EEG in Childhood and Adolescent Epilepsy Syndrome

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Childhood Epilepsy Syndrome

- Genetic Epilepsy With Febrile Seizures Plus
- Panayiotopoulos syndrome (early-onset childhood occipital epilepsy)
- Myoclonic-atonic (formerly astatic) epilepsy (Doose syndrome)
- Benign epilepsy with centrotemporal spikes (benign rolandic epilepsy)
- Late-onset childhood occipital epilepsy (Gastaut type)
- Epilepsy with myoclonic absences (Tassinari syndrome)
- Lennox-Gastaut syndrome
- Epileptic encephalopathy with continuous spike and wave in slow sleep
- Acquired epileptic aphasia (Landau-Kleffner syndrome)
- Childhood absence epilepsy (pyknolepsy)
- Generalized epilepsy with eyelid myoclonia (Jeavons syndrome)

Adolescence to adult onset

- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy with generalized tonic-clonic seizures alone
- Progressive myoclonus epilepsies
- Temporal lobe epilepsy with hippocampal sclerosis
- Autosomal dominant focal epilepsy with auditory features
- Autosomal dominant nocturnal frontal lobe epilepsy

Childhood Onset Epilepsy

Genetic Epilepsy With Febrile Seizures Plus

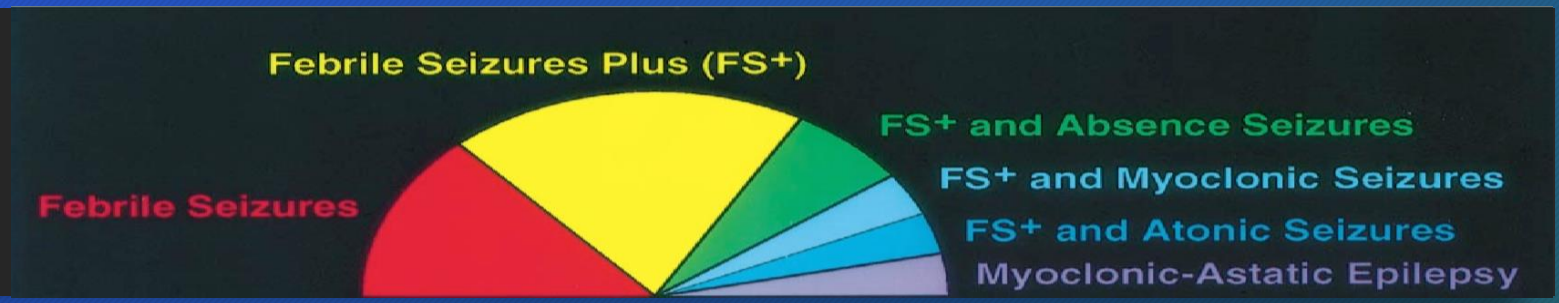
Genetic Epilepsy With Febrile Seizures Plus

- Familial epilepsy syndrome, affected individuals within a family typically have variety of epilepsy phenotypes, varying from simple FS and FS *plus*

- FS+ defined as either or both of the following:
  - FS extend beyond typical age range (before 3 mo or after 6 yr)
  - Occurrence of both febrile and afebrile GTCs

- GEFS+ family is defined as family with ≥ 2 individuals with GEFS+ phenotypes, at least one with FS or FS+

GEFS+



- Clinical spectrum
  - FS beyond age 6 (FS+)
  - FS+ with other seizure type: absence, myoclonic seizure, atonic seizure, focal seizure
  - Dravet and Doose syndromes may occur in GEFS+ family

Scheffer IE. Brain 1997;120:479-90

Genetic Epilepsy With Febrile Seizures Plus

- Mutations in *SCN1A* → Most common identified genetic cause of GEFS+ (19% of families)
- Other gene mutations: SCN1B channel, GABA-A receptor gene GABRG2
- EEG
  - Vary
  - May show irregular 2.5-4 Hz generalized spike and wave or PSW

Benign Epilepsy With Centrotemporal Spikes

Benign Rolandic Epilepsy (BRE)

- Nearly 25% of childhood epilepsies
- Age of onset is 4-11 yr, peak at age 7 to 8
- Normal cognition
- Seizure shortly after sleep onset or just before awakening
- Seizure during sleep in 80-90%, only while awake in <10%
- Infrequent and typically brief seizure (sec to min)

BRE: Seizure

- Major clinical findings
  - Orofacial motor signs
  - Speech arrest
  - Sialorrhea
  - Somatosensory symptom
- +/- Secondary generalization

EEG in BRE

- Normal BG
- High-voltage (100-300 microvolts), broad sharps, diphasic, centrotemporal region (rolandic spikes), often followed by slow wave
- Tangential dipole
  - Anterior positivity and centrotemporal negativity
- Potentiation during drowsy and non-REM sleep
- Bilateral independent or synchronous in ~1/3

BRE

- Presence of interictal spike discharges and frequency → deleterious impact on learning and memory??

