Benign and Malignant Rolandic and Occipital Spikes

Mary Connolly MB, FRCP(C)
Division of Pediatric Neurology
University of British Columbia

Objectives

- To review the differences between benign and malignant rolandic and occipital spikes
- To review the electroclinical features of idiopathic and symptomatic rolandic and occipital epilepsies
- To discuss differential diagnosis

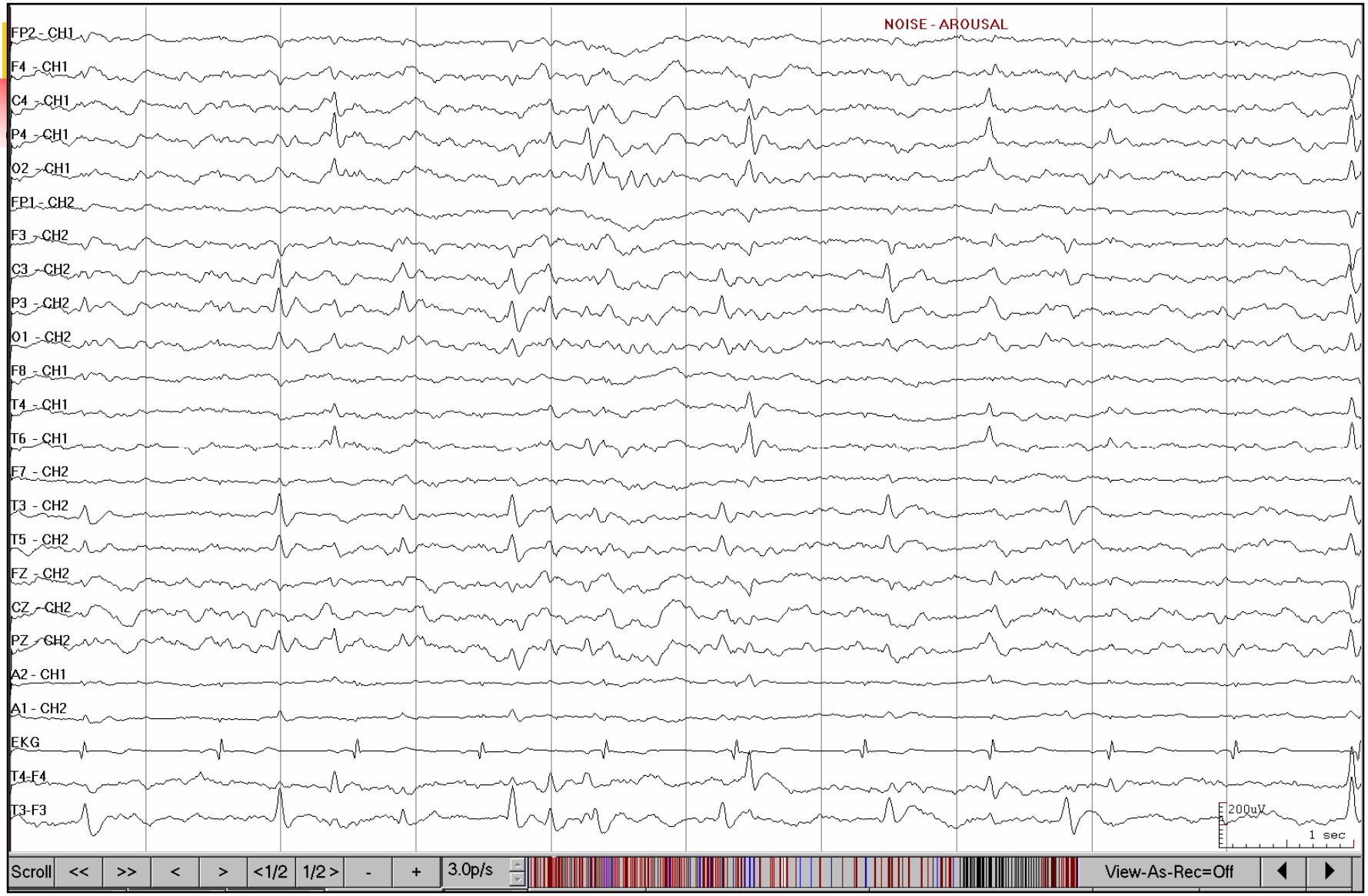
Disclosures

- None

History of Benign Rolandic Epilepsy

- EEG pattern - Gastaut in 1952
- Clinical pattern - Nayrac and Beaussart 1958
- EEG abnormality may occur without clinical seizures - Gibbs et al. 1954
- Genetic factors - Bray and Wiser 1964

Interictal Discharge



Benign Rolandic Epilepsy

- Most common partial epilepsy
- Population study: 6.2-10.7 per 100,000
- Onset 3-13 years
- M:F 1.5:1
- 10-13% have a single seizure
- 20% have frequent seizures
- 65% nocturnal ;15% nocturnal or diurnal ; 10-20% in waking state only

Description of Seizures

- **Somatosensory onset with unilateral parasthesiae of tongue, lips, gums or inner cheeks**
- **Unilateral tonic, clonic or tonic-clonic activity in face, lips, tongue, pharyngeal and laryngeal muscles**
- **Speech arrest or dysarthria**
- **Drooling**
- **Preservation of consciousness**

Description of Seizures

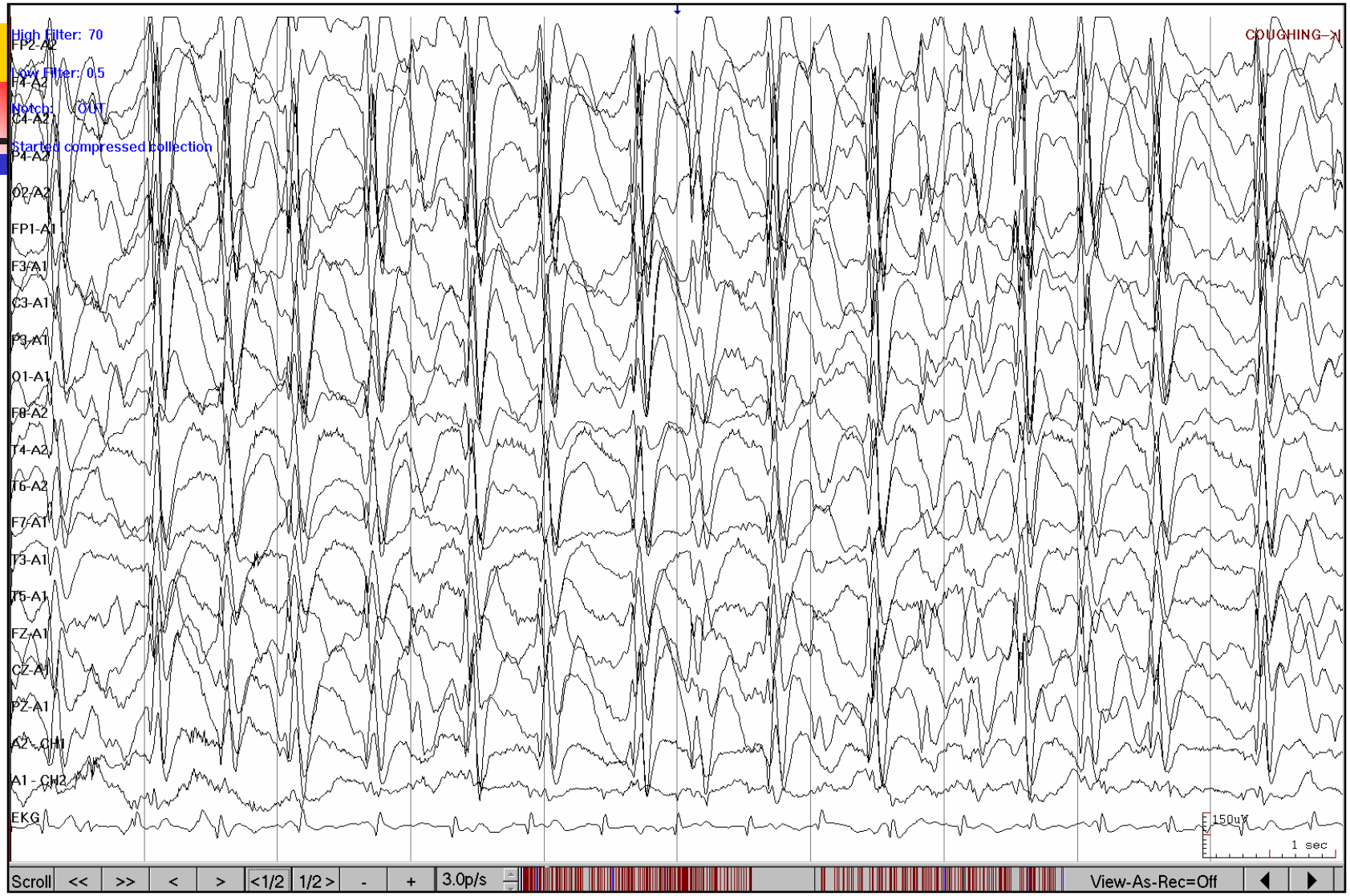
- **Simple partial hemifacial seizure with somatosensory aura**
- **Often associated loss of awareness**
- **Secondarily generalized seizure - onset not witnessed**

