4° CORSO RESIDENZIALE

EEG e POTENZIALI EVOCATI

22 – 27 NOVEMBRE 2021

Con il Patrocinio di

EEG nello *Status Epilepticus* peculiarità nel bambino

Gaetano Cantalupo

UNIVERSITÀ di VERONA

Sezione di NEUROPSICHIATRIA INFANTILE

Epilepsia, 6 (1965) 167–182

Treatment of Status Epilepticus with Diazepam (Valium)*

H. GASTAUT, R. NAQUET, R. POIRÉ AND C. A. TASSINARI

I. Generalized Status Epilepticus

A. Generalized convulsive status

(i) *Tonic–clonic status epilepticus* (grand mal status), or the classic status epilepticus of Calmeil (*2*) and Bourneville (*1*), with generalized tonic–clonic paroxysms repeated 1–5 times per hour, coma, progressive autonomic disturbances, and a fatal prognosis if not stopped quickly. We distinguish between primary tonic–clonic status with fits generalized from the start, and secondary tonic–clonic status with partial seizures secondarily generalized.

(ii) *Tonic status* usually occurring in childhood. Such attacks occur 4–20 times in an hour and may disappear or fade away until they can only be perceived on the EEG. Autonomic disturbances may increase and lead to death from asphyxia and circulatory collapse.

(iii) *Clonic status* only occurring in infancy as prolonged clonic attacks, lasting several hours or days and accompanied by coma.

(iv) *Myoclonic status* also occurring in childhood as continuously repeated fragmentary or massive myoclonus, lasting several hours or days with impaired consciousness of varying degree from subject to subject.

B. Non-convulsive generalized status epilepticus

This absence status is characterized by clouding of consciousness, which may be slight, with simple slowing of ideation and means of expression; or more marked and accompanied by confusion; or severe with drowsiness or even stupor. They may last for hours or days. These states correspond to what Lennox (*7*) suggests should be named *petit mal status*.

II. Unilateral Convulsive Status Epilepticus

These occur in young children, as hemiclonic attacks and may last for hours or days. They are always followed by hemiplegia, which fortunately usually passes off but may be permanent (H.H. syndrome), and sometimes may be accompanied by secondary epileptic attacks (H.H.E. syndrome).

III. Partial Status Epilepticus

This form is confined to adults with repetition at varying intervals of any type of partial epileptic seizures and usually presenting itself as Jacksonian motor status or jerks incessantly repeated in the same part of the body [epilepsia partialis continua of Kojewnikoff (*5*)].

50 years

Epilepsia, 56(10):1515–1523, 2015

A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus

*†‡Eugen Trinka, §Hannah Cock, ¶Dale Hesdorffer, #Andrea O. Rossetti, **Ingrid E. Scheffer, ††Shlomo Shinnar, ‡‡Simon Shorvon, and §§Daniel H. Lowenstein

Table 2. Axis 1: Classification of status epilepticus (SE)

(A) With prominent motor symptoms
- A.1 Convulsive SE (CSE, synonym: tonic–clonic SE)
  - A.1.a. Generalized convulsive
  - A.1.b. Focal onset evolving into bilateral convulsive SE
  - A.1.c. Unknown whether focal or generalized
- A.2 Myoclonic SE (prominent epileptic myoclonic jerks)
  - A.2.a. With coma
  - A.2.b. Without coma
- A.3 Focal motor
  - A.3.a. Repeated focal motor seizures (Jacksonian)
  - A.3.b. Epilepsia partialis continua (EPC)
  - A.3.c. Adversive status
  - A.3.d. Oculoclonic status
  - A.3.e. Ictal paresis (i.e., focal inhibitory SE)
- A.4 Tonic status
- A.5 Hyperkinetic SE

(B) Without prominent motor symptoms (i.e., nonconvulsive SE, NCSE)
- B.1 NCSE with coma (including so-called "subtle" SE)
- B.2 NCSE without coma
  - B.2.a. Generalized
    - B.2.a.a Typical absence status
    - B.2.a.b Atypical absence status
    - B.2.a.c Myoclonic absence status
  - B.2.b. Focal
    - B.2.b.a Without impairment of consciousness (aura continua, with autonomic, sensory, visual, olfactory, gustatory, emotional/psychic/experiential, or auditory symptoms)
    - B.2.b.b Aphasic status
    - B.2.b.c With impaired consciousness
  - B.2.c Unknown whether focal or generalized
  - B.2.c.a Autonomic SE

four axes:
1. Semiology
2. Etiology
3. EEG correlates
4. Age

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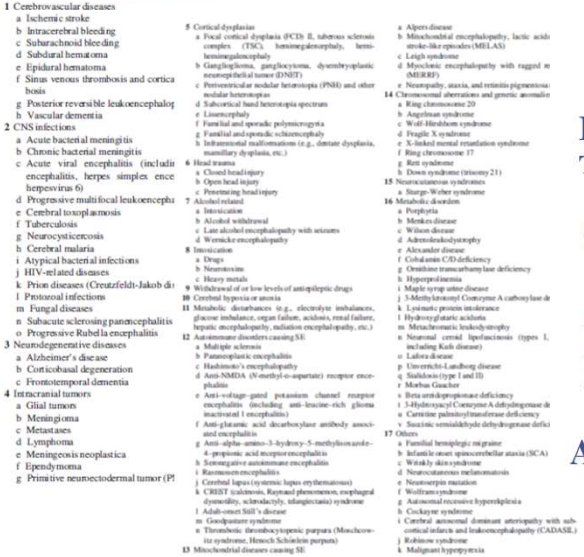
Axis 2: Etiology

Appendix 1: List of Etiologies That May Cause Status Epilepticus (Not Exhaustive)



1 Cerebrovascular diseases
a Ischemic stroke
b Intracerebral bleeding
c Subarachnoid bleeding
d Subdural hematoma
e Epidural hematoma
f Sinus venous thrombosis and cortica bosis
g Posterior reversible leukoencephalop
h Vascular dementia
2 CNS infections
a Acute bacterial meningitis
b Chronic bacterial meningitis
c Acute viral encephalitis (includir encephalitis, herpes simplex ence herpesvirus 6)
d Progressive multifocal leukoencepha
e Cerebral toxoplasmosis
f Tuberculosis
g Neurocysticercosis
h Cerebral malaria
i Atypical bacterial infections
j HIV-related diseases
k Prion diseases (Creutzfeldt-Jakob di
l Protozoal infections
m Fungal diseases
n Subacute sclerosing panencephalitis
o Progressive Rubella encephalitis
3 Neurodegenerative diseases
a Alzheimer's disease
b Corticobasal degeneration
c Frontotemporal dementia
4 Intracranial tumors
a Glial tumors
b Meningioma
c Metastases
d Lymphoma
e Meningeosis neoplastica
f Ependymoma
g Primitive neuroectodermal tumor (PI
5 Cortical dysplasias
a Focal cortical dysplasia (FCD: II, tuberous sclerosis complex (TSC), hemimegalencephaly, hemiheterotopia, hemimegalencephaly
b Ganglioglioma, gangliocytoma, dysembryoplastic neuroepithelial tumor (DNET)
c Periventricular nodular heterotopia (PNH) and other nodular heterotopias
d Subcortical band heterotopia spectrum
e Lissencephaly
f Familial and sporadic polymicrogyria
g Familial and sporadic schizencephaly
h Infratentorial malformations (e.g., dentate dysplasia, mamillary dysplasia, etc.)
6 Head trauma
a Closed head injury
b Open head injury
c Penetrating head injury
7 Alcohol related
a Intoxication
b Alcohol withdrawal
c Late alcohol encephalopathy with seizures
d Wernicke encephalopathy
8 Intoxication
a Drugs
b Neurotoxins
c Heavy metals
9 Withdrawal of or low levels of antiepileptic drugs
10 Cerebral hypoxia or anoxia
11 Metabolic disturbances (e.g., electrolyte imbalances, glucose imbalance, organ failure, acidosis, renal failure, hepatic encephalopathy, radiation encephalopathy, etc.)
12 Autoimmune disorders causing SE
a Multiple sclerosis
b Paraneoplastic encephalitis
c Hashimoto's encephalopathy
d Anti-NMDA (N-methyl-D-aspartate) receptor encephalitis
e Anti-voltage-gated potassium channel receptor encephalitis (including anti-leucine-rich glioma inactivated 1 encephalitis)
f Anti-glutamic acid decarboxylase antibody associated encephalitis
g Anti-alpha-amino-3-hydroxy-5-methylisoxazole-4-propionic acid receptor encephalitis
h Seronegative autoimmune encephalitis
i Rasmussen encephalitis
j Cerebral lupus (systemic lupus erythematosus)
k CREST (scleroosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, telangiectasia) syndrome
l Adult-onset Still's disease
m Goodpasture syndrome
n Thrombotic thrombocytopenic purpura (Moschcowitz syndrome, Henoch Schönlein purpura)
13 Mitochondrial diseases causing SE
a Alpers disease
b Mitochondrial encephalopathy, lactic acido stroke-like episodes (MELAS)
c Leigh syndrome
d Myoclonic encephalopathy with ragged re (MERRF)
e Neuropathy, ataxia, and retinitis pigmentosa
14 Chromosomal aberrations and genetic anomalie
a Ring chromosome 20
b Angelman syndrome
c Wolf-Hirshhorn syndrome
d Fragile X syndrome
e X-linked mental retardation syndrome
f Ring chromosome 17
g Rett syndrome
h Down syndrome (trisomy 21)
15 Neurocutaneous syndromes
a Sturge-Weber syndrome
16 Metabolic disorders
a Porphyria
b Menkes disease
c Wilson disease
d Adrenoleukodystrophy
e Alexander disease
f Cobalamin C/D deficiency
g Ornithine transcarbamylase deficiency
h Hyperprolinemia
i Maple syrup urine disease
j 3-Methylcrotonyl Coenzyme A carboxylase de
k Lysinuric protein intolerance
l Hydroxyglutaric aciduria
m Metachromatic leukodystrophy
n Neuronal ceroid lipofuscinosis (types 1, including Kufs disease)
o Lafora disease
p Unverricht-Lundborg disease
q Sialidosis (type I and II)
r Morbus Gaucher
s Beta ureidopropionase deficiency
t 3-Hydroxyacyl Coenzyme A dehydrogenase de
u Carnitine palmitoyltransferase deficiency
v Succinic semialdehyde dehydrogenase defici
17 Others
a Familial hemiplegic migraine
b Infantile onset spinocerebellar ataxia (SCA)
c Wrinkly skin syndrome
d Neurocutaneous melanomatosis
e Neuroserpin mutation
f Wolfram syndrome
g Autosomal recessive hyperekplesia
h Cockayne syndrome
i Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL)
j Robinow syndrome
k Malignant hyperpyrexia

Appendix 2: List of Specific Associations in Which SE Is an Integral Part of the Syndrome, the Entity, or Is a Symptom with Strong Clinical Implications (List Is Incomplete and Will Be Elaborated)

Absence status in Ring chromosome 20 syndrome.
Angelman syndrome.
Absence status epilepsy. [37]

Appendix 3: Previous Definitions

Axis 3: Electroencephalographic correlates

None of the ictal EEG patterns of any type of SE is specific. Epileptiform discharges are regarded as the hallmark, but with increasing duration of SE, the EEG changes and rhythmic nonepileptiform patterns may prevail. Similar EEG patterns, such as triphasic waves, can be recorded in various pathologic conditions, leading to substantial confusion in the literature. Although the EEG is overloaded with movement

A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus

*†‡Eugen Trinka, §Hannah Cock, ¶Dale Hesdorffer, #Andrea O. Rossetti, **Ingrid E. Scheffer, ††Shlomo Shinnar, ‡‡Simon Shorvon, and §§Daniel H. Lowenstein

Epilepsia, 56(10):1515–1523, 2015
doi: 10.1111/epi.13121

Table 5. SE in selected electroclinical syndromes according to age

SE occurring in neonatal and infantile-onset epilepsy syndromes
- Tonic status (e.g., in Ohtahara syndrome or West syndrome)
- Myoclonic status in Dravet syndrome
- Focal status
- Febrile SE

SE occurring mainly in childhood and adolescence
- Autonomic SE in early-onset benign childhood occipital epilepsy (Panayiotopoulos syndrome)
- NCSE in specific childhood epilepsy syndromes and etiologies (e.g., Ring chromosome 20 and other karyotype abnormalities, Angelman syndrome, epilepsy with myoclonic-atonic seizures, other childhood myoclonic encephalopathies; see Appendices 1–3)
- Tonic status in Lennox-Gastaut syndrome
- Myoclonic status in progressive myoclonus epilepsies
- Electrical status epilepticus in slow wave sleep (ESES)
- Aphasic status in Landau-Kleffner syndrome

SE occurring mainly in adolescence and adulthood
- Myoclonic status in juvenile myoclonic epilepsy
- Absence status in juvenile absence epilepsy
- Myoclonic status in Down syndrome

SE occurring mainly in the elderly
- Myoclonic status in Alzheimer's disease
- Nonconvulsive status epilepticus in Creutzfeldt-Jakob disease
- De novo (or relapsing) absence status of later life

These forms of SE may be encountered prevalently in some age groups, but not exclusively.

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Epileptic Disord 2005; 7 (3): 253-96

Nonconvulsive status epilepticus: Epilepsy Research Foundation Workshop Reports

Matthew Walker, Helen Cross, Shelagh Smith, Camilla Young, Jean Aicardi, Richard Appleton, Sarah Aylett, Frank Besag, Hannah Cock, Robert DeLorenzo, Franck Drislane, John Duncan, Colin Ferrie, Denson Fujikawa, William Gray, Peter Kaplan, Micheal Koutroumanidis, Mary O'Regan, Perrine Plouin, Josemir Sander, Rod Scott, Simon Shorvon, David Treiman, Claude Wasterlain, Udo Wieshmann

Proposed definition

«Nonconvulsive status epilepticus is a term used to denote a range of conditions in which electrographic seizure activity is prolonged and results in nonconvulsive clinical symptoms.»

Table 1. Classification scheme for NCSE.

NCSE in the neonatal period and infancy
- Neonatal NCSE
- NCSE in neonatal and infantile epilepsy syndromes
  - West Syndrome
  - Ohtahara syndrome
  - Severe myoclonic encephalopathies of infancy
  - Benign neonatal seizures (and benign familial neonatal seizures)
  - NCSE in other early neonatal and infantile epilepsies

NCSE in childhood
- NCSE in benign focal childhood epilepsy syndromes
- NCSE (often specific forms) in severe childhood epileptic encephalopathies/syndromes
  - Electrical status epilepticus in sleep (ESES)
  - Landau Kleffner Syndrome
  - NCSE in Dravet's syndrome
  - NCSE in Ring Chromosome X
  - NCSE in myoclonic syndromes of childhood
  - NCSE in Angelman's syndrome
  - Severe myoclonic encephalopathies of childhood
  - Myoclonic-astatic epilepsy

NCSE in childhood and adult life
- NCSE in the severe epileptic encephalopathies/syndromes (atypical absence and other forms of NCSE)
  - Lennox Gastaut syndrome
  - Other childhood epileptic encephalopathies
- NCSE in acute cerebral injury
  - Acute confusional states (including acute symptomatic partial SE)
  - NCSE in coma (including myoclonic status epilepticus in coma)
- NCSE in patients with epilepsy but without encephalopathy
  - Simple partial NCSE
    EPC and non-motor forms of simple partial NCSE
  - Complex partial status epilepticus
  - Absence status epilepticus in idiopathic generalised epilepsies
    Panyotopoulos syndrome, EMA, JME
  - Myoclonic status epilepticus in idiopathic generalised epilepsy
  - NCSE in the postictal phase of tonic clonic seizures
  - NCSE inpatients without epileptic encephalopathy/acute cerebral injury, which take the form of cognitive impairment or confusion, and which do not conform to the categories of simple or complex partial SE

Status epilepticus confined to adult life
- *De novo* absence status epilepticus of late onset

Boundary syndromes
- Cases with epileptic encephalopathy in whom it is not clear to what extent electrographic seizure activity is contributing to the clinical impairment
- Cases with acute brain injury in whom it is not clear to what extent electrographic seizure activity is contributing to the clinical impairment
- Cases with behavioural disturbances/psychosis in whom it is not clear to what extent electrographic seizure activity is contributing the clinical impairment

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Neonatal period/Infancy

- **NEONATAL SE DUE TO ACUTE INJURY**
- **SE IN NEONATAL-ONSET EPILEPSIES**
- **NEONATAL SYNDROMES DEFINED BY THE PRESENCE OF SE**
  - **EPILEPSY OF INFANCY WITH MIGRATING FOCAL SEIZURES**
  - **SELF-LIMITED (FAMILIAL) NEONATAL SEIZURES/EPILEPSY (*WATANABE-VIGEVANO SYNDROME*)**
  - **EARLY MYOCLONIC ENCEPHALOPATHY**
  - ***OHTAHARA SYNDROME***

ELSEVIER

Semin Pediatr Neurol 17:163-168; 2010

Seminars in Pediatric Neurology

2010

Neonatal Status Epilepticus

Russell Lawrence, MD,*,† and Terrie Inder, MD*,‡

Definition

….. One of the challenges in discussing neonatal SE is the lack of a definitive definition that is relevant to the newborn brain……

Neonatal seizures are also more difficult to diagnose clinically with many subtle signs not recognized at the bedside…….

Neonatal seizures **commonly occur in the setting of encephalopathy (85%)**; thus, it is challenging to recognize any return to baseline mental status between seizures…….

Thus, because of a current lack of clear consensus on the term “status epilepticus” in the newborn brain, we avoid this term in favour of a focus on recurrent and prolonged seizure activity in this population.

INVITED REVIEW

2012

J Clin Neurophysiol 2012;29: 441–448

Neonatal Seizures and Status Epilepticus

Nicholas S. Abend and Courtney J. Wusthoff†*

Key Words: **Seizures,** Neonate, Electroencephalography, Status epilepticus.

Summary: Neonatal seizures are common, often require EEG monitoring for diagnosis and management ……

A neonatal electrographic seizure is defined as a sudden, repetitive, evolving, and stereotyped event of abnormal electrographic pattern …….

The diagnosis of neonatal seizures relies heavily on the neurophysiologist's interpretation of EEG.

Diagnosing neonatal seizures can be quite challenging for two main reasons.
First, determining whether abnormal movements are the clinical manifestations of electrographic seizures is difficult.

Second, 80% to 90% of electrographic seizures do not have any associated clinical correlate and would not be identified without continuous EEG.

April 2013

INVITED REVIEW

J Clin Neurophysiol 2013;30: 106–114

Continuous EEG Monitoring in the Neonatal Intensive Care Unit

Bláthnaid McCoy† and Cecil D. Hahn*†*

slide from
Massimo Mastrangelo

Multichannel Conventional EEG (Routine or Continuous)
Amplitude-integrated EEG (aEEG)
Automated seizure detection algorithms (SDAs)

August 2013

Seminars in Fetal & Neonatal Medicine 18 (2013) 202–208

Contents lists available at SciVerse ScienceDirect

Seminars in Fetal & Neonatal Medicine

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

Monitoring neonatal seizures

Geraldine B. Boylan[a,*], Nathan J. Stevenson[a], Sampsa Vanhatalo[b]

Seizure burden in neonates can be very high, status epilepticus a frequent occurrence, and the majority of seizures do not have any clinical correlate.

2013

INVITED REVIEW

Diagnosing Neonatal Seizures and Status Epilepticus

Courtney J. Wusthoff

SEIZURE BURDEN AND STATUS EPILEPTICUS

When diagnosing neonatal seizures, and particularly when EEG monitoring investigates seizures over time, the clinical neurophysiologist is challenged to provide a **quantification of seizure burden**…...

While subjective terms such as "frequent" or "occasional" are sometimes used to describe seizure burden, these have no consistent definition between individuals and are less useful for communicating findings.

