EEG in Epileptic Syndrome

Surachai Likasitwattanakul, M.D.
Division of Neurology, Department of Pediatrics
Faculty of Medicine, Siriraj Hospital
Mahidol University

Epileptic syndrome

- Electroclinical syndrome
  - a complex of clinical features, signs, and symptoms that together define a distinctive, recognizable clinical disorder
- Constellation
  - are not exactly electroclinical syndromes in the same sense but which represent clinically distinctive constellations on the basis of specific lesions or other causes.
- Structural/metabolic epilepsies
  - secondary to specific structural or metabolic lesions or conditions but which do not, given our current understanding, fit a specific electroclinical patter
- Epilepsies with unknown causes
  - which in the past were termed ‘‘cryptogenic,’’ will now be referred to as being of ‘‘unknown’’ cause.

Berg, 2010

Electroclinical syndrome

- A complex of clinical features, signs, and symptoms that together define a distinctive, recognizable clinical disorder.
- These often become the focus of treatment trials as well as of genetic, neuropsychological, and neuroimaging investigations
- These are distinctive disorders identifiable on the basis of a typical age onset, specific EEG characteristics, seizure types, and often other features which, when taken together, permit a specific diagnosis

Berg, 2010

Benefit of epileptic syndrome diagnosis

- Rational decision making
  - Choice of investigation
  - Choice of treatment
    - Treat: with certain AED
    - Not treat: with certain reasons
  - Avoid certain precipitants
  - Prognostic information to an individual

Electroclinical syndromes arranged by age at onset

Neonatal period

- Benign familial neonatal epilepsy (BFNE)
- Early myoclonic encephalopathy (EME)
- Ohtahara syndrome

Infancy

- Epilepsy of infancy with migrating focal seizures
- West syndrome
- Myoclonic epilepsy in infancy (MEI)
- Benign infantile epilepsy
- Benign familial infantile epilepsy
- Dravet syndrome
- Myoclonic encephalopathy in nonprogressive disorders

Electroclinical syndromes arranged by age at onset

Childhood

- Febrile seizures plus (FS+)
- Panayiotopoulos syndrome
- Epilepsy with myoclonic atonic (previously astatic) seizures
- Benign epilepsy with centrotemporal spikes (BECTS)
- Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)
- Late onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences
- Lennox-Gastaut syndrome
- Epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS)
- Landau-Kleffner syndrome (LKS)
- Childhood absence epilepsy (CAE)

Electroclinical syndromes arranged by age at onset

Adolescence – Adult

- Juvenile absence epilepsy (JAE)
- Juvenile myoclonic epilepsy (JME)
- Epilepsy with generalized tonic–clonic seizures alone
- Progressive myoclonus epilepsies (PME)
- Autosomal dominant epilepsy with auditory features (ADEAF)
- Other familial temporal lobe epilepsies

Less specific age relationship

- Familial focal epilepsy with variable foci (childhood to adult)
- Reflex epilepsies

Role of EEG in epileptic syndrome

- Identifying of epileptic syndrome is important for appropriate management of epilepsy
- Diagnosis of epileptic syndrome is not solely based on EEG since it is only one of the integral part of epileptic syndrome
- However, EEG is still an important tool to diagnose epileptic syndrome especially in patients who have complex or inadequate clinical information

Practical points

- Attending physicians : physicians who order and take care of a patient
- Electroencephalographers : physicians who read EEG and interpret it in the given context
- In the situation whenever appropriate, the EEGers should provide a suggestion to the clinicians to aware of certain syndrome (again based on clinical information provided by attending physicians)

Case

- A 9-year-old girl presents with staring episodes lasting, per mother, a few minutes.
- She is diagnosed with complex partial seizure and prescribed with carbamazepine without benefit.
- She is then referred for appropriate treatment

Case

- A 14-year-old girl presented with frequent generalized tonic-clonic seizure.
- EEG shows polyspike-wave complex.
- Phenobarbital, as well as Phenytoin could not control the seizures.
- EEG as followed