- Pharmaceutical treatment only in some cases: CBZ, OXC, LEV, LTG, VPA
  - Very frequent events
  - Daytime events interfering with functioning
  - Secondary generalization

- Excellent outcome with long-term remission at age 14-16 yr

Myoclonic-Atonic Epilepsy

Doose syndrome

Myoclonic-Atonic Epilepsy

- Age onset: between 18 mo and 5 yrs of age, peak at 3 years

- Multiple seizure types
  - Myoclonus: large-amplitude symmetric jerks of arms, legs, neck, shoulders
  - Followed by loss of muscle tone and fall
  - Other seizures → absence, tonic-clonic, tonic, myoclonic, NCSE

- Typically normal cognition before onset of seizures (may maintain normal cognition)

Myoclonic-Atonic Epilepsy: EEG

- Normal BG, or BG slowing
- Recurrent paroxysms of generalized 2-3 Hz spike or polyspike and wave with admixed slow wave
- Parasagittal theta slowing
- Myoclonic events: bursts of 2-4 Hz epileptiform activity
- +/- Photoparoxysmal response
- +/- Nonconvulsive status epilepticus

Myoclonic-Atonic Epilepsy

- Prognosis: variable
  - Long-term remission in majority of patients
  - Some develop intractable epilepsy with intellectual impairment

Panayiotopoulos Syndrome

Panayiotopoulos Syndrome

- Early onset occipital seizure
- Onset: between 3 and 6 years of age (1-14 yrs)
- Normal physical and neuropsychological development

Panayiotopoulos Syndrome: Seizure

- Ictal emesis and other autonomic symptom
  - Pale, retching, cyanosis, mydriasis, miosis, incontinence, hypersalivation
- Eye deviation
- Impairment of consciousness 80-90%, usually intact at seizure onset
- Commonly end with hemiconvulsion, rarely with GTCs or SE
- Events usually long > 5 min, sometimes hours
- Episodes tend to be infrequent
- Long-term remission typically occurs within 2 years after onset

Panayiotopoulos Syndrome: EEG

- Occipital spikes bilateral, synchronous, often voltage asymmetry or unilateral
- High amplitude spike
- Occurred after eye closing, attenuated when eyes are opened
- Spikes can be shifting and multifocal

Late-onset Childhood Occipital Epilepsy (Gastaut Type)

Late-onset Childhood Occipital Epilepsy

- Peak between 8 and 11 year
- Visual seizure
  - Visual hallucinations, eye deviation/flutters, transient vision loss, ocular pain
- +/- secondary generalization and convulsive activity
- Ictal and postictal HA 30-50%: migraine like HA
- Seizures tend to be more frequent
- At least half experience long-term remission

Late-onset Childhood Occipital Epilepsy: EEG

- Classic appearance of bilateral occipital spike-and-wave discharges precipitated by eye closure and attenuated by eye opening

  (fixation off phenomenon)

Epilepsy With Myoclonic Absences (Tassinari Syndrome)

Epilepsy With Myoclonic Absences

- Characterized by very prominent rhythmic myoclonic or clonic activity during absence seizures
- Onset peaks at 7 years of age (1 to 12 years)
- GTCs and cognitive impairment associated with this syndrome

Etiology

- Family history of epilepsy is found in ~20% of cases
- 1/3 are idiopathic cases
- 2/3 related to symptomatic causes

Epilepsy With Myoclonic Absences: EEG

- Background EEG
  - Normal at onset
  - May deteriorate later
  - Abnormal in symptomatic patients
- Interictal: generalized spike wave and PSW
- Ictal: rhythmic generalized 3 Hz spike wave discharges of absence seizures
- Rhythmic myoclonic jerks of head, shoulders, and arms, time locked with spike-and-wave discharges

Epilepsy With Myoclonic Absences

- Tend to be pharmacoresistant
- Mental deterioration and evolution to LGS

Lennox-Gastaut Syndrome

Lennox-Gastaut Syndrome

- Multiple generalized seizure types, tonic being predominant
  - Core seizure: tonic, atonic, atypical absence
- EEG
  - Slow background
  - Slow (1.5-2.5 Hz) generalized synchronous spike-and-wave discharges
  - Paroxysmal fast activity during sleep, during tonic seizure
- Cognitive impairment

- *Onset is usually by age of 8 years, peak at 3-5 years*

Epileptic Encephalopathy With Continuous Spike and Wave in Slow Sleep

CSWS

- Electroclinical syndrome with epileptic encephalopathy
- Mixed generalized seizures, cognitive deterioration, EEG pattern of electrical status epilepticus in slow sleep (ESES)
- Seizures begin between 3 and 5 years of age

