
## Giovedì, 25 novembre

### Maturazione EEG

| | |
|---|---|
| 09:15 – 10:00 | Ontogenesi EEG – *A. Suppiej* |
| 10:00 – 10:45 | Pattern EEG patologici nel neonato pretermine/a termine – *S. Lori* |
| 10:45 – 11:30 | EEG nel bambino: peculiarità rispetto all'adulto – *P. Lanteri* |
| 11:30 – 12:00 | Pausa |
| 12:00 – 13:00 | Crisi epilettiche ed epilessie neonatali. Interpretazione della VideoEEG poligrafica *M. Mastrangelo* |

*Massimo Mastrangelo*

*Terapia intensiva post operatoria pediatrica – TIPO, IRCCS Policlinico San Donato*

*(UOC Neurologia pediatrica, Ospedale Buzzi, Milano)*


Gino Strada
Sesto san Giovanni, 21 aprile 1948
Rouen, 13 agosto 2021

**SOSTENIAMO EMERGENCY**

# Presentazione del Corso

…….. A distanza di circa 100 anni dalla sua prima descrizione ***l'EEG costituisce tuttora l principale metodica di studio funzionale del cervello*** e con l'enorme sviluppo delle tecniche digitali sono oggi possibili molteplici integrazioni con altri esami neurofisiologici e di neuroimmagine. Dal punto di vista clinico l'EEG rappresenta l'esame ***"gold standard" in ambito epilettologico, sia per la diagnosi che per il management delle svariate sindromi epilettiche***, ma oltre che per lo studio delle Epilessie l'EEG riveste notevole importanza per il ***neuromonitoraggio in area critica*** e in camera operatoria e per la ***valutazione sia clinica che prognostica di molte encefalopatie, acute e croniche, in diverse epoche della vita***……

Buon lavoro a tutti!

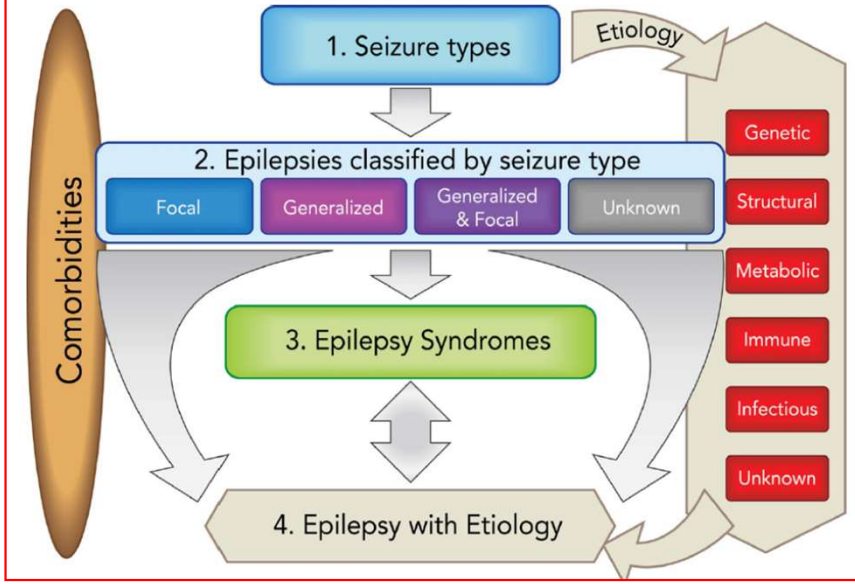
# ELETTROENCEFALOGRAFIA CLINICA

- ✓ **Crisi epilettiche neonatali**
- ✓ **Epilessie neonatali**
- ✓ **VideoEEG poligrafia**

## Sottotitolo

**Dalla semeiologia elettroclinica della crisi, alla diagnosi (eziologica e sindromica) del tipo di condizione epilettica**

1.




**2017**

2. il neonato, normale e patologico

3. l'EEG del neonato, normale e patologico

Prerequisiti:

Conoscenza di

1

e

2

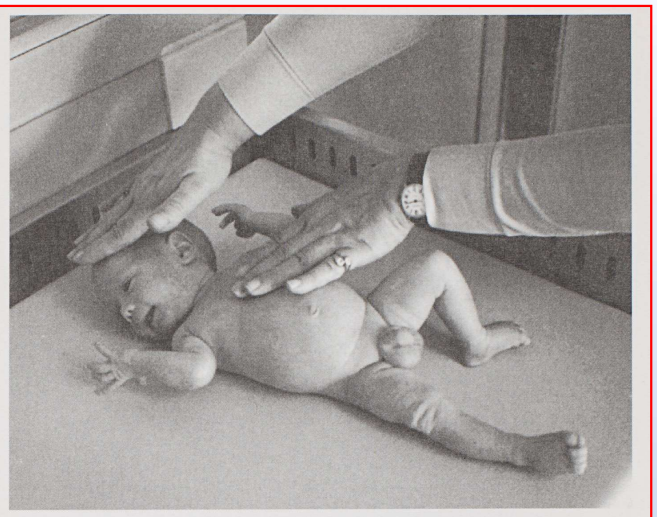


**Figura 7** Il riflesso tonico asimmetrico.

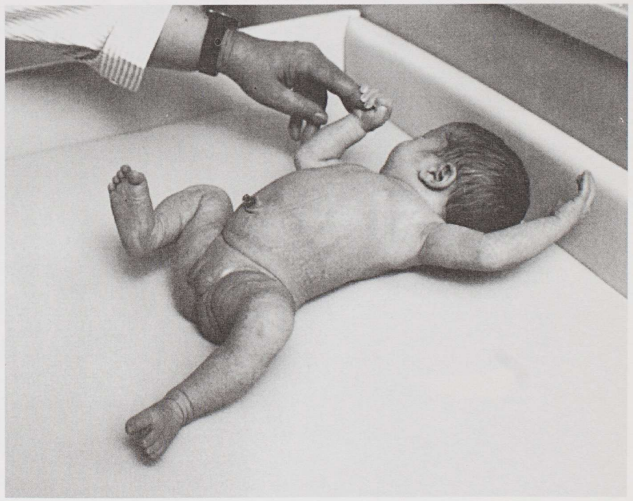


**Figura 8** Il grasp delle mani.

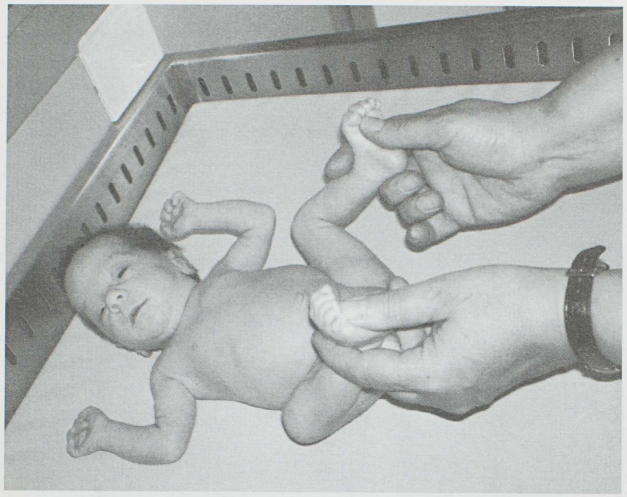


**Figura 9** Il grasp dei piedi.

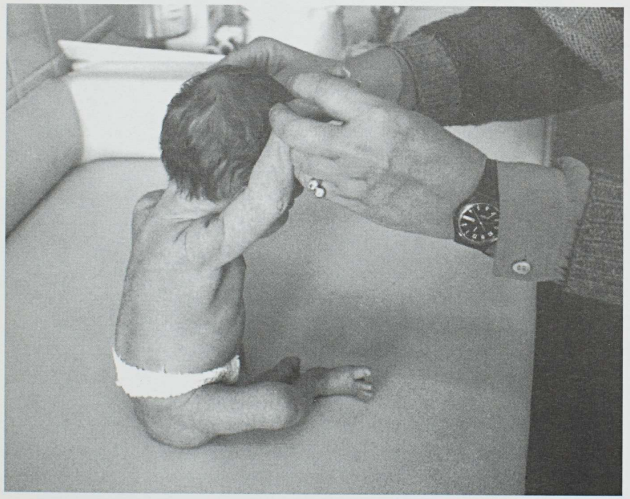
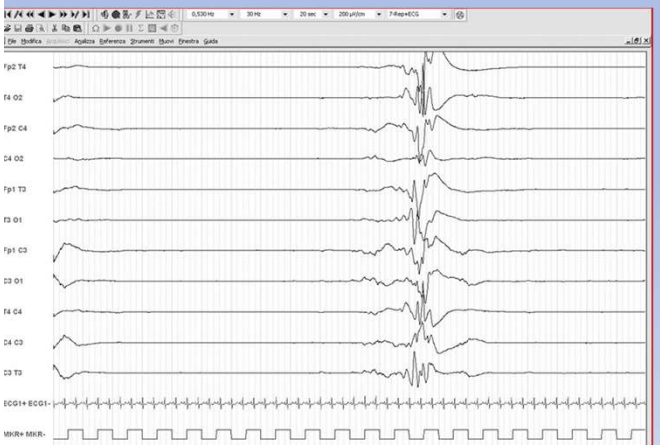


**Figura 10** Valutazione da seduto.

*(Prof.ssa Romana Negri)*



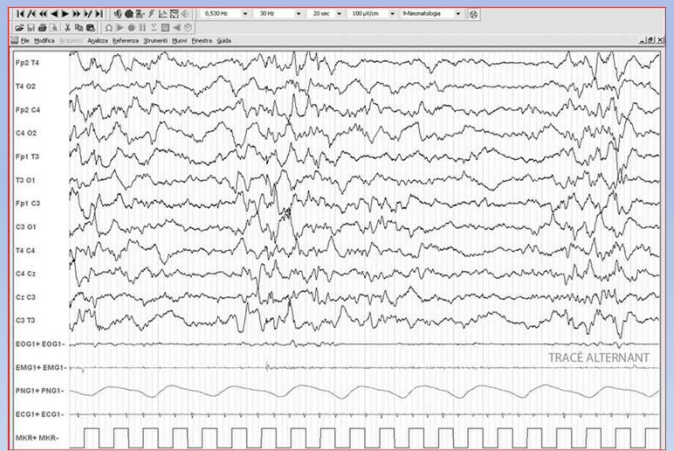


Figura 20 La valutazione del follow up.

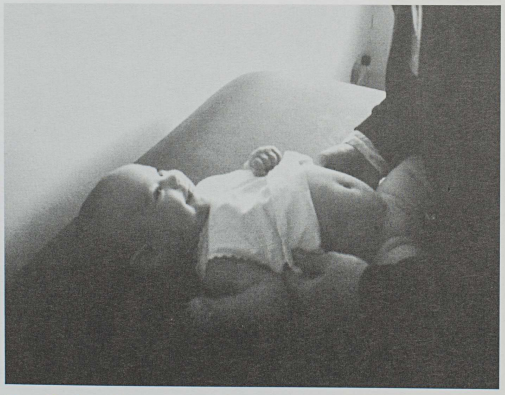
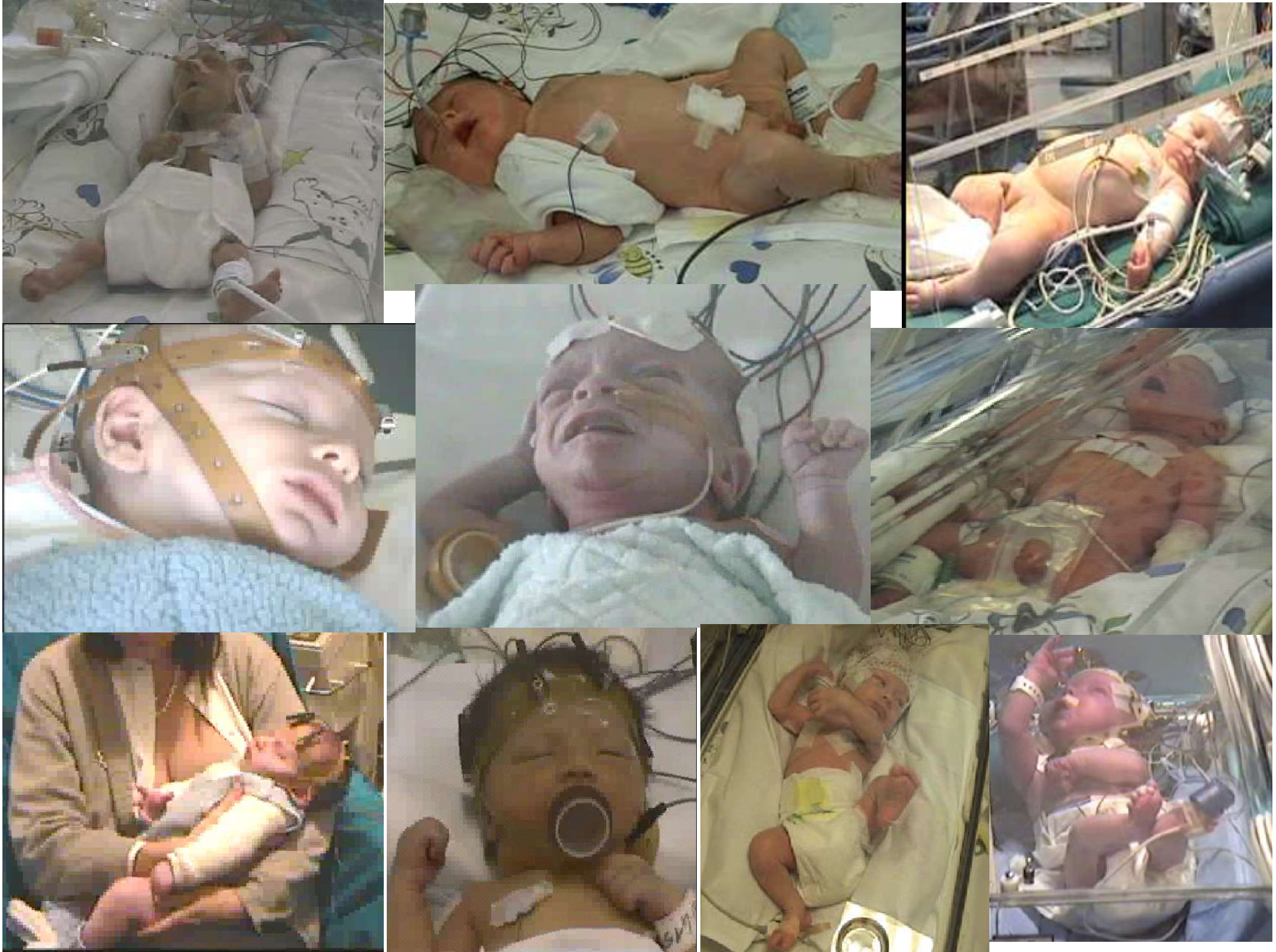


Figura 21 La madre spoglia il bambino.


*Nel neonato patologico ….. la registrazione EEG, videoEEG, long term monitoring a EEG/cEEG, ecc ecc, deve far parte dell'inquadramento neurologico*



Video_1

Video_2

Epilepsia

SPECIAL REPORT

The ILAE classification of seizures and the epilepsies: Modification for seizures in the neonate. Position paper by the ILAE Task Force on Neonatal Seizures

Ronit M. Pressler[1,2] | Maria Roberta Cilio[3] | Eli M. Mizrahi[4] | Solomon L. Moshé[5,6] | Magda L. Nunes[7] | Perrine Plouin[8] | Sampsa Vanhatalo[9] | Elissa Yozawitz[5,6] | Linda S. de Vries[10] | Kollencheri Puthenveettil Vinayan[11] | Chahnez C. Triki[12] | Jo M. Wilmshurst[13] | Hitoshi Yamamoto[14] | Sameer M. Zuberi[15]

**2021**

Video_1

Video_2

**MPNE**

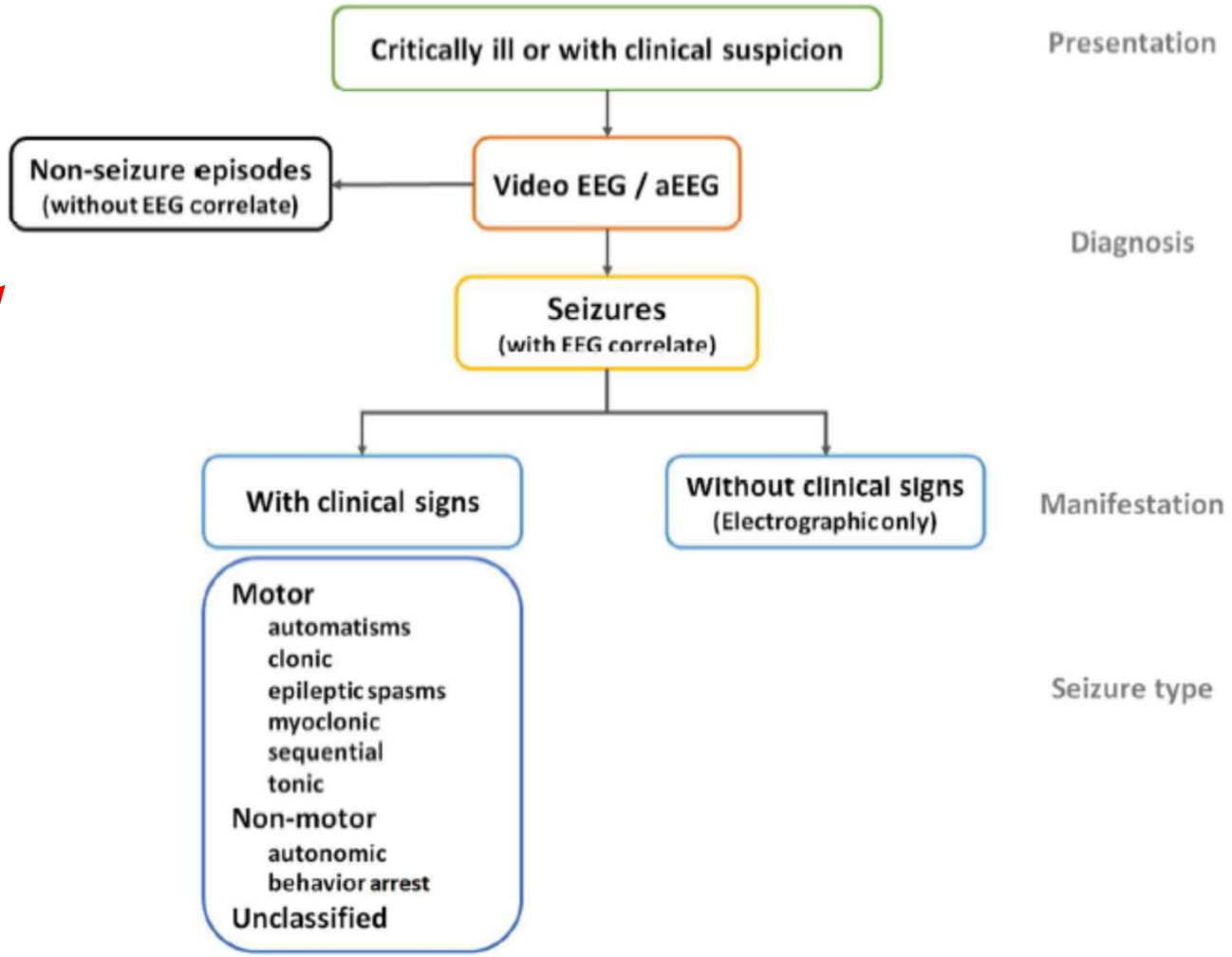
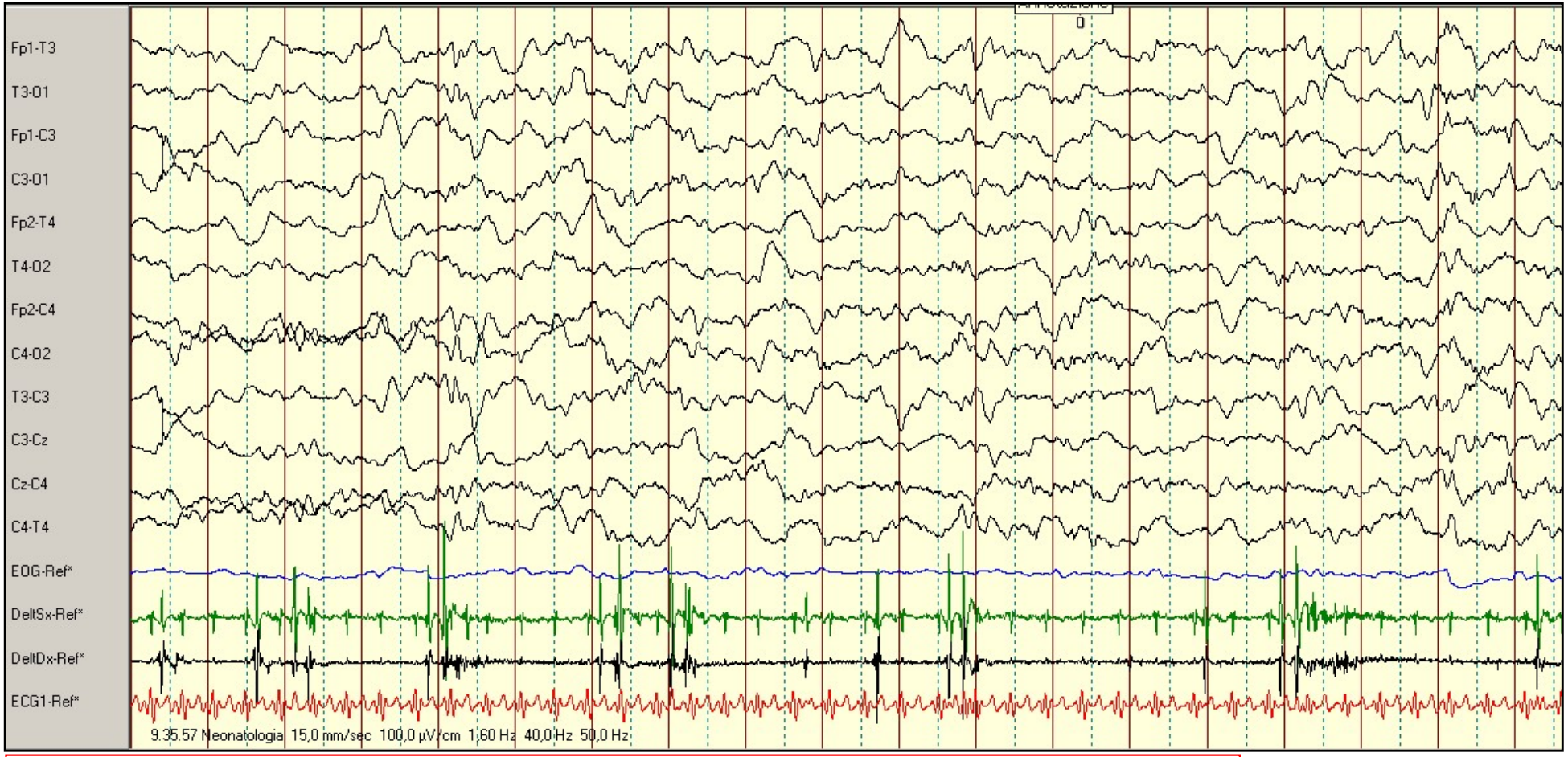



Figure 2: Proposed diagnostic framework of seizures in the neonatal period including classification of seizures. Adapted from 2017 ILAE seizure classification.3 Neonates present with discrete events suspected to be epileptic seizures or are critically ill (often ventilated, sedated and treated with muscle relaxants in intensive care).