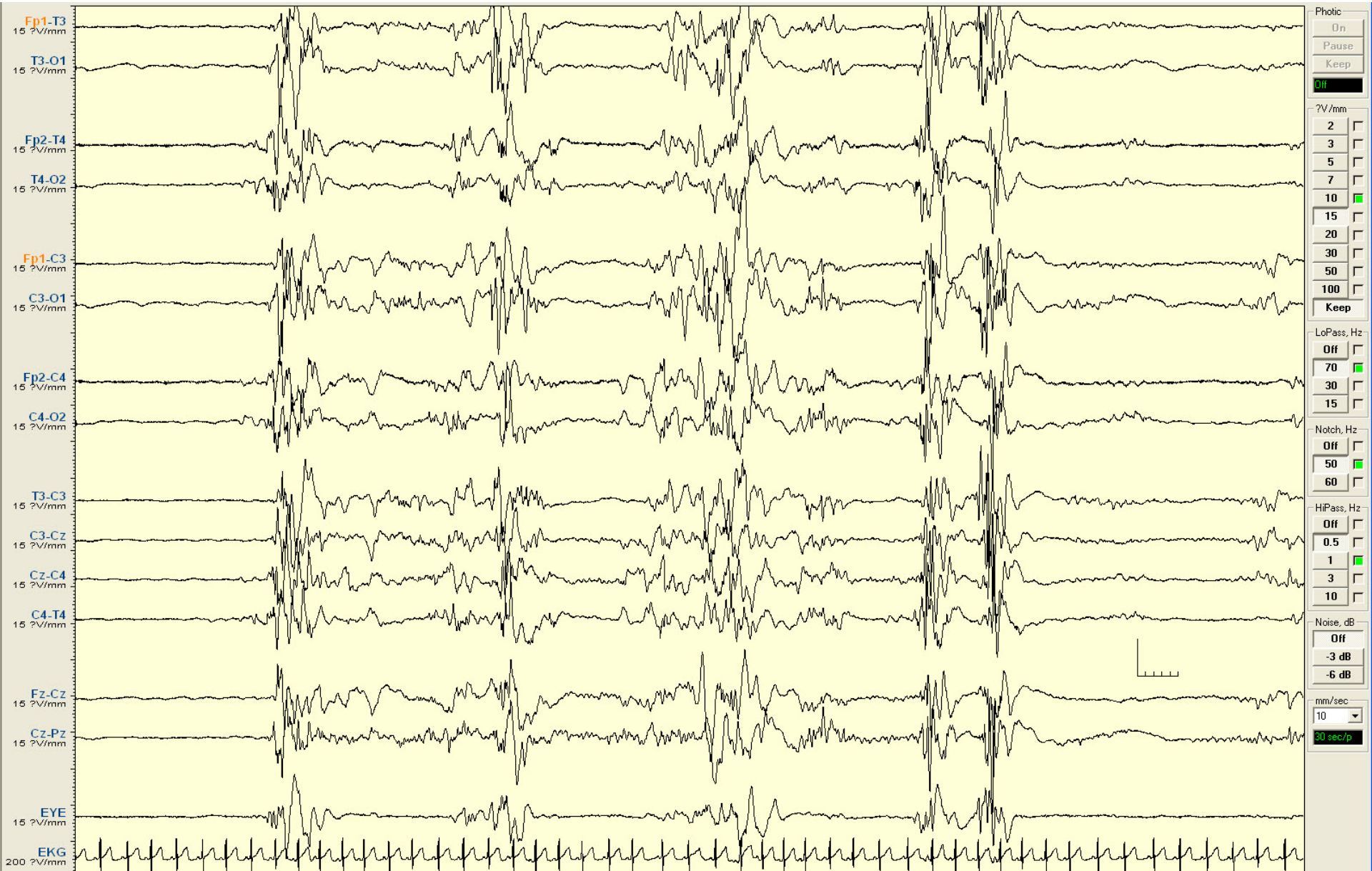
case

- A 9-year-old boy with seizure described, per mom, as generalized tonic-clonic seizure lasting 1-2 minutes
- This is his second seizures in 6 months.
- Normal examination
- EEG as followed

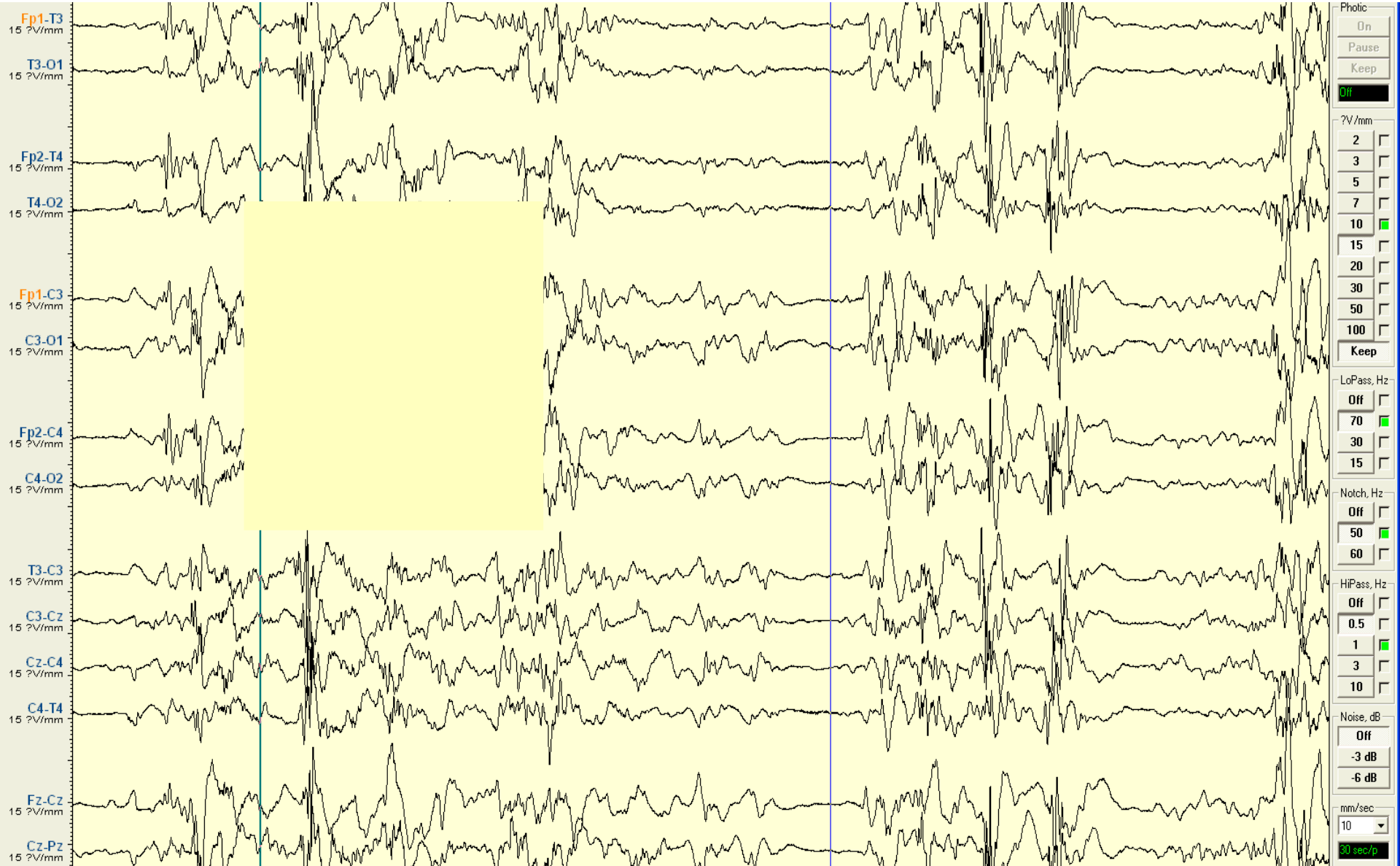
Otahara Syndrome

- Onset : Mainly in 1 month of life
- Seizure type : Tonic spasms
- Interictal EEG : Burst-suppression
- Ictal EEG : Diffuse synchronization, high amplitude slow wave or fast activity
- Causes : Structural brain lesions
- Course : Severe, progressive course
- Treatment : Difficult to treat