CSWS: EEG

- Usually slow diffuse (1.5-2.5 Hz) spike-wave discharges
- Traditional definition: spike-wave 85% of non-REM sleep
- Reducing percentage to ~50% for early diagnosis and therapy

CSWS

- Treatment usually attempted to improve EEG
- Epilepsy surgery may be indicated for resectable lesion
- Seizures typically remit by adolescence
- Neurocognitive sequelae persist

Acquired Epileptic Aphasia (Landau-Kleffner Syndrome)

LKS

- Acquired auditory agnosia as a presenting symptom, loss of understanding of previously familiar words or sounds
- Age of onset is typically 3-7 years (2 and 14 years)
- Language regression can be rapid or prolonged
- Attention and behavior problems and irritability are common

LKS

- Various infrequent seizure types, both generalized and focal
- ESES EEG predilection for perisylvian and posterior temporal
- Etiology
  - Unknown
  - Mutations of N-methyl-D-aspartate (NMDA) receptor subunit gene GRIN2A

Childhood Absence Epilepsy (Pyknolepsy)

CAE

- ~15% of childhood-onset epilepsy
- Typical age of onset between 4 and 10 yr (peak between 5 and 7 yr)
- Absence seizure:
  - Abrupt impairment of consciousness, behavioral arrest, staring, eye fluttering, automatisms
  - Immediate, complete recovery
  - Brief 10-30 seconds
  - Frequent, up to 100 times per day
  - Triggered by hyperventilation

EEG in CAE

- Normal background
- Bilaterally synchronous and symmetric regular 3 Hz spike-and-wave paroxysms of abrupt onset and offset
- Discharges tend to be frontal dominant
- A frequency range, especially at onset, of 3-4.5 Hz; and tend to slow down to 2-2.5 Hz by the end

EEG in CAE

- One hemisphere to show onset a few milliseconds before the other
- Fragmentary spike/PSW may be confined to one region, esp bifrontal
- Brief runs of spike-wave without clear clinical accompaniment
- Prominent runs of occipital intermittent rhythmic delta activity

CAE

- AEDs: ETX, VPA, with some role for CLZ and LTG
- Good outcome
- Seizure freedom reported in 57-74%

Generalized Epilepsy With Eyelid Myoclonia (Jeavons Syndrome)

Jeavons syndrome

- Onset childhood, peak 6-8 year
- Absence seizures with special features, eyelid myoclonia
- Brief absences (usually <10 sec) with prominent eye blinking and upward eye deviation, often triggered by eye closure
- Eyelid myoclonic and absence seizure may occur independently

Jeavons syndrome: EEG

- Ictal EEG: diffuse 3-6 Hz polyspike-and-wave complexes; may be precipitated by eye closure

- +/- generalized photoparoxysmal response

Jeavons syndrome

- AEDs used for primary generalized seizures (VPA, ETX, BZD)
- Prognosis is variable
  - Eyelid myoclonia can be frequent
  - GTCs may occur
  - Long-term remission is not the rule

Adolescent epilepsy syndrome

Adolescent to adult onset epilepsy syndrome

- Juvenile absence epilepsy
- Juvenile Myoclonic Epilepsy
- Epilepsy With Generalized Tonic-Clonic Seizures Alone
- Progressive Myoclonus Epilepsies
- Autosomal Dominant Nocturnal Frontal Lobe Epilepsy
- Autosomal Dominant Partial/Focal Epilepsy With Auditory Features
- Mesial Temporal Lobe Epilepsy With Hippocampal Sclerosis

Juvenile Absence Epilepsy

JAE

- Age of onset peaks at 15 years (10-19 years)
- Later onset than CAE without typical clustering
- More sporadic seizure and longer than in CAE
- Automatisms, speech arrest, loss of awareness
- +/- Myoclonic and GTCs (less prominent than JME)
- Triggers: sleep deprivation, alcohol, hyperventilation
- Typically pharmacoresponsive syndrome
- Lifelong requirement for medication

JAE EEG

- Paroxysms of generalized 3 Hz to 4 Hz spike- or polyspike-and-wave activity, faster at onset (4-5 Hz)

- Less organized, more fragmented spike wave than CAE

Juvenile Myoclonic Epilepsy

JME

- 5% to 10% of all epilepsy
- Onset is between 12 and 18 years of age
- Most constant clinical feature is myoclonic seizures
  - Predominantly involving upper extremities
  - Generally bilateral
  - Especially upon awakening
- Majority develop GTCs, usually shortly after awakening
- At least 1/3 experience absence seizures

JME: EEG

- Normal EEG background
- Abrupt paroxysmal generalized 4-6 Hz spike or PSW (fast spike and wave)
- +/- Focal spike
- Potentiated by sleep/sleep deprivation, alcohol use, menses, photic stimulation

JME

- Seizure freedom is achievable
- Life long treatment