Partial Epilepsies of Childhood

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Objectives

• Review most of the focal epilepsy syndromes in childhood and adolescence
• Review the electroencephalographic characteristics of these epilepsies
• Review treatment options.
Partial epilepsies of childhood

- Benign Neonatal Convulsions
- West Syndrome/Infantile spasms
- Otahara Syndrome
- Benign Rolandic Epilepsy (BECTS)
- Benign Occipital Epilepsy/Panayiatopolous Syndrome
- Dravet Syndrome
- Lennox Gastaut Syndrome (LGS) /LGs (spectrum)
- Sturge Weber Syndrome
- Hypothalamic hamartoma/Gelastic seizures
- Rasmussen Syndrome
- Landau Kleffner Syndrome
- Tuberous Sclerosis
- Neurofibromatosis
Benign Neonatal Convulsions

- Neonate or Infant
- Familial (chromosome 20) and non-familial (Fifth day fits)
- Otherwise healthy kids with no CNS risk factors or comorbidities
- Normal background EEG with bilateral independent mostly central spikes

Rolandic Epilepsy (BECTS)

- Age 3 to 15 years
- Semiology onset: sensorimotor oropharyngeal
- Not always benign
  - decrease school performance (30%)
    - ADD, impulsiveness, learning difficulties
    - auditory-verbal or visual spatial deficits
  - 20% Rx resistant, occasional SE.
- Normal background EEG with unilateral or bilateral independent centro-temporal spikes with shifting location, surface dipole and sleep activation
Rolandic Epilepsy (BECTS)

• Negative EEG predictors regarding behavior and cognition
  – Focal slowing
  – Abundance of wake and sleep epileptogenicity
  – Clusters of focal epileptiform discharges
  – Generalized 3 Hzs discharges
  – Paroxysmal negative or positive myoclonia.

Massa R. et al, NEUROLOGY 2001; 57: 1071-1079
Rolandic Epilepsy (BECTS)

- Treatment
  - Not always benign
  - Often associated cognitive problems when untreated
  - SUDEP?

- Oxcarbazepine, levetiracetam, pregabalin, topiramate, lamotrigine.

Massa R. et al, NEUROLOGY 2001; 57: 1071-1079
Panayiotopolous Syndrome

- Onset 3 to 6 years
- Prolonged episodes of mainly autonomic features (pallor, tachycardia, ictal emesis, eye deviation/nystagmus)
- EEG with multiple bilateral or unilateral independent epileptogenicity with occipital predominance
- Otherwise healthy children
- Usually outgrown in 2 years

Benign Occipital Epilepsy- Gastaut type

- Peak onset 8 to 11 (range 3 to 15 yrs)
- Otherwise healthy children
- Nystagmus, eye deviation or fluttering
- Blindness or occipital hallucinations
  - Elementary, multicolor, circular lasting 1 to 3 minutes (shorter than migraines).

Benign Occipital Epilepsy-Gastaut type

- Seizures are mainly diurnal and spontaneous but can be precipitated by TV, lights or videogames. They can be followed by severe headaches.
- EEG shows isolated occipital spikes or clusters activated by eye closure.
- Often persist into adulthood.
West Syndrome

- Infantile Spasms
- Developmental regression
- Hypsarrhythmia
  - First sleep “Hyps” them wake state
  - Multifocal high amplitude spikes
  - Disorganized background
  - Electrodecremental ictal events
Otahara Syndrome

- Neonatal or very early infancy onset
- Tonic spasms
  - 100 to 300 isolated spasms per day
  - 10 to 20 clusters per day
- Burst Suppression Pattern during wake and sleep cycles
  - Alternating periods of high amplitude sharp activity and periods of very attenuated or suppressed background

Otahara S, Yamatogi Y; Epilepsy Research 70 S (2006) S 58-S67
Otahara Syndrome

- Etiology usually structural abnormality and very rarely metabolic.
  - Lissencephaly, pachygyris, hemimegalencephaly, severe HIE.
- Treatment: ACTH, Zonisamide, Topiramate, Vigabatrin, VPA, ketogenic diet, benzodiazepines.

