelD-
MKR+ MKR-

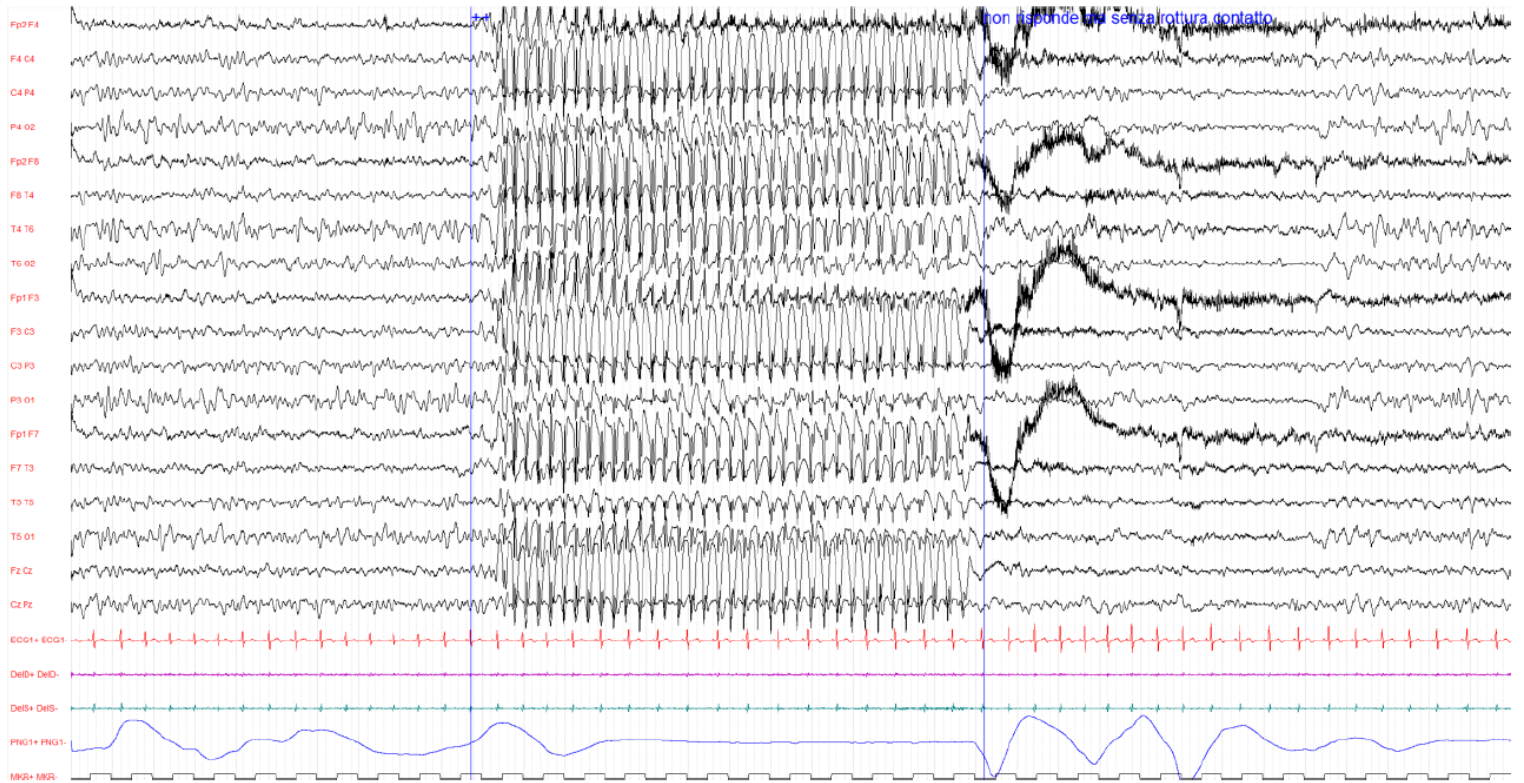
Assenza.



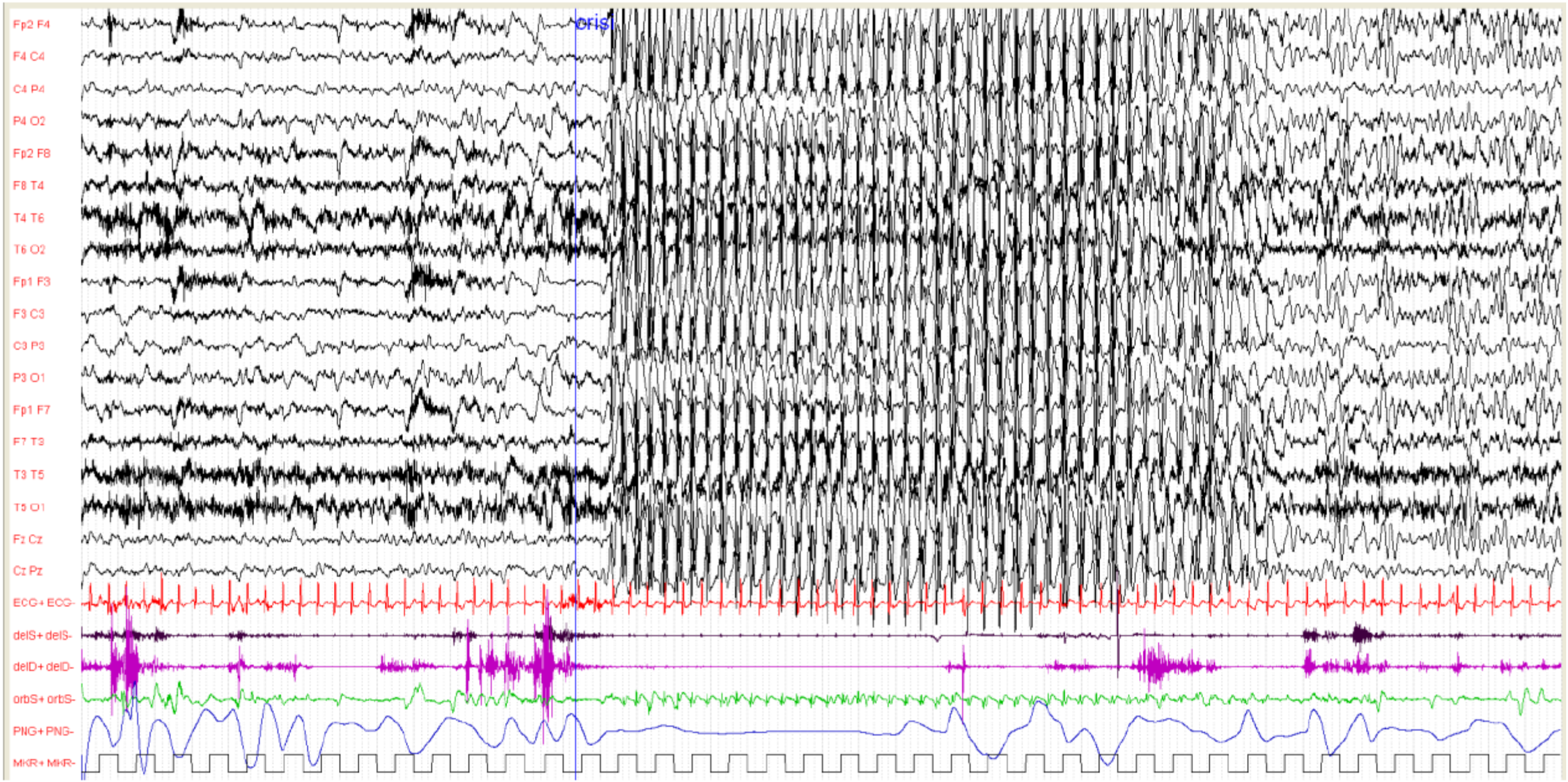
2 - durante Iperpnea assenza con mioclonie palpebrali



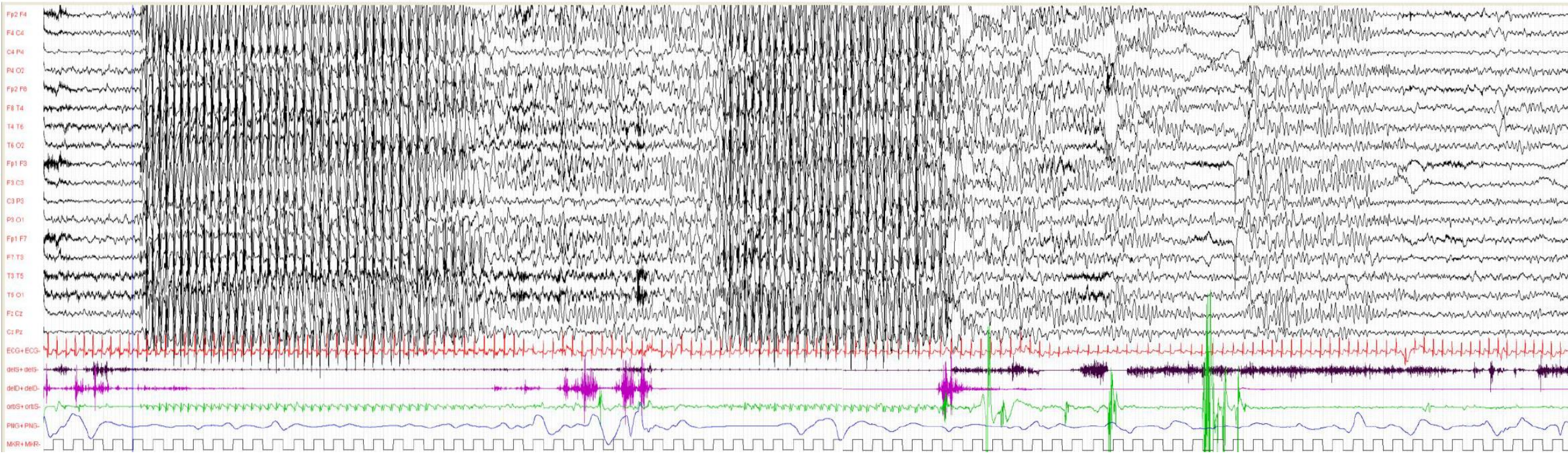
Veglia a riposo



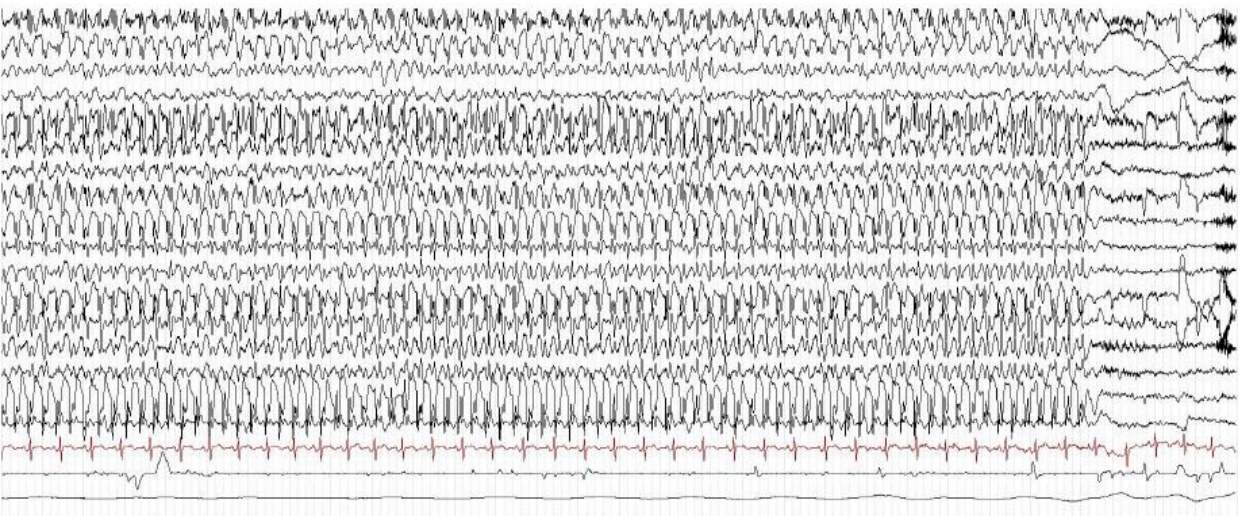
Analogo episodio di rottura di contatto durante iperpnea, si noti la pausa respiratoria nella precedente crisi non presente

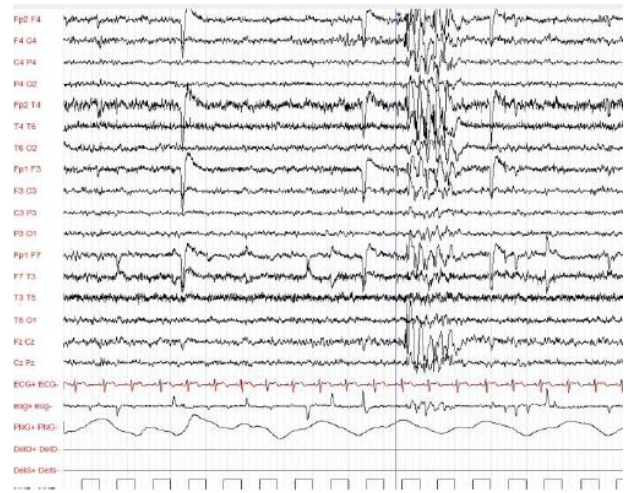


Crisi inizia e finisce, segue attività lenta post-critica.

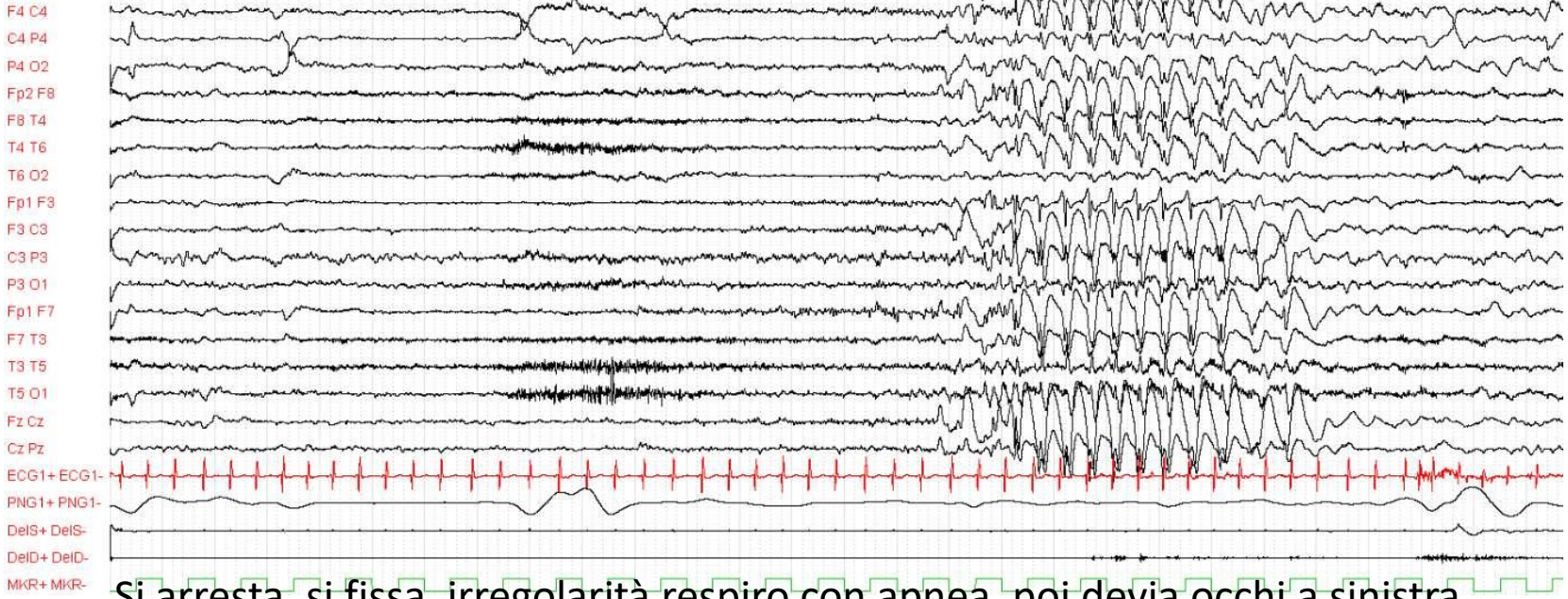

Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
delS+ delS-
delD+ delD-
orbS+ orbS-
PNG+ PNG-
MKR+ MKR-


Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
eog+ eog-
PNG+ PNG-
DelD+ DelD-

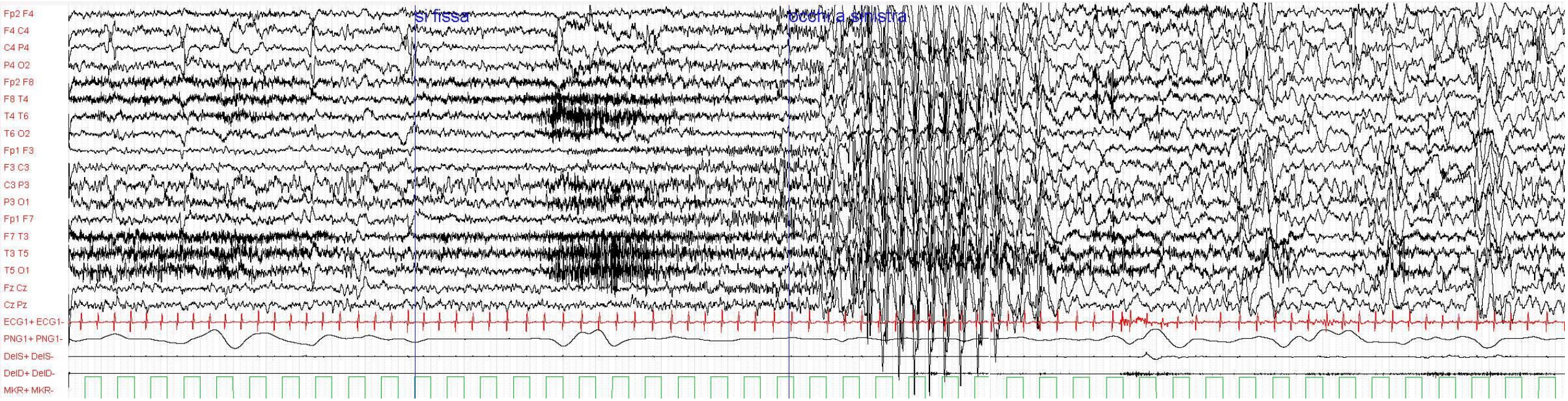



Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
F3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
eog+ eog-
PNG+ PNG-
DelD+ DelD-
DelS+ DelS-

ASSENZA FOCALE

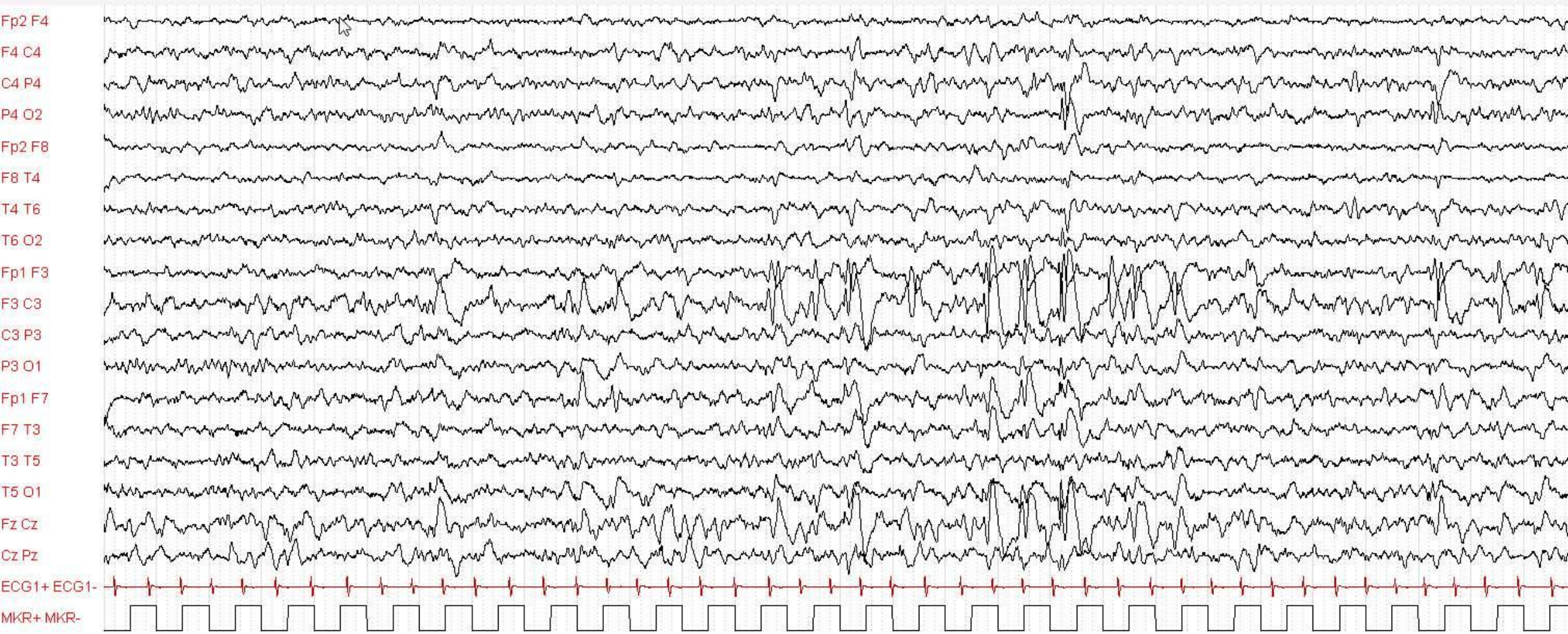


Si arresta, si fissa, irregolarità respiro con apnea, poi devia occhi a sinistra
Scarica rapida focale parietale sinistra e poi parossismi diffusi di punta-onda

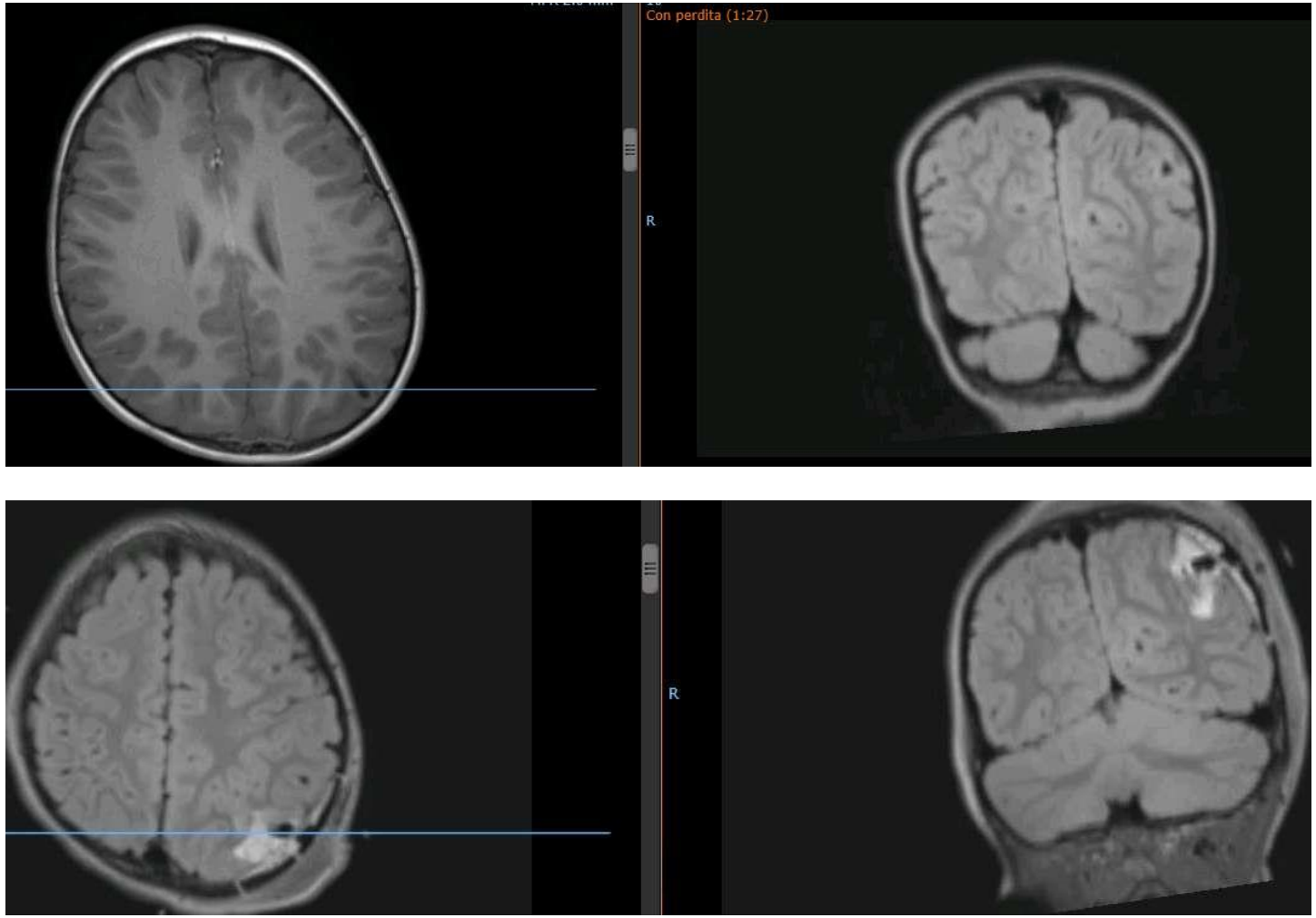


ASSENZA FOCALE

CF 1 EEG, sonno



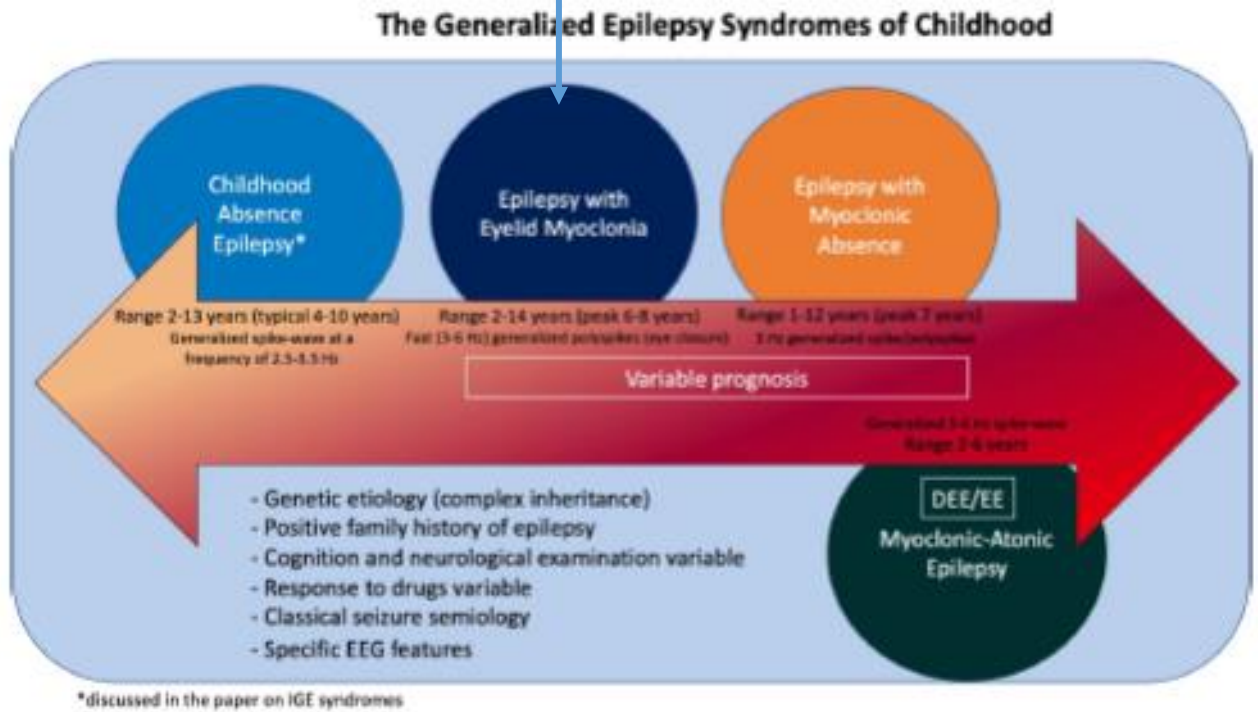
ASSENZA FOCALE



DIAGNOSI

Tumore glioneuronale di basso grado con caratteristiche morfologiche ed immunofenotipiche coerenti con tumore glioneuronale di basso grado polimorfo dei giovani (PLNTY).

