The ‘Fitting’ Child

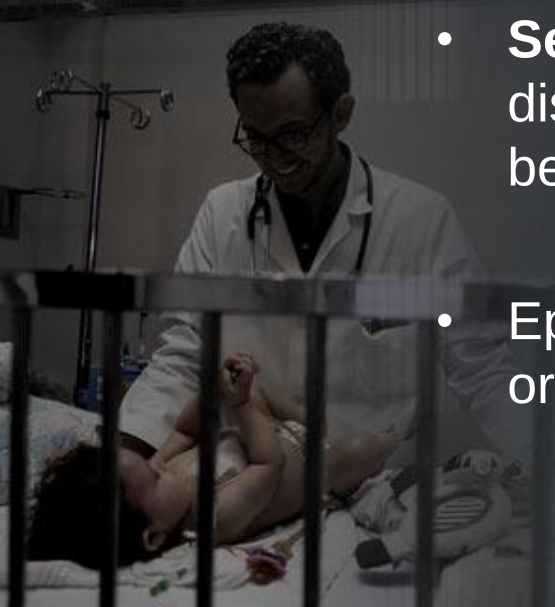
A/Prof Alex Tang

Objective

- Define relevant history taking and physical examination
- Classify the types of epilepsy in children
- Demonstrate the usefulness of investigations
- Define treatment strategies

Seizures in Children

- **Seizure**s are among the most common pediatric neurological disorders.
- The overall prevalence of epilepsy is approximately 1%
- 5% of all children experience febrile **seizure**s before the age of 6 years
- **Seizure**s are caused by an abnormal and excessive discharge of neurons, usually accompanied by behavioral or sensorimotor manifestations
- Epilepsy is defined classically as the occurrence of two or more unprovoked **seizure**s.

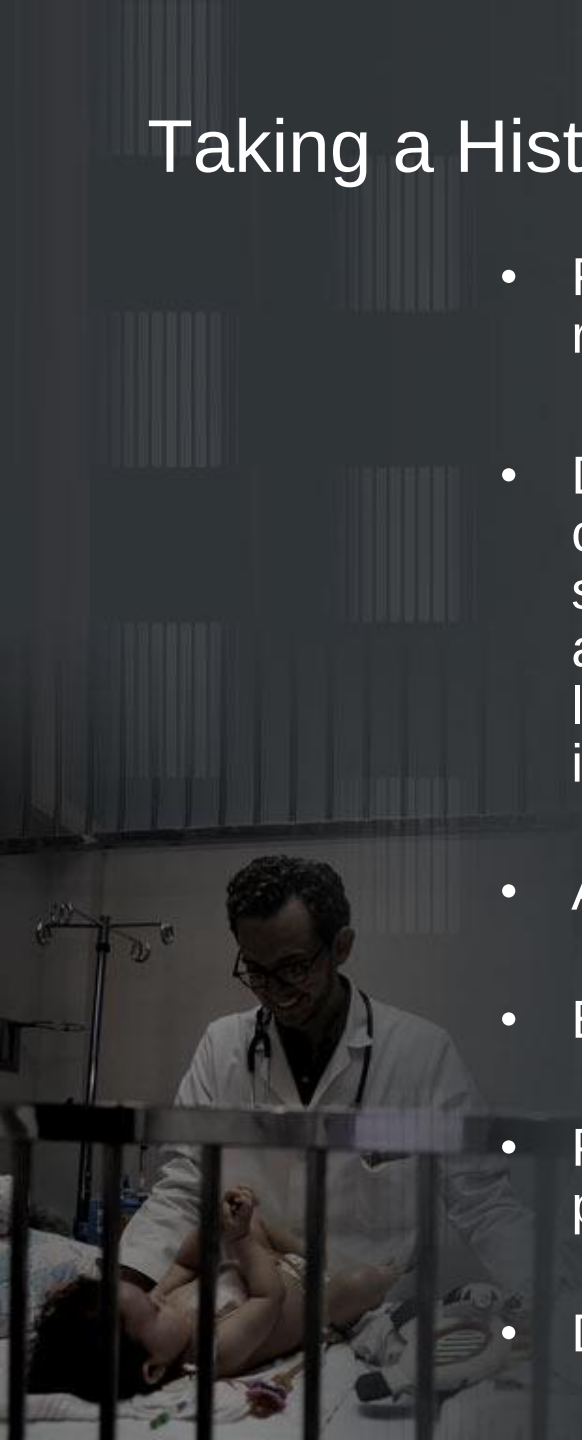


Taking a History

Taking a History

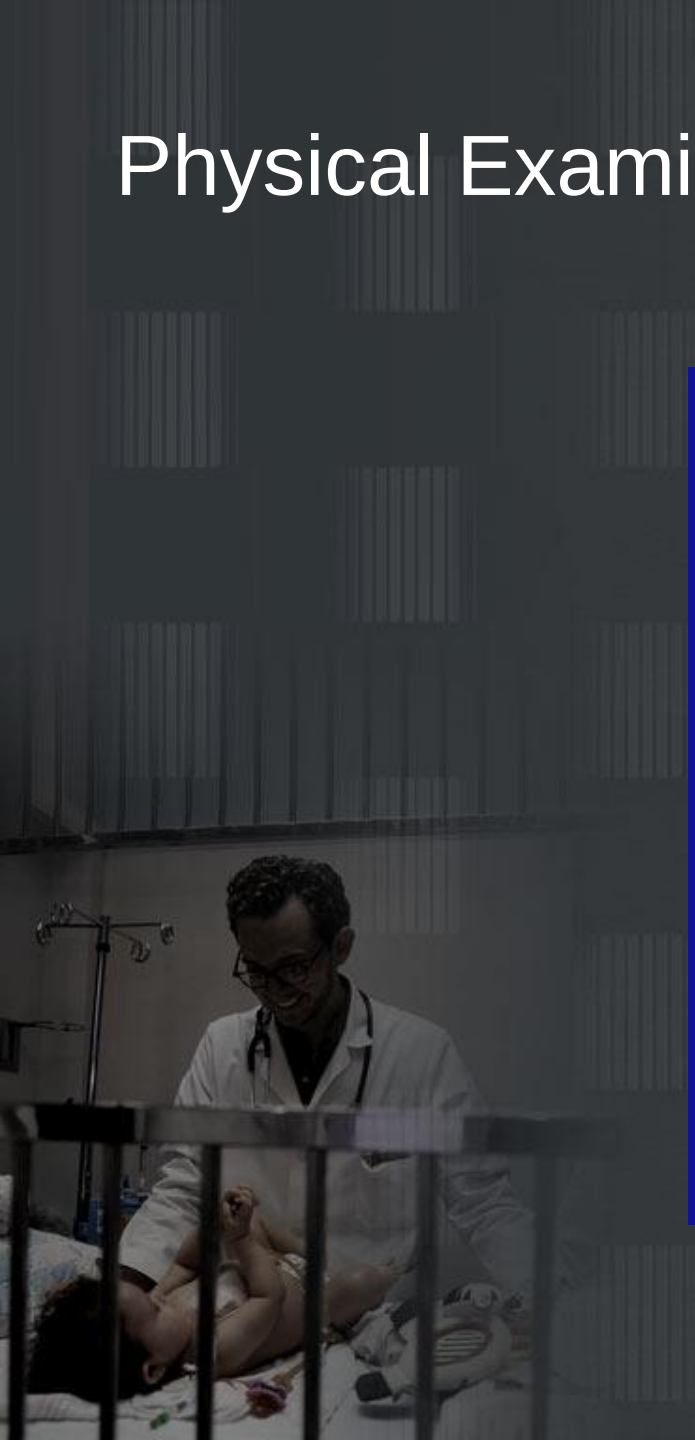
- Pregnancy history: Ultrasonography results, infections, medications, alcohol use, cigarette smoking, drug abuse, trauma, prematurity
- Prenatal history: Labor duration, spontaneous vaginal delivery or cesarean section, birth difficulties (resuscitation, intubation), birthweight, head circumference at birth
- Development: Fine motor, language, gross motor, and social skills
- School functioning
- General medical history: Head trauma, meningitis, stroke

Taking a History



- Family history: Epilepsy, febrile **seizure**s, mental retardations
- Description of the events: aura; motor (myoclonic or clonic jerk, hypertonia, atonia, chewing movements), sensory (somesthetic, auditive, visual, gustatory), autonomic, or psychologic phenomena; automatisms; level of consciousness; tongue-biting; fecal or urinary incontinence; episode length; postictal state
- Age at event onset
- Event frequency
- Precipitating factors: Fever, sleep deprivation, stress, photosensitivity, drugs, alcohol withdrawal, or others
- Diurnal and nocturnal patterns