**41.3 w CA > HIE + CLONIC, MYOCLONIC, SPASMS > PB 20 mg/Kg/EV > worsening > 4 > 5 > 6 mg/Kg/OS > worsening > 13 days old: recurrent spasms (associated to normal EEG)**




# Benign neonatal sleep myoclonus

**> PB withdrowal > clinical improvement > stop NEPE in 2 months**

**Seizures are the most common neurological emergency in the neonatal period occurring in 1 – 5 per 1000 live births**

1. **The majority of neonatal seizures are symptomatic** of an acute illness with an underlying etiology either documented or suspected

2. **Epilepsy syndromes may present in the neonatal period** and with the increasing availability of genetic testing **expanding numbers of neonatal epilepsies** with genetic and metabolic etiologies are recognized.

*Ronen et al,1999;Glass et al.,2016; Shellhaas et.,2017*

1943–1999

# CRISI

*La definizione dell'evento critico*

*nella sua evoluzione anatomo – elettro – clinica*

*è la strada che porta alla definizione*

*del tipo di epilessia (condizione epilettica)*

*del singolo paziente*

# Epileptic Seizure Type

# Tipo di Crisi Epilettica

Concetto ripreso dal 2001……

# 2001

*Epilepsia*, **42**(6):796–803, 2001
Blackwell Science, Inc.


ILAE Commission Report

# A Proposed Diagnostic Scheme for People with Epileptic Seizures and with Epilepsy: Report of the ILAE Task Force on Classification and Terminology

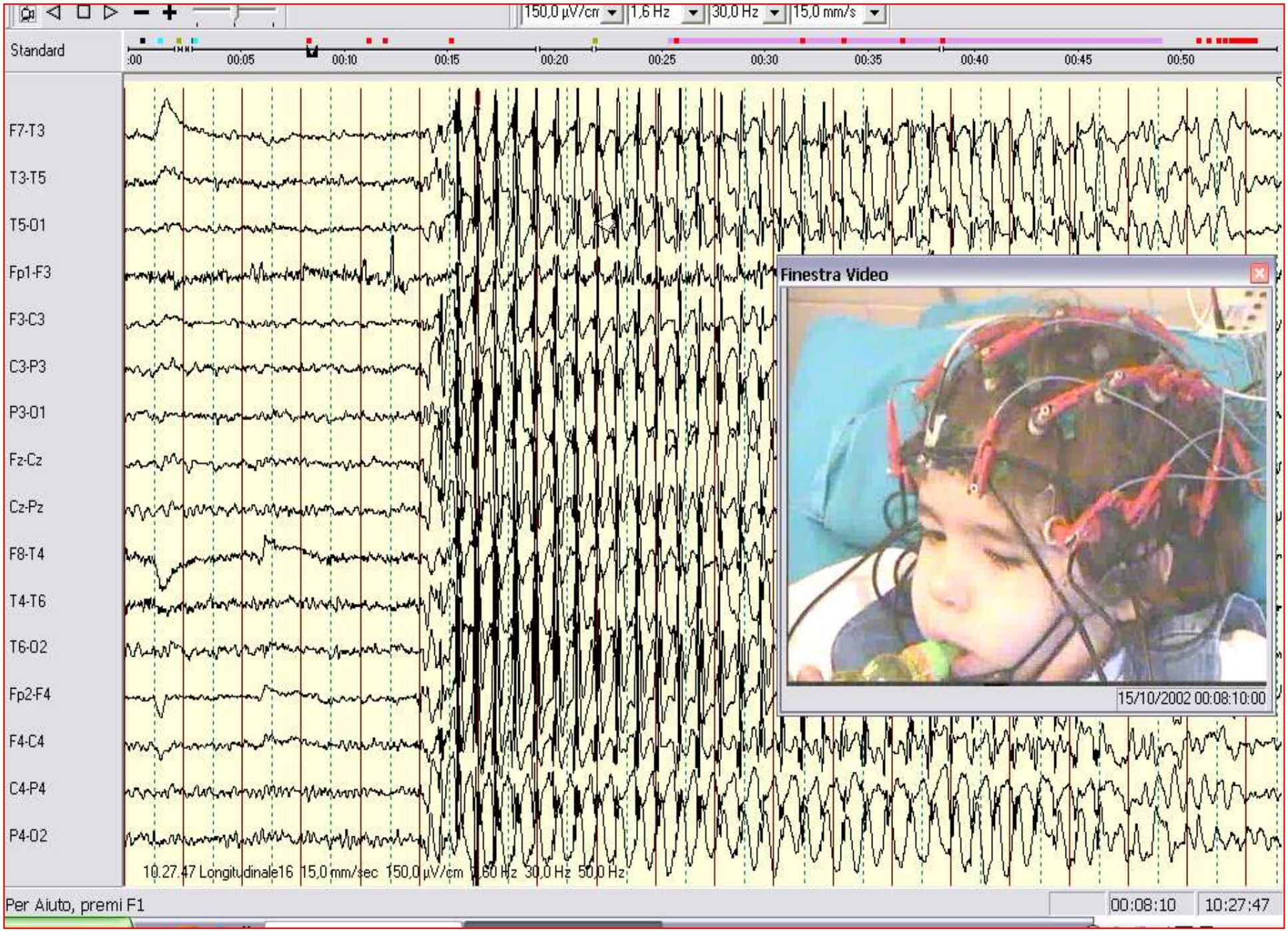
Jerome Engel, Jr.

| AXIS | 1 | > | 2 | > | 3 | > | 4 | > | 5 |
|---|---|---|---|---|---|---|---|---|---|
| SCHEME | Glossary | > | Seizure types | > | Epilesy Syndromes | > | Etiology | > | Impairment |

**Axis 2: is the *epileptic seizure type, or types*, experienced by the patient, derived from a List of Accepted Epileptic Seizure Types*, that *represent diagnostic entities with etiologic, therapeutic, and/or prognostic implications*. Localization within the brain should be specified when this is appropriate.**

*Epilepsia, 42(6), 2001

# LE CRISI – IL TIPO DI CRISI



ILAE POSITION PAPER

# ILAE 2017 Classification of Seizure Types Basic Version [1]

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

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

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

Focal Onset

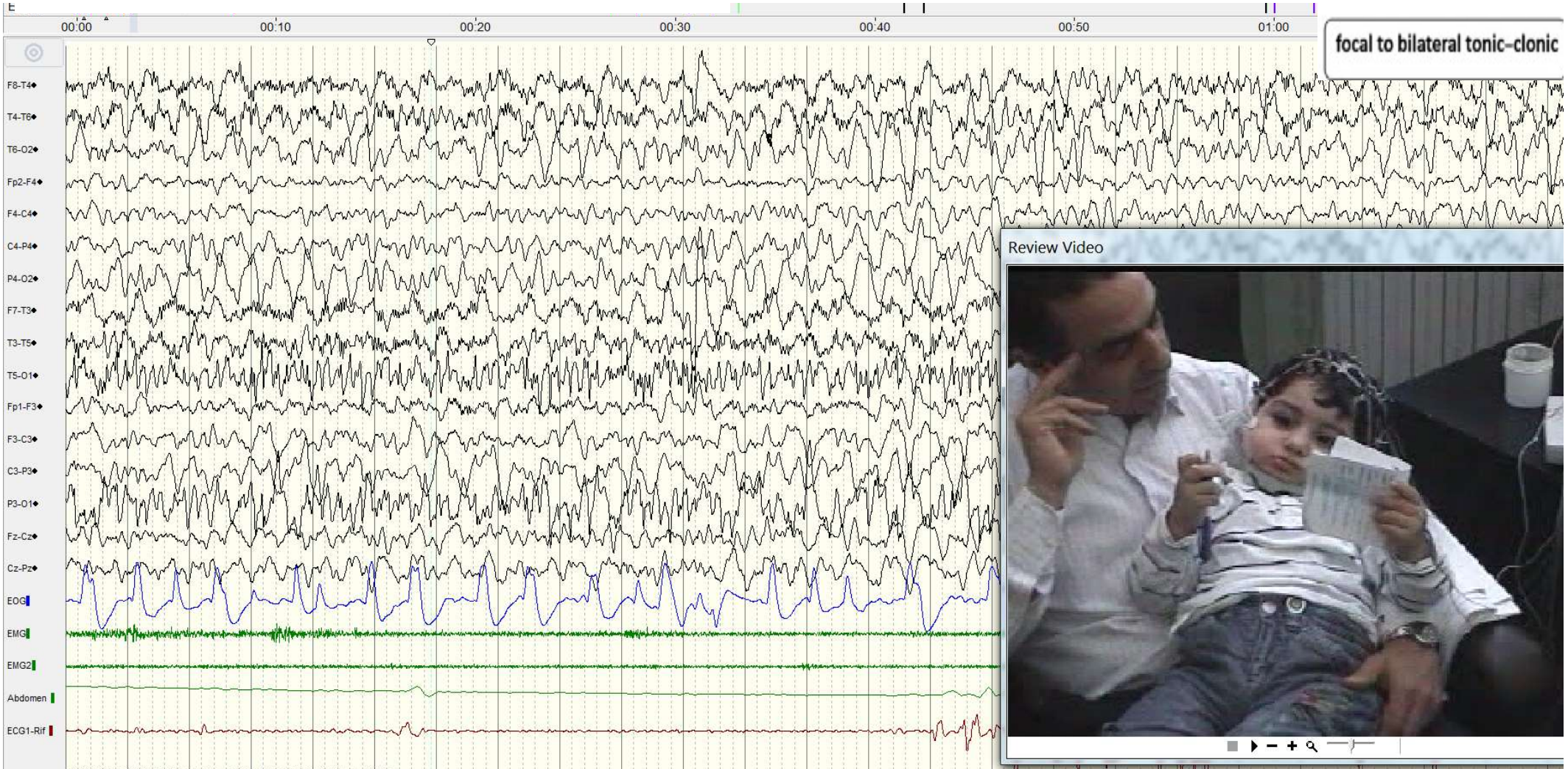
Aware | Impaired Awareness

Motor Onset

Nonmotor Onset

focal to bilateral tonic–clonic

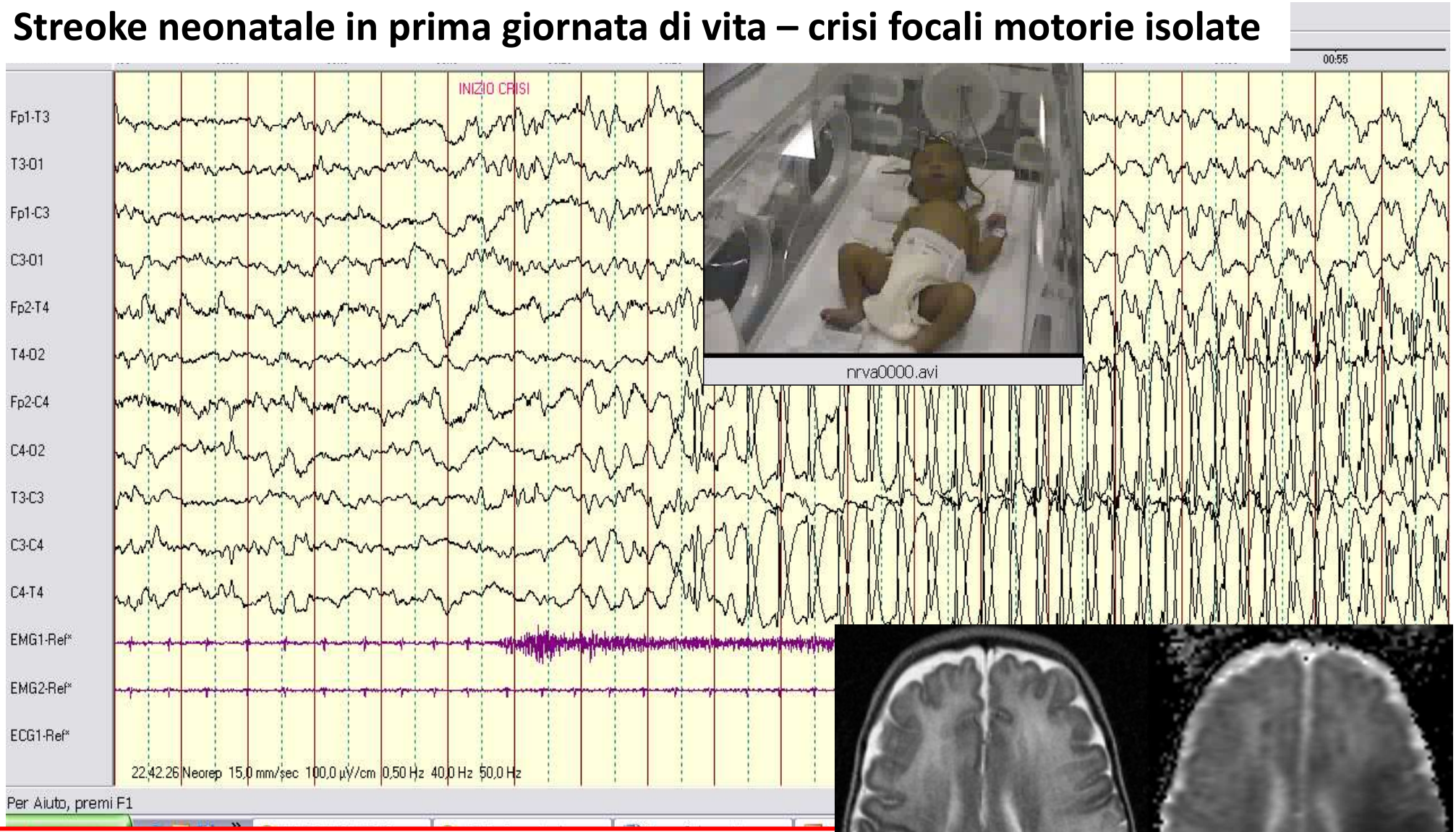
TM 3 anni – 17.4.2010 – ore 14.51



# LE CRISI – IL TIPO DI CRISI

## Neonato a termine

## Streoke neonatale in prima giornata di vita – crisi focali motorie isolate




**CLONIC SEIZURES**

**FOCAL SYMPTOMATIC (STROKE) ISOLATED SEIZURES**

***1. le crisi epilettiche neonatali***

***2. le epilessie neonatali***

***CRISI EPILETTICA:*** Scarica parossistica, eccessiva, di una popolazione neuronale, più o meno estesa (focale o generalizzata), che comporta la modificazione, parossistica, di un comportamento e/o di una funzione

***Epileptic Seizure Type***: An ictal event believed to represent a unique pathophysiological mechanism and anatomical substrate. This is a diagnostic entity with etiological, therapeutic, and prognostic implications. (new concept)

***Engel JJ, ILAE 2001***

## *EPILESSIA:*

condizione patologica cronica,

a prognosi estremamente variabile,

caratterizzata da eventi acuti che si ripetono,

le crisi,

(senza) fattori acuti scatenanti

## *Epileptic Disorder:*

a chronic neurological condition characterised by recurrent epileptic seizures.

## *Epilepsies:*

Those conditions involving chronic recurrent epileptic seizures that can be considered epileptic disorders.

***Blume et al., ILAE 2001***

# DIAGNOSI EPILETTOLOGICA IN EPOCA NEONATALE

(ILAE 2001, 2006, 2010, 2017)

*Manifestazioni parossistiche non epilettiche*

Crisi epilettiche "benigne" occasionali

Crisi epilettiche sintomatiche occasionali

Epilessia benigna familiare neonatale

EME encefalopatia mioclonica precoce

EIEE encefalopatia infantile epilet. precoce

MPSI crisi parziali migranti dell'infanzia (EIMFS)

*Epilessia neonatale sintomatica (causa nota)*

*Epilessia neonatale sintomatica (causa ignota > genetica)*

*DEE developmental and epileptic encephalopathy*

## Table 3. Electroclinical syndromes and other epilepsies

Epilepsia, 51(4):676–685, 2010

Electroclinical syndromes arranged by age at onset[a]

- 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
- Childhood
  - Febrile seizures plus (FS+) (can start in infancy)
  - 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)[b]
  - Landau-Kleffner syndrome (LKS)
  - Childhood absence epilepsy (CAE)
- 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

Distinctive constellations

- Mesial temporal lobe epilepsy with hippocampal sclerosis (MTLE with HS)
- Rasmussen syndrome
- Gelastic seizures with hypothalamic hamartoma
- Hemiconvulsion–hemiplegia–epilepsy

Epilepsies that *do not* fit into any of these diagnostic categories can be distinguished first on the basis of the presence or absence of a known structural or metabolic condition (presumed cause) and then on the basis of the primary mode of seizure onset (generalized vs. focal)

Epilepsies attributed to and organized by structural-metabolic causes

- Malformations of cortical development (hemimegalencephaly, heterotopias, etc.)
- Neurocutaneous syndromes (tuberous sclerosis complex, Sturge-Weber, etc.)
- Tumor
- Infection
- Trauma
- Angioma
- Perinatal insults
- Stroke
- Etc.

Epilepsies of unknown cause

Conditions with epileptic seizures that are traditionally not diagnosed as a form of epilepsy per se

- Benign neonatal seizures (BNS)
- Febrile seizures (FS)

[a]The arrangement of electroclinical syndromes does not reflect etiology.

[b]Sometime referred to as Electrical Status Epilepticus during Slow Sleep (ESES).

# CRISI & EPILESSIE AD ESORDIO NEONATALE

207 NEONATI CON CRISI EPILETTICHE

**165** Crisi neonatali sintomatiche acute (con Insulto cerebrale acuto)

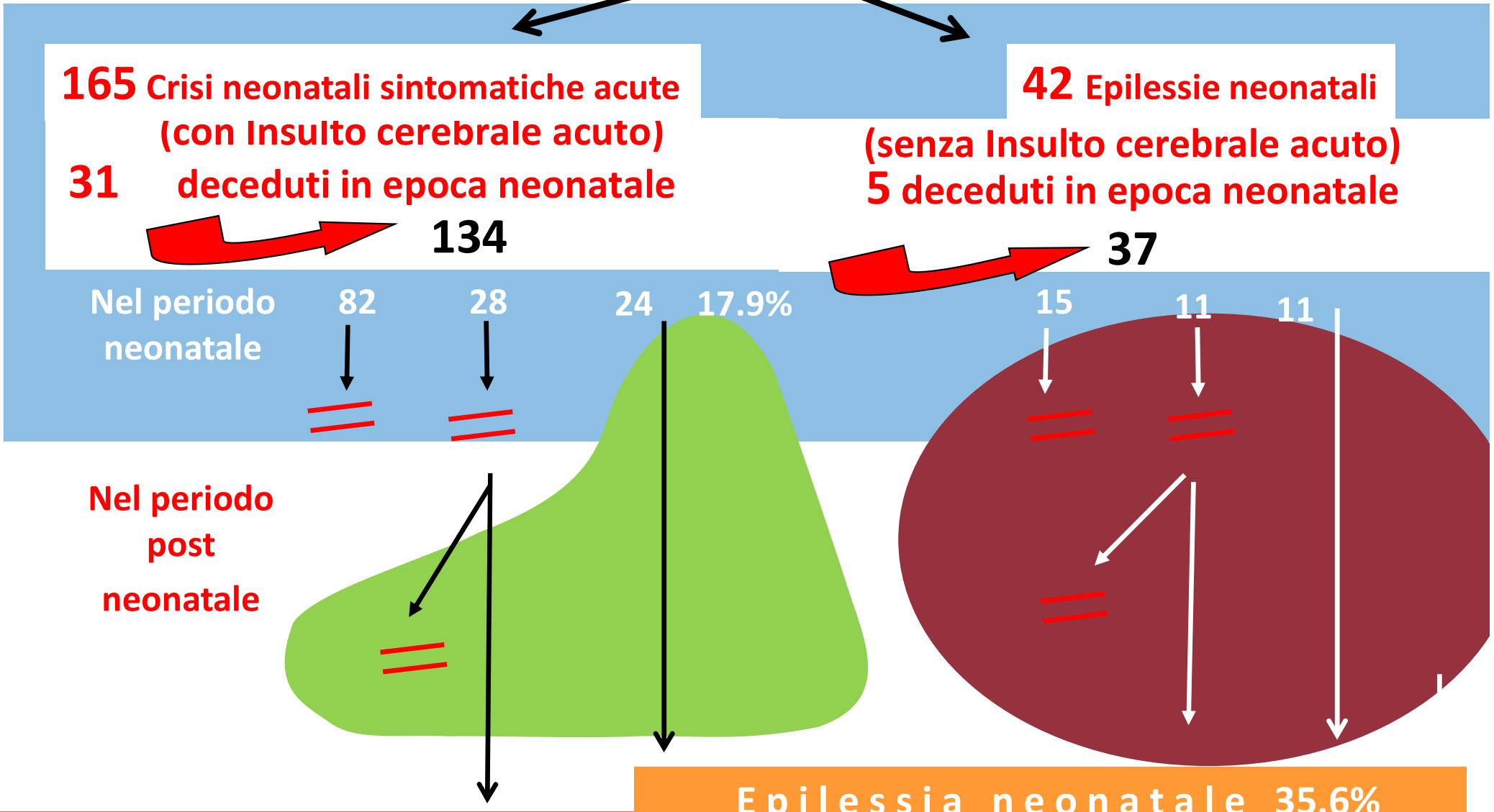
**31** deceduti in epoca neonatale

**134**

**42** Epilessie neonatali (senza Insulto cerebrale acuto)

**5** deceduti in epoca neonatale

**37**




**Epilessia post neonatale 20.9%**

**Epilessia neonatale 35.6%**

# LE CRISI EPILETTICHE NEONATALI SINTOMATICHE

*Epilepsia*, 51(4):671–675, 2010
doi: 10.1111/j.1528-1167.2009.02285.x

**SPECIAL REPORT**

## Recommendation for a definition of acute symptomatic seizure

*[1]Ettore Beghi, †Arturo Carpio, ‡Lars Forsgren, §Dale C. Hesdorffer, ¶Kristina Malmgren, #Josemir W. Sander, **Torbjorn Tomson, and §W. Allen Hauser

### Definition of acute symptomatic seizures

Acute symptomatic seizures are events, occurring in close temporal relationship with an acute CNS insult, which may be metabolic, toxic, structural, infectious, or due to inflammation. The interval between the insult and the seizure may vary according to the underlying clinical condition (see subsequent text). The term acute symptomatic seizure should be used instead of provoked seizure, reactive seizure, or situation-related seizure.

