Fits, faints, seizures and Epilepsy in childhood

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Statistics in developing countries

- Reliable incidence figures are harder
- Insurmountable logistical problems
- High incidence related to significant fraction of symptomatic epilepsy
- Infections eg. cysticercosis; perinatal complications
What is a seizure?

- Disturbance of brain function (disordered, excessive ‘electricity’)
- Paroxysmal; involuntary
- Different manifestations
  - loss/impairment of LOC
  - behavioural change
  - abnormal movements
  - odd sensations
“Non-seizure” - Paroxysmal events

- Not every paroxysmal event is a seizure
- Various events depending on age
- Examples (broad categories):
  - loss of consciousness
    (eg. Syncope/vasovag.)
  - motor/movements
    (eg. Shuddering att./tics)
  - respiratory events
    (eg. Breatholding/apnoea)
  - sleep related
    (eg. Hypnagogic/sleep terrors)
Episodic non-epileptic events *(infants and children)*

- Paroxysmal neonatal events
- Benign neonatal myoclonus
- Breath-holding spells
  - blue
  - pale
- Shuddering attacks
- Infantile gratification disorder; etc.
Benign neonatal sleep myoclonus

- Only during sleep
- Stops on awakening
- Face is spared
- Seizures do not awaken baby
- Normal EEG / normal development
Shuddering attacks

- Triggered by excitement / agitation mostly
- Short duration (seconds)
- Affect body, limbs and head
- No loss of consciousness
- No colour change
- Normal examination / EEG
Sequelae of misdiagnosis

- Labeling as “epileptic”/ stigma
- Drug treatment:
  - unnecessary
  - potentially harmful
  - costly
  - inconvenience
- Psycho-social
Diagnosis of a seizure

- History-based
- Parents or other witness
- EEG test
- Brain scan
Home videos / cell-phones

☐ Better seen than described

☐ Invaluable

☐ Is it a seizure?

☐ What kind of seizure?
Classification

SEIZURE TYPES

- Generalised – ‘major’ vs ‘minor’
- Focal – simple vs complex
Generalised seizures

- GTCS
- Tonic
- Atonic
- Myoclonic
- Absences
- ‘Drop’-attacks
Focal seizures

- **Simple** focal
  - eg. Motor (+/- jacksonian march)

- **Complex** focal / CPS

- With/without **secondary generalization**
Difference between fits and epilepsy

- One seizure does not make epilepsy
- Epilepsy is not equivalent to just many seizures
EPILEPSY

2 or more unprovoked seizures

A tendency for unprovoked recurring seizures
*(Simple)* Febrile seizures

- 6 months -> 6 years
- Febrile illness (source outside CNS)
- Generalized seizure
- Brief (<10-15 min.)
- Not recurrent during same febrile illness
- Well in-between
- Development not affected
Classification (of Epilepsies)

- Seizure type

- Cause can be:
  - Unknown/genetic
  - Symptomatic

- Age

- Epileptic syndrome

- ‘Good’ or ‘bad’ epilepsy
  (not mutually exclusive; all may apply!!!)
“Good” and “Bad” epilepsies

- Cause
- Development
- Response to treatment
- Remission
- Outcome
Epileptic encephalopathies in the neonate

- OHTAHARA SYNDROME
- EARLY MYOCLONIC ENCEPHALOPATHY

(with *BURST SUPPRESSION* on EEG)
Infantile spasms

- Often mistaken as ‘colic’
- Flexor or extensor
- Usually associated with a cry / upsetting
- Mostly on awakening
- Come in clusters
- NB: severe effects on development; ‘EMERGENCY’
Infantile spasms: treatment

- Investigate: cause?
- (TB screening)
- ACTH 20u, imi daily (10 days)- @ RXH
- Vigabatrin
- Valproate
- Clonazepam
- Developmental follow-up
Childhood onset

- Childhood absence epilepsy
- Benign Rolandic epilepsy
- Lennox-Gastaut Syndrome
- Landau-Kleffner / Acq. Epileptiform aphasia
Absence seizures “petit mal”