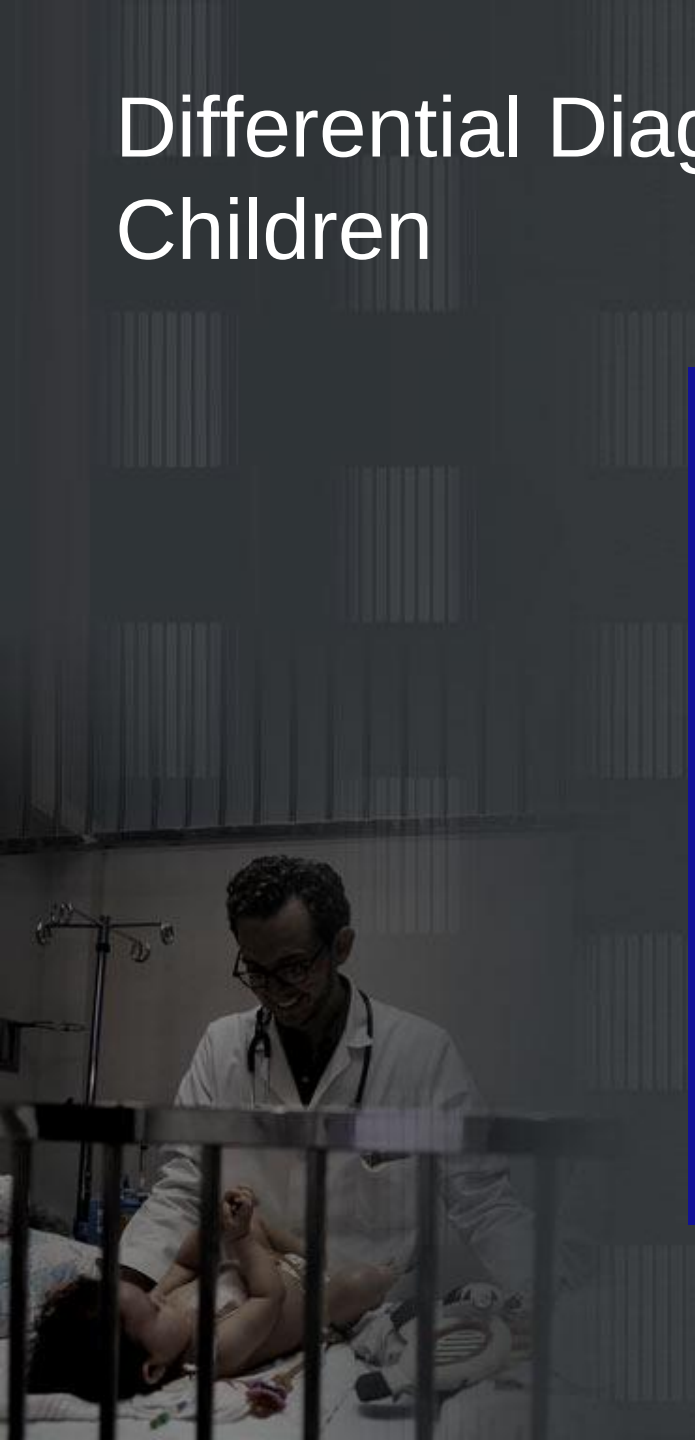
Physical Examination



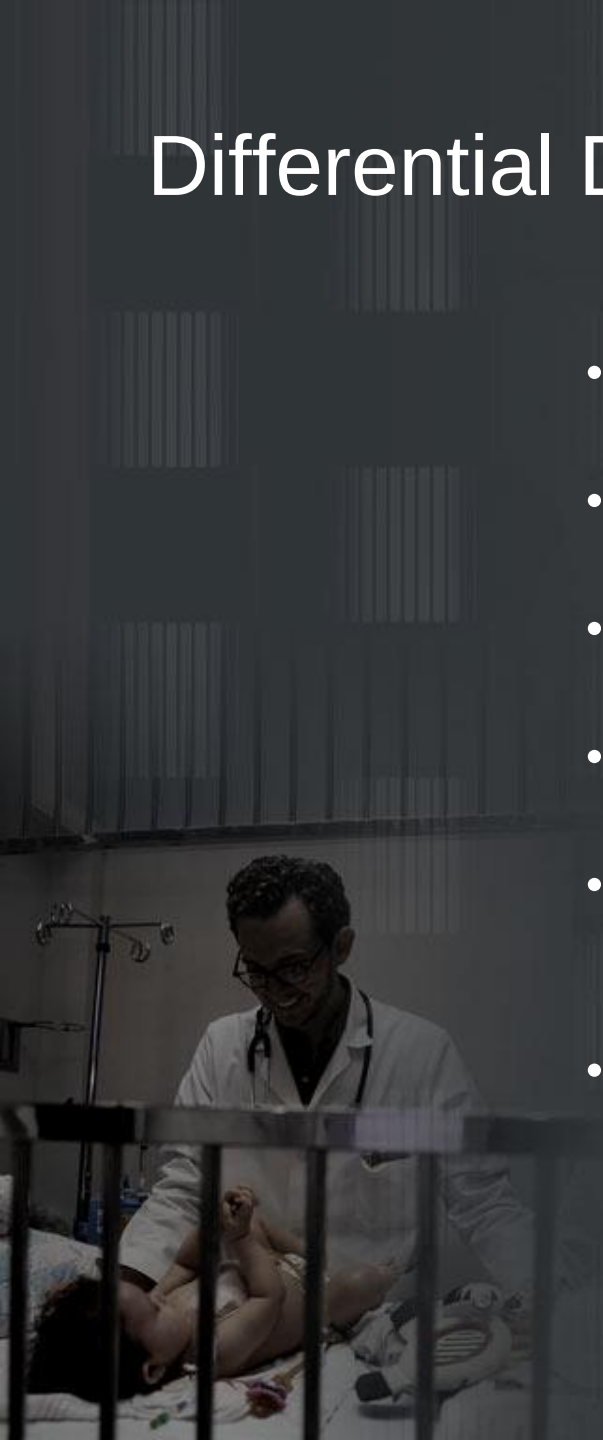
Physical Examination

- State of consciousness, language, social interactions
- Observation of the events (if possible); hyperventilation sometimes can provoke absence **seizure**s
- Global development
- Dysmorphic features, limb asymmetry, neurocutaneous skin findings, organomegaly
- Head circumference
- Neurologic examination: Cranial nerves, motor strength and tone, reflexes, sensory and cerebellar function tests, gait

Differential Diagnosis of Epilepsy in Children

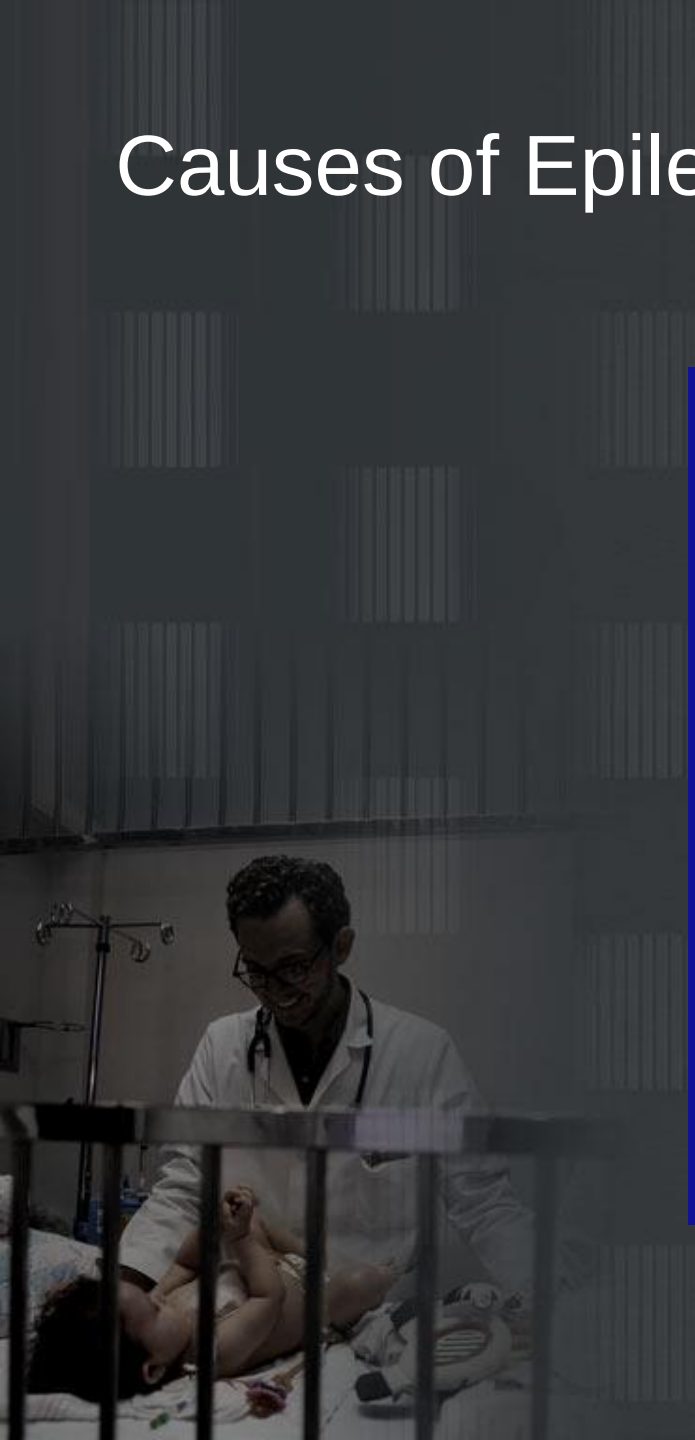


Differential Diagnosis of **Seizure**s

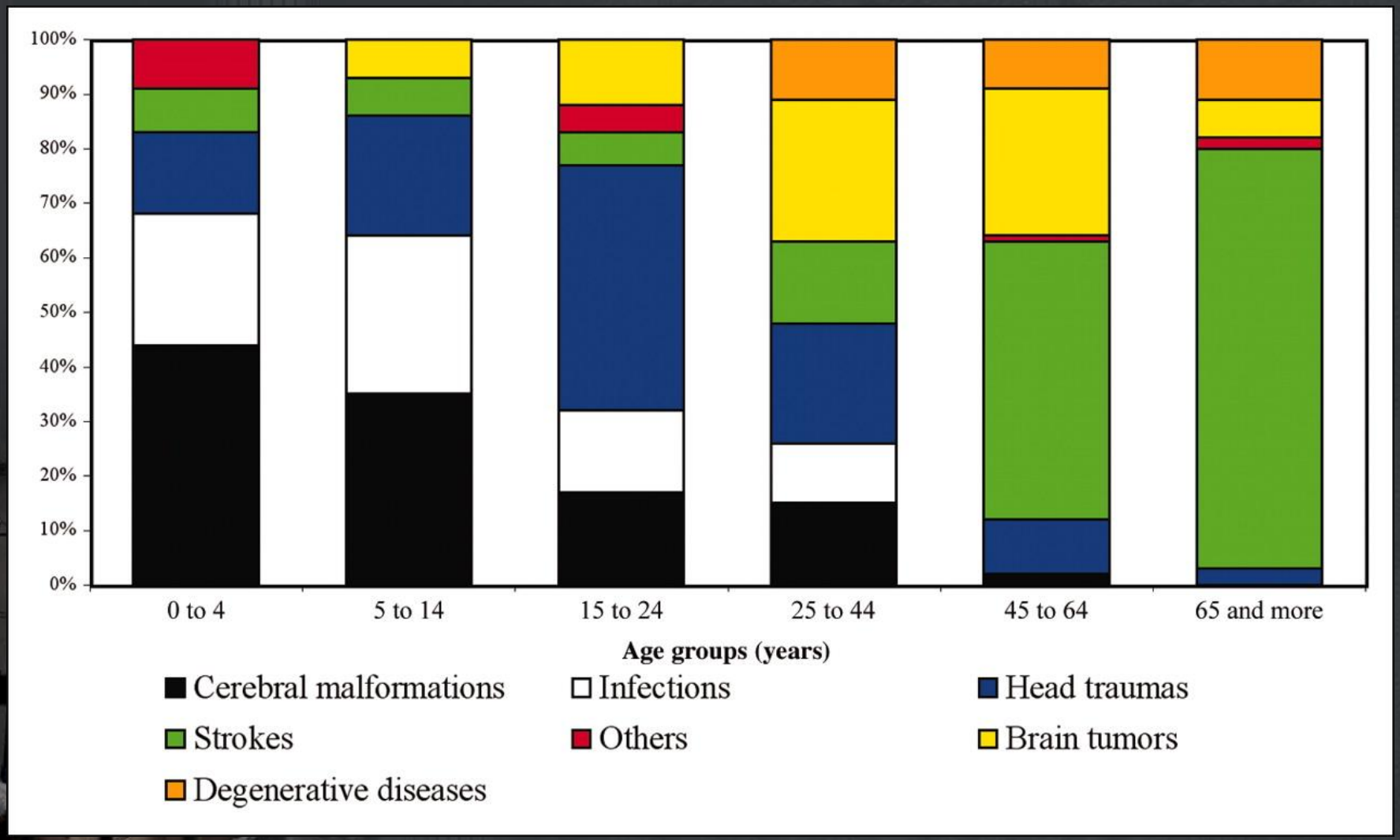


- Syncope
- Daydreaming
- Migraine
- Breath-holding spells
- Transient ischemic events
- Vestibular disorders
- Gastroesophageal reflux
- Movement disorders (tics, paroxysmal choreoathetosis)
- Psychotic hallucinations and delusions
- Nonepileptic events (pseudo**seizure**s)
- Panic attacks