*Suggestions are made to define acute symptomatic seizures as those events occurring within 1 week of .....*

### Definition of unprovoked seizures/epilepsy

Unprovoked seizures are defined as seizures occurring in the absence of a potentially responsible clinical condition or beyond the interval estimated for the occurrence of acute symptomatic seizures. Unprovoked seizures differ from acute symptomatic seizures in risk of seizure recurrence and mortality for several etiologies.

Epilepsia, 51(4):671–675, 2010

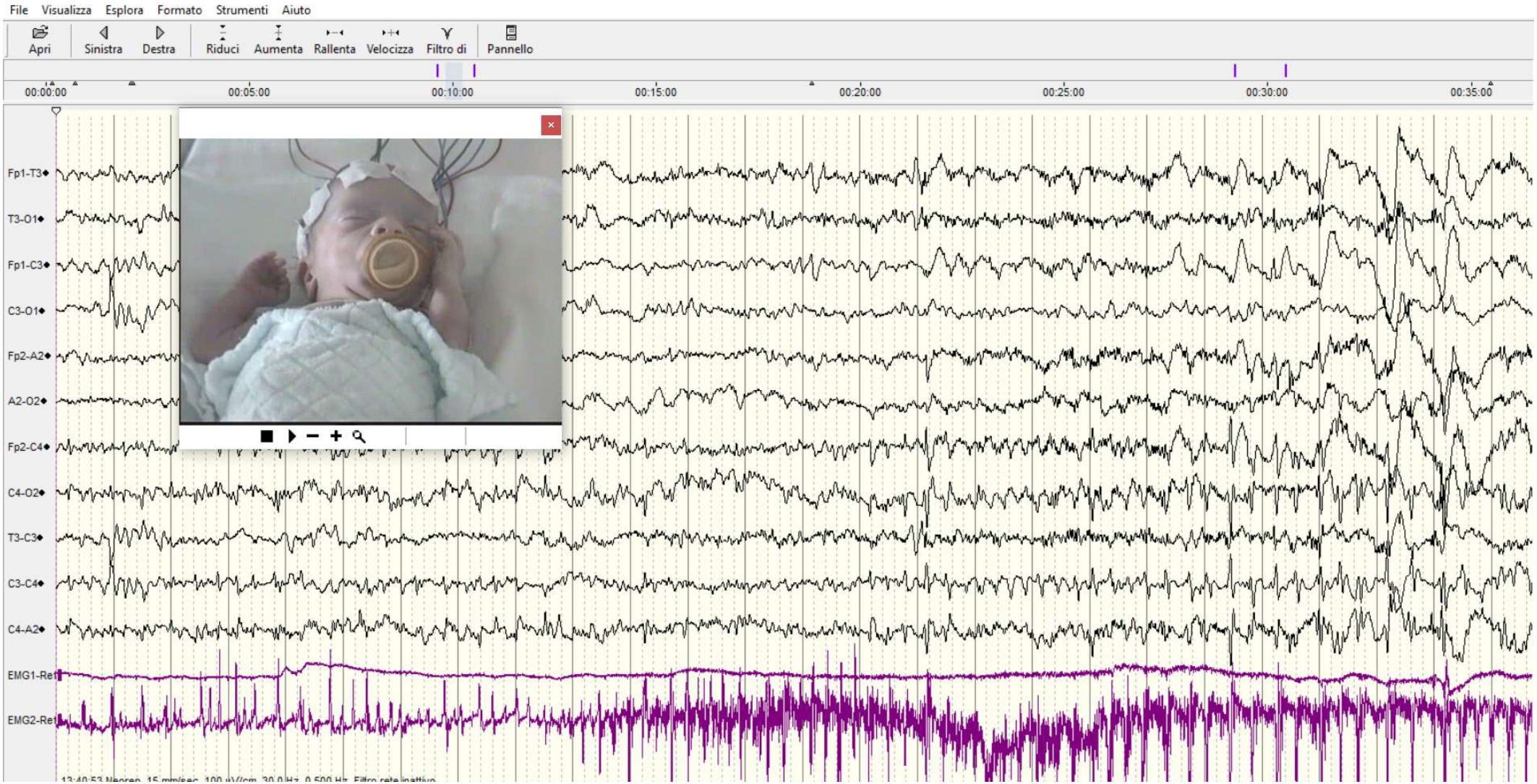
**Results:** ***An acute symptomatic seizure is defined as a clinical seizure occurring at the time of a systemic insult or in close temporal association with a documented brain insult***. Suggestions are made to define acute symptomatic seizures as ***those events occurring within 1 week (?)*** of stroke, traumatic brain injury, anoxic encephalopathy, or intracranial surgery; at first identification of subdural hematoma; at the presence of an active central nervous system (CNS) infection; or during an active phase of multiple sclerosis or other autoimmune diseases. ***In addition***, a diagnosis of acute symptomatic seizure should be made in the presence of ***severe metabolic derangements (documented within 24 h (?) by*** specific biochemical or hematologic abnormalities), drug or alcohol intoxication and withdrawal, or exposure to well-defined epileptogenic drugs

***Electroclinical syndrome:*** is a complex of clinical features, signs, and symptoms that together define a distinctive, recognizable clinical disorder.

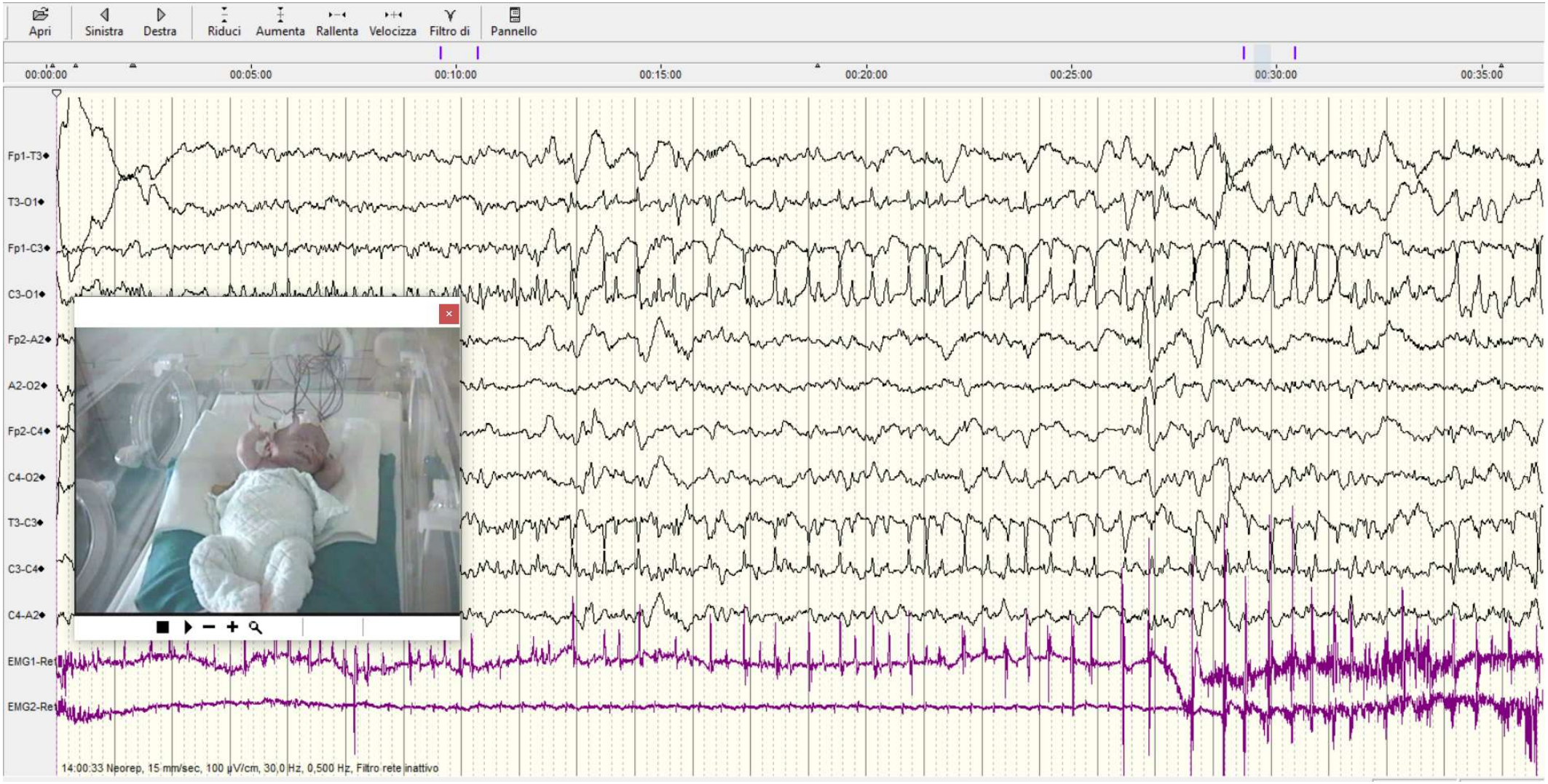
**Epilepsies associated with structural or metabolic conditions**: Previously, many such epilepsies were grouped together as "symptomatic focal epilepsies" and distinguished on the basis of localization……. Further organizations might consider type of lesion, age at onset, localization, seizure type, specific ictal and interictal EEG patterns, or other factors.

***Acute symptomatic seizure:*** a clinical seizure occurring at the time of a systemic insult or in close temporal association with a documented brain insult ..... ( *those events occurring within 1 week of ……* )

# *crisi epilettiche sintomatiche acute*

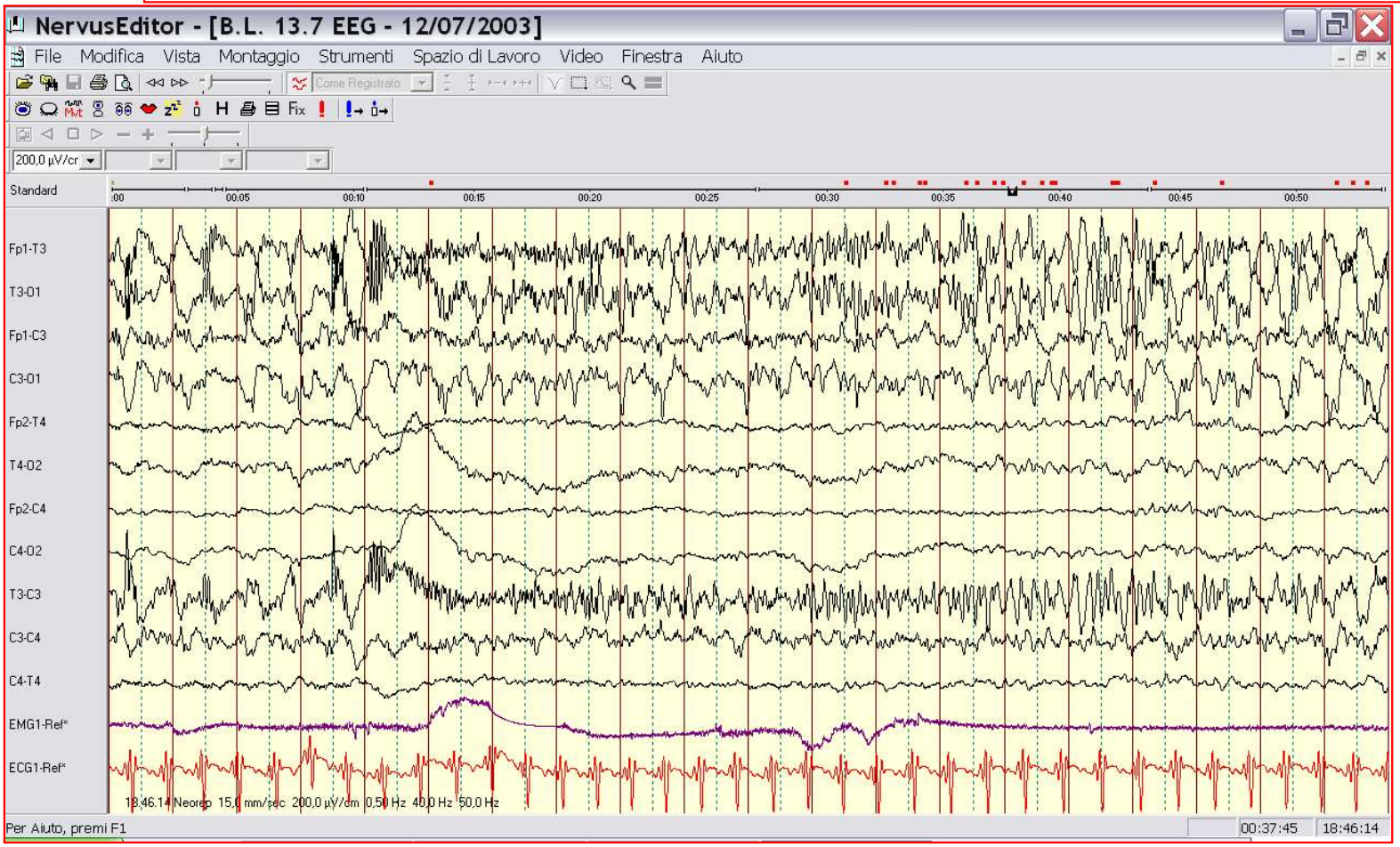


# *crisi epilettiche sintomatiche acute*



# *Epilessia focale sintomatica*

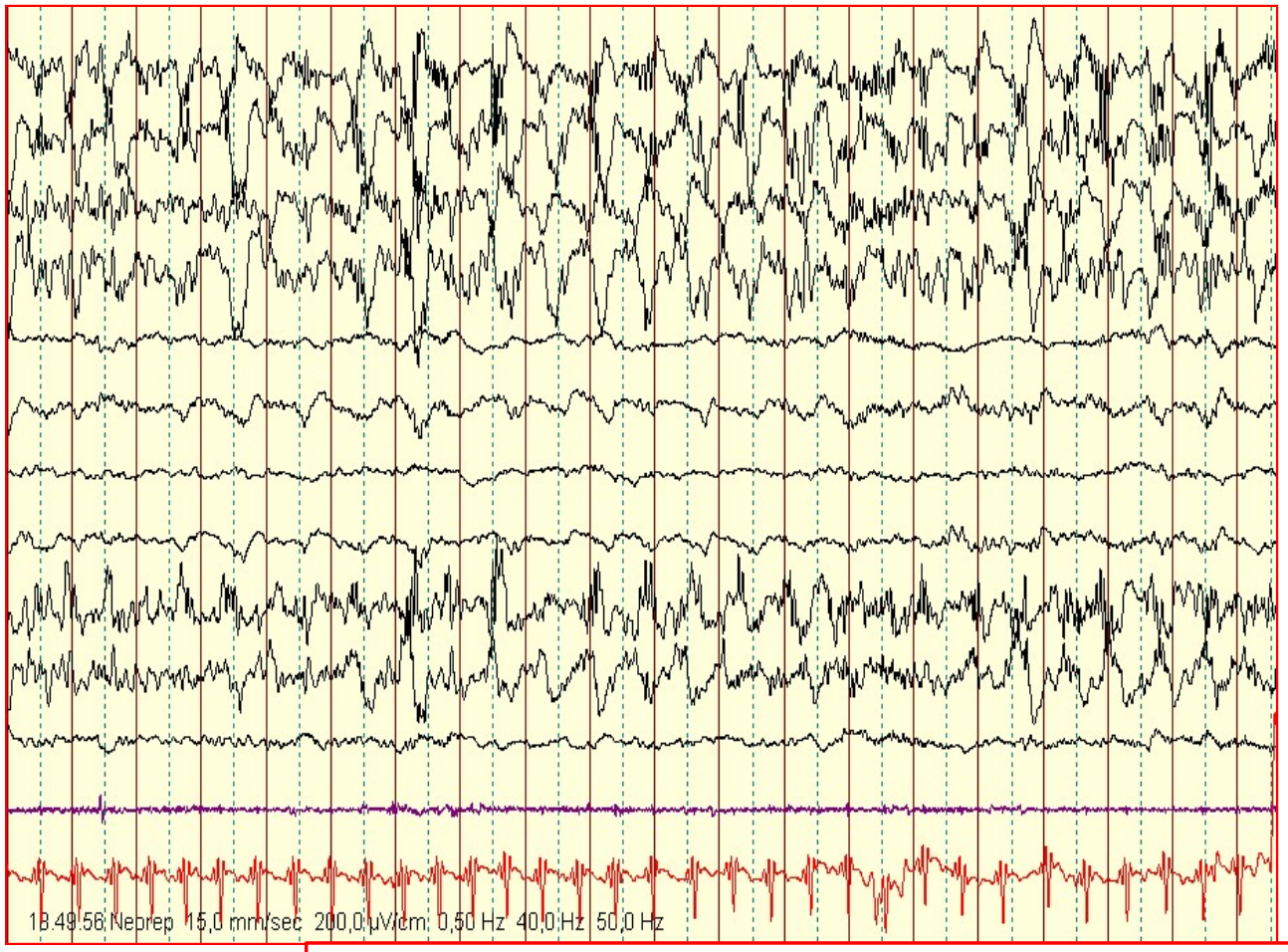
## 40 SETT EG – 40.4 SETT EC EMIMICROPOLIGIRIA A SINISTRA




- VEGLIA
- TRACCIATO COSTANTEMENTE ASIMMETRICO
- ATTIVITA' RAPIDA /P RAPIDE FOCALI SU TUTTO L'EMISFERO DI SINISTRA
- CRISI RICORRENTI FOCALI – SU Fp1 T3 ATTIVITA' BETA RECUTANTE DI BASSO VOLTAGGIO

# *Epilessia focale sintomatica*

## 40 SETT EG – 40.4 SETT EC EMIMICROPOLIGIRIA A SINISTRA




SONNO

TRACCIATO COSTANTEMENTE ASIMMETRICO

ATTIVITA' RAPIDA /P RAPIDE FOCALI A SINISTRA CHE SI VANNO A SOVRAPPORRE SOPRA ONDE LENTE FOCALI SU TUTTO L'EMISFERO DI SINISTRA

ASSENZA GRAFOELEMENTI FISIOLOGICI

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


**1987**

# The Exact Ictal and Interictal Duration of Electroencephalographic Neonatal Seizures

Robert R. Clancy and Agustin Legido

***Electrographic seizures were recognized as clear ictal events characterized by the appearance of sudden, repetitive, evolving stereotyped waveforms with a definite beginning, middle, and end.*** We chose an arbitrary minimum ictal duration of **10** s for inclusion in this study. The interictal periods were those portions of the tracing between the electrographic seizures in which no ictal patterns were seen.

NEUROLOGY 1987;37:1837-1844

**1987**

# Characterization and classification of neonatal seizures

Eli M. Mizrahi, MD, and Peter Kellaway, PhD

To characterize and classify neonatal seizures, we studied 349 neonates, ***using a portable, cribside EEG/polygraphic/video monitoring system.*** ***We recorded 415 clinical seizures in 71 infants; 11 other infants had electrical seizure activity without clinical accompaniments. Each seizure was analyzed in terms of its clinical character and its relationship to the presence of EEG seizure activity*****.....**

.....The clinical and background EEG features of infants whose seizures were not accompanied by EEG seizure activity suggest that these seizures may not be epileptic in character.