Video of Benign Rolandic Seizure

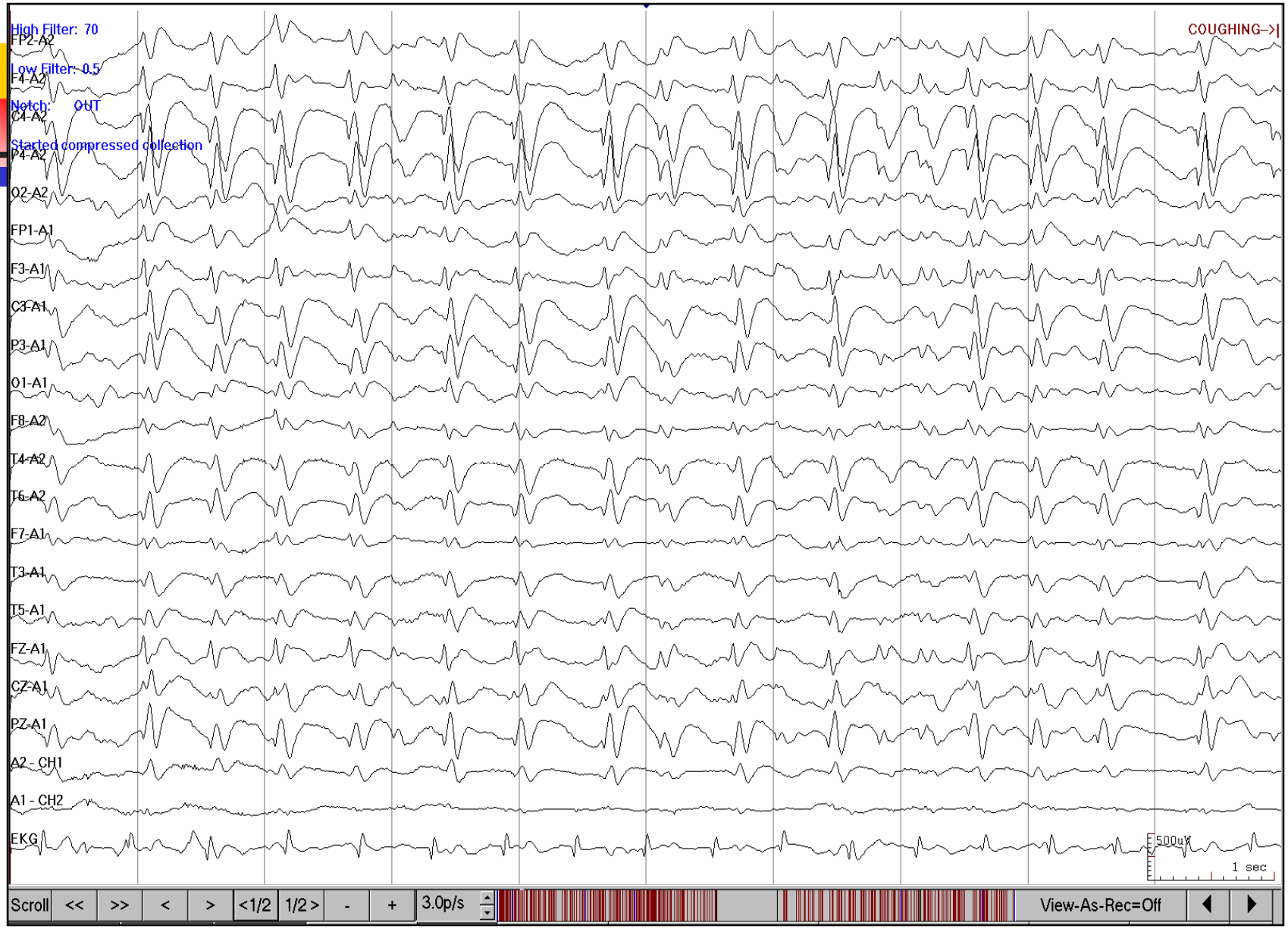
Rolandic Spike Dipoles

- Very common
- Only 9% of children with rolandic spike dipoles develop epilepsy
- Rolandic spike dipoles may occur in symptomatic rolandic epilepsy

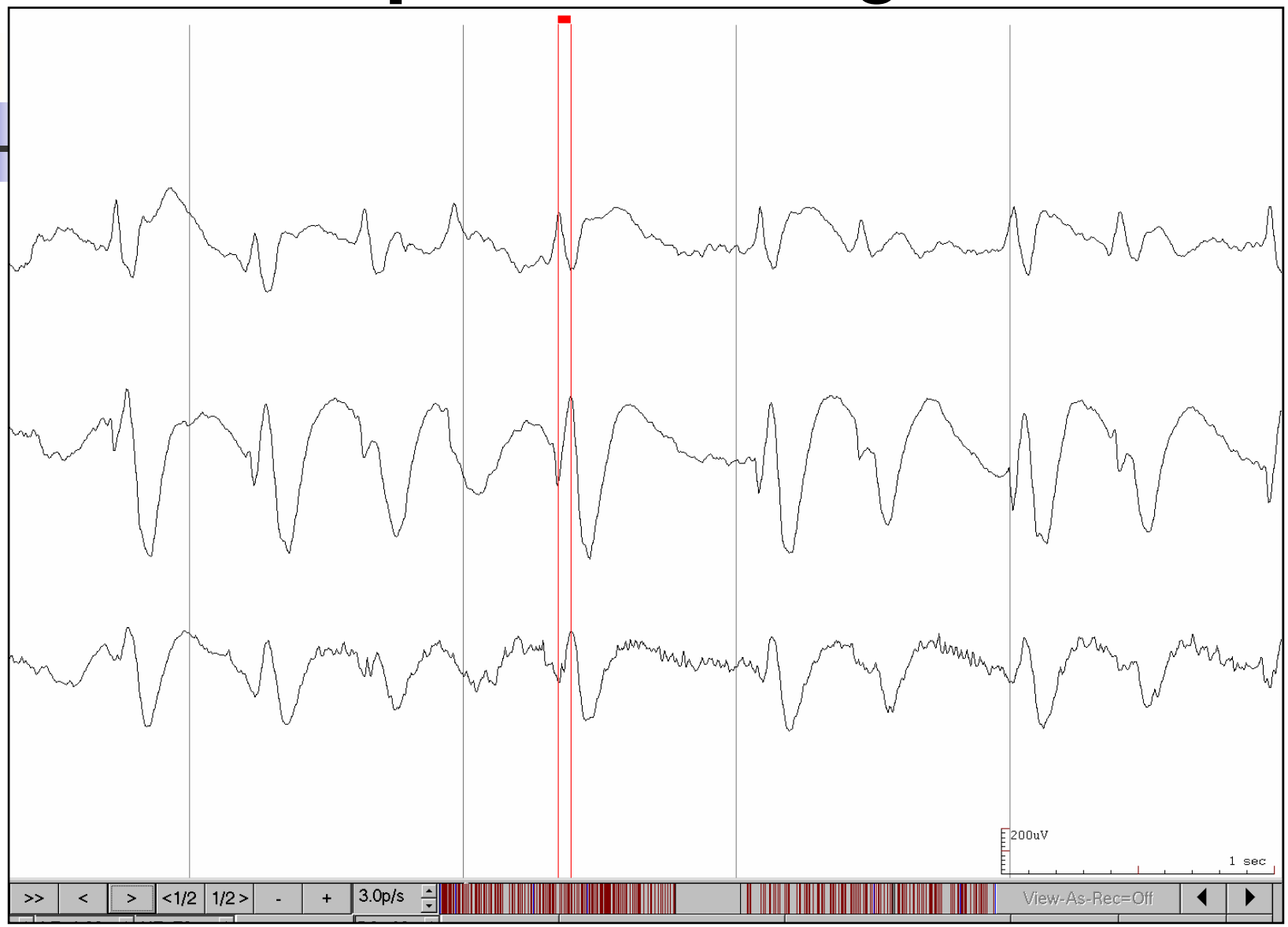
High Voltage Diffuse Rolandic Spikes



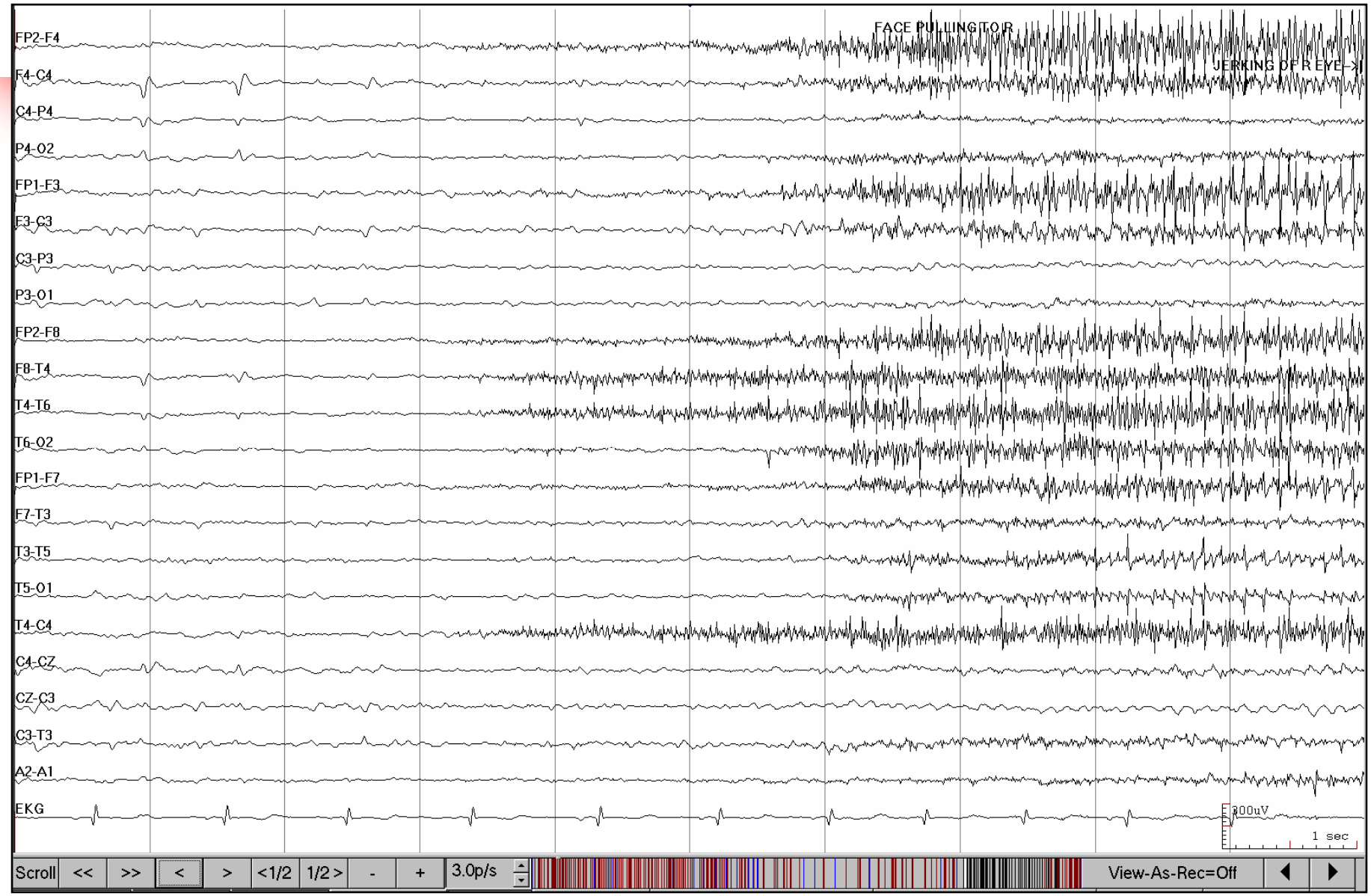
Sensitivity 50uV/mm



Ictal Dipole Discharges



Ictal EEG in Benign Rolandic Epilepsy



Ictal EEG contd

Neuropsychological Function in BREC

- **17 patients with BREC**
  - **7 – 14 years**
  - **12 on medication**
- **17 controls**
  - **age, sex and estimated intelligence**
- **Assessments**
  - **Neuropsychological testing**
  - **Parent and Teacher rating**