Causes of Epilepsy in Children



Proportional incidences for symptomatic epilepsies according to age and etiology

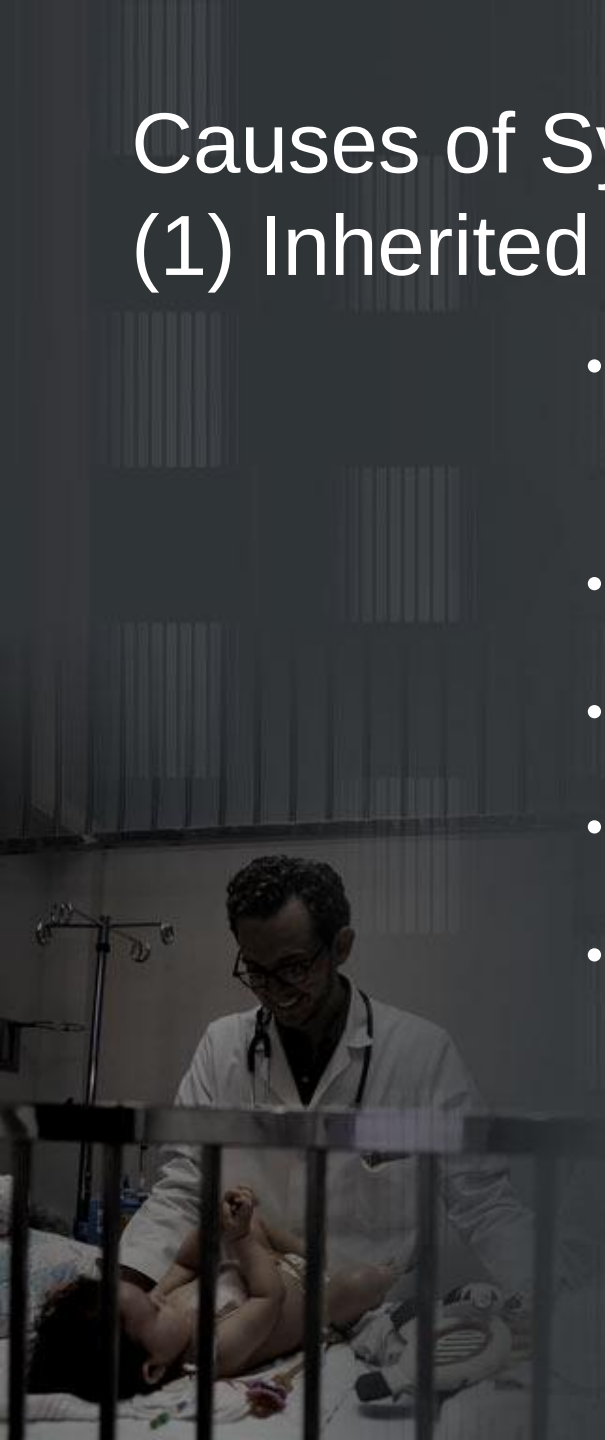


Adapted from Annegers JF. The epidemiology of epilepsy. In: Willie E, ed. *The Treatment of Epilepsy: Principles and Practice.* Philadelphia, Pa: Lea & Febiger; 2001:135.

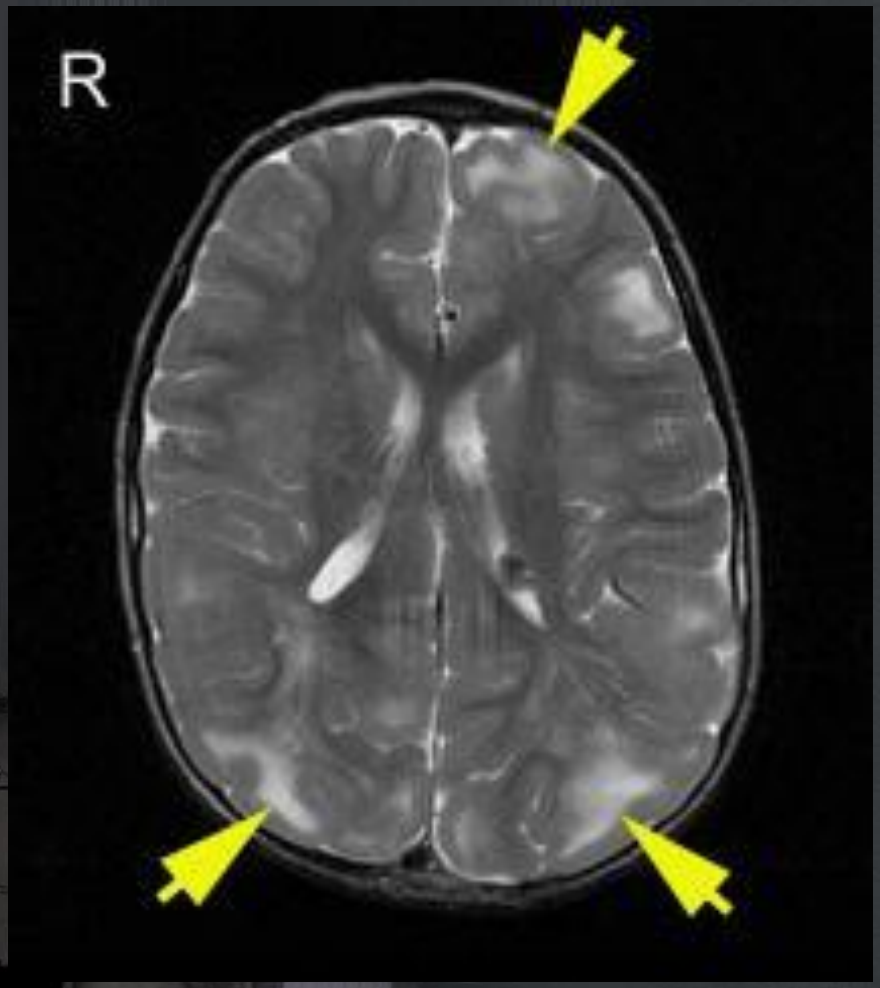
Causes of Symptomatic Epilepsy

(1) Inherited Genetic
(2) Congenital (inherited or acquired)
(3) Acquired

Causes of Symptomatic Epilepsy
(1) Inherited Genetic



- Channelopathies, defined as mutations of neuronal ion channels (eg, one sodium channel defect is associated with benign familial neonatal **seizure**s)
- Chromosomal abnormalities
- Mitochondrial DNA disorders
- Metabolic disorders
- Hereditary neurocutaneous disorders
  - —Tuberous sclerosis complex
  - —Neurofibromatosis
  - —Sturge Weber syndrome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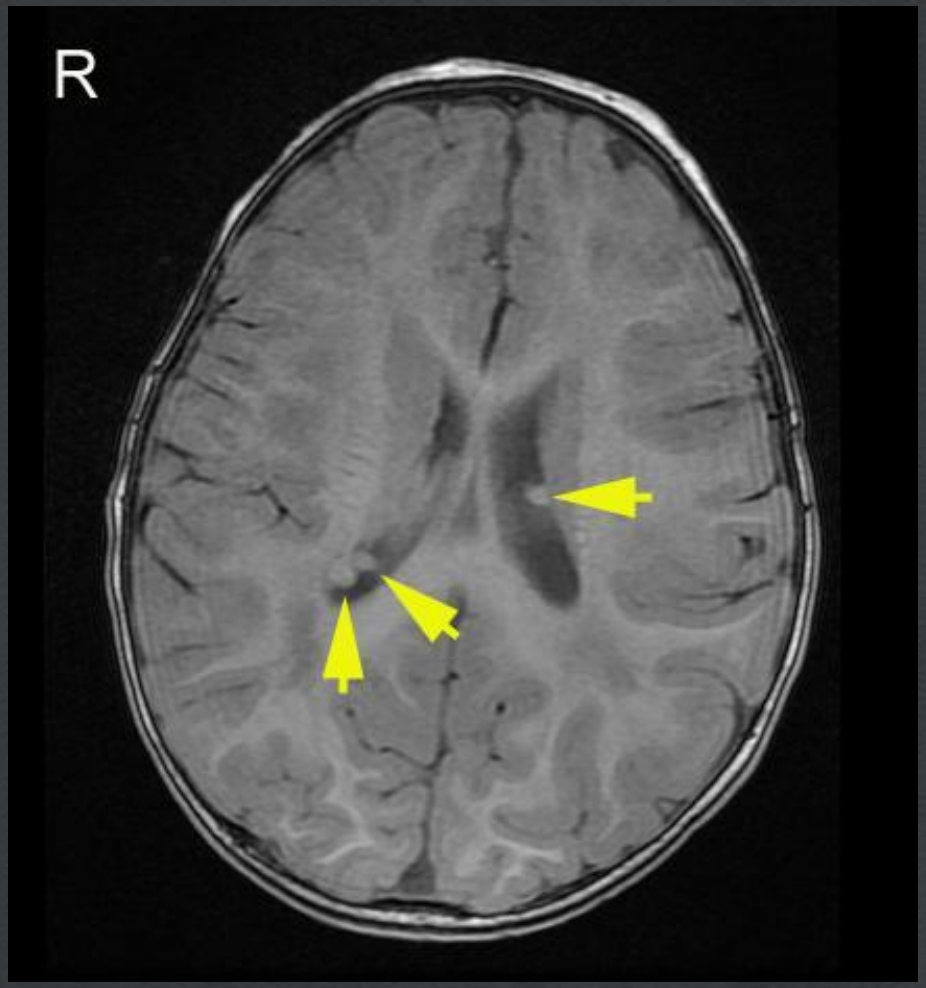
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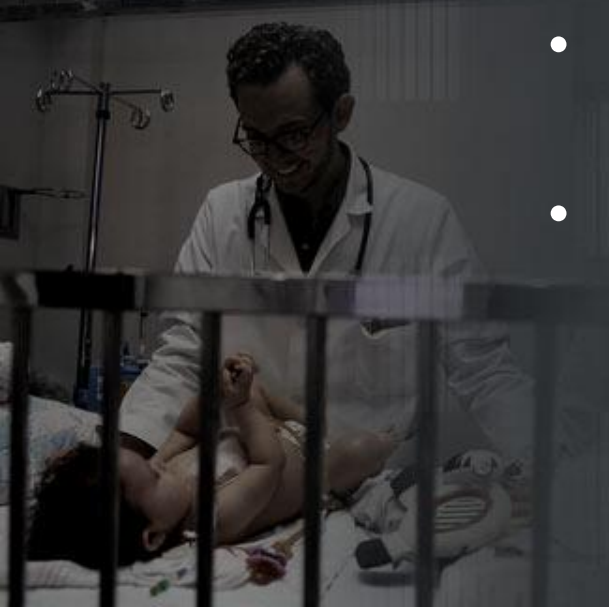
Causes of Symptomatic Epilepsy
(2) Congenital (Inherited or Acquired)



- Developmental cortical malformations
- Cerebral tumor
- Vascular malformations
- Prenatal injury

Causes of Symptomatic Epilepsy (3) Acquired

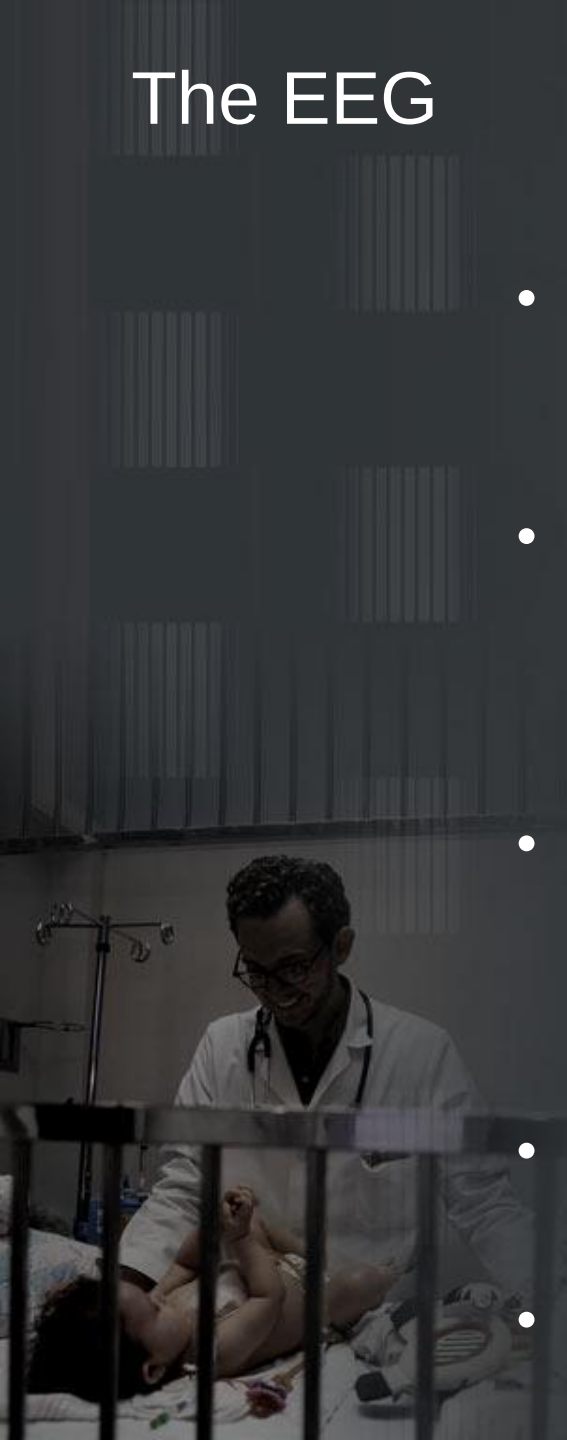
- Trauma
- Neurosurgery
- Infection
- Vascular disease
- Hippocampal sclerosis
- Tumors
- Neurodegenerative disorders
- Metabolic disorders
- Toxic disorders