Therefore, an objective quantification of seizure burden is preferred, although various methods have been used. Counting the absolute number of seizures in a recording is imprecise because both seizure durations and recording time may vary widely. Increasingly popular is the use of the "ictal fraction" to quantify seizures (*Pisani et al., 2008*). This sums the overall duration of all seizures in an epoch as a fraction of total recording time (*Temko et al., 2011*).

doi:10.1093/brain/awu042 Brain 2014: 137; 1429–1438 | 1429

BRAIN
A JOURNAL OF NEUROLOGY

Seizure burden is independently associated with short term outcome in critically ill children

Eric T. Payne,[1] Xiu Yan Zhao,[2] Helena Frndova,[3] Kristin McBain,[3] Rohit Sharma,[1] James S. Hutchison[3] and Cecil D. Hahn[1]

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Massimo Mastrangelo

Seizures are common among critically ill children, but their relationship to outcome remains unclear. We sought to quantify the relationship between electrographic seizure burden and short-term neurological outcome, while controlling for diagnosis and illness severity.
We prospectively evaluated all infants and children admitted to our paediatric and cardiac intensive care units who **underwent clinically ordered continuous video-electroencephalography monitoring ……**

Seizure burden was quantified by calculating the **maximum percentage of any hour** that was **occupied by electrographic seizures.**

doi:10.1093/brain/awu042 Brain 2014: 137; 1429–1438 | 1429

BRAIN
A JOURNAL OF NEUROLOGY

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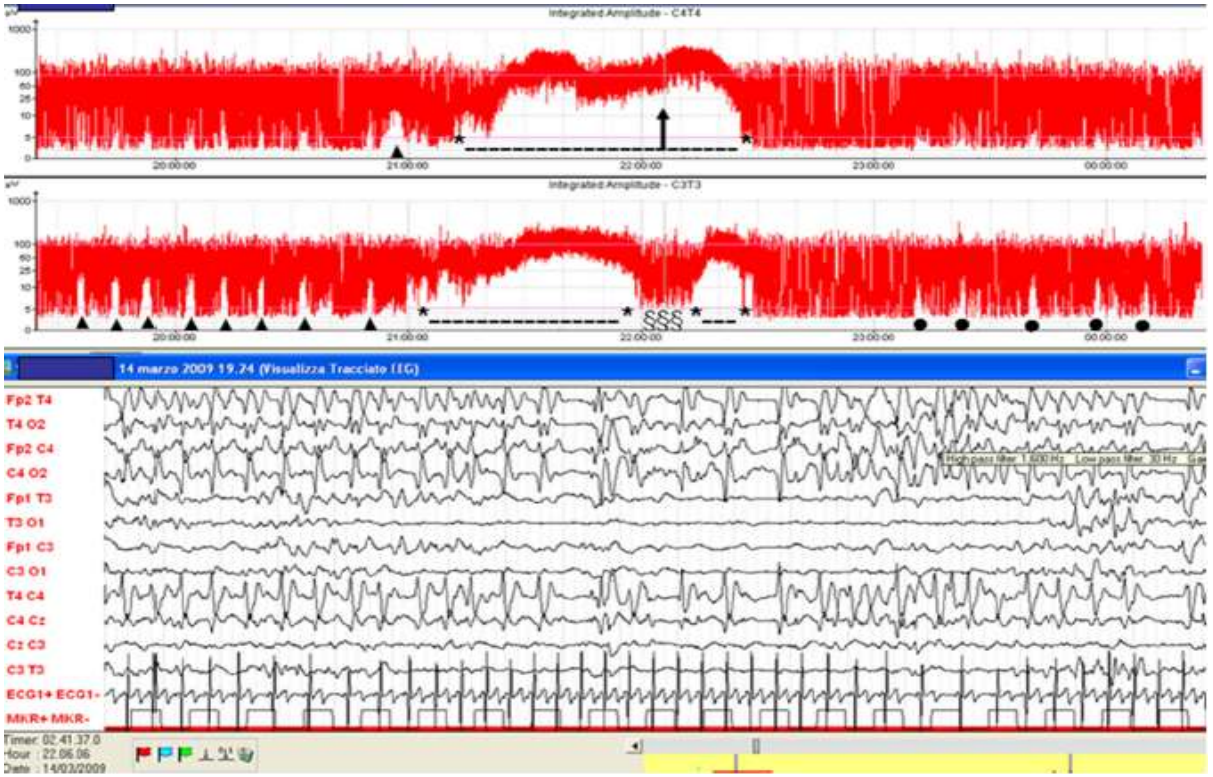
Massimo Mastrangelo

We conclude that in this cohort of critically ill children, increasing seizure burden was independently associated with a greater probability and magnitude of neurological decline.

Our observation that a **seizure burden of more than 12 min in a given hour** was strongly associated with neurological decline suggests that early antiepileptic drug management is warranted in this population, and identifies this seizure burden threshold as a potential therapeutic target.

NB: our study population represents a treated cohort: 81% of subjects received antiepileptic drugs before or during continuous video-EEG monitoring and 37% received at least three antiepileptic drugs.



‘‘Status epilepticus’’ (SE) aEEG pattern (*more than 1 discharge each 10 min of recording and/or a continuous discharges for equal or more than 15 min*): after 90 min with repetitive seizures (▲) on the left side (*less evident in the right hemisphere*), a continuous independent bilateral discharge occurs, lasting more than 60 min (*- - - - -*); at 22.06 SE persists on the right side (*–⬆—*) while stops in the left side (§§§), to recur here after 10 min with left flat EEG. Single seizures occur (●). The aEEG background pattern can be classified as ‘‘severely discontinuous normal without sleep–wake cycling’’.

‘‘Missed seizures’’ (MS) on aEEG recording (*#*), detected by cEEG traces in Cz-C3 (*——*): (i) the seizure lasts less than 15 s and (ii) the amplitude is less than 40 mcV. Ictal events of various lengths are recorded before and after the MS (~). The aEEG background pattern can be classified as ‘‘continuous extremely low voltage’’.

0031-3998/11/7005-0535
PEDIATRIC RESEARCH

Vol. 70, No. 5, 2011
Printed in U.S.A.


slide from
Massimo Mastrangelo

Risk Factors for Epilepsy in Children With Neonatal Encephalopathy

HANNAH C. GLASS, KAREN J. HONG, ELIZABETH E. ROGERS, RITA J. JEREMY, SONIA L. BONIFACIO, JOSEPH E. SULLIVAN, A. JAMES BARKOVICH, AND DONNA M. FERRIERO

In a multivariable analysis

adjusting for degree of encephalopathy and severe/near-total brain

injury, **status epilepticus** was independently associated with epilepsy.

lo SdM epilettico neonatale è dannoso, per cui:

- Va riconosciuto,
  - trattato
  - inquadrato
- Va seguito
  - monitoraggio aEEG/cEEG
- Va verificato l'effetto della terapia / terapie
  - Monitoraggio aEEG/cEEG
- Vanno verificati gli outcome (precoce e tardivo)
  - Mortalità
  - Epilessia post neonatale
  - Sviluppo psicomotorio

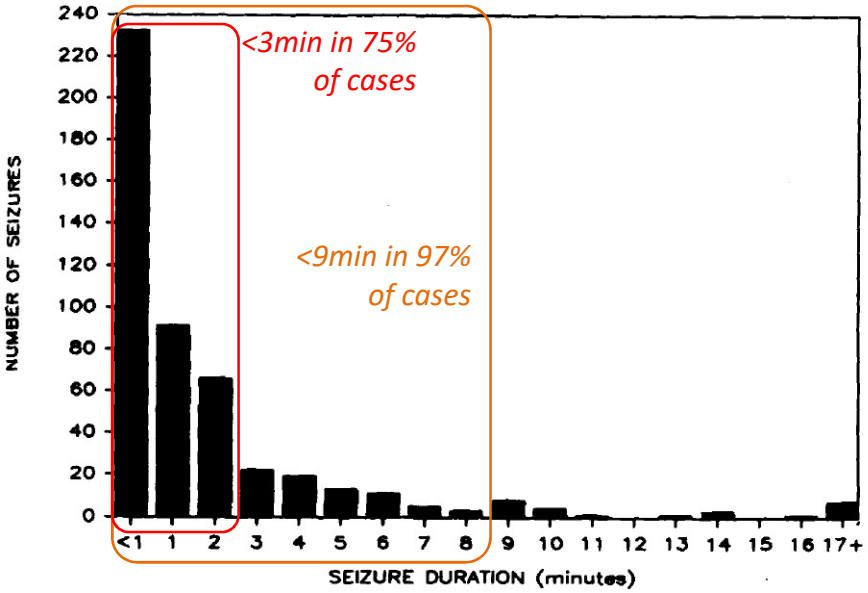
…… all'interno di condizioni epilettologiche omogenee di pazienti

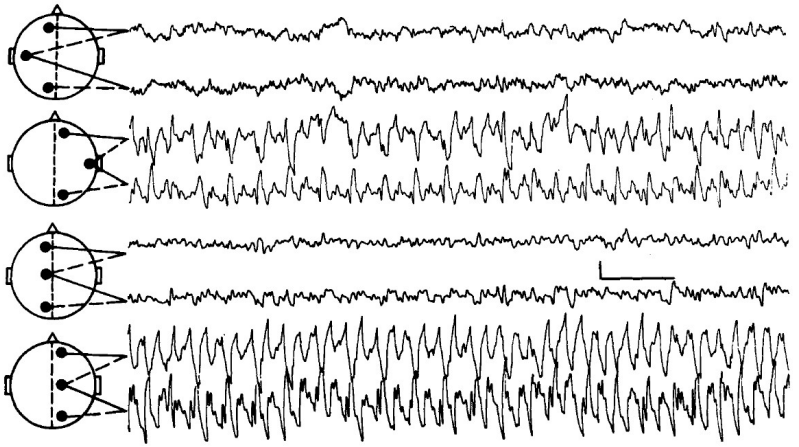
slide from
Massimo Mastrangelo

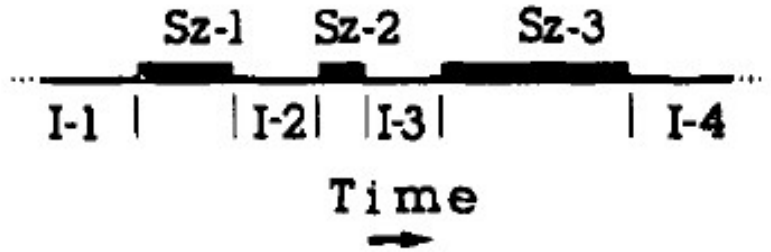
Epilepsia, 28(5):537–541, 1987
Raven Press, Ltd., New York

The Exact Ictal and Interictal Duration of Electroencephalographic Neonatal Seizures

Robert R. Clancy and Agustin Legido







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EUROPEAN JOURNAL OF PAEDIATRIC NEUROLOGY 19 (2015) 314–319

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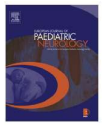

Orginal article

Neonatal status epilepticus: Differences between preterm and term newborns

Elena Pavlidis [a,*], *Carlotta Spagnoli* [a], *Annalisa Pelosi* [b], *Silvia Mazzotta* [a], *Francesco Pisani* [a]

5. Conclusions

Considering the existing data on NS duration and trend, and the adverse effects of prolonged and recurrent seizures on neonatal brain, it is advisable to adopt a shorter temporal criterion that allows early therapeutic intervention. It is also true that for NSE no standard treatment protocols exist, leading to a lack of a targeted intervention and no effective anticonvulsant drugs are available at the moment. However, before the right therapeutic approach, a correct definition of this clinical entity with a shorter temporal criterion would be the first step towards a proper treatment.

"repetition of clinical or subclinical seizures with abnormal interictal neurologic status, occurring for at least a **few hours**"

Monod et al. Arch Franc Pediatr 1969

"[(1) generalized high-voltage paroxysmal discharges OR (2) two or more discharges occurring every 10 s], persisting for at least **20 min**"

Mora et al. Clin Electroencephalogr 1984

"continuous seizure activity for at least **30 min** OR recurrent seizures for greater than or equal to 50% of the recording time (1-3 h)" [**30-90 min**].

Scher et al. Epilepsia 1993

"continuous EEG seizure-type activity in one or more channels for at least **4 h** or separated for only a short period by activity with frequent sharp waves or spikes"

Wertheim et al. Arch Dis Child 1994

"total seizure duration greater than **30 min** OR the sum of seizure duration and periodic discharges exceeded 50% of the EEG recording

Ortibus et al. Electroencephalogr Clin Neurophysiol 1996

(1) prolonged or frequently repeated seizures lasting more than **15 min**, AND (2) refractory to treatment with conventional anticonvulsants, (3) no response to glucose, Calcium and Magnesium (4) mechanical ventilation required during seizures, and (4) further therapeutic options for Midazolam or Lidocaine needed

Yamamoto et al. Brain Dev 2007

"repeated seizures longer than **1 hour**"

Co et al. Epilepsia 2007

EUROPEAN JOURNAL OF PAEDIATRIC NEUROLOGY 22 (2018) 757–762

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Official Journal of the European Paediatric Neurology Society

Review article

What is new: Talk about status epilepticus in the neonatal period

*Francesco Pisani, Elena Pavlidis**

Concerning the first time-point (t1), data show that the mean duration of seizures in neonates varies from a minimum of 2 min to a maximum of 19 min and some authors reported a maximum seizure duration of 6.3 min on average, being ≤ 9 min in 97% of cases and ≥30 min only in 0.4%.[12–16] However, differently from what recognized for older children,[17] a temporal duration after which a spontaneous resolution of the epileptic discharge is less probable has not yet been established in neonates.

For children >5 years old, an **operational definition** for convulsive SE has been proposed, referring to at least 5 min of continuous seizures or two or more discrete seizures between which there is incomplete recovery of consciousness.

Lowenstein. Epilepsia 1999

An operational definition should be applied also to neonates

Cross. Epileptic Disord 2014

operational definition:
when seizures are prolonged, recurrent and when they persist or recur during **5-15 min**

Mizrahi & Kellaway, 1998

AARHUS UNIVERSITY

Aarhus University Hospital
Department of Clinical Neurophysiology

Danish Epilepsy Centre
FILADELFIA

- **Seizures:**
  1. Electrographic:
     a. EDs ≥ 2.5 Hz (≥ 25 discharges /10s; ≥ 10s) or
     b. Evolving pattern (≥ 10s)
  3. Electroclinical:
     a. Time-locked clinical correlate (any duration) OR
     b. EEG and clinical improvement with an IV-AEDs
     (Only EEG improvement = possible NCSz /NCSE)

- **Status epilepticus**
     a. > 10 minutes *or*
     b. total duration of >20% (12 min) of any 60-minutes

Sandor.Beniczky@aarhus.rm.dk

(*J Clin Neurophysiol* 2013;30: 161–173)

INVITED REVIEW

American Clinical Neurophysiology Society Standardized EEG Terminology and Categorization for the Description of Continuous EEG Monitoring in Neonates: Report of the American Clinical Neurophysiology Society Critical Care Monitoring Committee

Tammy N. Tsuchida, Courtney J. Wusthoff,† Renée A. Shellhaas,‡ Nicholas S. Abend,§‖ Cecil D. Hahn,¶ Joseph E. Sullivan,# Sylvie Nguyen,** Steven Weinstein,* Mark S. Scher,†† James J. Riviello,‡‡ and Robert R. Clancy§‖*

TABLE 7. Seizures and Status Epilepticus

Seizures
Duration ≥10 seconds
Location
Diffuse)
Lateralized
Hemispheric: left and right
Focal
Frontal
Central
Temporal
Occipital
Vertex
Quadrant- anterior and posterior
Bilateral Independent
Multifocal
Migrating
Seizure Burden
Number of seizures per hour or
Summed duration of seizures divided by duration of epoch
Status epilepticus: summed duration of seizures totals ≥50% of a 1-hour epoch

ACNS – Terminology for cEEG in Neonates

Brief Rhythmic Discharges

This transient EEG pattern consists of evolving rhythmic patterns of electrical activity that share many characteristics with seizures but are very brief, with a duration of <10 seconds (Nagarajan et al., 2011; Oliveira et al., 2000; Shewmon, 1990). These have previously been alternatively described in the literature as BIRDs (brief ictal/interictal rhythmic/repetitive discharges) and BERDs (brief electrographic rhythmic discharges). Given that the true significance of these discharges is uncertain, the operational term "brief rhythmic discharges" (BRD) will be used here. They are usually seen in the context of an abnormal EEG background and/or confirmed electrographic seizures. Also, BRDs are rarely seen in isolation in a normal EEG. At this time, their pathologic significance is not fully understood. However, recent studies in adults suggested that clinical behavior changes can coexist with epileptiform discharges under 2 seconds in duration (D'Ambrosio et al., 2009). Similarly, a case series demonstrated similar mortality and neurologic disability for infants with BRDs as with seizures (Nagarajan et al., 2011). Further study is needed to better understand the basis and significance of BRD in the neonate.

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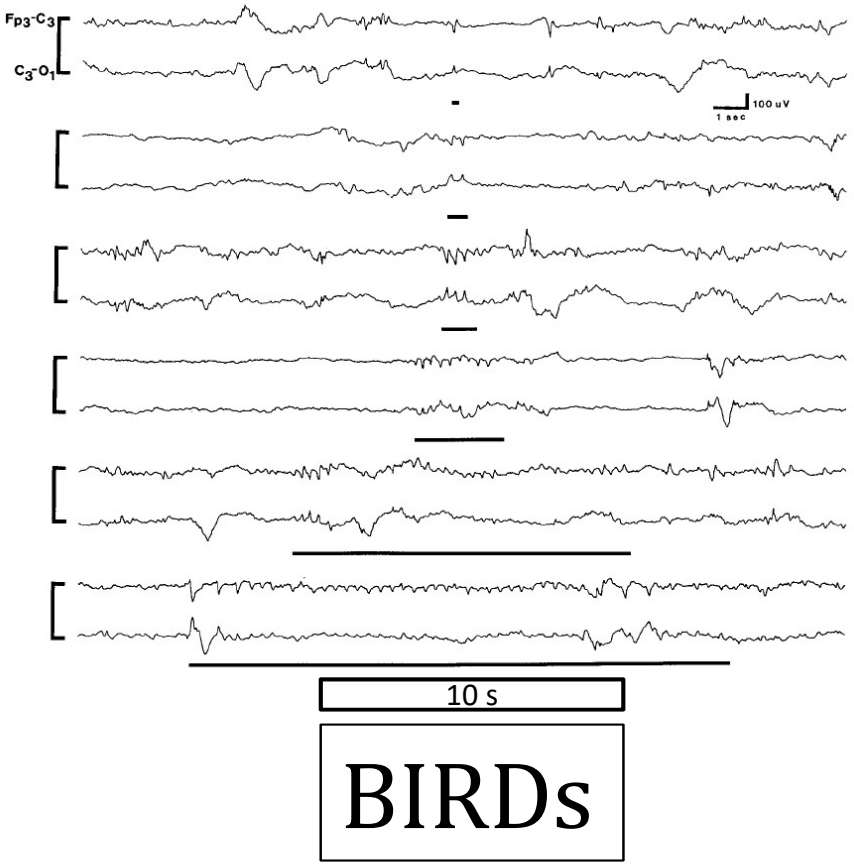
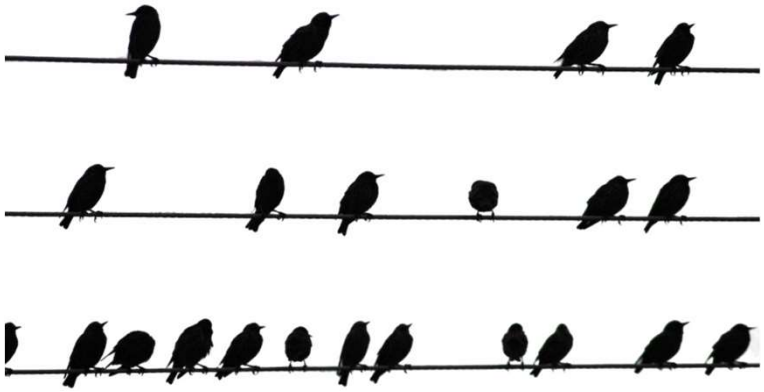


FIG. 7. Durational continuum of brief ictal rhythmic discharges (BIRDs) in a 1-month-old term infant with herpes encephalitis. Epochs are temporally distinct at the same pair of electrodes.

Journal of Clinical Neurophysiology
7(3):315–368, Raven Press, Ltd., New York

What Is a Neonatal Seizure? Problems in Definition and Quantification for Investigative and Clinical Purposes

D. Alan Shewmon



Over the years, this author has collected a number of neonatal EEGs demonstrating a continuum of durations of ictal discharges from a single waveform up to unequivocal (i.e., >10 s and evolving) seizures (Shewmon, 1983*a*). This phenomenon is illustrated in Figs. 7 and 8 and will be further illustrated in a manuscript now in preparation. Sometimes such discharges are called "larval seizures," but I prefer the term "brief ictal rhythmic discharge," which conveys more clearly that they are truly mini-seizures and not mere anlagen that must undergo metamorphosis before becoming seizures. The term is also advantagous for its acronymic properties (BIRD), in keeping with the traditional animal fetish of neurologists (viz., CAT scans, PET scans, and BAERs [sic]).

ELSEVIER

Clinical Neurophysiology 111 (2000) 1646–1653

www.elsevier.com/locate/clinph

Duration of rhythmic EEG patterns in neonates: new evidence for clinical and prognostic significance of brief rhythmic discharges

Andréa J. Oliveira[a,b], Magda L. Nunes[a,*], Lúcia M. Haertel[a], Fernando M. Reis[b], Jaderson C. da Costa[a,b]

Group	N	Abnormal neurodevelopmental outcome	Epilepsy	Death
No-RD	53	9 (17.0%)	8 (15.1%)	1 (1.9%)
BRD	29	14 (48.3%)**	5 (17.2%)	3 (10.3%)
LRD	31	12 (38.7%)*	7 (22.6%)	2 (6.5%)

[a] $*P < 0.05$, $**P < 0.01$ vs. No-RD (chi-square test).