Epilepsy with Eyelid Myoclonia

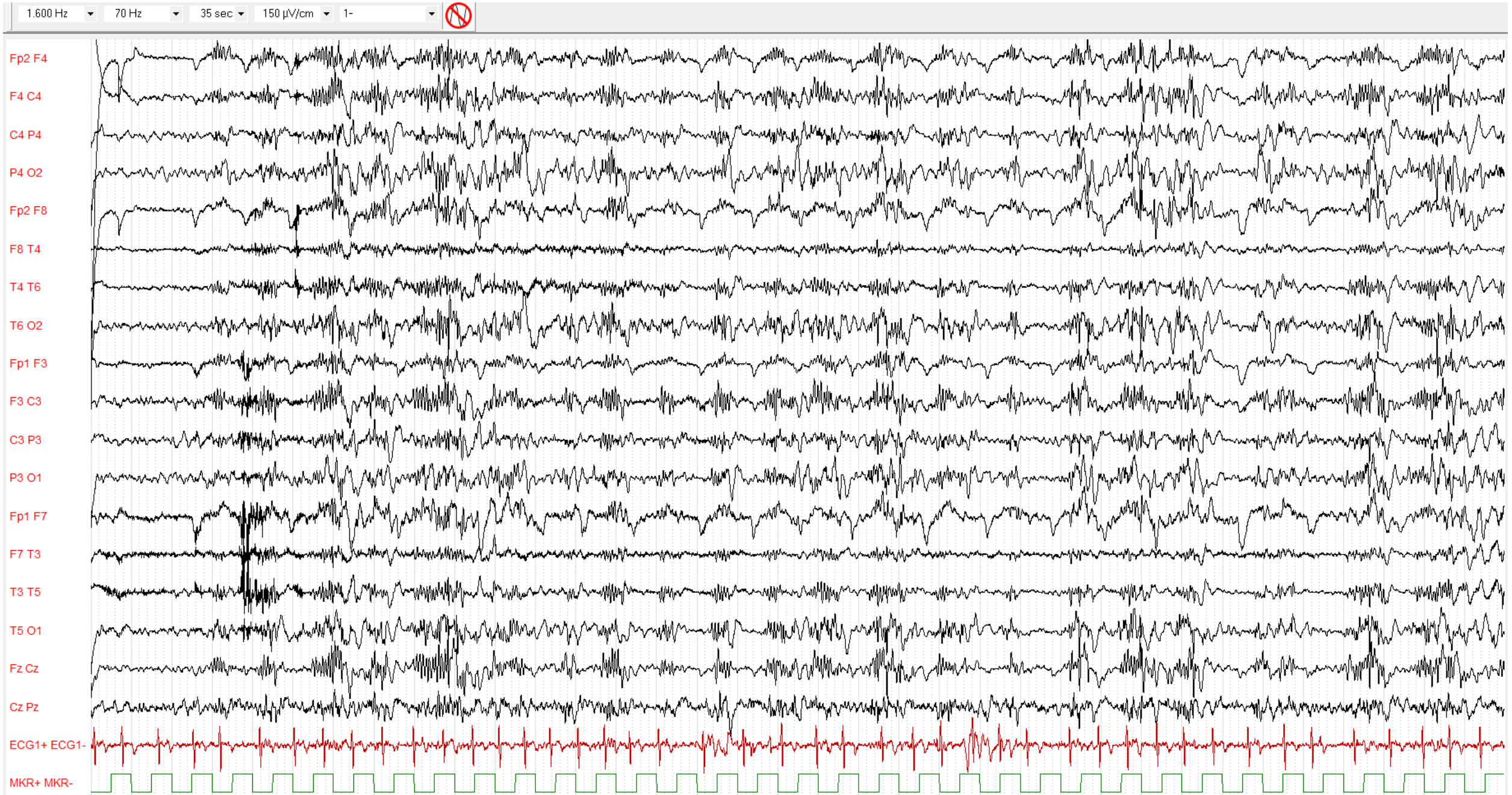


Epilepsy with Eyelid Myoclonia

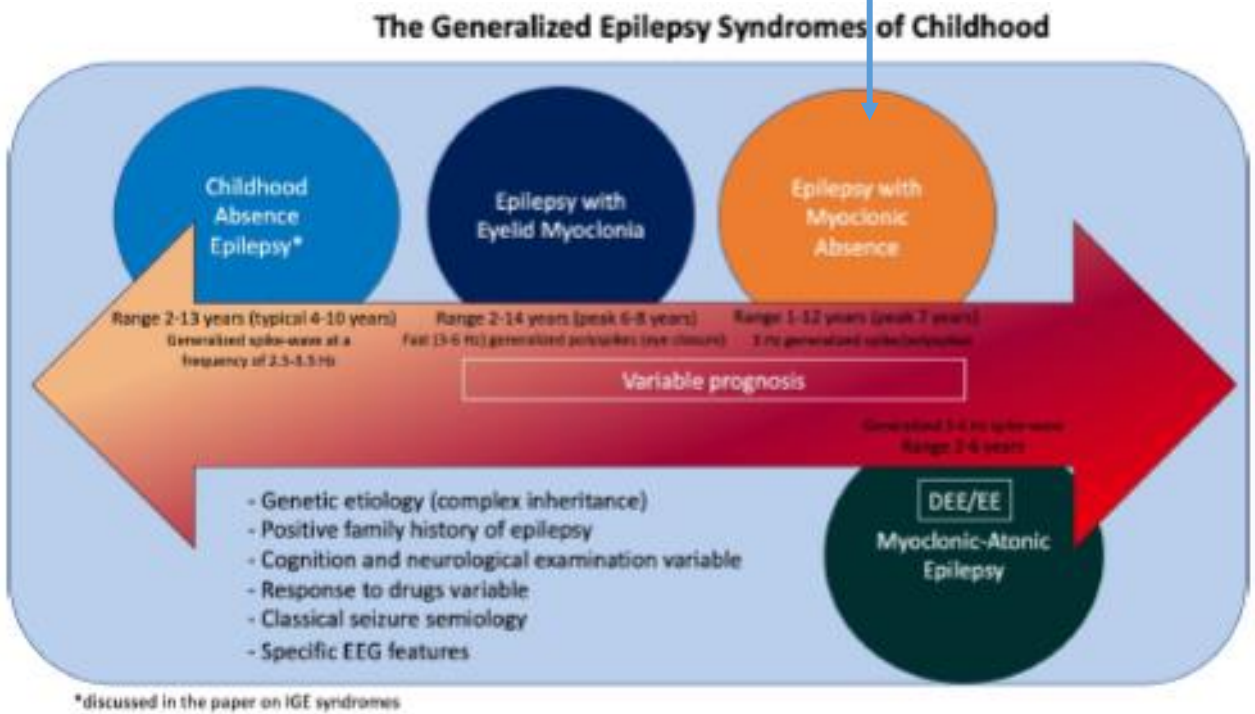
Epilepsy with Eyelid Myoclonia

Fer Cri 12 anni

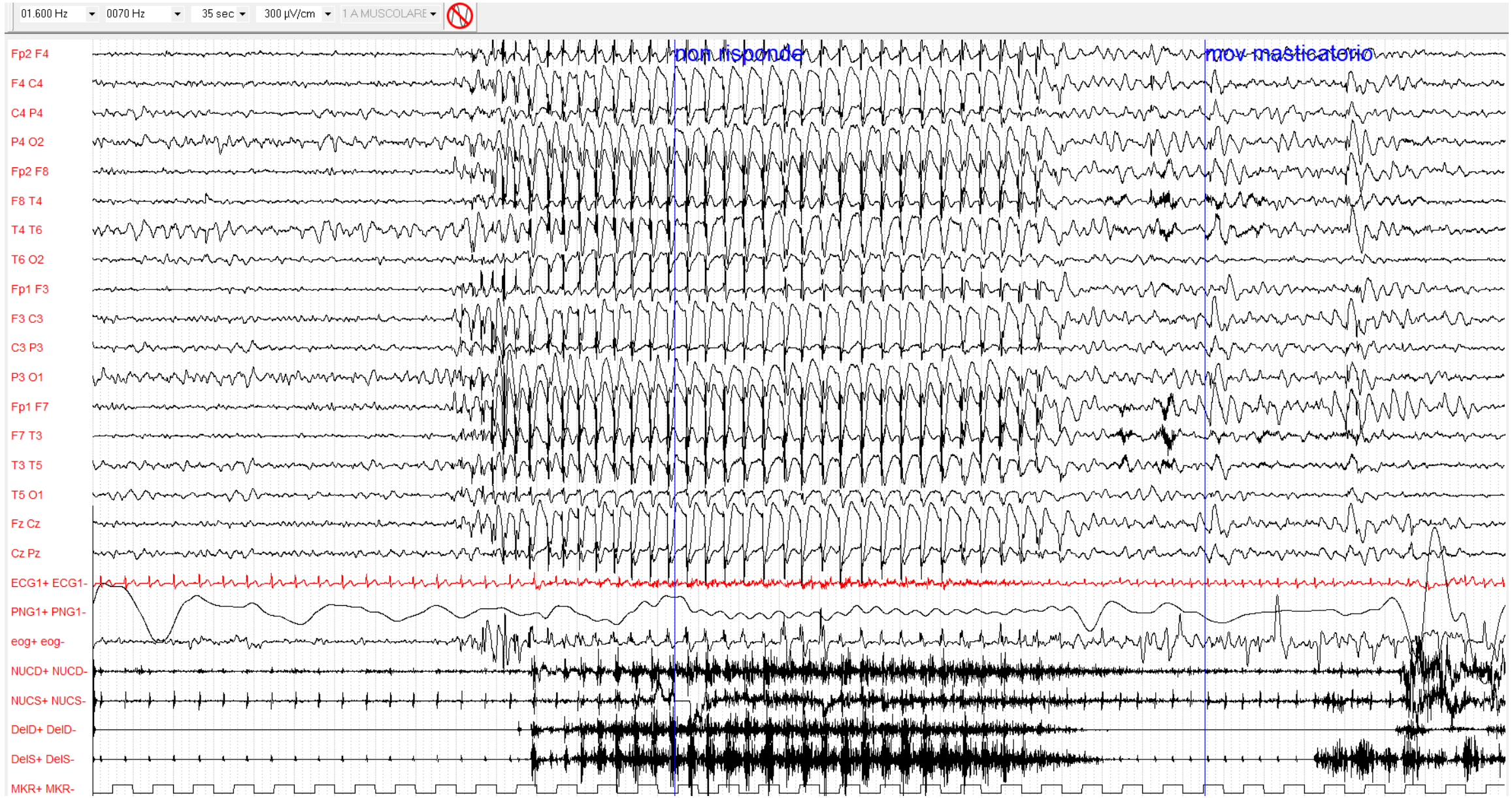
stato di eyelid myoclonia

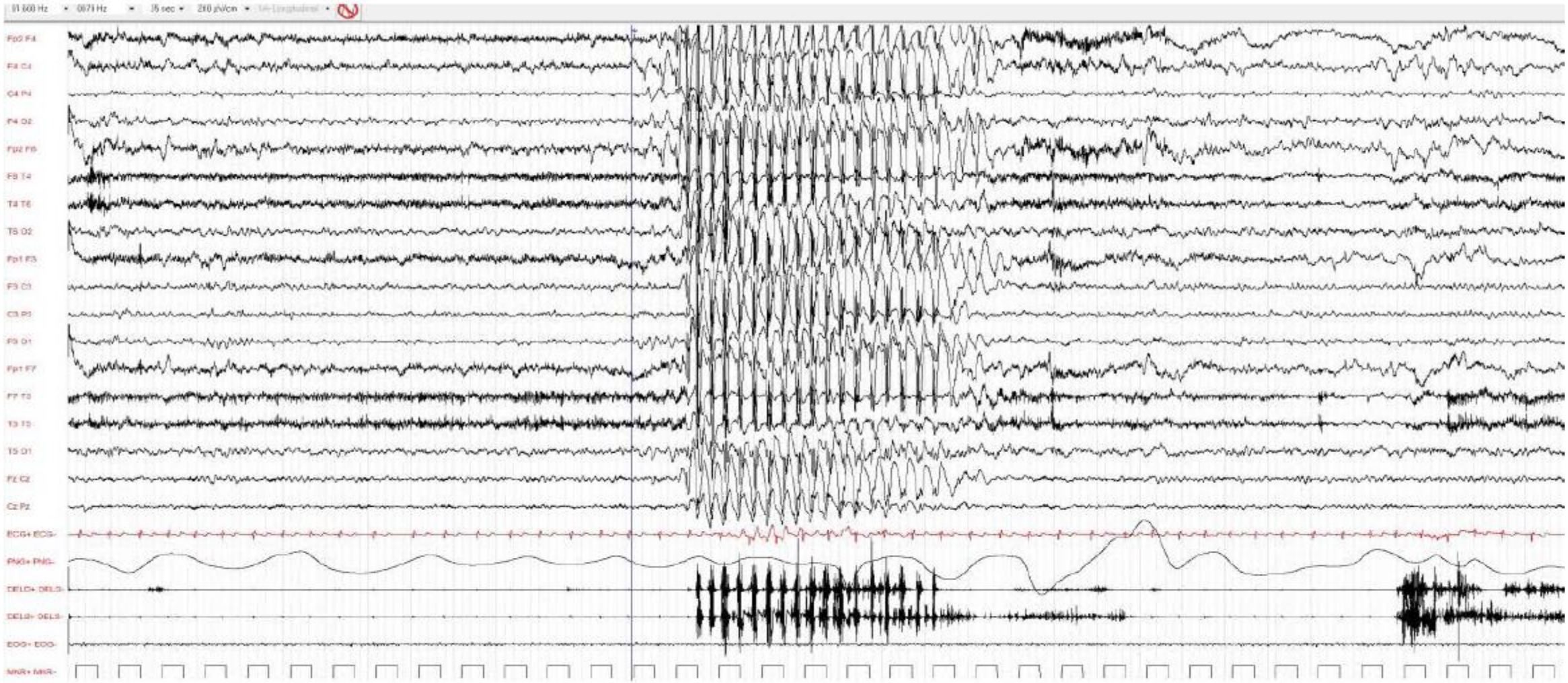


Epilepsy with Myoclonic Absence

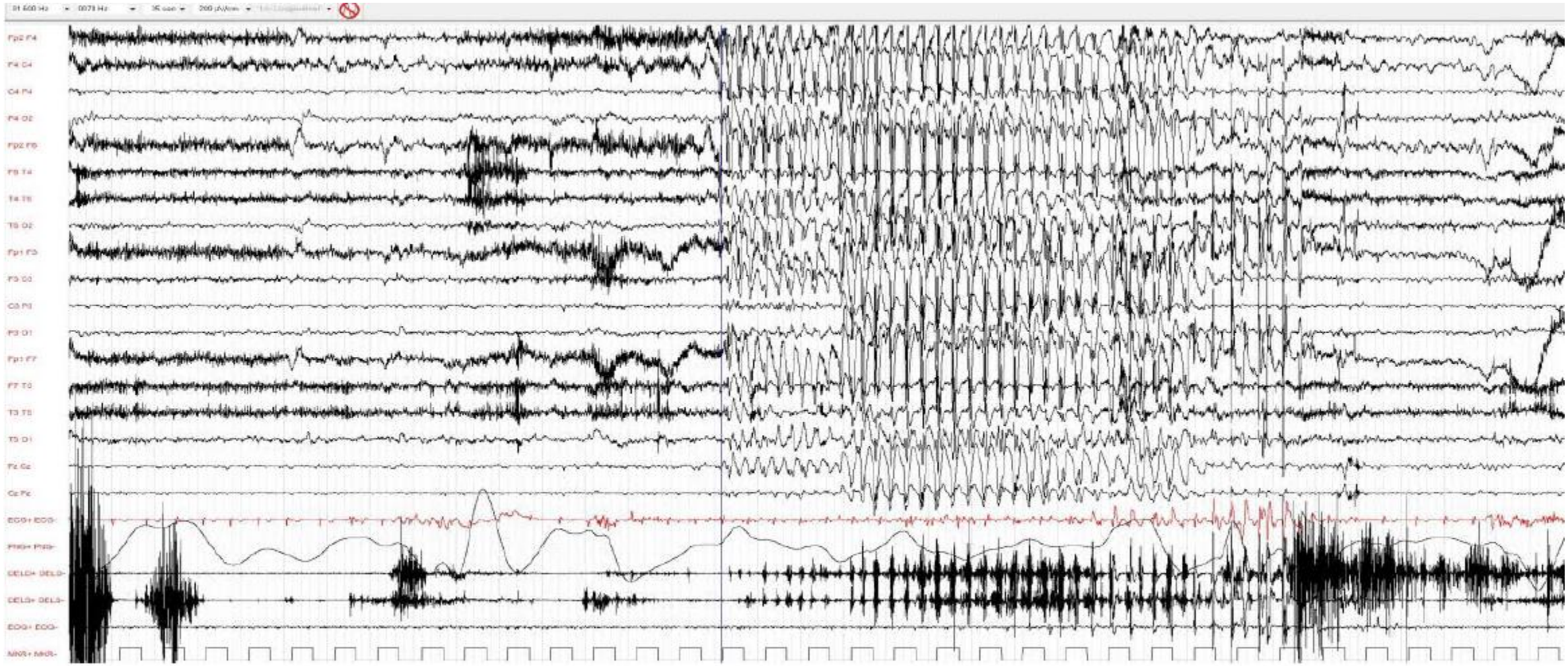


Pol Ann 8aa Assenza mioclonica



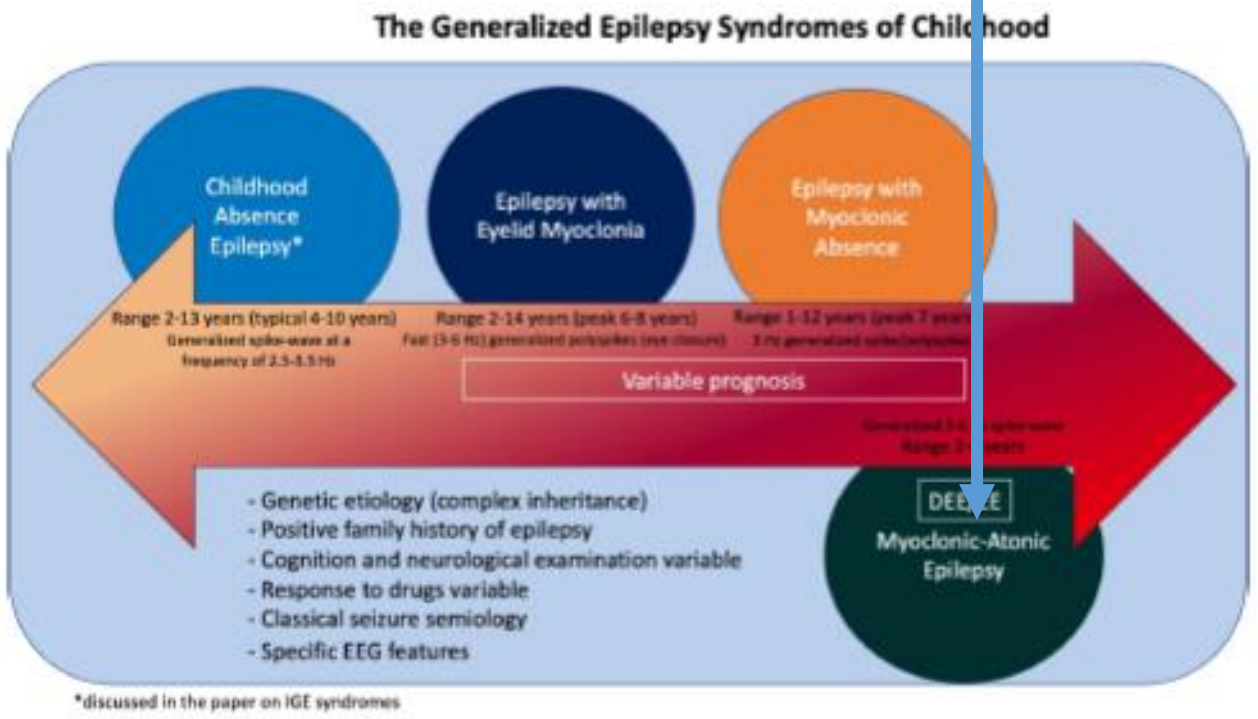


7 - crisi di assenza mioclonica

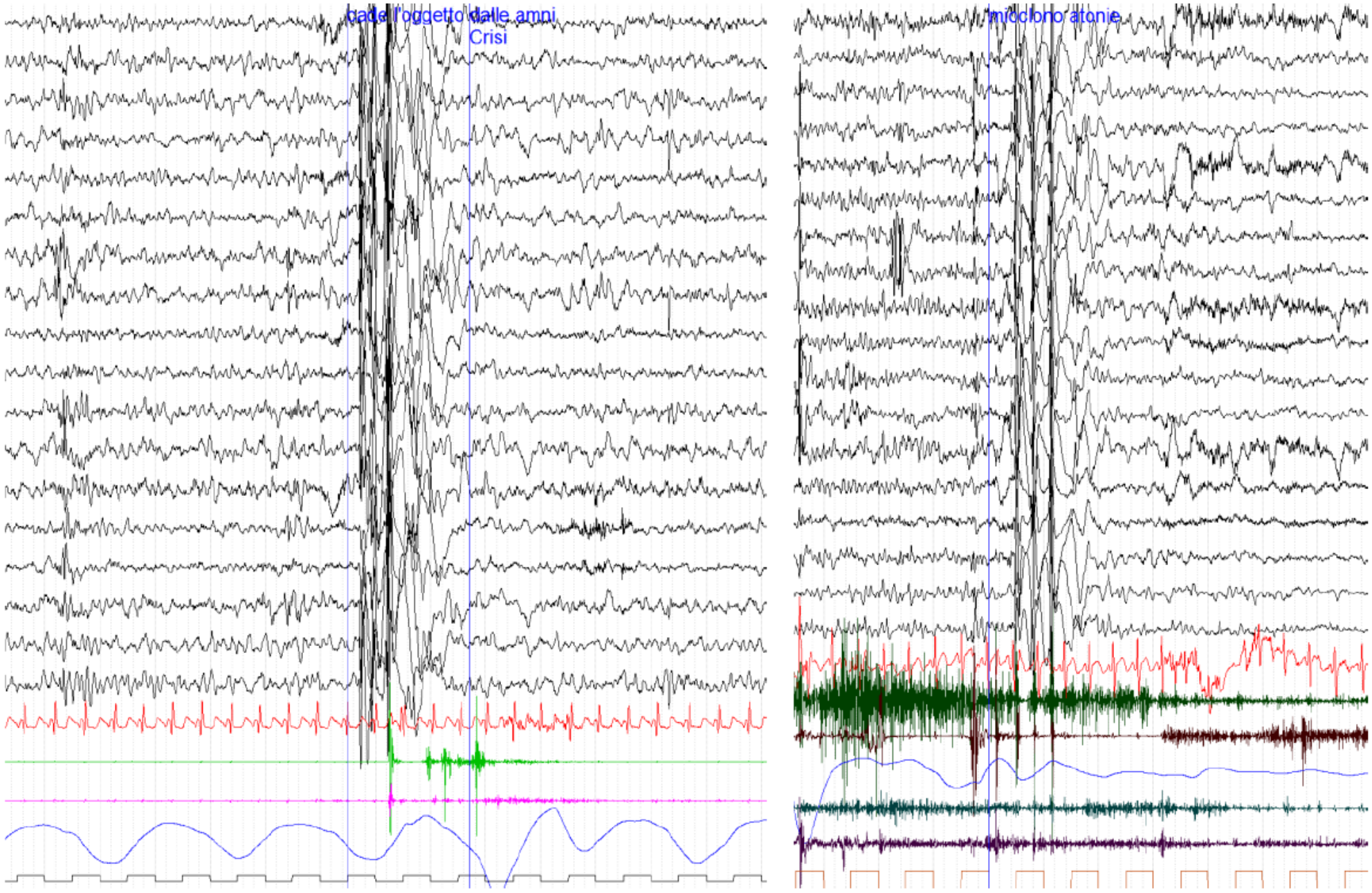


8 - assenza mioclonica in due fasi

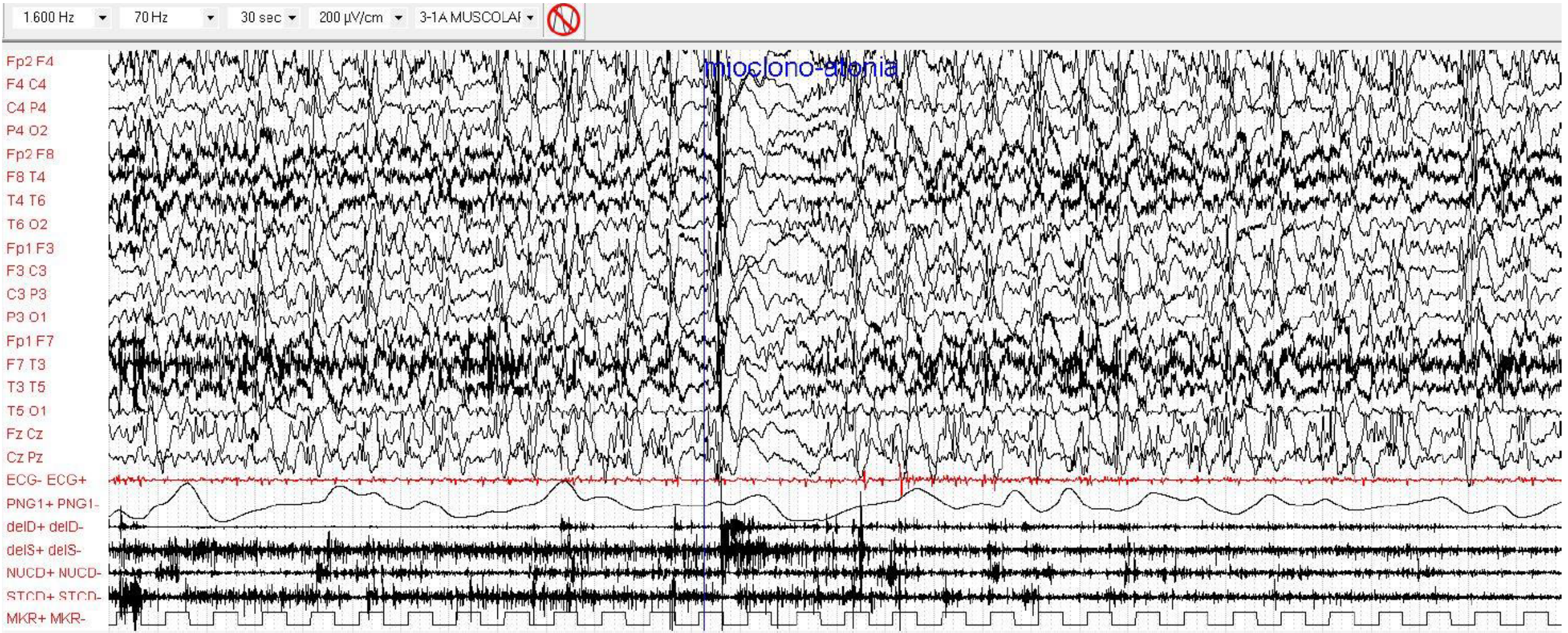
Myoclonic-Atonic Epilepsy
Previoulsy Myoclonic-Astatic Epilepsy



LaM Glo 3aa5m Mioclono-Atonia

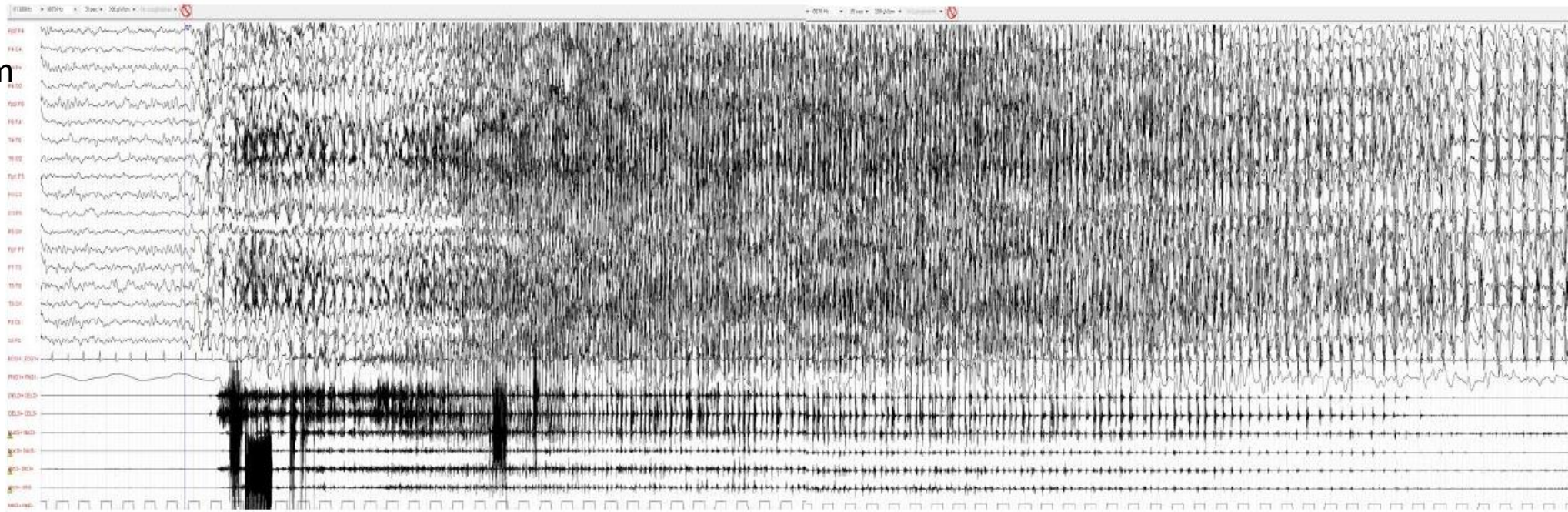


Dif Gio Mioclono atonia in MAE



durante un parossismo critico episodio di mioclono-atonia (visibile sui muscoli posteriori del collo)

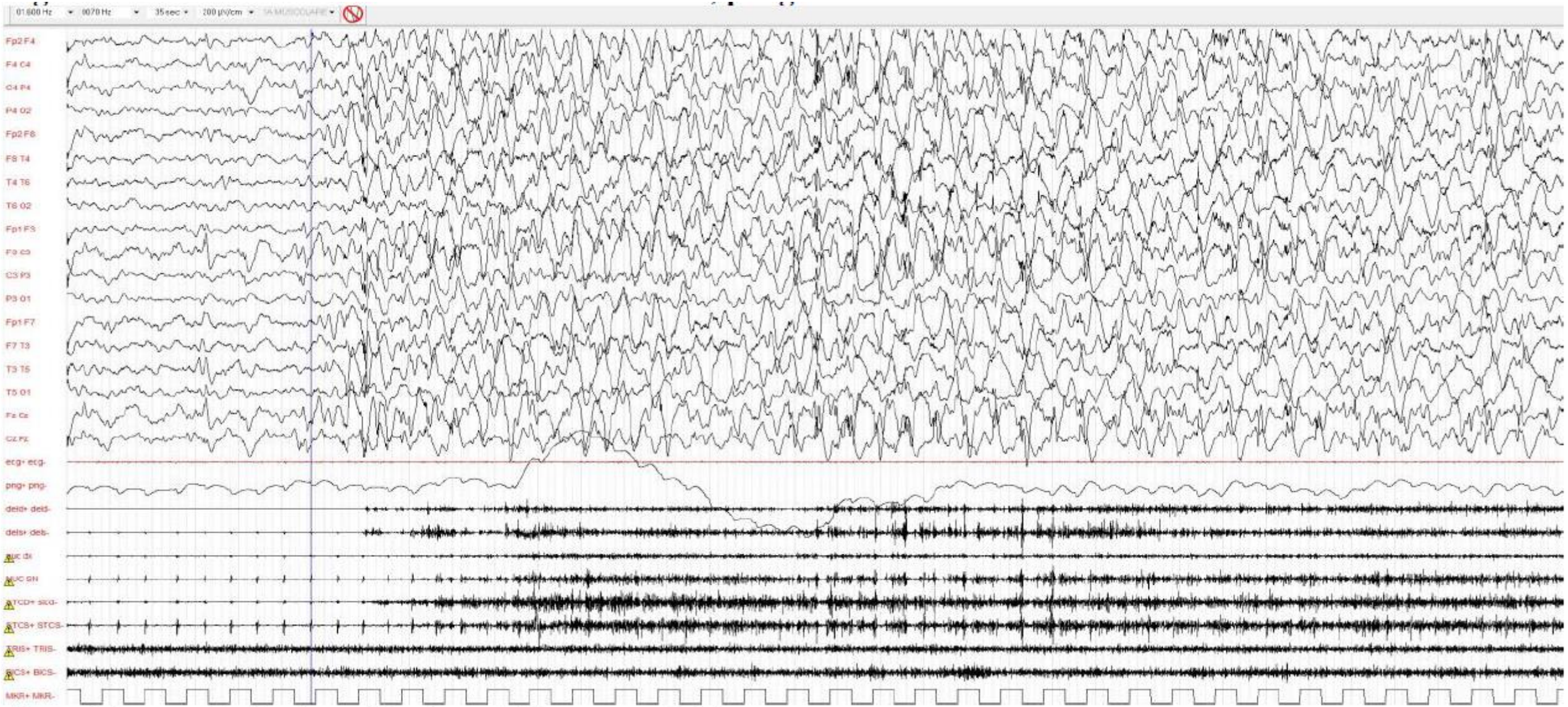
DF.G. Epilessia con crisi mioclono-atoniche 3aa8m
Presenza anche di crisi generalizzate motorie
Mioclono-tonico-cloniche



13 - ore 4.14 notte del 25.2

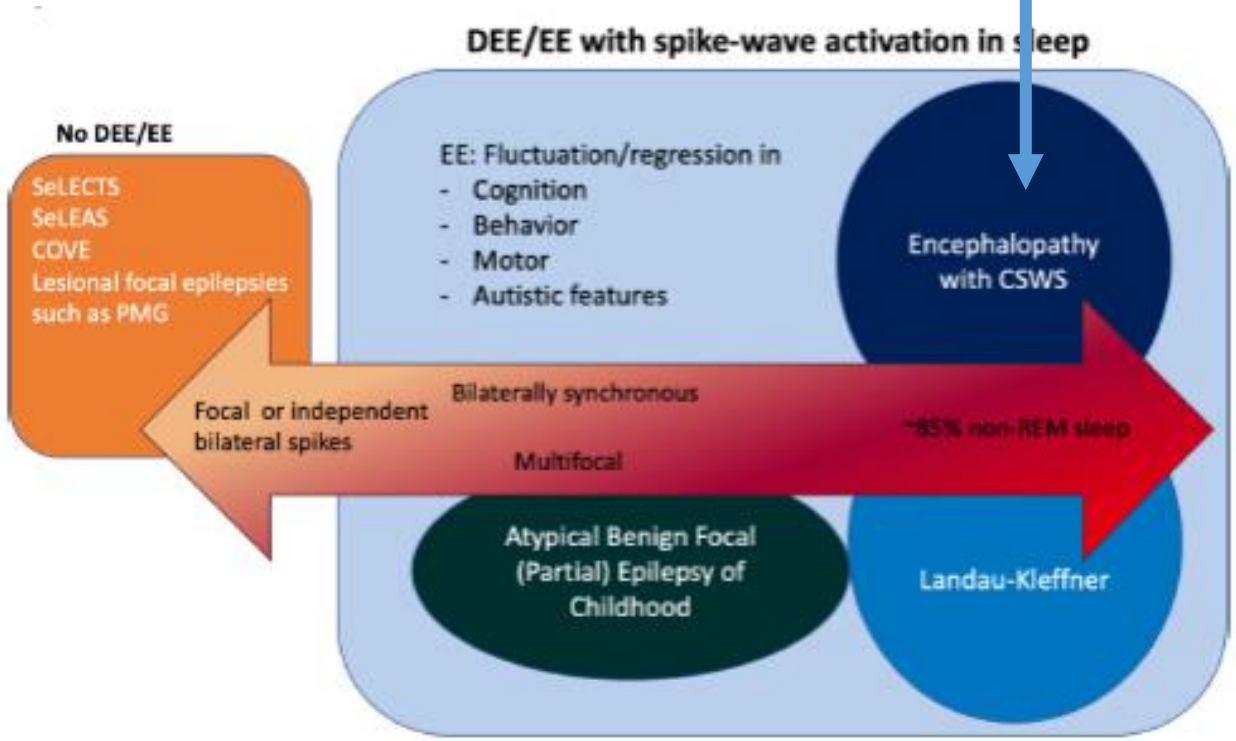
14 - continua crisi convulsiva con PGES Postictal Generalized EEG Suppression della durata di circa 30 secondi associata a gasping respiratorio e recupero con EEG discontinuo

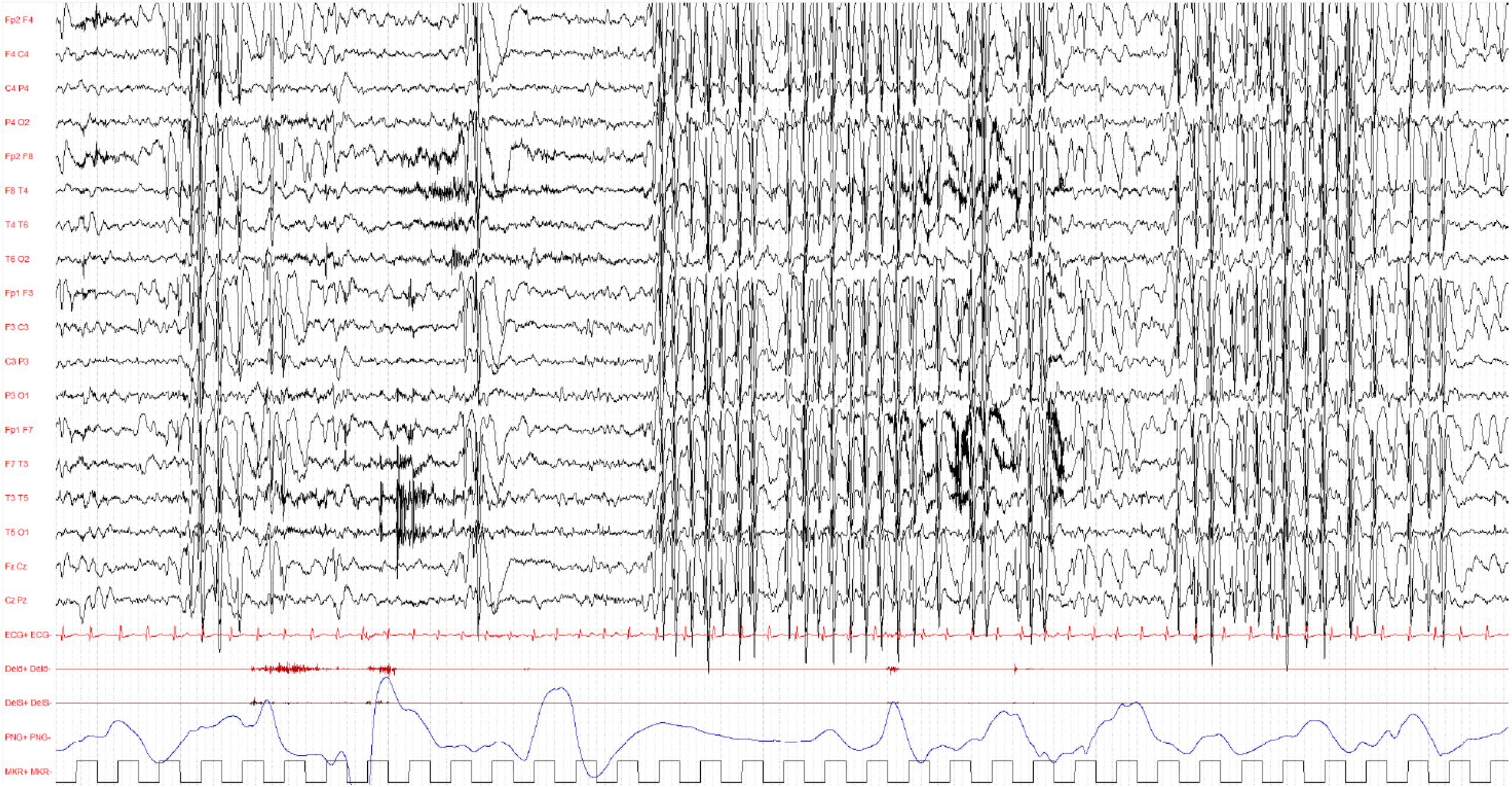
DF.G. Epilessia con crisi mioclono-atoniche 3aa8m - stato mioclonico