Croona et al Dev Med Child Neurol 1999;41:813

Neuropsychological Function in BREC

- **Significant differences in**
  - **Auditory-verbal learning**
  - **Memory**
  - **Executive function**
  - **Parent and Teacher Rating**
    - **Parents - distractibility, impulsivity,**
    - **Teachers – reading comprehension**

Croona et al Dev Med Child Neurol 1999;41:813

Evolution of Benign Rolandic Epilepsy

- 35 children
- 6 monthly assessments
  - clinical
  - EEG
  - neuropsychology
- First seizure to recovery

Massa et al Neurology 2001;57:1071-1079

Prospective Study of BREC

- Decline mainly involved
  - **Performance IQ**
  - **Sustained attention**
  - **Working memory**
  - **Executive functioning.**

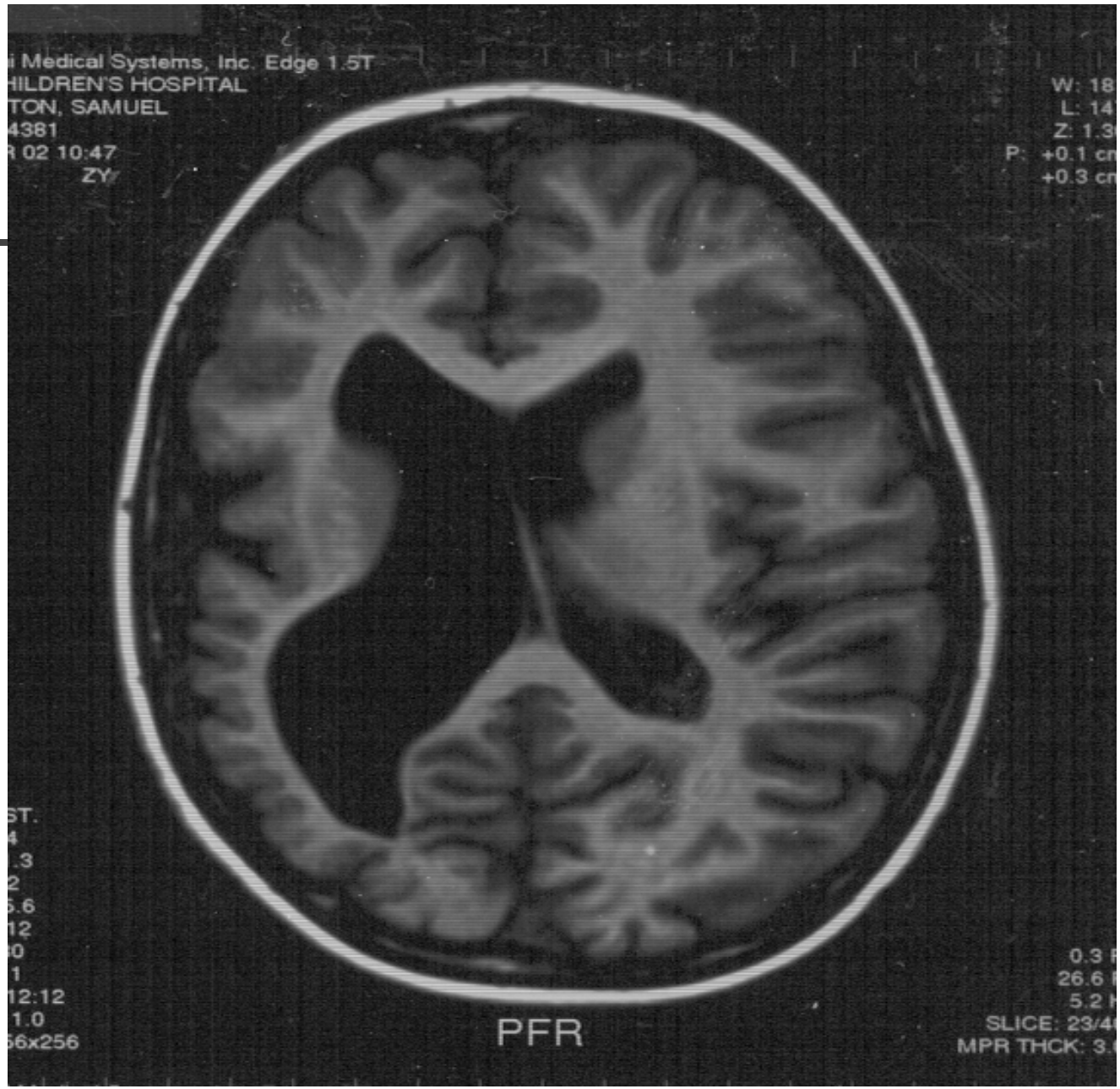
EEG Features Predictive of Poor Prognosis

EEG Abnormality	P value
Intermittent slow wave focus	<0.001
Asynchronous bilateral spike wave foci	<0.001
Rhythmic clusters of spike-wave	<0.001
Generalized 3-4 Hz spike waves	<0.05
Atonia, myoclonia correlates with SW	<0.05

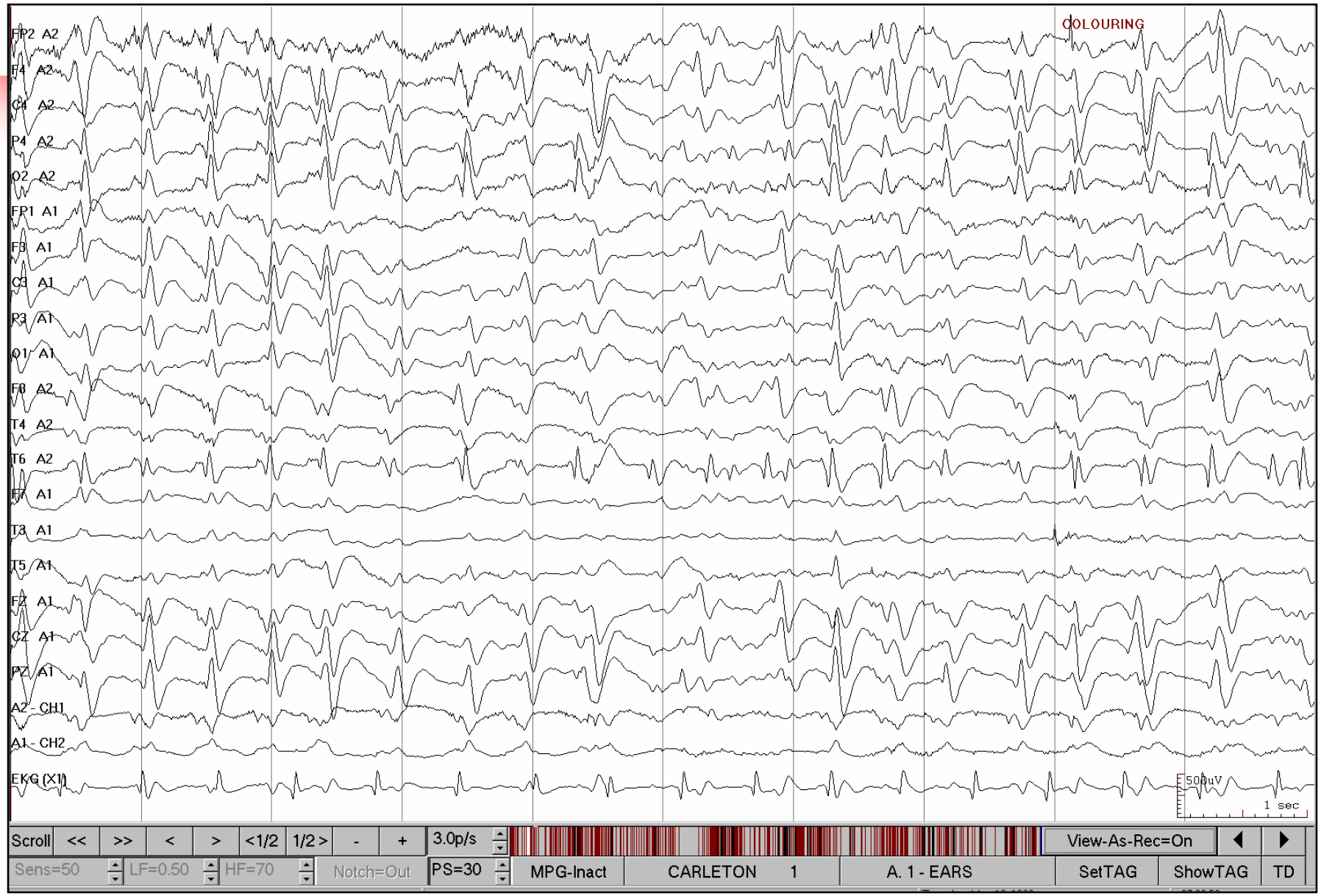
3 of 5 features in 10 children for at least 6 months