Original Article

Journal of Child Neurology
26(12) 1529-1533

DOI: 10.1177/0883073811409750
http://jcn.sagepub.com
SAGE

Brief Electroencephalography Rhythmic Discharges (BERDs) in the Neonate With Seizures: Their Significance and Prognostic Implications

Lakshmi Nagarajan, MD[1,2,3], Linda Palumbo, BSc[1], and Soumya Ghosh, PhD[3]

Table 1 *(modified)*

Paroxysmal Rhythmic Activity on EEG	No. of Babies	Mortality	Neurologically Normal (survivors)
BERDs only	9	22.2%	28.6%
BERDs+EEG Szs	11	18.2%	22.2%
EEG Sz only	32	21.8%	36%
P values		.96	.73

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Neonatal period/Infancy

- **NEONATAL SE DUE TO ACUTE INJURY**
- **SE IN NEONATAL-ONSET EPILEPSIES**
- **NEONATAL SYNDROMES DEFINED BY THE PRESENCE OF SE**
  - **EPILEPSY OF INFANCY WITH MIGRATING FOCAL SEIZURES**
  - **SELF-LIMITED (FAMILIAL) NEONATAL SEIZURES/EPILEPSY**
  - **EARLY MYOCLONIC ENCEPHALOPATHY**
  - ***OHTAHARA SYNDROME***

EPILEPSY OF INFANCY WITH MIGRATING FOCAL SEIZURES

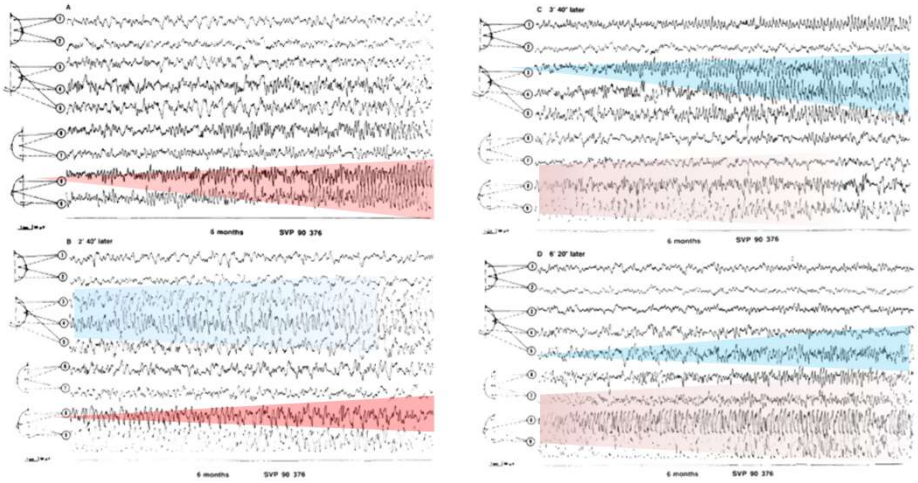
Epilepsia, 36(10):1017–1024, 1995

Migrating Partial Seizures in Infancy: A Malignant Disorder with Developmental Arrest

*G. Coppola, †‡P. Plouin, *‡C. Chiron, ‡O. Robain, and *‡O. Dulac

TABLE 1. *Various criteria of "Migrating Partial Seizures in Infancy" in the present series*

Major syndrome criteria	Number of infants Meeting Each Criteria
1. Normal development before first seizure	12 of 14
2. Onset of seizures before 6 months	13 of 14
3. Focal motor seizures at onset	10 of 14
4. Multifocal seizures becoming nearly continuous	14 of 14
5. Intractability to conventional AEDs	3 of 14 died 2 of 13 survivors seizure-free at 1 year 11 of 13 survivors at 1–5 years
6. Lack of definable etiology	14 of 14
7. Severe psychomotor delay	11 of 13 survivors at 1 year



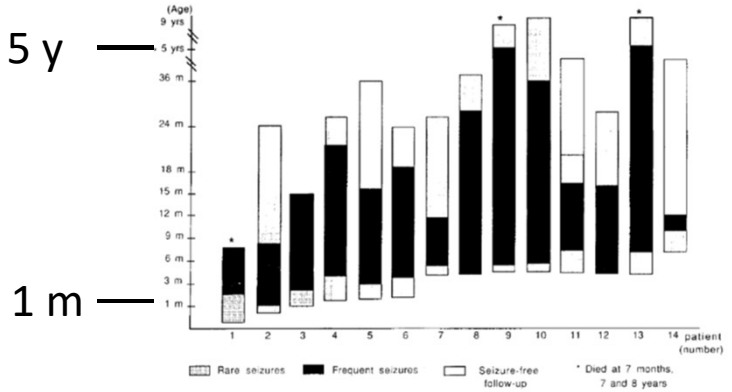


FIG. 1. Age of onset of seizures and evolution.

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Original Article

Journal of Child Neurology
Volume 23 Number 5
May 2008 497-506
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Migrating Focal Seizures in Infancy: Analysis of the Electroclinical Patterns in 17 Patients

Roberto Horacio Caraballo, MD, Elena Fontana, MD, Francesca Darra, MD, Laura Cassar, MD, Francesca Negrini, MD, Elena Fiorini, MD, Hugo Arroyo, MD, Stella Ferraro, MD, Natalio Fejerman, MD, and Bernardo Dalla Bernardina, MD

Table 1. Electroclinical Features, Imaging Findings, and Evolution of

Patient	Age at Onset (d)	Type of Seizures at Onset	CPMS	Electroclinical Patterns	Ictal EEG Finding				
1	40	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in F				
2	1	CFS	-	Independent and bilateral asynchronous	δ-θ activity in TO-R				
3	5	CFS-MC	-	Alternating, independent and bilateral asynchronous	Flattening, fast activ in FT-R				
4	60	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in F				
5	35	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in F				
6	29	CFS-MC	-	Alternating, independent and bilateral asynchronous	Flattening, fast activity in FT-R	-	Moderate increase CSF spaces	Hypotonia, microcephaly, SPR	seizures Died
7	18	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in RR	Yes	Moderate ventricular enlargement	Hypotonia, microcephaly, SPR	Daily seizures
8	58	CFS-MC	-	Alternating, independent and bilateral asynchronous	Flattening, fast activity in FT-R	Yes	Moderate ventricular enlargement	Hypotonia, MPR	Sporadic seizures
9	19	CFS	-	Independent and bilateral asynchronous	δ-θ activity in TO-R	Yes	MTS	Hypotonia, microcephaly, SPR	Sporadic seizures
10	46	CFS	-	Independent and bilateral asynchronous	δ-θ activity in TO-R	Yes	Normal	Hypotonia, microcephaly, MPR	Daily seizures
11	8	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in RR	-	Normal	Hypotonia, microcephaly, SPR	Died
12	8	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in RR	Yes	Mild ventricular enlargement	Hypotonia, microcephaly, SPR	Daily seizures
13	59	CFS-MC	-	Alternating, independent and bilateral asynchronous	Flattening, fast activity in FT-R	Yes	Mild increase CSF spaces	Hypotonia, microcephaly, SPR	Daily seizures
14	48	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in RR	Yes	Moderate increase CSF spaces	Hypotonia, microcephaly, SPR	Seizure free
15	38	CFS	-	Independent and bilateral asynchronous	δ-θ activity in TO-R	Yes	Mild increase CSF spaces	Microcephaly SPR	Daily seizures
16	52	FMS	Yes	Independent and bilateral asynchronous	Spikes and S-W in RR	Yes	Mild ventricular enlargement	Hypotonia, microcephaly, SPR	Daily seizures
17	55	CFS	-	Independent and bilateral asynchronous	δ-θ activity in TO-R	Yes	Moderate ventricular enlargement	Hypotonia, SPR	Died

NOTE: FMS = focal motor seizures; CFS = complex focal seizures; MC = motor component; CPMS = continuous partial motor seizures; S-W = sharp waves; RR = Rolandic region; TO-R = temporo-occipital region; FT-R = frontotemporal region; SE = status epilepticus; MTS = mesial temporal sclerosis; SPR = severe psychomotor retardation; MPR = moderate psychomotor retardation; EEG = electroencephalography; PND = paroxysmal nocturnal dyspnea; CSF = cerebrospinal fluid.

7/17 neonatal onset

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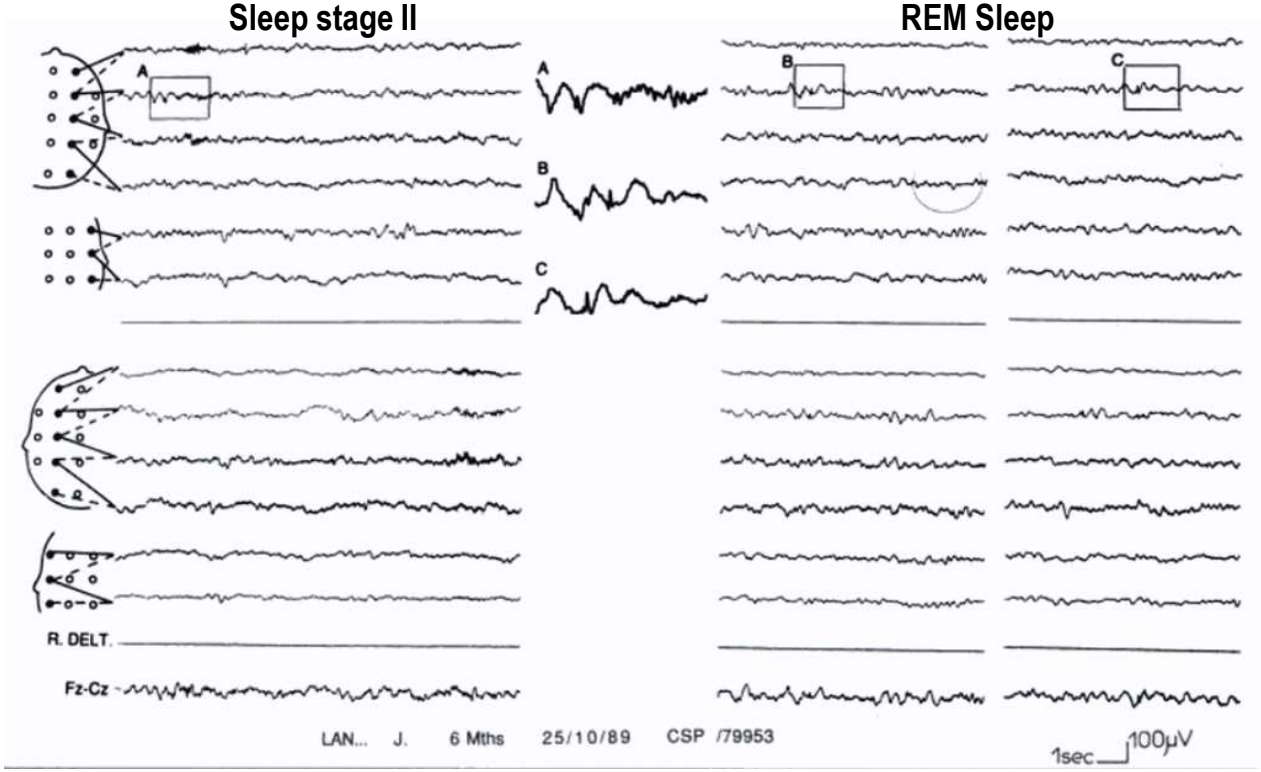
SELF-LIMITED (FAMILIAL) NEONATAL SEIZURES/EPILEPSY

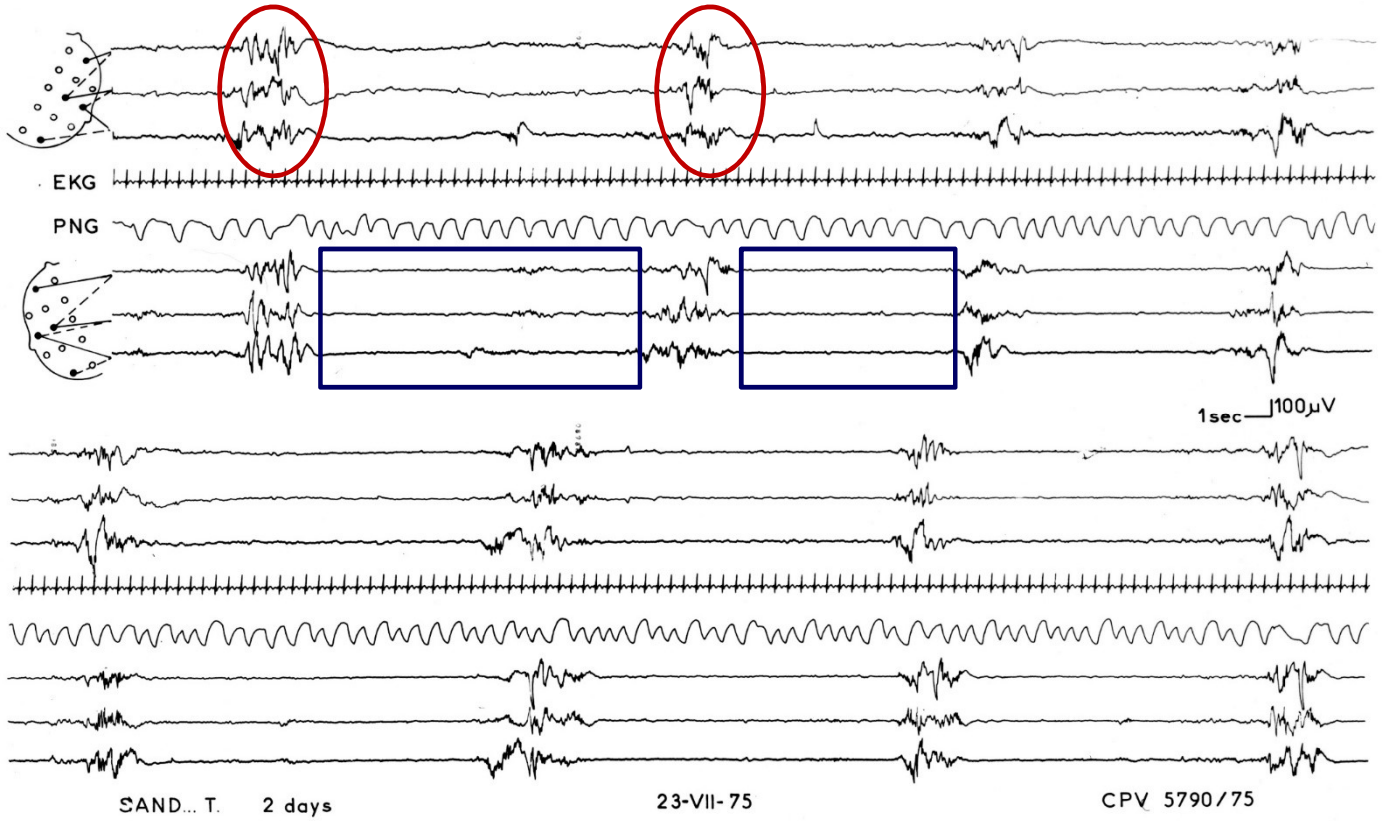
Epileptic Syndromes in Infancy, Childhood and Adolescence *(4th edn)*, *J. Roger, M. Bureau, Ch. Dravet, P. Genton, C.A. Tassinari & P. Wolf*
©2005 John Libbey Eurotext Ltd, pp. 171–179.



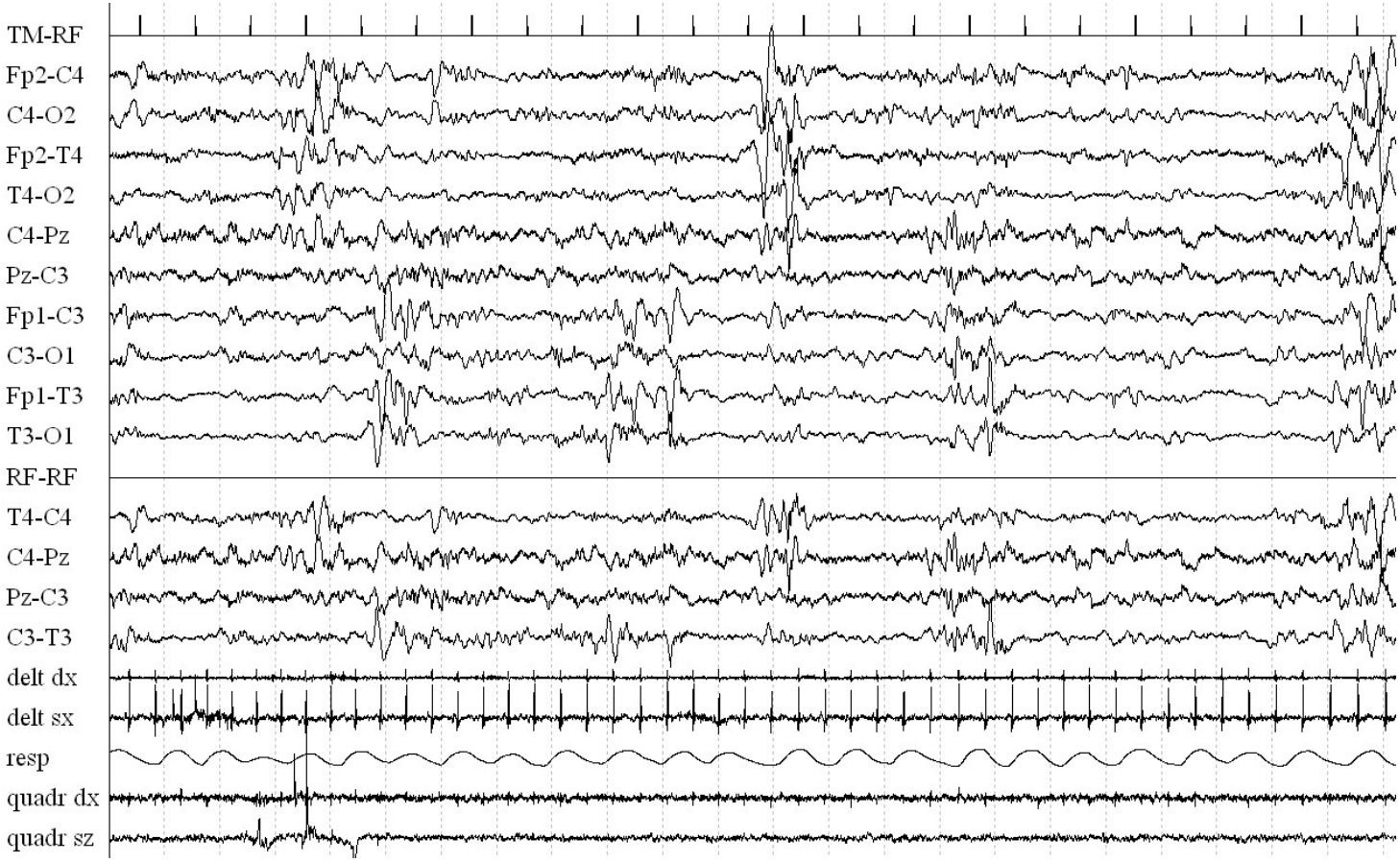
Idiopathic and/or benign localization-related epilepsies in infants and young children

Federico VIGEVANO and Michelle BUREAU*

SUPPRESSION-BURST



Early Myoclonic Encephalopathy (EME)


TM-RF
Fp2-C4
C4-O2
Fp2-T4
T4-O2
C4-Pz
Pz-C3
Fp1-C3
C3-O1
Fp1-T3
T3-O1
RF-RF
T4-C4
C4-Pz
Pz-C3
C3-T3
delt dx
delt sx
resp
quadr dx
quadr sz

Early Infantile Epileptic Encephalopathy (EIEE)
or *OHTAHARA SYNDROME*

Infancy/Childhood

- **FEBRILE SE IN**
  - **OTHERWISE NORMAL CHILDREN**
  - ***DRAVET SYNDROME***
  - **PCDH19-RELATED EPILEPSY**
  - **(F.I.R.E.S.)**
- **EPILEPSY SYNDROMES WITH *CHARACTERISTIC* SE TYPES**
  - ***SELF-LIMITED EPILEPSY WITH AUTONOMIC SEIZURES (PANAYIOTOPOULOS SYNDROME)***
  - ***MYOCLONIC-ATONIC EPILEPSY (DOOSE SYNDROME)***
  - ***LENNOX-GASTAUT SYNDROME***
  - **RING20-RELATED EPILEPSY**
  - **ATYPICAL SELF-LIMITED FOCAL EPILEPSIES OF CHILDHOOD**
- **EPILEPSY SYNDROMES DEFINED BY THE PRESENCE OF SE**
  - **ENCEPHALOPATHY with STATUS EPILEPTICUS during SLEEP (*TASSINARI ENCEPHALOPATHY*)**
  - **MYOCLONIC STATUS IN NON-PROGRESSIVE ENCEPHALOPATHIES (*DALLA BERNARDINA SYNDROME*)**

FEBRILE SE

Epilepsia, **37**(1):31–35, 1996

Prolonged Nonepileptic Twilight State with Convulsive Manifestations After Febrile Convulsions: A Clinical and Electroencephalographic Study

Naoki Yamamoto

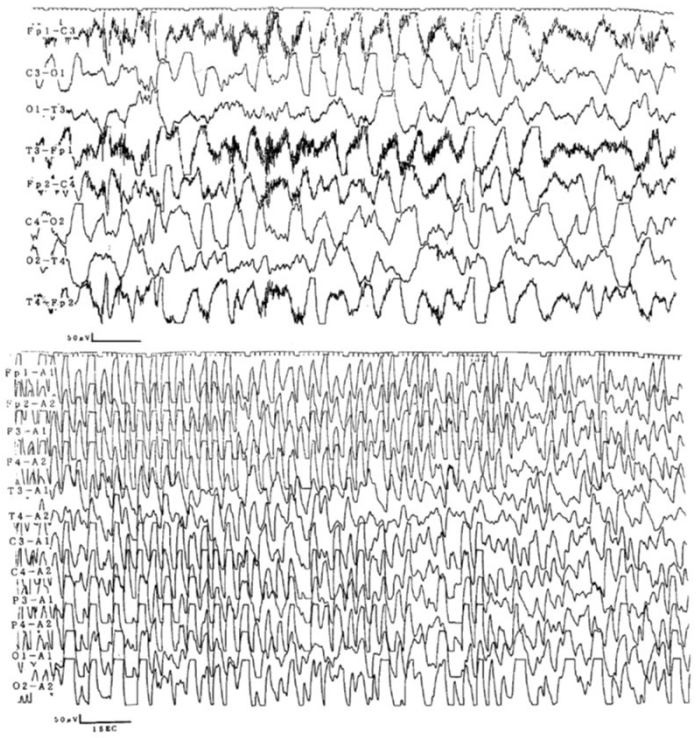
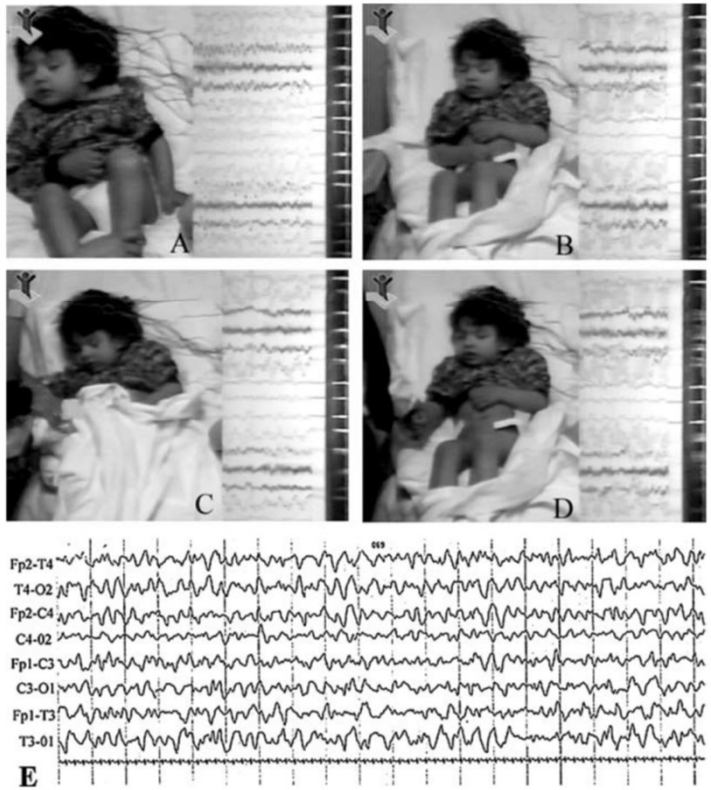


FIG. 2. *Patient 8.* Nonepileptic twilight states with convulsive manifestations (NETC). Diffuse rhythmic theta activity is evident. Clinically, consciousness was impaired, limb tone was increased, and upward eye turning was noted.

