Epilepsy & Fits in Children

Dr. Toh Teck Hock
MBBS (Melb.), FRCPCH(UK), Dip Derm (Spore), AM (Mal), Cert. Com. Child Health (Melb)
Hospital Sibu Paediatric Refresher Course
9 April 2017

Definition - Seizure
- paroxysmal involuntary disturbance of brain function
- abnormal and excessive discharge of a set of neurons in the brain
- may be manifested as:
  - an impairment or LOC,
  - abnormal motor activity,
  - behavioural abnormalities,
  - sensory disturbances, or
  - autonomic dysfunction
- seizures do not constitute diagnosis

Definition - Epilepsy
- recurrent unprovoked seizures (convulsion / fits) unrelated to fever or to an acute cerebral insult
- Epileptic syndrome – a complex of signs and symptoms that define a unique epilepsy
- Syndromes – classified based on seizure type, clinical context, EEG features and neuroimaging

Epidemiology
- Seizures are common: 3-5 % of children
  - by age 20 – 4 to 5% of population
  - by age 80 – 8 to 10% of population
- Epilepsy
  - occurs in 0.5-1.0% of the population;
  - begins in childhood 60% of the time

Names
- Fit
- Convulsion
- Seizure
- Funny turn
- Spell
- Faint
- Epilepsy

Diagnostic Approach
- Is it an epileptic seizures?
- If epileptic, is it partial/focal) or generalized seizures? (seizure semiology)
- If recurrent seizures, is it idiopathic or symptomatic? (epileptic syndrome)

Most important tool is history and video
Semiology of the event

Description of seizures

- **Semiology of event**
  - **Aura**
    - epigastric discomfort
  - Pain
  - feeling of fear

Description of seizures

- **Semiology of event**
  - **Ictal phase**
    - Seizures (type and duration)
      - vocal – cry or gasp, slurring of words, garbled speech
      - motor – head or eye turning, eye deviation, posturing, jerking (rhythmic), stiffening, automatons purposeless repetitive movements
      - Symptoms during fits
        - respiration – change in breathing pattern, cessation of breathing, cyanosis
        - autonomic – pupillary dilatation, drooling, RR/HR changes, incontinence, pallor, vomiting
      - State of consciousness (impaired or retained)

Description of seizures

- **Semiology of event**
  - **Post-ictal state**
    - Amnesia for events
    - Confusion
    - Lethargy
    - Sleepiness
    - Headache and muscle aches
    - Transient focal weakness (Todd’s paresis)
    - Nausea or vomiting

Seizure Types

- **Partial / focal seizures**
  - Simple partial seizures
  - Complex partial seizures
  - Partial seizures with secondary generalization
- **Generalized seizures**
  - Not all seizures are “GTC”
  - Tonic – increase tone or rigidity
  - Atonic – flaccidity or movement-less during a fit
  - Clonic – rhythmic muscles contraction & relaxation
  - Myoclonus – shock-like contracture of a muscle
  - Absence

Dr. Toh Teck Hock, Sibu, 9 April 2017
Other Histories
- Assoc. features suggesting underlying cause:
  - developmental delay / intellectual impairment = degenerative CNS disease
  - family history
  - neurocutaneous signs / symptoms
  - constitutional symptoms (vomiting, FIT) = primary metabolic disease
  - abnormal behaviour = autism
- Define factors that may have provoked the seizure (e.g. fever, concurrent illness)
- Precipitating events other than illness - photic stimulation, trauma, toxins, sleep deprivation, environmental stimuli
- Time of occurrence (nocturnal / daytime)
- Frequency and last seizure

Epileptic or Non-epileptic?
- 13 months old boy
dystonic quadriplegic CP from severe perinatal asphyxia
- recurrent prolonged episodes of severe tonic spasm of limbs with arching of the back
- treated as seizures

Opisthotonus (Sandifer Syndrome)
- a tetanic spasm
  - spine and extremities are bent with convexity forward.
  - body resting on the head and the heels.
  - opistho– G. tonos, tension, stretching

Differential diagnosis – Paroxysmal Non-epileptic events
- Childhood
  - Benign paroxysmal vertigo
  - Breath holding spells
  - Tics and ritualistic movements
  - Paroxysmal choreoathetosis
    - movement disorder
    - episodes or attacks of involuntary movements of the limbs, trunk, and facial muscles.
  - Parasomnias
    - hypnagogic jerks, night terrors, sleep walking, head banging

- Childhood and Adolescence
  - Vasovagal syncope
  - Migraine
  - Panic attacks
  - Pseudo seizures
  - Day dreaming
  - Narcolepsy
    - periods of sudden extreme daytime sleepiness
    - +/- sudden loss of muscle tone (cataplexy), hallucination, sleep paralysis (can’t move or speak while waking up)