Simon

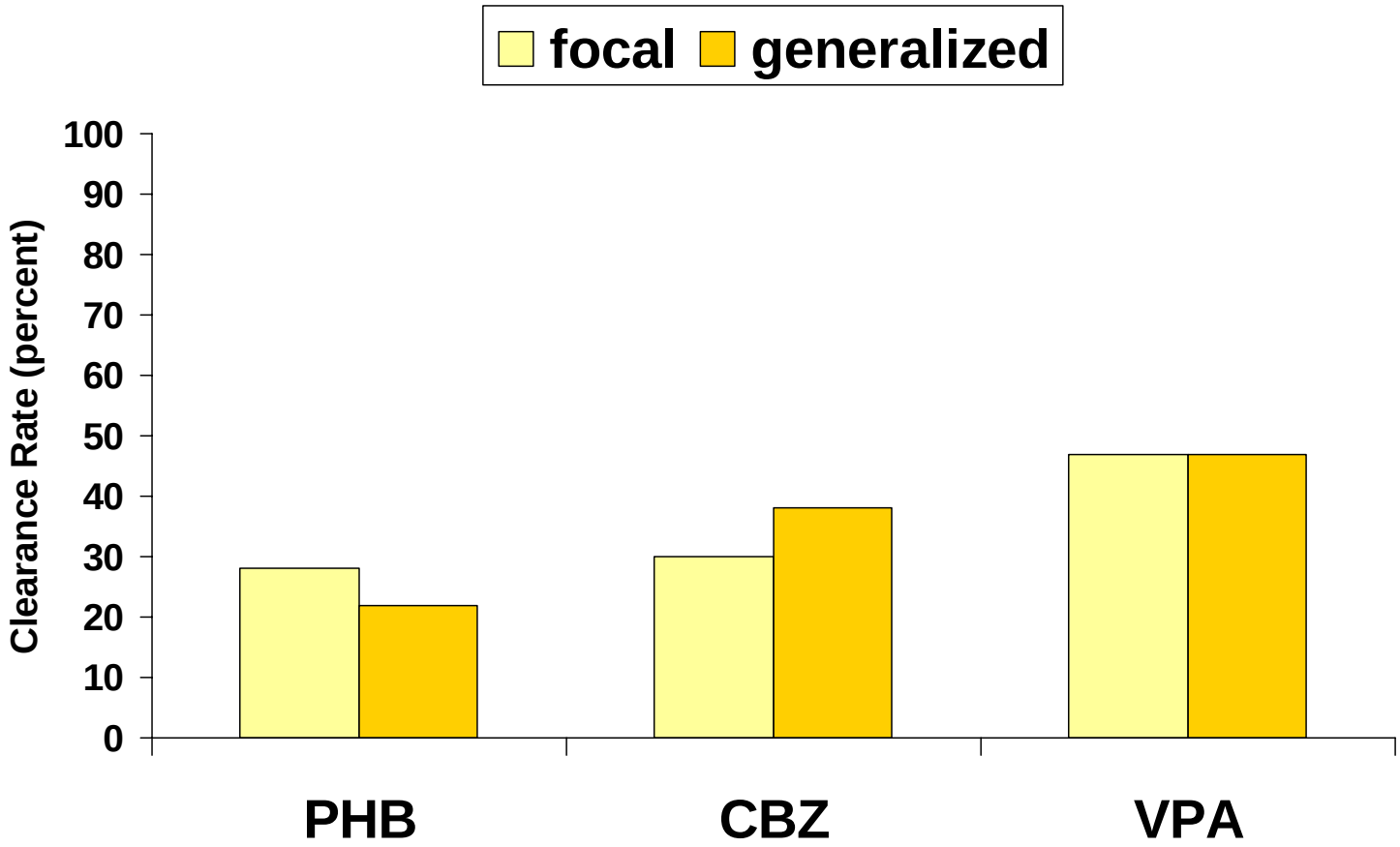
- Antenatal ultrasound – right pachygyria
- Mild left hemiplegia
- 5 X GTC with fever – 18 months to 5 years
- Intermittent L facial and speech dificulty for up to 30 mins at 5 years


Medical Systems, Inc. Edge 1.5T
HILDREN'S HOSPITAL
TON, SAMUEL
4381
02 10:47
ZY
W: 18
L: 14
Z: 1.3
P: +0.1 cm
+0.3 cm
ST.
.3
.6
12
12:12
1.0
6x256
0.3
26.6
5.2
SLICE: 23/4
MPR THCK: 3.
PFR

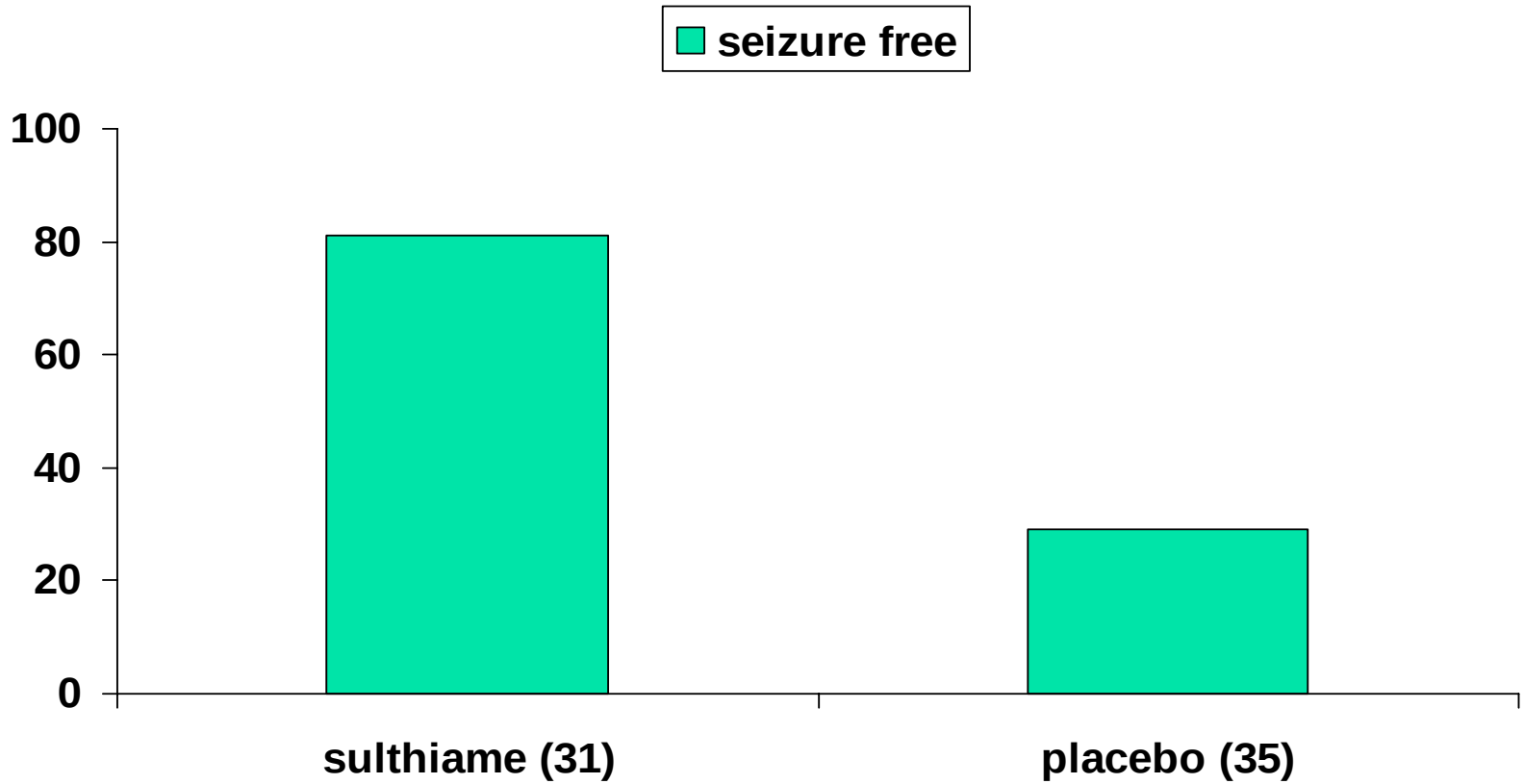
Simon's EEG


COLOURING
FP2 A2
F4 A2
C4 A2
P4 A2
O2 A2
FP1 A1
F3 A1
C3 A1
P3 A1
O1 A1
F8 A2
T4 A2
T6 A2
F7 A1
T3 A1
T5 A1
FZ A1
CZ A1
PZ A1
A2 - CH1
A1 - CH2
EKG (X1)
500uV
1 sec
Scroll
<<
>>
<
>
<1/2
1/2>
-
+
3.0p/s
View-As-Rec=On
Sens=50
LF=0.50
HF=70
Notch=Out
PS=30
MPG-Inact
CARLETON 1
A. 1 - EARS
SetTAG
ShowTAG
TD

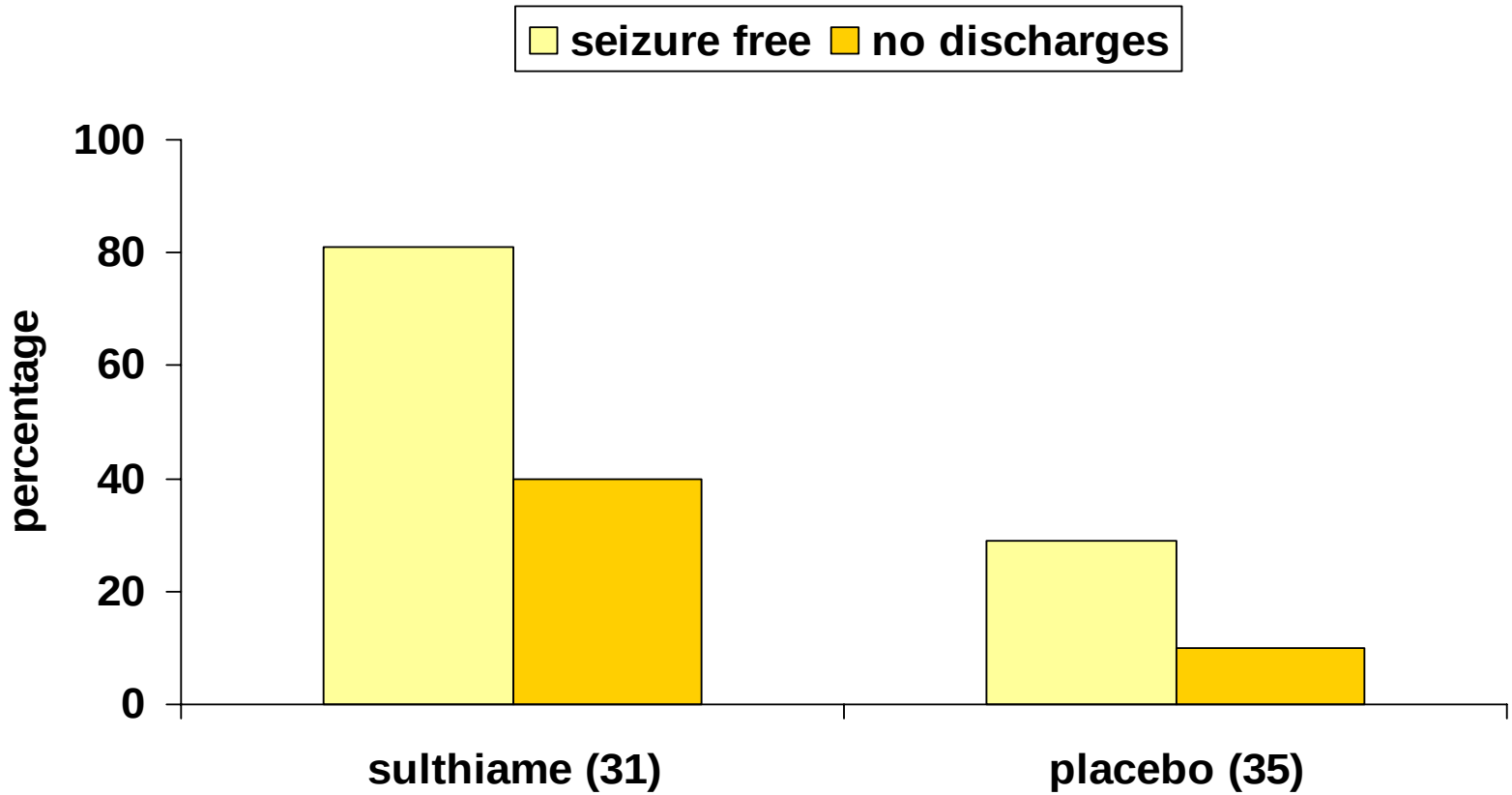
Effect of AEDs on Interictal Spikes in Children