Epilepsia, **47**(6):1079–1081, 2006

Occurrence of a Prolonged Nonepileptic Motor Status after a Febrile Seizure

*Nicola Specchio, *Raffaella Cusmai, †Josiv Volkov, ‡Paolo Montaldo, and *Federico Vigevano



ORIGINAL ARTICLE

Non-convulsive febrile status epilepticus mimicking the postictal state after a febrile seizure: ictal electroclinical and evolutive study

Jacopo Proietti[1,2], Elena Fiorini[1,3], Lisa Meneghello[1,4], Gaetano Cantalupo[1,3,5], Elena Fontana[1,3], Tommaso Lo Barco[5,6], Bernardo Dalla Bernardina[3], Francesca Darra[1,3,5]

Epileptic Disorders *in press*

DRAVET SYNDROME

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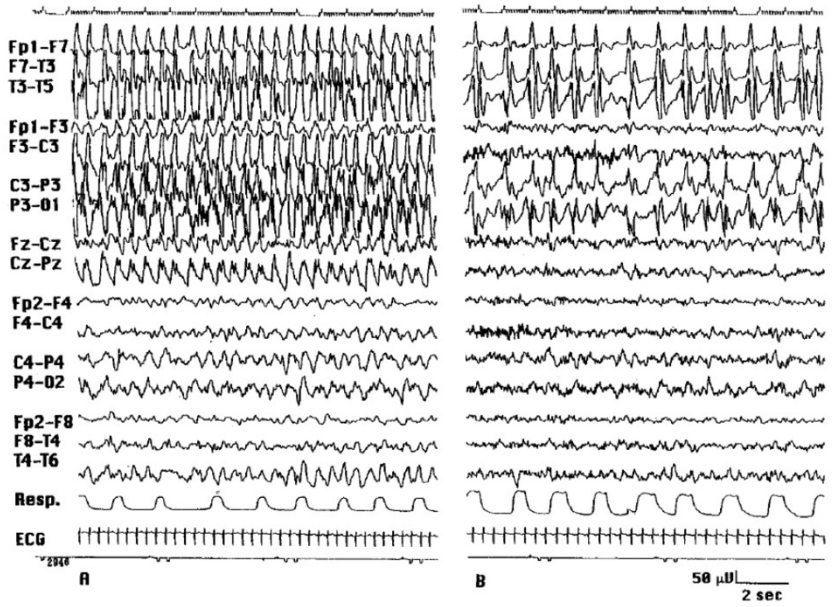
Epilepsia, 37(10):1020–1022, 1996
Lippincott-Raven Publishers, Philadelphia

Brief Communication

''Obtundation Status (Dravet)'' Caused by Complex Partial Status Epilepticus in a Patient with Severe Myoclonic Epilepsy in Infancy

Shuji Wakai, Masami Ikehata, Hiroshi Nihira, Nozomi Ito, Hirofumi Sueoka,
*Yoshitaka Kawamoto, *Hiroyuki Hayasaka, and Shunzo Chiba

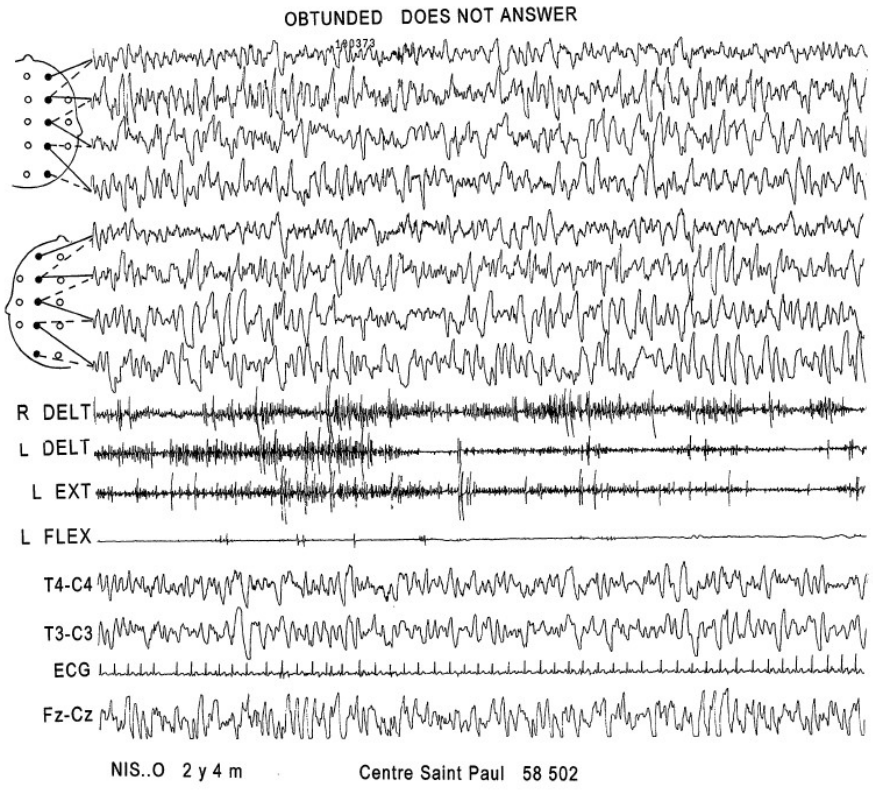


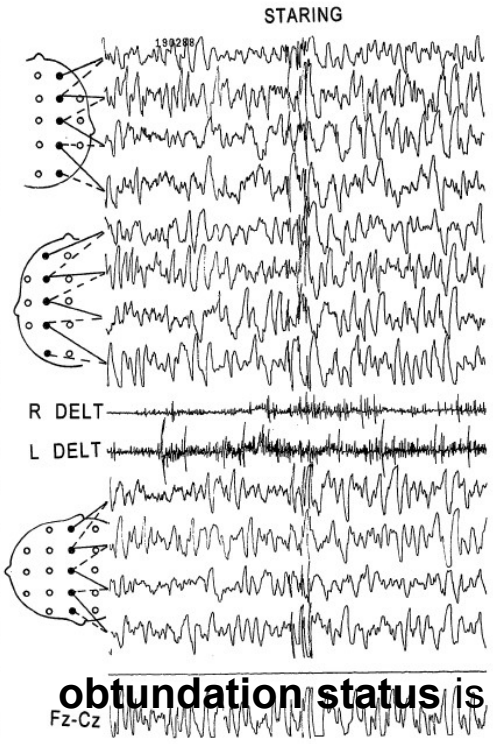
obtundation status is caused either by

(a) complex partial status

(b) atypical absence status

(c) a type of non-convulsive epileptic seizure related to *"post-grand mal exhaustion"*





Dravet C, Bureau M, Roger J. Severe myoclonic epilepsy in infants. In: Roger J, Bureau M, Dravet C, Dreifuss FE, Perret A, Wolf P, editors. Epileptic syndromes in infancy, childhood and adolescence, 2nd ed.. London: John Libbey, 1992. pp. 75–88.

obtundation status is caused either by

(a) complex partial status

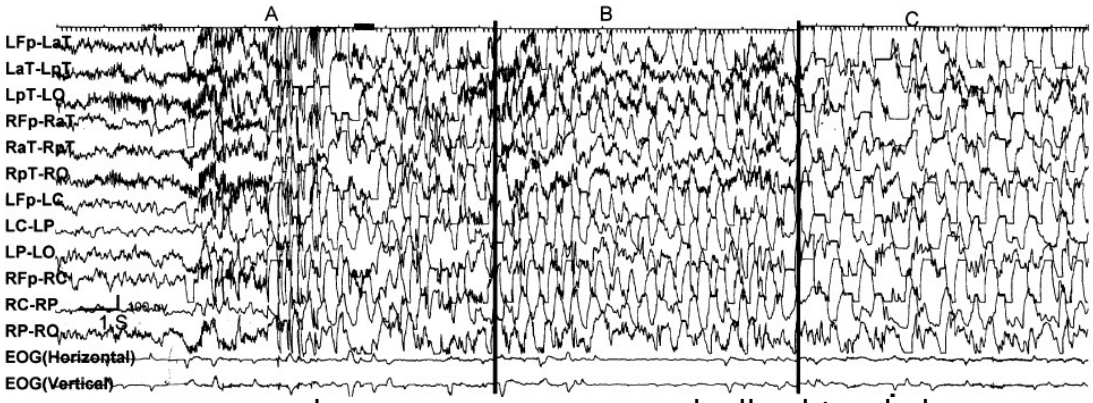
(b) atypical absence status

(c) a type of non-convulsive epileptic seizure related to *"post-grand mal exhaustion"*

Brain & Development 23 (2001) 736–748

Severe myoclonic epilepsy in infants – a review based on the Tokyo Women's Medical University series of 84 cases

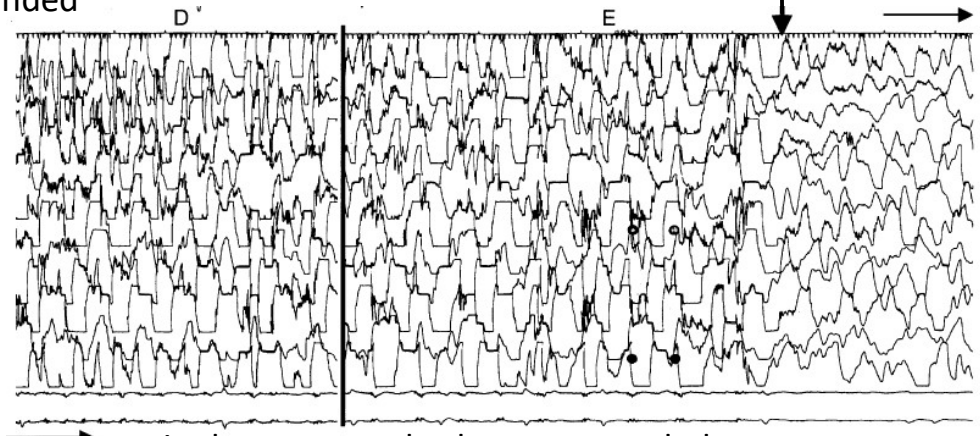
Hirokazu Oguni[a,*], Kitami Hayashi[a], Yutaka Awaya[b], Yukio Fukuyama[c], Makiko Osawa[a]



normal

gradually obtunded

started to move



30 min later

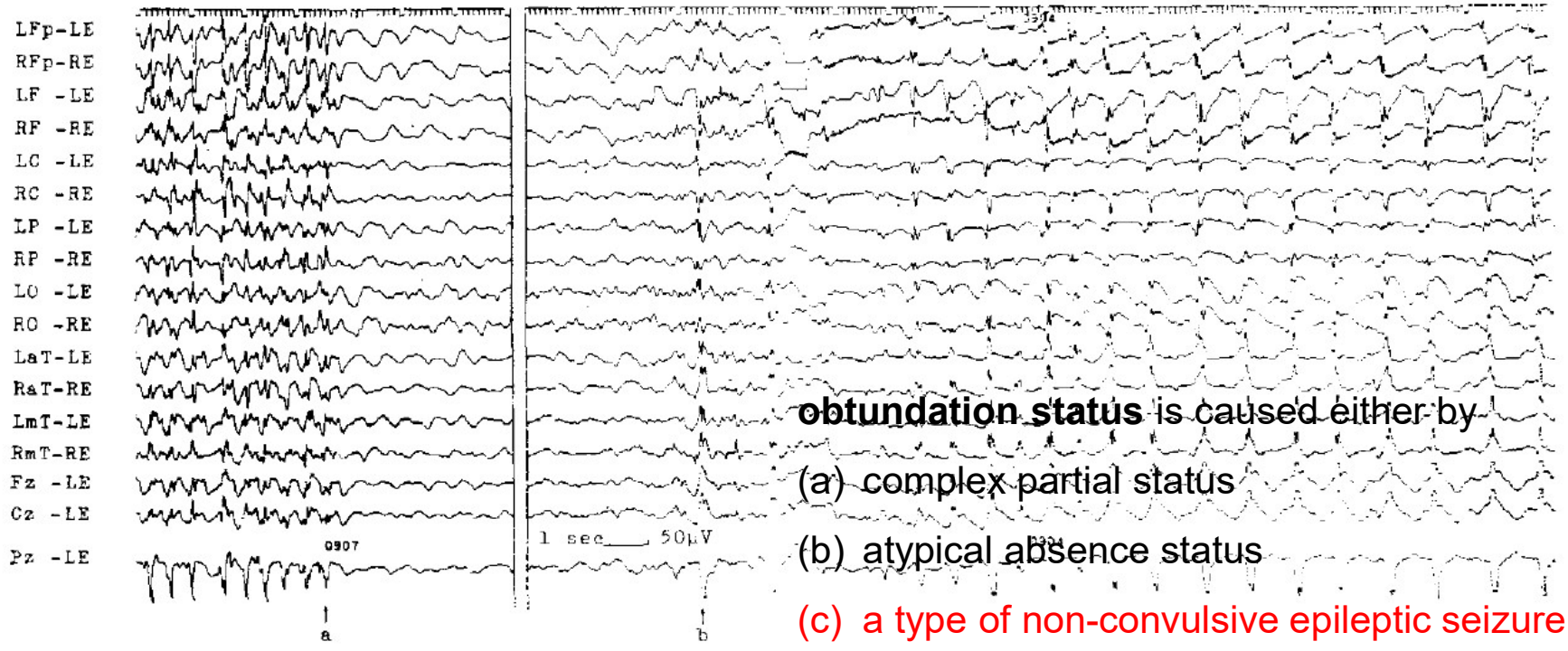
motionless

both arms extended

A Peculiar State Observed in 4 Patients with Severe Myoclonic Epilepsy in Infancy

Sumie Yasuda, M.D., Masako Watanabe, M.D., Tateki Fujiwara, M.D., Kazuichi Yagi, M.D. and Masakazu Seino, M.D.

The Japanese Journal of Psychiatry and Neurology, Vol. 43, No. 3, 1989



obtundation status is caused either by

(a) complex partial status

(b) atypical absence status

(c) a type of non-convulsive epileptic seizure related to *"post-grand mal exhaustion"*

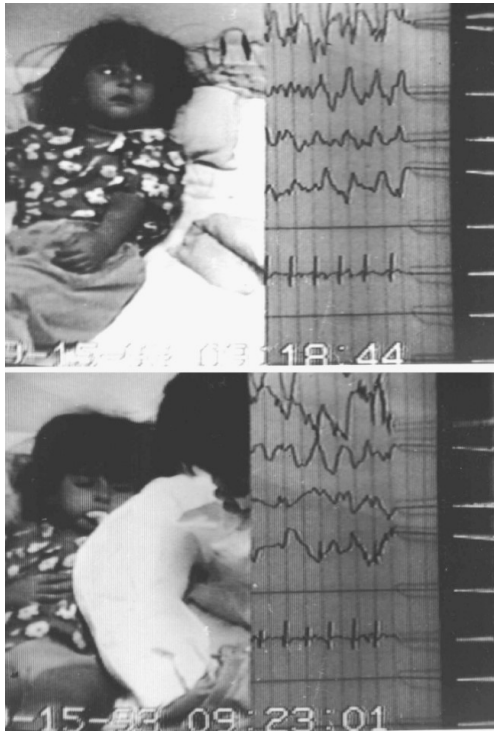
PCDH19-RELATED EPILEPSY

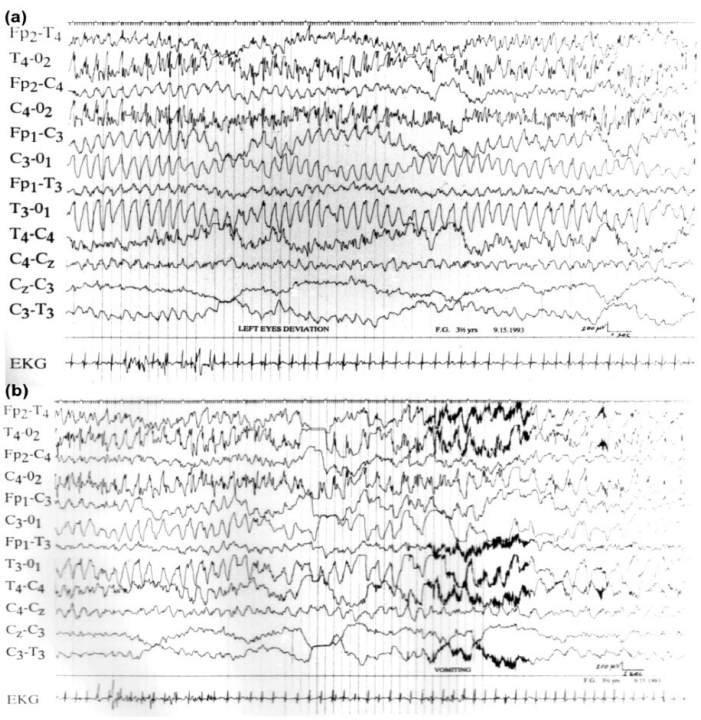
PANAYIOTOPOULOS SYNDROME

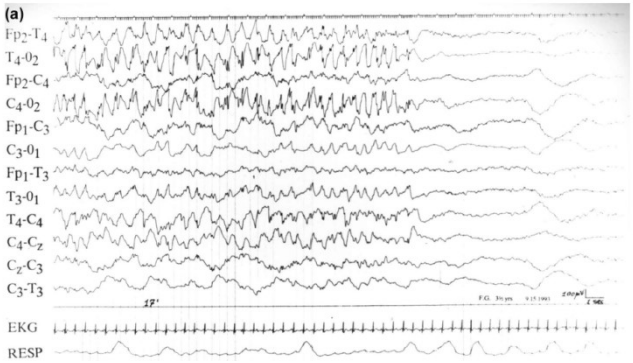
Early onset benign occipital susceptibility syndrome: video-EEG documentation of an illustrative case

Federico Vigevano*, Maria Luisa Lispi, Stefano Ricci

Clinical Neurophysiology 111, Suppl. 2 (2000) S81–S86







Epilepsy & Behavior 19 (2010) 383–393

Contents lists available at ScienceDirect

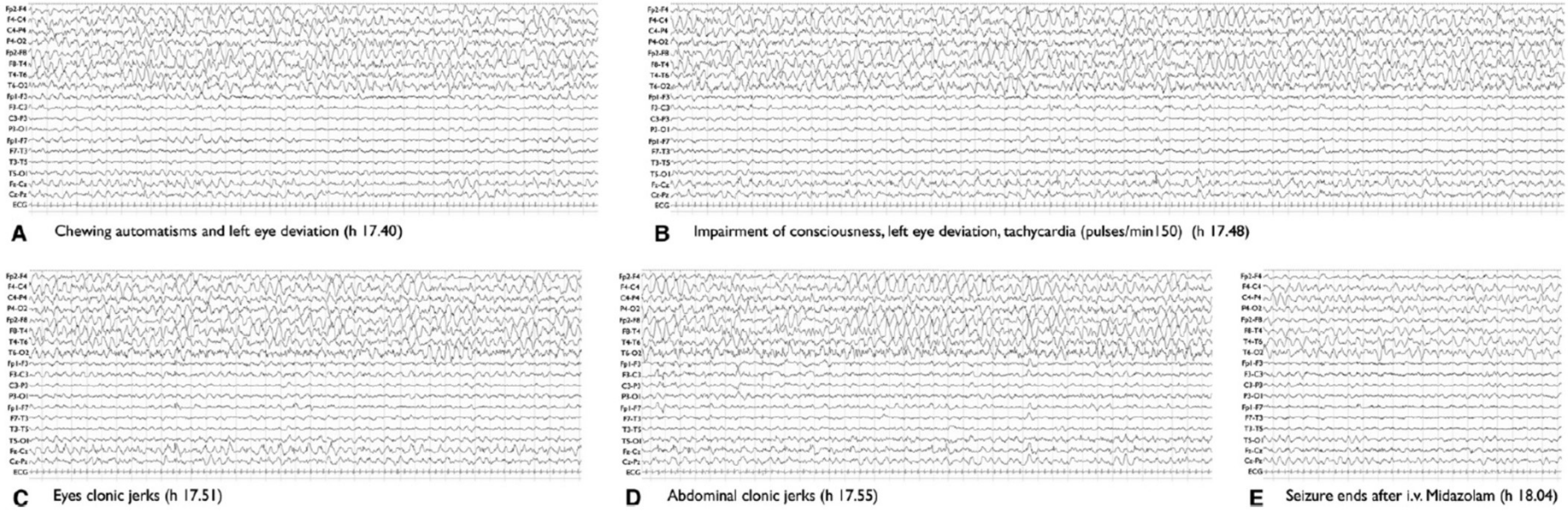
Epilepsy & Behavior

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

Documentation of autonomic seizures and autonomic status epilepticus with ictal EEG in Panayiotopoulos syndrome