12 - crisi con mioclonie e riduzione della responsività

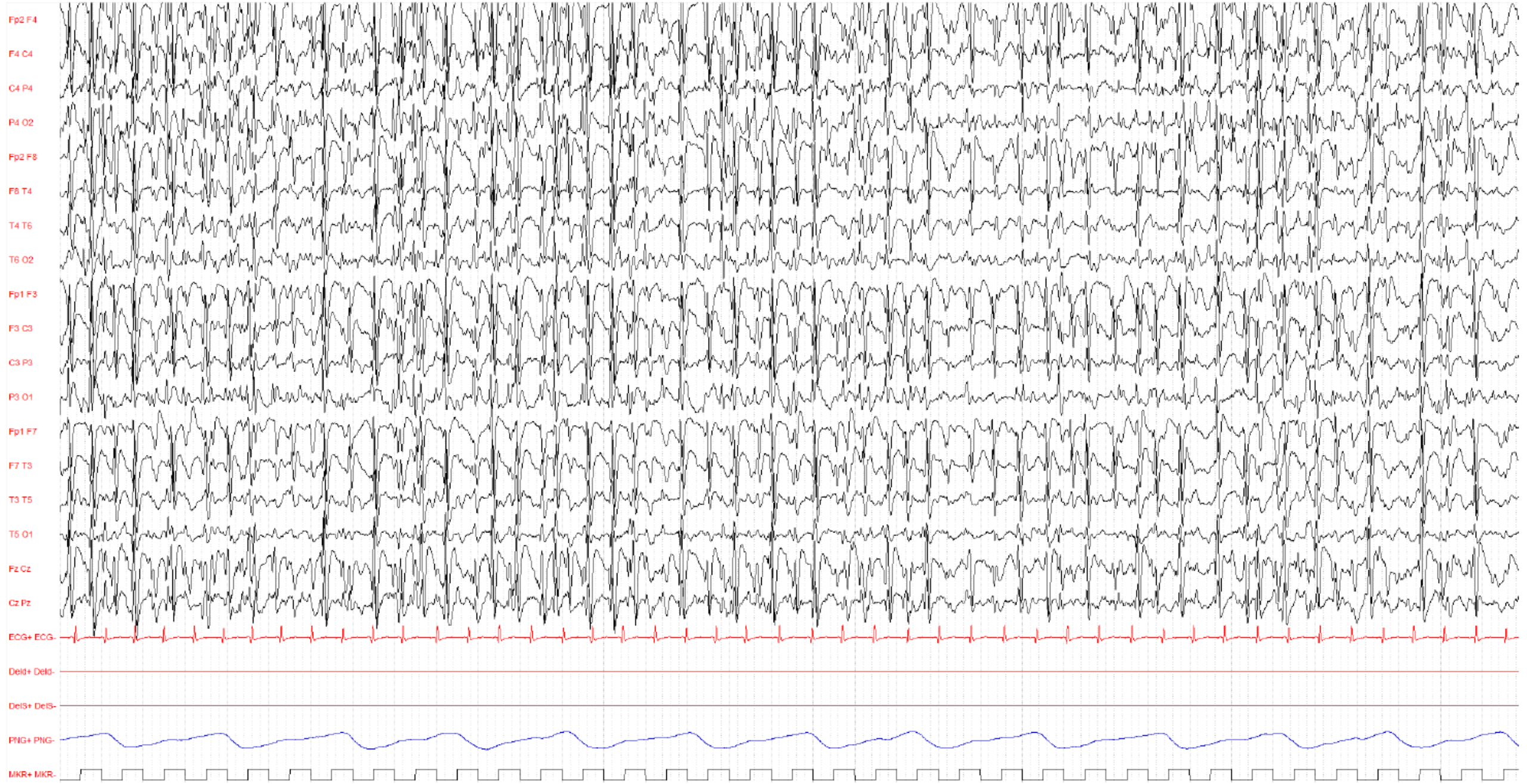
Encephalopathy with Continuous Spike-and-Wave during Sleep (CSWS)





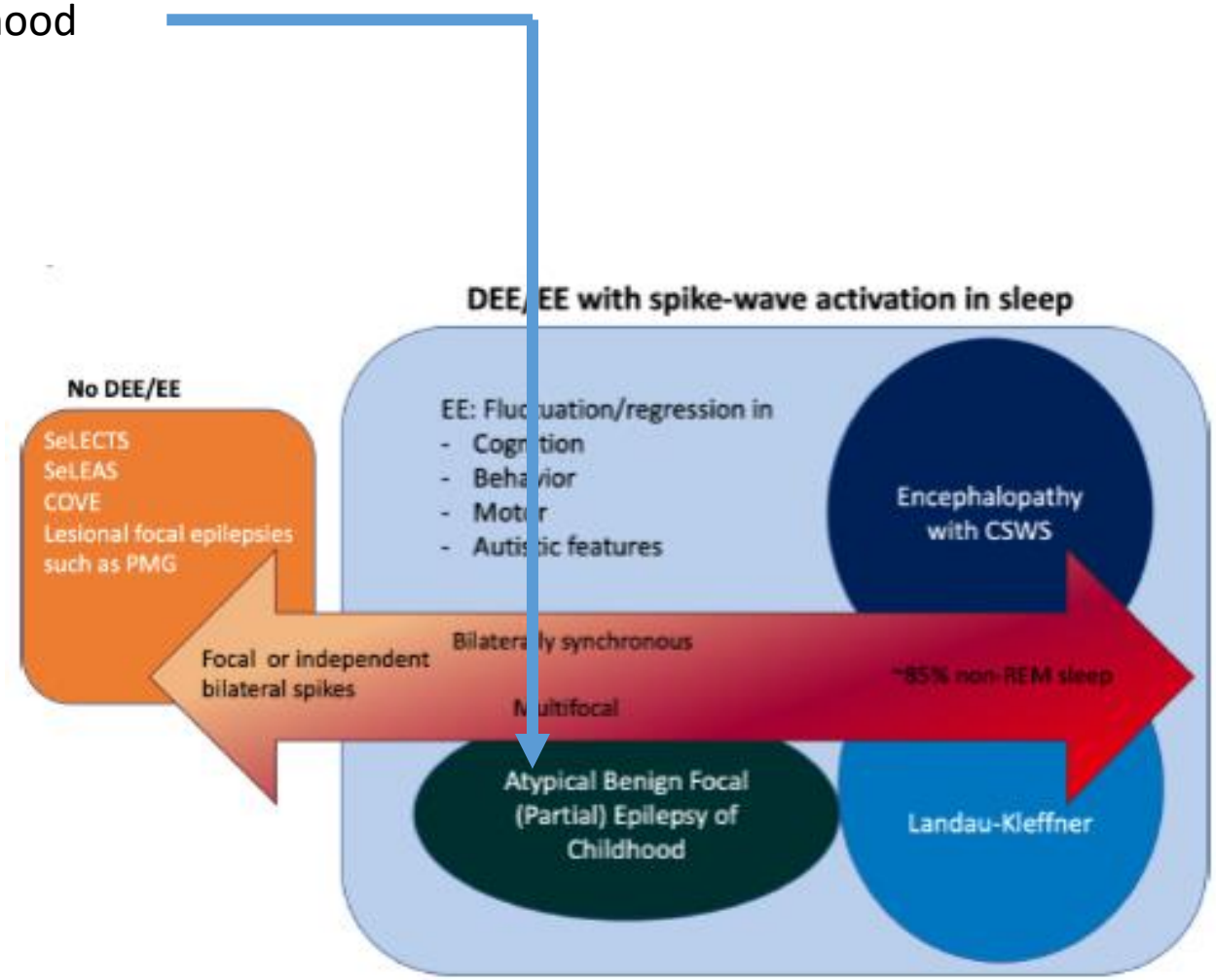
3 - ore 11.58 supina vede la TV apparentemente non segni clinici

5 - ore 23.08 a dieci minuti dall'addormentamento

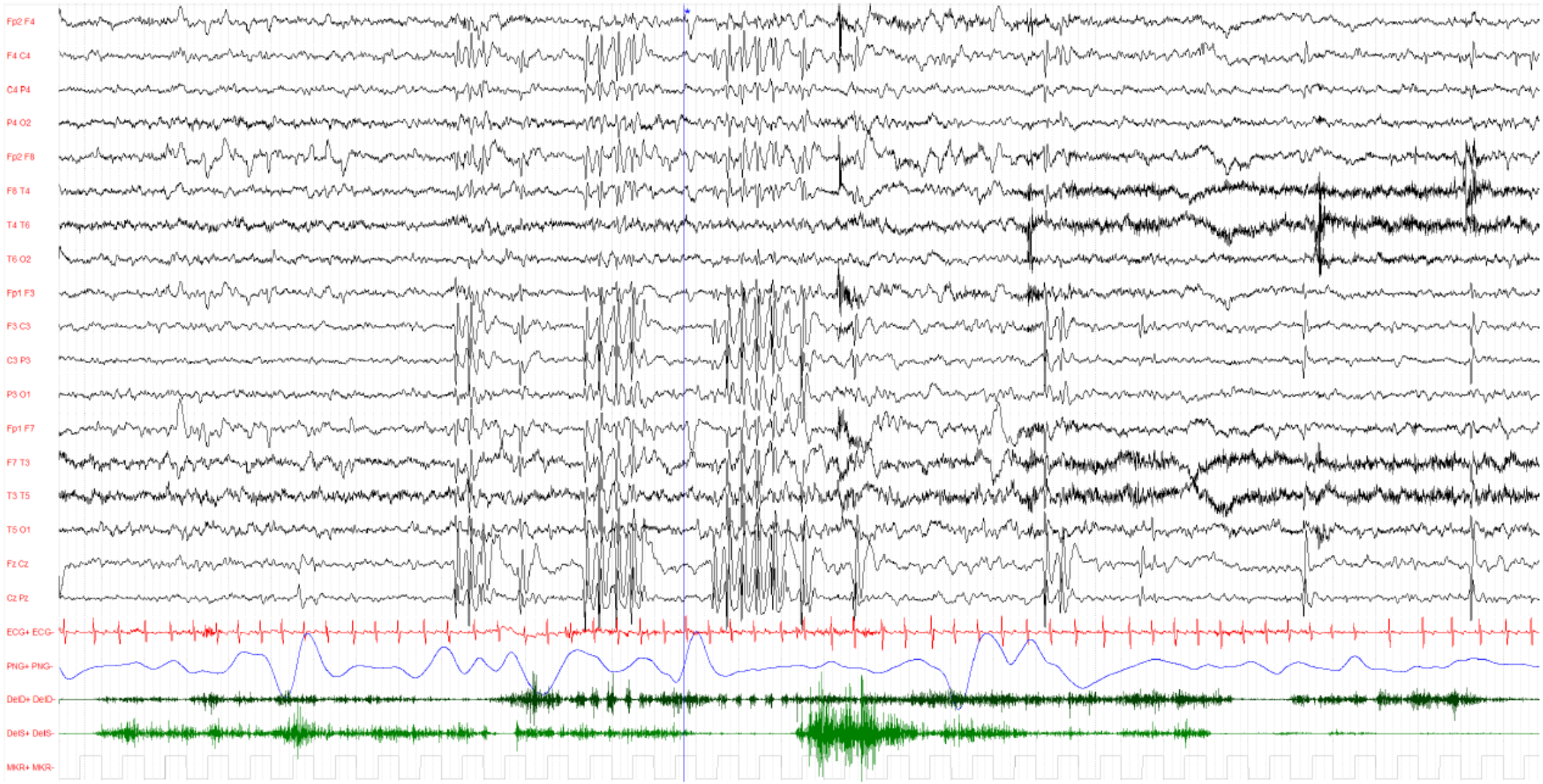


6 - ore 12.57 a 50 minuti dall'addormentamento

Atypical Benign Focal Epilepsy of chidlhood
Negative Myoclonus, CSWS

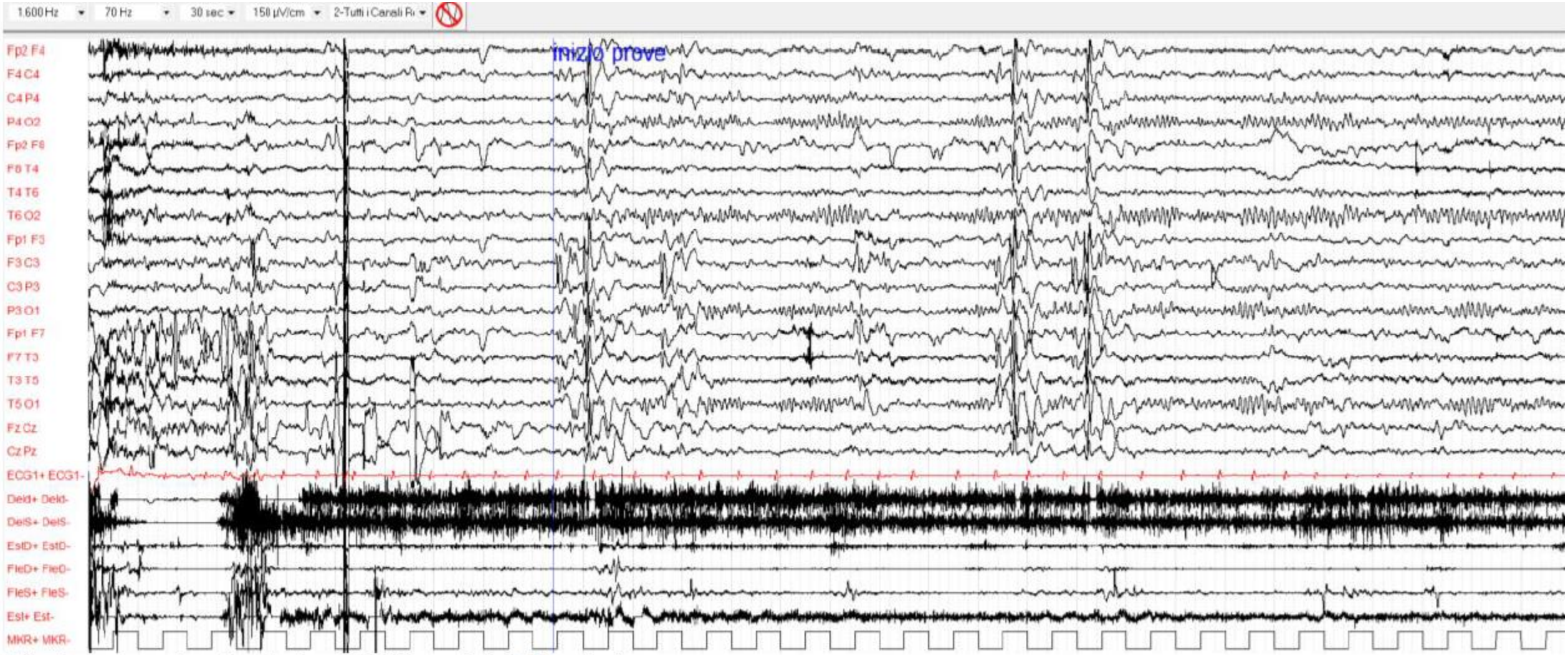


Sor Mar 5aa10m distrubid ell’equilibrio a rencente insorgenza Mioclono negativo



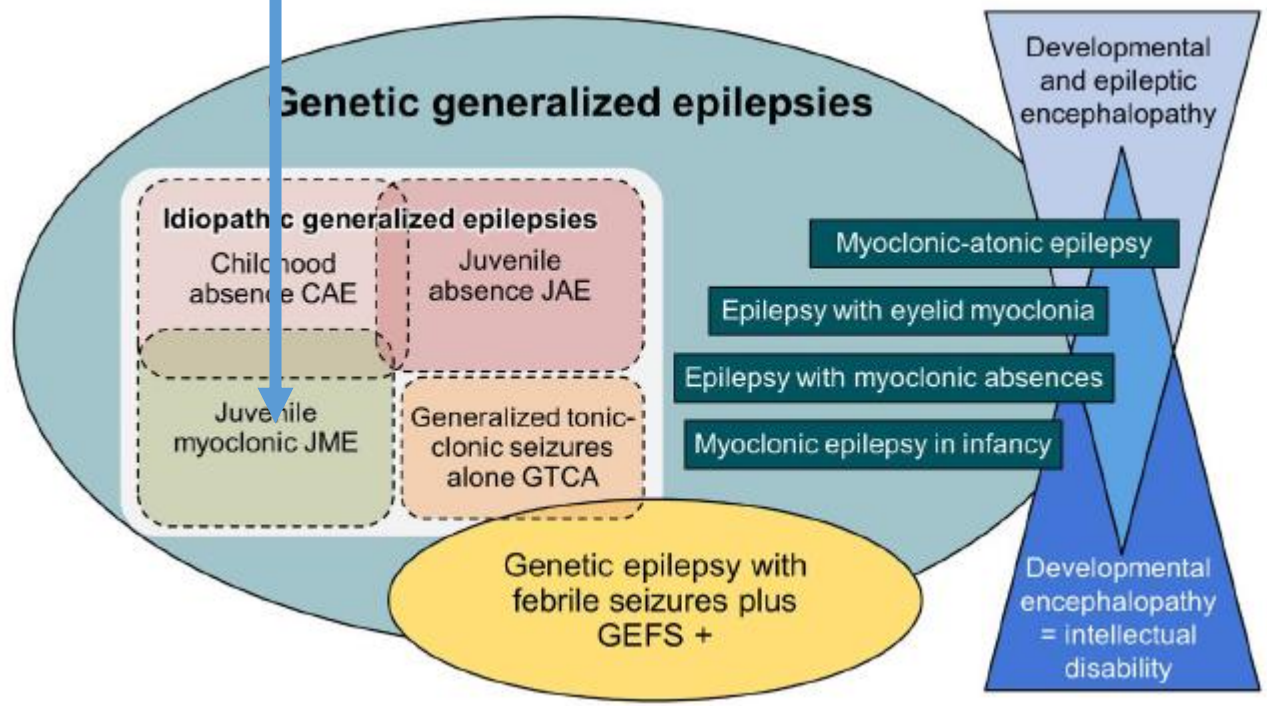
6 - mioclono negativo arto superiore destro e tronco

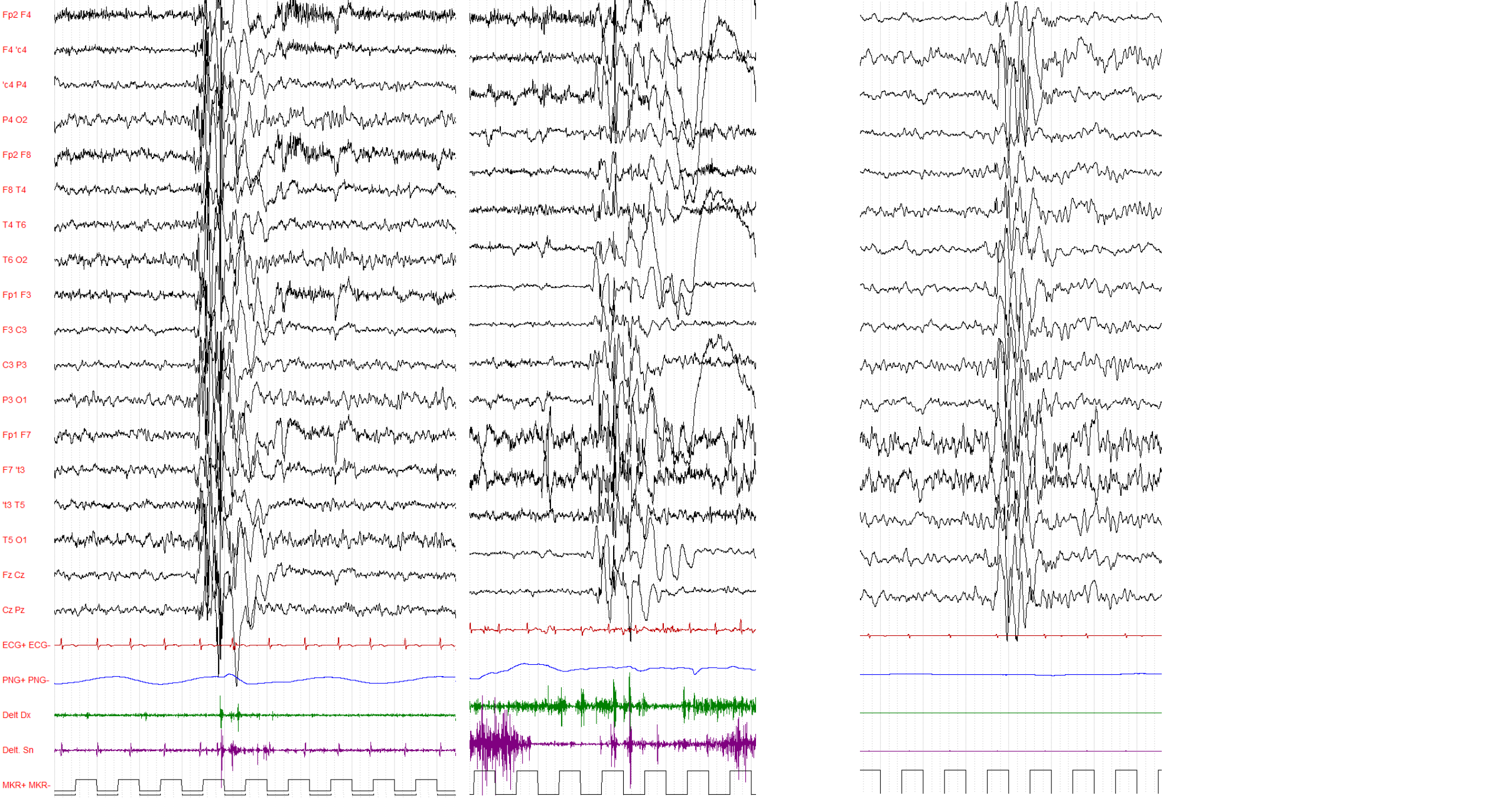
Val Sar 7aa5m Epilessia Rolandica, da qualche mese ipostenia AS dx



Mioclono negativo focale ben visibile sul deltoide destro.

Juvenile Myoclonic Epilepsy

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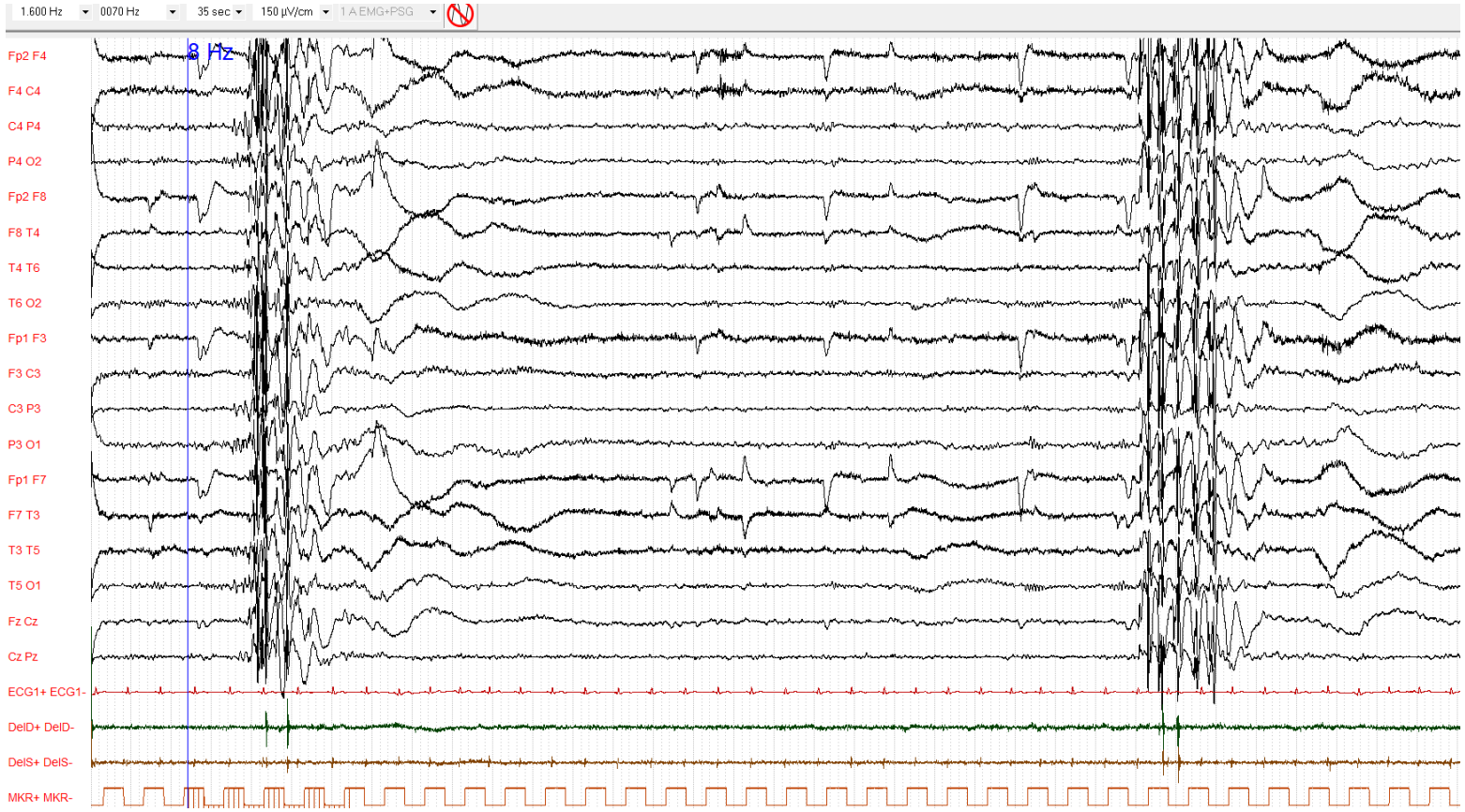
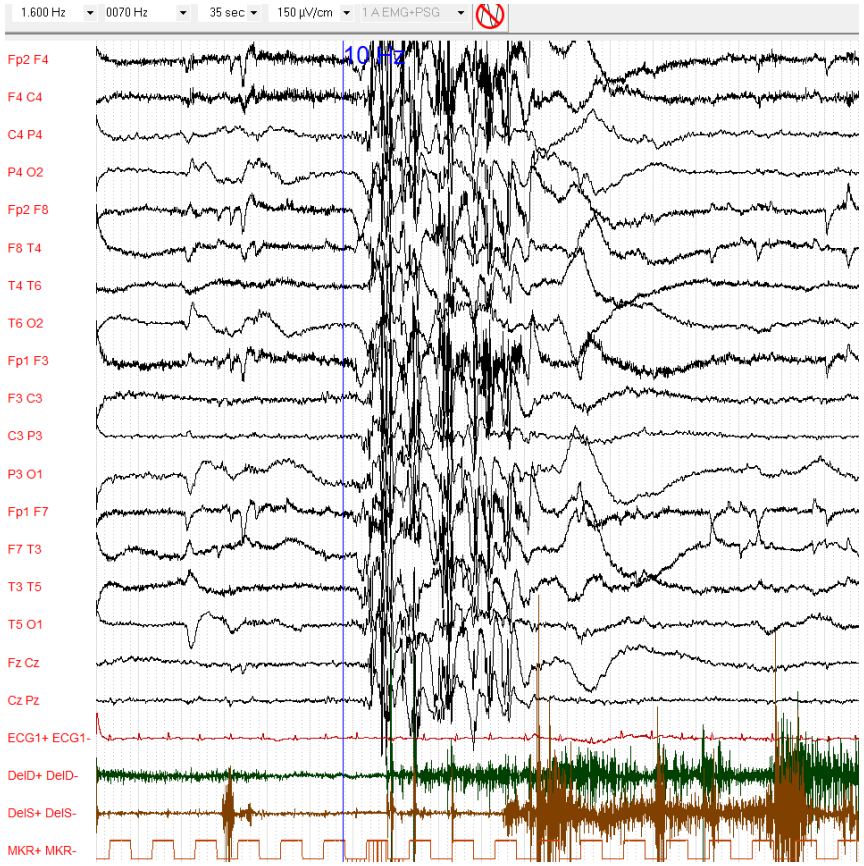
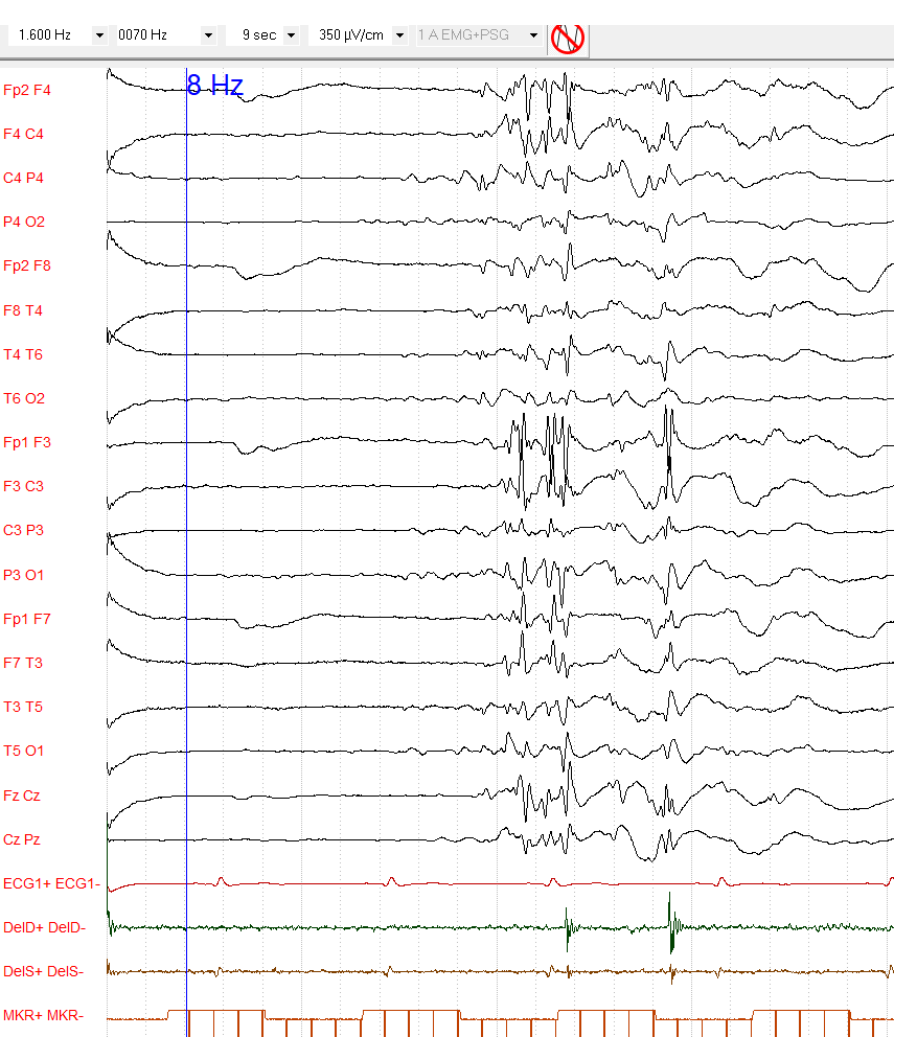
'c4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 't3
't3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
PNG+ PNG-
Delt Dx
Delt. Sn
MKR+ MKR-


1.600 Hz
0070 Hz
35 sec
150 µV/cm
1 A EMG+PSG
8 Hz
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-


1.600 Hz
0070 Hz
35 sec
150 µV/cm
1 A EMG+PSG
10 Hz
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-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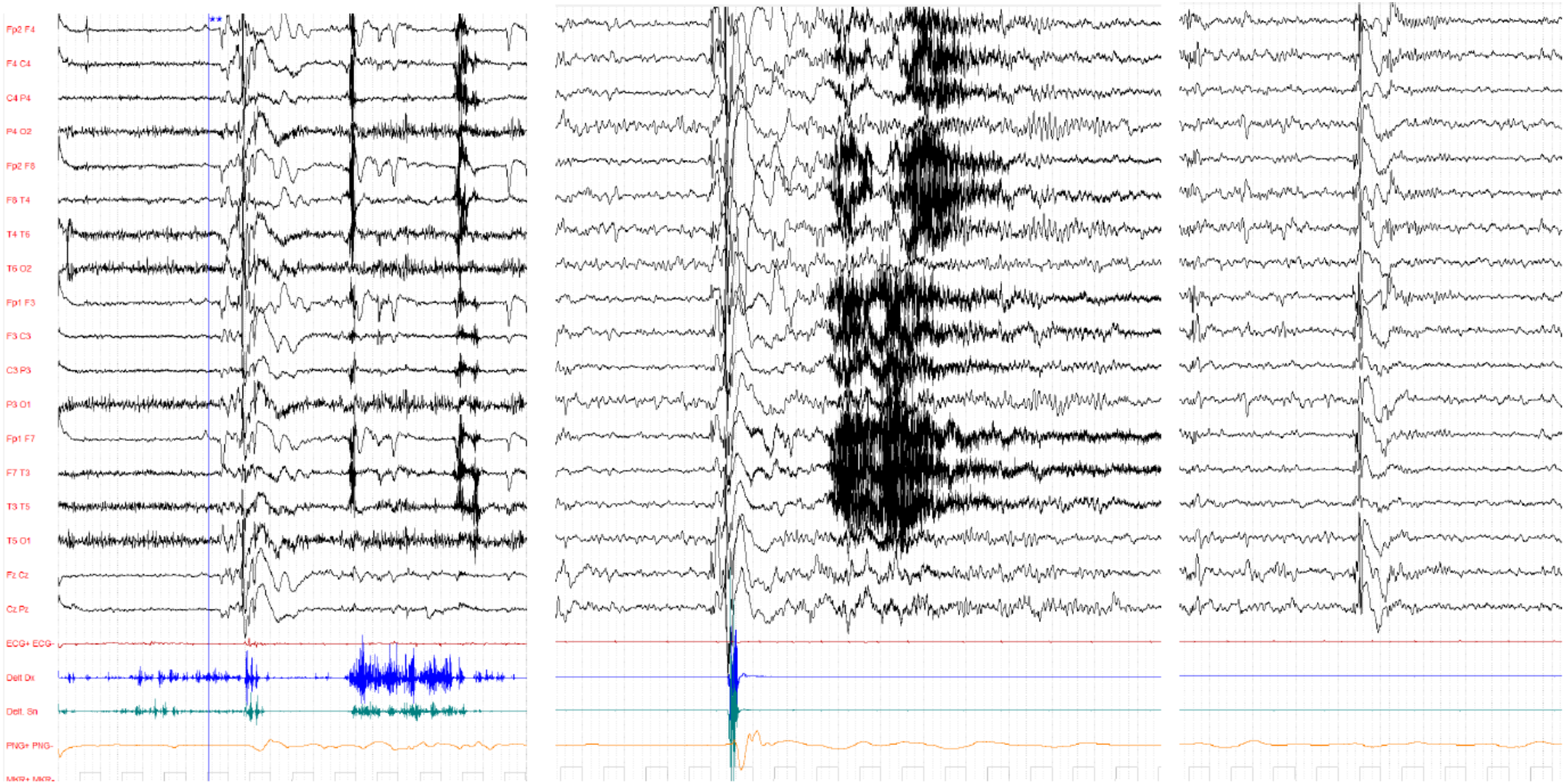
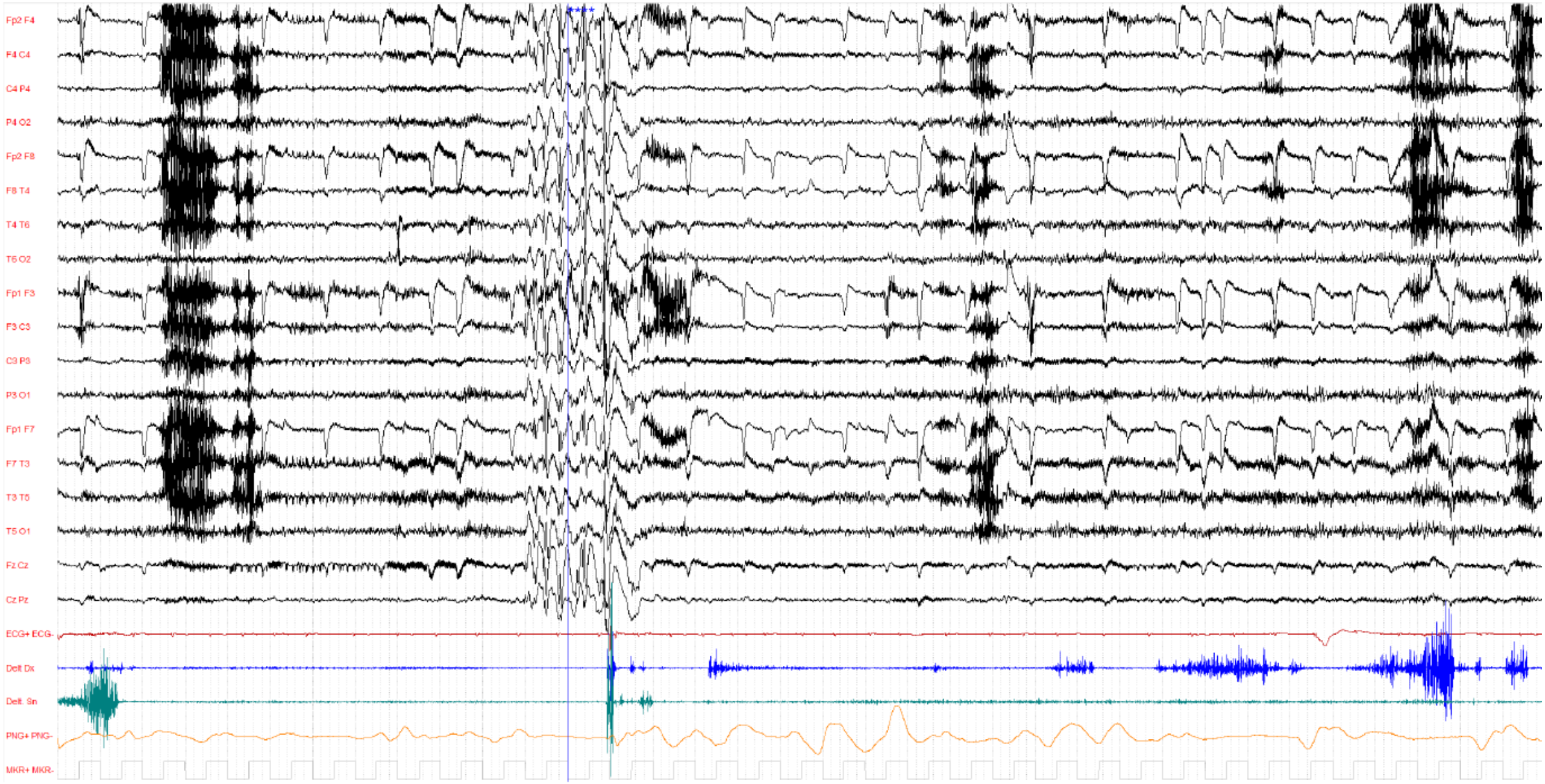
0070 Hz
9 sec
350 µV/cm
1 A EMG+PSG
8 Hz
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-