Sulthiame in Benign Rolandic Epilepsy



Sulthiame in Benign Rolandic Epilepsy



Rating et al. Epilepsia 2000;41:1284

Drugs which Suppress Interictal Discharges

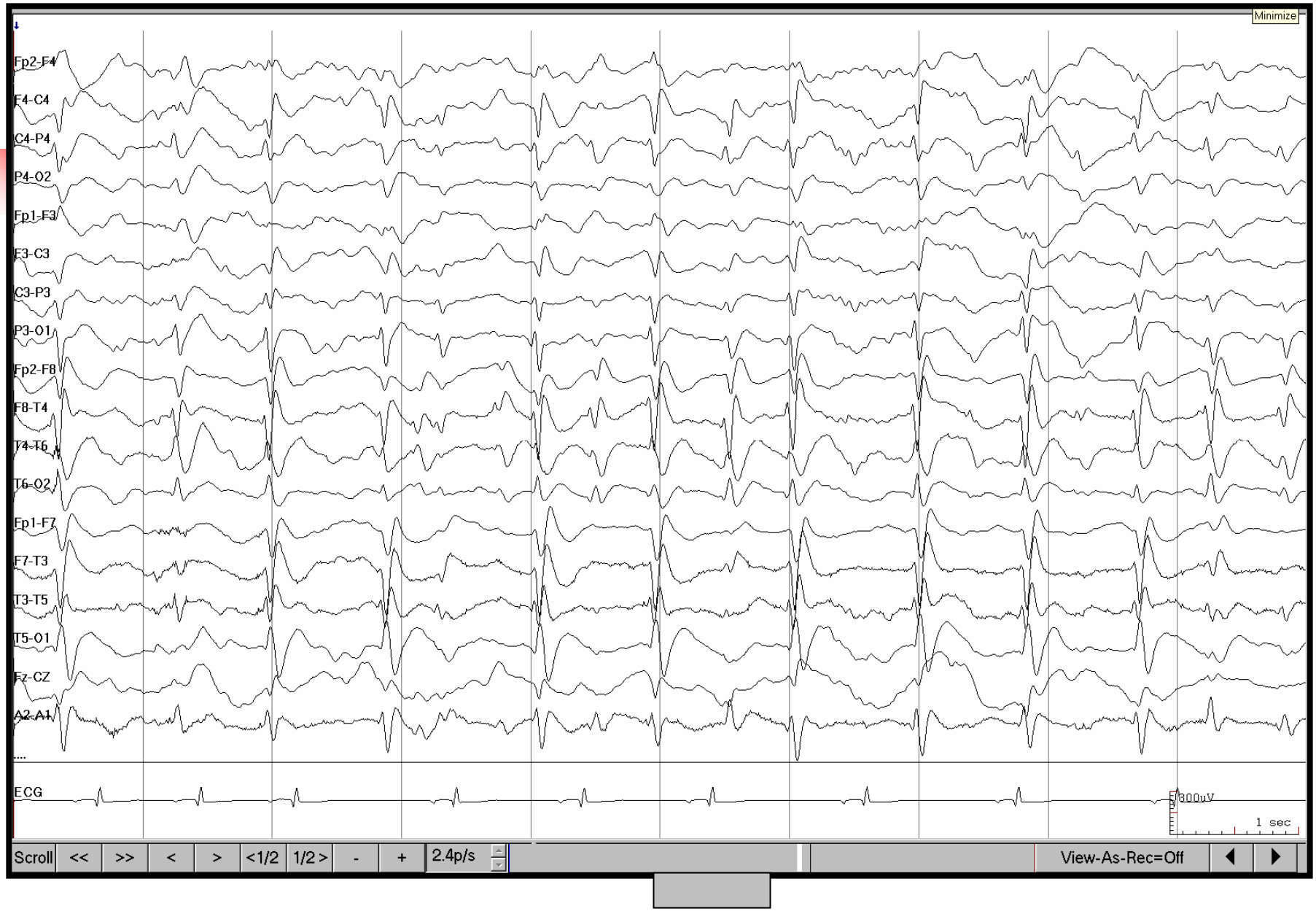
- Sulthiame
- Diazepam
- Valproic acid
- Lamotrigine
- Corticosteroids
  - Landau-Kleffner
  - Continuous Spike-Wave in Slow Sleep

Malignant Rolandic Spikes

- Christine developmentally normal
- Referred at 9 years of age
- At 7.5 years gradual deterioration in speech
- No clinical seizures
- Initially diagnosed with “Selective mutism”

Landau-Kleffner Syndrome

- **Difficulty understanding speech and sounds**
- **Gradual deterioration in ability to speak**
- **More distractible in school**
- **Behavior problems**

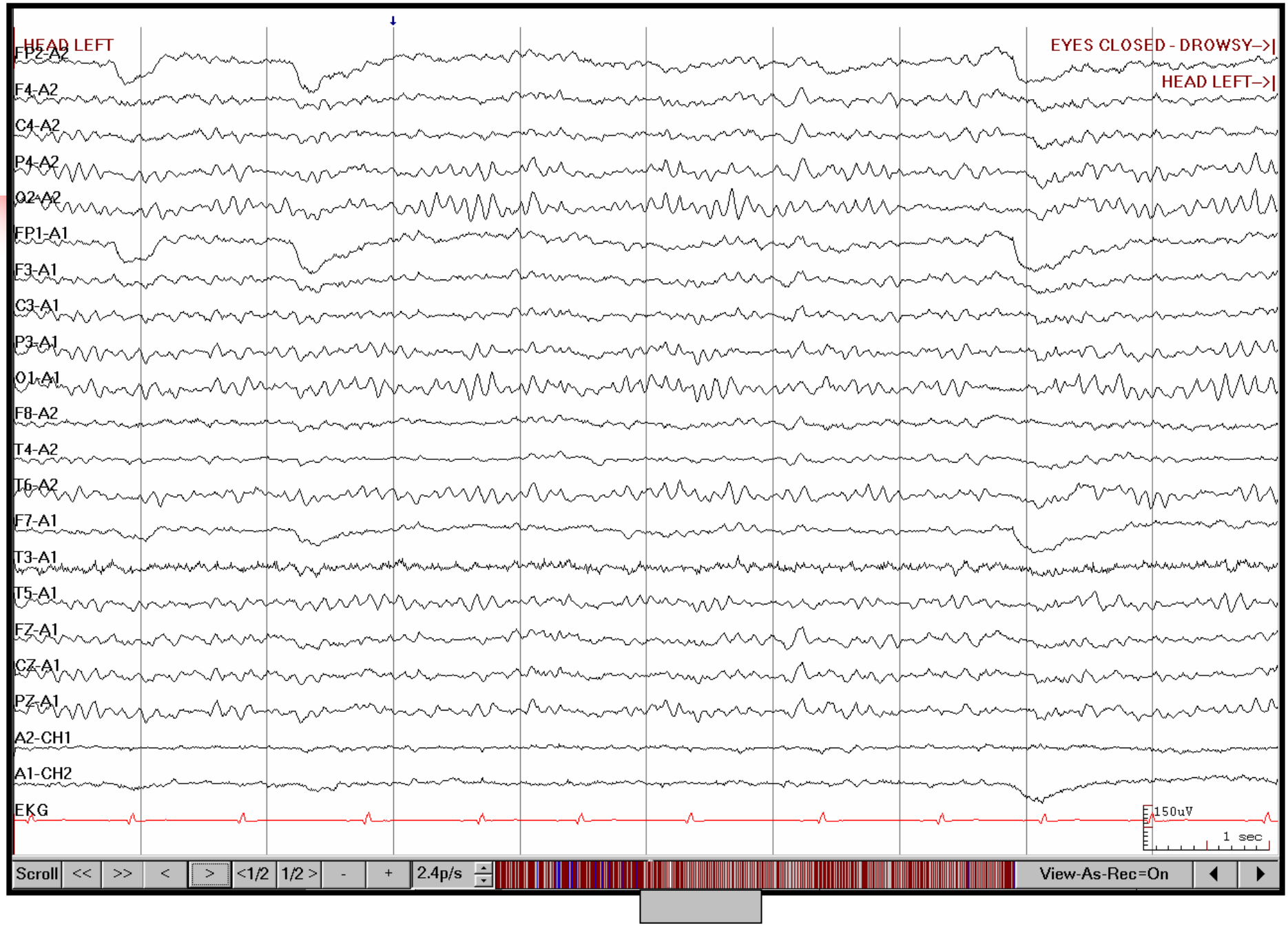

Minimize
Fp2-F4
F4-C4
C4-P4
P4-O2
Fp1-F3
F3-C3
C3-P3
P3-O1
Fp2-F8
F8-T4
T4-T6
T6-O2
Fp1-F7
F7-T3
T3-T5
T5-O1
Fz-CZ
A2-A1
ECG
300uV
1 sec
Scroll
<<
>>
<
>
<1/2
1/2>
-
+
2.4p/s
View-As-Rec=Off