Nicola Specchio [a,*], Marina Trivisano [b], Dianela Claps [a], Domenica Battaglia [c], Lucia Fusco [a], Federico Vigevano [a]

40’ after clinical onset



A Chewing automatisms and left eye deviation (h 17.40)

B Impairment of consciousness, left eye deviation, tachycardia (pulses/min150) (h 17.48)

C Eyes clonic jerks (h 17.51)

D Abdominal clonic jerks (h 17.55)

E Seizure ends after i.v. Midazolam (h 18.04)

Epilepsy & Behavior 19 (2010) 383–393

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Documentation of autonomic seizures and autonomic status epilepticus with ictal EEG in Panayiotopoulos syndrome

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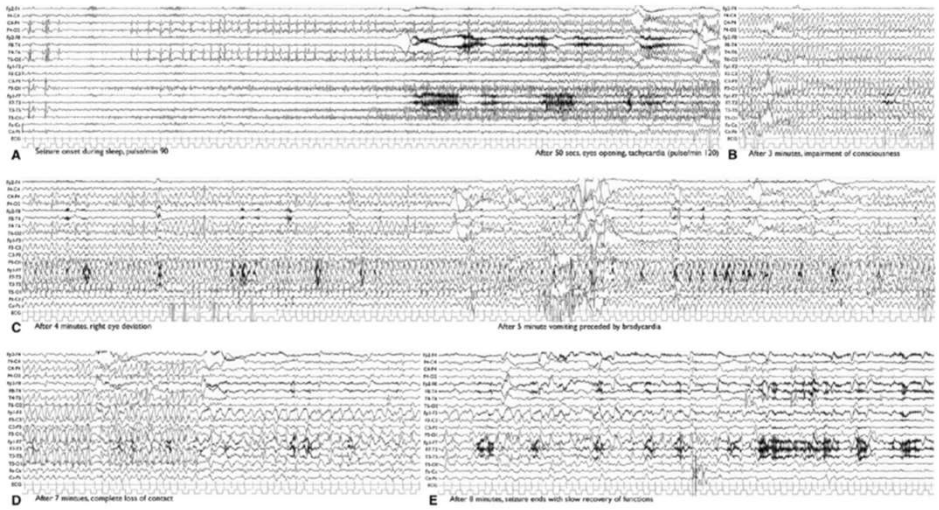

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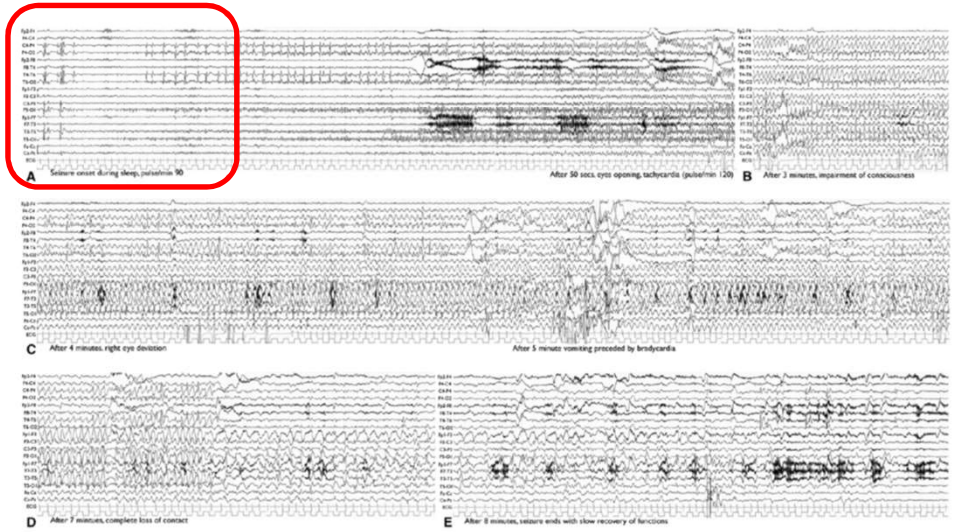
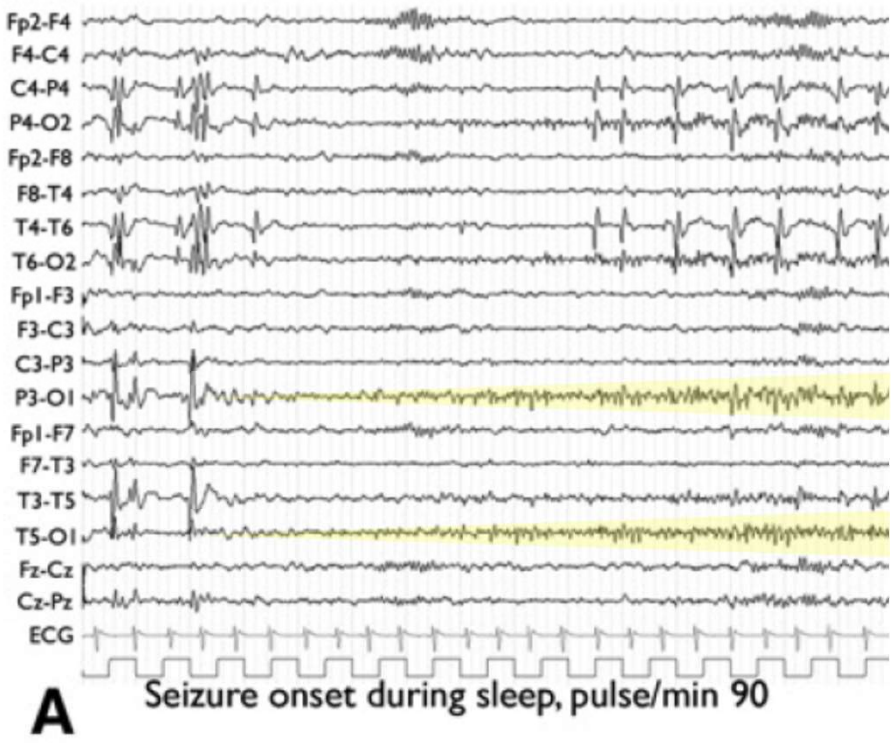
Nicola Specchio [a,*], Marina Trivisano [b], Dianela Claps [a], Domenica Battaglia [c], Lucia Fusco [a], Federico Vigevano [a]





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Epilepsy & Behavior 7 (2005) 543–547
Epilepsy & Behavior
www.elsevier.com/locate/yebeh

Case Report

Recurrent autonomic status epilepticus in Panayiotopoulos syndrome: Video/EEG studies

Michael Koutroumanidis *, Shaun Rowlinson, Sue Sanders

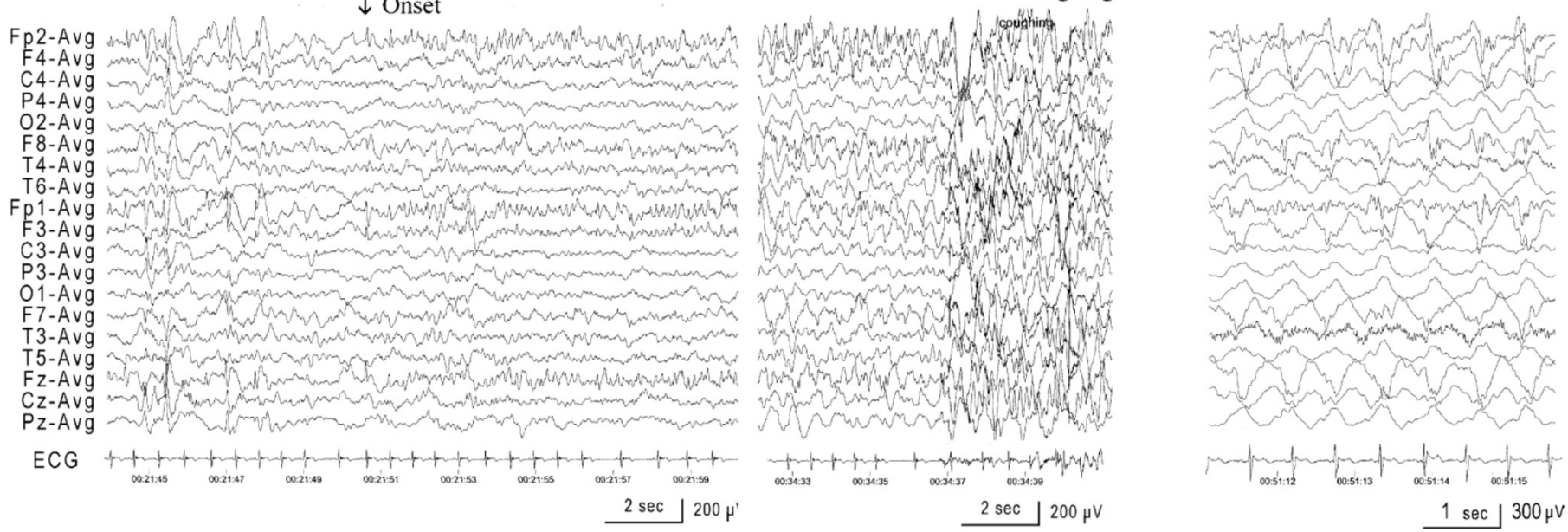
00:21 ↓ Onset

+13'

00:34 ↓ Coughing

+20'

00:51 Difficult to communicate



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Case Report

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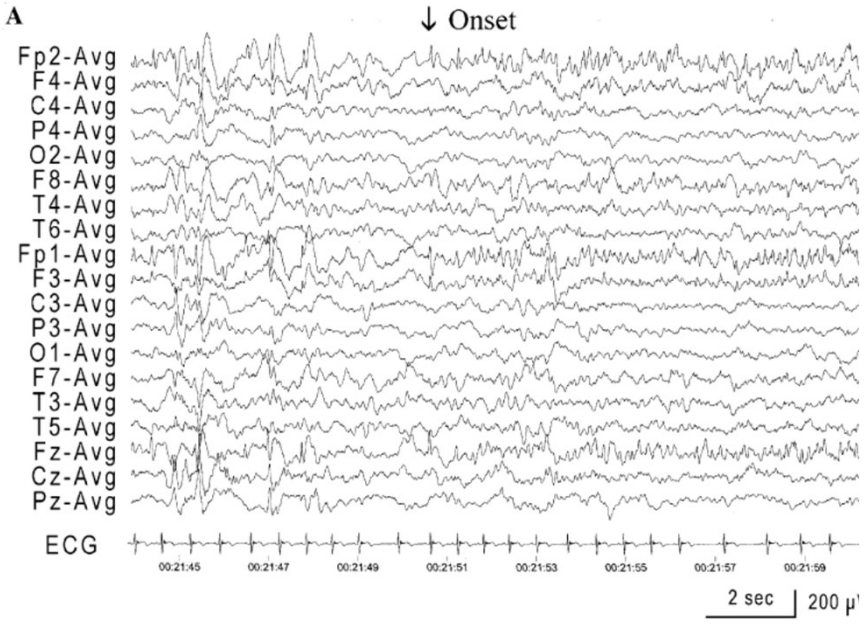
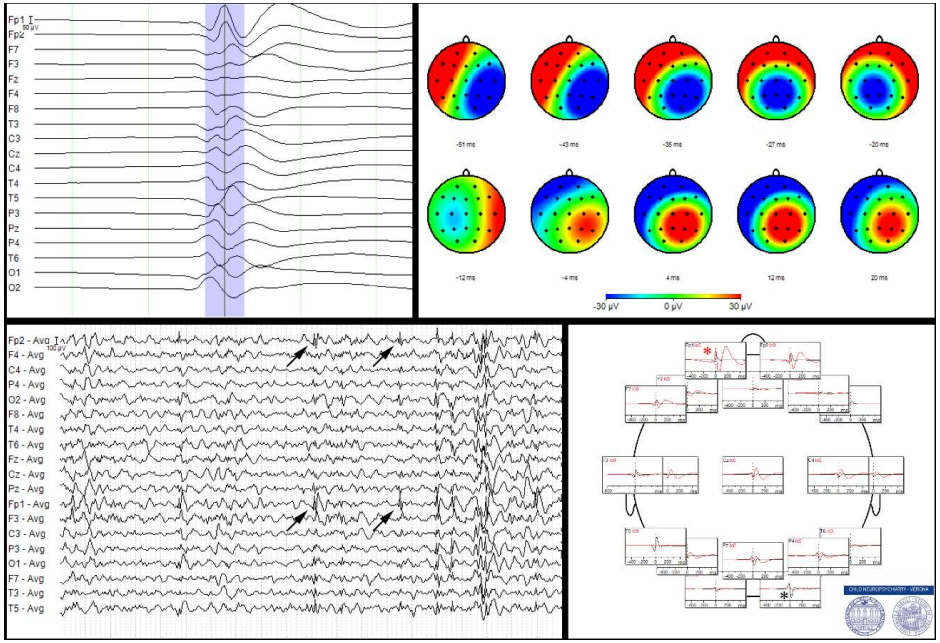
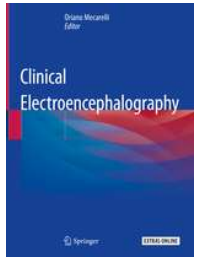


«*Oguni-Leal phenomenon*» in PS



Gardella & Cantalupo. Focal *"Idiopathic"* Epilepsies of infancy. In: Mecarelli 2019



DOOSE SYNDROME

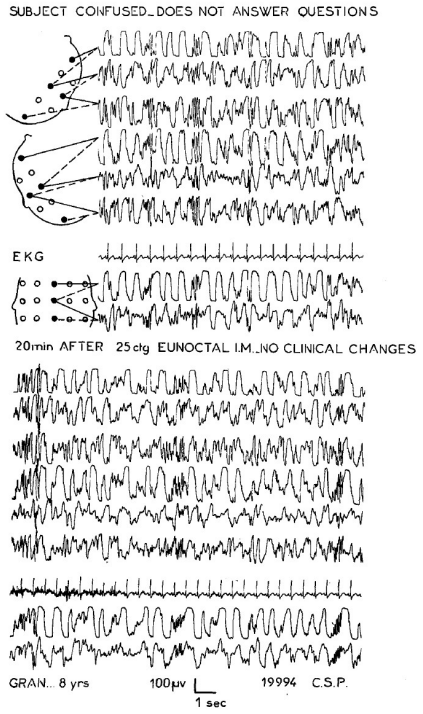
LENNOX-GASTAUT SYNDROME

Rev. E.E.G. Neurophysiol. clin., 1985, 15: 361 – 368

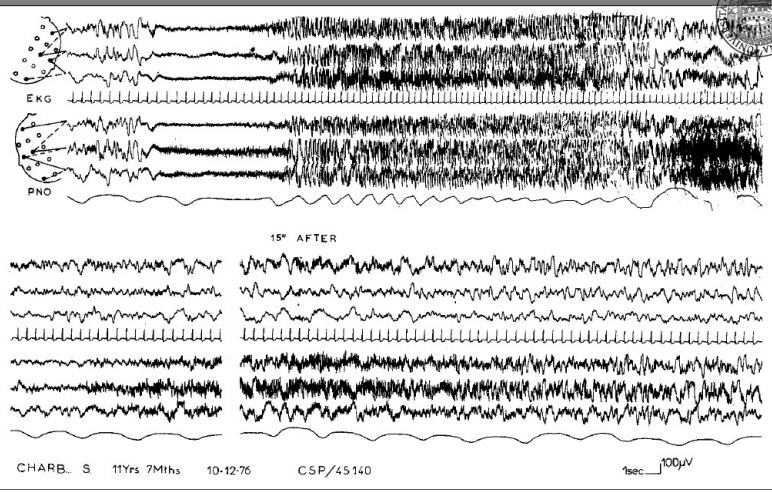
LES ETATS DE MAL DANS LE SYNDROME DE LENNOX–GASTAUT

C. DRAVET, O. NATALE, A. MAGAUDDA, J.L. LARRIEU,
M. BUREAU, J. ROGER et C.A. TASSINARI

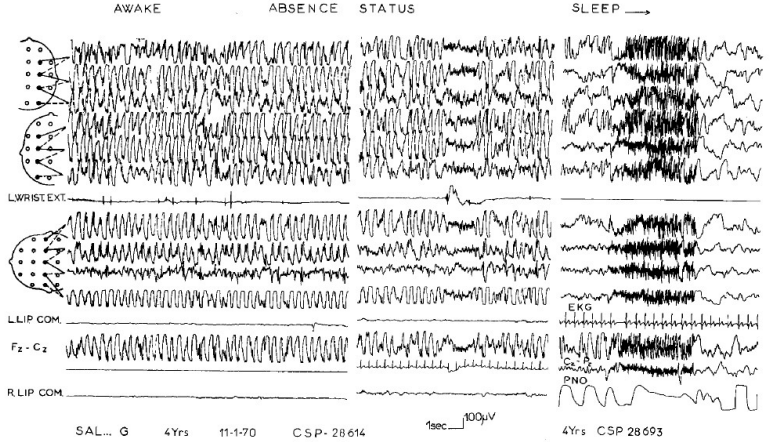
Atypical Absence Status



Tonic Status



Absence Status with interspersed Tonic seizure series



UNIVERSITÀ di VERONA

Rev. E.E.G. Neurophysiol. clin., 1985, 15: 361 – 368

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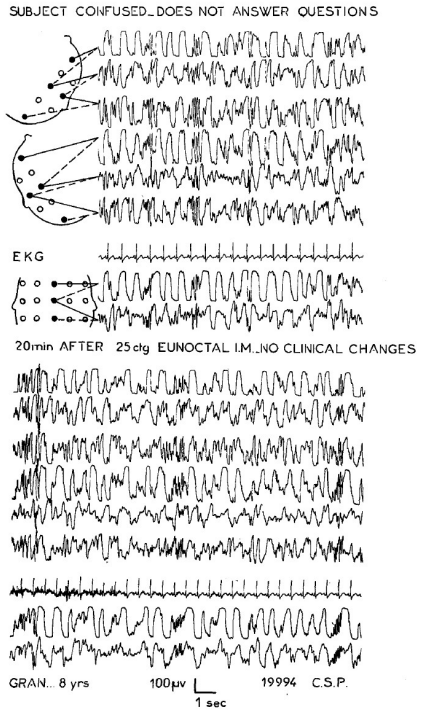
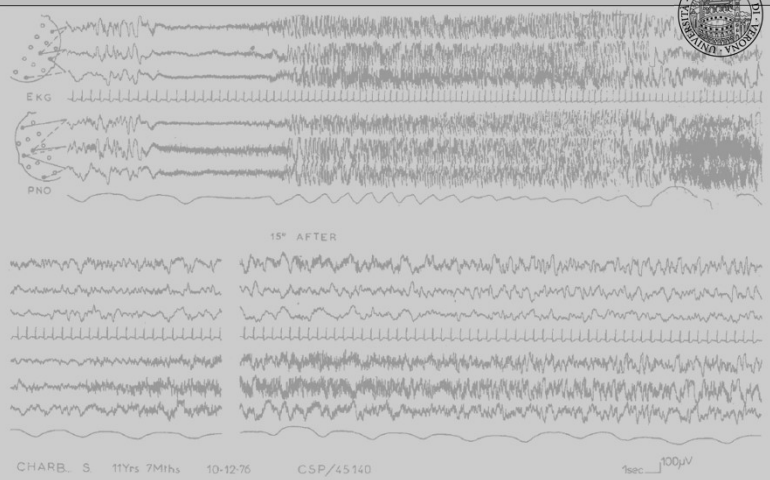
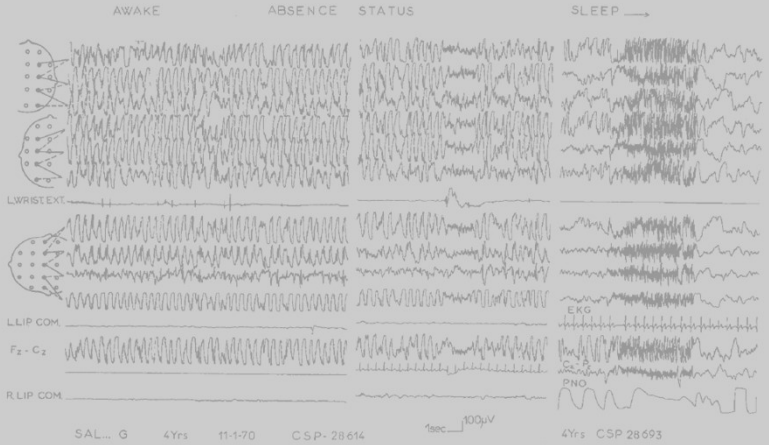