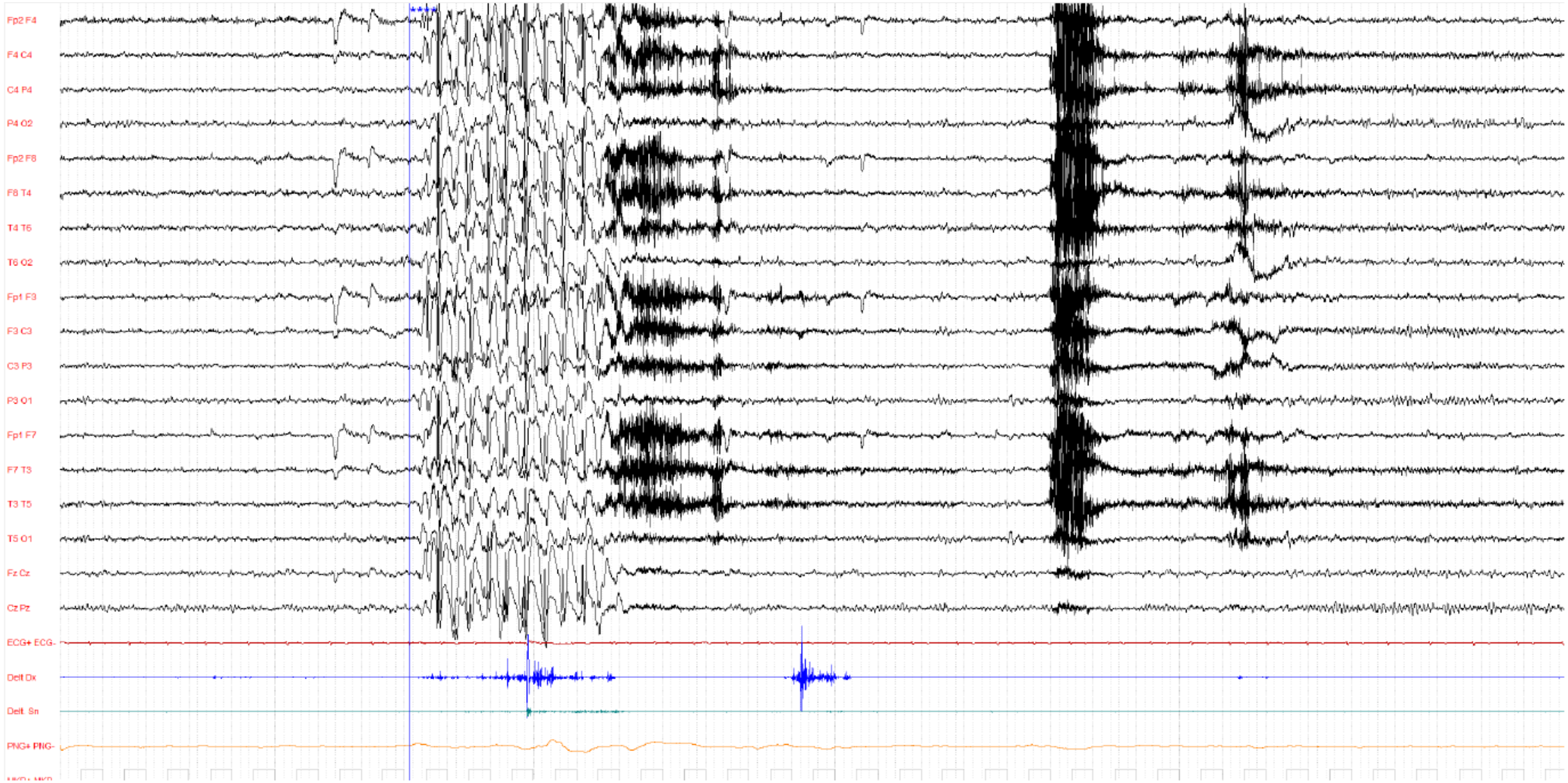

1.600 Hz
0070 Hz
9 sec
350 µV/cm
1 A EMG+PSG
10 Hz
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-



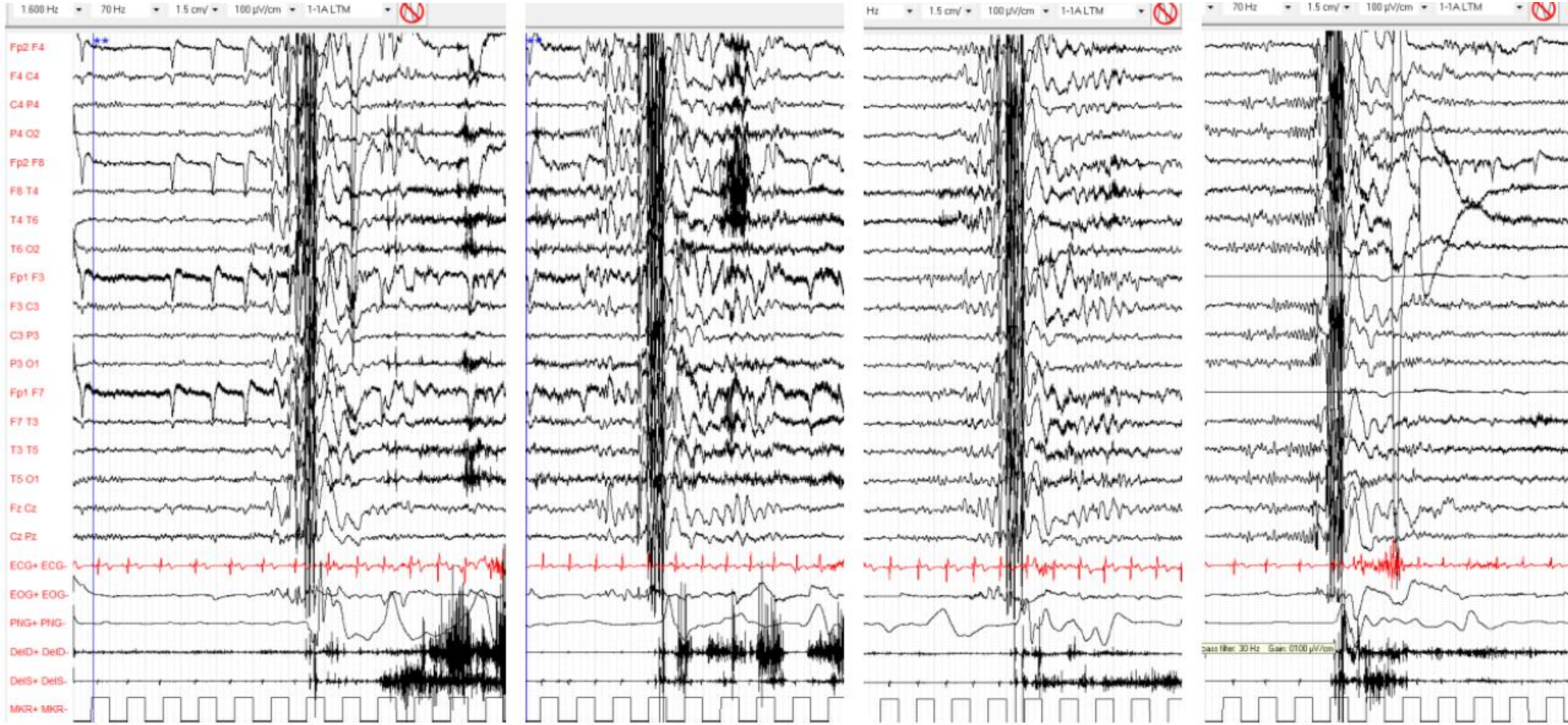
3 - parossismi con mioclonie in veglia, in addormentamento, in sonno senza mioclonie



4 - mioclonia in veglia

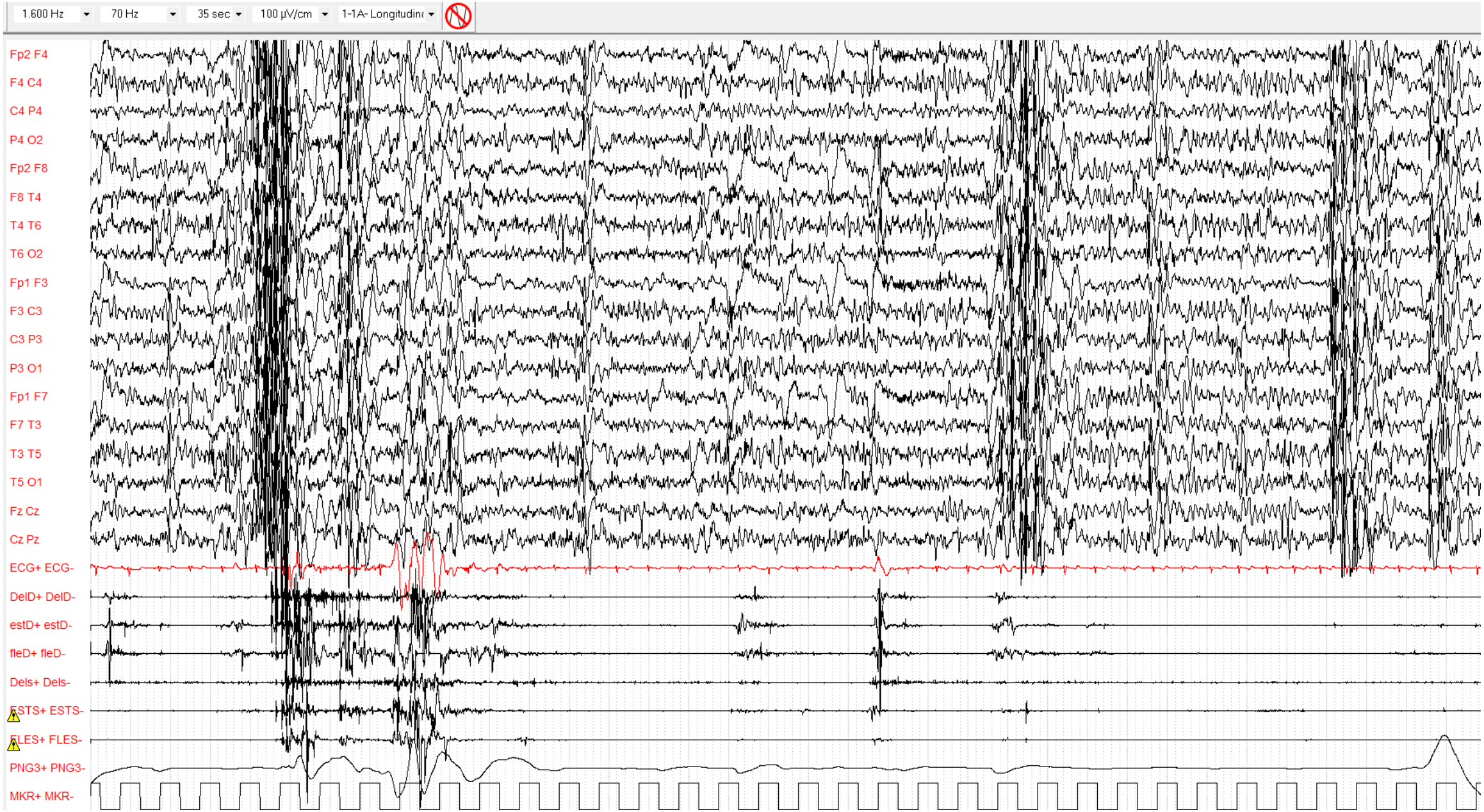


5 - alla apparenza nulla solo mioclonia molto leggera

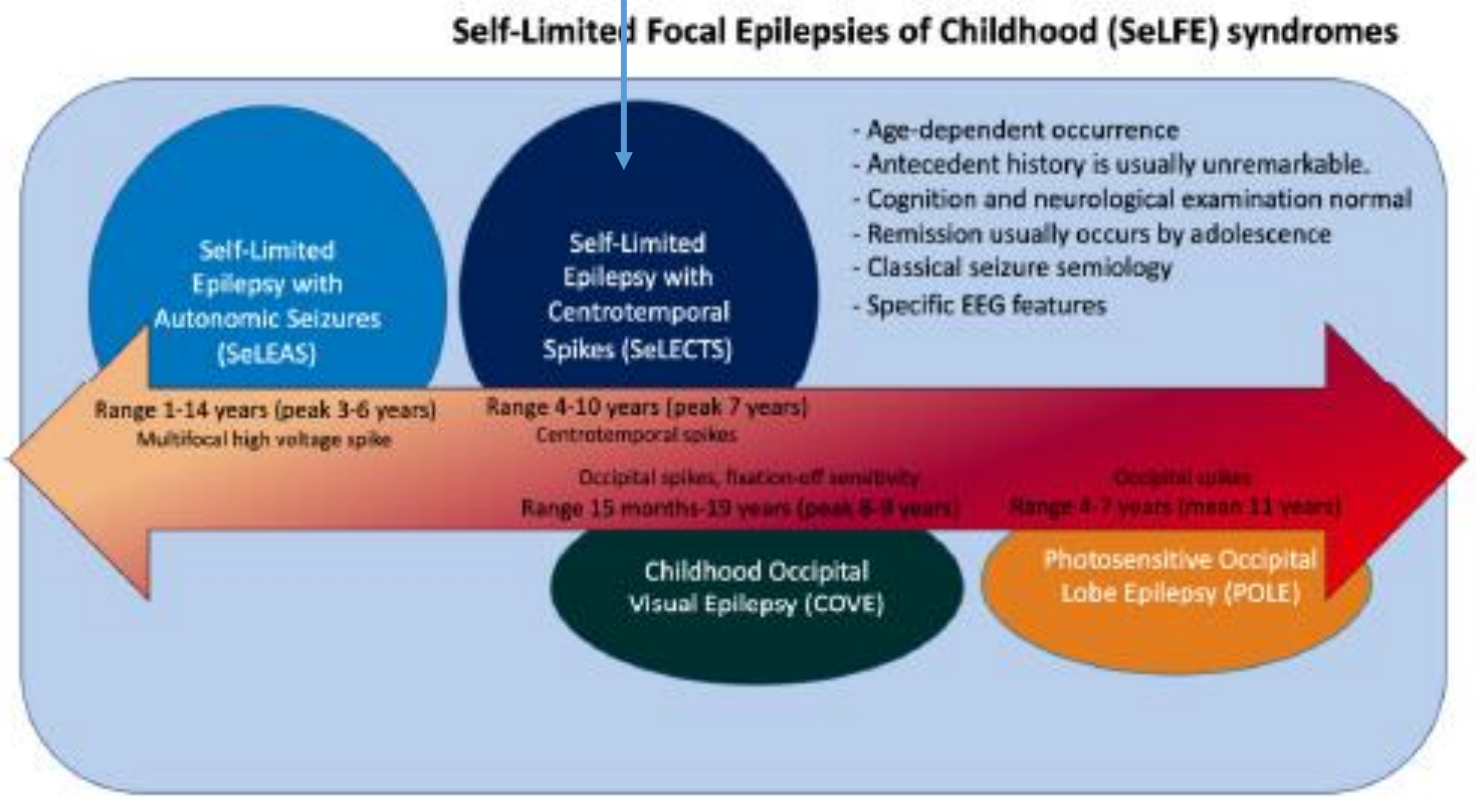


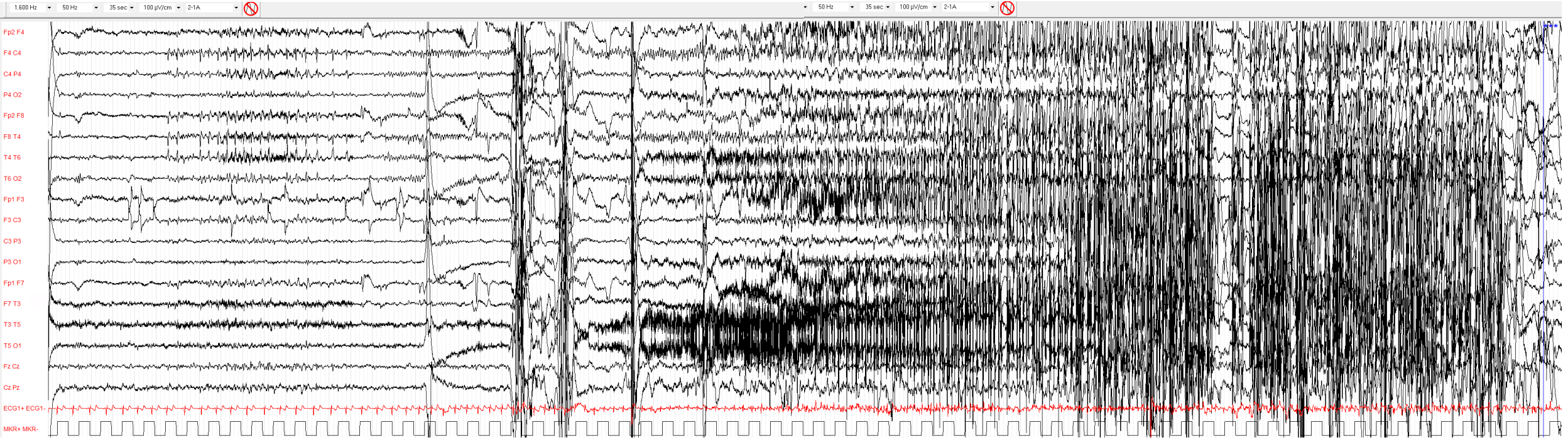
8 - grappolo di mioclonie alle 06:30 del mattino al risveglio

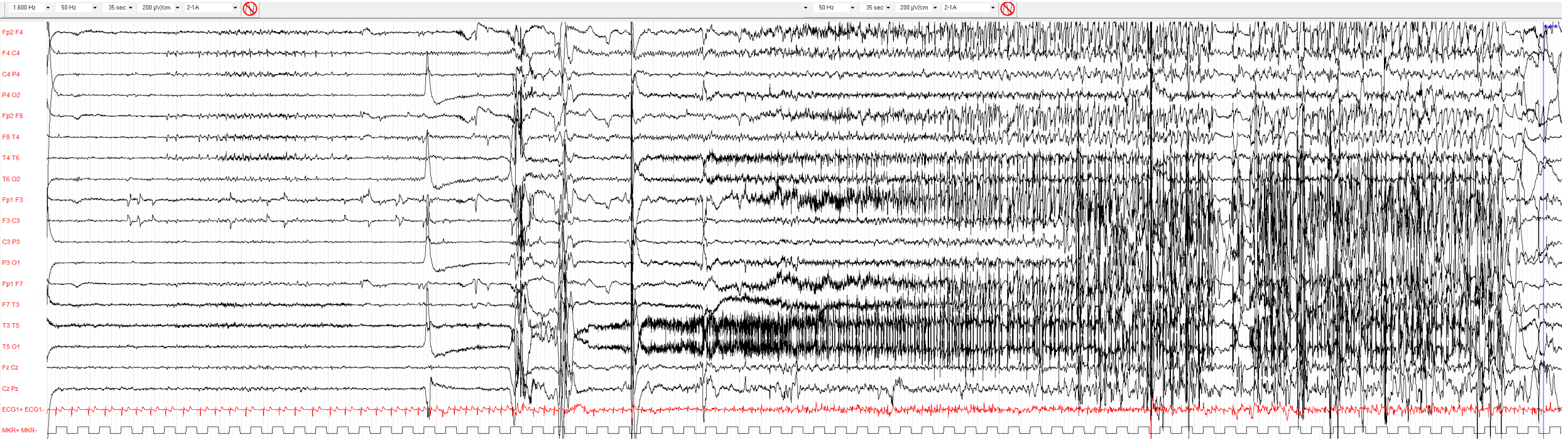
Lafora

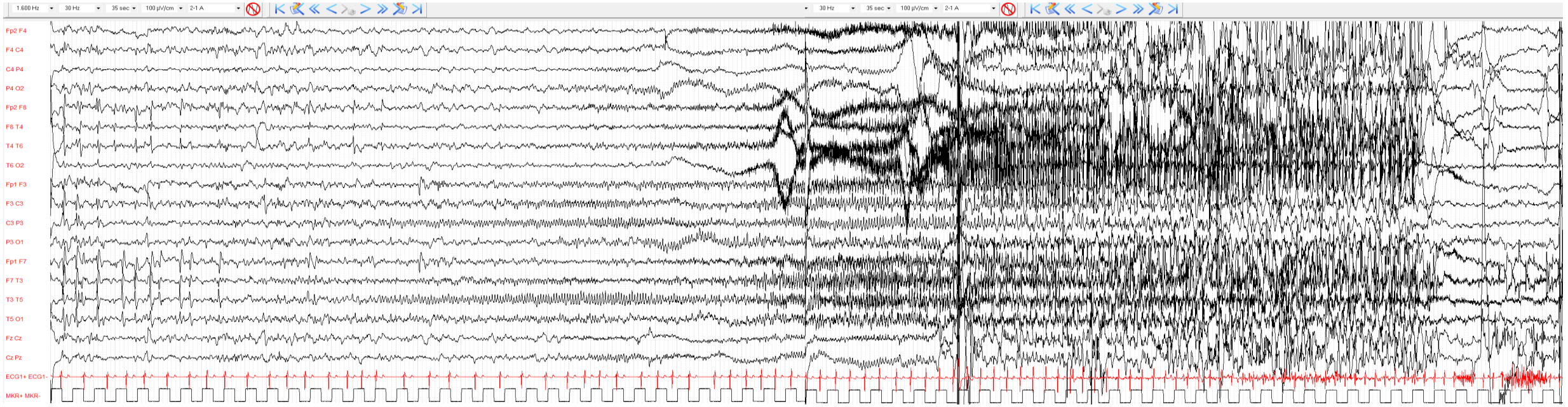

1.600 Hz
70 Hz
35 sec
100 µV/cm
1-1A- Longitudina
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
DelD+ DelD-
estD+ estD-
fleD+ fleD-
Dels+ Dels-
ESTS+ ESTS-
FLES+ FLES-
PNG3+ PNG3-
MKR+ MKR-

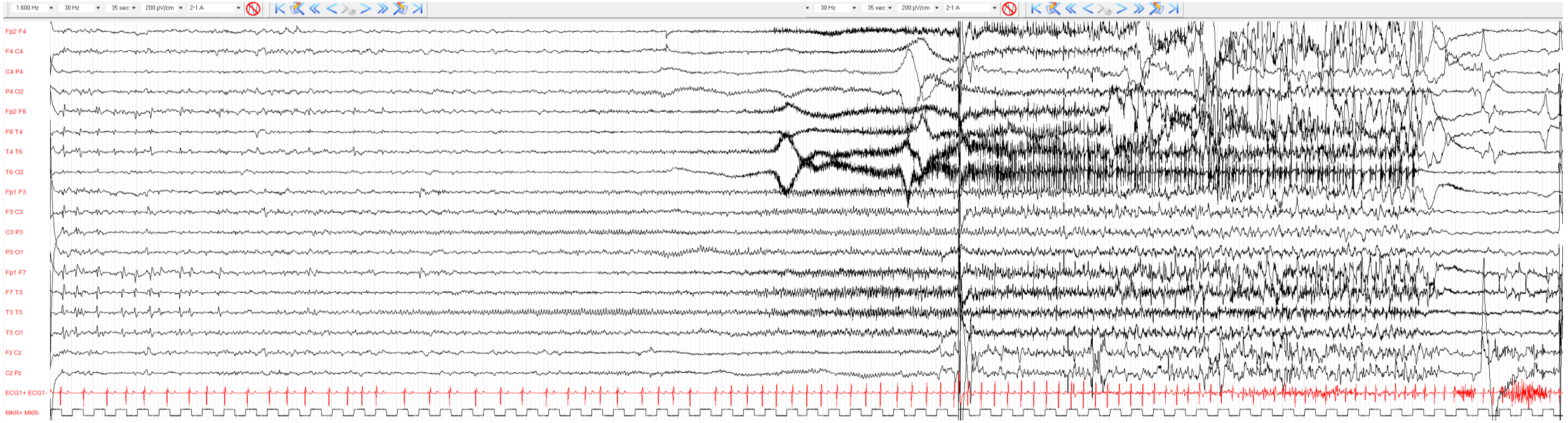
Self-Limited Epilepsy with Centrotemporal Spikes
Ex Rolandic Epilepsy or
Focal Idiopatic Epilepsy with Centrotemporal Spikes



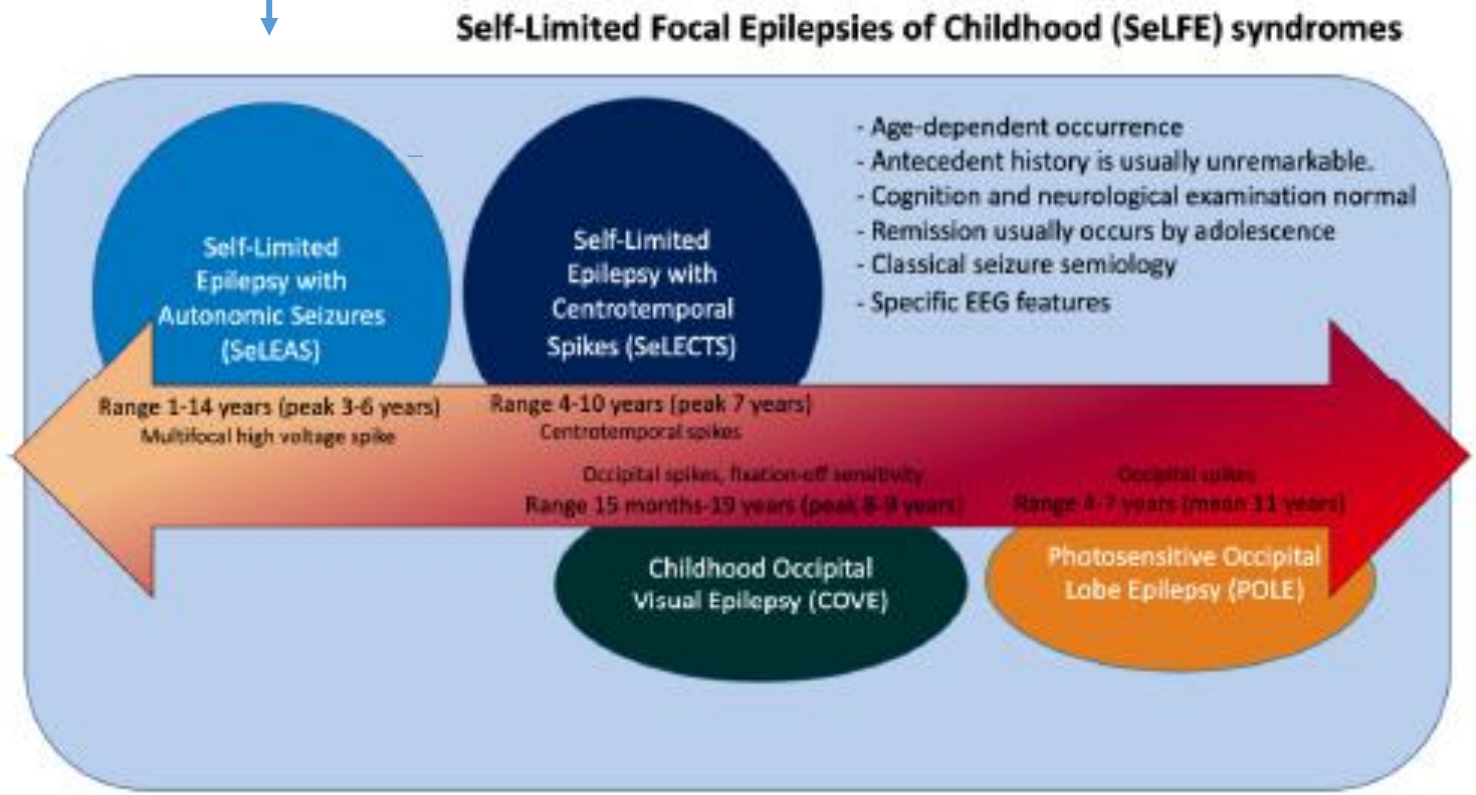

1.600 Hz
50 Hz
35 sec
100 µV/cm
2-1A
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
MKR+ MKR-

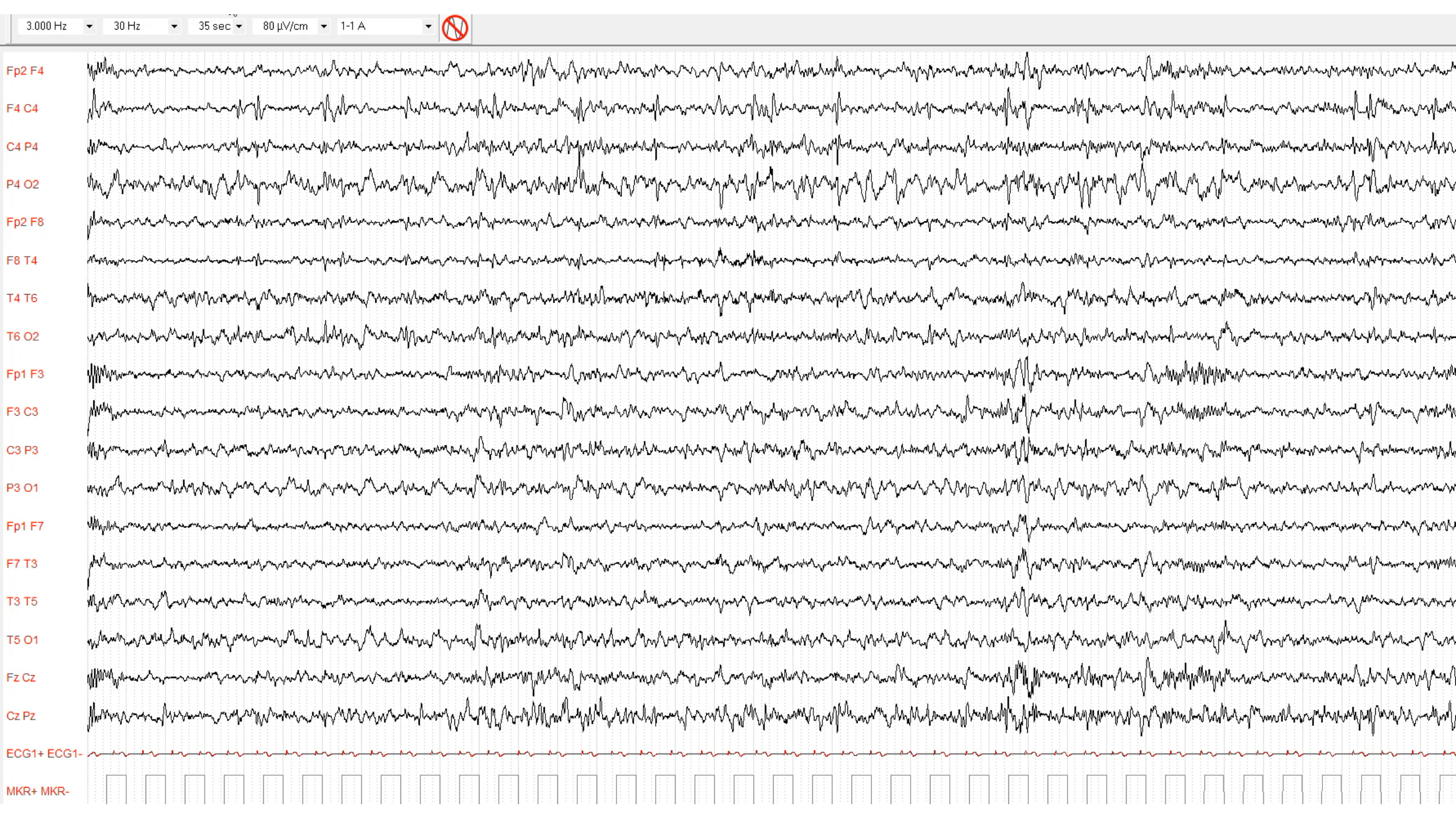

1.600 Hz
50 Hz
35 sec
200 µV/cm
2-1A
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
MKR+ MKR-


1.600 Hz
30 Hz
35 sec
100 µV/cm
2-1 A
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
MKR+ MKR-

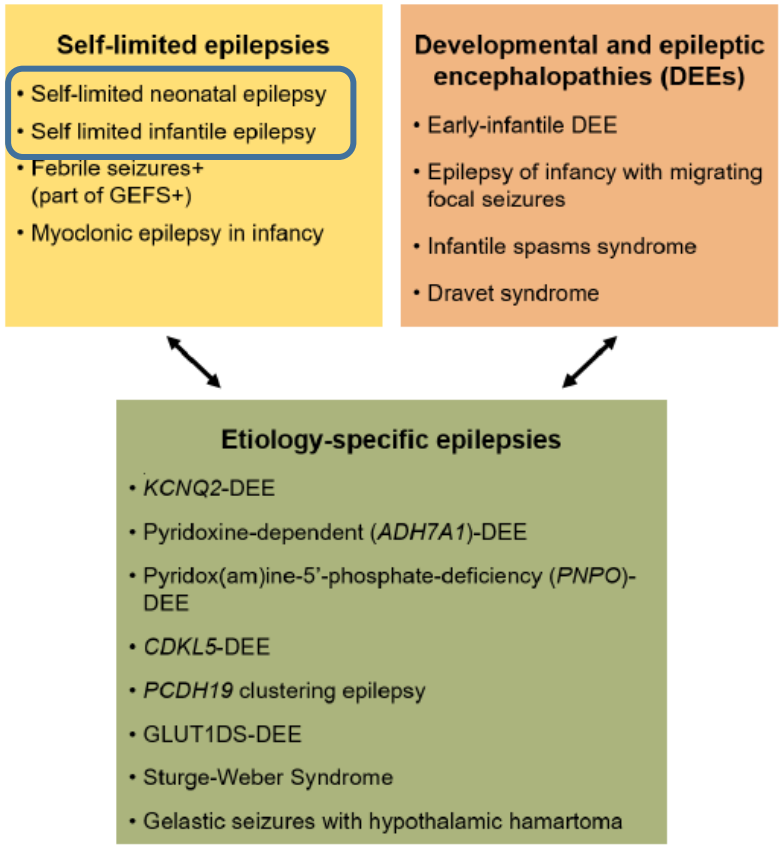

1.600 Hz
30 Hz
35 sec
200 µV/cm
2-1 A
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
MKR+ MKR-

Self-limited Epilepsy with Autonomic Seizures
Ex Panayiotopoulos Syndrome or
Focal Idiopatic Epilepsy woth Autonomic Seizures