Investigations

Investigations

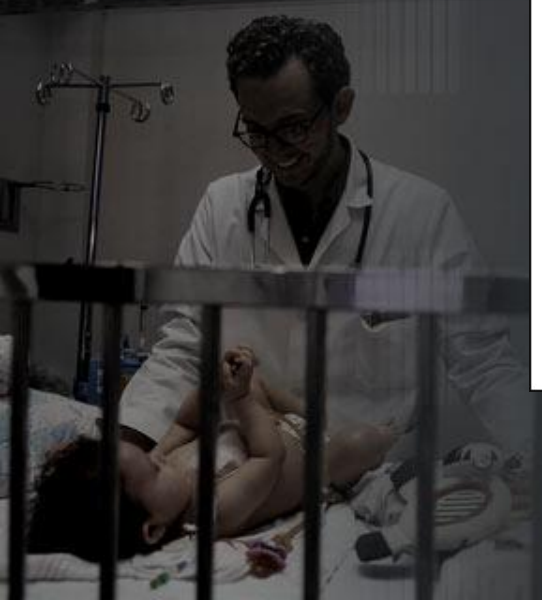
- Electrophysiology
  - Electroencephalogram (EEG)
- Brain imaging
  - CT head
  - MRI
  - PET
  - SPECT
  - fMRI
- Video
- Neuropsychological evaluation

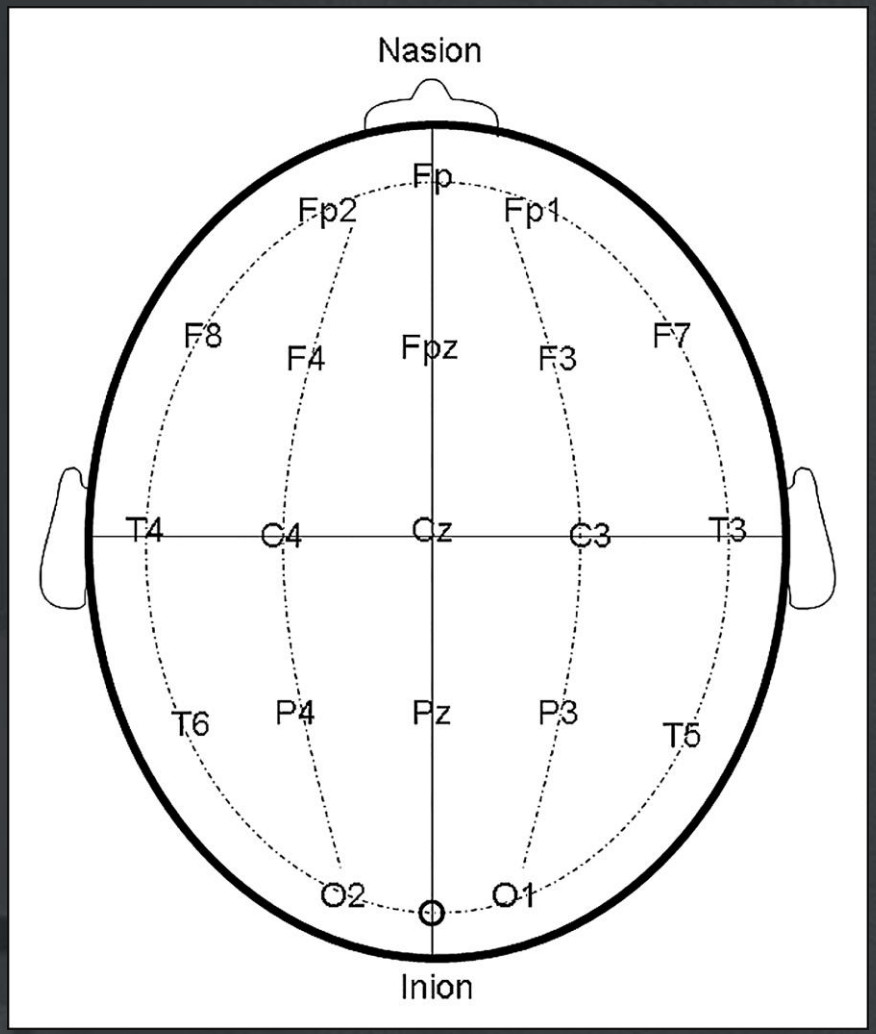
The EEG



- a normal EEG is noted in 10% to 20% of children who have epilepsy
- Hyperventilation can trigger epileptic discharges in 80% of patients who have generalized absence epilepsy
- photic stimulation induces EEG abnormalities in up to 40% of patients who have generalized epilepsy
- a sleep-deprived EEG
- Long-term video-EEG monitoring

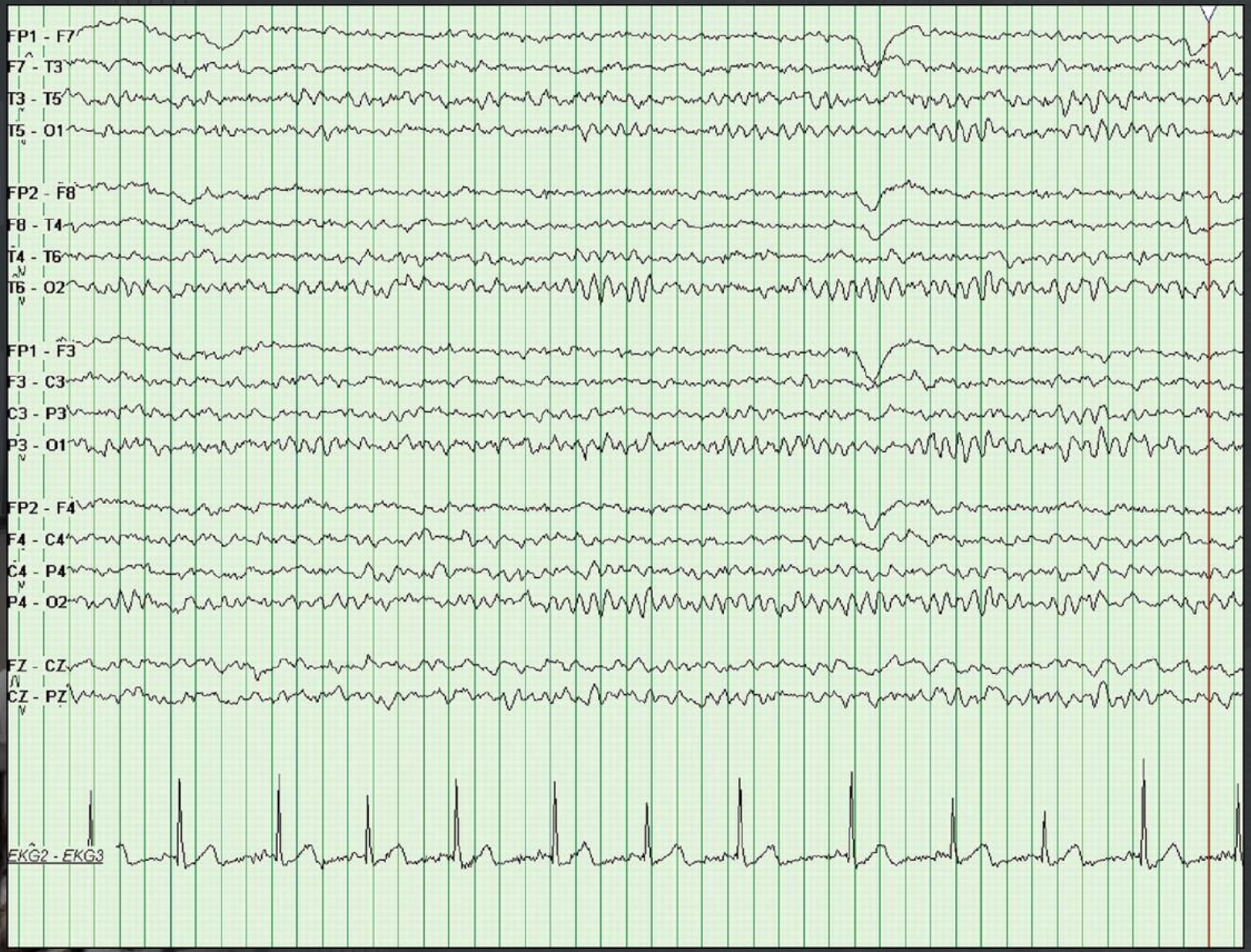
Scalp electrode positions for EEG





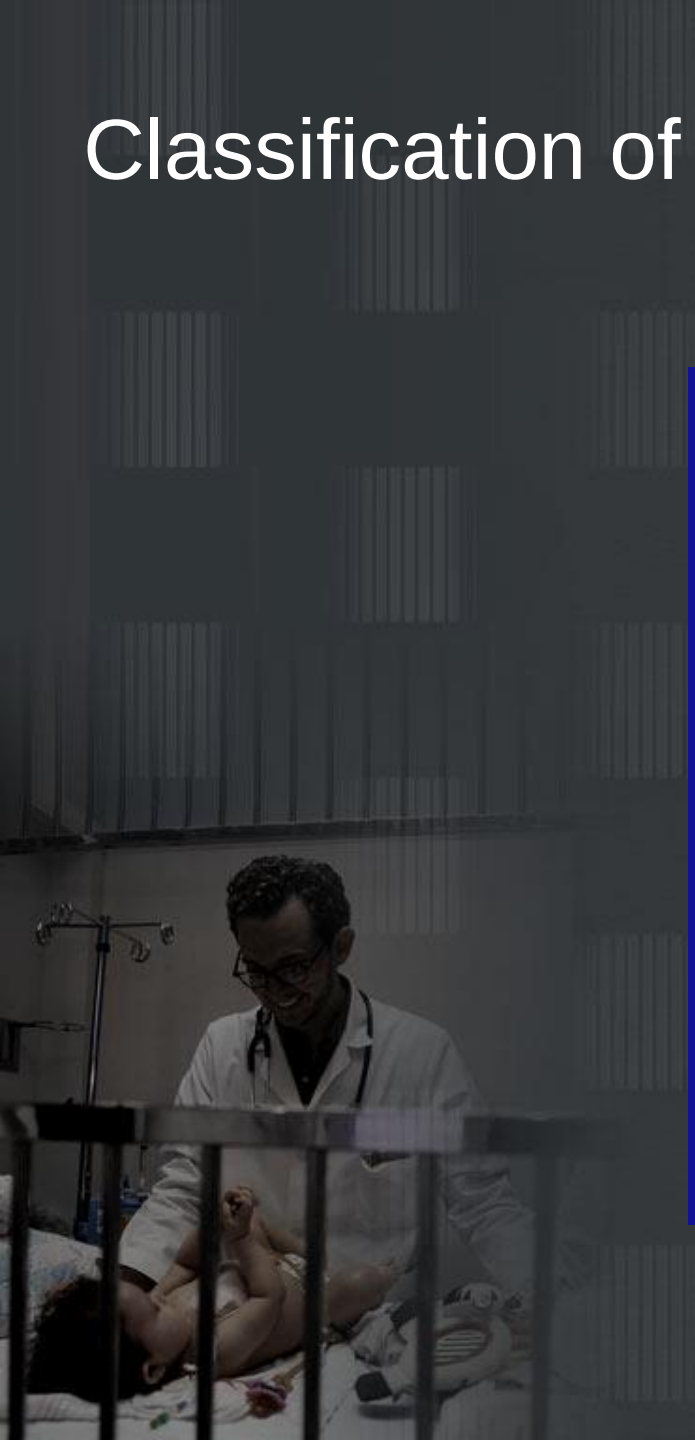
Major, P. et al. Pediatrics in Review 2007;28:405-414