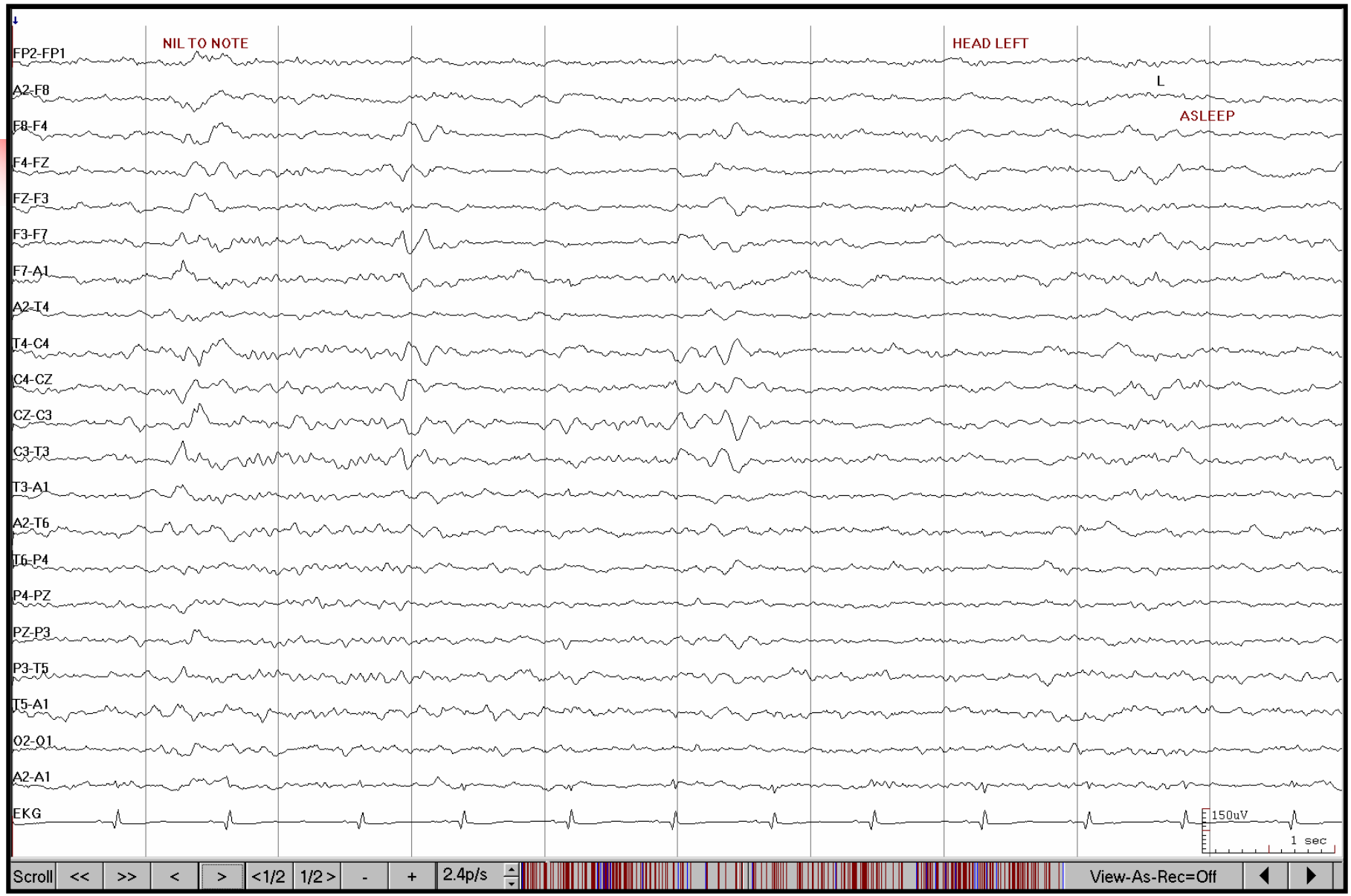
Christine: Treatment

- Prednisone 2mg/kg
- Sulthiame added 3 months post diagnosis
  - Sulfonamide derivative
  - Mechanism of action
    - Carbonic anhydrase inhibition
    - Blocks sodium channel

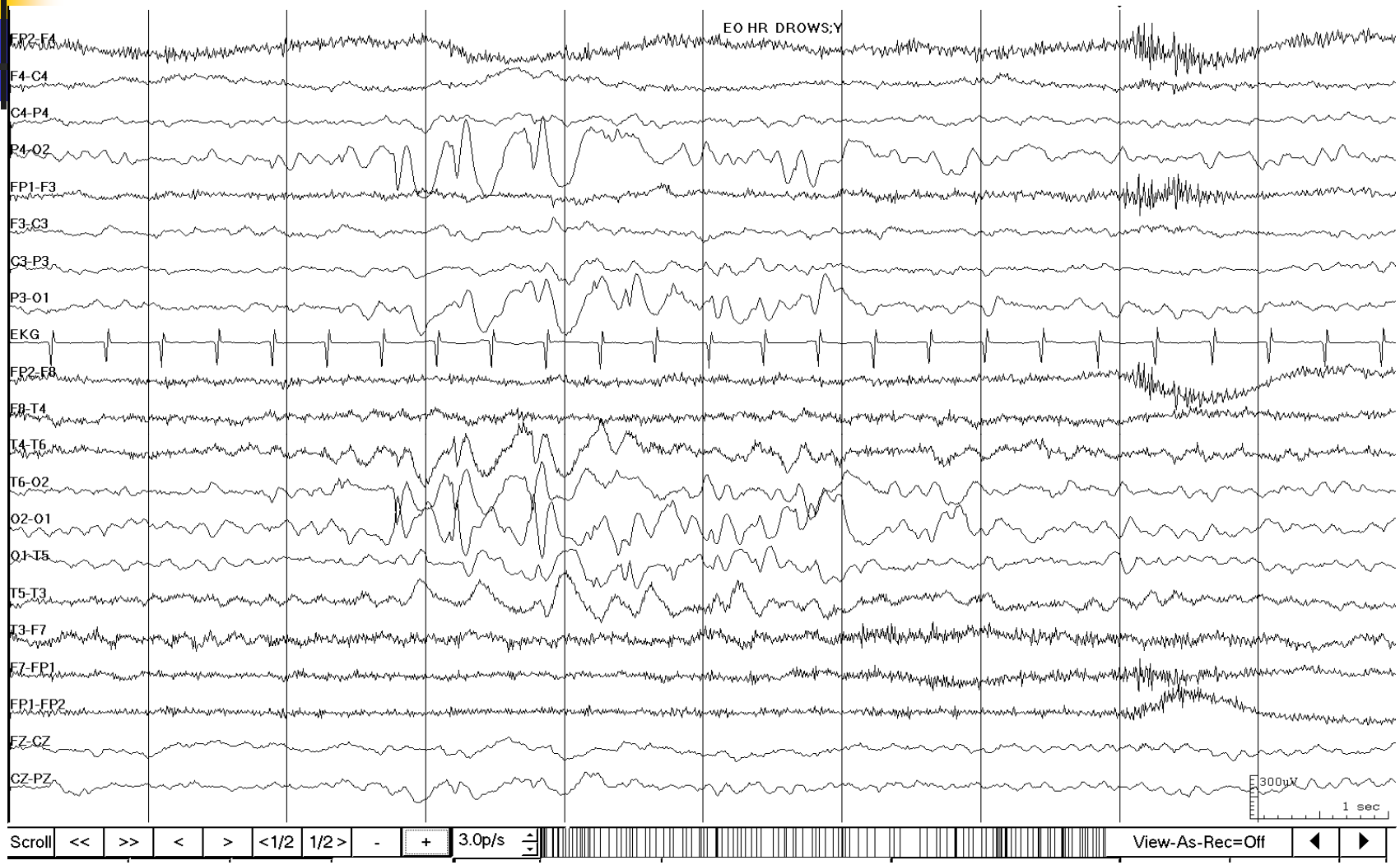
Christine - Evolution

- Dramatic improvement clinically
- 10 months following diagnosis speaking well
- Presently – 6.5 years since diagnosis
  - Mild comprehension difficulty in a noisy environment
  - EEG normal
- Now off medication for 5 years


HEAD LEFT
EYES CLOSED - DROWSY->|
HEAD LEFT->|
FP2-A2
F4-A2
C4-A2
P4-A2
O2-A2
FP1-A1
F3-A1
C3-A1
P3-A1
O1-A1
F8-A2
T4-A2
T6-A2
F7-A1
T3-A1
T5-A1
FZ-A1
CZ-A1
PZ-A1
A2-CH1
A1-CH2
EKG
150uV
1 sec
Scroll
<<
>>
<
>
<1/2
1/2 >
-
+
2.4p/s
View-As-Rec=On


NIL TO NOTE
HEAD LEFT
L
ASLEEP
FP2-FP1
A2-F8
F8-F4
F4-FZ
FZ-F3
F3-F7
F7-A1
A2-T4
T4-C4
C4-CZ
CZ-C3
C3-T3
T3-A1
A2-T6
T6-P4
P4-PZ
PZ-P3
P3-T5
T5-A1
O2-O1
A2-A1
EKG
150uV
1 sec
Scroll
<<
>>
<
>
<1/2
1/2>
-
+
2.4p/s
View-As-Rec=Off

OCCIPITAL SPIKES



Elizabeth

- 3.5 year old girl
- Onset of seizures at 3 years
  - Staring, eyes and head to left
  - Unresponsive, limp, pale
  - Vomiting
  - Duration 12-20 minutes
- Normal development and exam
- Maternal grandmother epilepsy

Panayiotopoulos Syndrome

- 3-5 years (range 1-14 years)
- Nocturnal seizures in 2/3
- Tonic eye deviation, vomiting
- Visual symptoms rarely reported
- Prominent autonomic features

Panayiotopoulos Syndrome

- Seizures prolonged, status in 1/3
- Infrequent seizures
- Prognosis excellent
- Seizures rare after 13 years
- Children may develop rolandic epilepsy
- EEG: multifocal posterior quadrant epileptiform discharges

Gastaut Syndrome

- **Brief seizures characterized by visual hallucinations or ictal blindness**
- **Children 4-16 years**
- **5% had symptoms in adulthood**

EEG Features

- Normal background
- High amplitude spike-wave (80%) or sharp waves (20%) over the occipital and or posterior temporal area
- Discharges occur rhythmically
- May disappear on eye-opening in 94%
- 38%: generalized spike-wave or centrotemporal spikes

Clinical Features

- Amaurosis in 52%
- Phosphenes in 45%
- Complex visual hallucinations in 14%
- Visual illusions in 14%
  - micropsia
  - palinopsia

Clinical Features

- Hemiclonic seizures in 43%
- Complex partial seizures in 14%
- Generalized tonic-clonic seizures 13%
- Other features in 25%
  - dysphasia
  - dysesthesiae