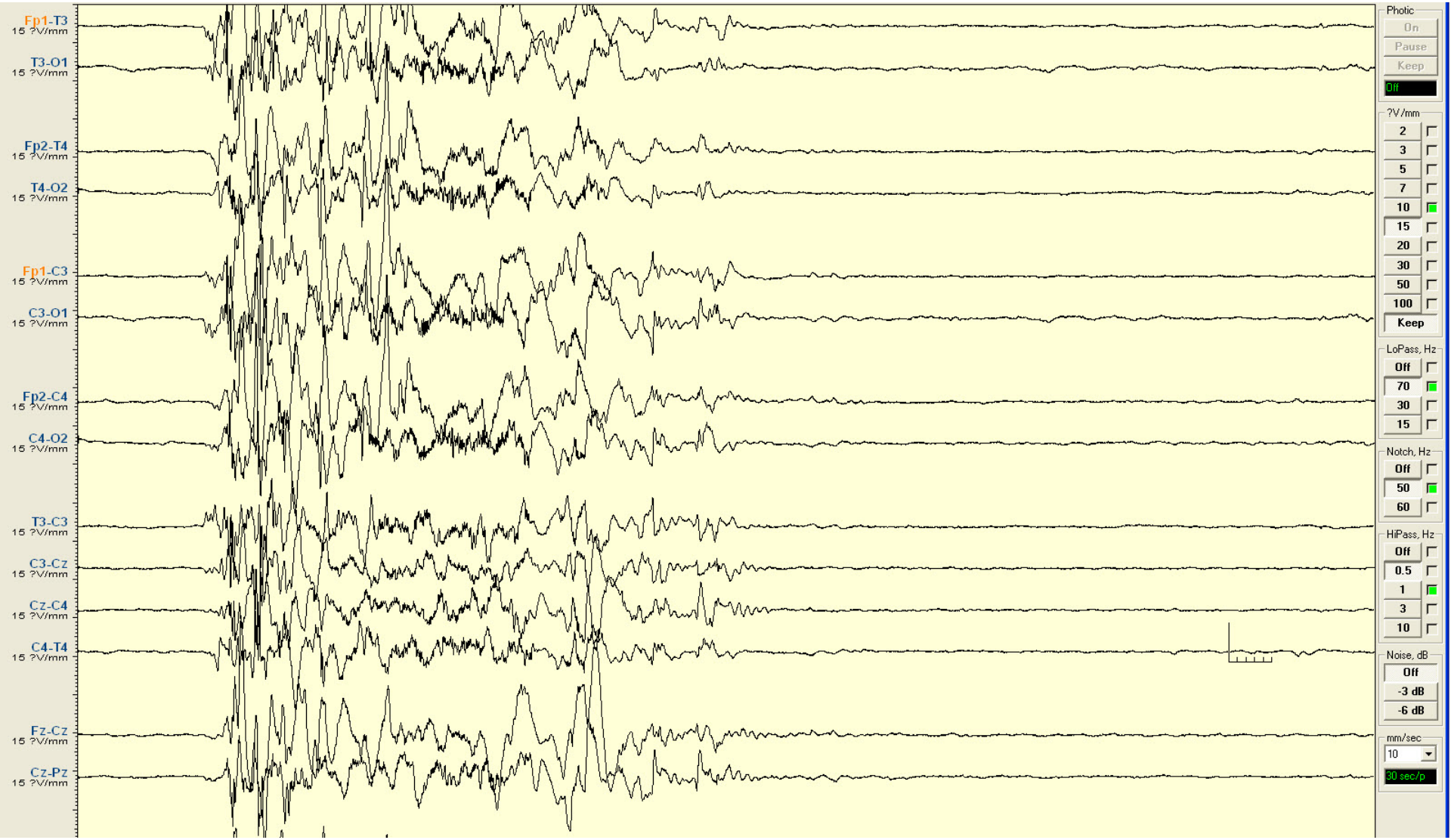
Burst-suppression in 30 sec/page



Ictal of tonic spasm



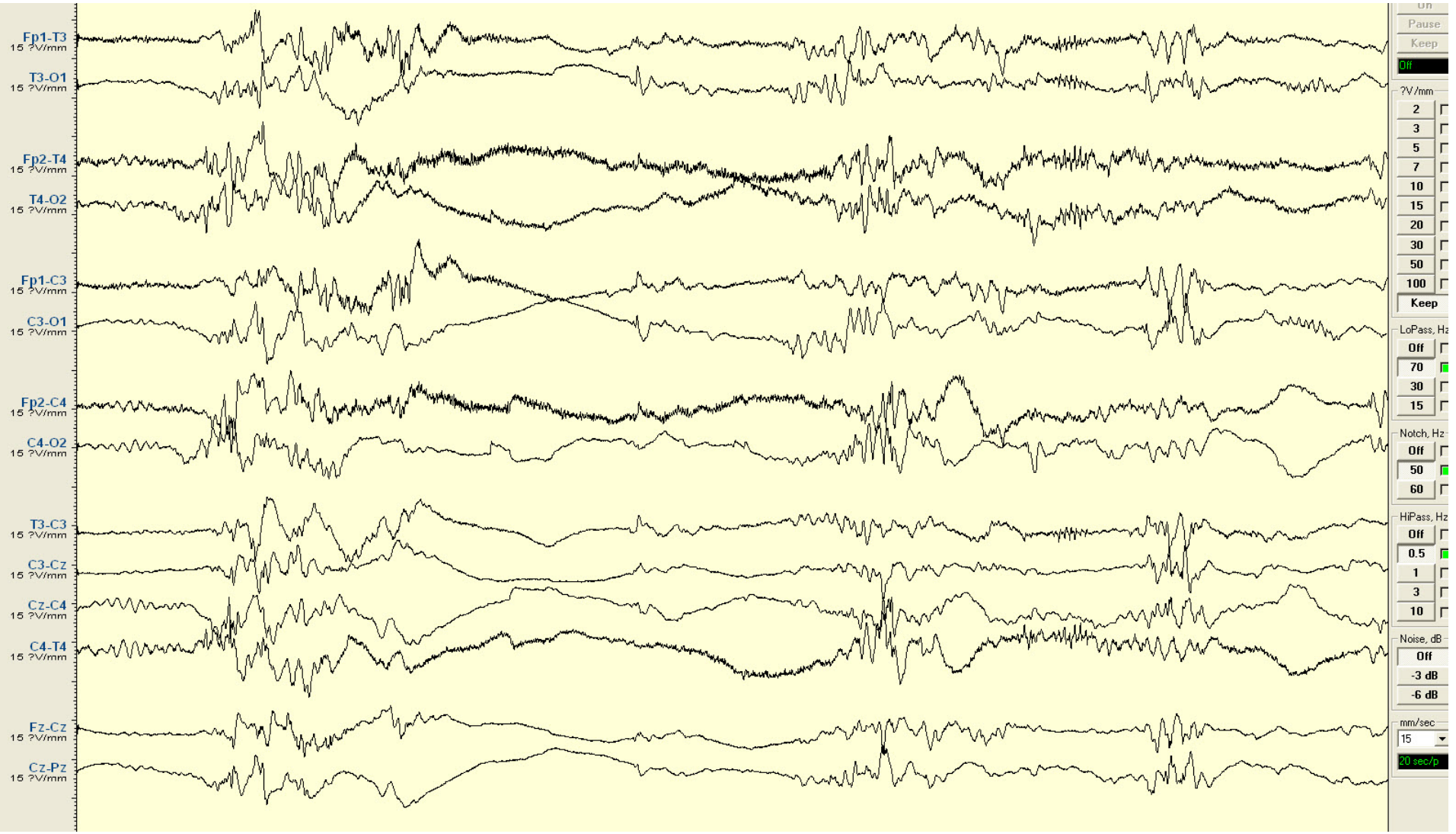
Long burst and suppression



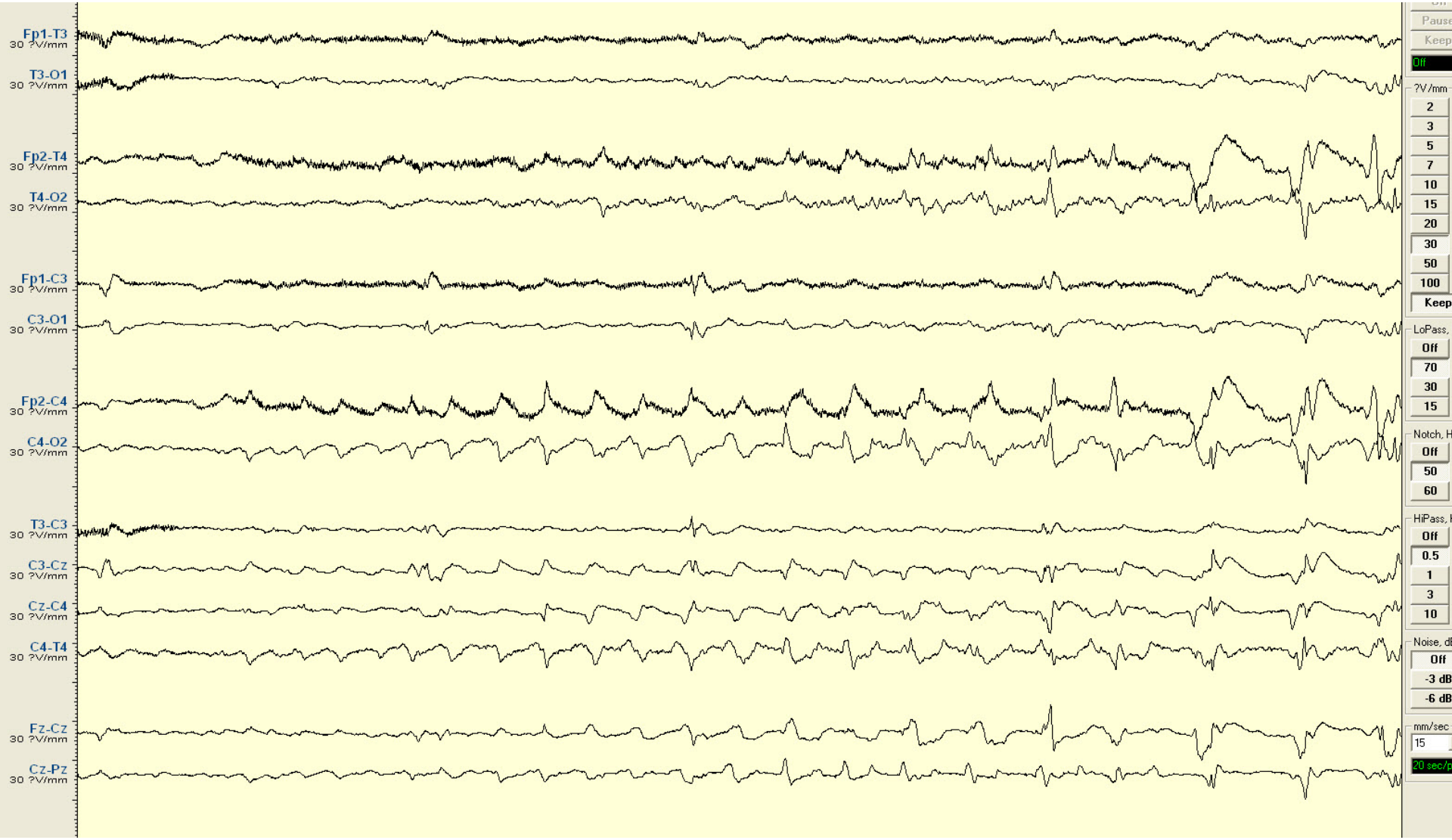
Early myoclonic encephalopathy (EME)

- Onset : Mainly in 1 month of life
- Seizure type : fragmentary myoclonus, focal seizure, tonic spasm
- Interictal EEG : Burst-suppression
- Ictal EEG : Diffuse synchronization, high amplitude slow wave or fast activity
- Causes : Metabolic disorders (NKH)
- Course : Severe, progressive course
- Treatment : Difficult to treat