Normal EEG tracing showing a reactive posterior alpha (9-Hz) rhythm in an 8-year-old boy who has no history of seizures



Major, P. et al. Pediatrics in Review 2007;28:405-414

Classification of Epilepsy in Children



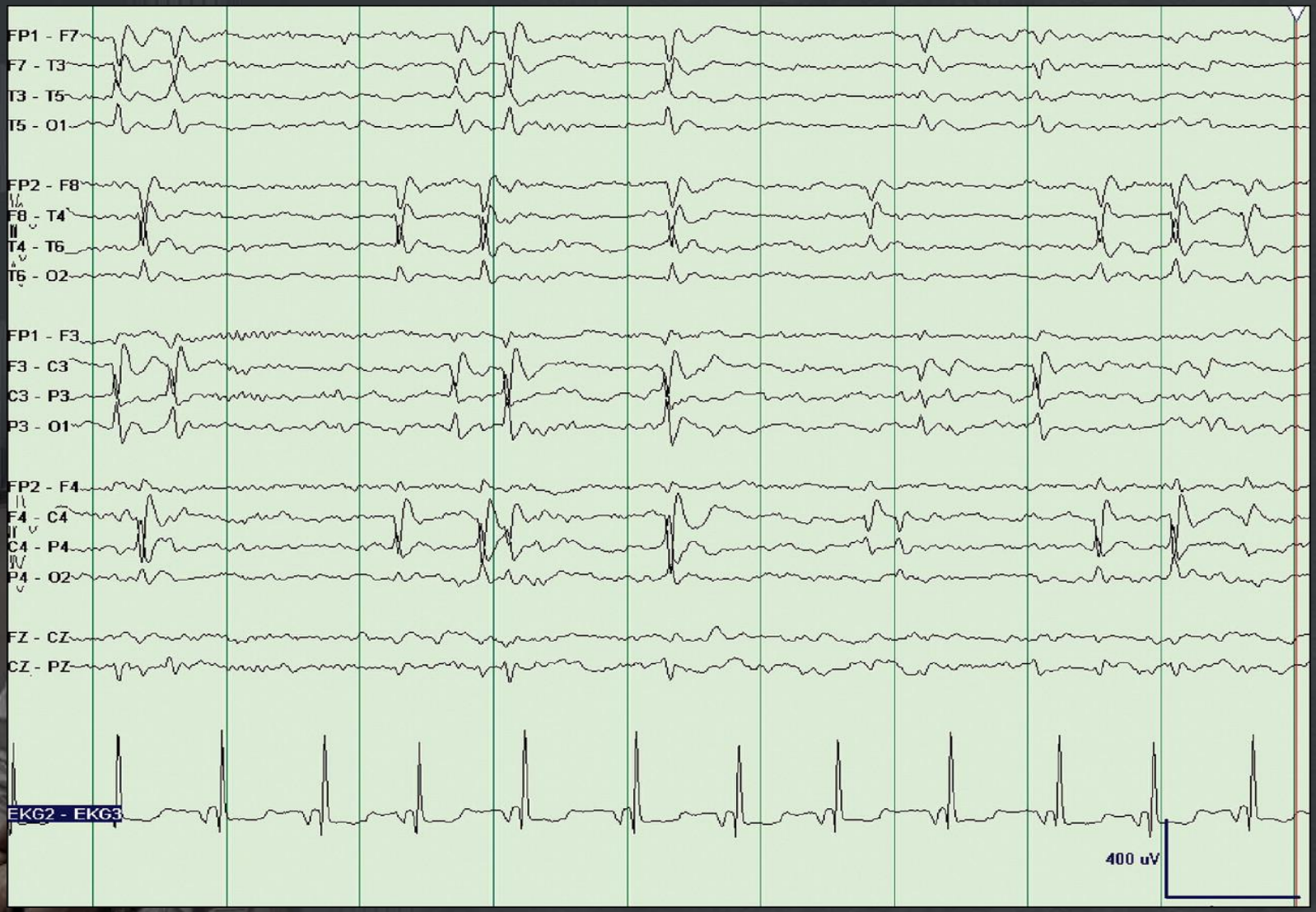
International Classification of Epileptic Seizures

(1) Partial (Focal, Localized) Seizures
(2) Generalized Seizures
(3) Unclassified Seizures

Localization-related (Focal, Local, Partial)

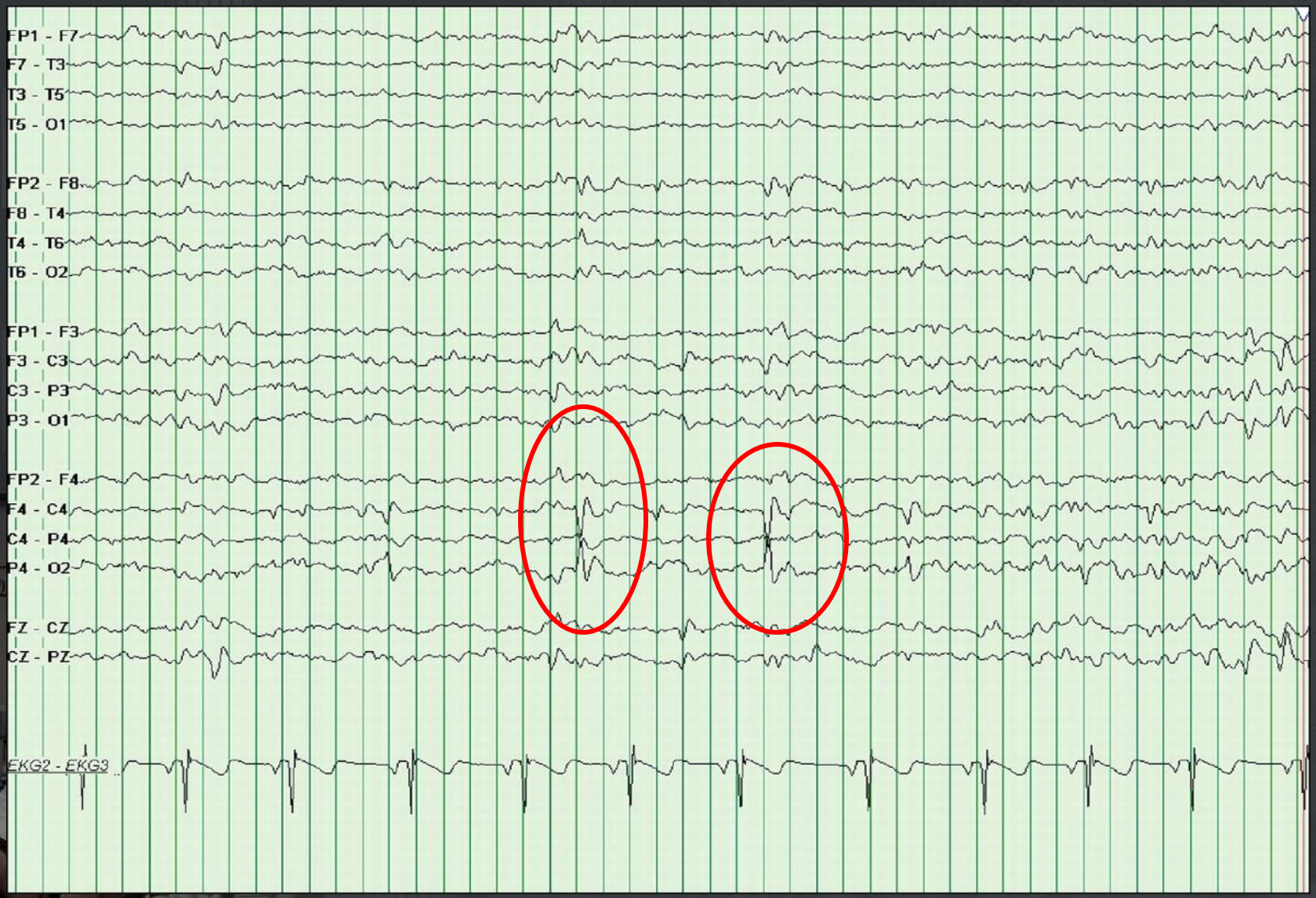
- Idiopathic (primary)
  - —Benign childhood epilepsy with centrotemporal spikes (Benign Rolandic Epilepsy)
  - —Childhood epilepsy with occipital paroxysms
  - —Primary reading epilepsy
- Symptomatic (secondary)
  - —Temporal lobe epilepsies
  - —Frontal lobe epilepsies
  - —Parietal lobe epilepsies
  - —Occipital lobe epilepsies
  - —Chronic progressive epilepsia partialis continua of childhood
  - —Syndromes characterized by **seizure**s that have specific modes of precipitation
- Cryptogenic, defined by
  - —**Seizure** type
  - —Clinical features
  - —Anatomic localization

EEG tracing showing frequent independent left and right centrotemporal spikes in an 8-year-old child who has benign partial epilepsy with centrotemporal spikes (also called benign rolandic epilepsy)



Major, P. et al. Pediatrics in Review 2007;28:405-414

EEG tracing showing a right centroparietal spike (spikes observed in P4-O2, C4-P4, and F4-C4 leads) in a 12-year-old girl who has partial epilepsy