3.000 Hz
30 Hz
35 sec
80 µV/cm
1-1 A
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
MKR+ MKR-

SEL-LIMITED EPILEPSY
1) NEONATE/INFANTILE



ILAE Classification & Definition of Epilepsy Syndromes in the Neonate and Infant: Position Statement by the ILAE Task Force on Nosology and Definitions

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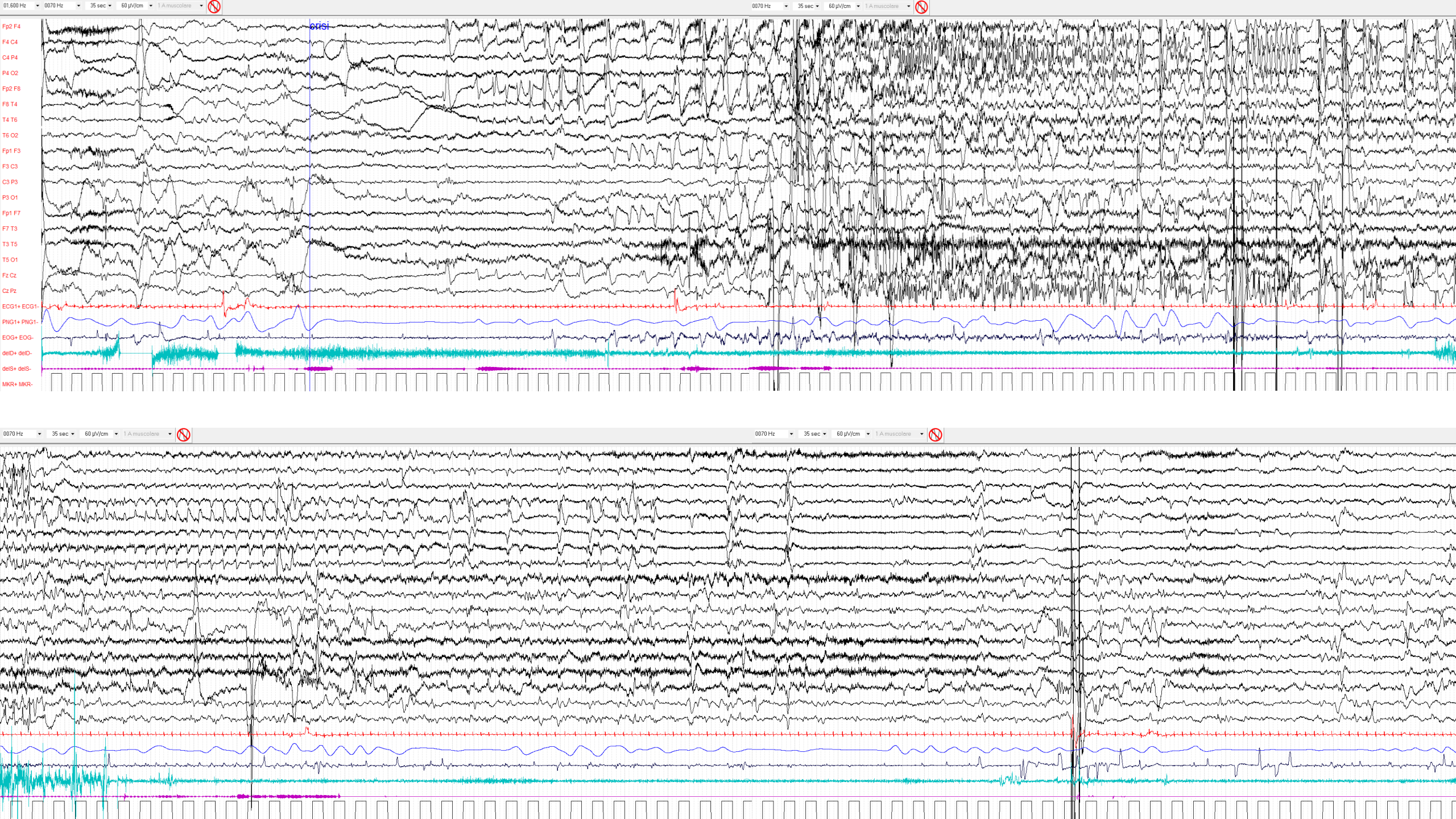
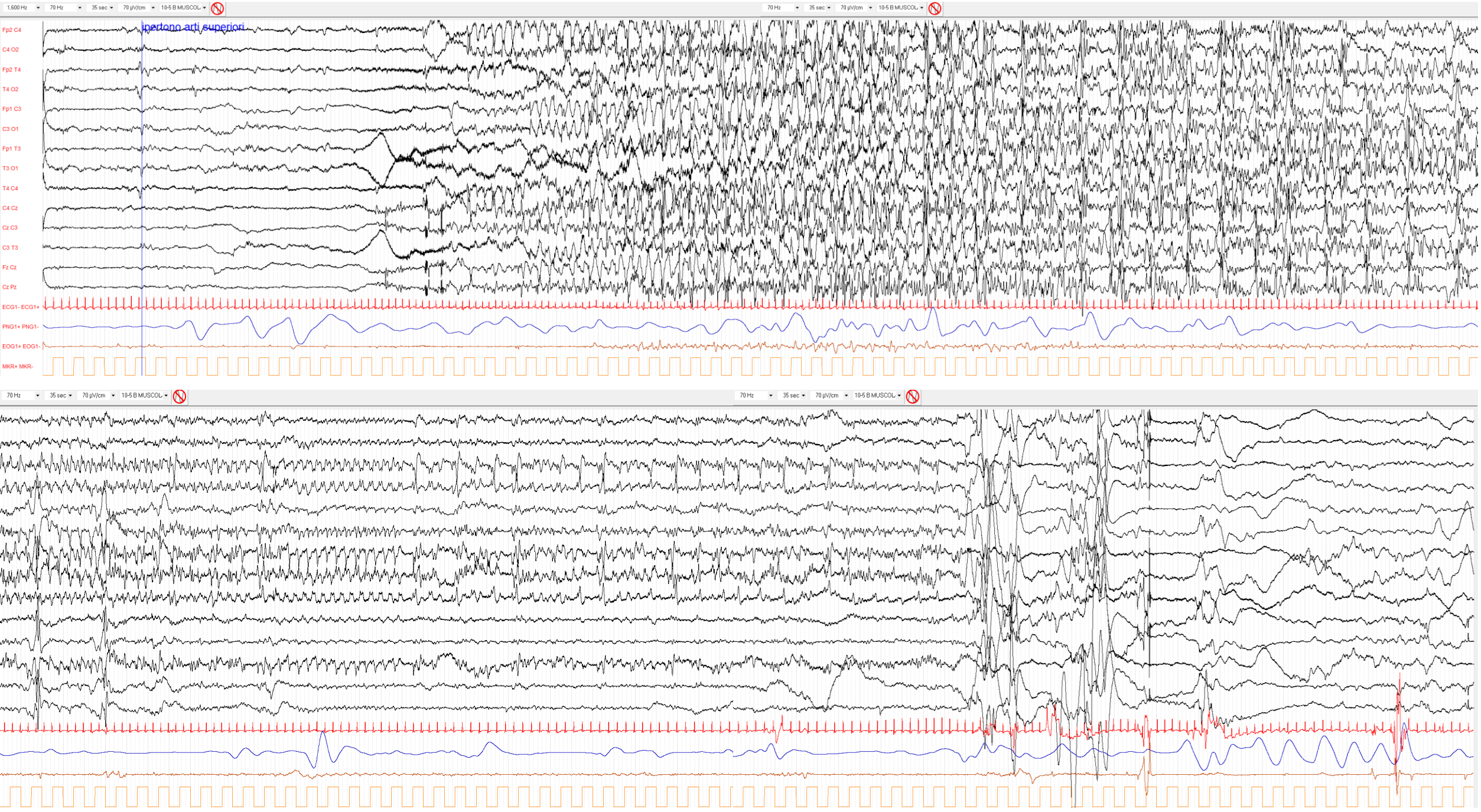
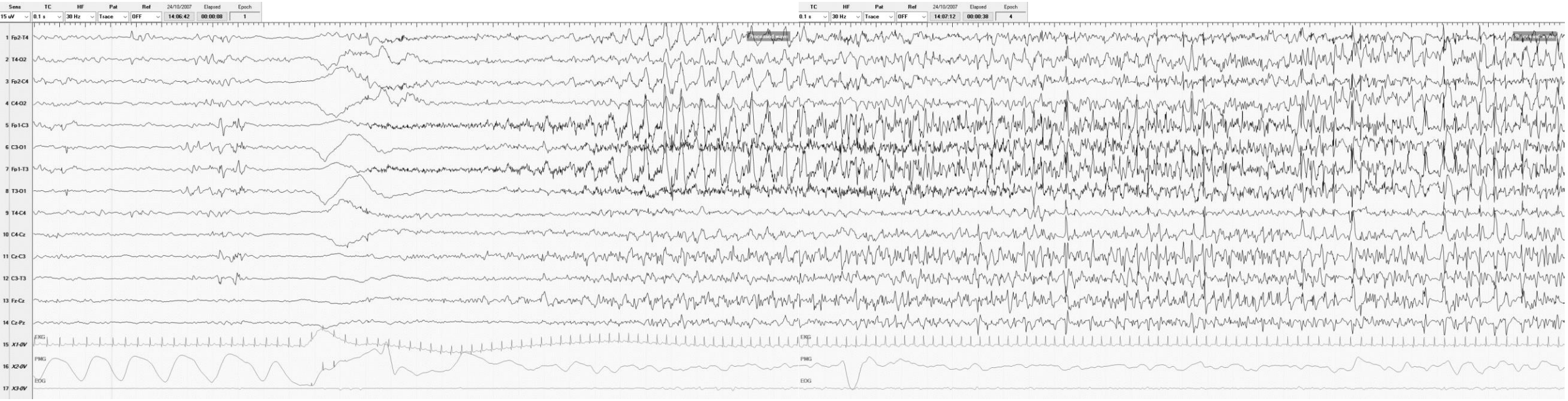
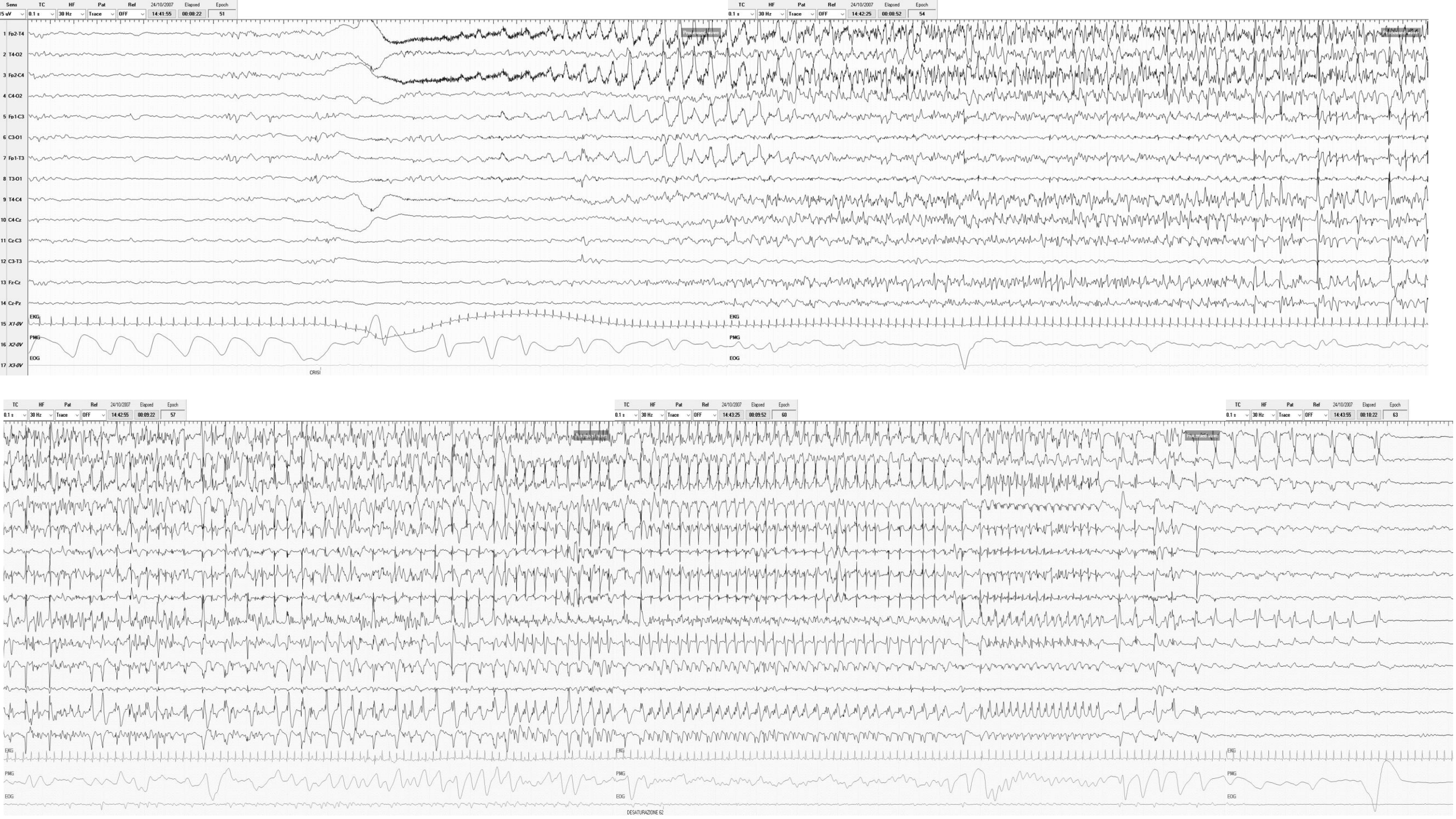

01,600 Hz
0070 Hz
35 sec
60 µV/cm
1 A muscolare
crisi
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
PNG1+ PNG1-
EOG+ EOG-
delD+ delD-
delS+ delS-
MKR+ MKR-

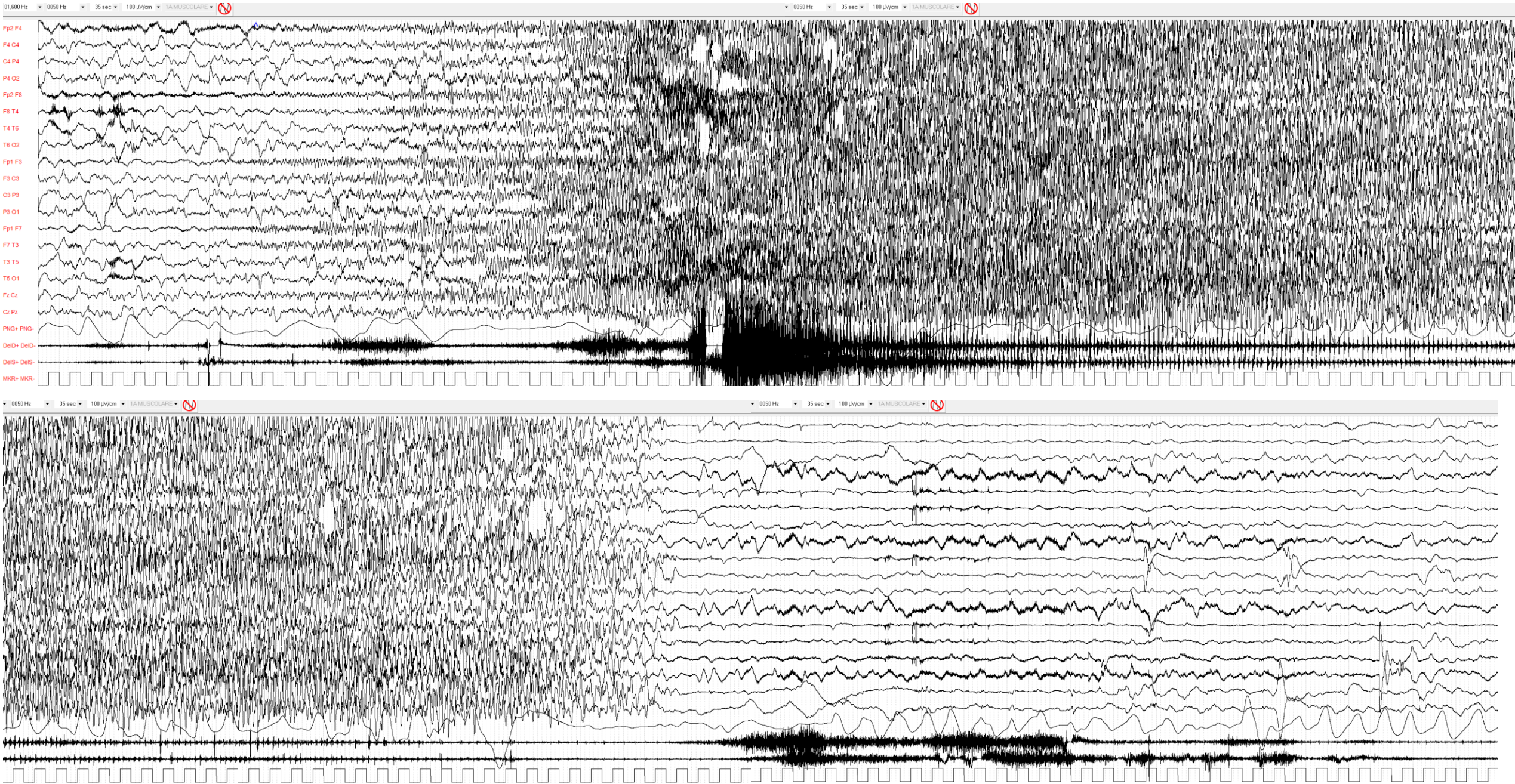

1,600 Hz
70 Hz
35 sec
70 µV/cm
10-5 B MUSCOL
ipertono arti superiori
Fp2 C4
C4 O2
Fp2 T4
T4 O2
Fp1 C3
C3 O1
Fp1 T3
T3 O1
T4 C4
C4 Cz
Cz C3
C3 T3
Fz Cz
Cz Pz
ECG1- ECG1+
PNG1+ PNG1-
EOG1+ EOG1-
MKR+ MKR-


Sens
15 uV
TC
0.1 s
HF
30 Hz
Pat
Trace
Ref
OFF
24/10/2007
14:06:42
Elapsed
00:00:08
Epoch
1
TC
0.1 s
HF
30 Hz
Pat
Trace
Ref
OFF
24/10/2007
14:07:12
Elapsed
00:00:38
Epoch
4
1 Fp2-T4
2 T4-O2
3 Fp2-C4
4 C4-O2
5 Fp1-C3
6 C3-O1
7 Fp1-T3
8 T3-O1
9 T4-C4
10 C4-Cz
11 Cz-C3
12 C3-T3
13 Fz-Cz
14 Cz-Pz
15 X1-0V
16 X2-0V
17 X3-0V
EKG
PMG
EOG


TC
0.1 s
HF
30 Hz
Pat
Trace
Ref
OFF
24/10/2007
14:07:42
Elapsed
00:01:08
Epoch
7
TC
0.1 s
HF
30 Hz
Pat
Trace
Ref
OFF
24/10/2007
14:08:12
Elapsed
00:01:38
Epoch
10
EKG
PMG
EOG
SA02 47


Sens
TC
HF
Pat
Ref
24/10/2007
Elapsed
Epoch
15 uV
0.1 s
30 Hz
Trace
OFF
14:41:55
00:08:22
51
14:42:25
00:08:52
54
1 Fp2-T4
2 T4-O2
3 Fp2-C4
4 C4-O2
5 Fp1-C3
6 C3-O1
7 Fp1-T3
8 T3-O1
9 T4-C4
10 C4-Cz
11 Cz-C3
12 C3-T3
13 Fz-Cz
14 Cz-Pz
15 X1-0V
16 X2-0V
17 X3-0V
EKG
PMG
EOG
CRISI
14:42:55
00:09:22
57
14:43:25
00:09:52
60
14:43:55
00:10:22
63
DESATURAZIONE 62

Female 6 mnths, PRRT2 mutation, focal seizures to bilateral tonic-clonic with PGES



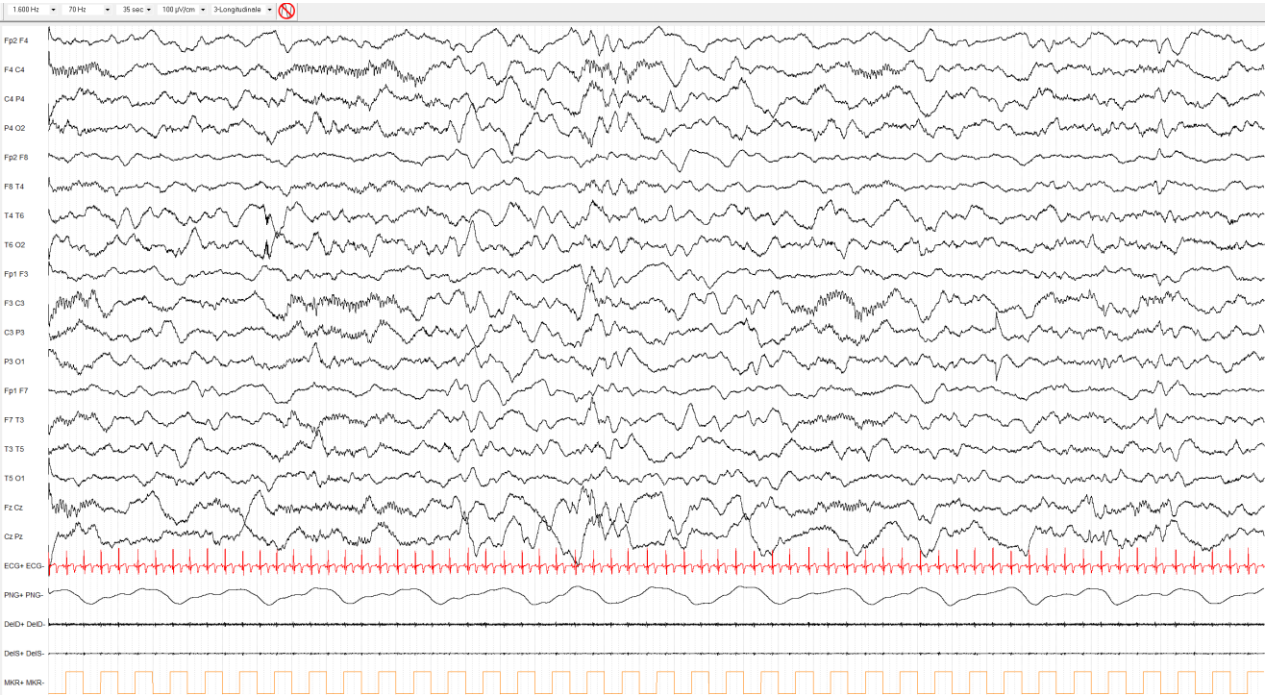
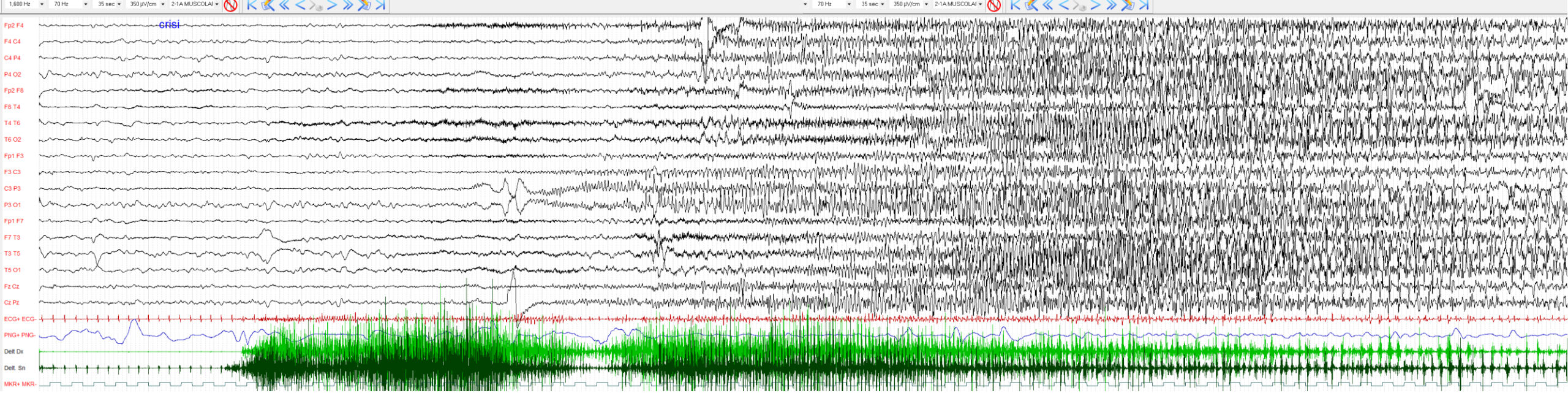


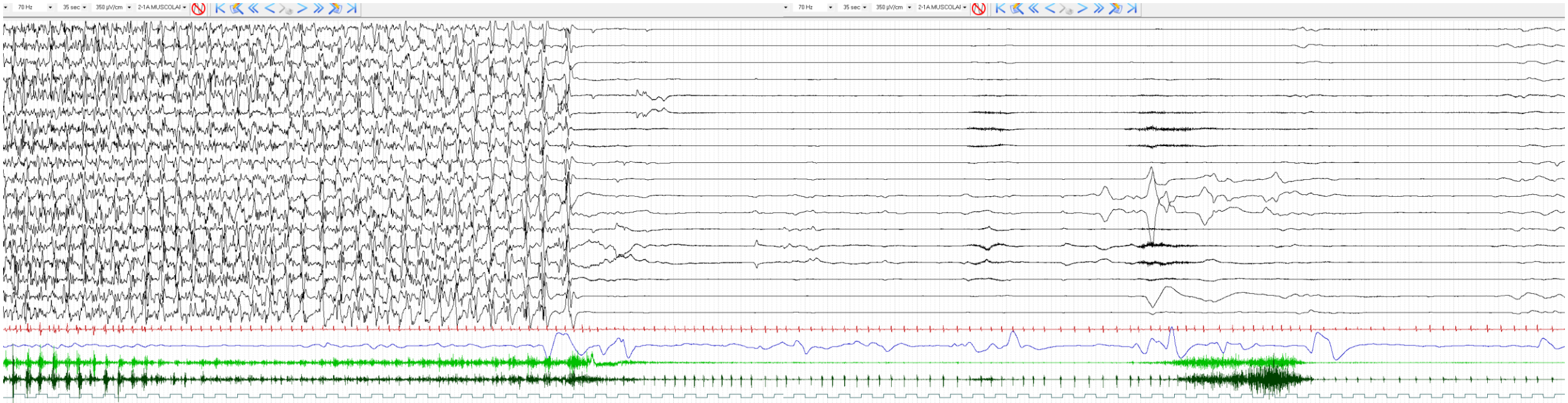
Figure 1c
Well organized sleep, phase N2 NREM
Spindles are evident symmetric and
at time asynchrounous
No epileptiform abnormalities in the sleep interictal EEG

Figure 1d
During wakefulness the theta-beta pysiological activity
is intermingled with delta waves, expecially over the bilateral
posterior regions
No epileptiform abnotmalities are evident in the wakefulness interictal EEG

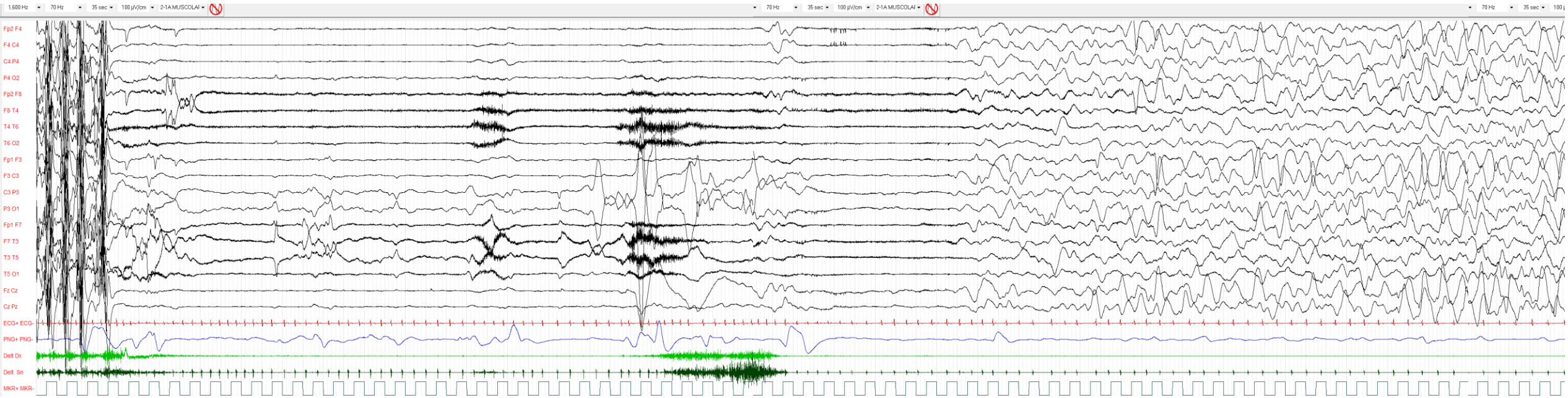
Female 6 mnths, PRRT2 mutation, focal seizures to bilateral tonic-clonic with PGES



TrFi 13 mesi, 4 convulsioni neonatali Pb per 4 mesi, poi a 13 mesi grappolo di 13 crisi in 36 ore, CBZ per 4 mesi
Poi benessere - Mutazione KCNQ2 Crisi focal to bilateral tonic-clonic con PGES




70 Hz
35 sec
350 µV/cm
2-1A MUSCOLAI
70 Hz
35 sec
350 µV/cm
2-1A MUSCOLAI


1.600 Hz
70 Hz
35 sec
100 µV/cm
2-1A MUSCOLAI
70 Hz
35 sec
100 µV/cm
2-1A MUSCOLAI
70 Hz
35 sec
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
PNG+ PNG-
Delt Dx
Delt. Sn
MKR+ MKR-

Crisi sintomatiche acute da infezioni, stroke, traumi: Focali cloniche
Crisi in DEE o in Self limited: Focali toniche

Neonatal seizures: When semiology points to etiology

Marta Elena Santarone, Nicola Pietrafusa, Lucia Fusco*

Seizure: European Journal of Epilepsy 80 (2020) 161–165

Acute Symptomatic
- Focal Motor Clonic 30/31 (p>0.05)
  - Stroke 13/13
  - HIE 8/9
  - Infective 9/9

Remote Symptomatic
- Focal Motor Tonic 19/20 (p>0.05)
  - DEE 11/11
  - Self-limited neonatal 4/4
  - Malformative 2/2
  - Metabolic 2/2

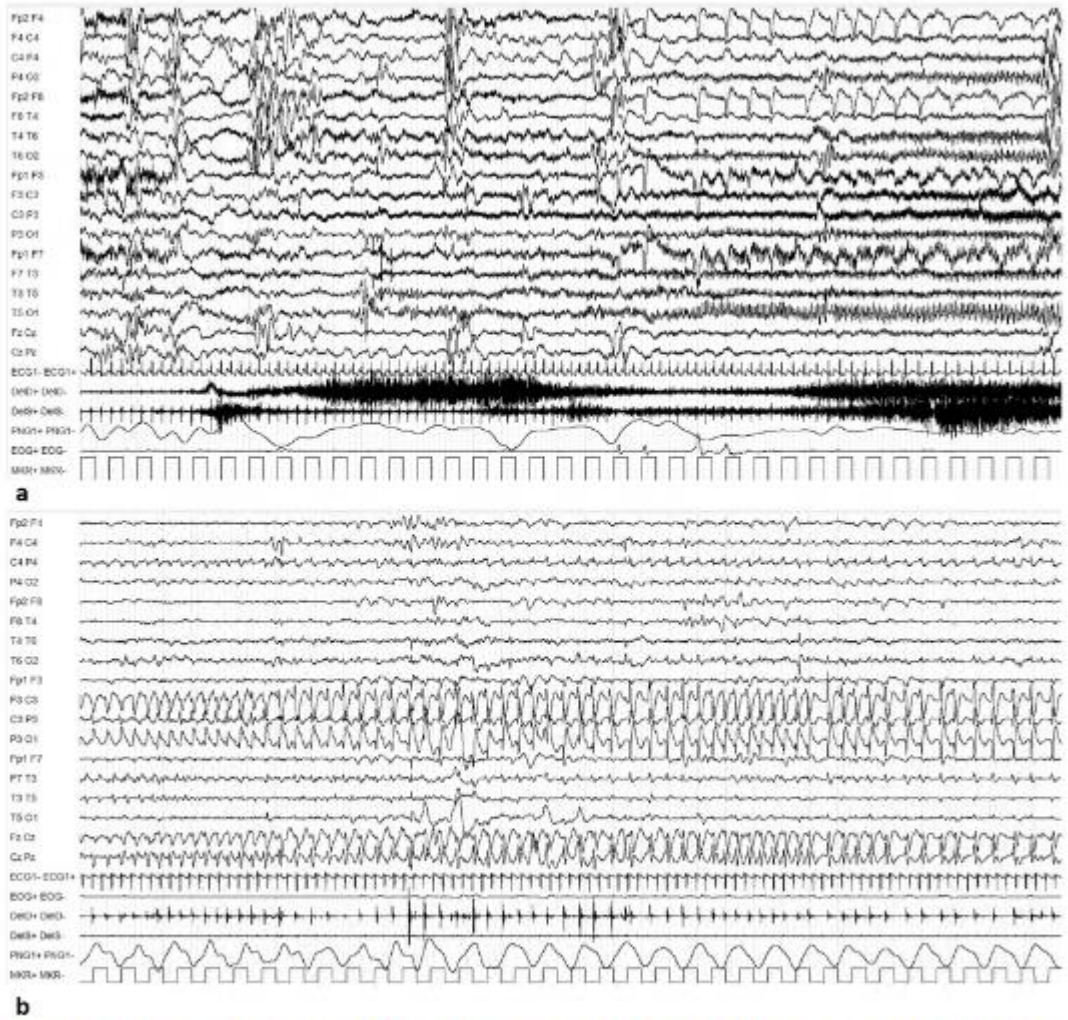
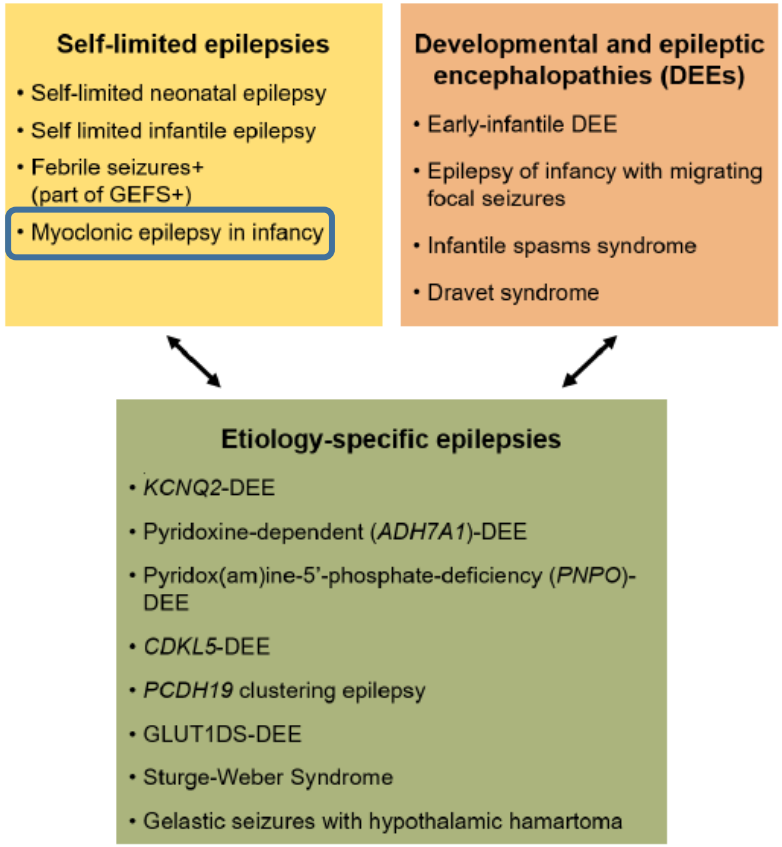


Fig. 2. a) Focal tonic seizure with a clearcut discharge beginning over the left posterior regions in a 27 days old newborn with KCNQ2-related DEE. LFF 1,600 Hz, HFF 70 Hz, speed 1.5 cm/sec, amplitude 70μV/cm. b) Right focal clonic seizure associated to a left central EEG discharge in a 21 days old newborn with infective encephalitis. LFF 1,600 Hz, HFF 70 Hz, speed 1.5 cm/sec, amplitude 250μV/cm.