2021

Received: 8 August 2020 | Revised: 23 December 2020 | Accepted: 23 December 2020

DOI: 10.1111/epi.16815

Epilepsia®

SPECIAL REPORT

# The ILAE classification of seizures and the epilepsies: Modification for seizures in the neonate. Position paper by the ILAE Task Force on Neonatal Seizures

Ronit M. Pressler[1,2] | Maria Roberta Cilio[3] | Eli M. Mizrahi[4] | Solomon L. Moshé[5,6] | Magda L. Nunes[7] | Perrine Plouin[8] | Sampsa Vanhatalo[9] | Elissa Yozawitz[5,6] | Linda S. de Vries[10] | Kollencheri Puthenveettil Vinayan[11] | Chahnez C. Triki[12] | Jo M. Wilmshurst[13] | Hitoshi Yamamoto[14] | Sameer M. Zuberi[15]

## 2 | INTRODUCTION

Seizures are the most common neurological emergency in the neonatal period, occurring in 1–5 per 1000 live births ……

*Epilepsia*. 2021;62:615–628.

**Key points**

- The International League Against Epilepsy (ILAE) presents a new classification and framework for seizures in the neonatal period in line with 2017 ILAE classifications.
- It emphasizes the key role of electroencephalography (EEG) for the diagnosis of seizures in this age group.
- Seizures are considered focal at onset, and thus a division into focal and generalized is unnecessary.
- Seizures can occur with clinical manifestations or without clinical manifestations (electrographic-only).
- Descriptors are determined by the predominant clinical feature and divided into motor, non-motor, and sequential.

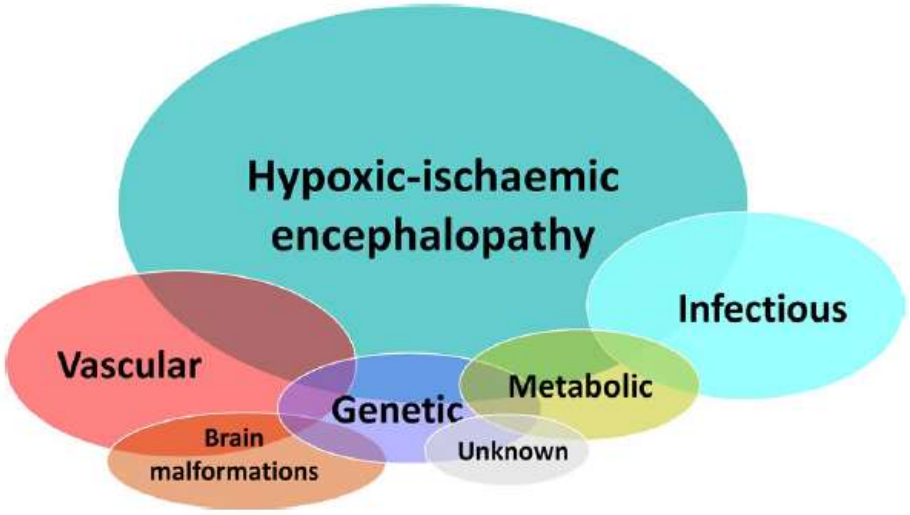



**FIGURE 1** Relative occurrences of common etiologies of neonatal seizures in term infants. Adapted from [3–5,8,81,82]

*Epilepsia*. 2021;62:615–628.



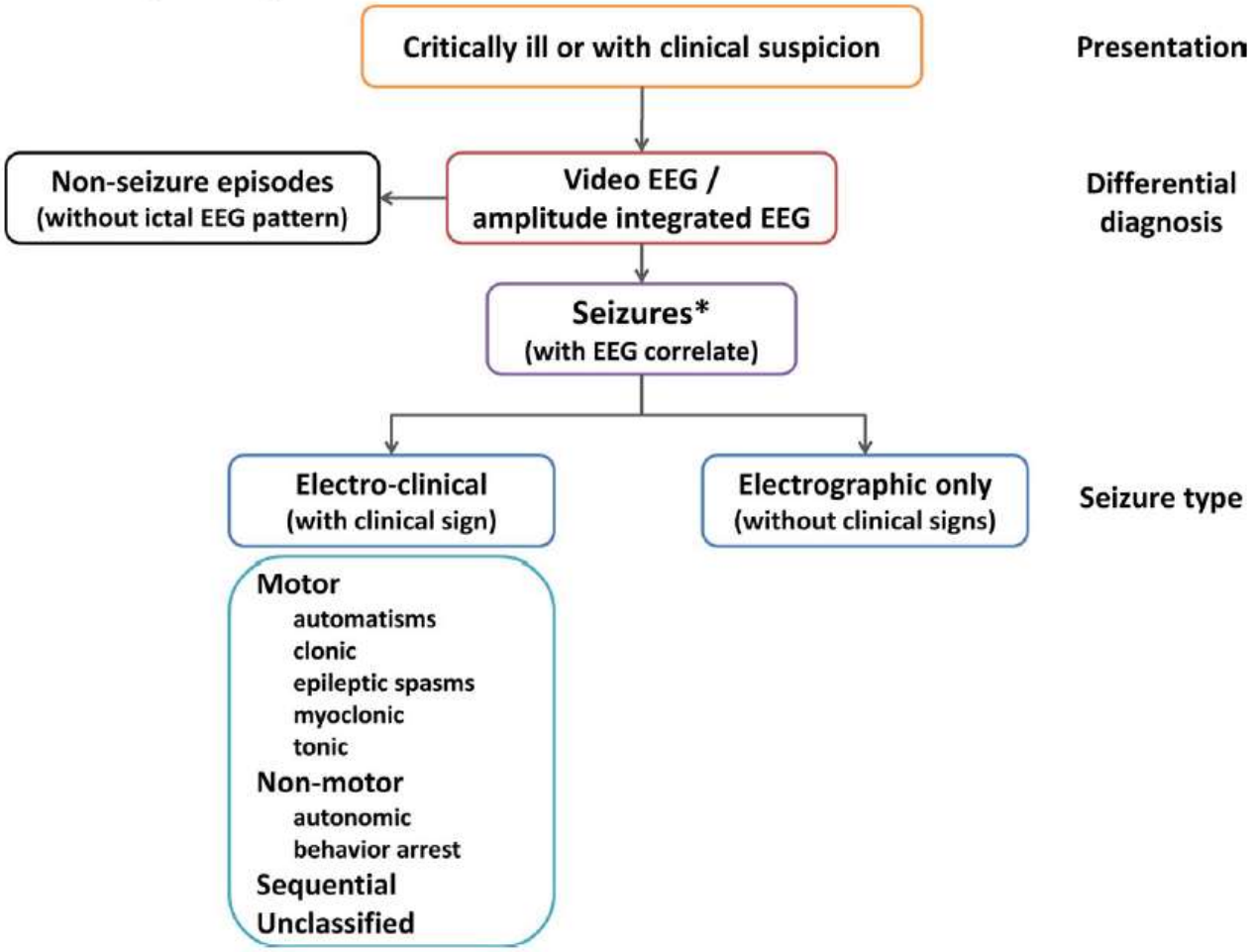



**FIGURE 2** Diagnostic framework of seizures in the neonatal period including classification of seizures. Adapted from 2017 ILAE seizure classification[7] Neonates present with discrete events suspected to be epileptic seizures or are critically ill (often ventilated, sedated, and treated with muscle relaxants in intensive care). *If no EEG available refer to global alignment of immunization safety assessment in pregnancy levels of diagnostic certainty (Figure 5)

*Epilepsia*. 2021;62:615–628.

**TABLE 1** Integration with the 2017 ILAE Classification of Seizures and considerations for neonates.

| Type | Description[6,7] | Special considerations | Clinical context of seizure type |
|---|---|---|---|
| Automatisms | A more or less coordinated motor activity usually occurring when cognition is impaired. This often resembles a voluntary movement and may consist of an inappropriate continuation of preictal motor activity. | Typically oral in neonates. Behavior in term and preterm infants may mimic ictal automatisms, thus EEG / aEEG mandatory. | Seen in HIE and preterm infants. Often part of sequential seizures. |
| Clonic | Jerking, either symmetric or asymmetric, that is regularly repetitive and involves the same muscle groups. | Seizure type, which is more reliably diagnosed clinically. | Typical seizure type in neonatal stroke or cerebral hemorrhage. May be seen in HIE. |
| Epileptic spasms | A sudden flexion, extension, or mixed extension–flexion of predominantly proximal and truncal muscles that is usually more sustained than a myoclonic movement but not as sustained as a tonic seizure. Limited forms may occur: Grimacing, head nodding, or subtle eye movements. | Brief in neonates, thus may be difficult to differentiate from myoclonic seizures without EMG channel. May occur in clusters. | Rare. May be seen in inborn errors of metabolism or early-infantile DEE. |
| Myoclonic | A sudden, brief (< 100 msec) involuntary single or multiple contraction(s) of muscles(s) or muscle groups of variable topography (axial, proximal limb, distal). | Clinically difficult to differentiate from non-epileptic myoclonus, requires EEG, ideally with EMG channels. | Typical seizure type in inborn errors of metabolism and preterm infants. May also be seen in early-infantile DEE. |
| Tonic | A sustained increase in muscle contraction lasting a few seconds to minutes. | Focal, unilateral or bilateral asymmetric. Generalized tonic posturing not of epileptic origin. | Typical seizure type early-infantile DEE and genetic neonatal epilepsies. |
| Autonomic | A distinct alteration of autonomic nervous system function involving cardiovascular, pupillary, gastrointestinal, sudomotor, vasomotor, and thermoregulatory functions. | May involve respiration (apnea). EEG / aEEG mandatory. | Rare in isolation. Seen in intraventricular hemorrhage as well as temporal or occipital lobe lesions. Also described in early-infantile DEE. |
| Behavioral arrest | Arrest (pause) of activities, freezing, immobilization, as in behavior arrest seizure. | EEG / aEEG mandatory. | Rare as an isolated seizure type. More commonly seen as part of sequential seizure. |
| Sequential seizure | This term is used in the instruction manual for the ILAE 2017 operational classification of seizure types for events with a sequence of signs, symptoms, and EEG changes at different times.[6] | No predominant feature can be determined, instead the seizure presents with a variety of clinical signs. Several features typically occur in a sequence, often with changing lateralization within or between seizures. | Often seen in genetic epilepsies such as self-limited neonatal epilepsy or KCNQ2 encephalopathy. |
| Electrographic-only seizure | Subclinical, without clinical manifestation. | EEG / aEEG mandatory. | Often seen in preterm infants, HIE (particularly in those with basal ganglia/thalamus injury), critically ill and neonates undergoing cardiac surgery. |
| Unclassified seizure type | Due to inadequate information or unusual clinical features with inability to place in other categories. | EEG / aEEG mandatory. | |

Abbreviations: aEEG, amplitude-integrated EEG; early infantile DEE, early infantile developmental and epileptic encephalopathy; EEG, electroencephalography; EMG, electromyography; HIE, hypoxic-isch
ILAE International League Against Epilepsy; msec, millisecond.

*Epilepsia*. 2021;62:615–628.

**TABLE 2** Descriptors of motor seizures in the neonatal period

| Seizure type | Descriptors |
|---|---|
| Automatisms | Unilateral<br>Bilateral asymmetric Bilateral symmetric |
| Clonic seizures | Focal<br>Multifocal<br>Bilateral |
| Epileptic spasms | Unilateral<br>Bilateral asymmetric Bilateral symmetric |
| Myoclonic seizures | Focal<br>Multifocal<br>Bilateral asymmetric<br>Bilateral symmetric |
| Tonic seizures | Focal<br>Bilateral asymmetric<br>Bilateral symmetric |

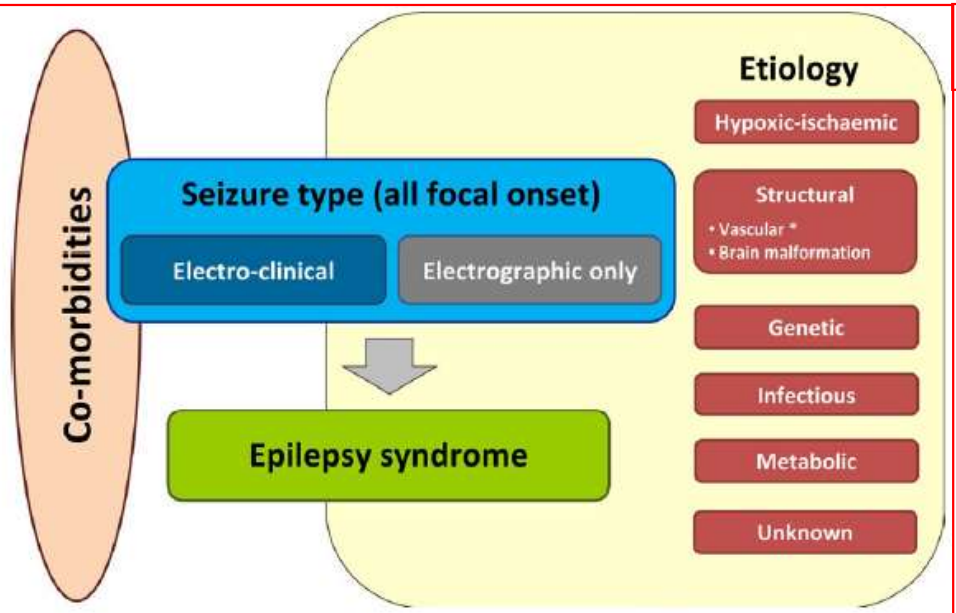
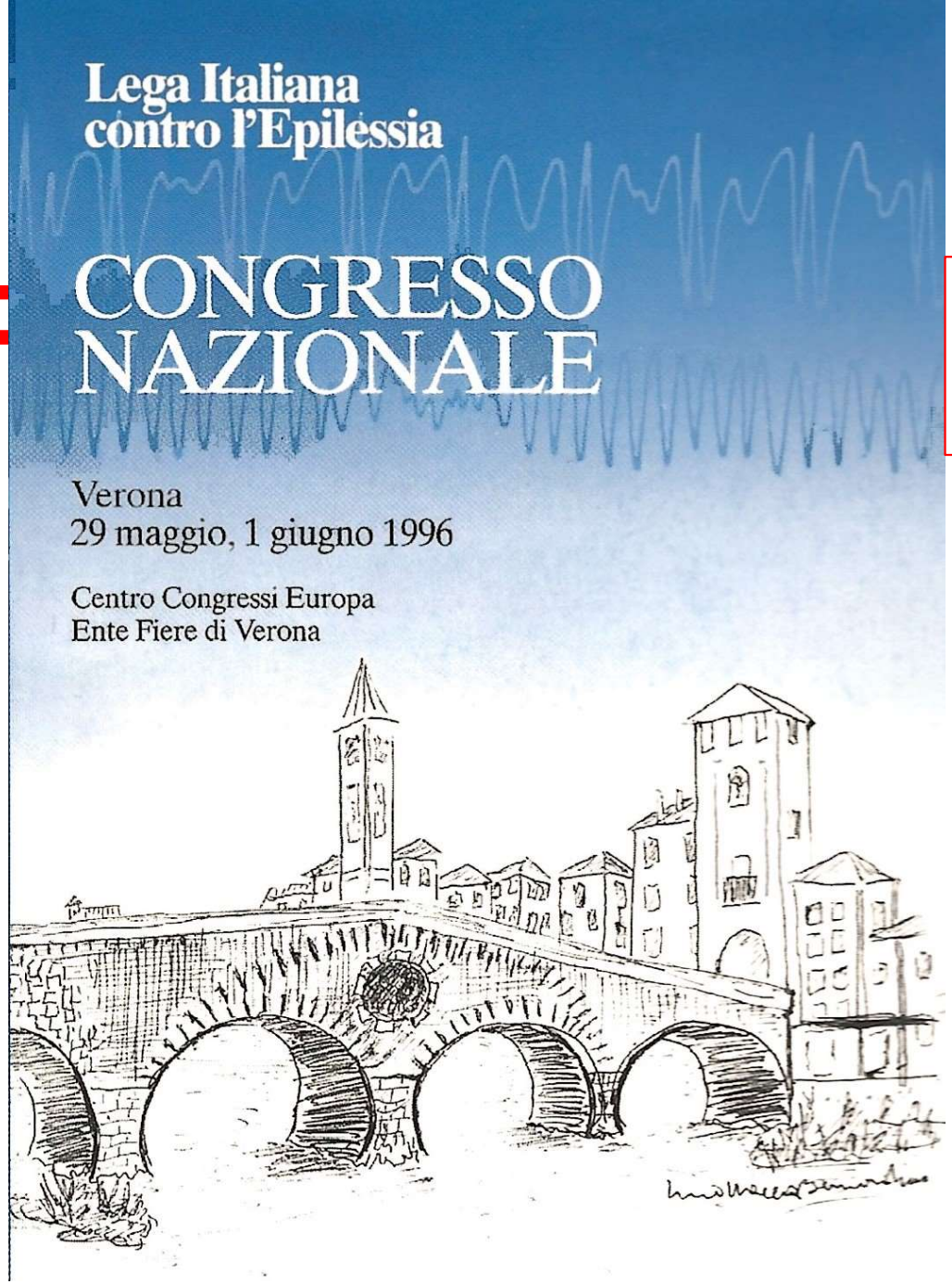



**FIGURE 3** Framework for neonatal seizures and epilepsy

2021

| | Clinical context of seizure type | Current data review |
|---|---|---|
| Automatisms | Described as common seizure type in HIE and preterm infants. [13,52,53,74] | Very rare in isolation (<1%). |
| Clonic | Described as acute symptom of neonatal stroke or cerebral hemorrhage. [13,53,54,75-77] | Typical for vascular etiologies in term infants, also commonly seen in HIE. |
| Myoclonic | Typical seizure type in early myoclonic encephalopathy, but may be seen in other etiologies too, particularly in genetic epilepsy syndromes and preterm infants. Also seen in HIE and inborn errors of metabolism. [13,42,55,56,68,69,72,78] | Typical in inborn error of metabolism. |
| Sequential seizure | Often seen in channelopathies such as BFNE or KCNQ2 encephalopathy, but may be seen with other etiologies. [63,64,69,79] | Typical in genetic etiologies. |
| Spasms | Described in association with inborn error of metabolism or Ohtahara syndrome, here usually in form of tonic spasms (STXBP1, hemimegalencephaly). [42,56,58,60,61,78,80,] | Rare, mostly in inborn error of metabolism. |
| Tonic | Typical seizure type in early infantile epileptic encephalopathy (Ohtahara syndrome), but also seen in other epileptic encephalopathies and genetic neonatal epilepsies (eg KCNQ2 and KCNQ3 mutations). May be seen in HIE. [13,56,57,63,65,69,72,78,81] | Typical in genetic etiologies. |
| Autonomic | Described in intraventricular hemorrhage as well as temporal or occipital lobe lesions, typically as apnea and cyanosis. Also described in Ohtahara syndrome. [13,64,66,81-84] | Overall rare in isolation (<5%). |
| Behavioral arrest | Arrest (pause) of activities, freezing, immobilization. | Very rare in isolation (<1%). |
| Electrographic only seizures | Typical in preterm infants, HIE (particularly in those with basal ganglia/thalamus injury) and neonates undergoing cardiac surgery. [6,11-13,17,85,86] | Most common seizure type in preterm infants, HIE and infectious causes. |

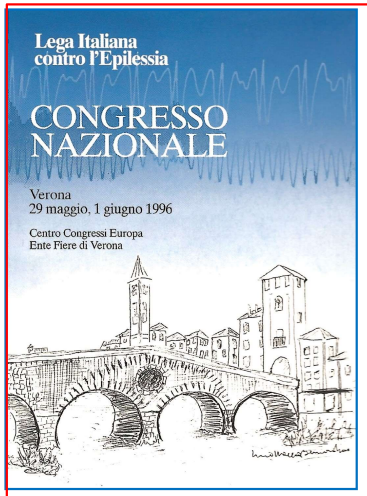
**Table 5: Clinical relevance of seizure types in the neonatal period: review of literature and own data.**




1996

**Crisi miocloniche in epoca neonatale**

M.Mastrangelo, P.Faldini, P.Sergi, C.Vegni, GC.Pastorino




**Myoclonic seizures in the neonatal period.**

SUMMARY

The electroclinical findings in 17/68 newborns with myoclonic seizures are reported. Correlation between myoclonic seizures and ictal EEG activity is variable: it is always evident in focal myoclonic seizures; myoclonic jerks are generally related to the EEG bursts but with a slightly variable latency; segmental asynchronous erratic myoclonias are never related to the paroxysmal EEG activity. The neurological status is always very poor**, i. e. acute encephalopathy**, and always partial electrical or electroclinical sezures are associated to myoclonic seIzures. ***The clinical course has been severe in 15/17, with epilepsy and psychomotor impairment. Among newborns with neonatal seizures, the myoclonic-seizure pattern seems to be highly predictive of unfavourable outcome.***

**2005**

Seizure (2005) 14, 304—311

# Epileptic seizures, epilepsy and epileptic syndromes in newborns: A nosological approach to 94 new cases by the 2001 proposed diagnostic scheme for people with epileptic seizures and with epilepsy

Massimo Mastrangelo [a,*], Andrea Van Lierde [b], Milena Bray [c], Giancarlo Pastorino [a], Antonio Marini [b], Fabio Mosca [c]

Conclusions: Compared to the 1989 ILAE classification, …… the 2001 ILAE diagnostic scheme, applied at the end of the neonatal period, offers a variety of approaches to classification, ***allowing an early distinction between epilepsy and single or isolated clusters of seizures, with therapeutic and prognostic implications.***

**Epilepsia, 51(4):676–685, 2010**
Report of the ILAE Commission on Classification and Terminology

**ILAE DEFINITION OF EPILEPTIC SEIZURE (Fischer et al., 2005):**

"a transient occurrence of signs and or symptoms due to abnormal excessive or synchronous neuronal activity in the brain".