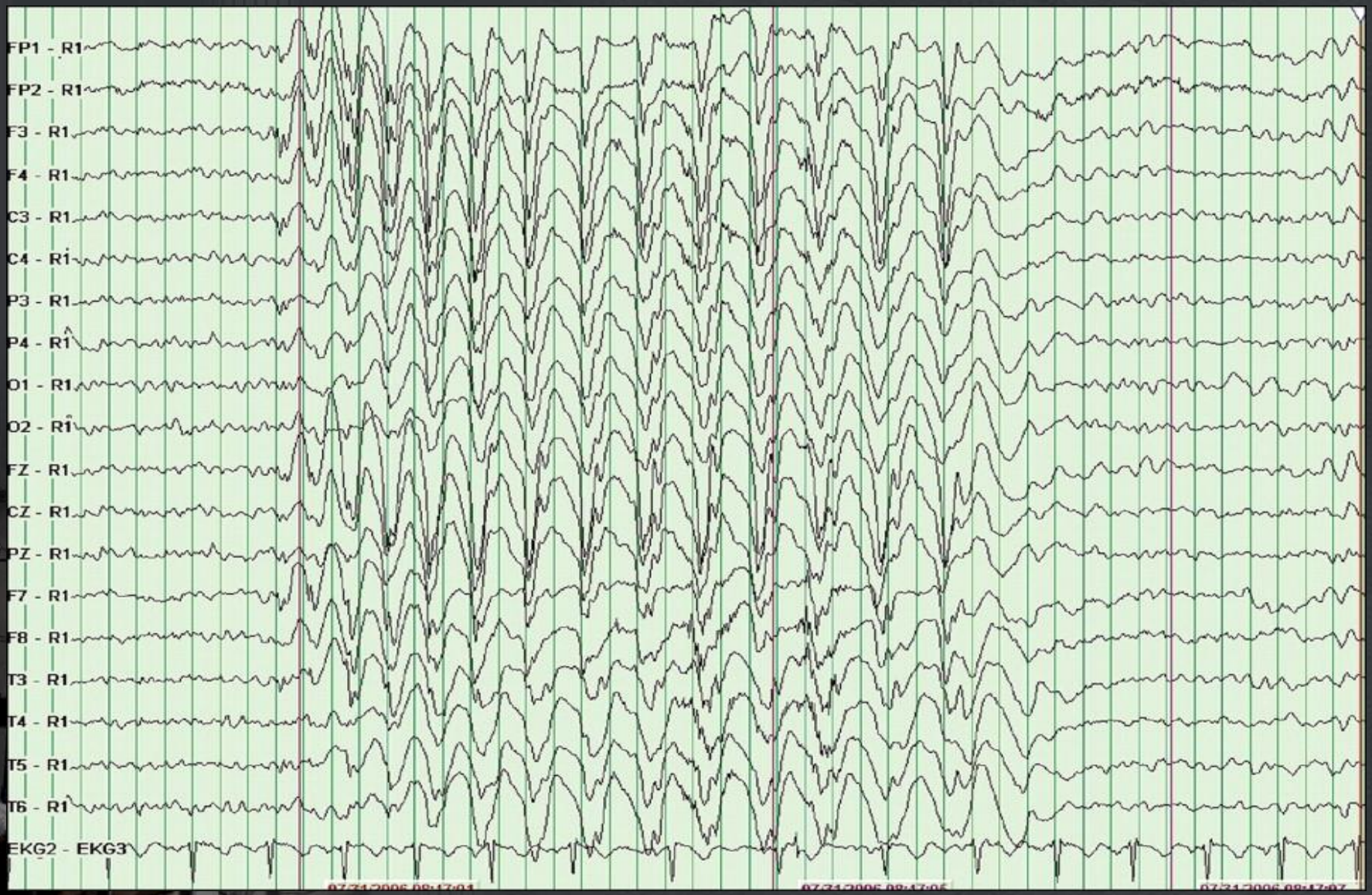


Major, P. et al. Pediatrics in Review 2007;28:405-414

(2) Generalized **Seizure**s (Convulsive or Non-convulsive)

- Absence **seizure**s
  - —Typical absences
  - —Atypical absences
- Myoclonic **seizure**s
- Clonic **seizure**s
- Tonic **seizure**s
- Tonic-clonic **seizure**s
- Atonic **seizure**s

EEG tracing showing a generalized 3-Hz spike and wave discharge lasting 6 seconds in a 7-year-old girl who has generalized absence epilepsy



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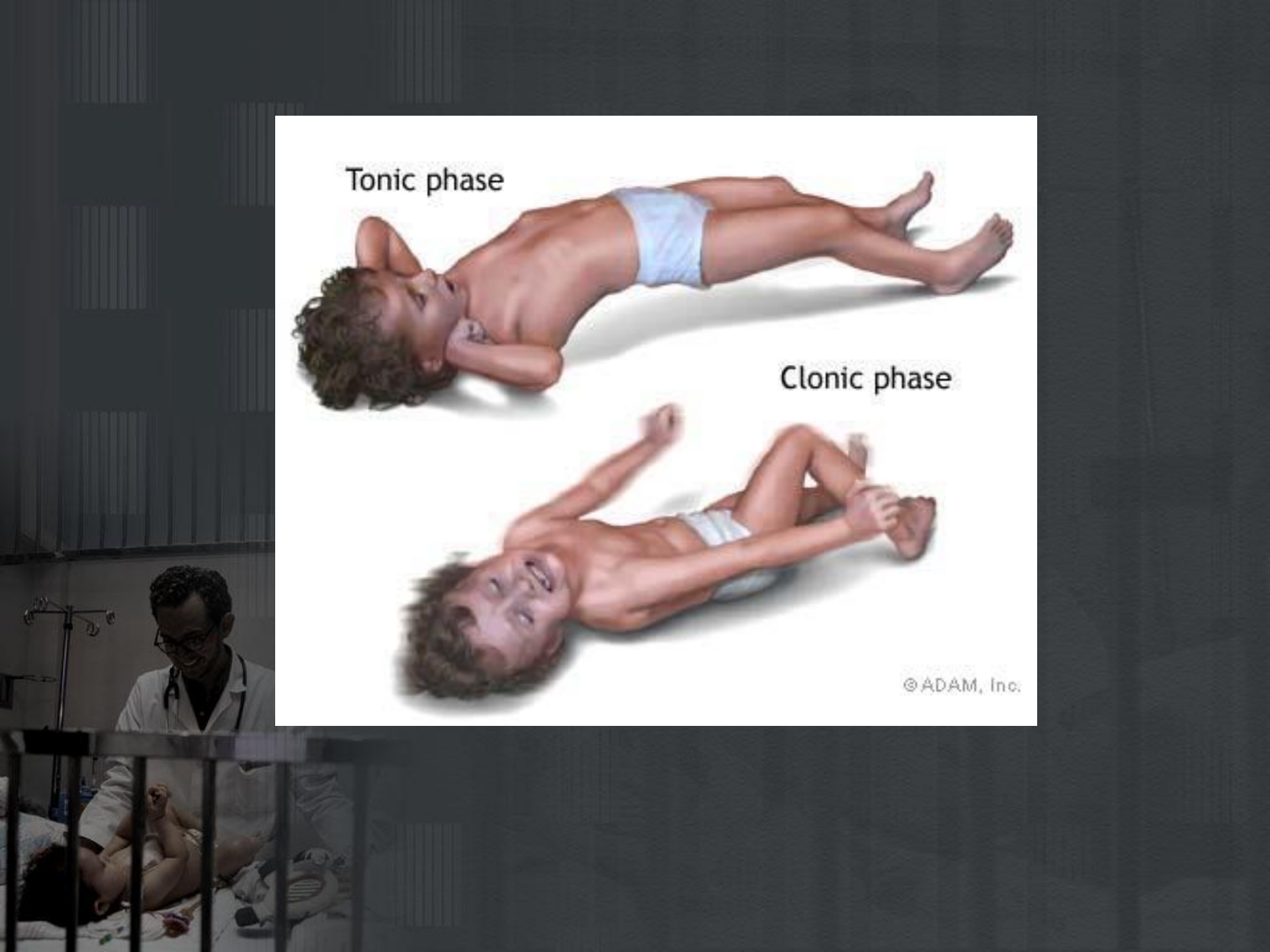
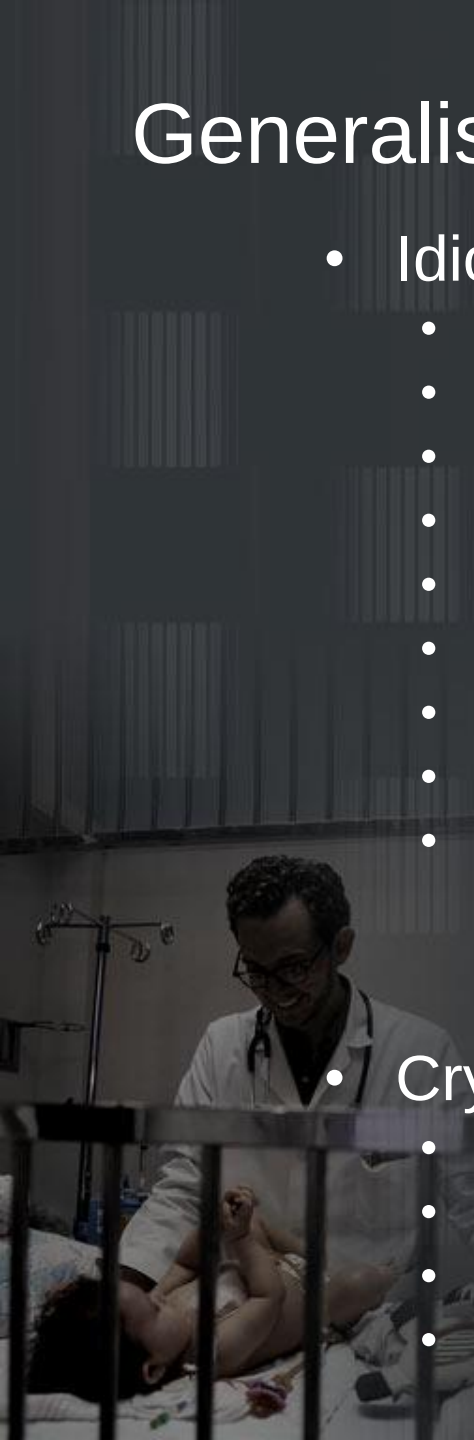
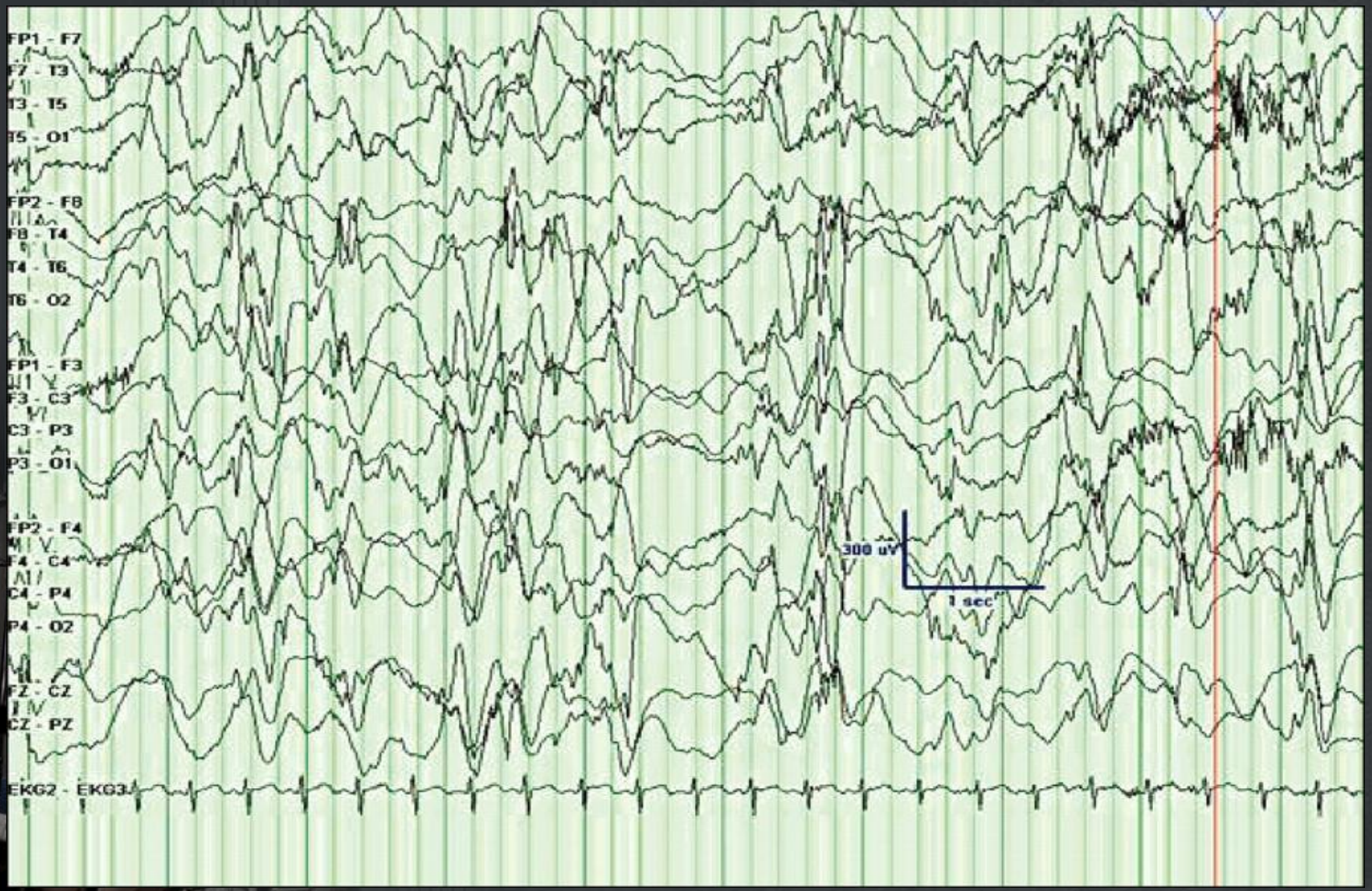

Tonic phase
Clonic phase
©ADAM, Inc.