Tonic Status



Absence Status with interspersed Tonic seizure series



Rev. E.E.G. Neurophysiol. clin., 1985, 15: 361 – 368

LES ETATS DE MAL DANS LE SYNDROME DE LENNOX–GASTAUT

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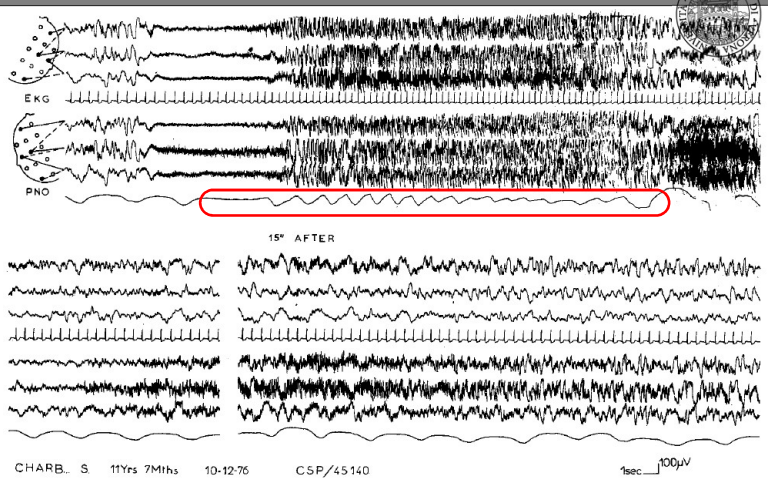
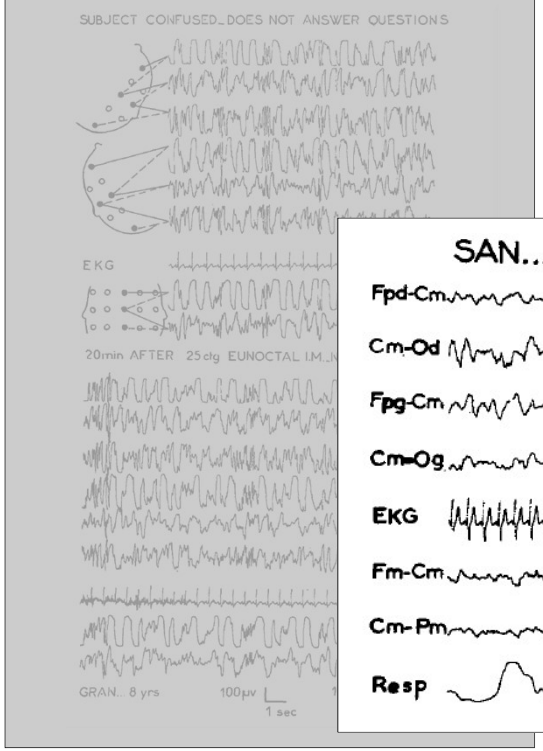
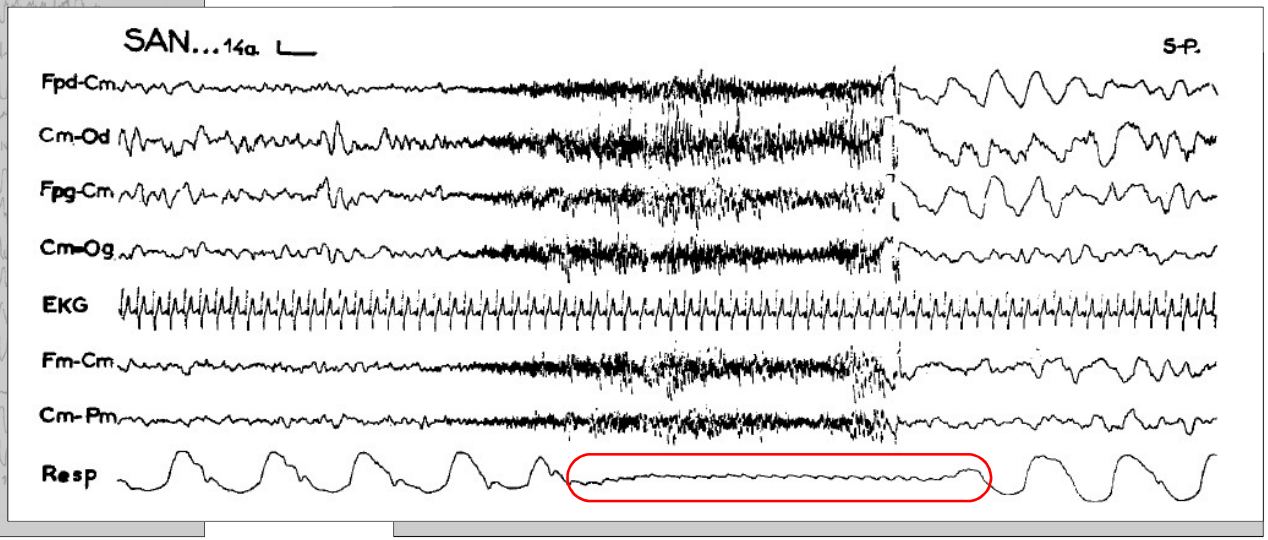
M. BUREAU, J. ROGER et C.A. TASSINARI

Tonic Status



Atypical Absence Status





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LES ETATS DE MAL DANS LE SYNDROME DE LENNOX–GASTAUT

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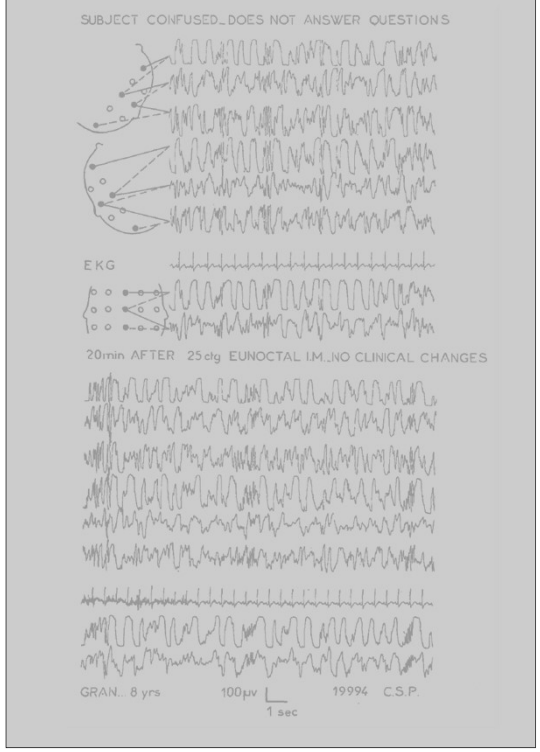
Atypical Absence Status



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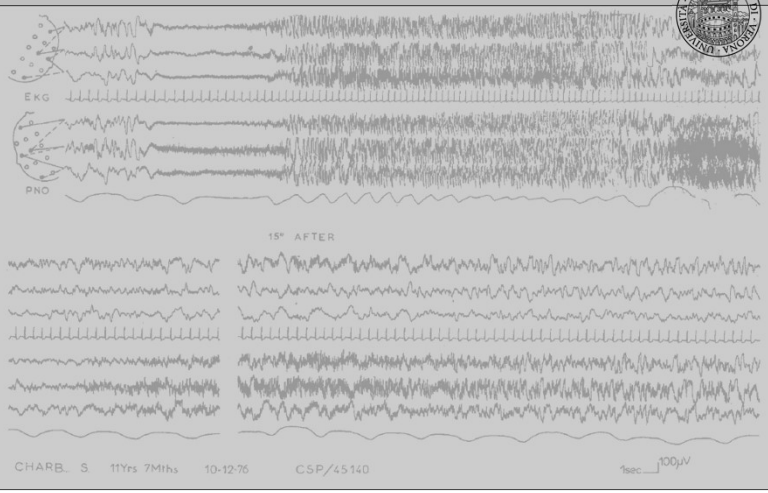

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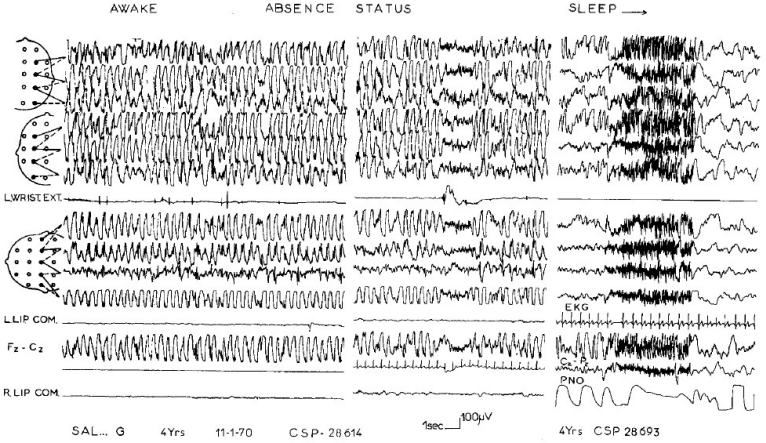
Absence Status with interspersed Tonic seizure series



Rev. E.E.G. Neurophysiol. clin., 1985, 15: 361 – 368

LES ETATS DE MAL DANS LE SYNDROME DE LENNOX–GASTAUT

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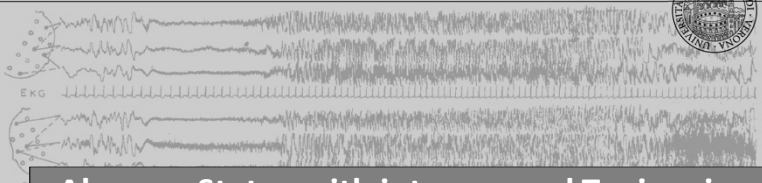
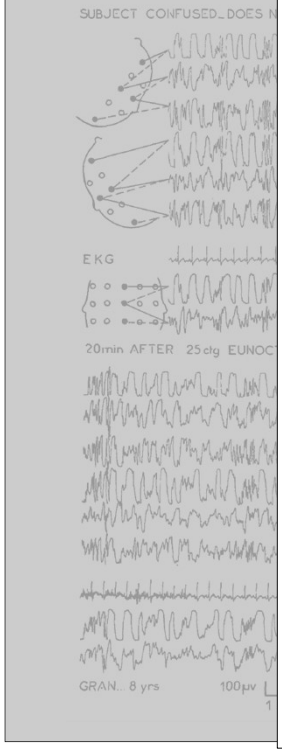
Tonic Status

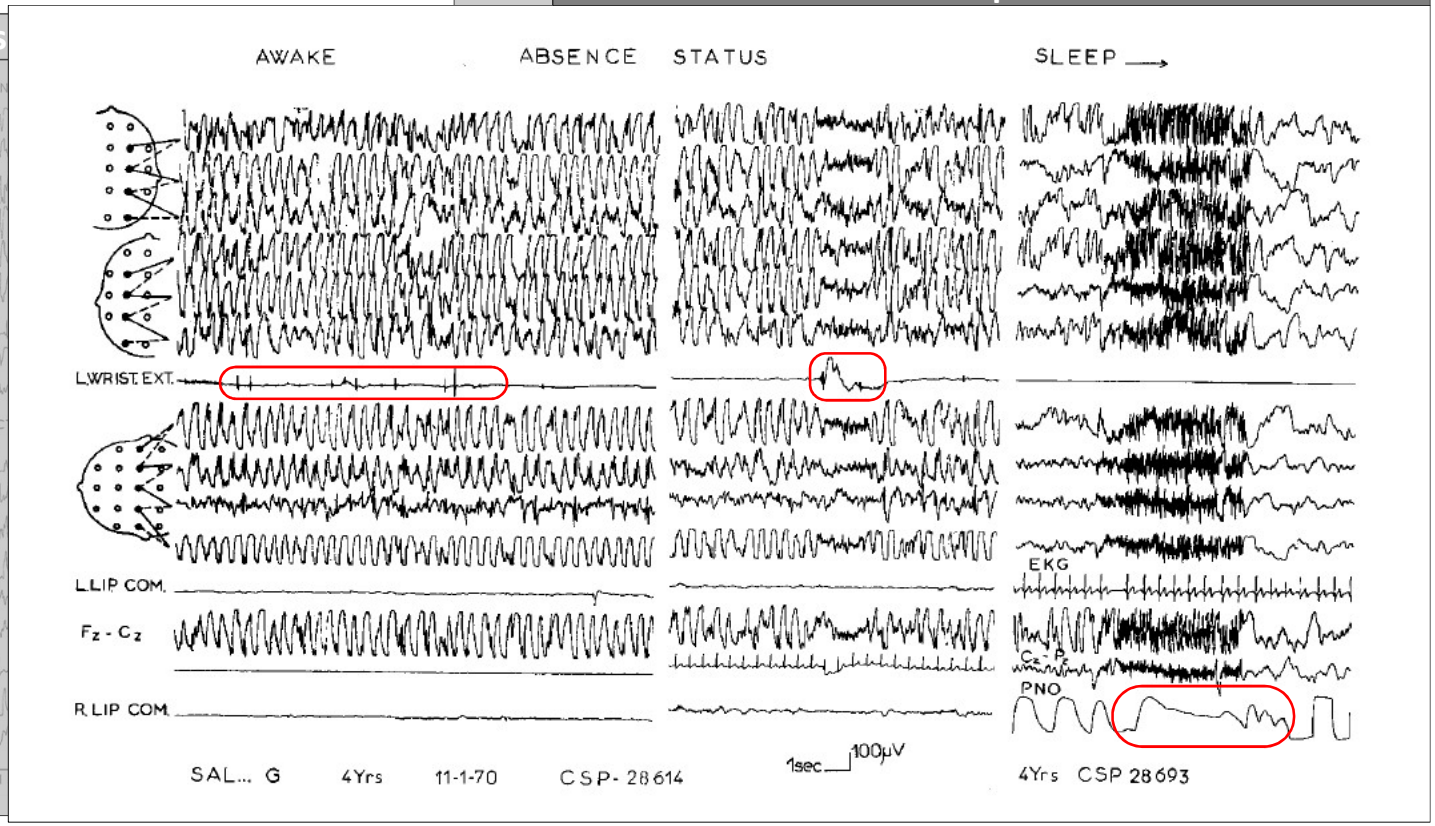


UNIVERSITÀ di VERONA

Absence Status with interspersed Tonic seizure series

Atypical Abs





*Super-refractory **Tonic-Myoclonic Status Epilepticus** with insidious onset and catamenial recurrence in Epilepsy with Myoclonic atonic seizures (Doose syndrome)*

J. Proietti[1], T. Lo Barco[1], G. Rizzi[1], E. Santangelo[1], E. Parrini[2], C. Bonin[3], G. Cantalupo[1], E. Fontana[1], F. Darra[1], E. Fiorini[1], B. Dalla Bernardina[1]

[1]U.O.C. Neuropsichiatria Infantile, Azienda Ospedaliera Universitaria Integrata, Verona.
[2] Laboratorio di Neurogenetica, Azienda Ospedaliero-Universitaria "A. Meyer", Firenze.
[3]U.O.C. Ostetricia e Ginecologia B, Azienda Ospedaliera Universitaria Integrata, Verona.

*CLINICAL CASES AND REVIEWS IN EPILEPSY **(under review)***

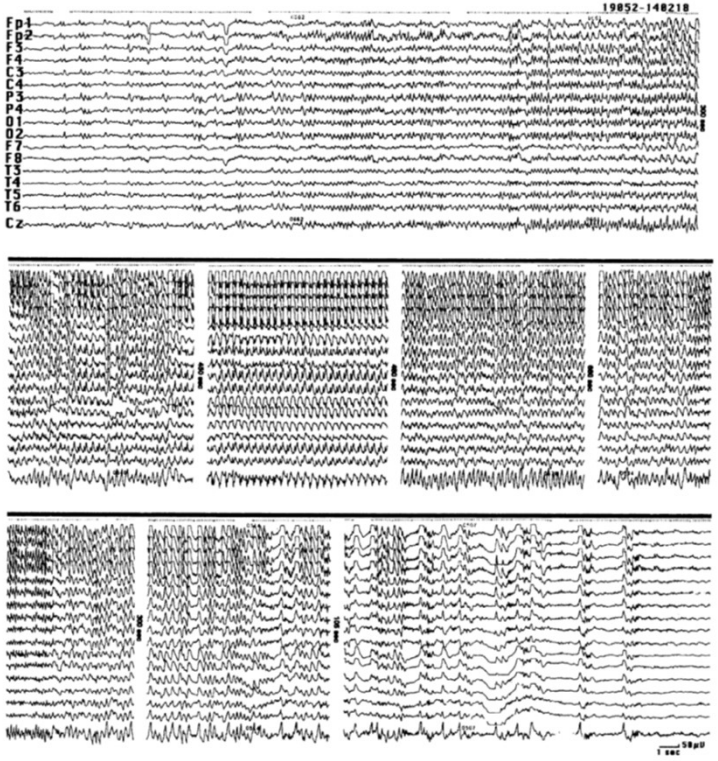
RING20-RELATED EPILEPSY

Brain (1997), **120**, 939–953

Ring chromosome 20 and nonconvulsive status epilepticus

A new epileptic syndrome

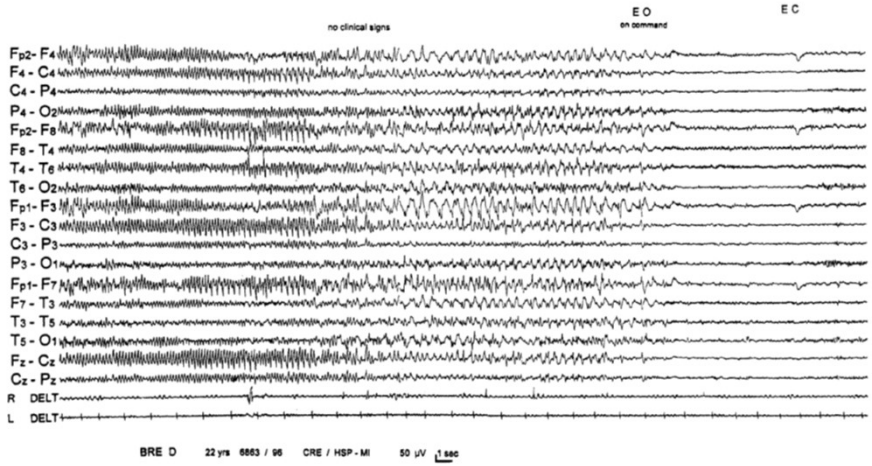
Y. Inoue,[1] T. Fujiwara,[1] K. Matsuda,[1] H. Kubota,[1] M. Tanaka,[1] K. Yagi,[1] K. Yamamori[1] and Y. Takahashi[2]



Epilepsia, **39**(9):942–951, 1998

Chromosome 20 Ring: A Chromosomal Disorder Associated with a Particular Electroclinical Pattern

Maria Paola Canevini, Vincenzo Sgro, *Orsetta Zuffardi, Raffaele Canger, ‡Romeo Carrozzo, §Elena Rossi, ||David Ledbetter, †Fabio Minicucci, Aglaia Vignoli, Ada Piazzini, Loredana Guidolin, Amalia Saltarelli, and ¶Bernardo dalla Bernardina



ATYPICAL SELF-LIMITED FOCAL EPILEPSIES OF CHILDHOOD

- Convulsive (hemiclonic) SE
- NCSE
  - Atypical absences +/- Myoclonic phenomena
  - Opercular Status
  - ESES (*Tassinari Encephalopathy*)

Epilepsia, 28(4):351–355, 1987
Raven Press, Ltd., New York

Status Epilepticus of Benign Partial Epilepsies in Children: Report of Two Cases

Natalio Fejerman and A. Marcelo Di Blasi

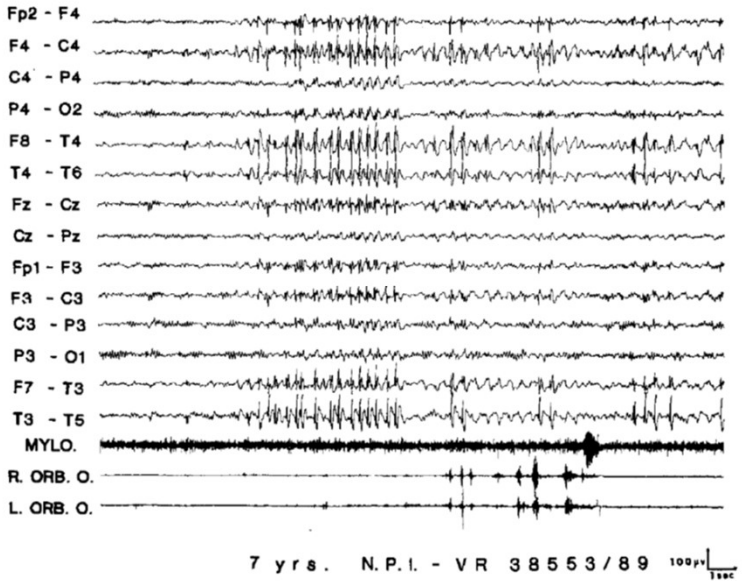
In summary, we believe that there are neither clinical nor theoretic reasons to refute the possibility that a neurologically normal child with BPEC might have clinical and electrical signs of SE corresponding to the type of epilepsy suffered and have a complete SE remission without neuropsychologic deterioration. We have no explanation for the lack of response to antiepileptic drug therapy and the dramatic improvement after steroid treatment in our two cases. Finally, we are aware that a longer follow-up of these cases is needed to support the hypothesis that episodes of SE can exceptionally occur in patients with BPEC-R without changing the prognosis of this condition.