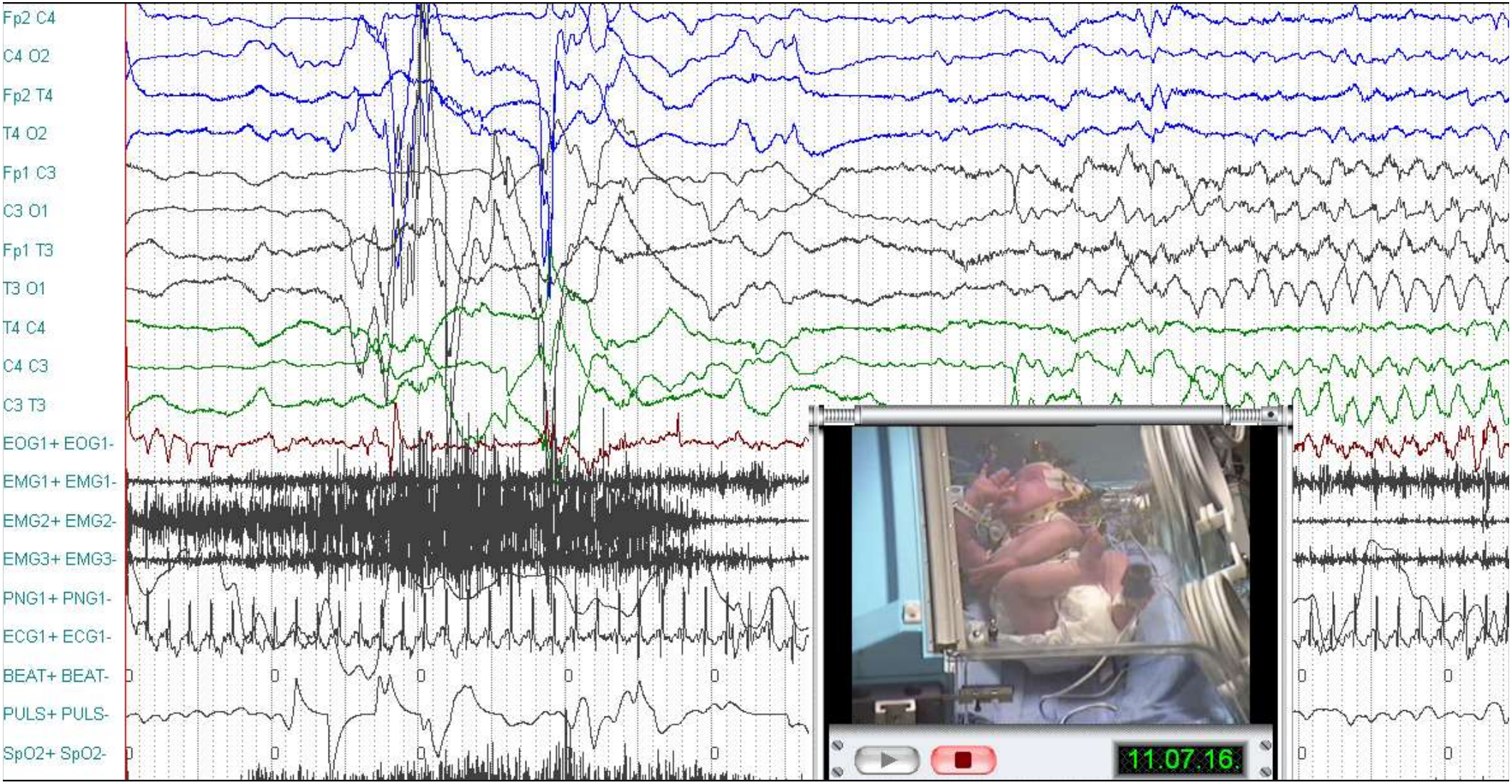
# Nel neonato

***Electroclinical seizures***

***Electrical-only seizures***

**Neonato a termine**

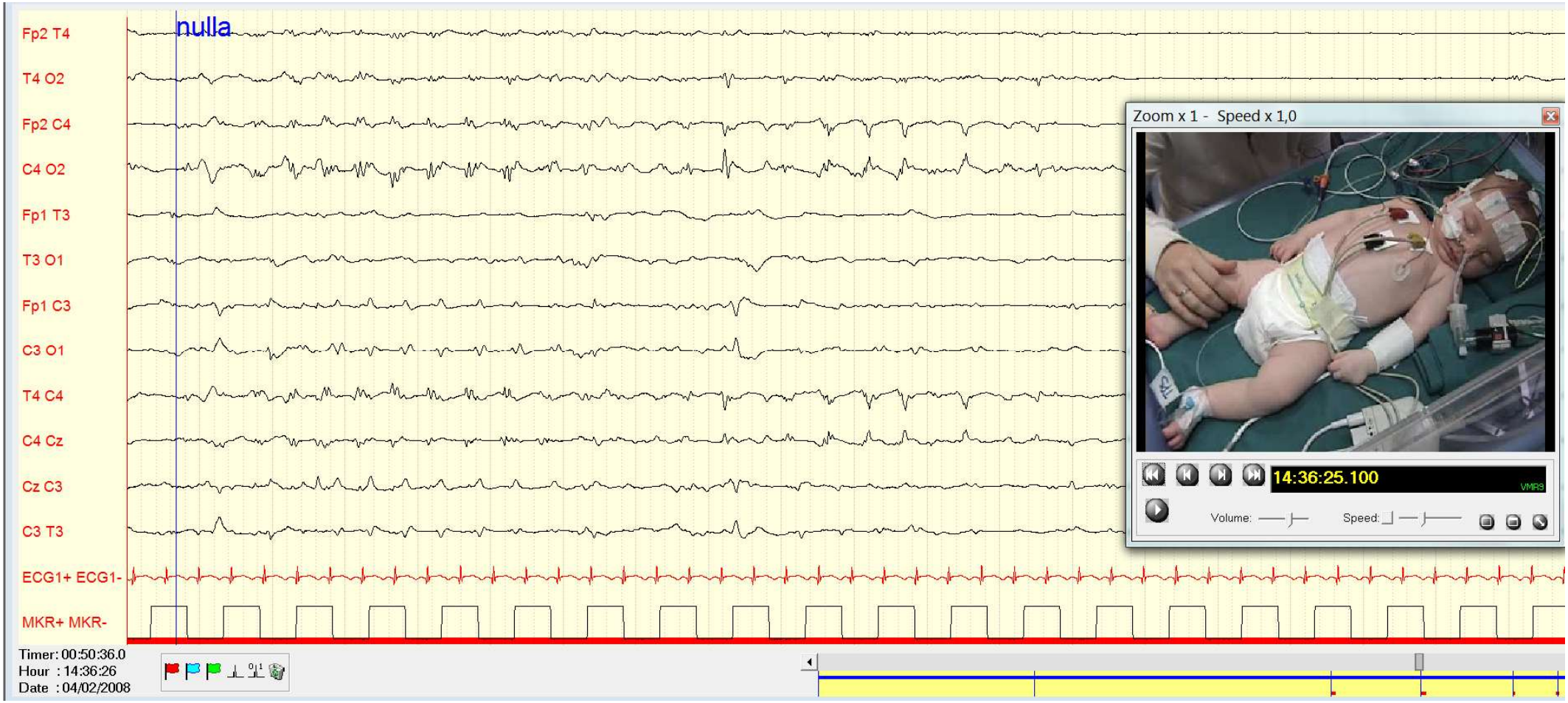
**encefalopatia epilettica + stato di male, a 5 giorni**




**Electroclinical seizure**

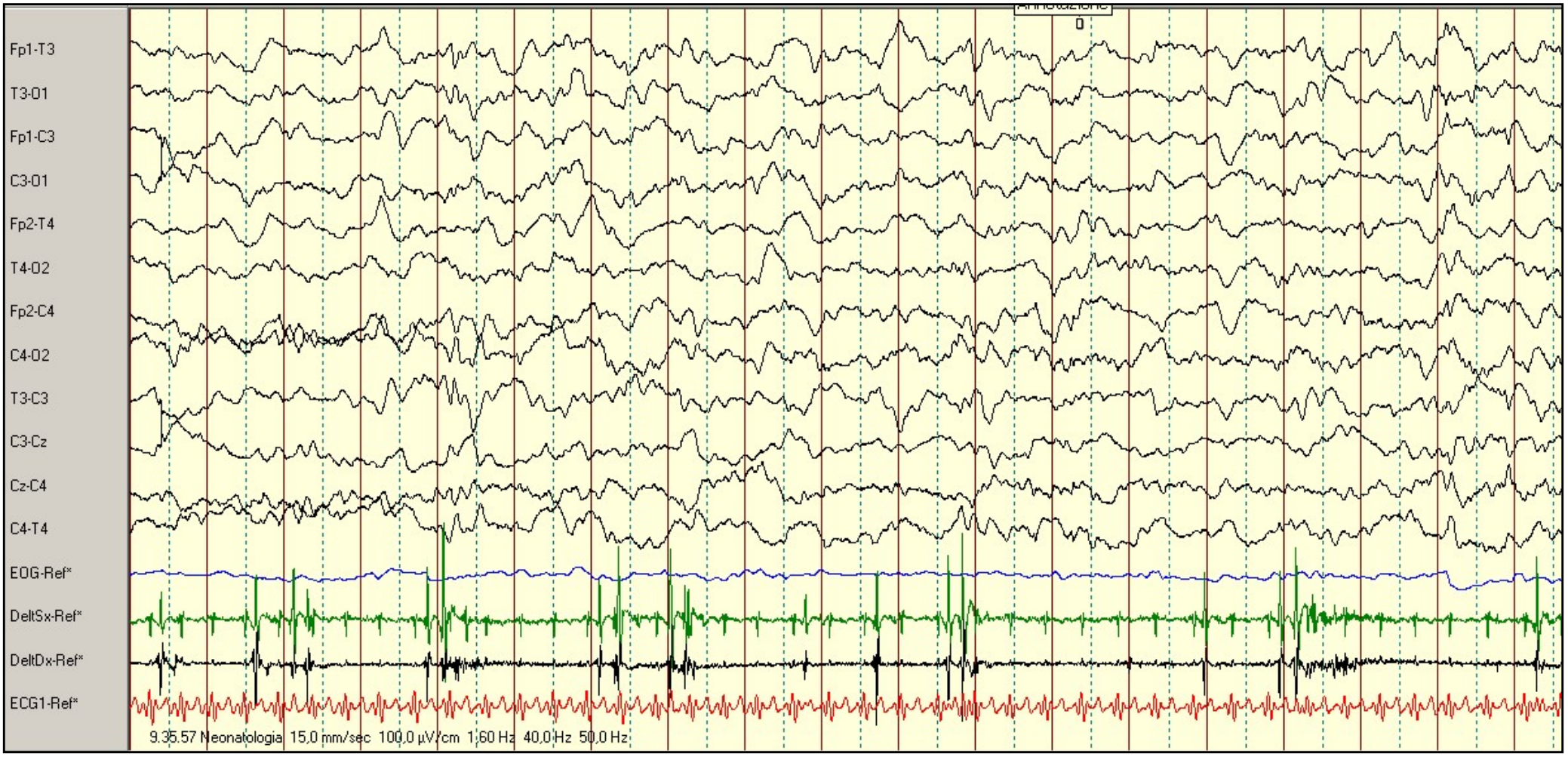
# Neonato a termine

## encefalite da streptococco beta + stato di male, a 25 giorni



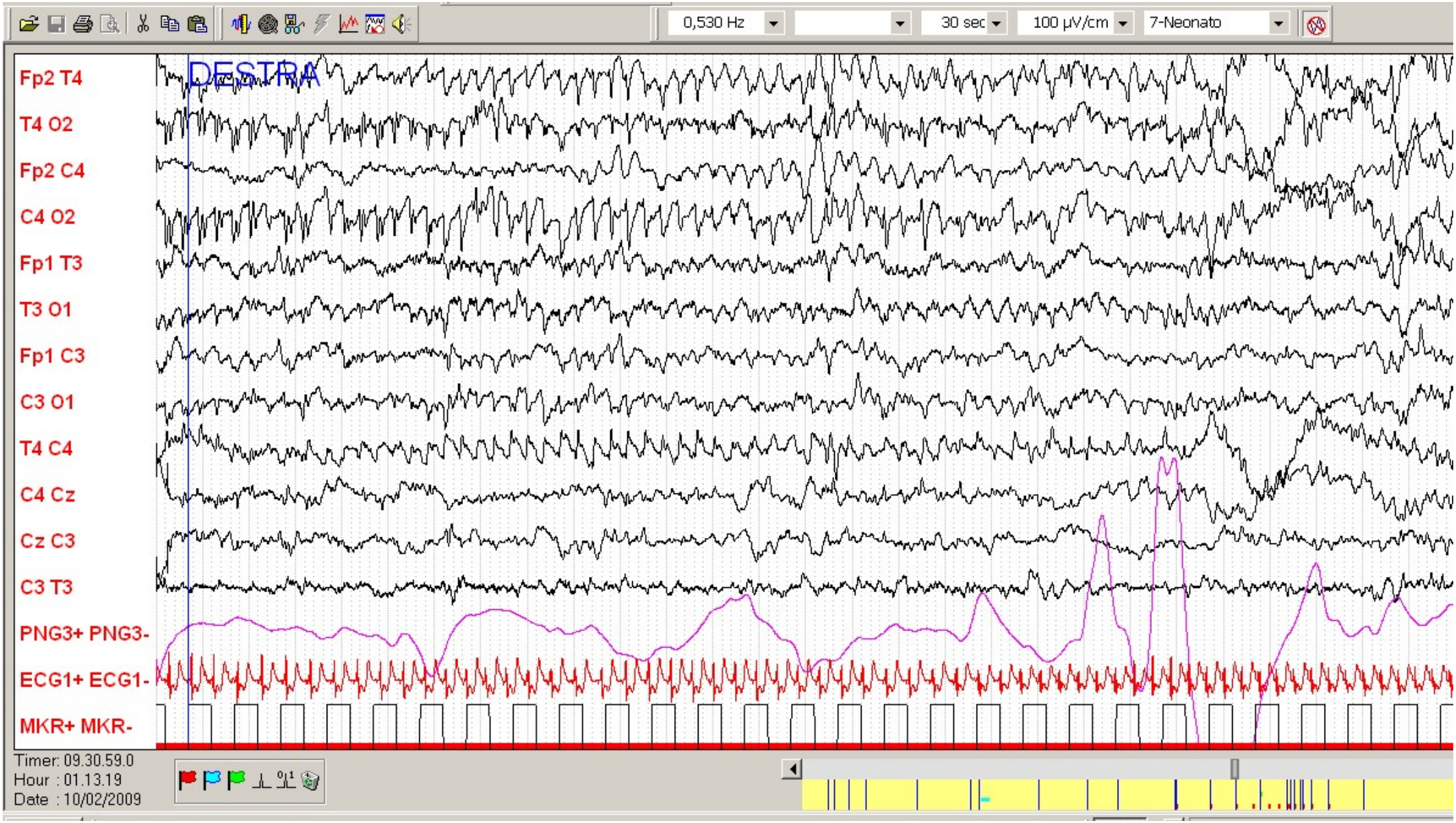

**Electrographic only seizure**

**41.3 sett EC > HIE + CRISI NEONATALI CLONICHE E MIOCLONICHE > PB 20 mg/Kg EV > peggioramento > 4 > 5 > 6 mg/Kg/OS > peggioramento > > a 13 giorni MIOCLONIE BILATERALI ISOLATE E IN SALVE**




**Manifestazioni parossistiche non epilettiche (mioclono ipnico benigno neonatale)**

# 39 sett EG 41 sett EC crisi durante la notte / sonno




**Unclassified seizure**

# Most epileptic seizure consists of symptoms that evolve as the seizure discharge spread to involve new cortical areas

**Hans O. Lüders; Claudio Munari**

The ILAE Classification of Seizures & the Epilepsies:
Modification for Seizures in the Neonate.
Proposal from the ILAE Task Force on Neonatal Seizures

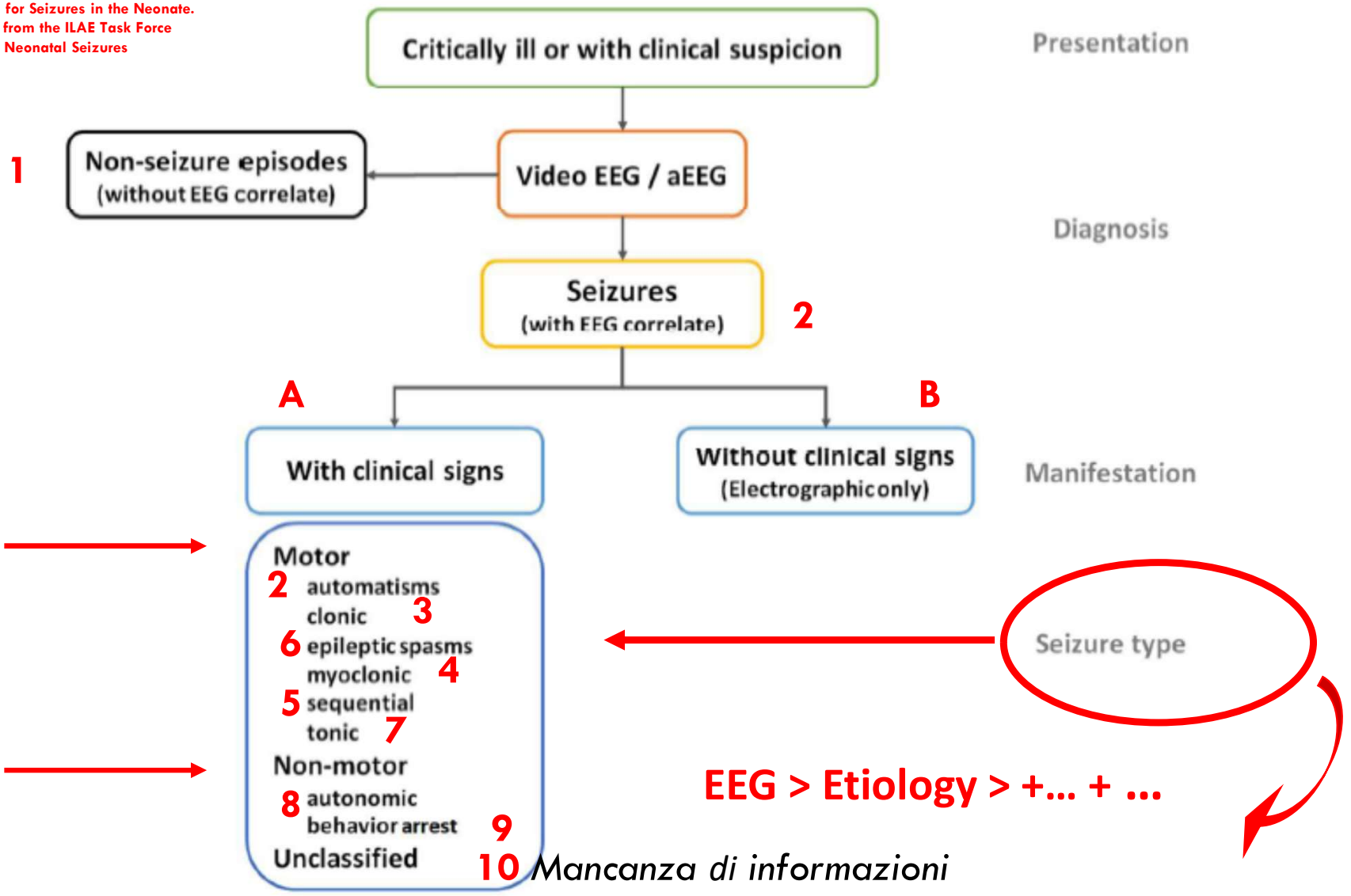



Figure 2: Proposed diagnostic framework of seizures in the neonatal period including classification of seizures. Adapted from 2017 ILAE seizure classification.3 Neonates present with discrete events suspected to be epileptic seizures or are critically ill (often ventilated, sedated and treated with muscle relaxants in intensive care).

*Video ..... **Mod. MM***

Seminars in Fetal & Neonatal Medicine 20 (2015) 149–153

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

Review

Continuous long-term electroencephalography: The gold standard for neonatal seizure diagnosis

Renée A. Shellhaas*

Practice points:

- *Continuous video-EEG monitoring is the gold standard for the diagnosis of neonatal seizures…….*
- Newborn infants with suspected or confirmed acute brain injury, especially those with concomitant encephalopathy, are at high risk for seizures and should be considered for EEG monitoring.