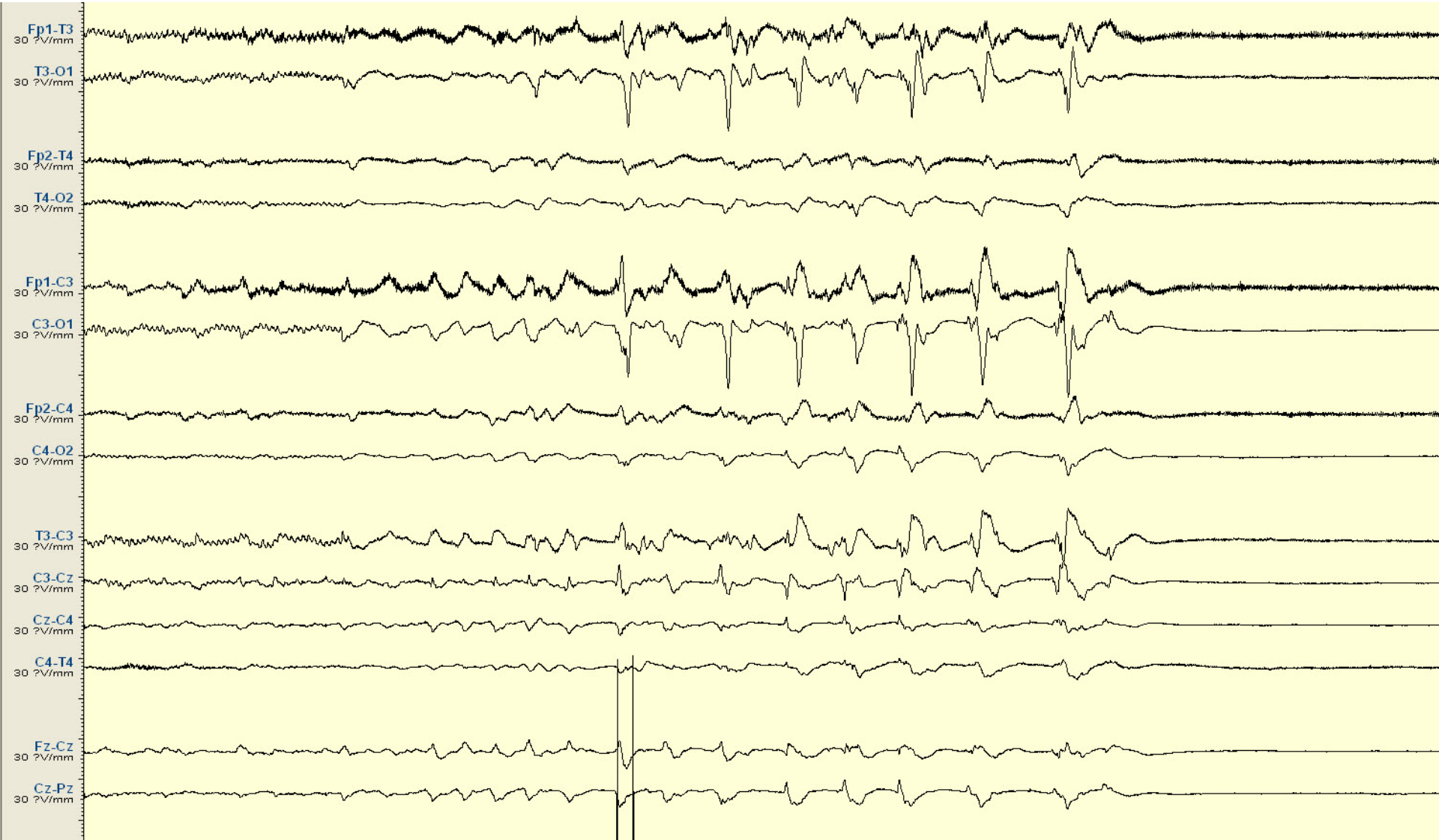
Burst and suppression 30 sec/page



Focal right


Fp1-T3
30 ?V/mm
T3-O1
30 ?V/mm
Fp2-T4
30 ?V/mm
T4-O2
30 ?V/mm
Fp1-C3
30 ?V/mm
C3-O1
30 ?V/mm
Fp2-C4
30 ?V/mm
C4-O2
30 ?V/mm
T3-C3
30 ?V/mm
C3-Cz
30 ?V/mm
Cz-C4
30 ?V/mm
C4-T4
30 ?V/mm
Fz-Cz
30 ?V/mm
Cz-Pz
30 ?V/mm
Pause
Keep
Off
?V/mm
2
3
5
7
10
15
20
30
50
100
Keep
LoPass,
Off
70
30
15
Notch, H
Off
50
60
HiPass, H
Off
0.5
1
3
10
Noise, dB
Off
-3 dB
-6 dB
mm/sec
15
20 sec/p