Examination
- ABC and resuscitation
- Search for underlying organic cause
- BP
- Growth parameters (HC, Ht, WI)
- Dysmorphic
- Neurocutaneous sign – café-au-lait spots, nevus flammeus, shagreen patch, adenoma sebaceum
- Hepatosplenomegaly – storage / metabolic disease
- Localizing neurologic sign – SOL in the brain
- Hyperventilation for 3-4 min -> absence

Investigation
- Electrolytes
  - FBC, BUSE, glucose, Ca, Mg
  - Not helpful if febrile convulsion
  - base on individual clinical circumstances such as vomiting, diarrhea, dehydration, failure to return to baseline alertness
- Chromosomal study & genetic tests
- Toxicology screen
  - drug exposure
  - substance abuse
- Lumbar puncture
  - limited value in first afebrile seizure
  - only considered when concern about meningitis / encephalitis
Investigation
- Brain imaging (CT, MRI)
- Epilepsy in infant, except febrile seizure
- Focal epilepsy except benign Rolandic epilepsy
- Developmental delay or regression
- Intractable epilepsy

Electroencephalography +/- video
- Support clinical diagnosis of epileptic seizures
- Classify epileptic syndrome – partial/generalized and specific syndrome
- Localisation of seizure foci in intractable case
- Selection of AED and rarely monitor AED
- Determine prognosis

Hypsarrhythmias (awake record)
West Syndrome
- Infantile Spasm + Hypsarrhythmias + developmental arrest / delay
- Treatment:
  - Vigabatrin
  - ACTH / Prednisolone
  - Benzodiazepine
  - Pyridoxine
  - Valproate
  - Topiramate

Infantile spasm – after treatment
Benign Rolandic Epilepsy (BECTS)
epilepsy of childhood with centrotemporal spikes
Benign Rolandic Epilepsy - Rx

- Nil if not > 2 fits or if the seizures > 6 months apart
- AED: Monotherapy with
  - VPA 20-30 mg/kg/day
  - CBZ CR / Clobazam o.n. for nocturnal seizure
  - CBZ may lead to status epilepticus / CSWS
- Wean off AED 1-2 years after seizure control, even when EEG has not normalized completely

Absence Epilepsy: 3/s spike wave complexes

- Hyperventilation
- Absence attack

Childhood Absence Epilepsy

- Onset: 4-8 years old
- Typical absence attacks
  - Frequent episodes that last for 5-15 s
  - Onset and termination abrupt
  - Do not have motor phenomena except eyelid flickering and mild change in muscle tone
  - Hyperventilation effective to precipitate attack
- May have GTCS in about 10-30% of cases
- Ictal EEG: 3 Hz generalized spike wave complexes
- Rx: VPA or/+ ETX, +LTG

Sodium Valproate – Side Effect

- Common
  - GI symptoms
  - Weight gain
  - Transient hair loss
  - Tremor
  - Behavioural problem
  - Thrombocytopenia
- Rare
  - Encephalopathy
  - Pancreatitis
  - Hepatic failure (1: 20,000)

Do not do EEG for...

- Syncope
  - Unless syncope with TC seizures
  - Cardiogenic syncope
    - Ask for exercise ECG and echo
- Febrile seizures
  - Even if complex or recurrent febrile convulsion
  - Paroxysmal non-epileptic events
    - If they are typical

Treatment

- The decision to treat must be based on a risk-benefit assessment
- Risk of another seizure against risk of chronic AED therapy
- Take into account both medical issues and patient and family preference
Medical therapy

Commonly used
• Sodium valproate
• Carbamazepine
• Phenobarbitone
• Phenytin
• Benzodiazepam group (clobazam, diazepam)

Less common
• Topiramate
• Lamotrigine
• Levetiracetam
• Vigabatrin
• Ethosuximide
• Oxacarbazepine
• Zonisamide
• Stripentol
• Febamate

Principles of Epilepsy Rx with AEDs

• Treat seizures, not EEG
• Start low and go slow
• Aim for monotherapy
• Push AED to clinical effect or high dose
• Monitor AED level (PHT, Phenobarb) (CBZ: useful)
• Monitor TWBC, Na+ if on high dose CBZ
• Combine AED with different mode of actions

Bonus Question:

A 3-year-old girl presents with a blistering rash on her face and body.

Differential Diagnoses?

- AED: Carbamazepine, Lamotrigine
- Other AEDs
- Medications, including NSAIDs, sulfonamides, and allopurinol

Drugs known to a/w SJS

Social Issues

- Parents and patient education
- Risks in daily life
  - Special home precautions for dangerous seizures (LGS, Drop attacks)
  - Drowning while bathing
  - Swimming allowed with 1 to 1 supervision
  - Cycling, car driving, rock climbing
- Need to balance btw acceptable risks and excessive protectionism

Parent education

- do not panic
- turn head down and lateral
- do not insert anything into the mouth
- time the duration
- do not try to restrain the seizure/ make sure the surrounding is safe
- prepare to bring to hospital or call ambulance if the seizure do not stop