Brain & Development 36 (2014) 363–371

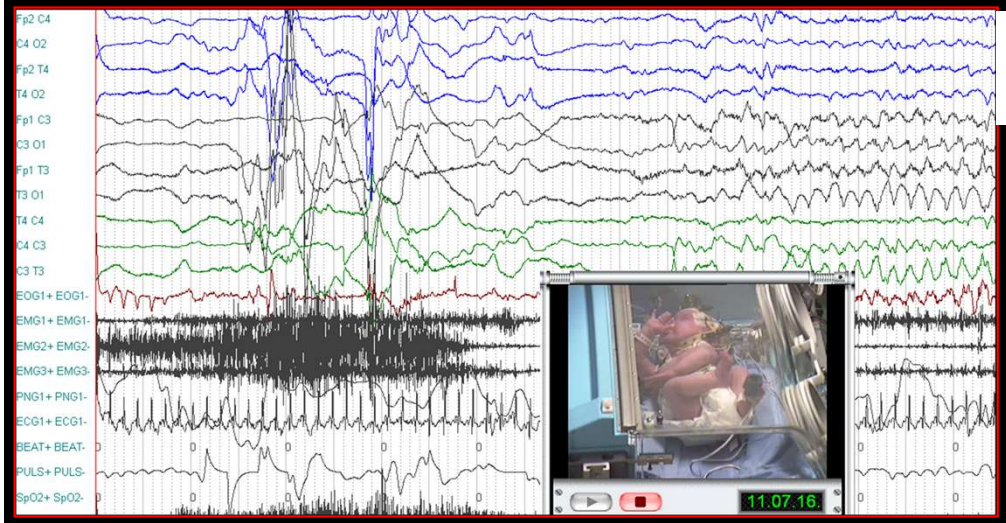
Review article

Neurophysiological aspects of neonatal seizures

Kazuyoshi Watanabe*

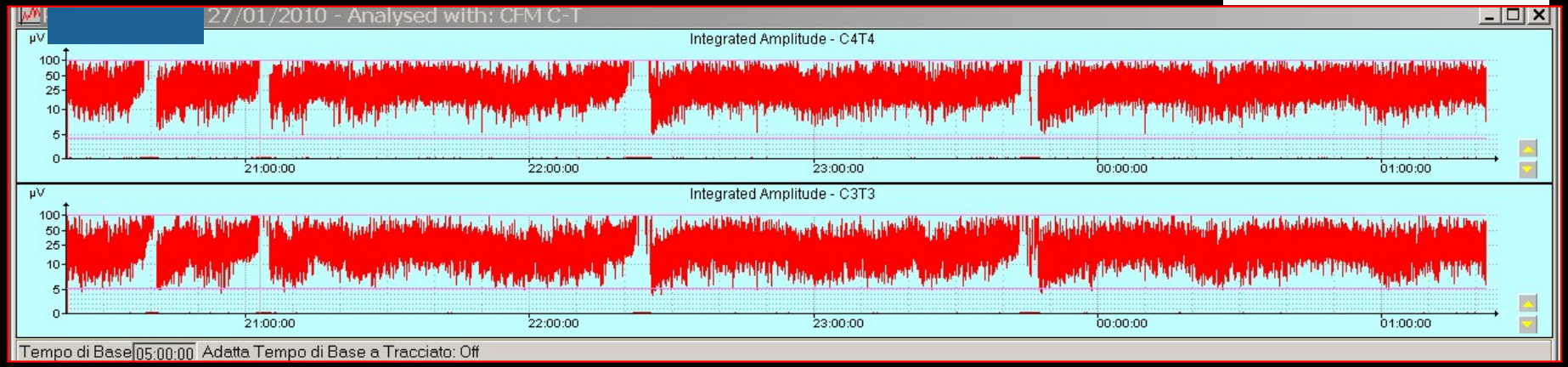
"Recently, *amplitude-integrated EEG (aEEG) has been increasingly used and proved useful in neonatal intensive care units (NICU) for the management of neonatal seizures.*

***It does not replace, but is supplementary to standard EEG***"

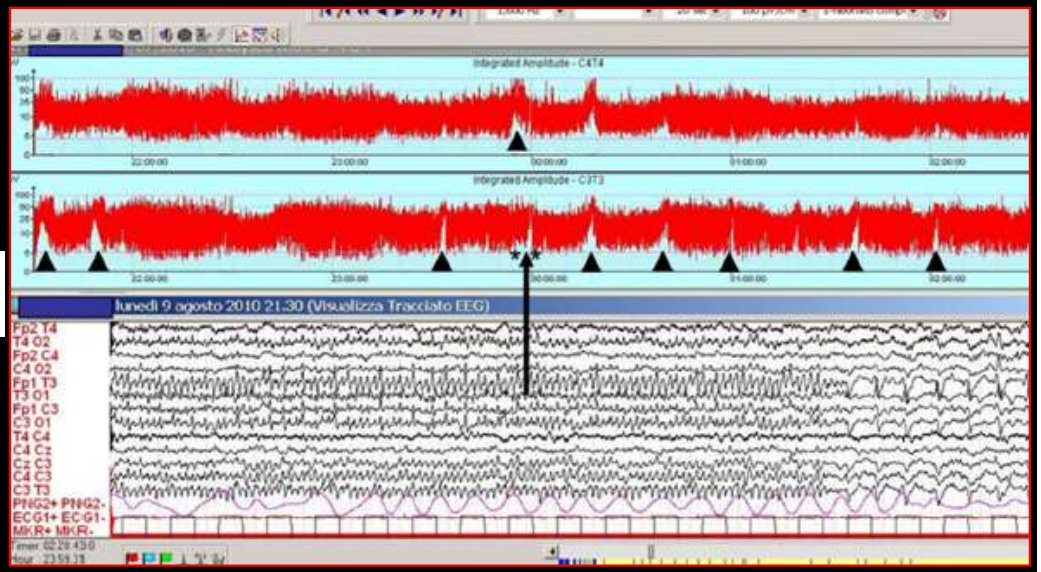



EEG ……

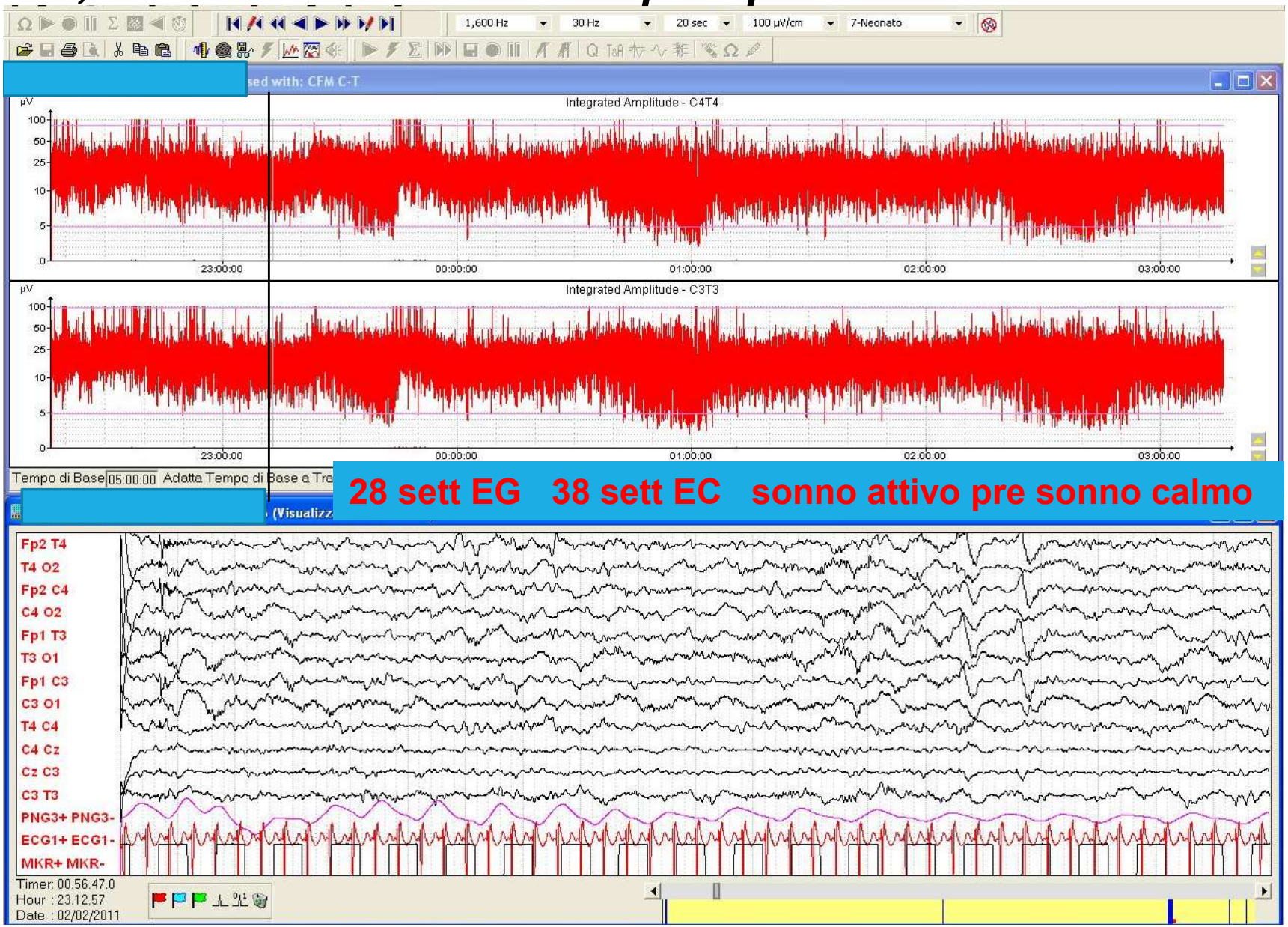
aEEG ……




EEG + aEEG

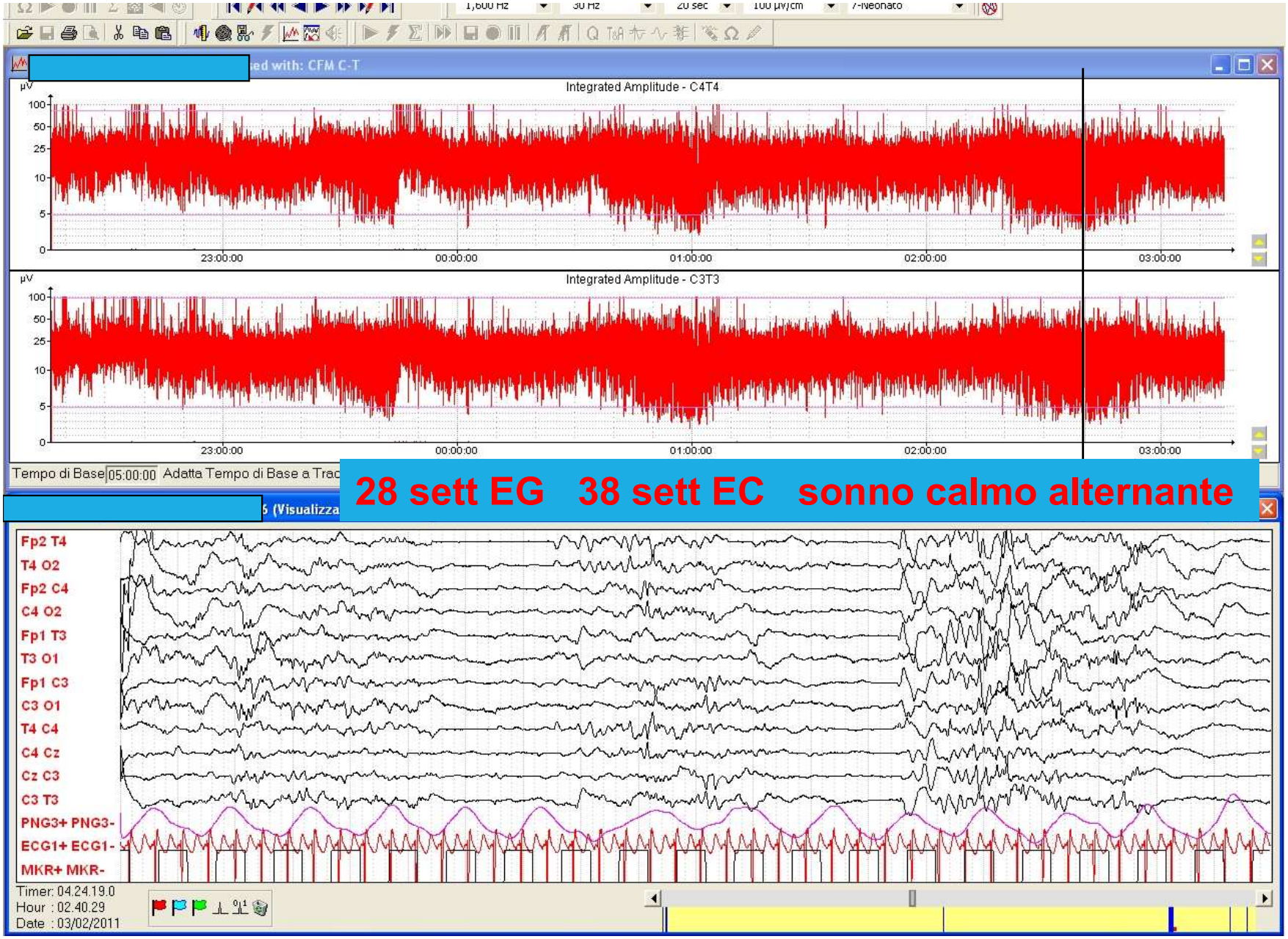


# *FR, a 10 settimane ricoverato per spasmi*




***Concordanza: aEEG e cEEG > attività di fondo normale - sonno REM***

## *Registrazione combinata EEG / aEEG – encefalopatie neonatali – crisi epilettiche*




## *Concordanza: aEEG e cEEG > attività di fondo normale - sonno alternante*

2013

Seizure 22 (2013) 703–707

# Acute neonatal encephalopathy and seizures recurrence: A combined aEEG/EEG study

Massimo Mastrangelo [a], Isabella Fiocchi [a], Paola Fontana [b], Gaetano Gorgone [c], Gianluca Lista [b], Vincenzo Belcastro [d,*]

**Purpose:** To evaluate amplitude-integrated EEG (aEEG) in comparison with conventional (cEEG) for the identification of electrographic seizures in neonates with acute neonatal encephalopathies.

Demographic characteristics of study subjects.

| | |
|---|---|
| Patient | 28 |
| Gestational age (wk) and birth | 39.4 ± 1.6 |
| Sex (M/F) | 18M/10F |
| Birth weight (g) | 3250 ± 705 |
| Primary diagnosis (*n*) | |
| HIE | 9 |
| Epilepsy | 6 |
| Intracranial haemorrhage | 5 |
| Meningitis/encephalitis | 4 |
| Genetic/metabolic disease | 4 |
| Total no. of EEG recordings | 31 |
| Duration of recording (h) | 15.5 ± 3.0 |
| No. of EEG recordings | 31 |
| No. of recordings containing seizures | 25 |
| Seizure types (*n*; %) | |
| Status epilepticus | (7; 22.6) |
| Repetitive seizures | (12; 38.6) |
| Single seizures | (6; 19.4) |
| No seizures (*n*; %) | (6; 19.4) |

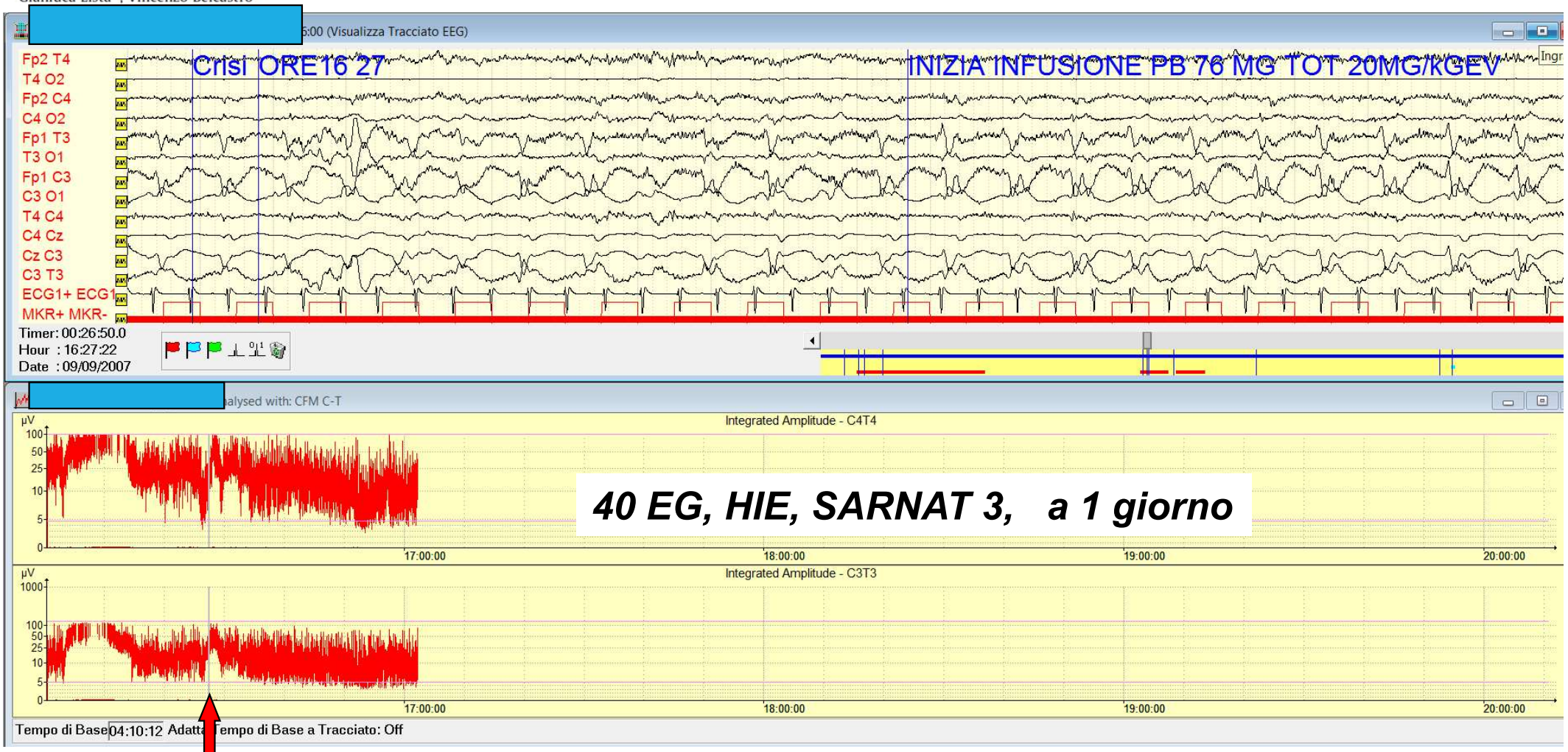
**Conclusions:** In our study the observers identified SE in 100% of the reviewed traces using raw aEEG alone, thus aEEG might represent a useful tool to detect SE in the setting of NICU. SS may not be reliably identified using aEEG alone. Simultaneous recording of the raw cEEG/aEEG provides a good level of sensitivity for the detection of neonatal electrographic seizures.

Seizure 22 (2013) 703–707

# Acute neonatal encephalopathy and seizures recurrence: A combined aEEG/EEG study

Massimo Mastrangelo[a], Isabella Fiocchi[a], Paola Fontana[b], Gaetano Gorgone[c], Gianluca Lista[b], Vincenzo Belcastro[d,*]

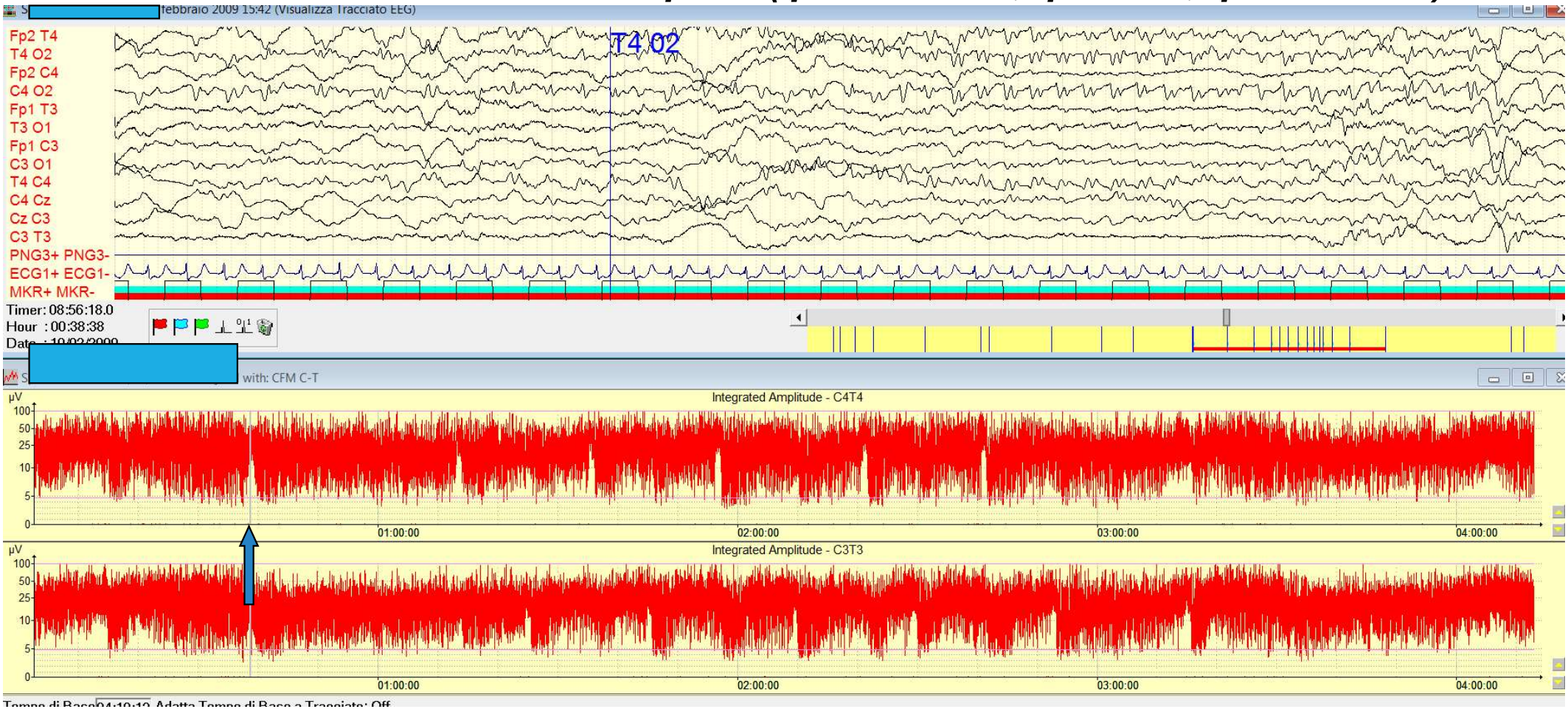





***40 EG, HIE, SARNAT 3, a 1 giorno***

***<u>TRAINING</u> ..... si esamina con il neonatologo la registrazione combinata cEEG / aEEG, in funzione di definire la morfologia del <u>pattern aEEG</u> in relazione al <u>pattern cEEG</u> > critico (crisi isolate, ricorrenti, SdM, attività di fondo, artefatti……)***