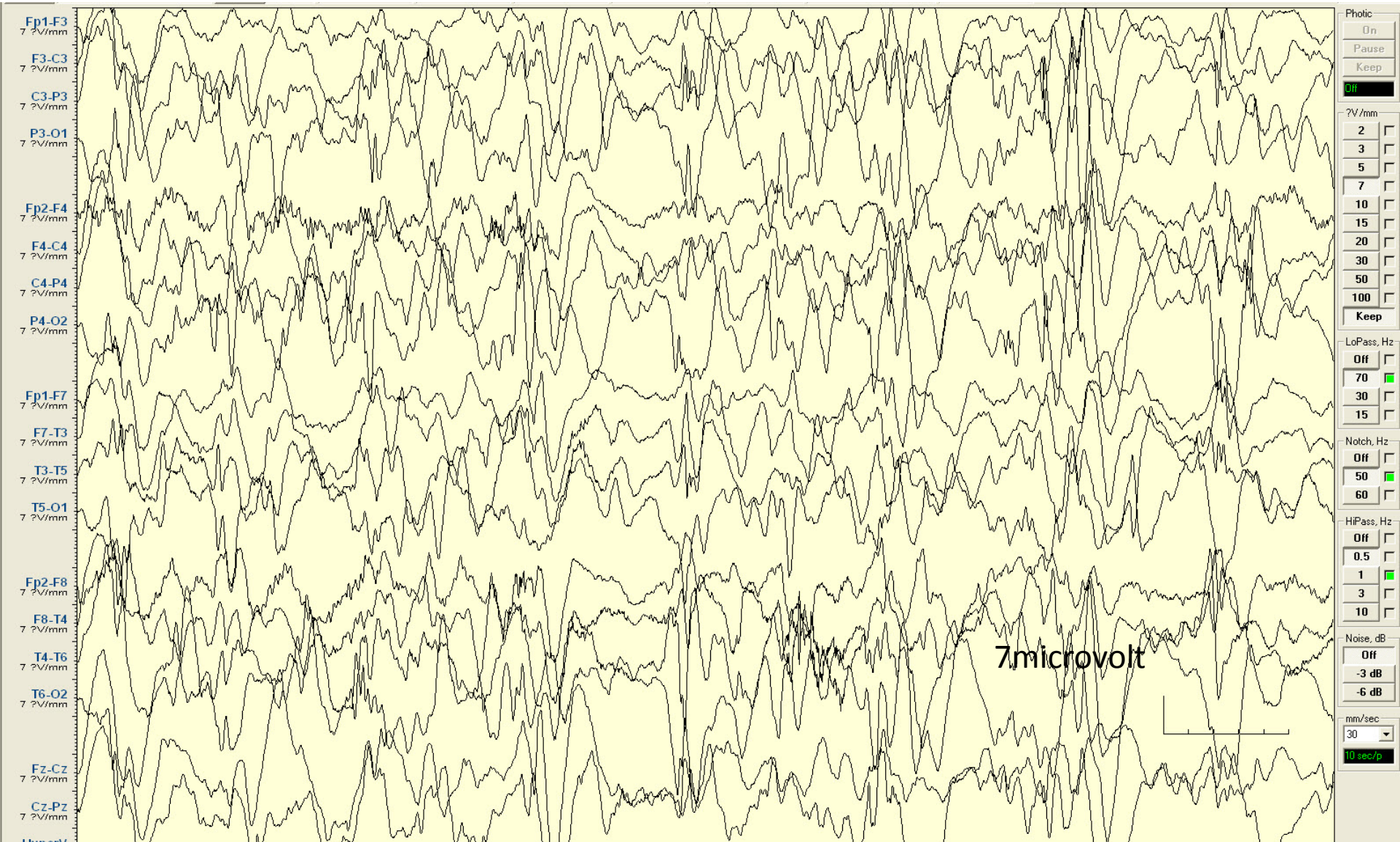
Focal left


Fp1-T3
30 ?V/mm
T3-O1
30 ?V/mm
Fp2-T4
30 ?V/mm
T4-O2
30 ?V/mm
Fp1-C3
30 ?V/mm
C3-O1
30 ?V/mm
Fp2-C4
30 ?V/mm
C4-O2
30 ?V/mm
T3-C3
30 ?V/mm
C3-Cz
30 ?V/mm
Cz-C4
30 ?V/mm
C4-T4
30 ?V/mm
Fz-Cz
30 ?V/mm
Cz-Pz
30 ?V/mm

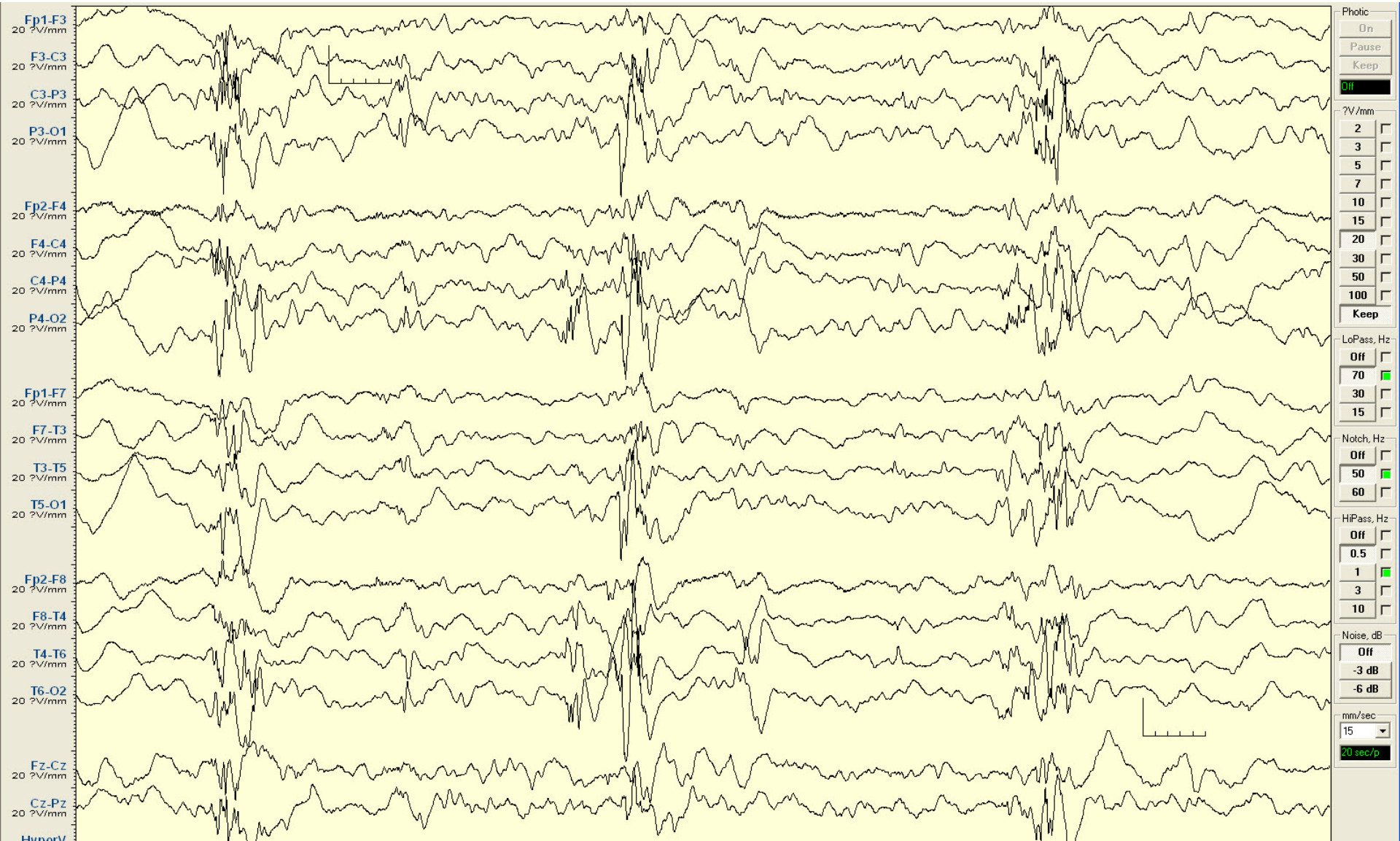
West syndrome

- Onset : 3-7 months
- Seizure type : Epileptic spasm
- Interictal EEG : Hypsarrhythmia
- Ictal EEG : Electrodecremental response
- Causes : CNS malformation, insults, TSC
- Course : Self-limited
  develop other types of seizures
- Treatment : Vigabatrin, difficult to treat