Clinical Features

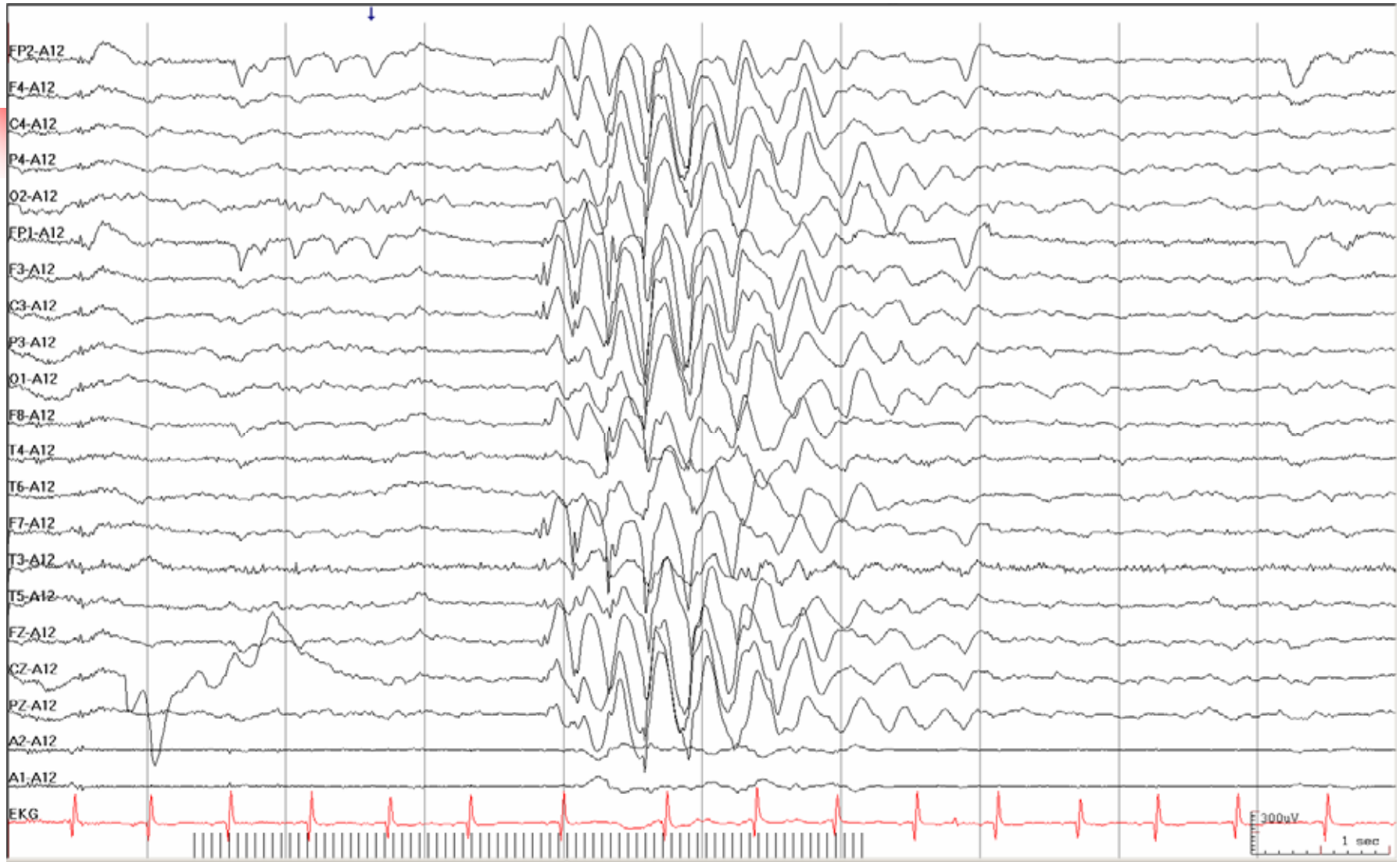
- Post-ictal headache in 33%
- Nausea in 17%
- No clear precipitating factors
- Features may be difficult to differentiate from migraine

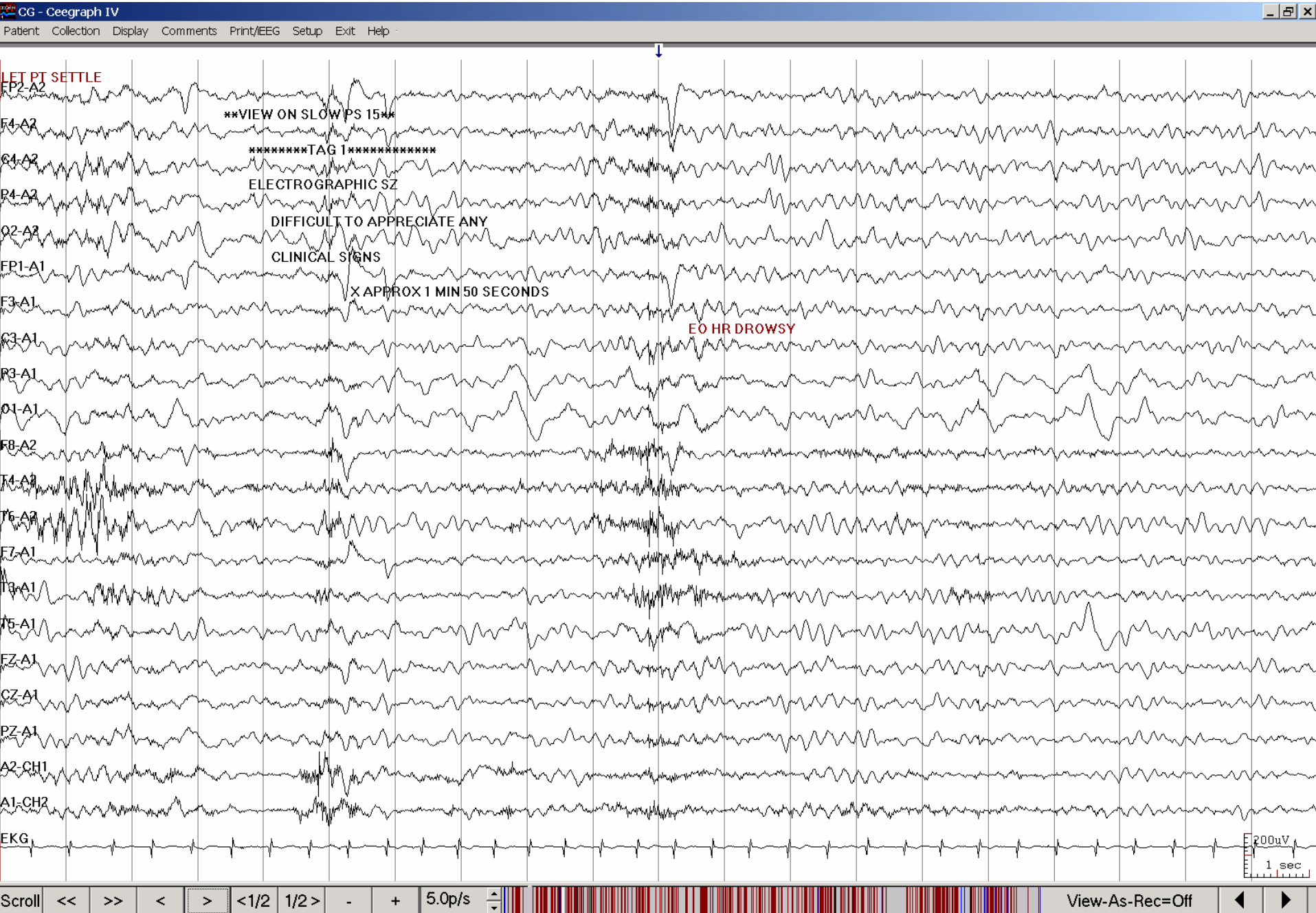
Prognosis of Gastaut Syndrome

- Complete seizure control in 60%
- Remission in late adolescence although up to 5% of adults may continue to have seizures
- Diagnosis of this condition is difficult

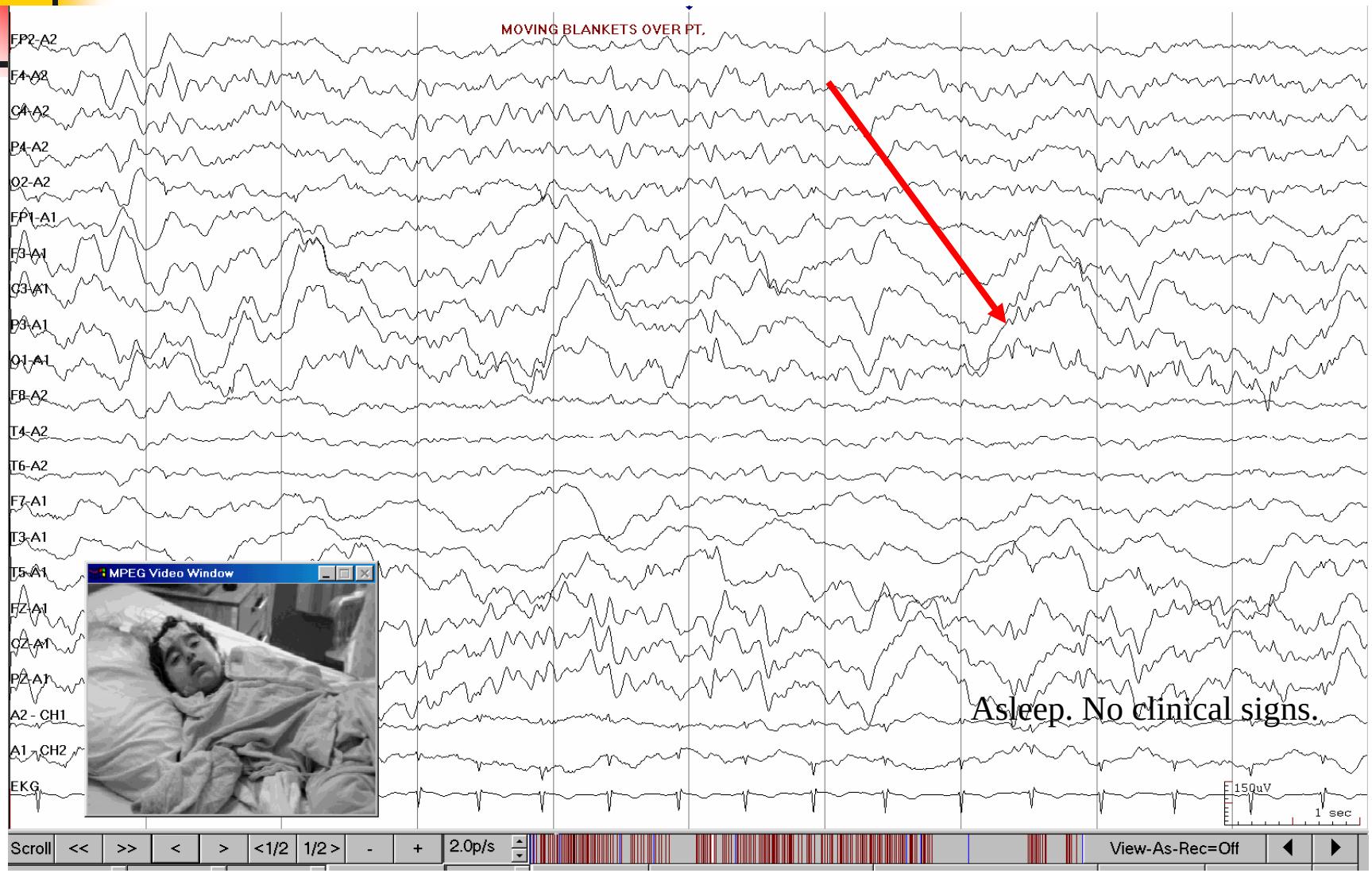
Idiopathic OLE with Photosensitivity

- ***Guerrini et al. (Epilepsia 1995;36:883-891)***
- **5-17 years of age**
- **Seizures induced by light, visual hallucinations, tonic head and eye deviation, nausea, headache, may be aware**