## *SM – 40 EG Crisi nella prima settimana > controllo (DZP,PB,PHT )> a 17 giorni non crisi ma condizione di encefalopatia (iporeatttività, ipotonia, ipomotricità)*




## *Crisi a semeiologia clinica minima o nulla, non riconosciute dal personale del TIN.*

***Concordanza: aEEG > CRISI - cEEG > CRISI***

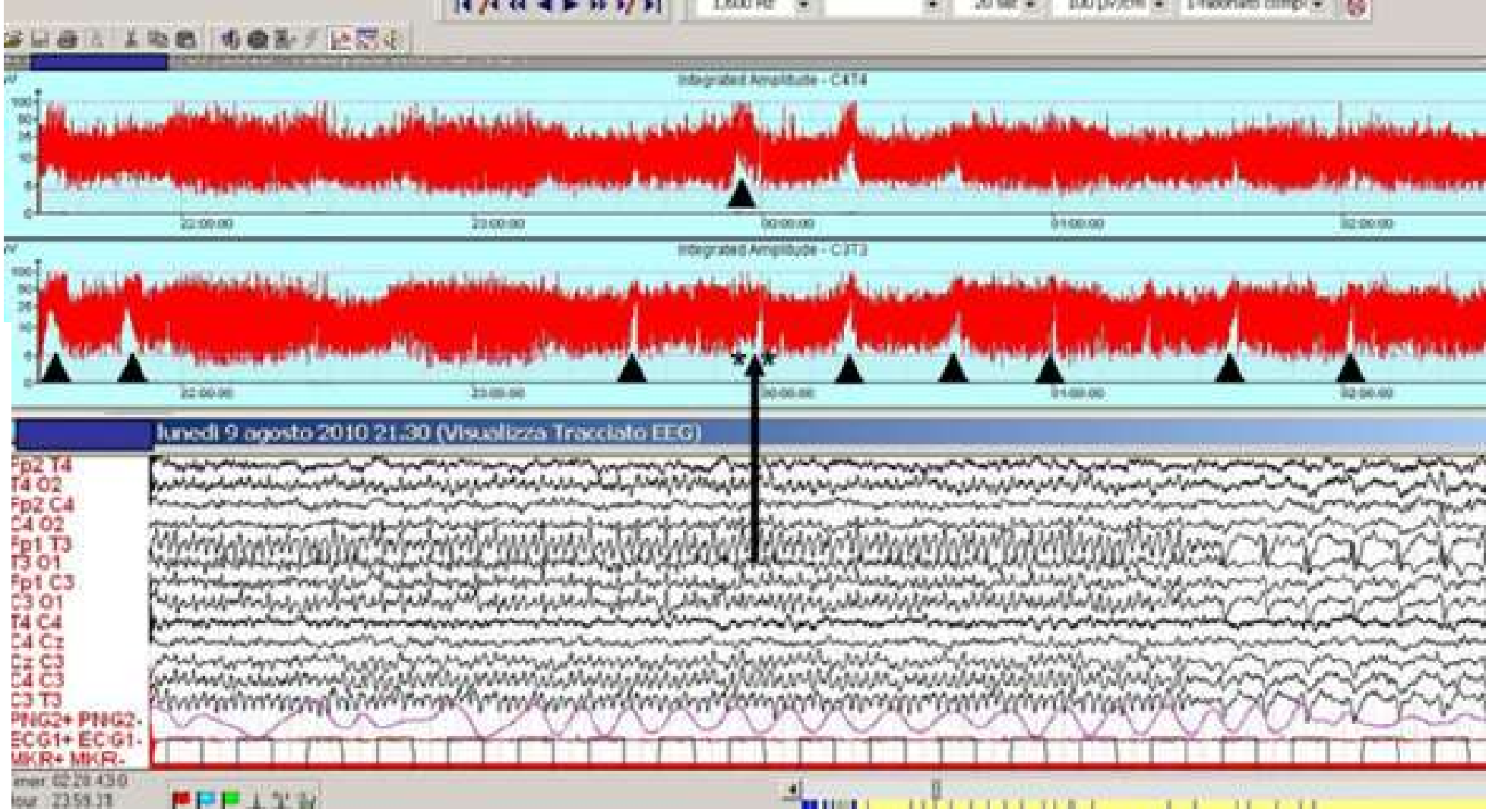
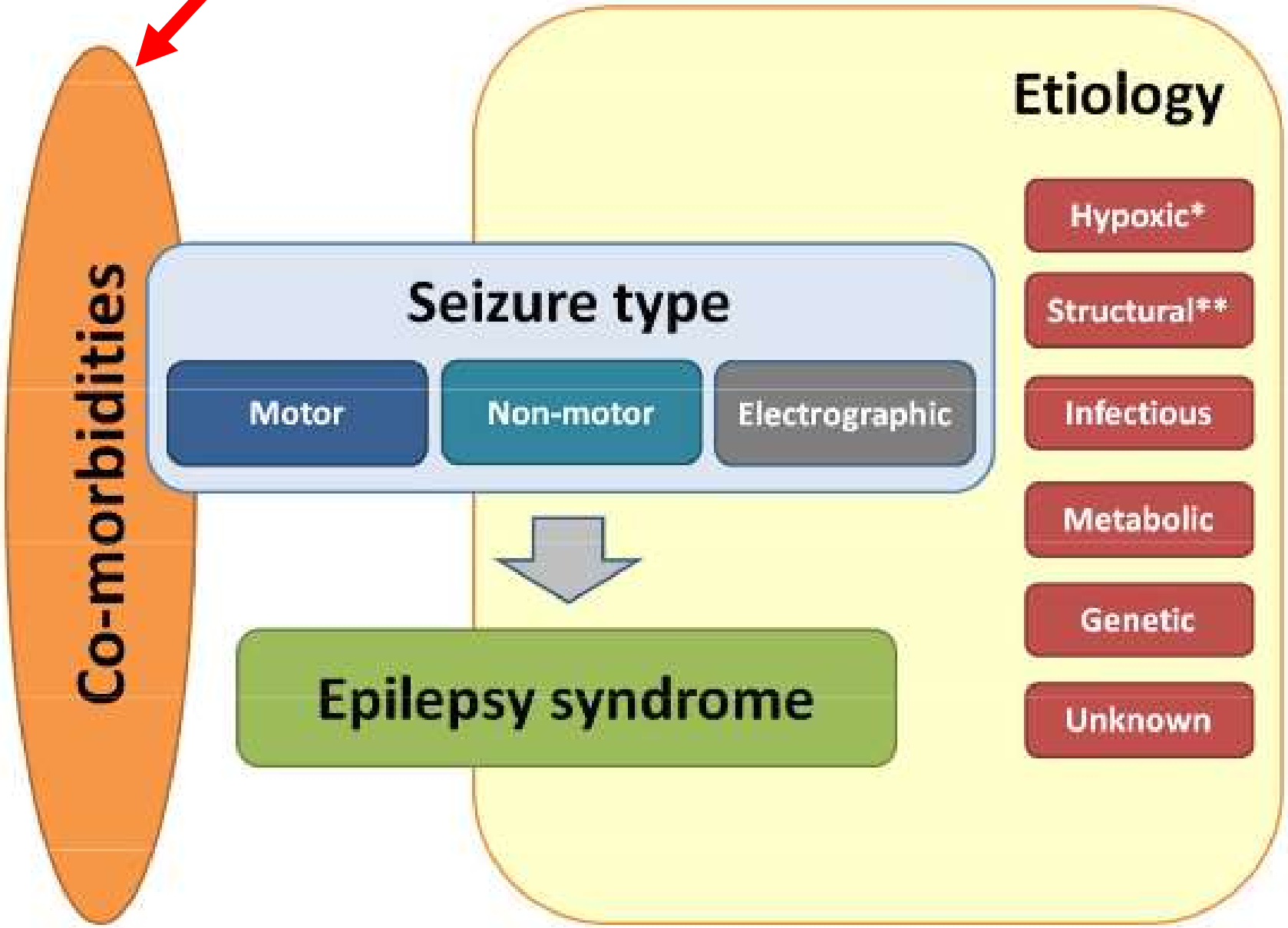


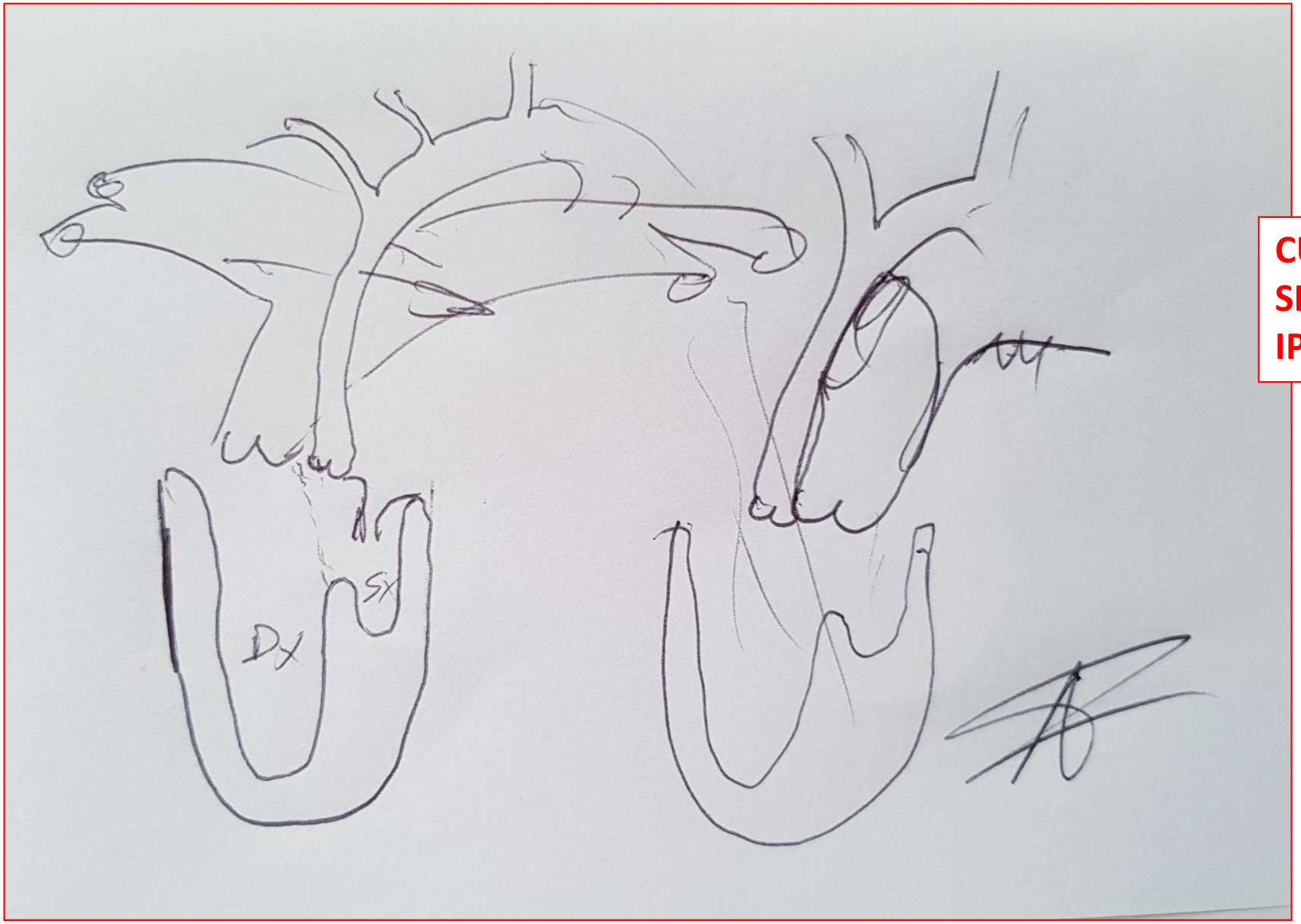

Fig. 2. “Repetitive seizures” (RS) aEEG pattern (more than 1 discharge during a 30 min epoch but not more than one event each 10 min): from 22.30 to 02.10, ten seizures (~) recur, lasting 2–5 min each.

At 22.59 one of these is shown (*"*), lasting less than 2 min, strictly associated to a clear left ictal discharge on cEEG. The aEEG background pattern can be classified as ‘‘continuous normal voltage with sleep–wake cycling’’, with some short periods of discontinuity.

# Co-morbidità

2021





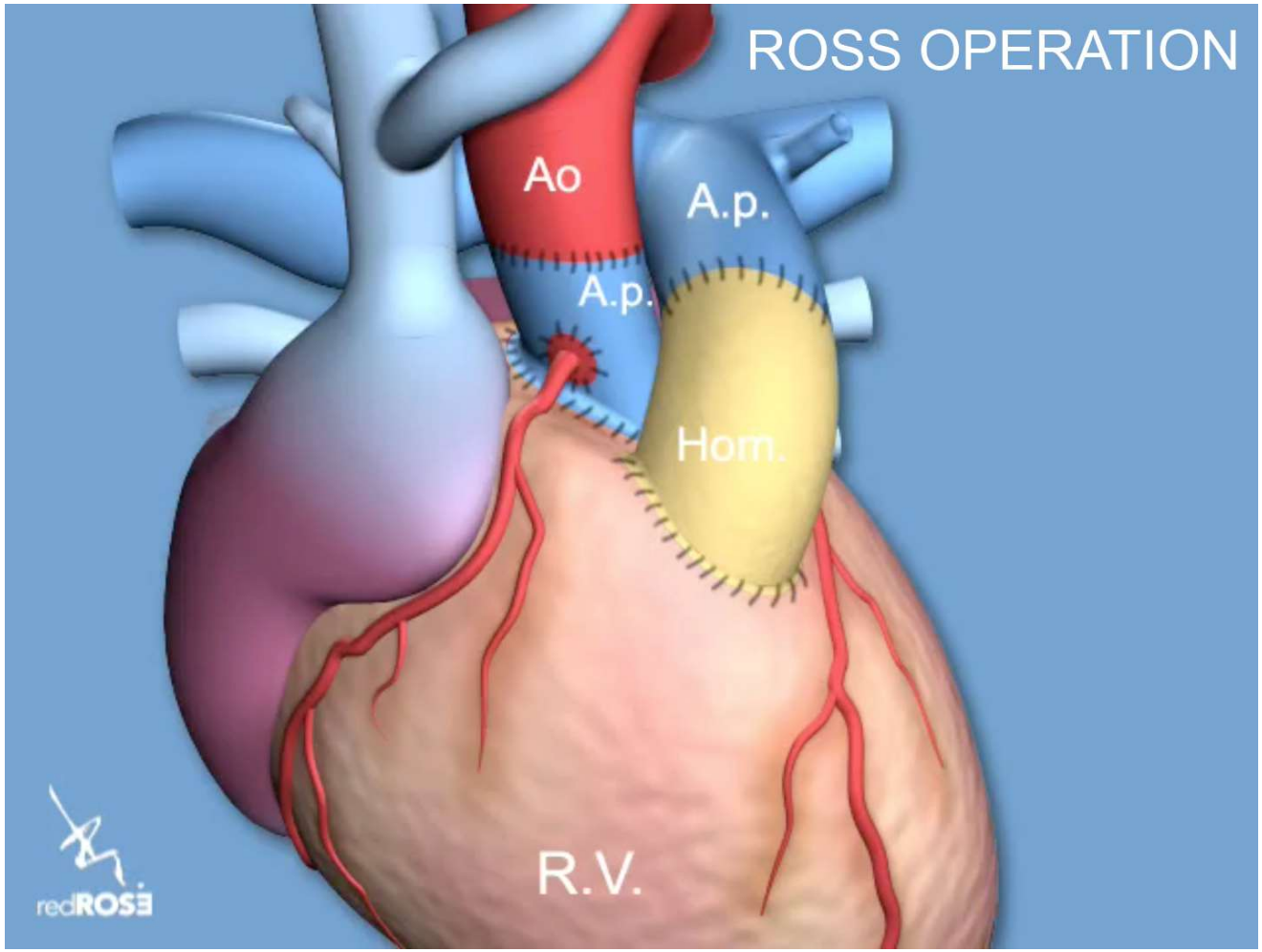

**CUORE SINISTRO IPOPLASICO**

## *Monitoraggio cEEG / a EEG e cardiopatie congenite chirurgiche*

***CUORE SINISTRO IPOPLASICO***. Questa cardiopatia, nonostante i progressi clinici, risulta ancora gravata da mortalità elevata; la sopravvivenza a 1 anno è del 63.5%, a 5 anni del 58.6% e a 10 anni del 54.6%. Solo il 32.6% raggiunge i 15 anni di vita (Best e coll. Frontiers, Pediatric 2021).

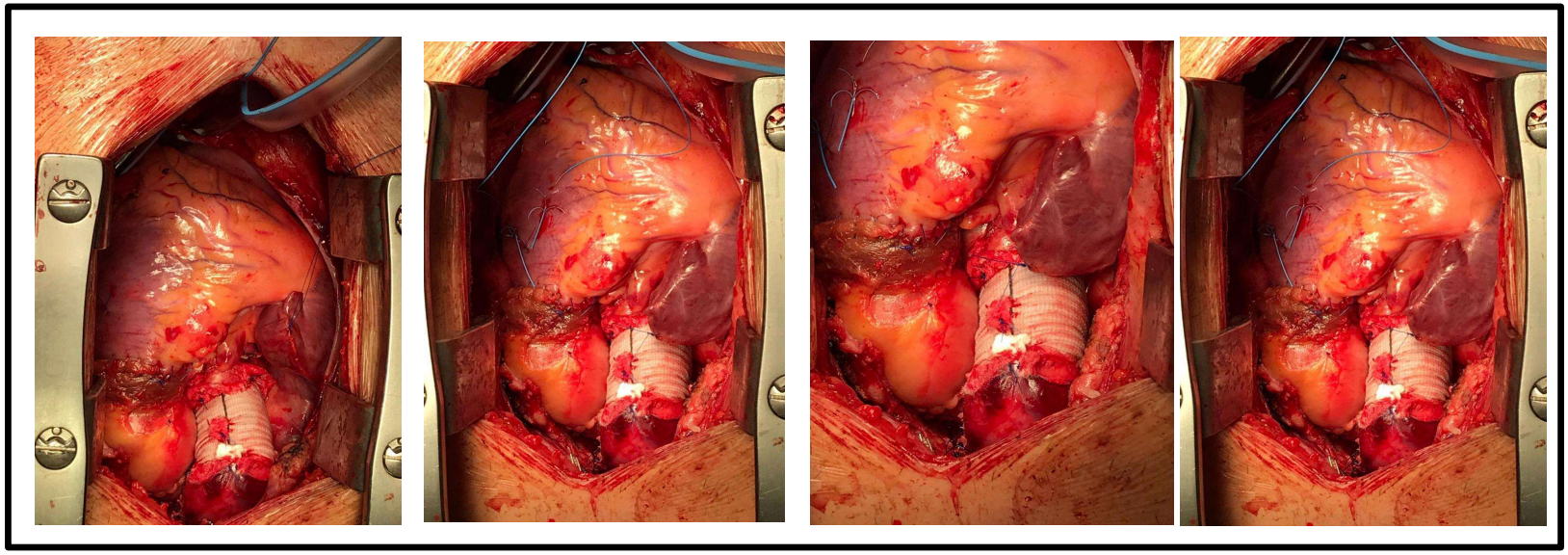
Anche dal punto di vista neurologico, la sindrome del cuore sinistro ipoplasico, come altre cardiopatie congenite complesse, presenta un'elevata incidenza di alterazioni dello sviluppo della sostanza bianca, già riconosciute in epoca fetale. In particolare, in questa patologia, la risonanza magnetica fetale mostra alterazioni della sostanza bianca in circa il 40-50% dei feti (*Ultrasound Obstet Gynecol* 2014; J Thorac Cardiovasc Surg 2021).

I soggetti con ventricolo sinistro ipoplasico sono ad elevato rischio di danno cerebrale post-natale, legato all'ipossia ma anche ai plurimi interventi, in particolare durante il secondo stadio di correzione, quando si rende necessaria la circolazione extracorporea. I principali danni riscontrati sono a carico della sostanza bianca, in particolare prima degli interventi chirurgici, mentre l'infarto cerebrale è la lesione più comune post-operatoria, in particolare nel territorio della cerebrale media e anteriore (J Thorac Cardiovasc 2018). I danni cerebrali esitano nel 40% dei pazienti in un interessamento motorio da moderato a severo (Pediatric Cardiology 2019. Tra i fattori di rischio per un outcome neuroevolutivo peggiore vi è un peso alla nascita inferiore a 2500 gr (Circulation 2012).