Generalised Seizures

- Idiopathic (primary)
  - —Benign neonatal familial convulsions
  - —Benign neonatal convulsions
  - —Benign myoclonic epilepsy in infancy
  - —Childhood absence epilepsy (pyknolepsy)
  - —Juvenile absence epilepsy
  - —Juvenile myoclonic epilepsy (Janz syndrome)
  - —Epilepsies with grand mal **seizure**s on awakening
  - —Other generalized idiopathic epilepsies
  - —Epilepsies with **seizure**s precipitated by specific modes of activation
- Cryptogenic or symptomatic
  - —West syndrome (infantile spasms)
  - —Lennox-Gastaut syndrome
  - —Epilepsy with myoclonic-astatic **seizure**s
  - —Epilepsy with myoclonic absences



EEG showing hypsarrhythmia in a 9-month-old girl who has infantile spasms



Major, P. et al. Pediatrics in Review 2007;28:405-414

Generalised Seizures

- Special syndromes
  - Situation-related **seizure**s
    - —Febrile convulsions
    - —Isolated **seizure**s or isolated status epilepticus
  - **Seizure**s occurring only with an acute or toxic event, due to factors such as alcohol, drugs, eclampsia, and nonketotic hypoglycemia

Febrile **Seizure**s

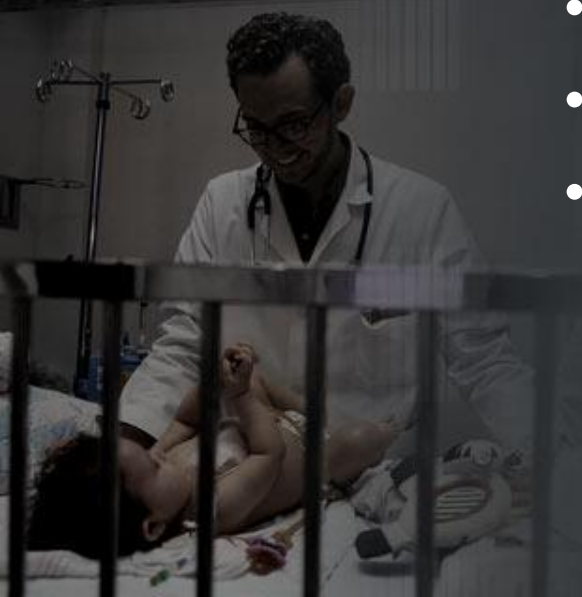
- **Seizure** occurrence between ages 3 months and 6 years of age
- Normal development and normal neurologic examination findings
- Duration <15 min
- Generalized tonic-clonic **seizure**
- Only one **seizure** during one febrile episode
- No postictal deficit (eg, Todd paralysis)
- Not caused by a central nervous system infection

(3) Unclassified Epileptic **Seizure**s

- With both generalized and focal **seizure**s
  - —Neonatal **seizure**s
  - —Severe myoclonic epilepsy in infancy (Dravet syndrome)
  - —Epilepsy with continuous spike and waves during slow-wave sleep
  - —Acquired epileptic aphasia (Landau-Kleffner syndrome)
  - —Other undetermined epilepsies
- Without unequivocal generalized and focal features

Adapted from the Commission on Classification and Terminology of the International League Against Epilepsy. Proposal for revised clinical and electroencephalographic classification of epileptic **seizure**s. *Epilepsia*. 1981;22:489–501.

Neonatal Convulsion etiologies

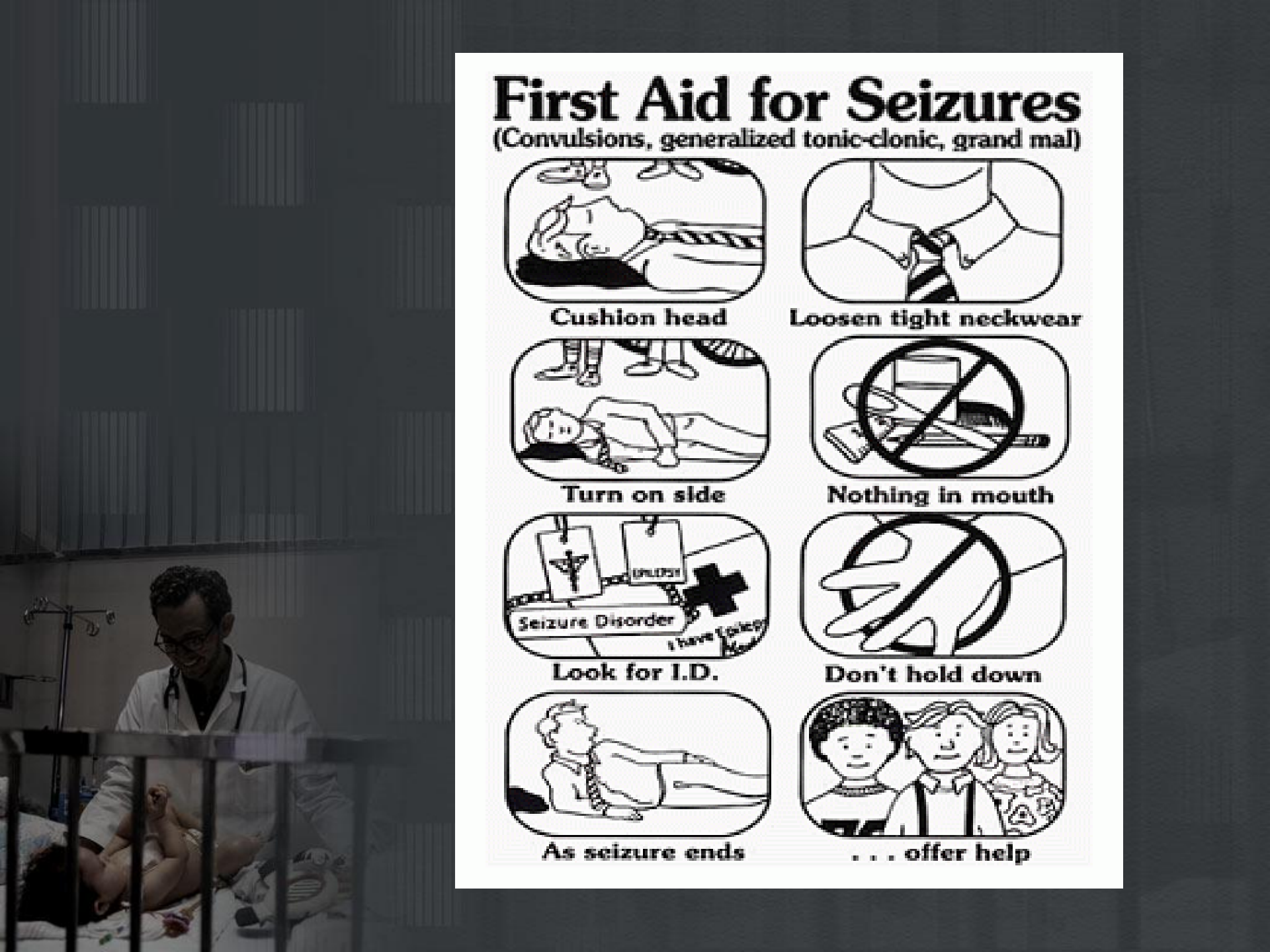


- Hypoxic-ischemic encephalopathy
- Intraventricular hemorrhage
- Subarachnoid hemorrhage
- Hypoglycemia
- Hypocalcemia
- Intracranial infection
- Cerebral dysgenesis

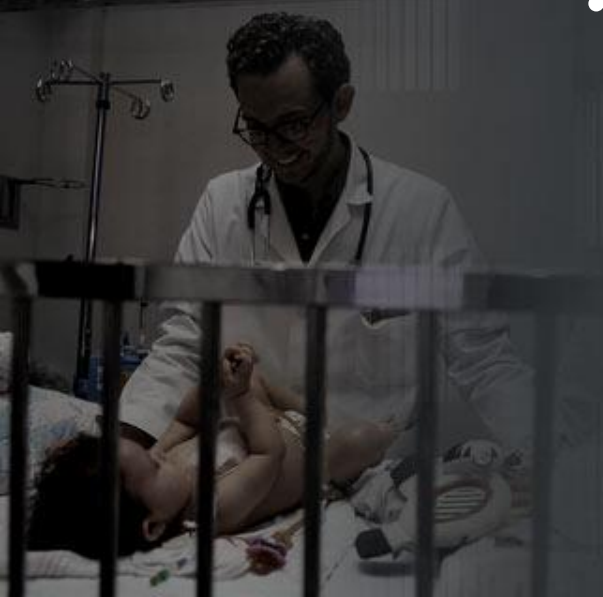
Management

Treatment

- Educate parents and child about epilepsy
  - Precipitating factors
  - Seizure first aid
  - Sports
- Treatment strategies
  - Anti-epileptic drugs
  - Special diets
  - Surgery
  - Vagus nerve stimulation


First Aid for Seizures
(Convulsions, generalized tonic-clonic, grand mal)
Cushion head
Loosen tight neckwear
Turn on side
Nothing in mouth
Seizure Disorder
Look for I.D.
Don't hold down
As seizure ends
. . . offer help

Anti-epileptic drugs

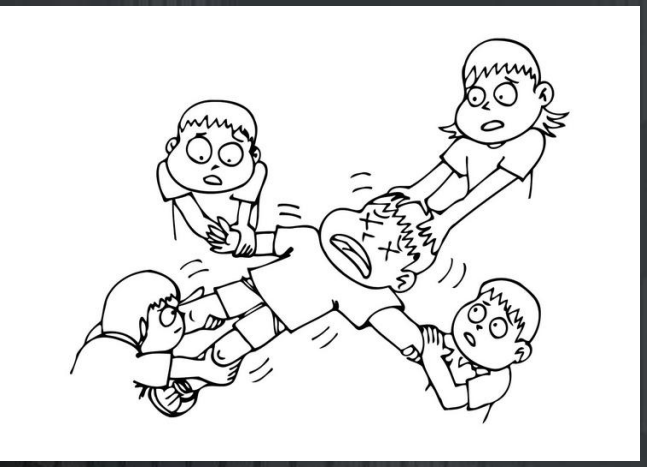


- Benzodiazepines
- Valporate
- Carbamazepine
- Phenytoin
- Phenobarbitone
- Ethosuximide
- Primidome
- Gabapentin
- Tiagabine
- Oxcarbezepine
- Lamotrigine
- Topiramate