Otahara S, Yamatogi Y; Epilepsy Research 70 S (2006) S 58-S67
Dravet Syndrome

• “Severe myoclonic epilepsy in infancy”
• Typical and borderline (“spectrum”)
• Onset at age 1 year
  – Febrile or afebrile generalized or focal sz.
  – Myoclonic, atypical absence, partial szs.
• Normal cognition followed by developmental regression
• 70- to 80% of this phenotype has a SCN1A mutation

Dravet, C; Epilepsia 52 (suppl 2) 3-9, 2011
Dravet Syndrome

- Seventy to eighty % of the severe phenotype has a SCN1A mutation
- Mutations in PCDH19, the gene encoding the protocadherin 19 on the X chromosome, were discovered in some of the SCN1A-negative female patients presenting with a clinical picture resembling the borderline SMEI, which was described as “Epilepsy and mental retardation limited to females (EFMR)”

Dravet, C; Epilepsia 52 (suppl 2) 3-9, 2011
Dravet Syndrome

• Incidence 1.4% of epilepsies below 15 years.
• Worsened by drugs blocking sodium channels
  – Phenytoin, Carbamazepine, Lamotrigine, Oxcarbazepine
• Treatment
  – Clobazam, Valproic acid, Levetiracetam, Rufinamide, Topiramate, Bromides, Stiripentol, Vagal Nerve stimulation, ketogenic diet

Dravet, C; Epilepsia 52 (suppl 2) 3-9, 2011
Lennox- Gastaut Syndrome

• Epileptic encephalopathy
  – Same etiology as west syndrome, which often preceds LGS.

• Triad
  – Multiple seizure types including drop attacks (atonic or tonic in nature)
  – 1.5 to 2.5 spike and wave discharges
  – Cognitive impairment

• High Mortalitiy
  – Skull fractures (epidural, subdurals), Status epilepticus, SUDEP.

Arzimanoglu A et al, Lancet Neurol 2009; 8:82-93
Lennox- Gastaut Syndrome

• Treatment
  – Clobazam vs Rufinamide
  – Lamotrigine
  – Valproic acid
  – Felbamat
  – ACTH vs methylprednisolone
  – Ketogenic diet
  – Callsotomy
  – Vagal Nerve Stimulation

Arzimanoglu A et al, Lancet Neurol 2009; 8:82-93
Rasmussen’s encephalopathy

• Onset 2 to 14 years.
• Usually normal before onset.
• Partial seizures with normal or relatively normal EEG and normal MRI at onset.
• Evolution into Epilepsia partialis continue, hemiparesis, aphasia or dysarthria and progressive brain hemiatrophy by MRI.
Rasmussen’s encephalopathy

- Antiseizure medications fail to provide complete control.
- Immunomodulation fails to arrest progression. Palliative?
- Functional hemispherectomy.
Sturge Weber Syndrome

- Only one in six port-wine stains are associated with intracranial leptomeningeal hemangionatosis
- Intracranial disease associated with
  - Epilepsy
  - Glaucoma
  - Microinfarcts with local hemosiderosis and progressive loss of function
Sturge Weber Syndrome

• Treatment:
  – Anticonvulsants
  – Aspirin
  – Good hydration during acute illnesses/fever
  – Early surgery if refractory to medical treatment
    • Focal resection if not hemiparetic
    • Functional hemispherectomy if hemiparetic
Tuberous sclerosis

- Wide spectrum
- Clinical findings:
  - Hypomelanotic skin lesions
  - Facial angiofibromas
  - Shagreen patches
  - Subependimal hamartomas
  - Cortical tubers
  - Giant Cell astrocytomas
  - Lung, heart or kidney involvement
Tuberous sclerosis

- Treatment
- West syndrome:
  - Vigabatrin
  - ACTH, Methylprednisolone, Topiramate, VPA
- Partial seizures:
  - Oxcarbazepine, Levetiracetam, Pregabalin, Lamotrigine, Topiramate, Vigabatrin, Clobazam, KD, lesionectomies, VNS.
Other partial epilepsies

- Landau-Kleffner
- Gelastic seizures
- Neurofibromatosis