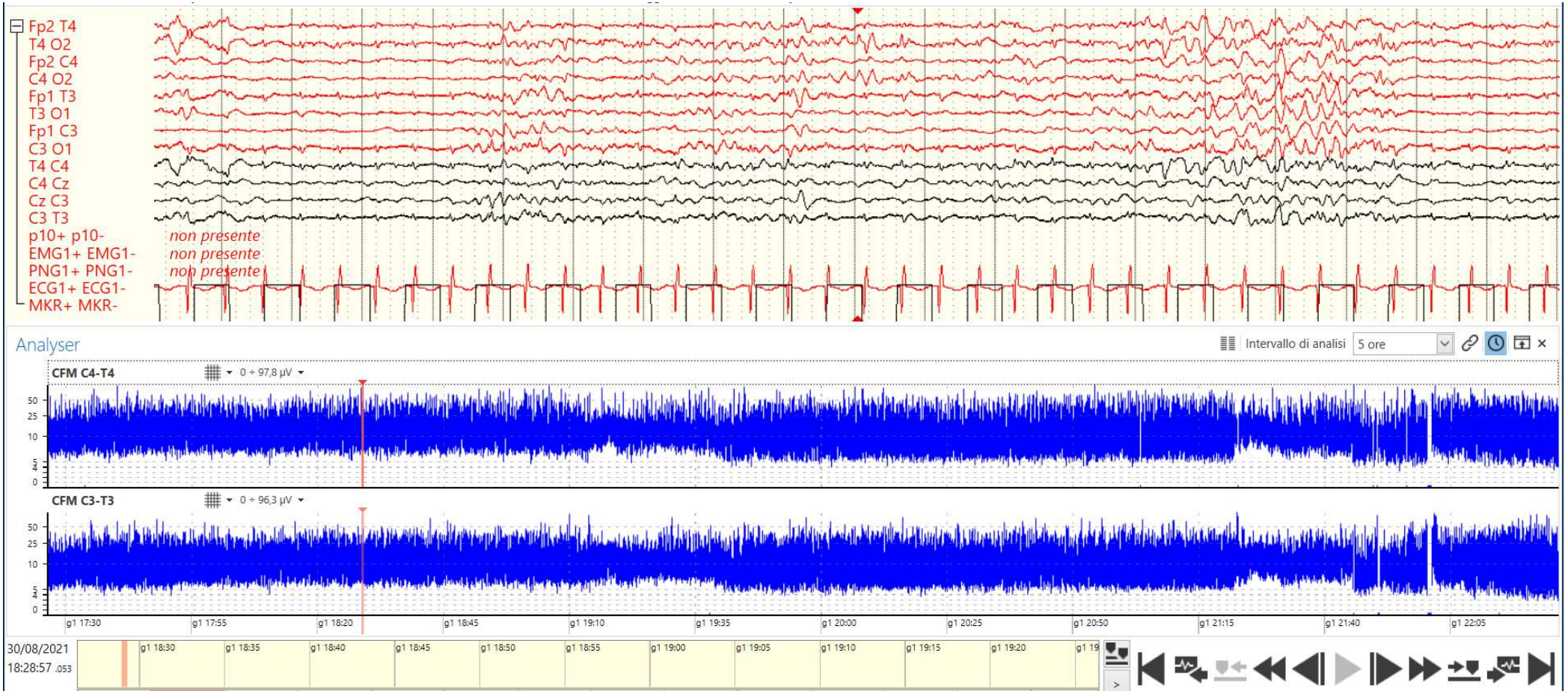

Ti invio questa foto della ROSS dove è ben evidenziato il concetto dell'intervento con la polmonare azzurra che va al posto dell'aorta ascendente e homograft in giallo che va  al posto della polmonare.
In rosso si vedono anche le coronarie reimpiantate.
Cordiali saluti.
Alessandro

# *Monitoraggio cEEG / a EEG e cardiopatie congenite chirurgiche*

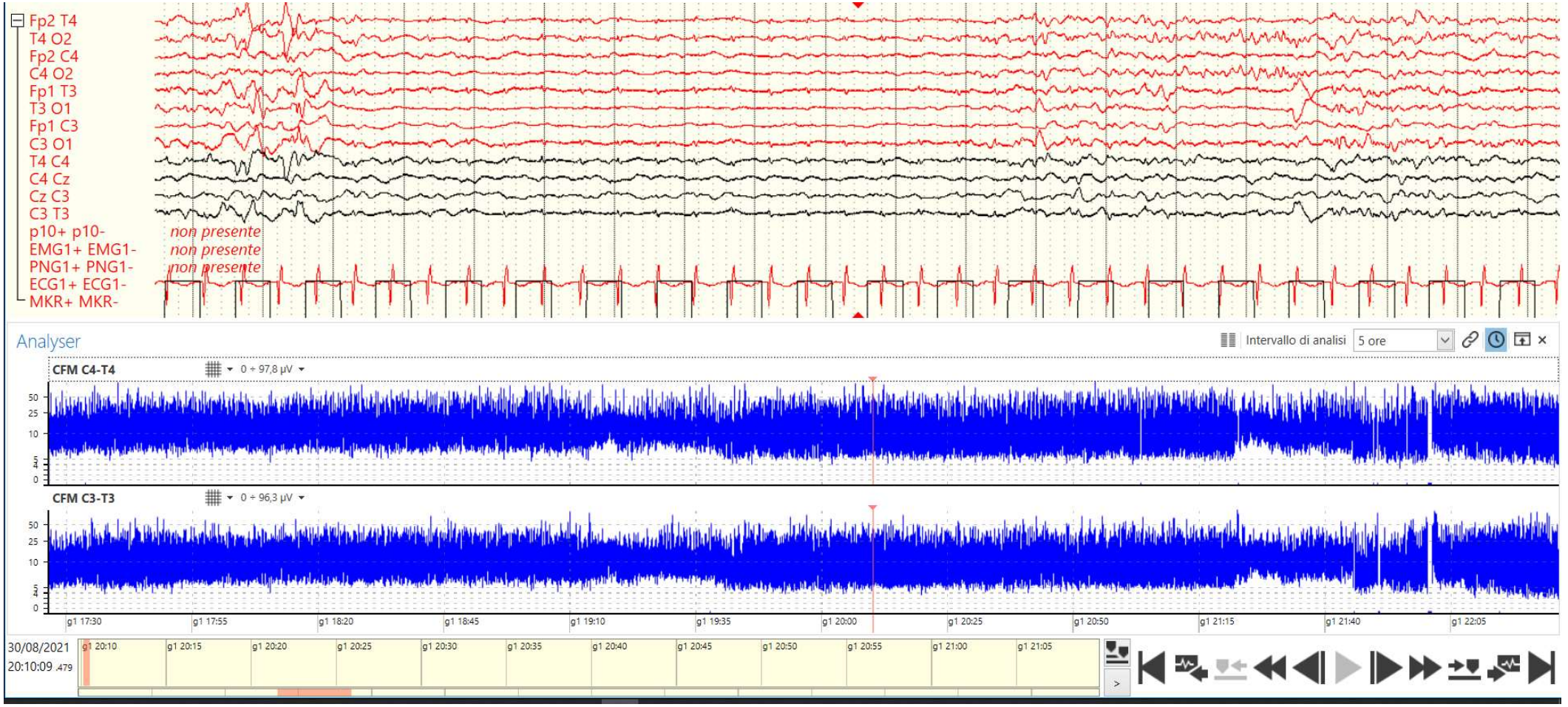


**CUORE SINISTRO IPOPLASICO – INTERVENTO SECONDO ROSS** La Ross operation è un intervento cardio-chirurgico molto complesso che prende il nome dal chirurgo che per primo l'ha eseguita nel 1967, Donald Ross, a Londra, in Inghilterra. Tecnicamente l'intervento consiste nel "traslocare" la valvola polmonare di un cuore in posizione aortica. La complessità dell'intervento consiste nella necessità di un vero e proprio "smontaggio" della valvola polmonare con annesso una porzione di ventricolo destro, dal resto di quest'ultimo e di andarla a ricollocare nella sede della valvola aortica. Il "gap" di tessuto che si viene a creare (che consiste in una porzione di ventricolo destro, nella valvola polmonare ed un segmento di arteria polmonare) viene colmato con uno eguale ma da donatore umano (generalmente da cadavere), o per mezzo di tessuti artificiali (condotti polmonari) ……, in circolazione extracorporea (ECMO) …….

# *Monitoraggio cEEG / a EEG  e cardiopatie congenite chirurgiche*



# *Monitoraggio cEEG / a EEG  e cardiopatie congenite chirurgiche*



# Video

# MONITORAGGIO INTENSIVO NEL NEONATO

## DIAGNOSI E RICONOSCIMENTO CRISI & SDM NEONATALE


DIAGNOSI CLINICA NO

+

EEG ?

MONITORAGGIO

cEEG / aEEG

100%

# Conclusioni

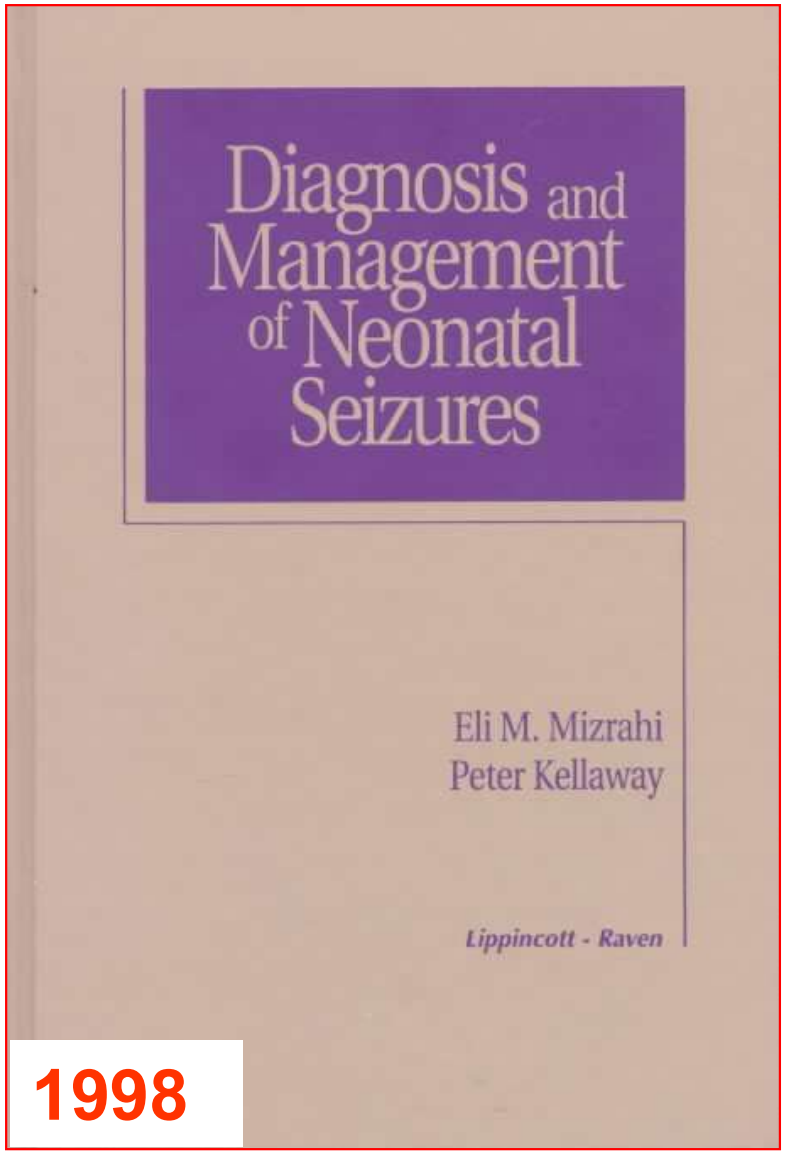
Neurology 1987;37(12):1837- 44

## Characterization and classification of neonatal seizures

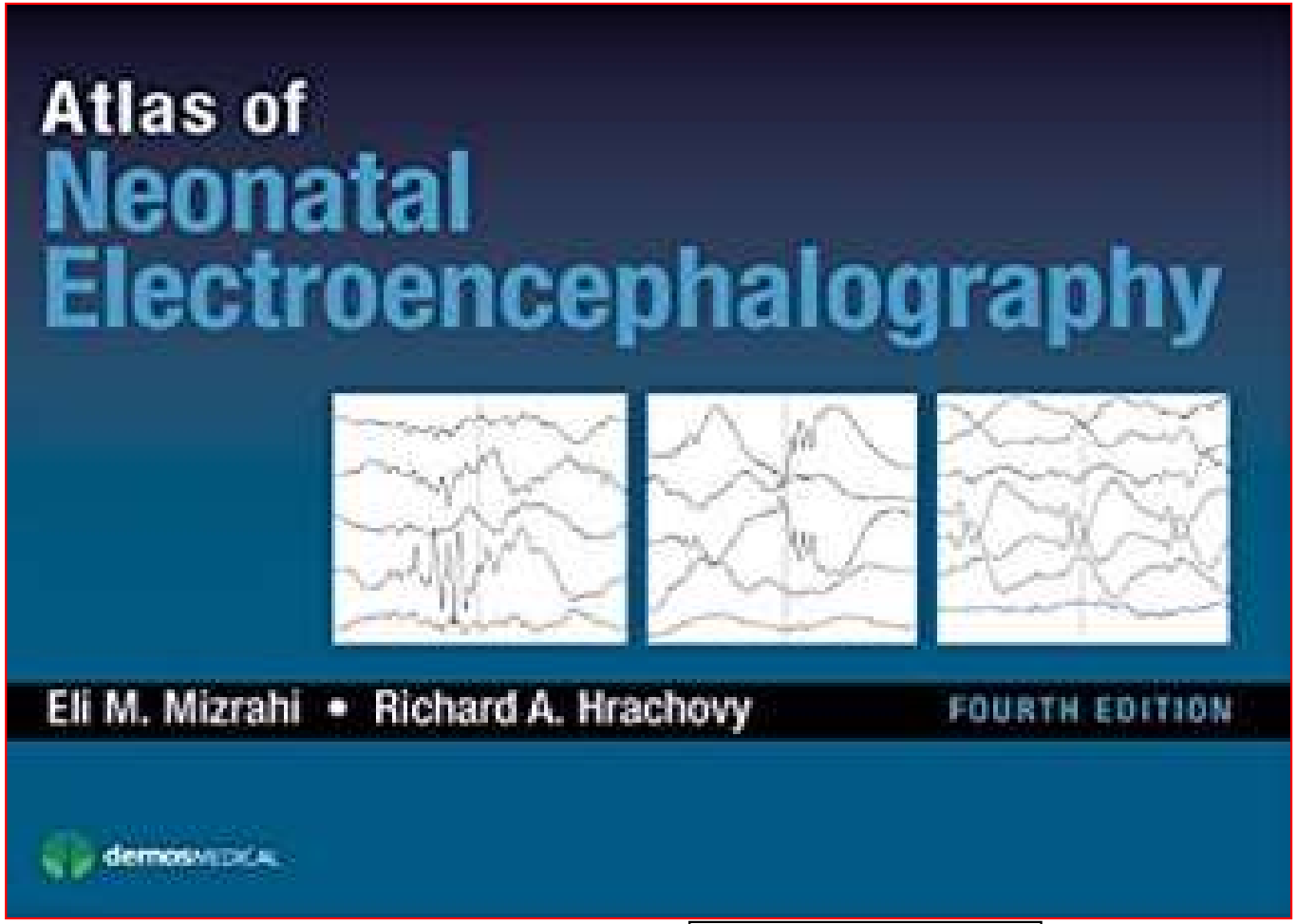
Mizrahi EM, Kellaway P.

To characterize and classify neonatal seizures, we studied 349 neonates, ***using a portable, cribside EEG/polygraphic/video monitoring system. We recorded 415 clinical seizures in 71 infants; 11 other infants had electrical seizure activity without clinical accompaniments. Each seizure was analyzed in terms of its clinical character and its relationship to the presence of EEG seizure activity*****.....**

**EEG features of infants whose seizures were not accompanied by EEG seizure activity suggest that these seizures may not be epileptic in character.**

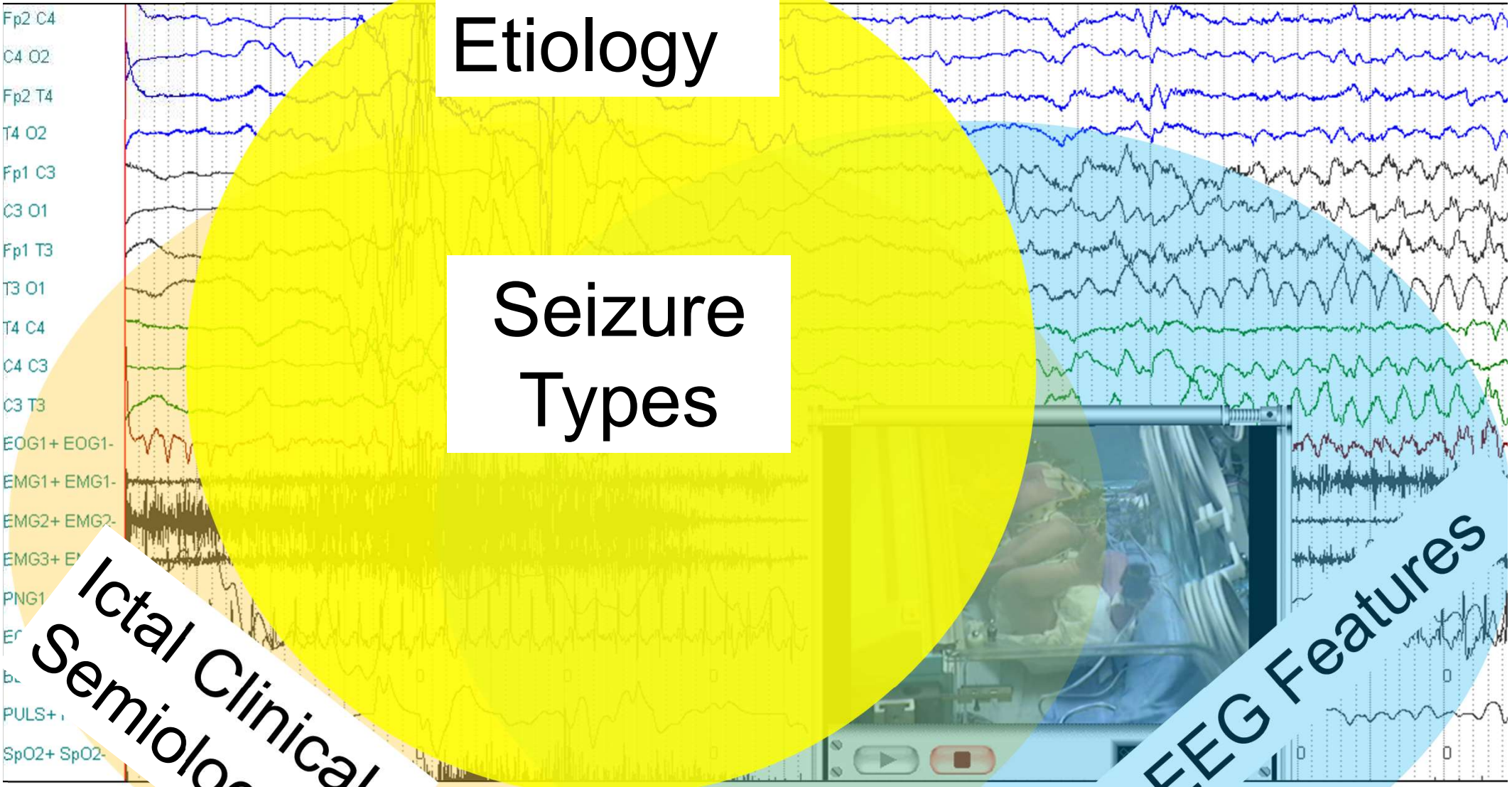


# Conclusioni




**Dic. 2015**

# Conclusioni



*Grazie per l'attenzione .....*

*..... per chi avesse domande o volesse chiarimenti,*

*la mia mail è*
*massimo.mastrangelo2411@gmail.com*

*Milano, 24 novembre 2021*

# La refertazione

*Ne parliamo oggi pomeriggio*

*.....*

## "REFERTAZIONE EEG NEONATALE"

NEL REFERTO DEVE ESSERE SEMPRE SEGNALATO:
- NOME DEL TECNICO DI NEUROFISIOPATOLOGIA CHE HA ESEGUITOO LA REGISTRAZIONE

- NOME COGNOME DEL BAMBINO
- ETA' GESTAZIONALE + ETA' CONCEZIONALE
- DATA ESAME
- NUMERO ESAME
- TEMPO DI REGISTRAZIONE
- MODALITA' DI REGISTRAZIONE > VIDEO, POLIGRAFIA, ecc
- MOTIVO PER IL QUALE VIENE ESEGUITA LA REGISTRAZIONE

**1)DESCRIZIONE**

- CARATTERISTICHE DELL'ATTIVITA' DI FONDO:

SE A) CONTINUA oppure B) DISCONTINUA

A) SE CONTINUA: STATI RICONOSCIBILI SI' / NO
PRESENZA FIGURE FISIOLOGICHE
POLIGRAFIA COMPORTAMENTALE CONSONA CON GLI STATI
DESCRIZIONE DELLE ATTIVITA' ELETTRICHE
SIMMETRIA E SINCRONIA

B) SE DISCONTINUA: DURATA DEI BURSTS > MIN - MAX
DURATA TRATTI INATTIVI > MIN - MAX
SINCRONIA DEI BURSTS
CARATTERISTICHE DEI BURSTS
TRACCIATO FISSO O VARIABILE
REATTIVITA' AGLI STIMOLI

> COMPORTAMENTO DEL BAMBINO NEI TRATTI CON ATTIVITA' ELETTRICA CONTINUA
> " " " " ATTIVITA' ELETTRICA DISCONTINUA

C) CRISI: NO

C1) CRISI: SI
REGISTRATE CON VIDEO O SENZA
DESCRIZIONE PATTERN CLINICO
DESCRIZIONE PATTERN ELETTRICO
DURATA
EVENTUALE SEIZURE BURDEN NEGLI STATI DI MALE

D) MANIFESTAZIONI MOTORIE NON EPILETTICHE: NO

D1) MANIFESTAZIONI PAROSSISTICHE NON EPILETTICHE: SI'
REGISTRATE CON VIDEO O SENZA;
DESCRIZIONE DEGLI EVENTI
PATTERN ELETTRICO CORRISPONDENTE

E) APNEE: NO

E1) APNEE SI:
REGISTRAZIONE CON UNO / DUE RESPIRI > NASO E BOCCA
DURATA APNEE
CARATTERISTICHE POLIGRAFICHE: SE CENTRALI O PERIFERICHE

**2) CONCLUSIONI:** NORMALE /ANOMALIE LIEVI /MEDIE / GRAVI
CRISI SI' / NO
MPNE SI' / NO

**3) INTERPRETAZIONE** IN RELAZIONE AL QUESITO PER IL QUALE E' STATO RICHISTO L'ESAME

Milano, ....., 2020 Firma .....