SEL-LIMITED EPILEPSY
2) MYOCLONIC EPILEPSY OF INFANCY



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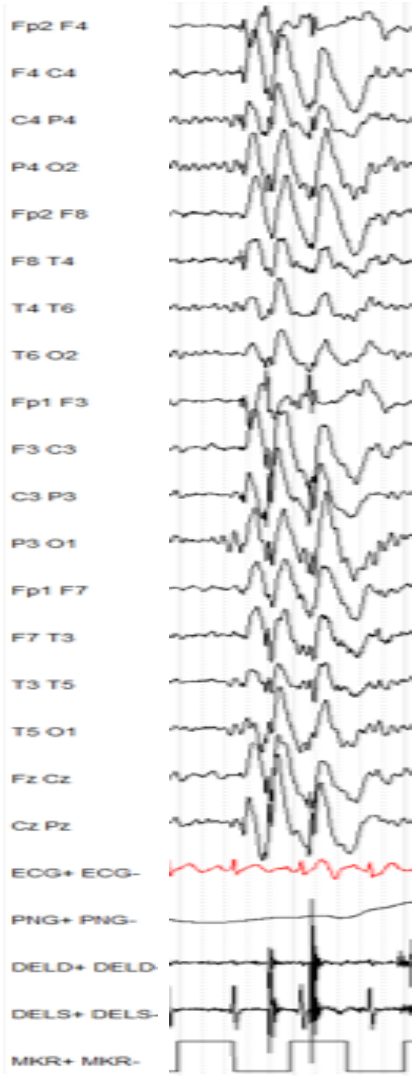
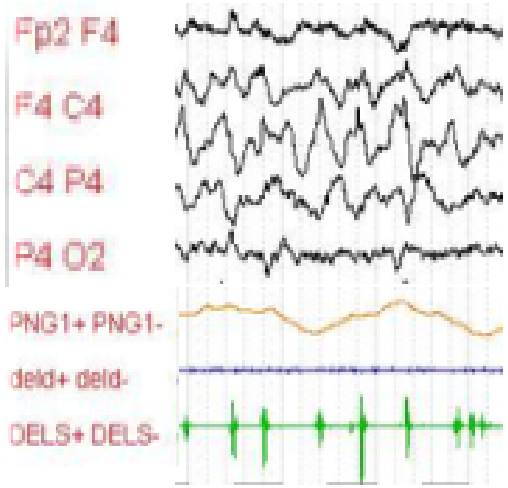
Il mioclono è chiamato EPILETTICO quando si associa a una controparte EEG epilettiforme sia diffusa sia focale

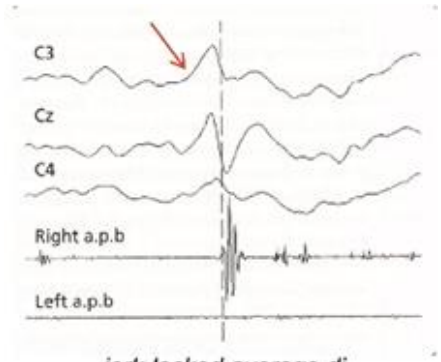
- scarica epilettiforme diffusa nel mioclono generalizzato
- punte focali centrali controlaterali, nel mioclono corticale focale



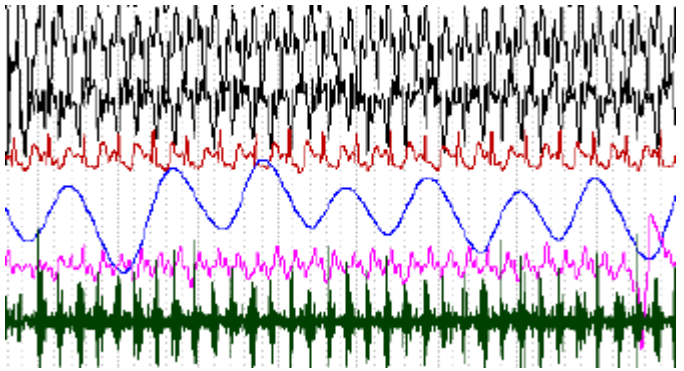
Mioclono Generalizzato



Mioclono Focale

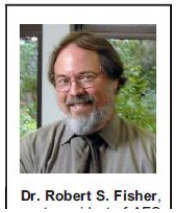


jerk-locked average di 25 mioclonie sull'abp dx



Instruction manual for the ILAE 2017 operational classification of seizure types

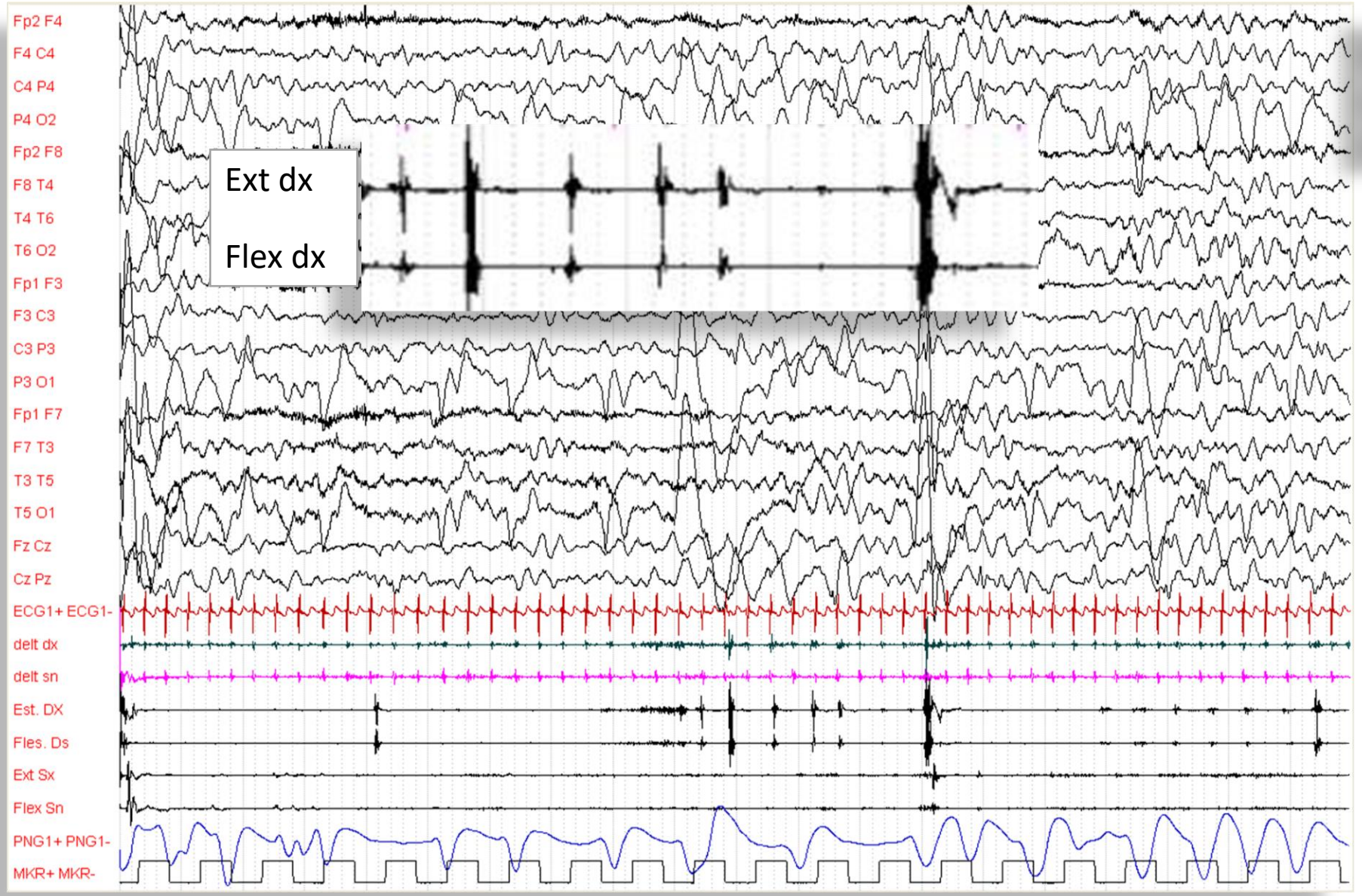
Epilepsia, 58(4):531–542, 2017



Dr. Robert S. Fisher

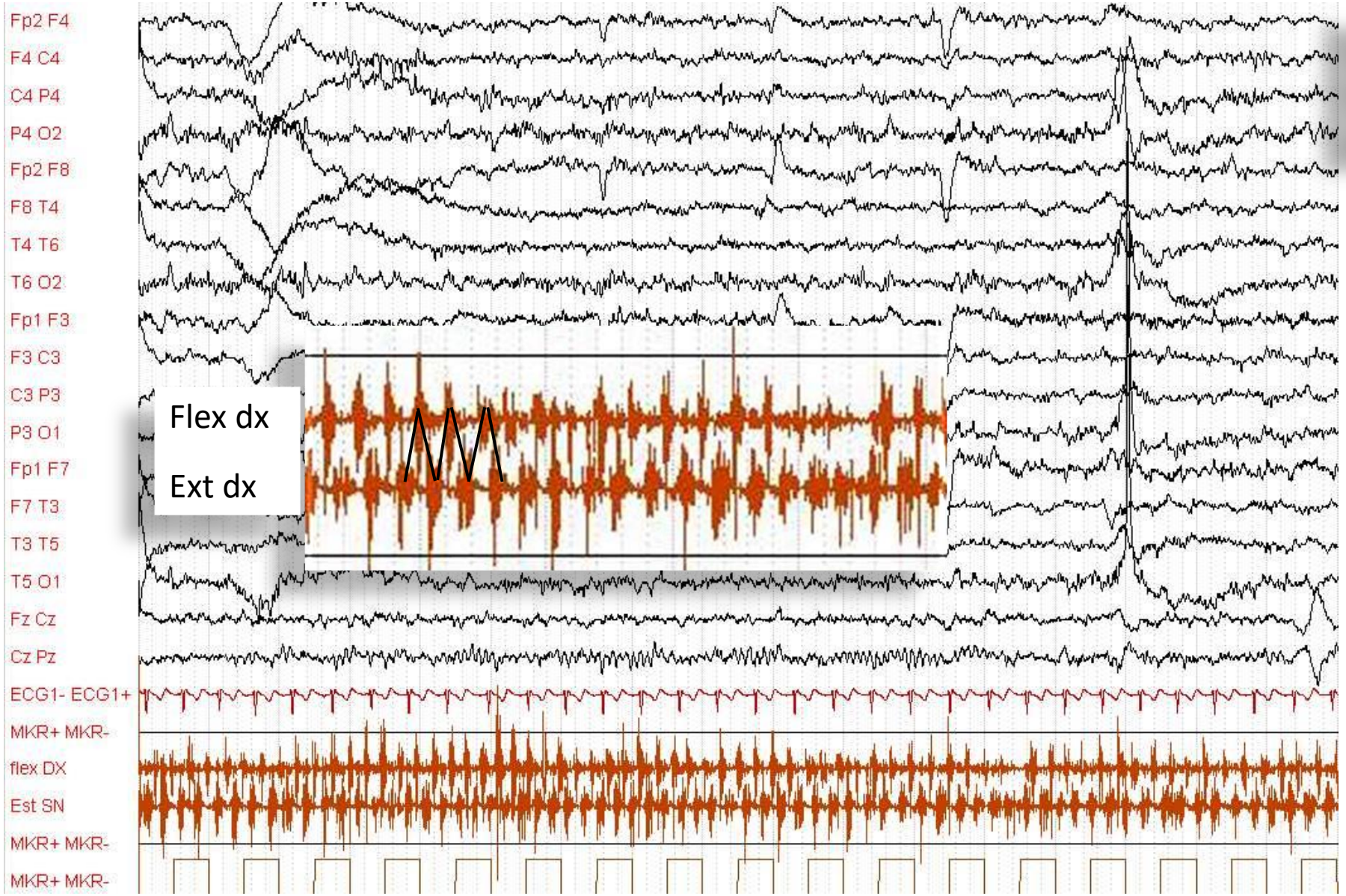
The distinction between clonic and myoclonic is somewhat arbitrary, but clonic implies sustained, regularly spaced stereotypical jerks, whereas, myoclonus is less regular and in briefer runs.

Bambino Gesù
OSPEDALE PEDIATRICO



Myoclonus:
co-contraction
agonist/antagonist muscles

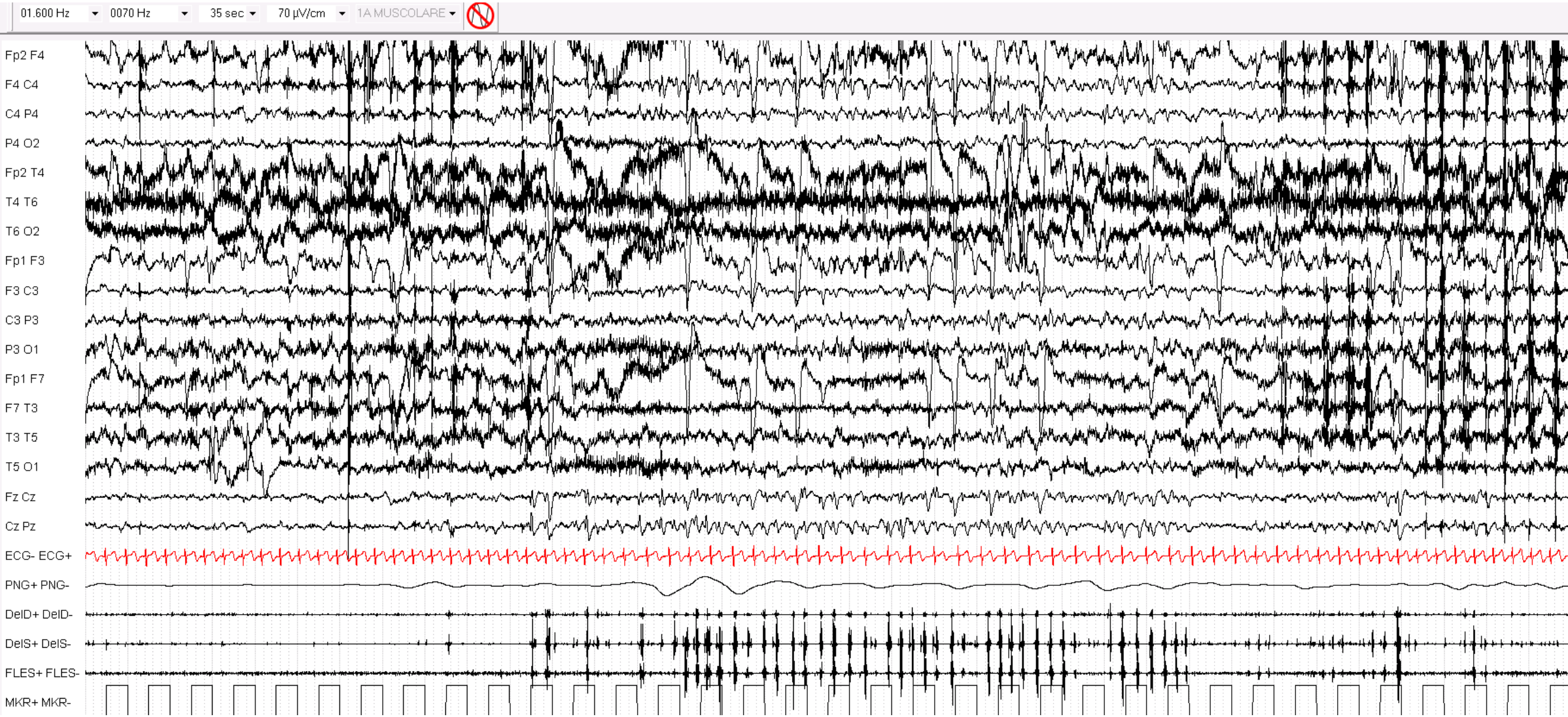
Bambino Gesù
OSPEDALE PEDIATRICO



Tremor:
Triangular contraction of agonist/antagonist muscles

Str Car 8 anni

Esito HIE - mioclono corticale focale in brevi crisi



EEG: technical notes

EOG LFF: 15 Hz
HFF: 10.000
GAIN: 200 µ

RESP LFF: 0,53 H
HFF: 15 Hz
GAIN: 200 µv/cm

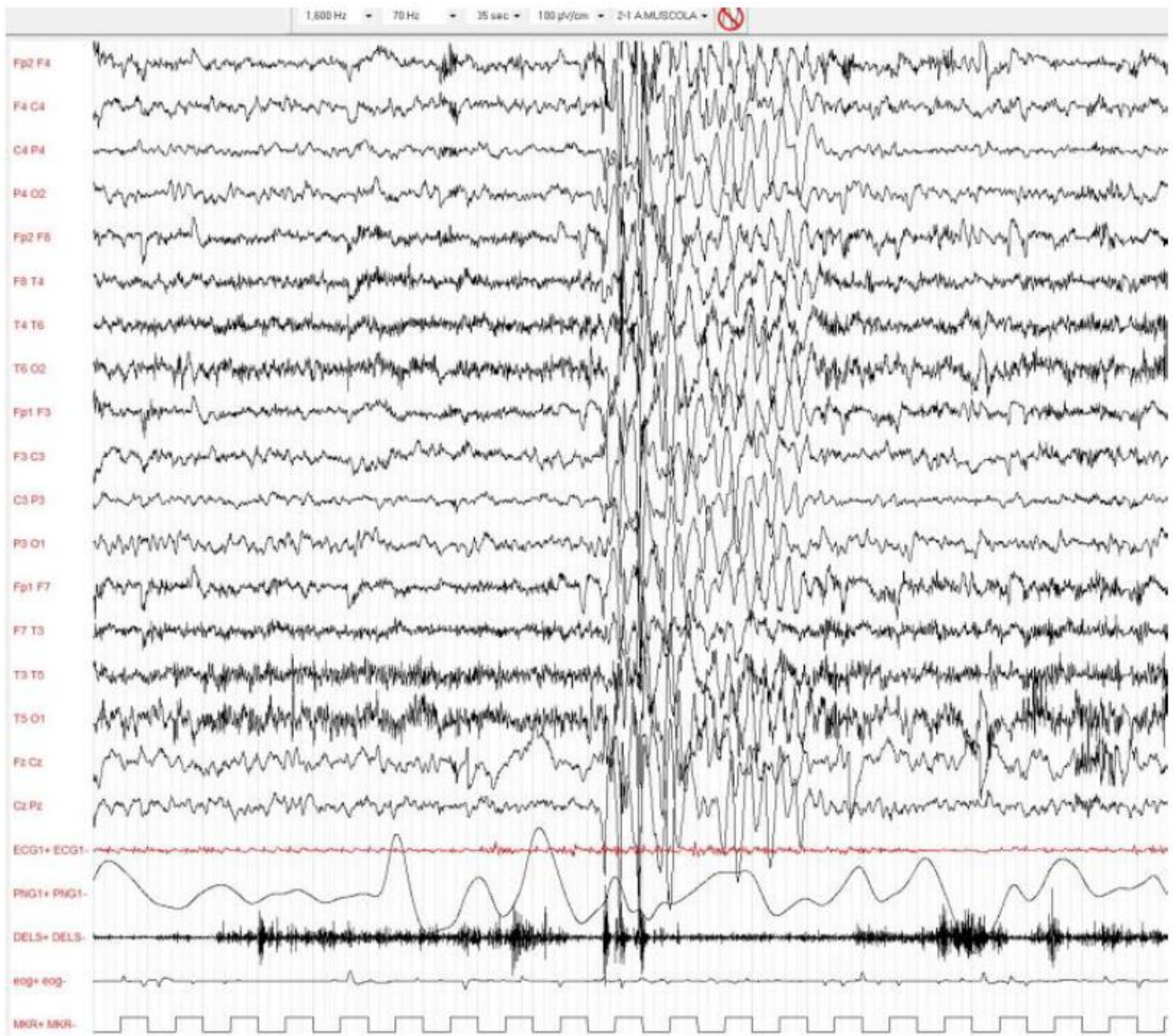
MUSCLE LFF: 53 Hz
HFF: 400 Hz
GAIN: <30 µv/cm

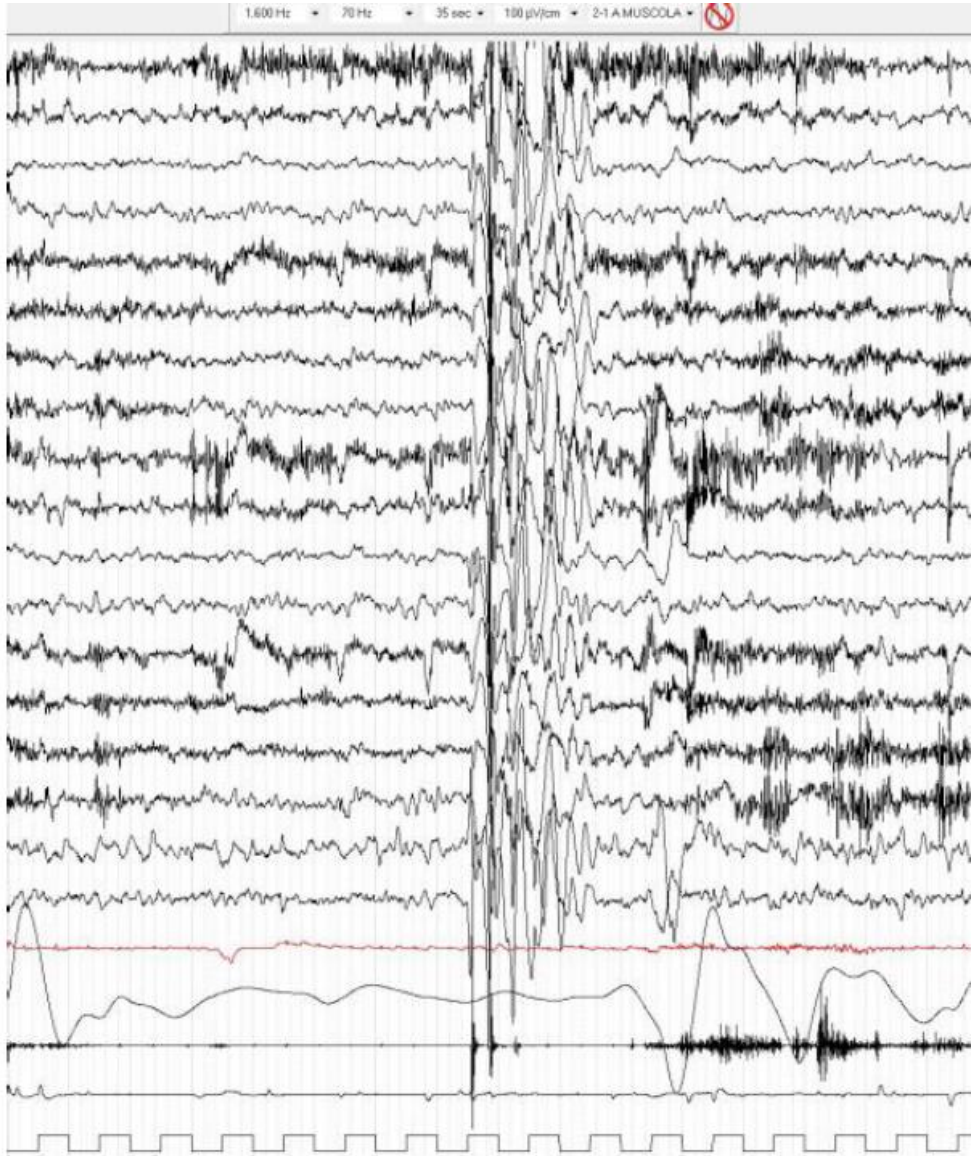
MUSCLE LFF: 53 Hz
HFF: 400 Hz
GAIN: <30 µv/cm

/cm
HFF: 15 Hz
GAIN: >800 µv/cm

Bambino Gesù
OSPEDALE PEDIATRICO

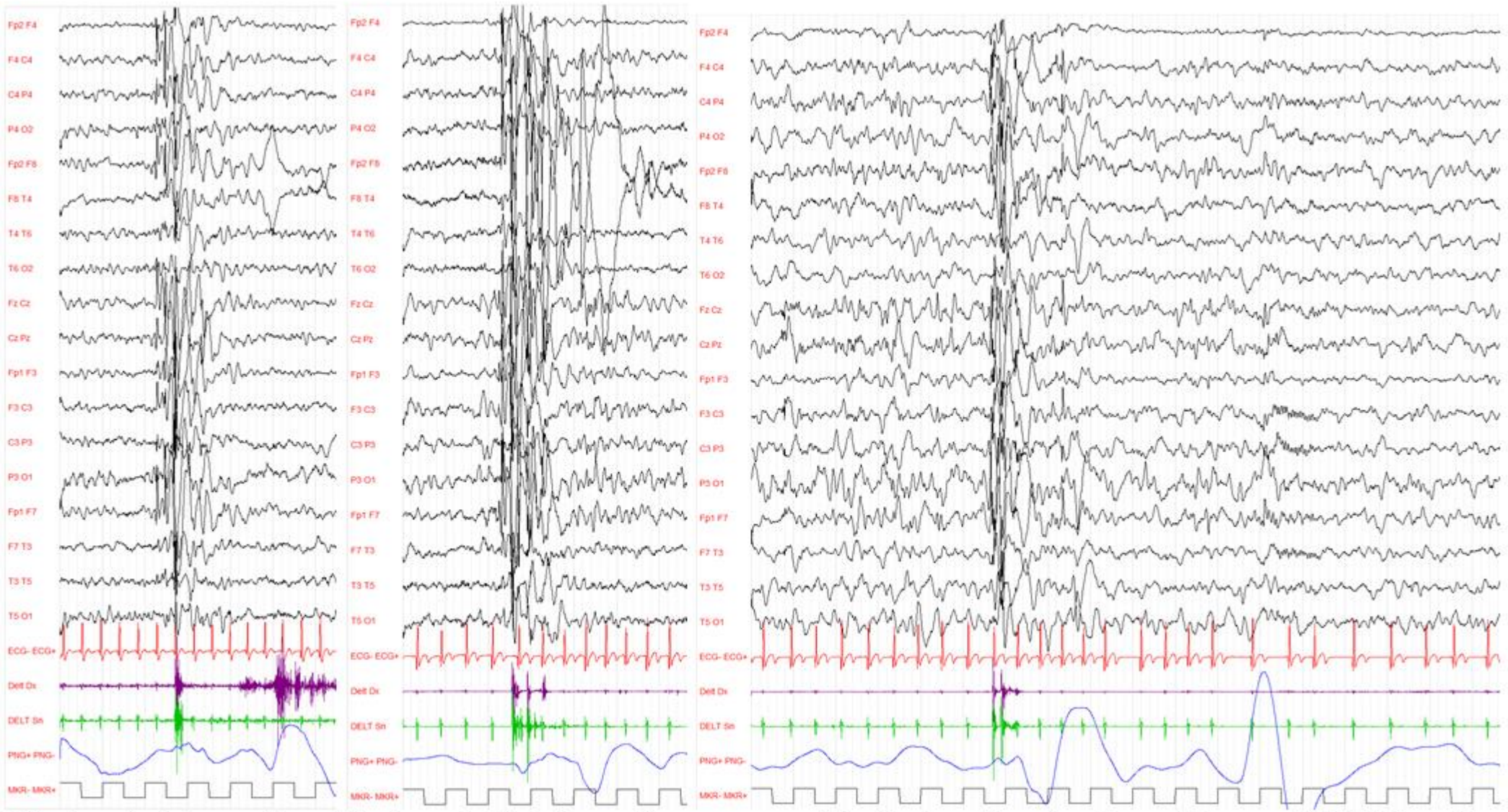
Ada Bry 16 mesi Epilessia Mioclonica della Infanzia





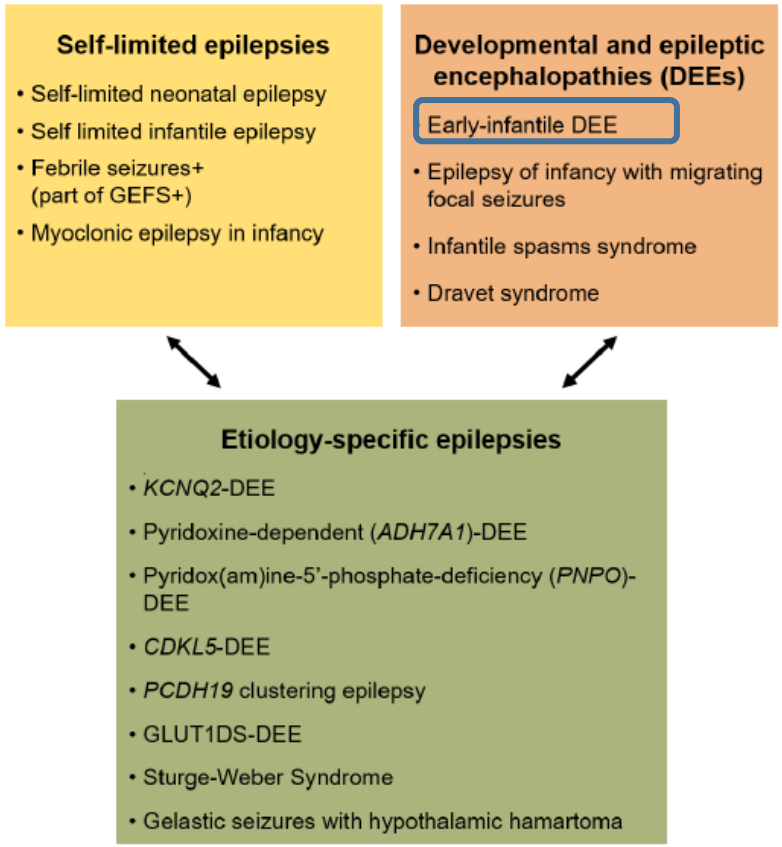
6 - mioclonie in veglia alla apparenza spontanee

Mec Fil 15 mesi, Epilessia Mioclonica in Ritardo di sviluppo
Mutazione KBG ANHRD11



Mioclono isolato veglia Mioclono in triplette veglia Mioclonie nella prima fase del sonno NREM fase II

DEVELOPMENTAL AND EPILEPTIC ENCEPHALOPATHIES (DEE)
1) EARLY INFANTILE



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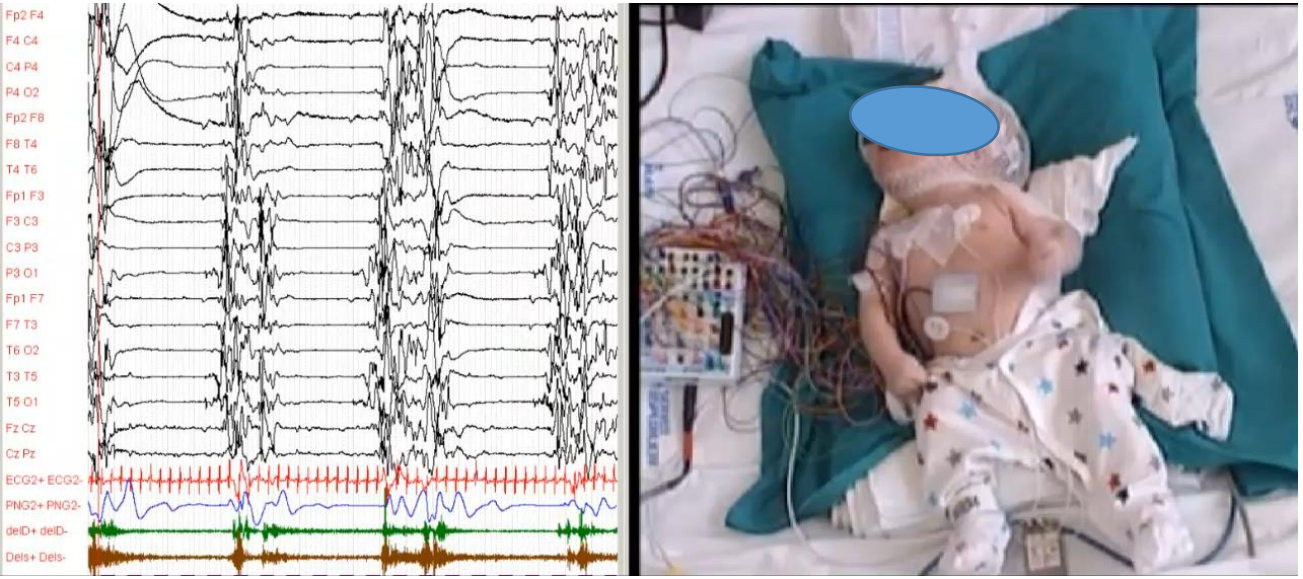
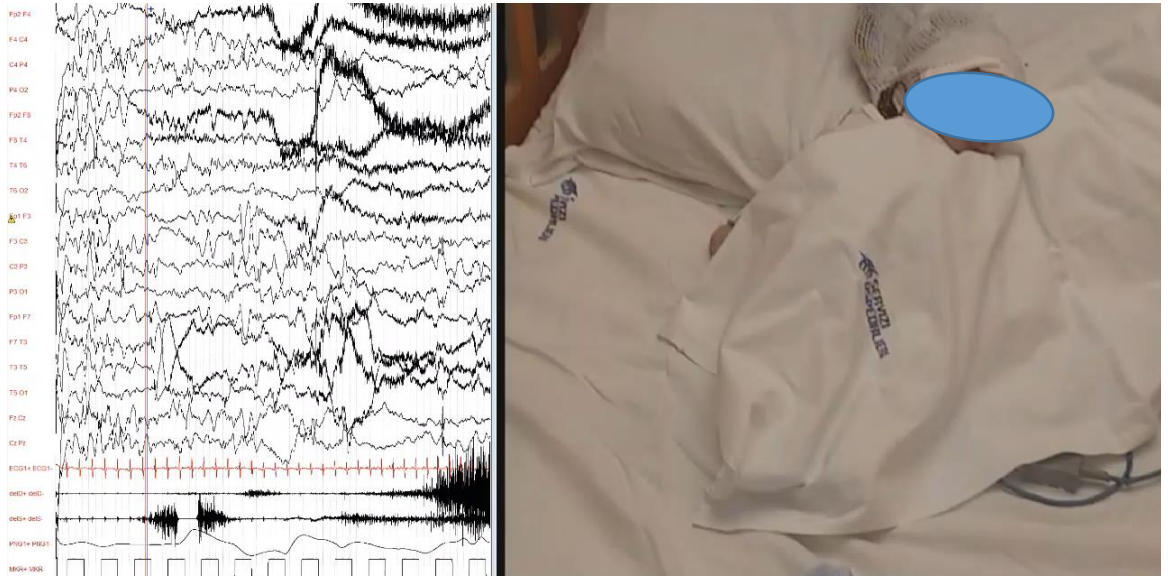
Genetics and Genotype–Phenotype Correlations in Early Onset Epileptic Encephalopathy with Burst Suppression