Morikawa T, Seino M, Osawa T, Yagi H. Cinq enfants avec décharges de pointes ondes continues pendant le sommeil. In: Roger J, et al. eds. *Les syndromes épileptiques de l'enfant et de l'adolescent*. London: John Libbey Eurotext, 1984:210–7.

Epilepsia, 32(3):329–334, 1991
Raven Press, Ltd., New York

Status Epilepticus in Benign Rolandic Epilepsy Manifesting as Anterior Operculum Syndrome

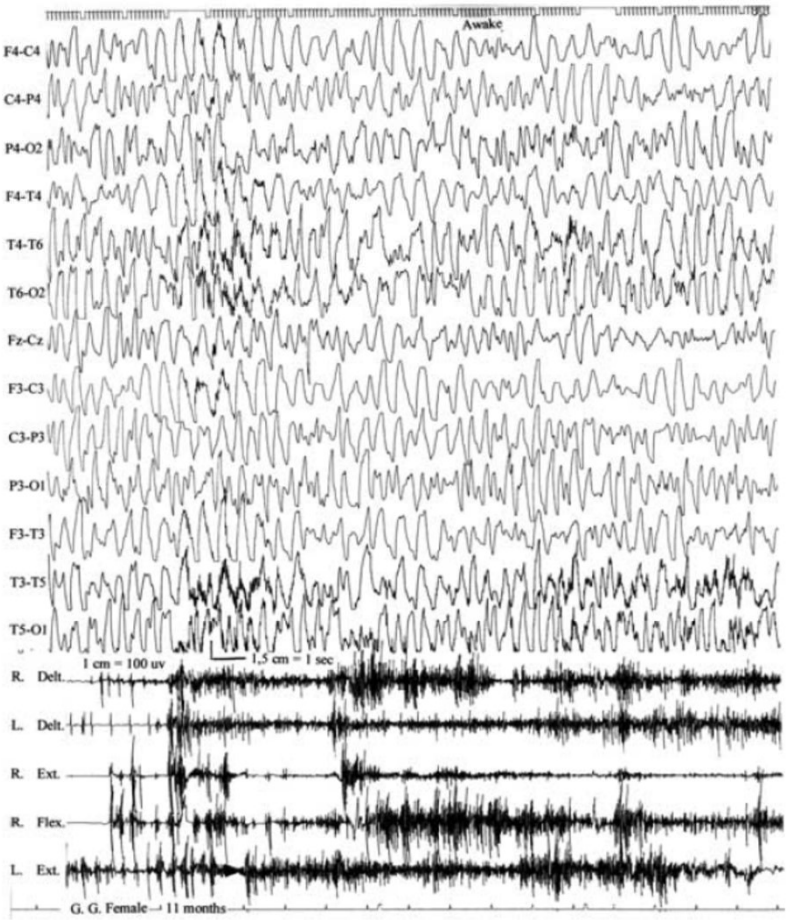
Vito Colamaria, Vincenzo Sgrò, *Roberto Caraballo, Marina Simeone, Emanuele Zullini, Elena Fontana, Rossella Zanetti, Rosal Grimau-Merino, and Bernardo Dalla Bernardina



Boulloche J, Le Luyer B, Husson A, Le Roux P. Dysphagie et troubles du langage: manifestations d'un état de mal épileptique a pointes temporales. *Proceedings IVème Congrès de la Société Européenne de Neurologie Pédiatrique*. Barcelona, novembre 1989:131.

Dalla Bernardina B, Chiamenti C, Capovilla G, Colamaria V. Benign partial epilepsies in childhood. In: Roger J, Dravet C, Bureau M, Dreifuss FE, Wolf P, eds. *Epileptic syndromes in infancy, childhood and adolescence*. London: John Libbey Eurotext, 1985*a*:137–49.

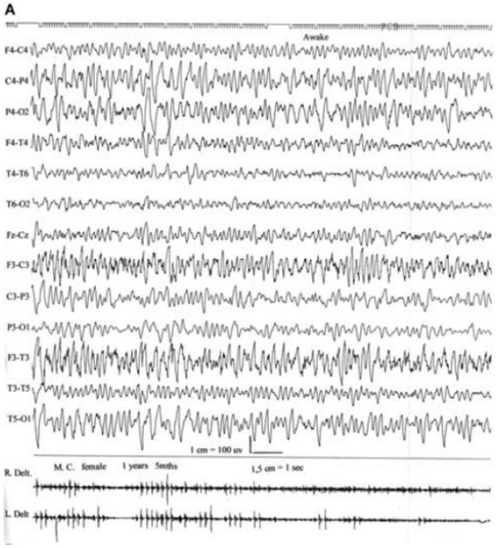
MYOCLONIC STATUS IN NONPROGRESSIVE ENCEPHALOPATHIES OR DALLA BERNARDINA SYNDROME

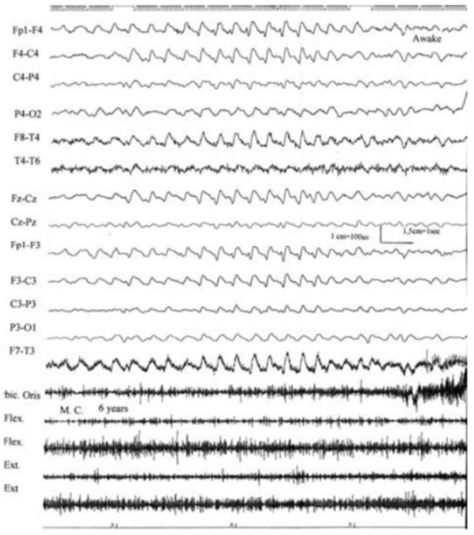


Epilepsia, **48**(1):107–113, 2007
Blackwell Publishing, Inc.

Myoclonic Status in Nonprogressive Encephalopathies: Study of 29 Cases

Roberto Horacio Caraballo, Ricardo Oscar Cersósimo, Alberto Espeche, Hugo Antonio Arroyo, and Natalio Fejerman





Other rare conditions

1986, Vol. 17, pp. 75–80, © Hippokrates Verlag GmbH

neuropediatrics

Progressive Neuronal Degeneration of Childhood with Liver Disease ("Alpers' Disease"): Characteristic Neurophysiological Features

S. G. Boyd
Ann Harden
J. Egger
G. Pampiglione

In 9 patients, one of whom had been seen during an earlier phase, EEGs were taken when frequent, usually focal seizures had become a prominent feature of the illness. In all 9 cases an EEG taken at this stage of the illness showed unusual and strikingly similar abnormalities. These consisted of very slow activity (1 c/s or less) of very high amplitude (200 microvolts to about one millivolt), mixed with polyspikes which were of much lower amplitude and which had a rather distinctive configuration (Figs. 1 bottom, 2, 3). These polyspikes were often more prominent over one area, including the occipital cortex (Fig. 4). Clinical attacks were usually contralateral to the hemisphere showing focal discharges. However, surface polyEMG recordings in three cases did not show a clear correlation between the groups of muscle action potentials and the EEG discharges. In six cases, the slow activity was grossly

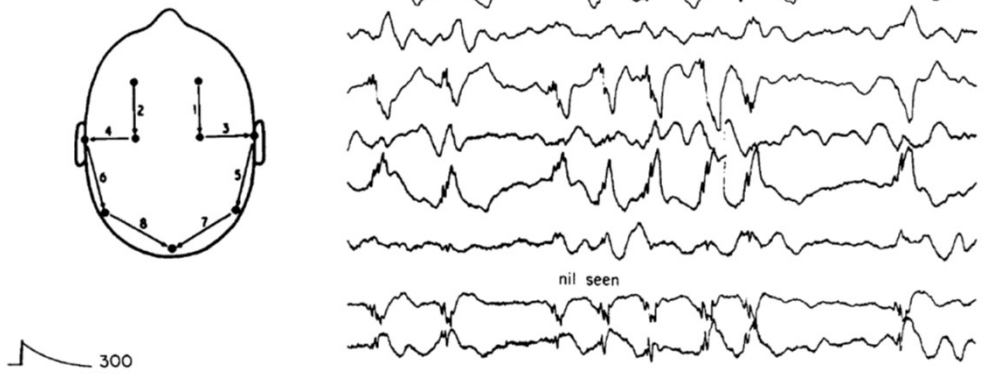
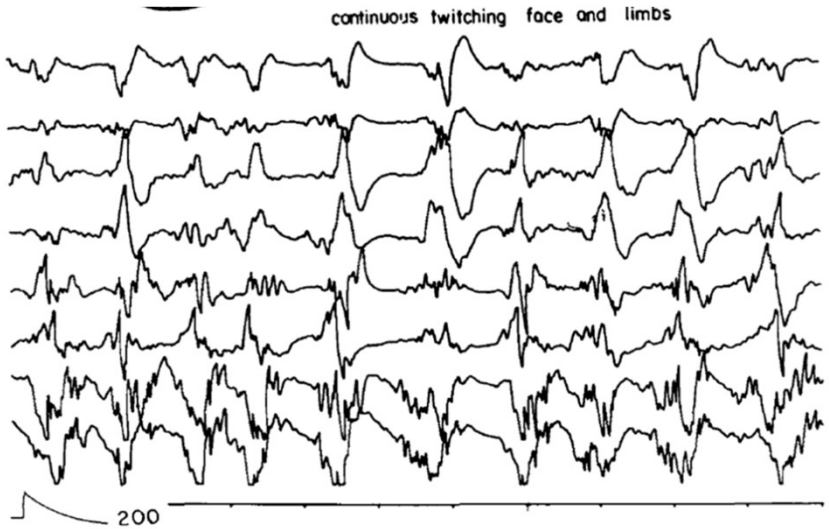


Fig. 2 (Case 10) This 6-year-old boy had frequent left sided twitching. Note the marked asymmetry in the EEG with high amplitude slow activity over the right hemisphere. Lower amplitude polyspikes are seen particularly over the right temporal and mid occipital regions

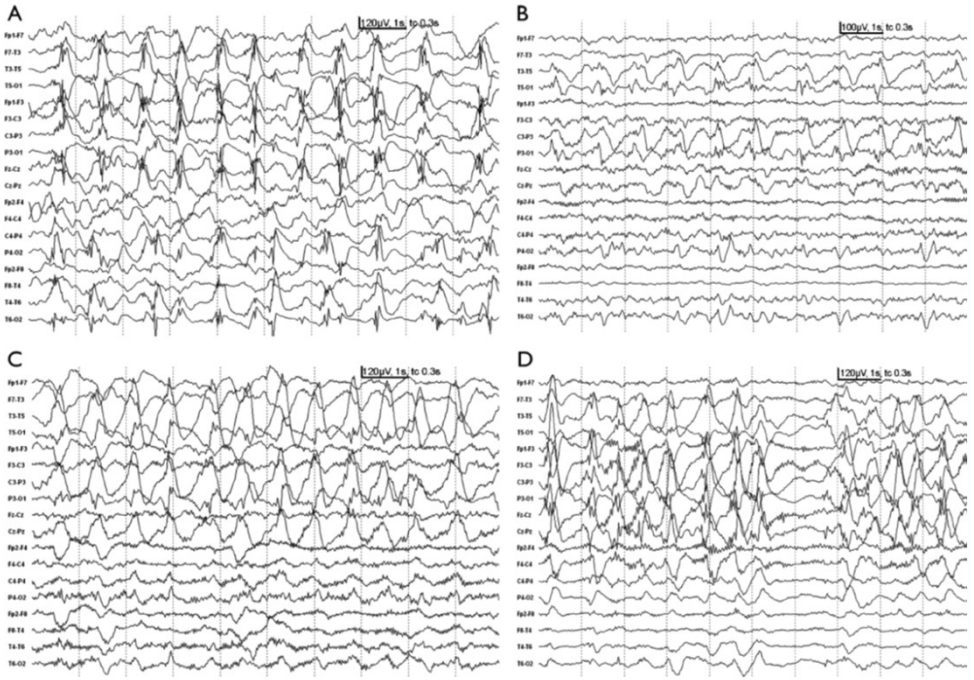


Epilepsia, 50(6):1596–1607, 2009
doi: 10.1111/j.1528-1167.2008.01877.x

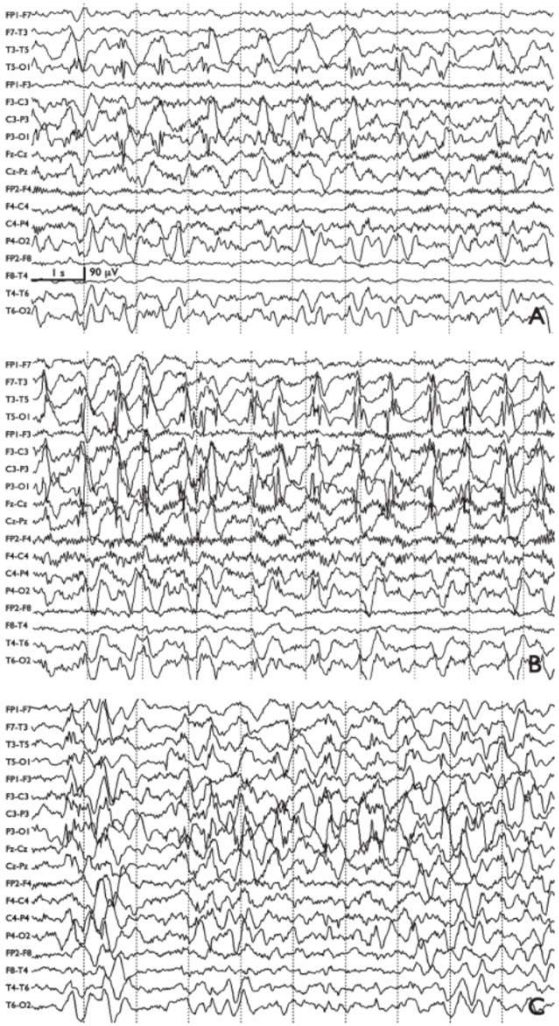
FULL-LENGTH ORIGINAL RESEARCH

Status epilepticus in children with Alpers' disease caused by *POLG1* mutations: EEG and MRI features

*Nicole I. Wolf, †Shamima Rahman, ‡Bernhard Schmitt, §Jan-Willem Taanman, †Andrew J. Duncan, ¶Inga Harting, ‡Gabriele Wohlrab, *Friedrich Ebinger, *Dietz Rating , and *Thomas Bast



Rhythmic
High-
Amplitude
Delta with
superimposed
(poly)**S**pikes
(**RHADS**)



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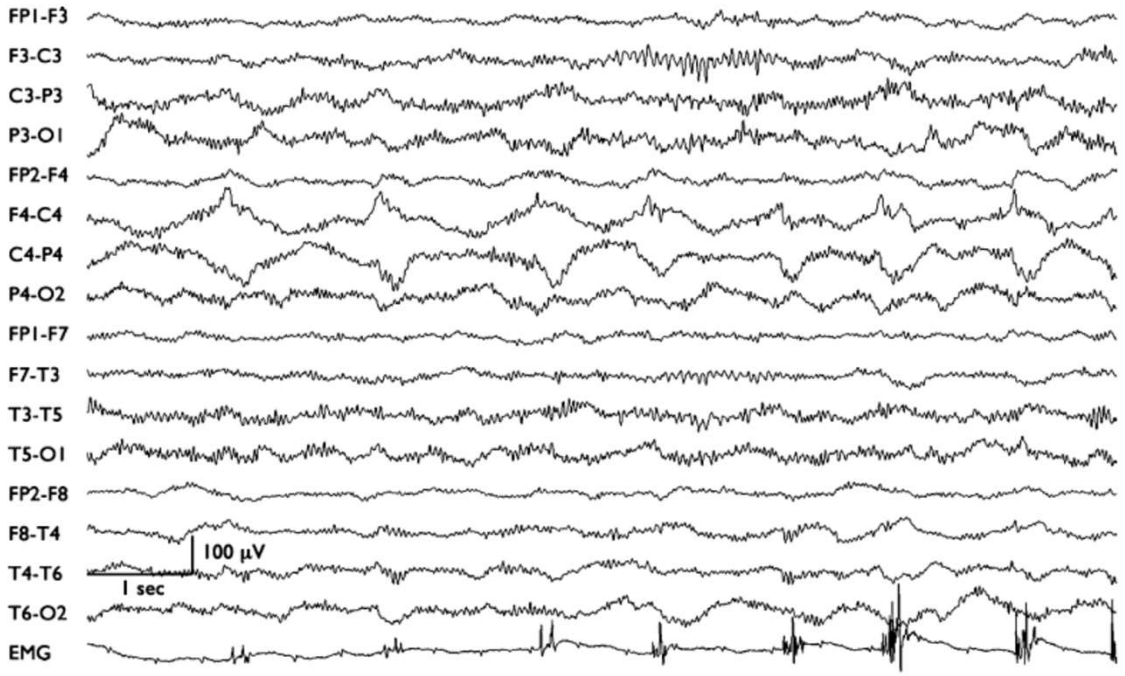
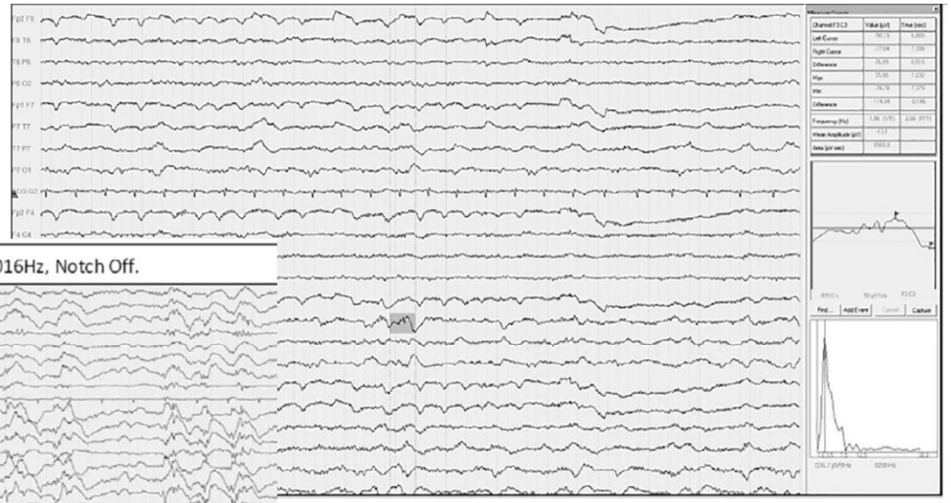


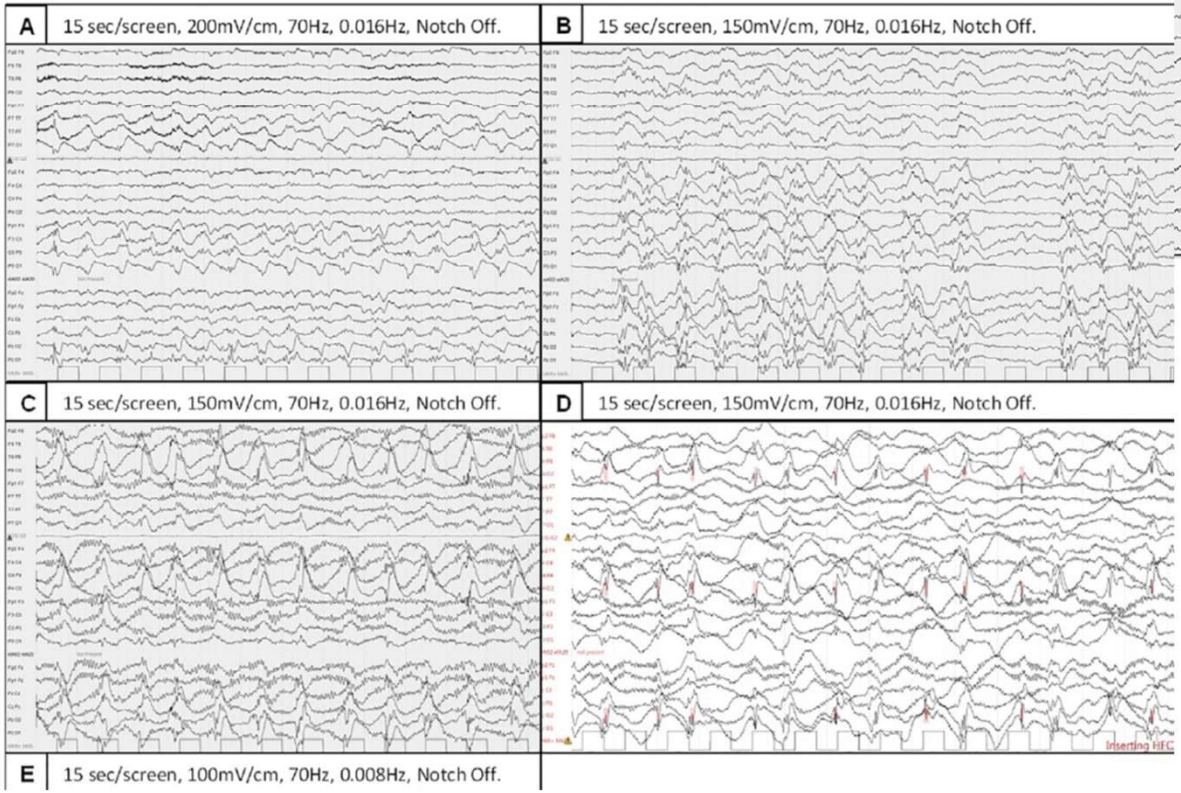
Figure 4.
Electroencephalography (EEG) during sleep (stage II) of Patient 4 at disease onset. Sleep spindles are lacking over the right central region. With a maximum over C4, there is a continuous slowing and sharp waves (PLDs) followed by a myoclonic jerk.
Epilepsia © ILAE