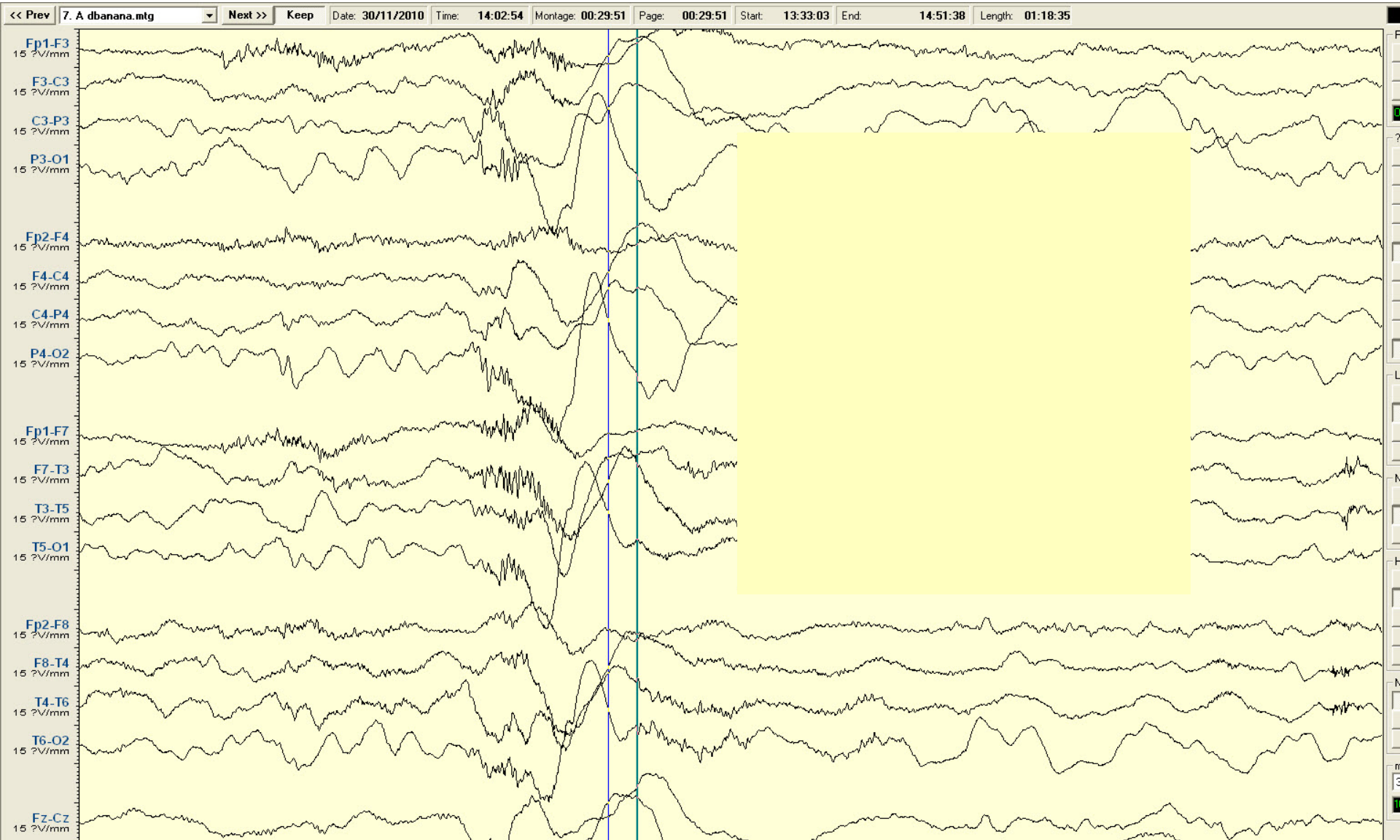
Hypsarrhythmia



Burst-suppression sleep



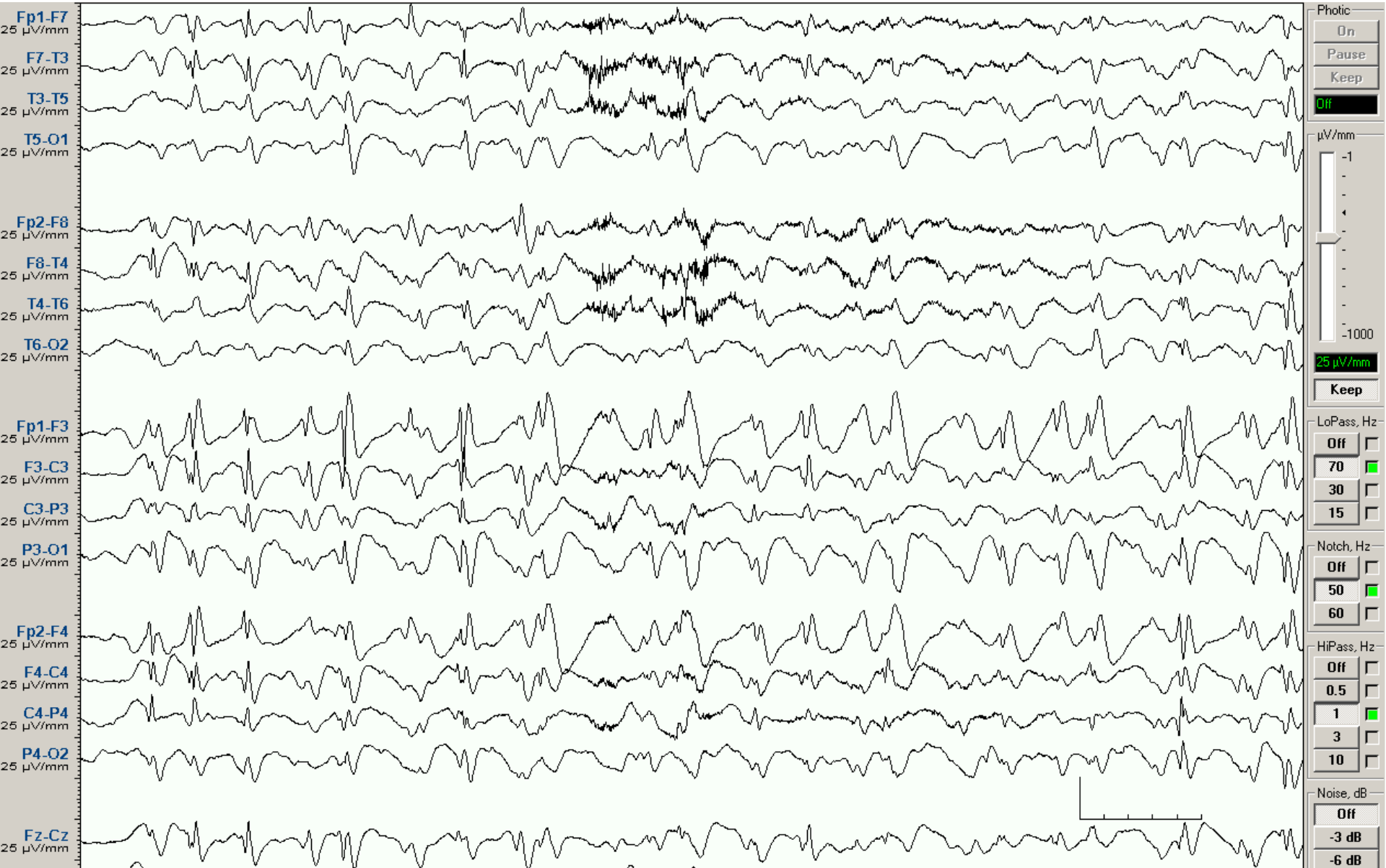
Ictal EEG: electrodecremental



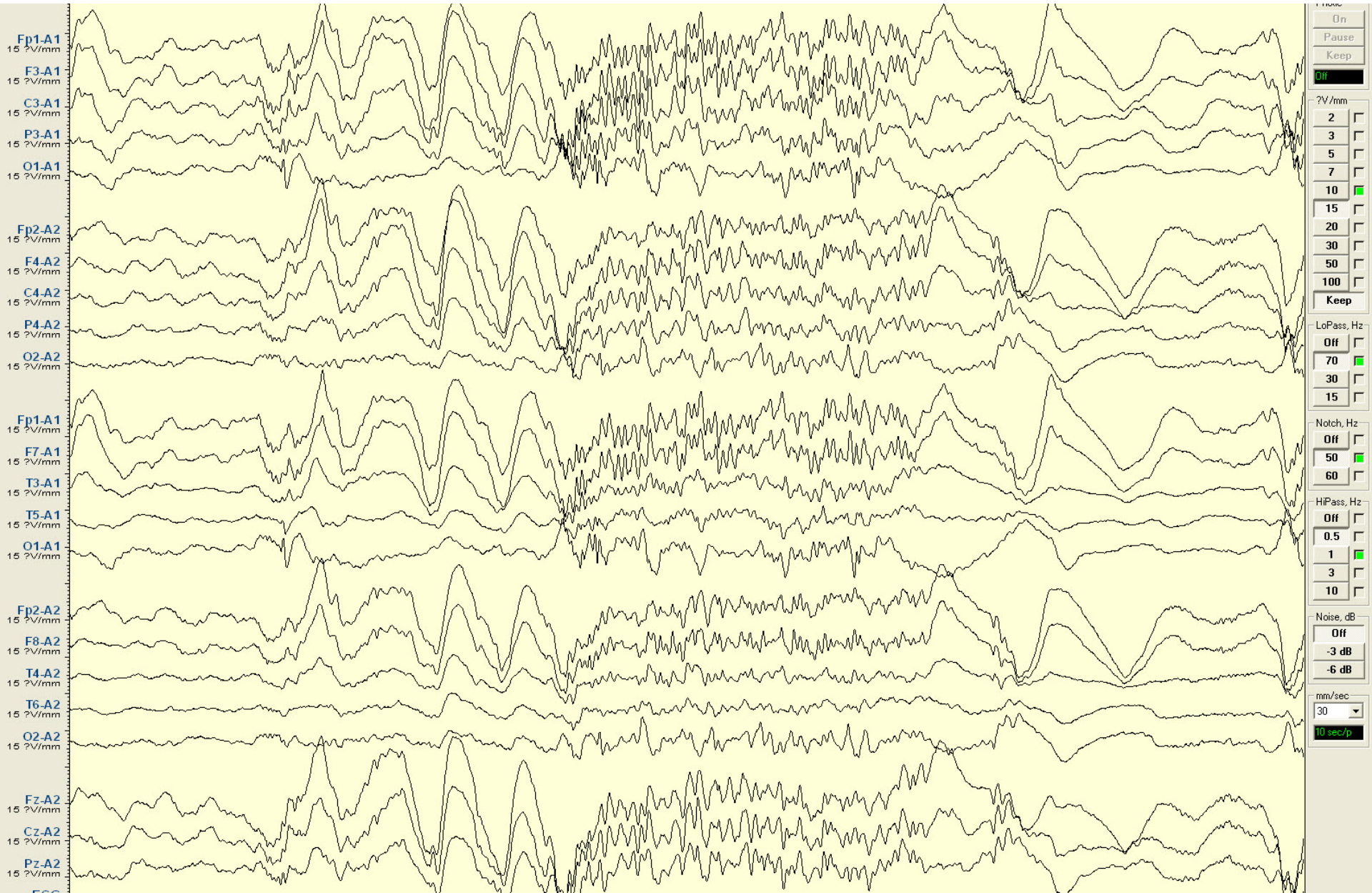
Lennox-Gastaut syndrome

- Onset :
- Seizure type : GT, Atonic, Atypical absence
- Interictal EEG : Awake: Slow spike-wave
  : Sleep: fast rhythmic wave
- Ictal EEG : Depends on seizure types
- Causes : CNS malformation/insults
- Course : Severe, difficult to control
- Treatment : VPA, TPM, LEV, ZNS
-

Slow-spike wave complexes



Paroxysmal fast activity



Focal epileptiform discharges