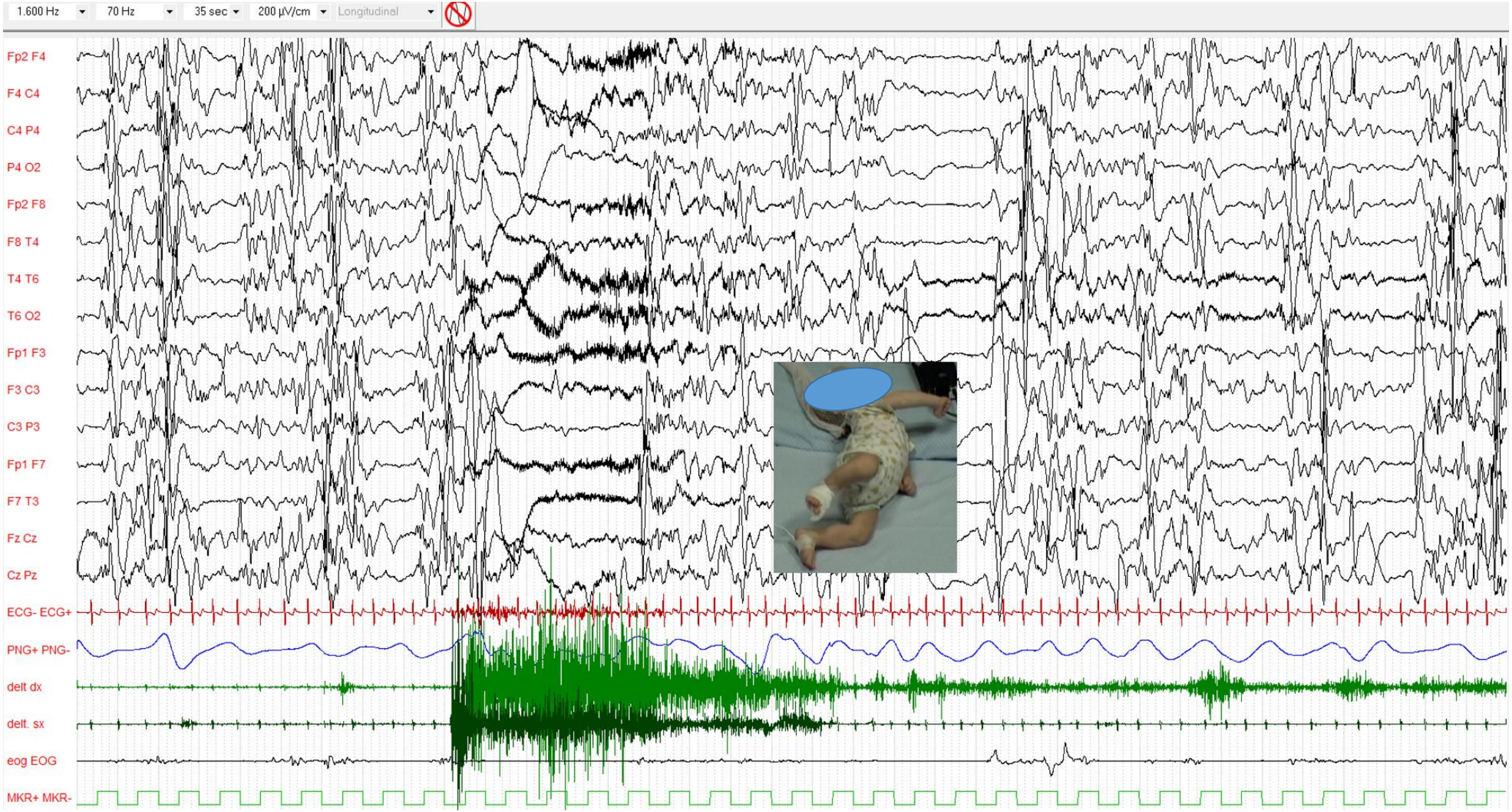
Heather E. Olson, et al., Annals of Neurology

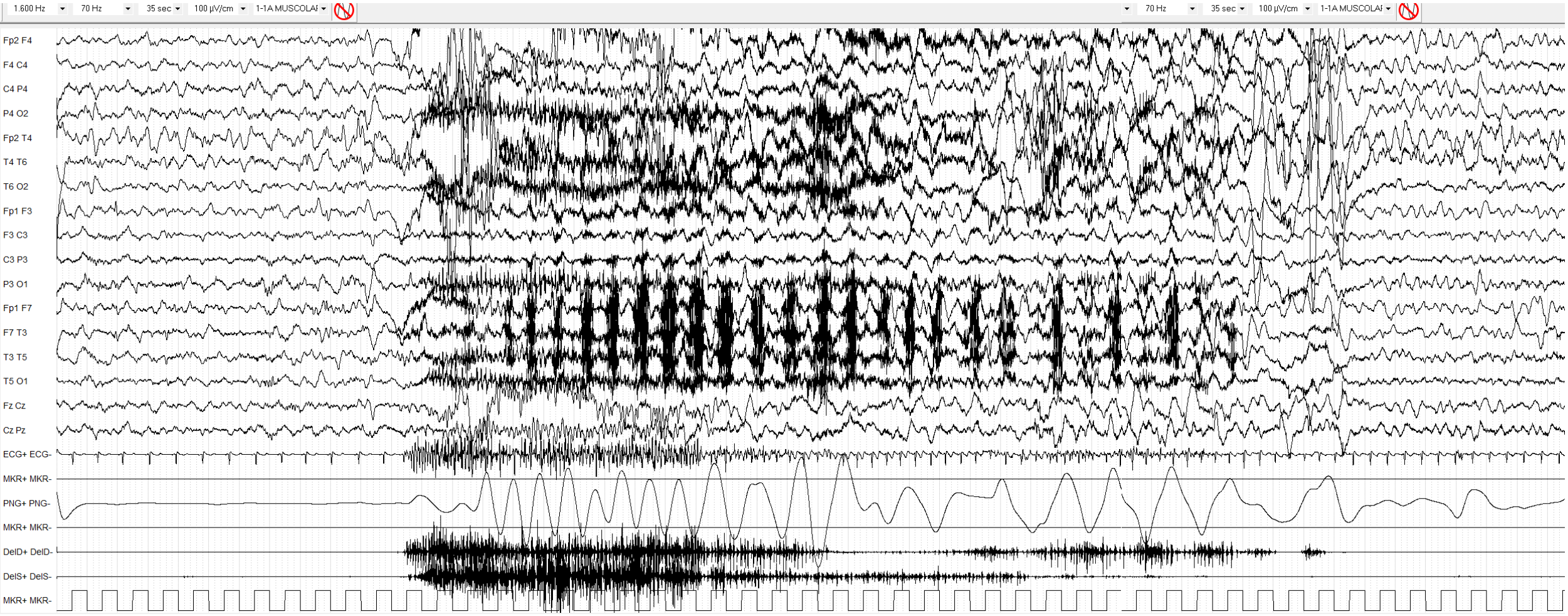
Results: In 17 of 28 (61%) patients with confirmed early burst suppression, we identified variants predicted to be pathogenic in *KCNQ2* (n = 10), *STXBP1* (n = 2), *SCN2A* (n = 2), *PNPO* (n = 1), *PIGA* (n = 1), and *SEPSECS* (n = 1). In 3 of 5 (60%) patients without confirmed early burst suppression, we identified variants predicted to be pathogenic in *STXBP1* (n = 2) and *SCN2A* (n = 1). The patient with the homozygous *PNPO* variant had a low cerebrospinal fluid pyridoxal-5-phosphate level. Otherwise, no early laboratory or clinical features distinguished the cases associated with pathogenic variants in specific genes from each other or from those with no prior genetic cause identified.
Interpretation: We characterize the genetic landscape of epileptic encephalopathy with burst suppression, without brain malformations, and demonstrate feasibility of genetic diagnosis with clinically available testing in >60% of our cohort, with *KCNQ2* implicated in one-third. This electroclinical syndrome is associated with pathogenic variation in *SEPSECS*.

ANN NEUROL 2017;81:419–429





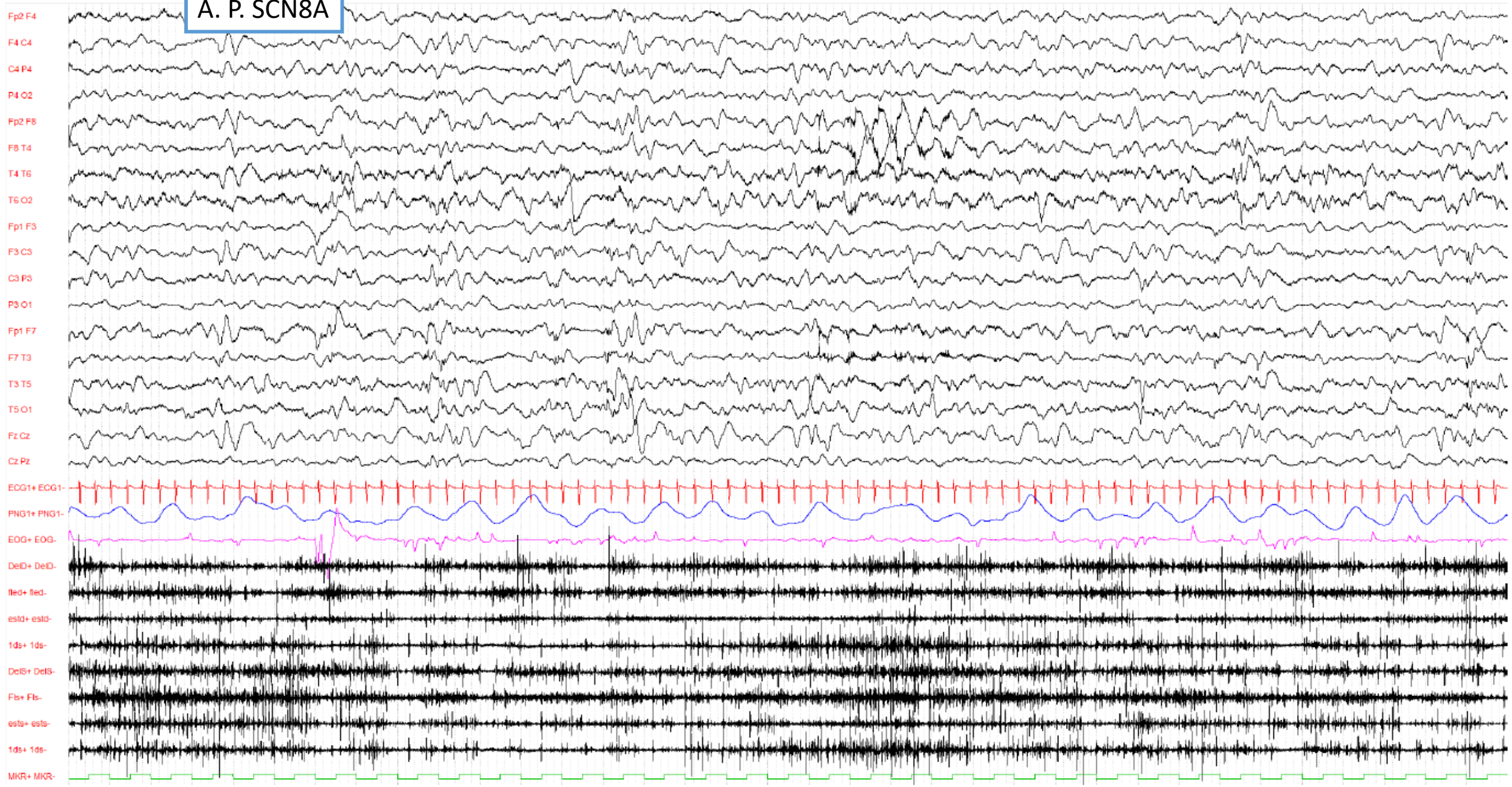

1.600 Hz
70 Hz
35 sec
200 µV/cm
Longitudinal
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
Fp1 F7
F7 T3
Fz Cz
Cz Pz
ECG- ECG+
PNG+ PNG-
delt dx
delt. sx
eog EOG
MKR+ MKR-


1.600 Hz
70 Hz
35 sec
100 µV/cm
1-1A MUSCOLAI
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
MKR+ MKR-
PNG+ PNG-
MKR+ MKR-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-

STXBP1

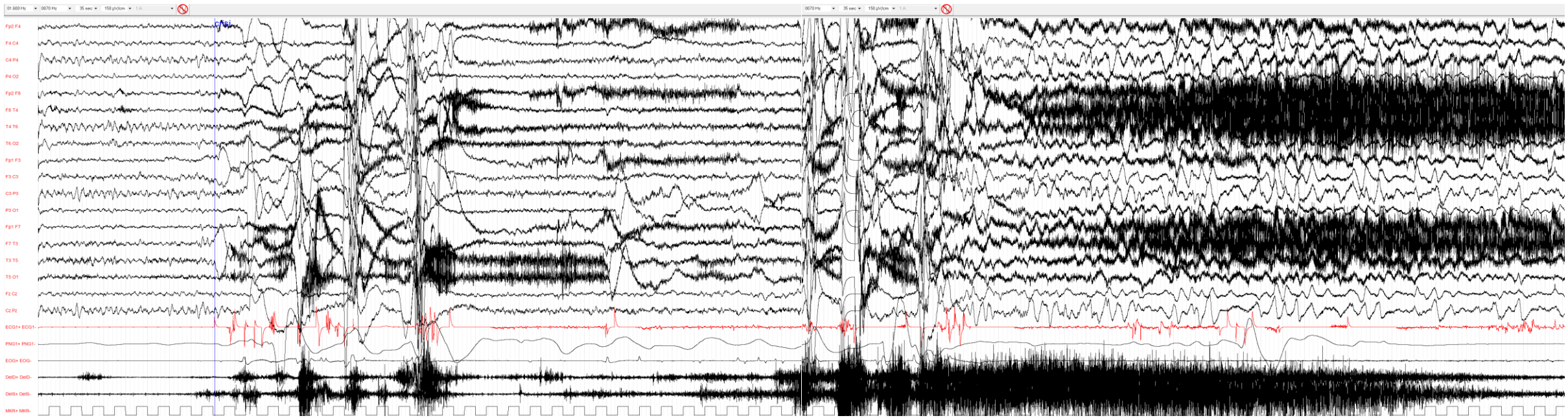
Giu Jac 2 mesi esordio epilessia mutazione TBD1D24

A. P. SCN8A

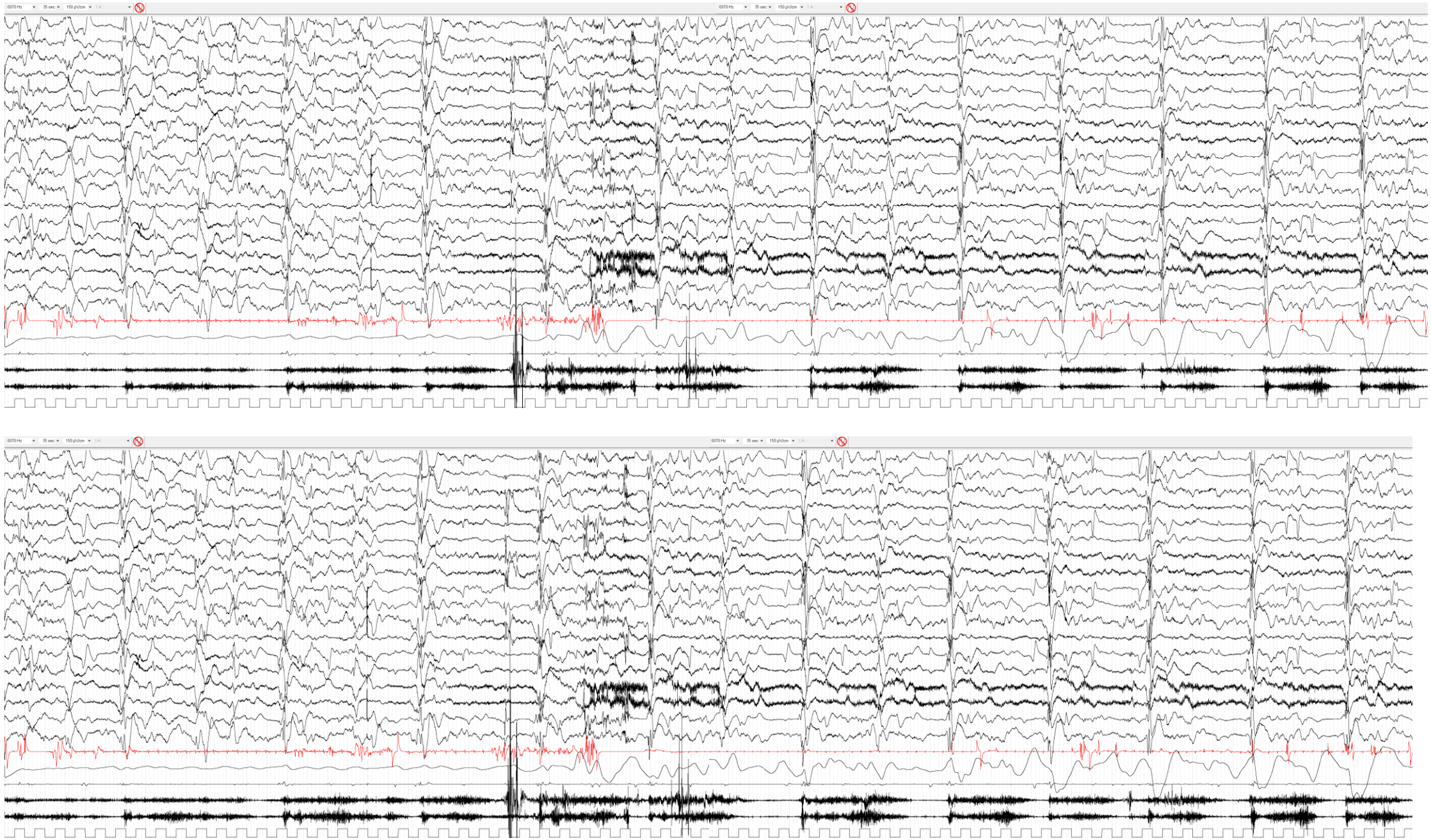


1- veglia mioclono parcellare multifocale a alta frequenza

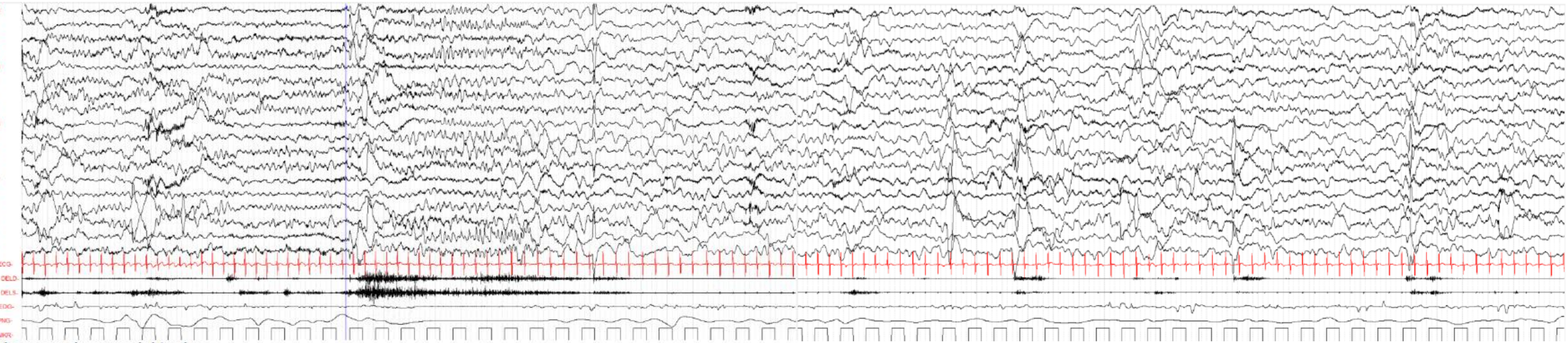
WFDM CDKL5


Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
PNG1+ PNG1-
EOG+ EOG-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-

WFDM CDKL5



LomRidLu CDKL5

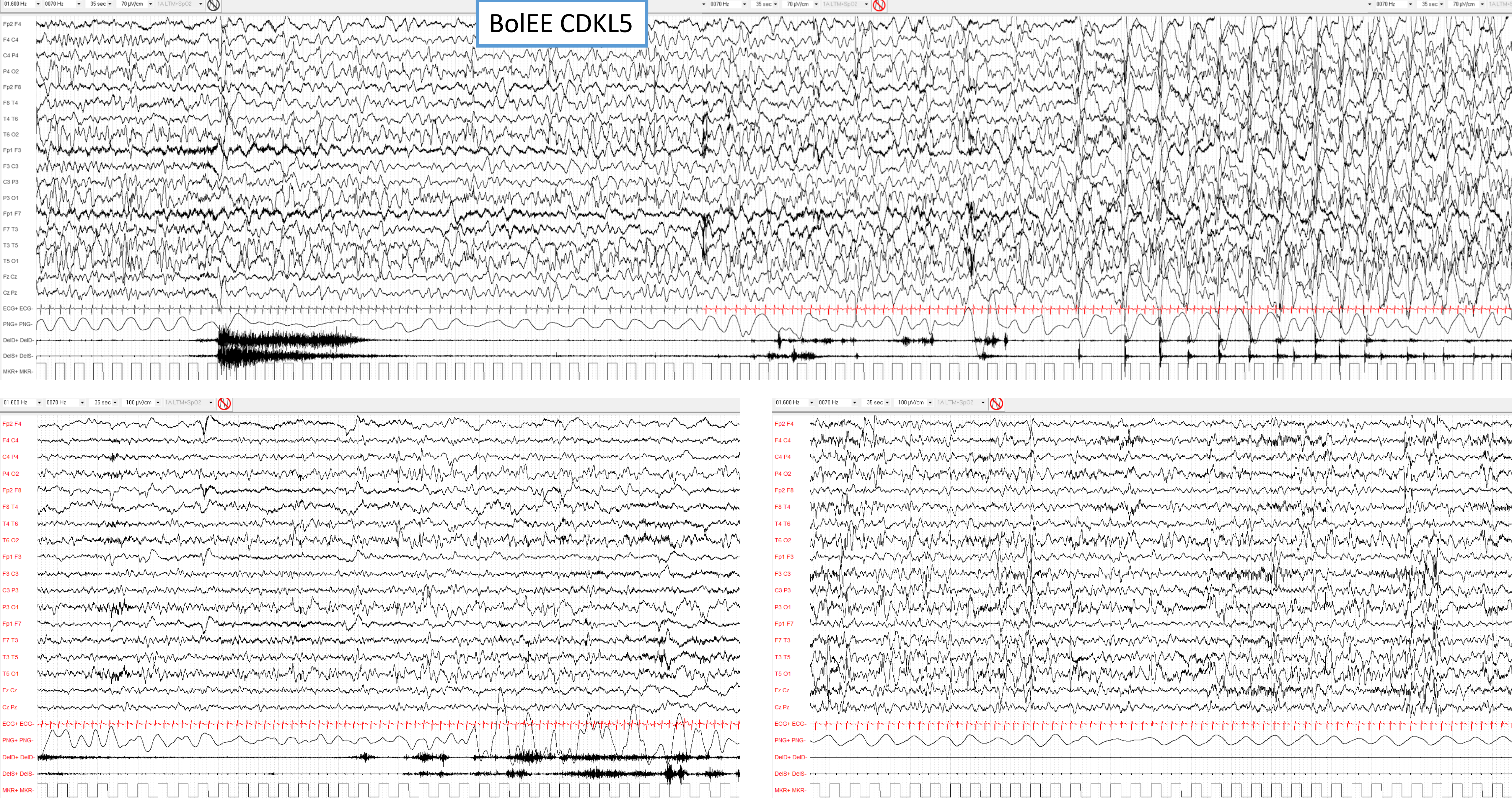


zio grappolo con crisi tonica.

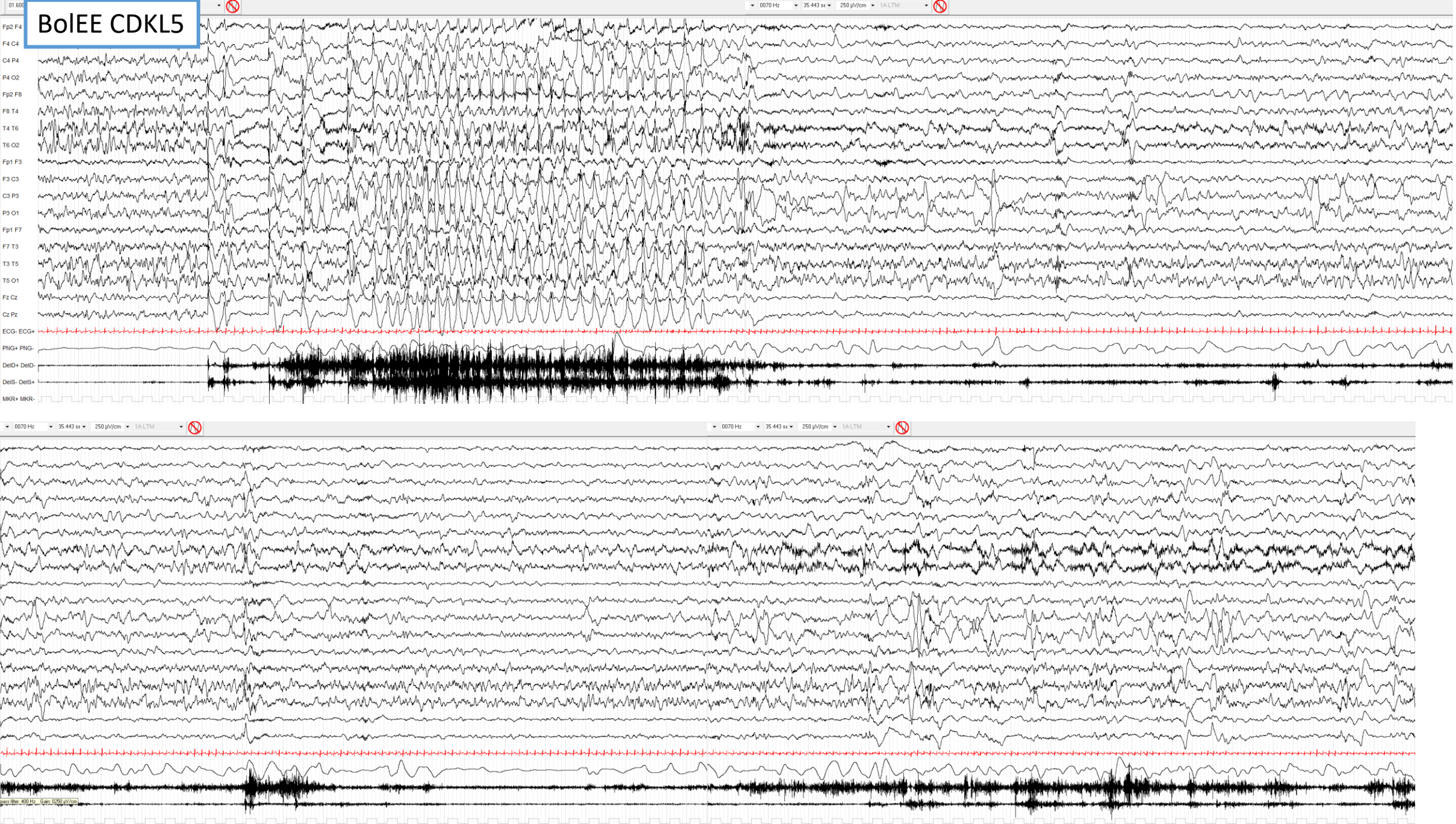
Seguono micolono-spasmi peridici.

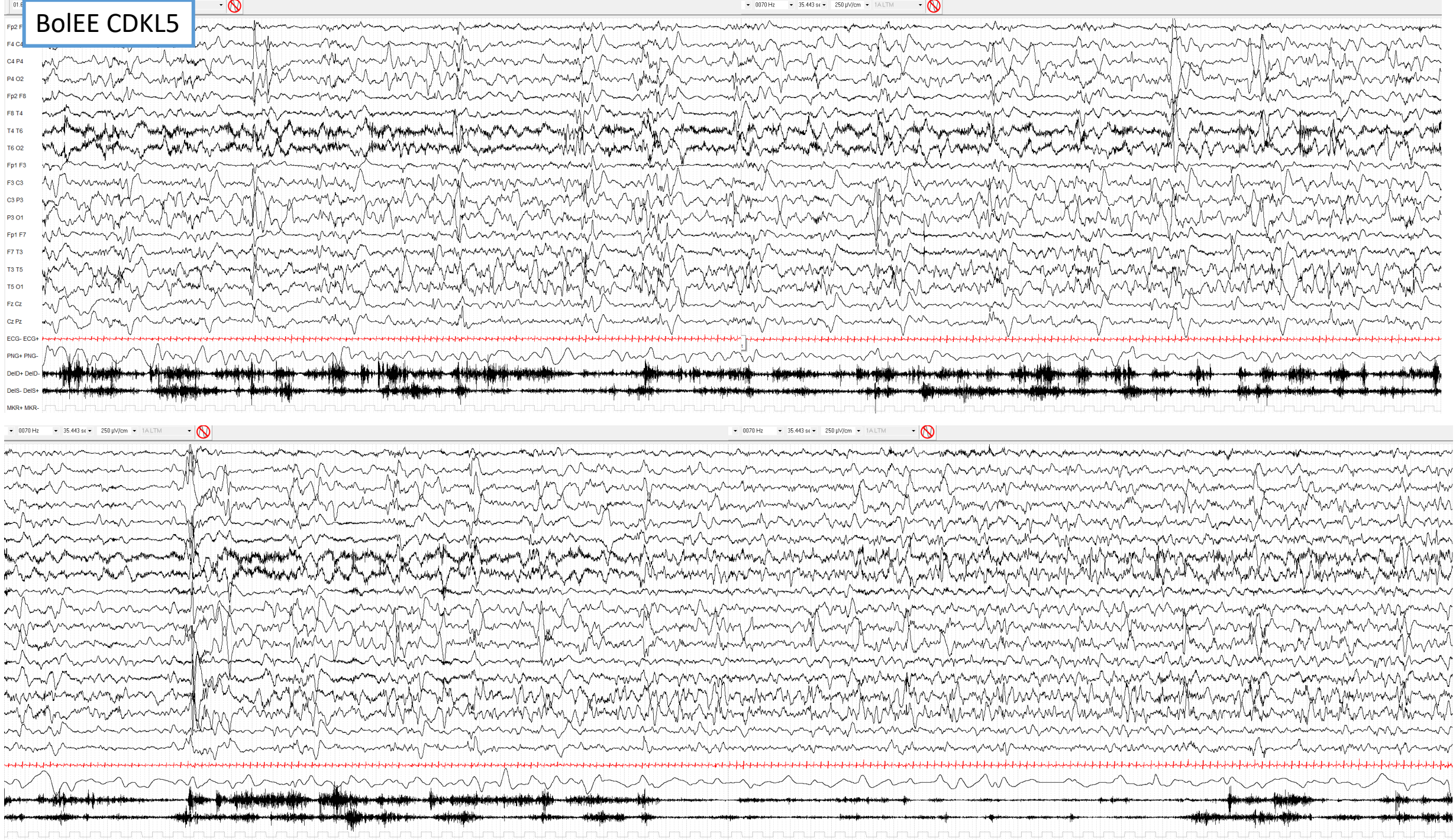
Ancora.