Clinical Neurophysiology 129 (2018) 2127–2131

Specific EEG markers in POLG1 Alpers' syndrome

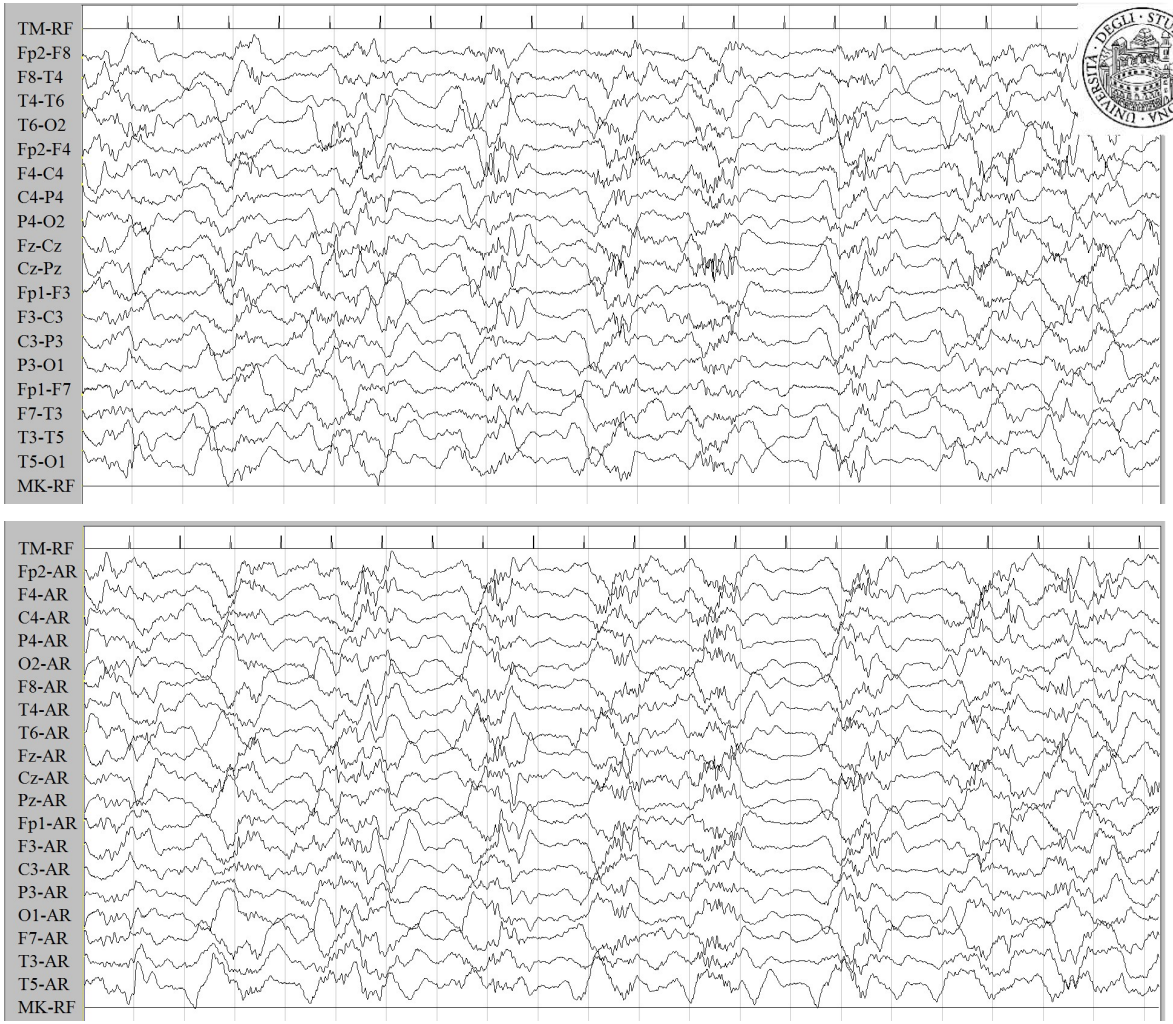
Anouk van Westrhenen [a,b], Elisabeth A. Cats [d], Bart van den Munckhof [a], Sandra M.A. van der Salm [a,c], Nico W. Teunissen [a], Cyrille H. Ferrier [a], Frans S.S. Leijten [a,*], Karin P.W. Geleijns [a]





Four video recordings were lacking (patient 1 and 6) or had incomplete imaging of the patient (patient 7 and 8). In two patients a co-occurrence of clinical epileptic signs (myoclonus of right leg and foot in patient 1 and 4 respectively) and RHADS was found [Table 1]. Patient 3 showed visual agnosia along with nearly continuous RHADS in the occipital region. Subsequent EEGs displayed no RHADS and clinical records described restored vision.

Our results suggest that RHADS are a highly specific marker of AHS, and can (dis)appear over time. Using our criteria, RHADS can be well detected by the non-expert clinician. The appearance of correlating clinical epileptic symptoms and ripples indicate that RHADS signify an epileptic phenomenon. Further studies should give more insight in the pathophysiology of RHADS in AHS.



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Sezione di NEUROPSICHIATRIA INFANTILE

Epilepsia, 56(Suppl. 1):97-98, 2015
doi: 10.1111/epi.13241

p0379

A NEW CASE OF ALPERS SYNDROME WITH RAPID ONSET OF PARTIAL STATUS EPILEPTICUS: NEURORADIOLOGICAL AND ELECTROPHYSIOLOGICAL EVOLUTION

A. Iodice, S. Ferrari†, C. Spagnoli*, L. Pinelli‡, C. Vezzoli§, G. Cantalupo¶, B. Dalla Bernardina¶, P. Accorsi**, L. Giordano***

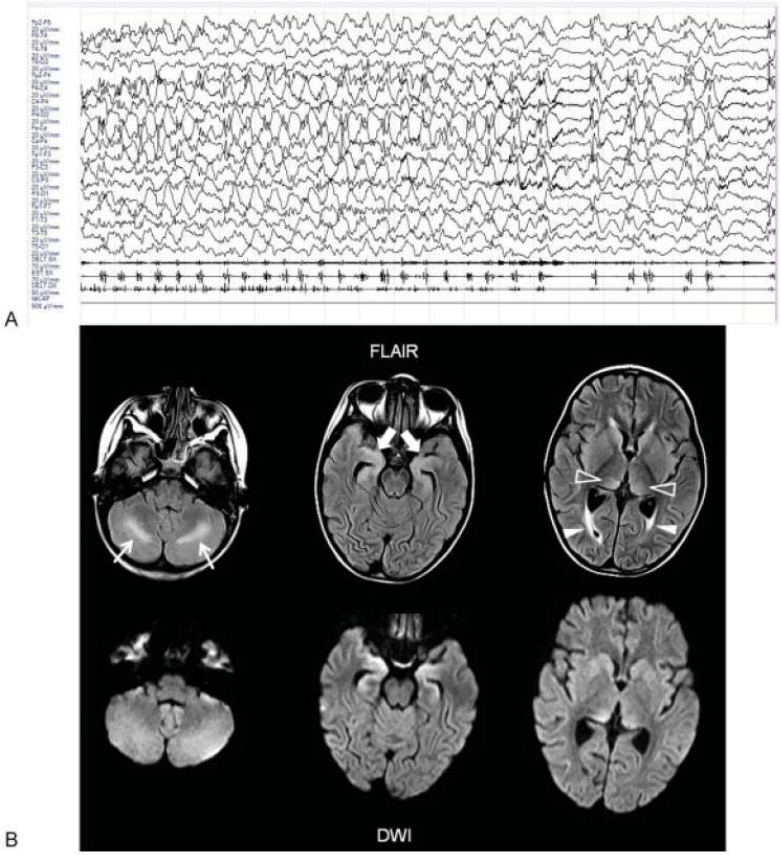
Sistema Socio Sanitario
Regione Lombardia
ASST Spedali Civili

Epilepsia, 56(Suppl. 1):97-98, 2015
doi: 10.1111/epi.13241

p0379

A NEW CASE OF ALPERS SYNDROME WITH RAPID ONSET OF PARTIAL STATUS EPILEPTICUS: NEURORADIOLOGICAL AND ELECTROPHYSIOLOGICAL EVOLUTION

A. Iodice, S. Ferrari†, C. Spagnoli*, L. Pinelli‡, C. Vezzoli§, G. Cantalupo¶, B. Dalla Bernardina¶, P. Accorsi**, L. Giordano***



Case Report

A Case of Alpers–Huttenlocher Syndrome Due to a New POLG1 Mutation with Rapid Onset of Partial Status Epilepticus: Serial Neuroradiological and Neurophysiological Evaluation

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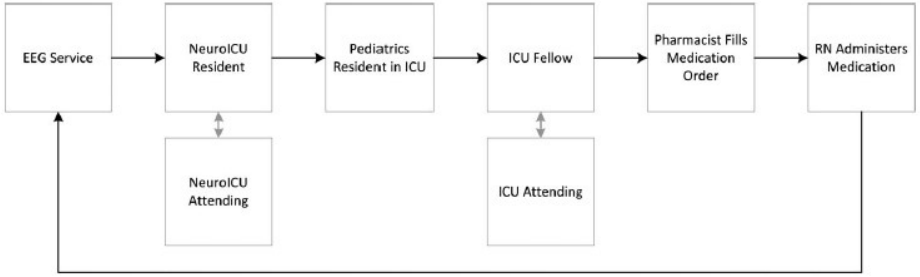
EEG role in SE TREATMENT

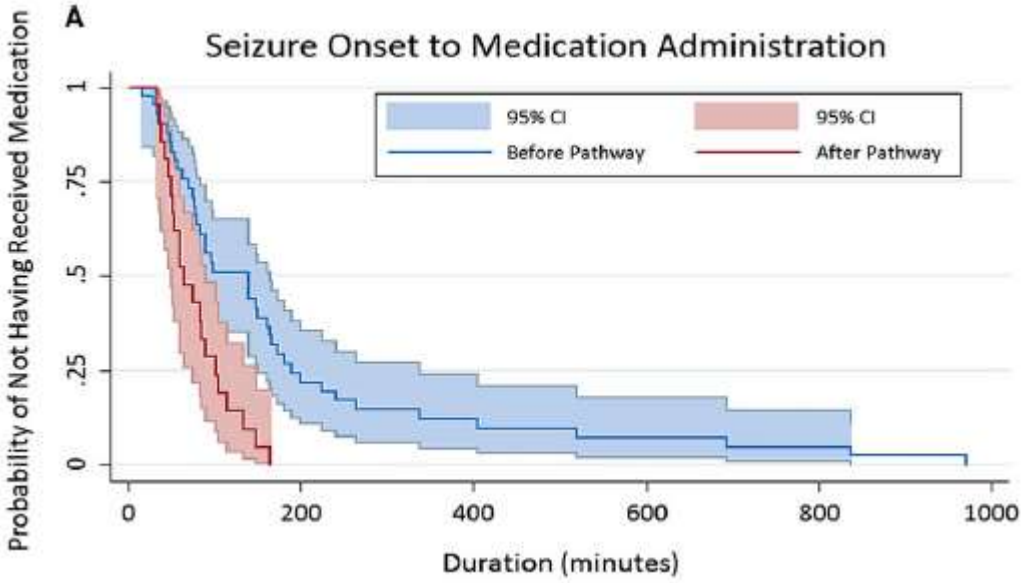
FULL-LENGTH ORIGINAL RESEARCH

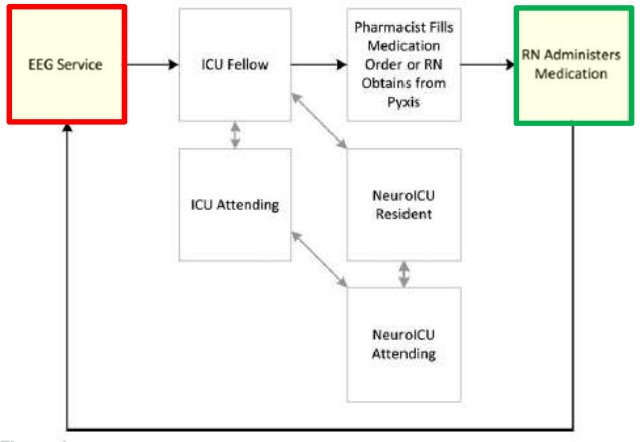
Impact of an ICU EEG monitoring pathway on timeliness of therapeutic intervention and electrographic seizure termination

*†Ryan P. Williams, *†Brenda Banwell, ‡Robert A. Berg, *†Dennis J. Dlugos, §Maureen Donnelly, *†Rebecca Ichord, *†Sudha Kilaru Kessler, *Jane Lavelle, *†Shavonne L. Massey, ¶Jennifer Hewlett, *Allison Parker, §Alexis A. Topjian, and *†§Nicholas S. Abend

Epilepsia, 57(5):786–795, 2016
doi: 10.1111/epi.13354







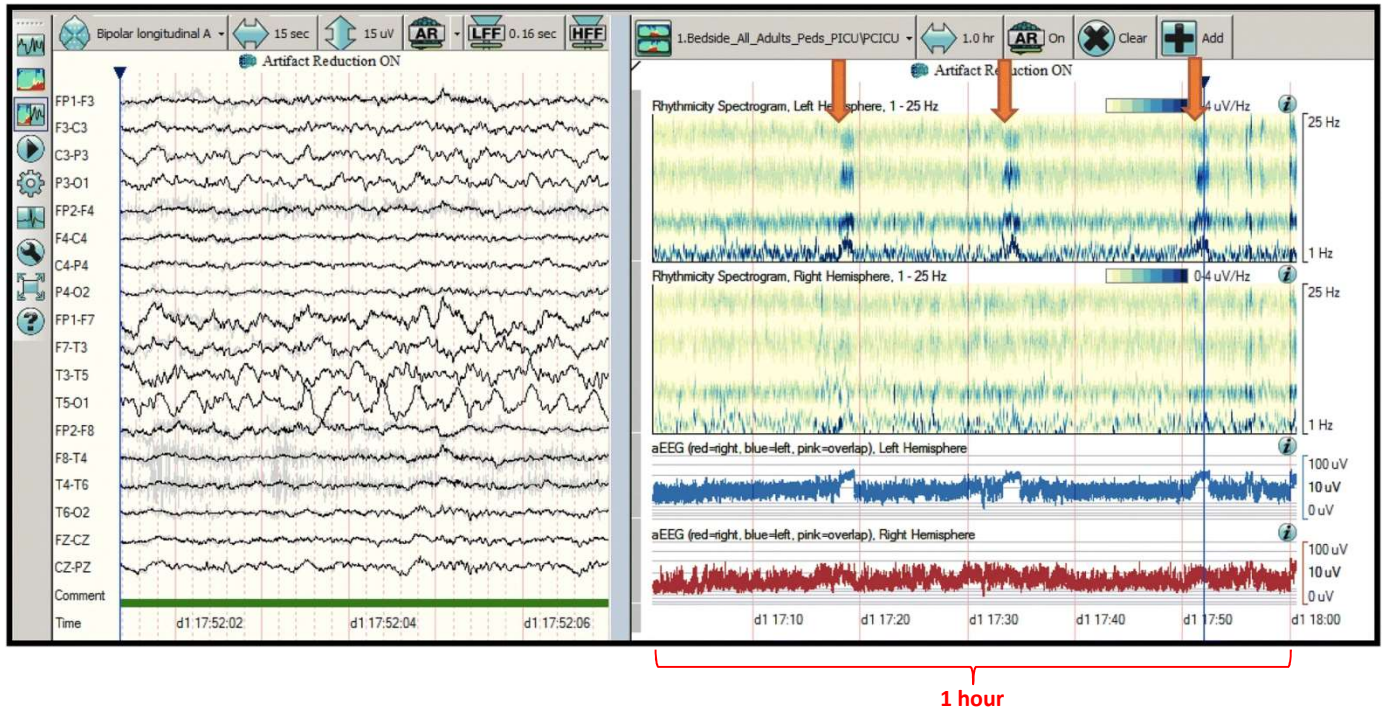
Neurocrit Care (2019) 31:312–320
https://doi.org/10.1007/s12028-019-00673-z

NEUROCRITICAL CARE SOCIETY

ORIGINAL WORK

A Trial of Real-Time Electrographic Seizure Detection by Neuro-ICU Nurses Using a Panel of Quantitative EEG Trends

Jennifer H. Kang[1*], G. Clay Sherill[1], Saurabh R. Sinha[1,2] and Christa B. Swisher[1]

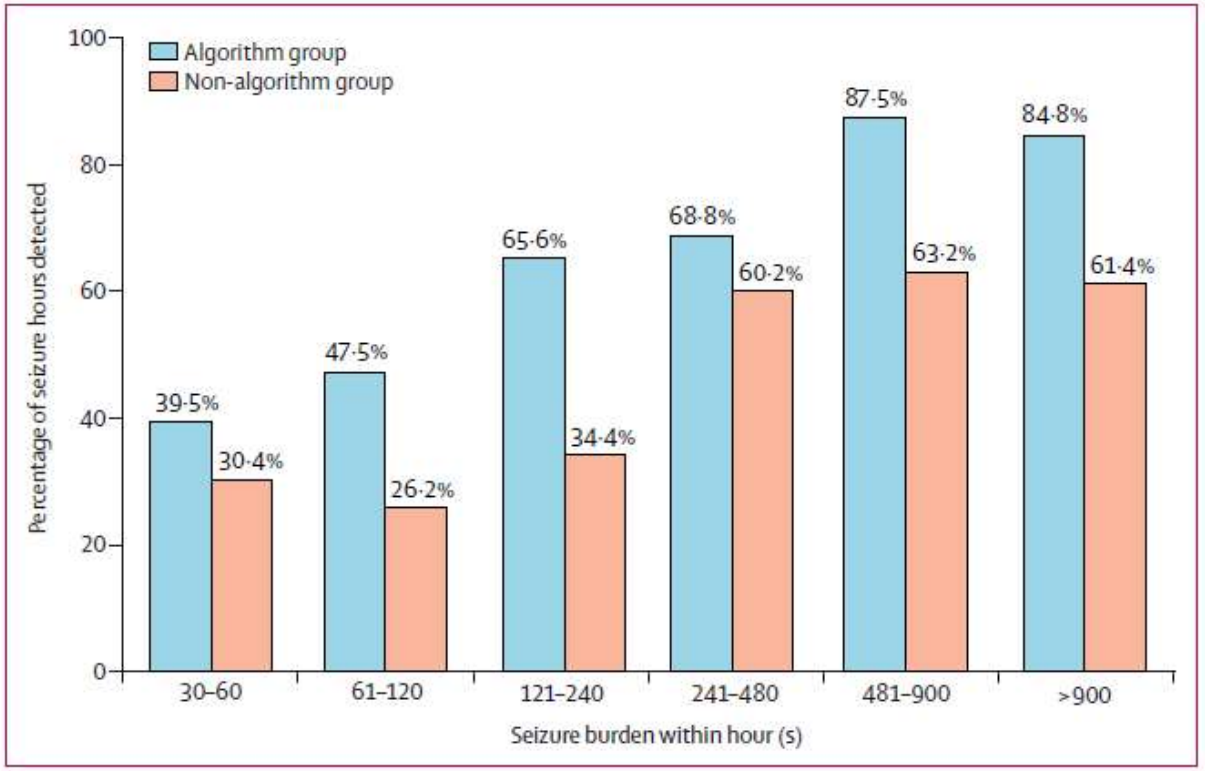


Lancet Child Adolesc Health 2020

Published Online
August 27, 2020

A machine-learning algorithm for neonatal seizure recognition: a multicentre, randomised, controlled trial

Andreea M Pavel, Janet M Rennie, Linda S de Vries, Mats Blennow, Adrienne Foran, Divyen K Shah, Ronit M Pressler, Olga Kapellou, Eugene M Dempsey, Sean R Mathieson, Elena Pavlidis, Alexander C van Huffelen, Vicki Livingstone, Mona C Toet, Lauren C Weeke, Mikael Finder, Subhabrata Mitra, Deirdre M Murray, William P Marnane, Geraldine B Boylan



Remote Teamwork Management of NORSE During the COVID-19 Lockdown

Alberto Cossu, MD*, Tommaso Lo Barco, MD*, Francesca Darra, MD, Elena Fontana, MD, Elena Fiorini, MD, Martina Marangone, Paolo Biban, MD, Bernardo Dalla Bernardina, MD, and Gaetano Cantalupo, MD

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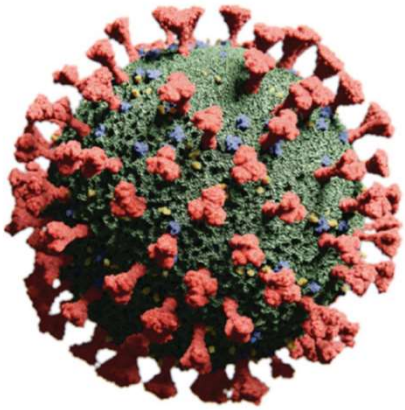

Grazie

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