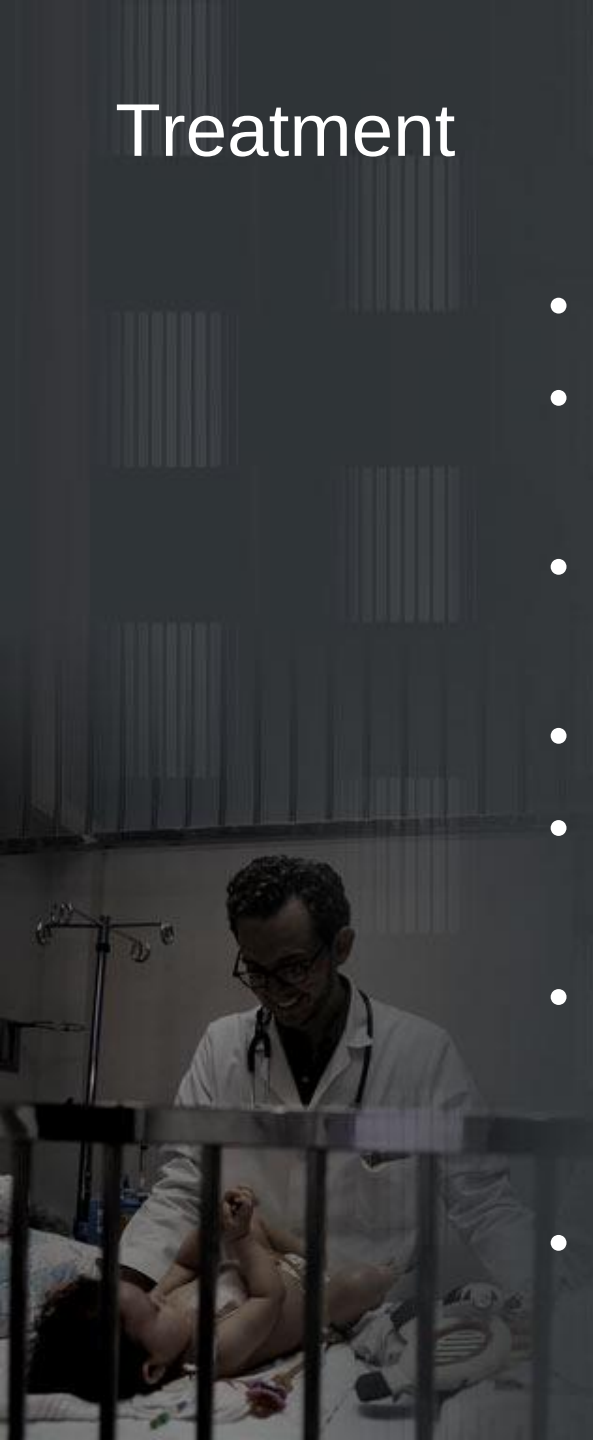
Anti-epileptic drugs

- When to start?
- Monitoring
- When to stop?

Treatment of a Seizure

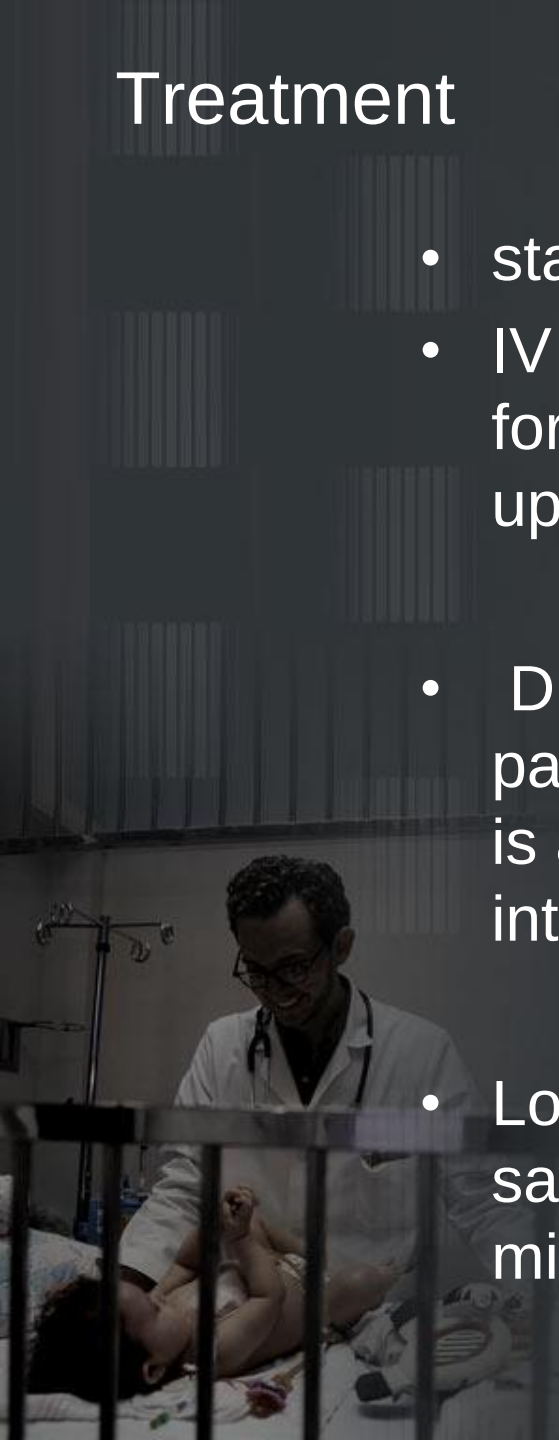


Treatment



- secure the airway
- placed on his or her side to prevent aspiration
- make sure that the upper respiratory airway is free
- oxygen is by mask
- Blood pressure and electrocardiographic monitoring
- blood should be drawn for a complete blood count, electrolytes, blood glucose, calcium, and magnesium and toxicologic screen
- intravenous (IV) line should be placed

Treatment



- start treatment with a benzodiazepine
- IV lorazepam at a dose of 0.1 mg/kg (0.15 mg/kg for patients already receiving a benzodiazepine) up to a maximum of 4 mg

- Diazepam at a dose of 0.3 mg/kg (0.5 mg/kg for patients already receiving a benzodiazepine) also is a good option and can be administered intravenously, intrarectally, or endotracheally

- Lorazepam or diazepam can be repeated at the same dose if the seizure does not stop after 5 minutes

Treatment

- The second step is to use IV phenytoin or phenobarbital
- Phenytoin is given at a dose of 20 mg/kg intravenously up to a maximum of 1,250 mg
- Phenobarbital is administered at a dose of 10 to 20 mg/kg, up to a maximum of 300 mg.

Status Epilepticus

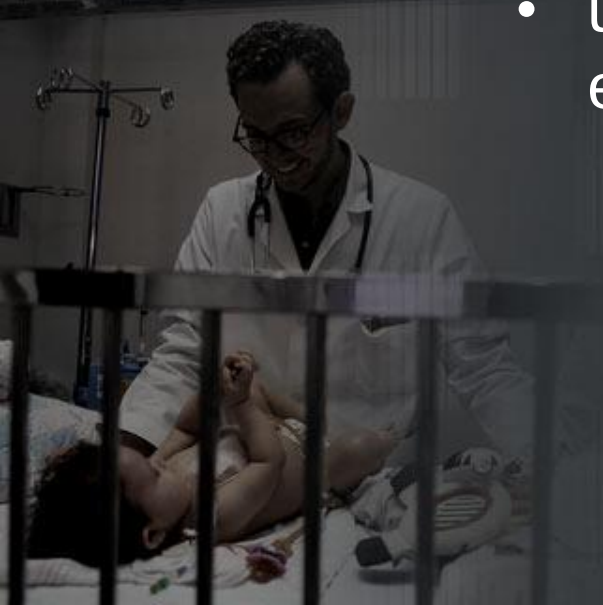
- a continuous **seizure** or the occurrence of serial **seizure**s, between which there is no return of consciousness, lasting more than 30 minutes

- a continuous **seizure** lasting more than 30 minutes potentially can harm the brain

- increased metabolic demand by constantly discharging neurons produces regional oxygen insufficiency that causes cell damage and necrosis

Treatment

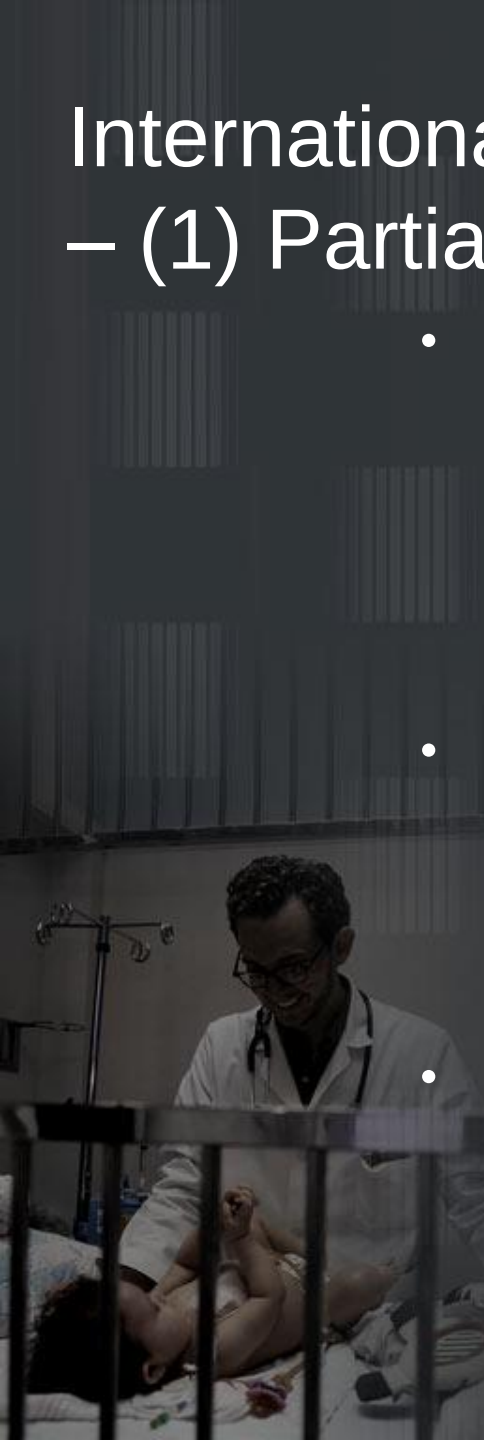
- the third step is to induce a "barbiturate coma."
- intubation is mandatory, and an anesthesiologist should be involved
- use midazolam, valproic acid, or other anti-epileptic drugs



Objective

- Define relevant history taking and physical examination
- Classify the types of epilepsy in children
- Demonstrate the usefulness of investigations
- Define treatment strategies

International Classification of Epileptic **Seizure**s – (1) Partial (Focal, Localized) **Seizure**



- Simple partial **seizure**s
  - —With motor signs
  - —With somatosensory or special sensory systems
  - —With autonomic symptoms and signs
  - —With psychic symptoms
- Complex partial **seizure**s
  - —Simple partial onset followed by impairment of consciousness
  - —With impairment of consciousness at onset
- Partial **seizure**s evolving to secondarily generalized **seizure**s
  - —Simple partial **seizure**s evolving to generalized **seizure**s
  - —Complex partial **seizure**s evolving to complex partial **seizure**s evolving to generalized **seizure**s

Generalised Seizures

- Symptomatic (secondary)
  - —Nonspecific cause
    - –Early myoclonic encephalopathy
    - –Early infantile epileptic encephalopathy with suppression burst
    - –Other symptomatic generalized epilepsies
  - —Specific syndromes
    - –Epileptic **seizure**s may complicate many disease states

Generalised Seizures

- With both generalized and focal **seizure**s
  - —Neonatal **seizure**s
  - —Severe myoclonic epilepsy in infancy (Dravet syndrome)
  - —Epilepsy with continuous spike and waves during slow-wave sleep
  - —Acquired epileptic aphasia (Landau-Kleffner syndrome)
  - —Other undetermined epilepsies
- Without unequivocal generalized and focal features