BolEE CDKL5

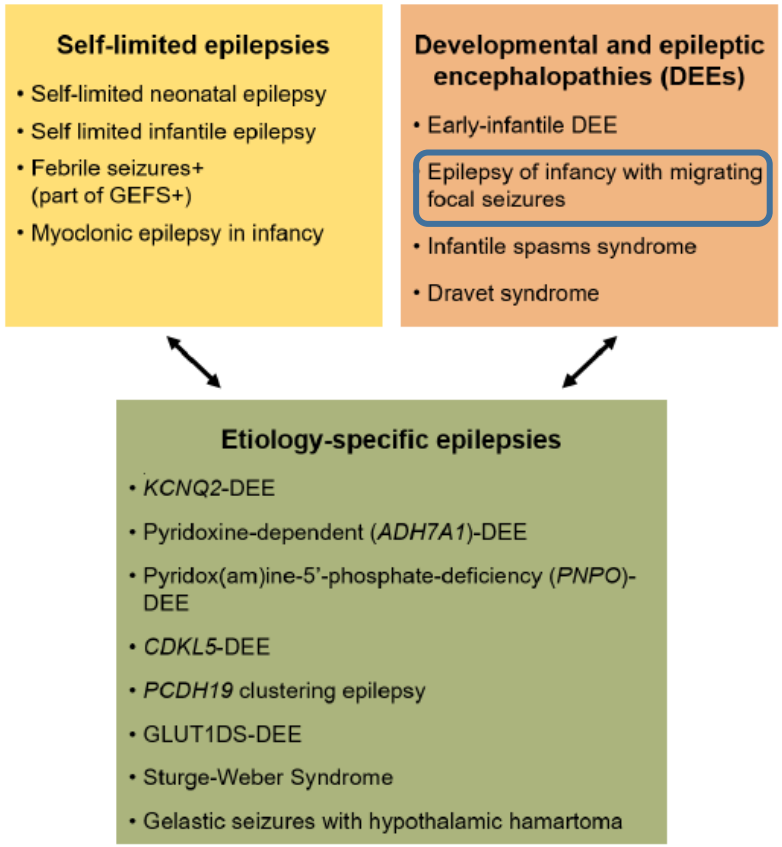

01.600 Hz
0070 Hz
35 sec
70 µV/cm
100 µV/cm
1A LTM+SpO2
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG+ ECG-
PNG+ PNG-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-

BolEE CDKL5


Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG- ECG+
PNG+ PNG-
DelD+ DelD-
DelS- DelS+
MKR+ MKR-


BolEE CDKL5
0070 Hz
35.443 se
250 µV/cm
1A LTM
Fp2 F
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG- ECG+
PNG+ PNG-
DelD+ DelD-
DelS- DelS+
MKR+ MKR-

DEVELOPMENTAL AND EPILEPTIC ENCEPHALOPATHIES (DEE)
2) MIGRATING FOCAL SEIZURES



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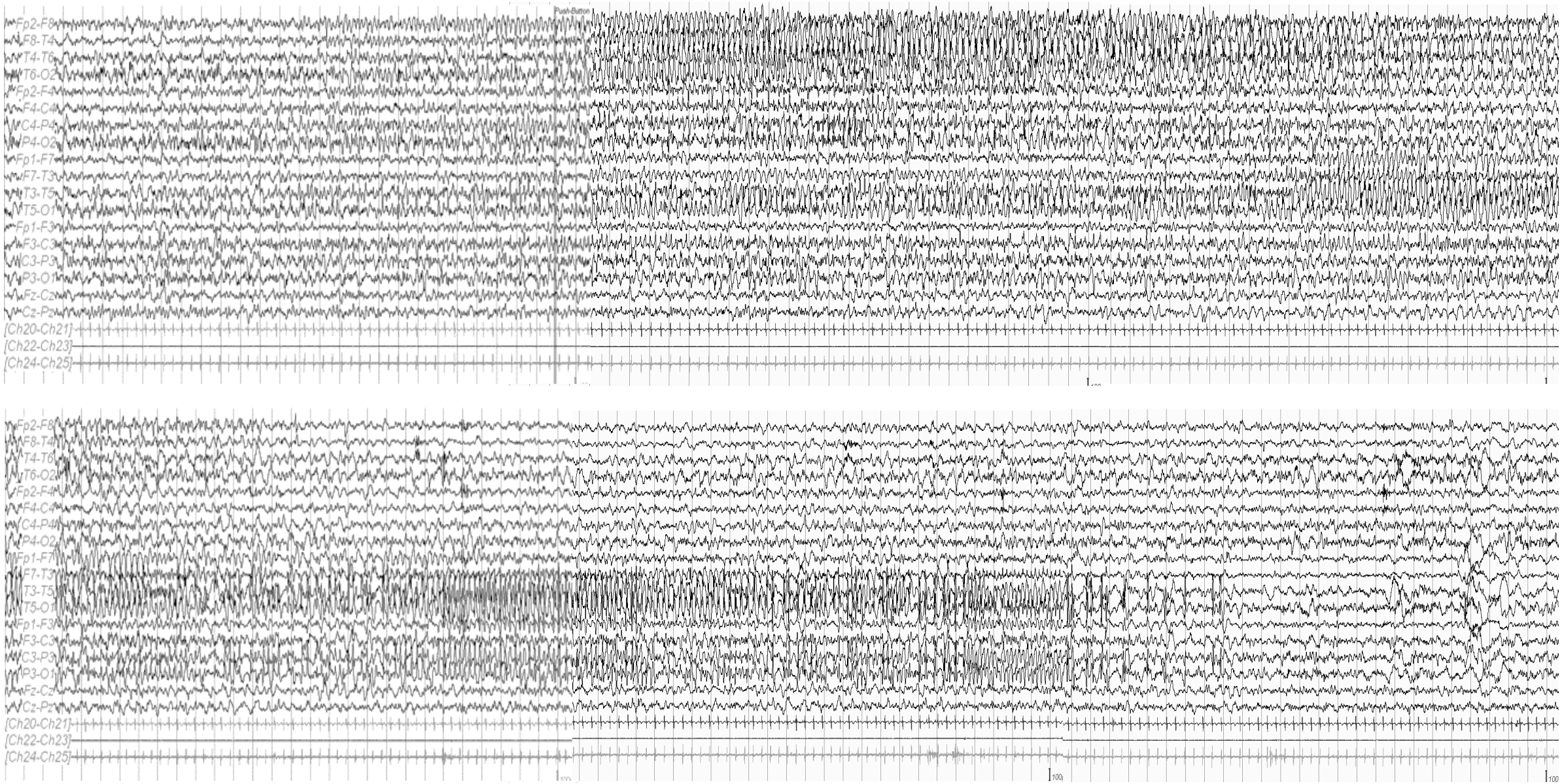
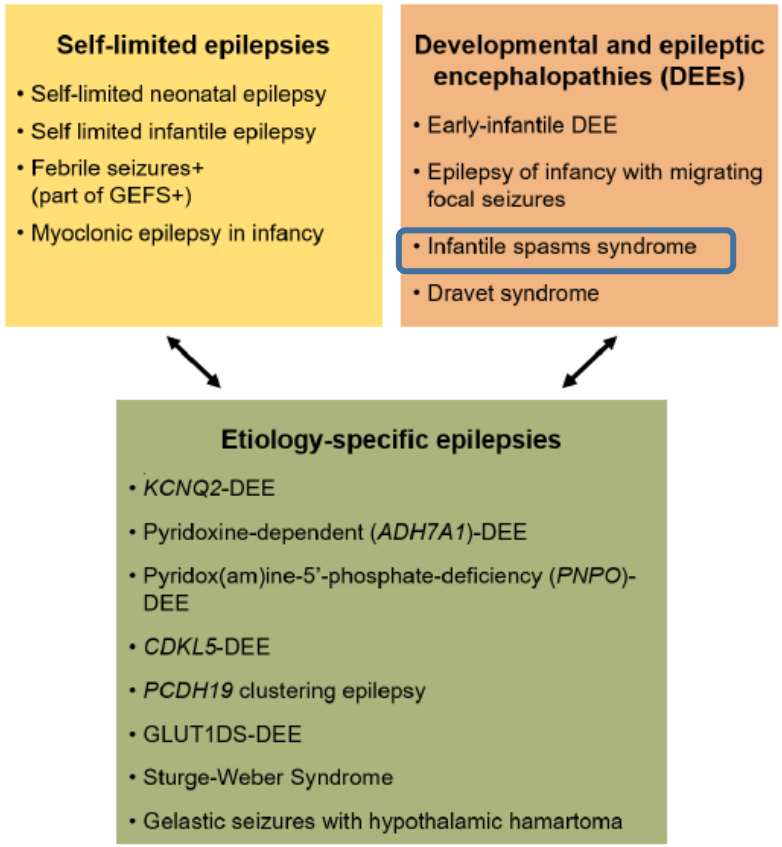
1.600 Hz
70 Hz
35 sec
150 µV/cm
2-1A+PNG+EOG
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG1+ ECG1-
EOG+ EOG-
DELD+ DELD-
DELS+ DELS-
PNG1+ PNG1-
MKR+ MKR-

MPSI - Mir.Lat., 4 months, focal seizure, minimal signs



DEVELOPMENTAL AND EPILEPTIC ENCEPHALOPATHIES (DEE)
3) INFANTILE SPASMS SYNDROME



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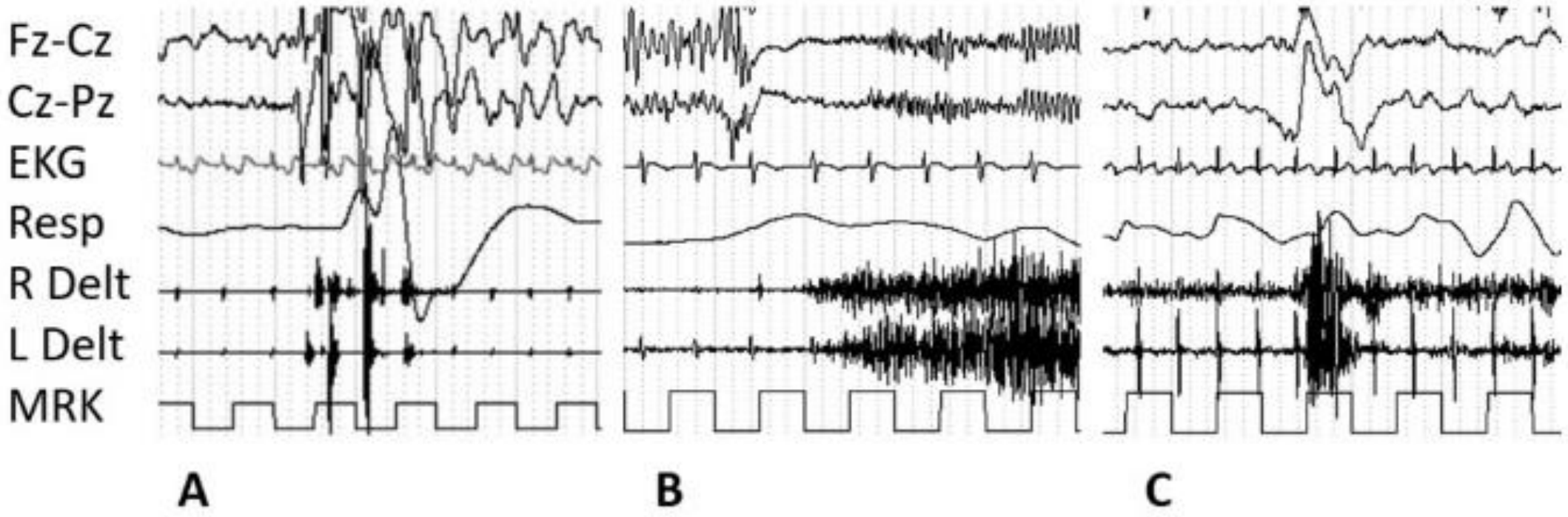


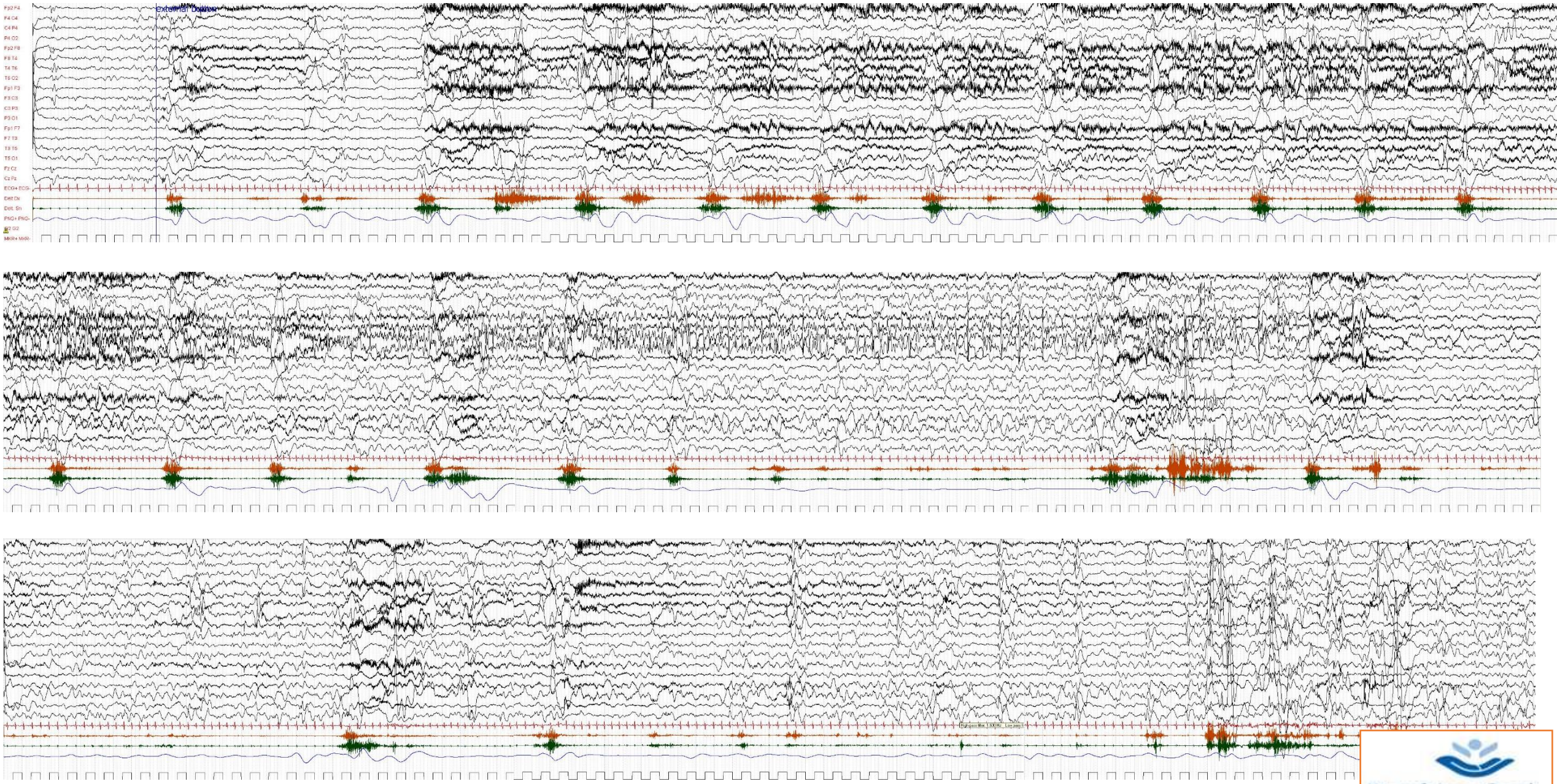
Mioclonia massiva

Crisi tonica

Spasmo Epilettico

Fusco et al, Epilepsy and Behavior 2020

Modified from Epilepsia 1993, Fusco e Vigevano

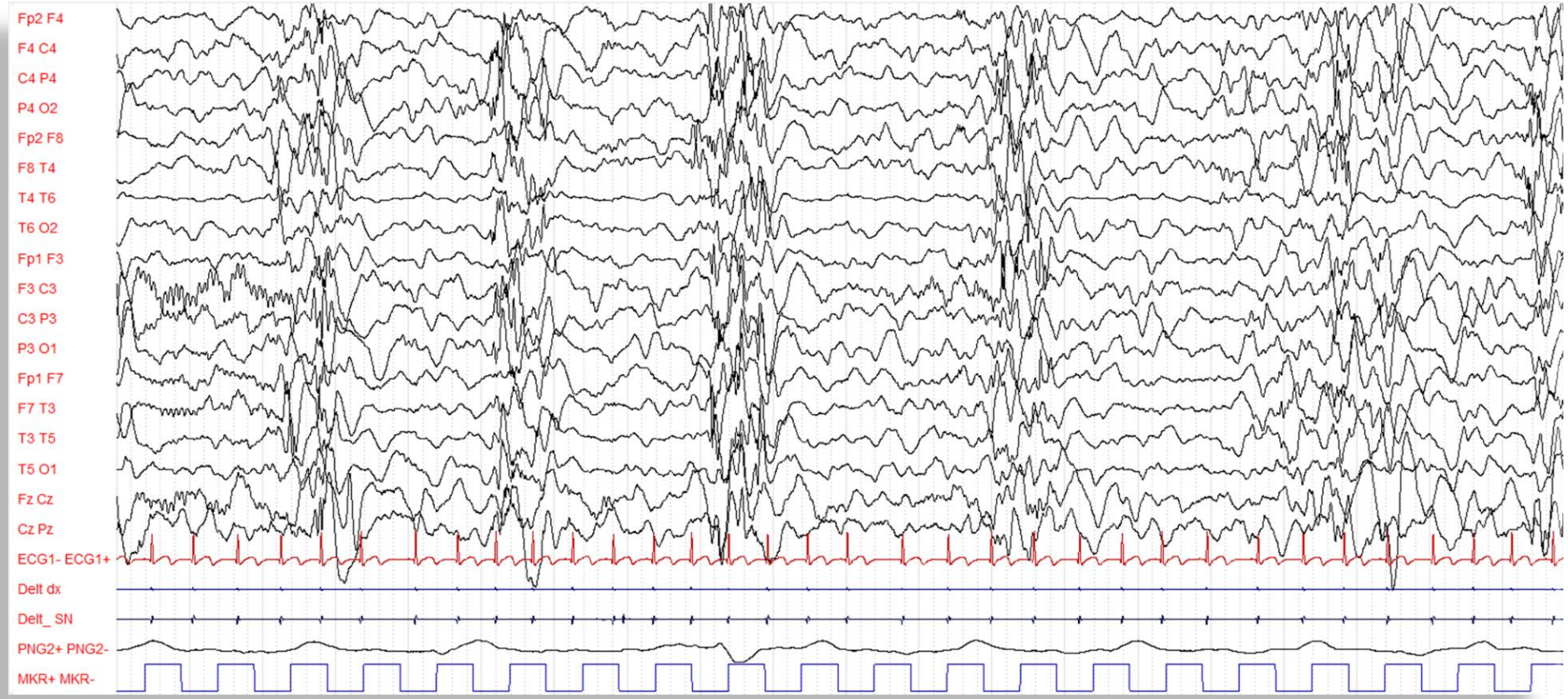


Bambino Gesù
OSPEDALE PEDIATRICO

INTERICTAL EEG

Bambino Gesù
OSPEDALE PEDIATRICO

During sleep: alternating paroxysmal pattern



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Ictal EEG patterns

- Slow wave
- Sharp wave
- Attenuation
- Fast activity

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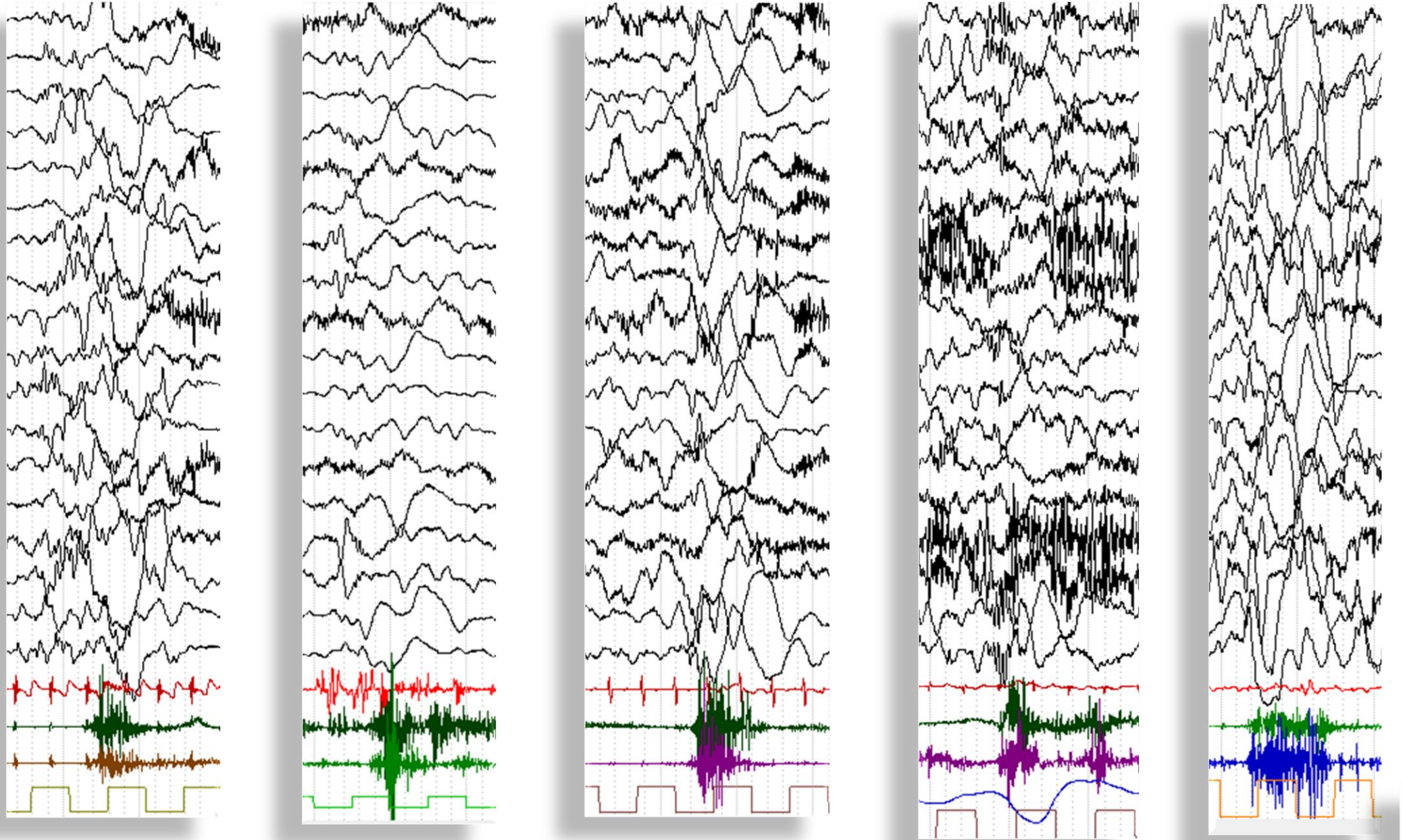
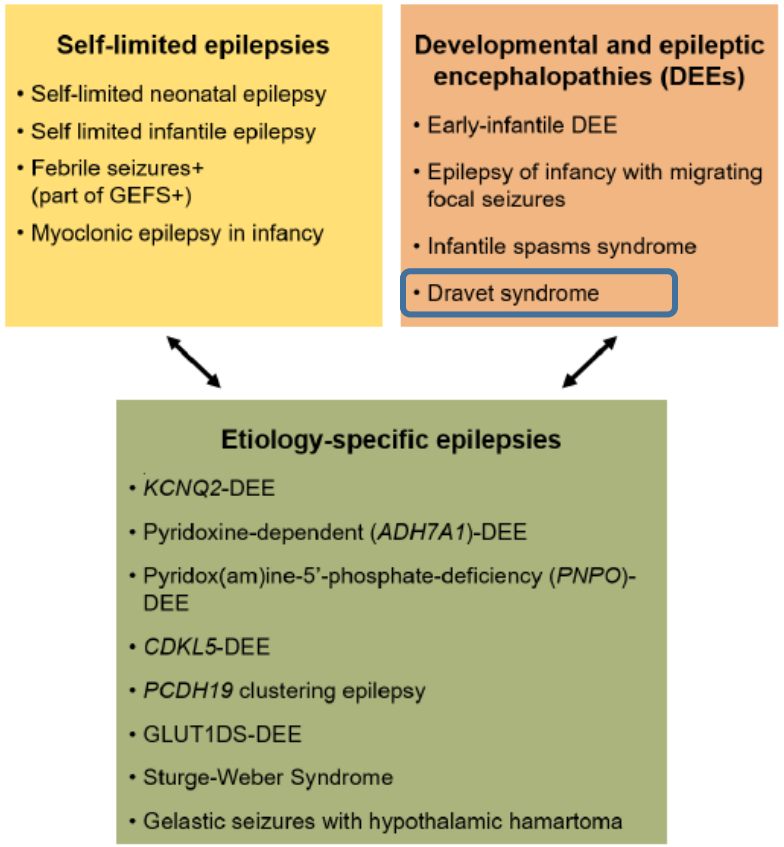
OSPEDALE PEDIATRICO

Pattern ictale dello spasmo epilettico



Bambino Gesù
OSPEDALE PEDIATRICO

DEVELOPMENTAL AND EPILEPTIC ENCEPHALOPATHIES (DEE)
4) DRAVET SYNDROME



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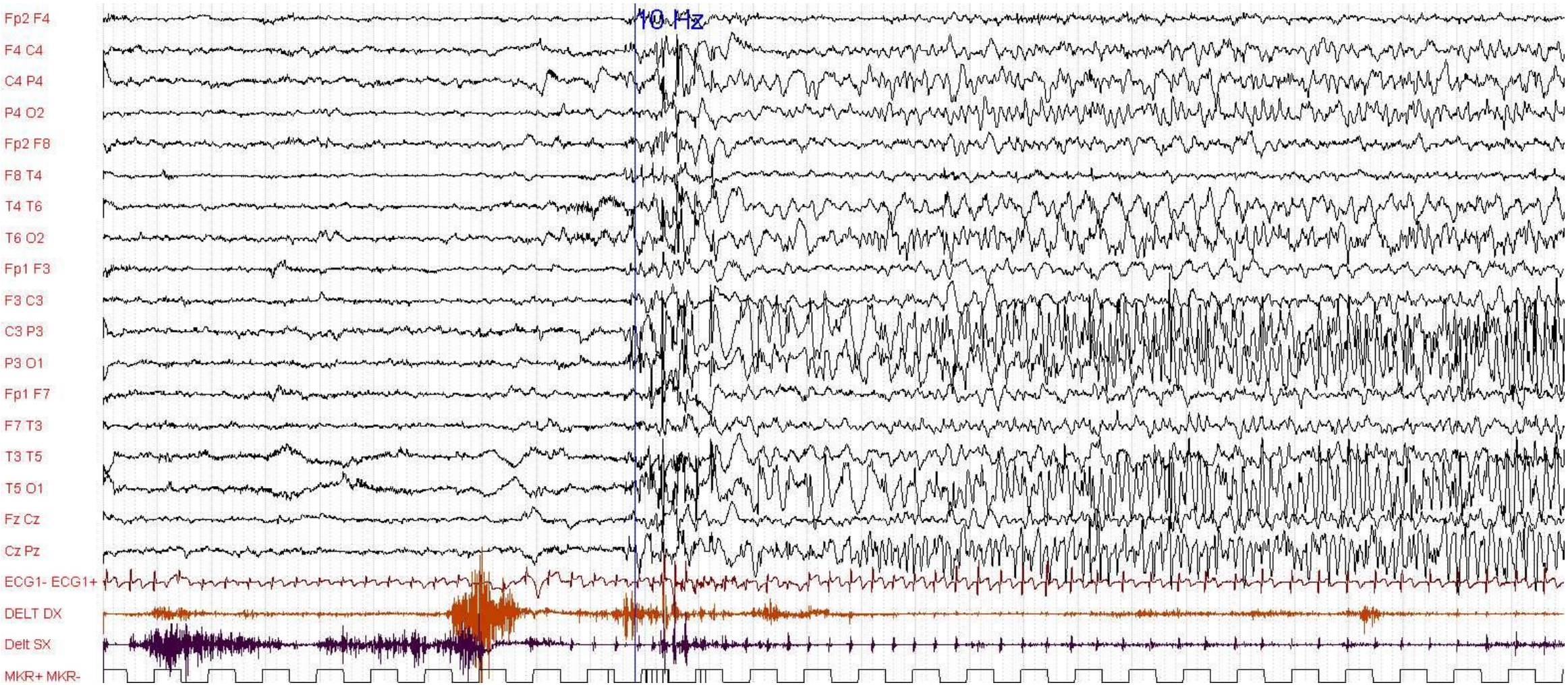
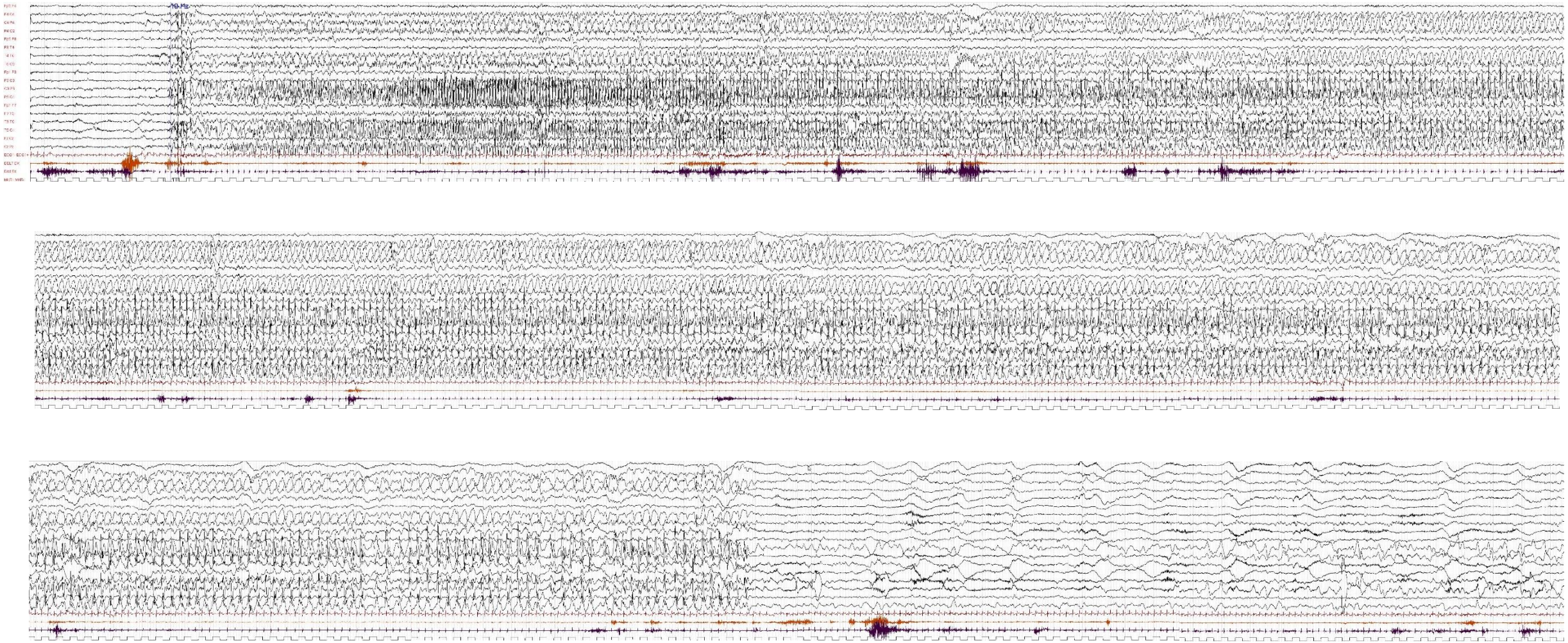
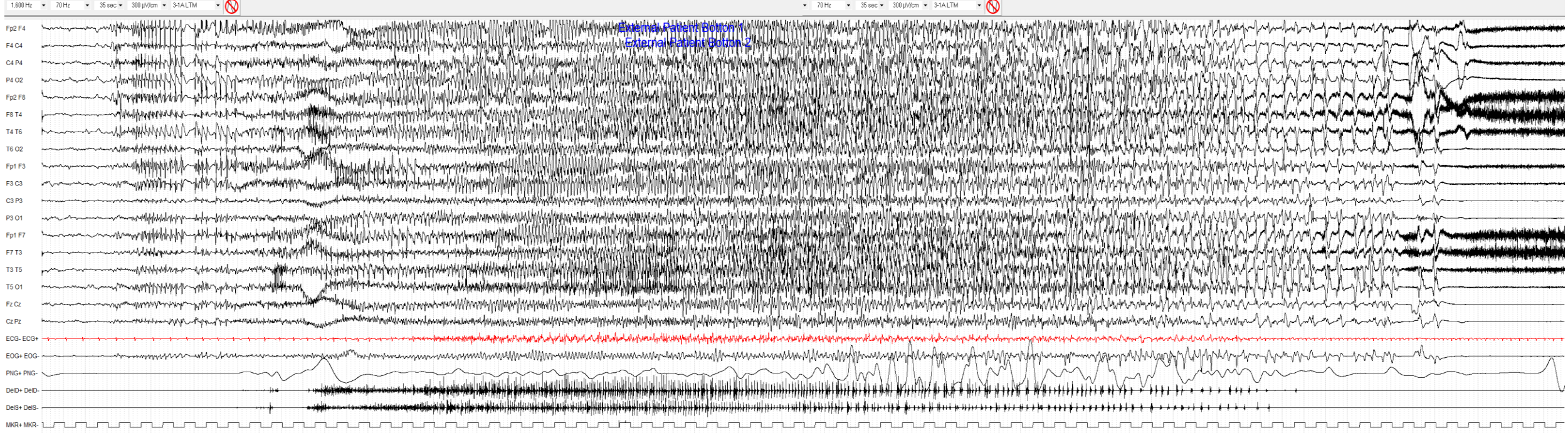
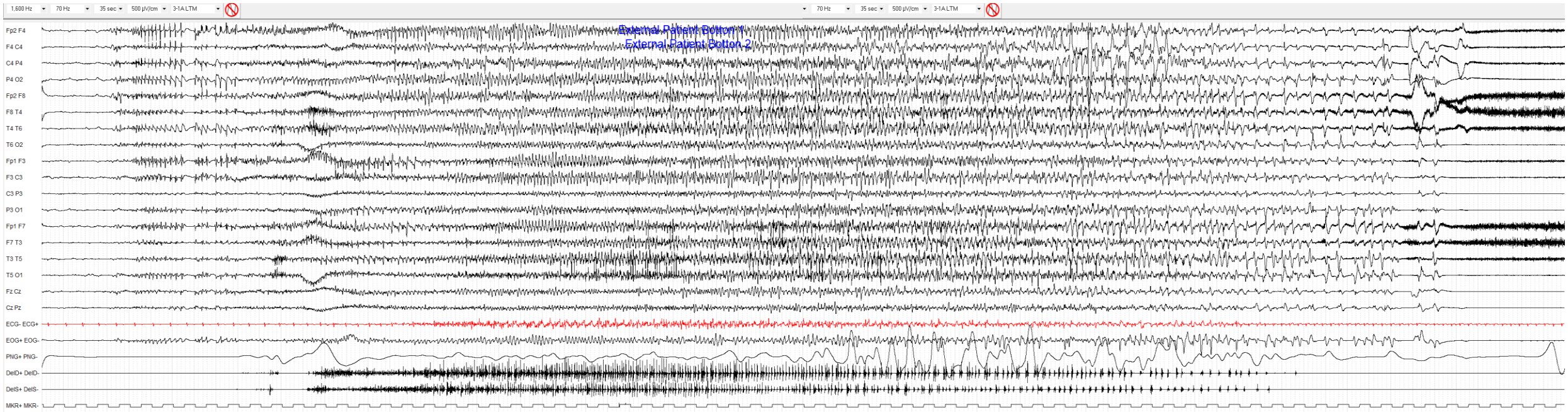
Mor Sar 14 mesi



Mor Ser 14 mesi durata crisi 5minuti dopo sli




1,600 Hz
70 Hz
35 sec
300 µV/cm
3-1A LTM
External Patient Button 1
External Patient Button 2
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG- ECG+
EOG+ EOG-
PNG+ PNG-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-


1,600 Hz
70 Hz
35 sec
500 µV/cm
3-1A LTM
External Patient Button 1
External Patient Button 2
Fp2 F4
F4 C4
C4 P4
P4 O2
Fp2 F8
F8 T4
T4 T6
T6 O2
Fp1 F3
F3 C3
C3 P3
P3 O1
Fp1 F7
F7 T3
T3 T5
T5 O1
Fz Cz
Cz Pz
ECG- ECG+
EOG+ EOG-
PNG+ PNG-
DelD+ DelD-
DelS+ DelS-
MKR+ MKR-

lucia.fusco@opbg.net

Corso di Perfezionamento in EEG pediatrico
Università di Tor Vergata – OPBG (20 CFU)
AA 2021-2022
Scadenza bando 15/01/2022
www.scuoladipediatria.it
lucia.fusco@opbg.net

Children'Hospital Bambino Gesù
Rome