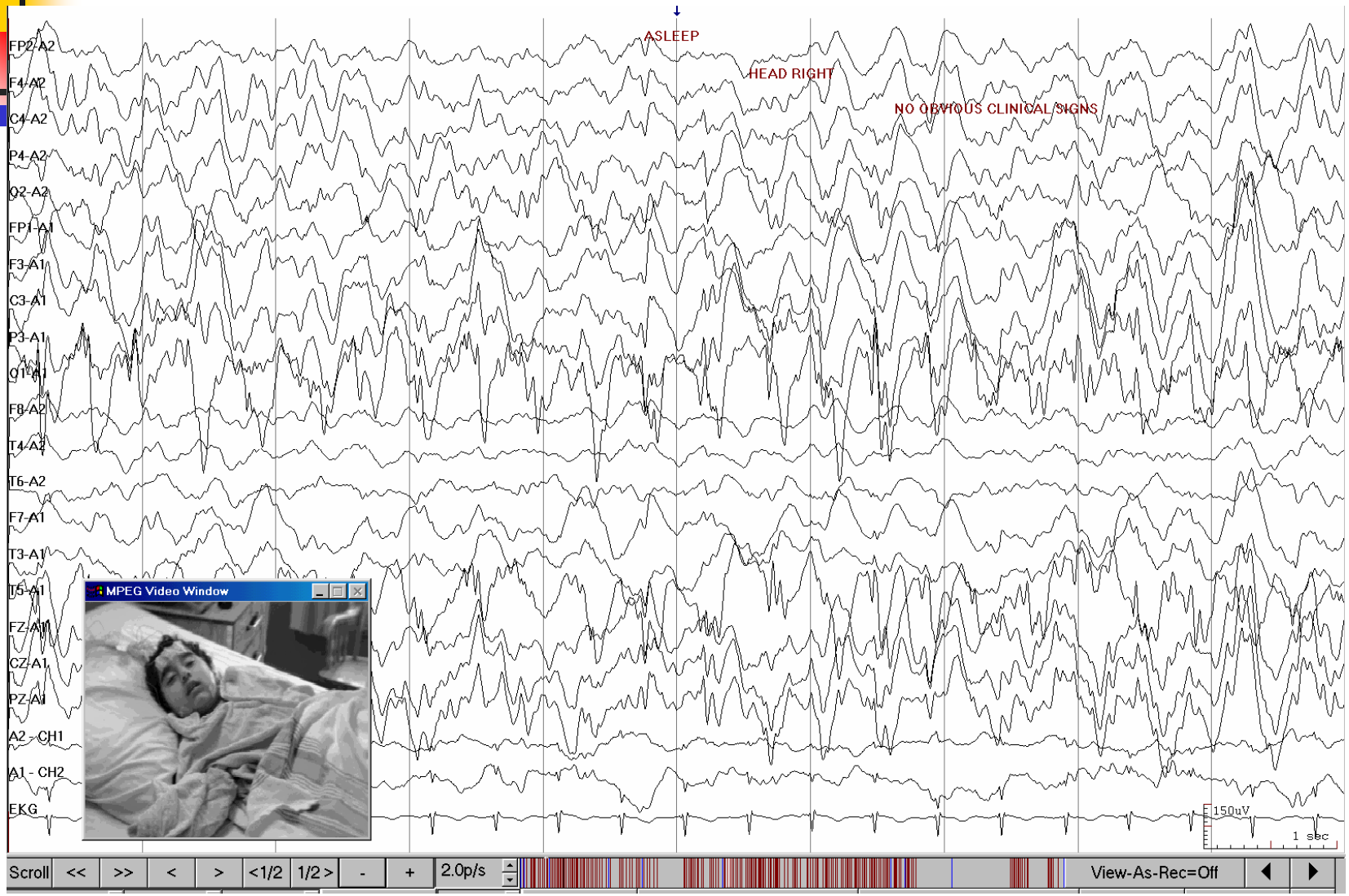

FP2-A12
F4-A12
C4-A12
P4-A12
O2-A12
FP1-A12
F3-A12
C3-A12
P3-A12
O1-A12
F8-A12
T4-A12
T6-A12
F7-A12
T3-A12
T5-A12
FZ-A12
CZ-A12
PZ-A12
A2-A12
A1-A12
EKG
300uV
1 sec


CG - Ceegraph IV
Patient Collection Display Comments Print/EEG Setup Exit Help
LET PT SETTLE
FP2-A2
F4-A2
C4-A2
P4-A2
O2-A2
FP1-A1
F3-A1
C3-A1
P3-A1
O1-A1
F8-A2
T4-A2
T6-A2
F7-A1
T3-A1
T5-A1
FZ-A1
CZ-A1
PZ-A1
A2-CH1
A1-CH2
EKG
VIEW ON SLOW PS 15
********TAG 1**********
ELECTROGRAPHIC SZ
DIFFICULT TO APPRECIATE ANY
CLINICAL SIGNS
X APPROX 1 MIN 50 SECONDS
EO HR DROWSY
200uV
1 sec
Scroll << >> < > <1/2 1/2 > - + 5.0p/s View-As-Rec=Off

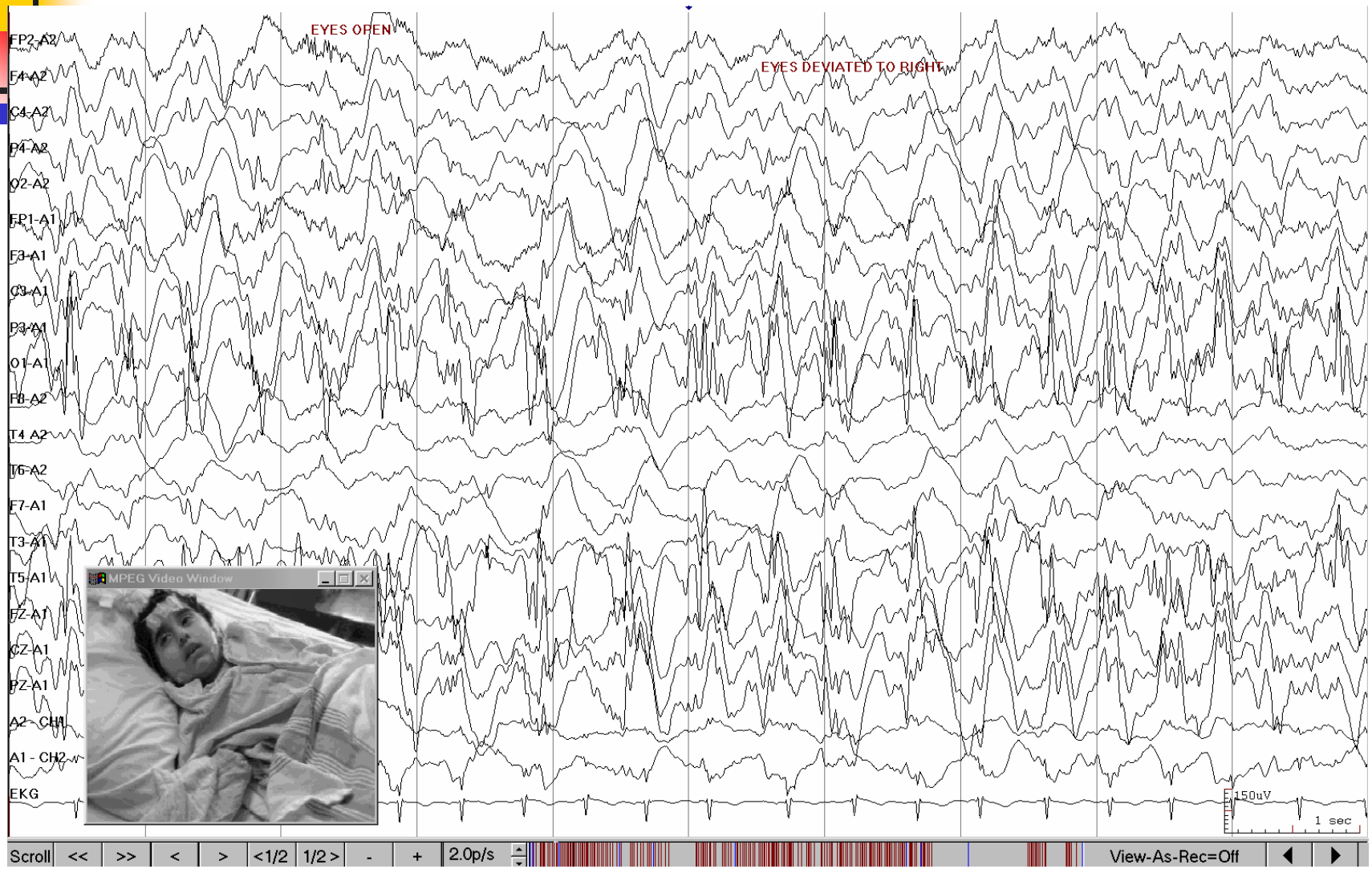
Symptomatic Occipital Epilepsy: Ictal onset



Evolution of Seizure



4 MINUTES INTO SEIZURE: EYES OPEN & TO THE RIGHT



Challenges in Occipital Lobe Epilepsy

- 66 children with OLE (BC Children's Hospital series – Schrader et al. submitted)
- 21 Symptomatic
- 12 Probable symptomatic
- 33 idiopathic
  - Panayiotopoulus syndrome (n=9)
  - Gastaut syndrome (n=12)
  - Overlap (n=11)
  - Idiopathic OLE with photosensitive epilepsy (n=1)

BC Children's Hospital Series

- **Predictors of abnormal MRI**
  - **Early age of seizure onset**
  - **Abnormal neurological examination**
  - **No difference in clinical semiology between idiopathic and symptomatic group**

Key points

- Benign rolandic epilepsy is most common partial epilepsy in children
- Rolandic spikes may occur in children without epilepsy
- Rolandic spikes may occur in symptomatic epilepsies
- Cognitive and behaviour changes may occur with interictal spikes

Key Points

- 3 variants of idiopathic epilepsy with occipital or posterior epileptiform discharges
- Panayiotopoulos syndrome
  - is under recognized and now categorized as an autonomic epilepsy
  - rolandic spikes / epilepsy may develop
- Clinical features do not differentiate symptomatic and idiopathic occipital lobe epilepsy