- Very common between 5-8 years of age
- Sudden onset; motion arrest
- Loss of consciousness
- Staring, eye blinking
- Duration: seconds
- Abrupt cessation
- Easy to treat (usually)
- Usually resolve in adolescence
“Staring spells”

- Daydreaming (inattention)
- Absence seizures
- Complex partial seizures
Lennox-Gastaut Syndrome

- Severe epilepsy
- Multi-seizure types
- Developmental delay/regression
- Can evolve from infantile spasms
- Many causes
- Very abnormal EEG
- Often different kinds of medication required
- Prognosis not great
Things to remember

- Epilepsy rarely comes alone
- Common to have additional disabilities
- Learning difficulties / Dev delay
- Behavioural problems / ADHD
- Focal neurology
- **Impact on quality of life often not the actual seizures but the way it changes the child’s (and family’s) lifestyle and future**
ADHD - Risk in Epilepsy

- Boys
- Family history
- Intellectual Disability /LD
- Frontal lobe disease
- Treatment resistant epilepsy
- Drugs eg. Phenobarbitone, benzodiazepines
Adolescent issues & Transition to adulthood

- Seizure frequency
- Compliance
- Medication “handling” (hormones)
- Contraceptives
- Independence
- School-leaving / employment
- Driving
- Late nights
- Experimenting with drugs/alcohol
Management Issues

- Investigations
- Counselling/education
- Treatment
When to request an EEG.

- After 2 or more seizures
- Urgent EEG - *Sub-clinical status* - *Infantile spasms*
- Awake studies OK in most
- Absence epilepsy – need to be awake; for HV
- Sleep studies for difficult cases
  - CPS
  - Myoclonic
  - Normal awake
- NB normal study does not exclude epilepsy
When to refer for neuroimaging

- Focal seizures
- Focal neurology
- Reduced GCS (not recovering)
- *(i.e. Concern of SOL, cerebral malformation, infective focus, etc)*
Who to tell?

- Family
- Teachers
- Carers
General principles

- **Education** and support of parents
  - diagnosis uncertainty
  - what to do during a seizure?
  - is my child going to die?

- Address educational, learning and behaviour problems (cormorbidities)

- Take into account myths, prejudices, misunderstandings to inform an adapted yet full explanation
Modalities

- Drug vs non-drug treatment
- Conventional AE drugs vs non-conventional treatments
Phenobarb (oral)

- Problems with ADHD
- Learning difficulties
- Useful in neonates, CP
**carbamazepine**

- Focal seizures
- Aggravates myoclonic
- Sedation
- Cognitive disturbances
- Rash
- Slow titration – start @5mg/kg/day (max. 20mg/kg)
- Controlled release prep’s better
valproate

- Mainly generalised seizures
- Especially useful for mixed seizure types
- Usually 1st line in syndromic epilepsies
- Does not aggravate other seizures
- Remember hepatotoxicity, esp in v.young
- Increased appetite and weight-gain
topiramate

- Partial and generalised
- 2nd line (*usually add-on*)
- Useful in syndromes (eg LGS)
- Cognitive s/e; *somnolence* at higher doses; wt.loss; urolithiasis (CAI)
- Titrate very slowly
- 0.5 – 10mg/kg/day (aver.: 1-5mg/kg)
**lamotrigine**

- Spectrum of activity similar to epilim
- Generalised and partial sz
- Can exacerbate myoclonic sz
- “Start small -> go slow”
- Initial dose 0.25-0.5mg/kg/day
- Can give daily or bd
- Max. dose with valproate is 5mg/kg/day
- RASH!!
levetiracetam

- Broad spectrum
- Twice daily regimen
- Minimal side effects
- ‘novel’ mechanism of action
- Start @10-20mg/kg up to 40-60mg/kg
- Can make some children “nasty”- *pyridoxine*
Outgrowing it / Stopping medicines
Discontinuation

- About 70% of children with epilepsy who become seizure free for 1-2 years can successfully stop medication
  
  *(Berg and Shinnar’1991/4; Dooley et al., 1982)*

- No > benefit if meds continued for 5 years

- If failed weaning – about 50% become Sz-free after restarting Rx with 70% success
  
  *(Hollowach-Thurston et al, 1982)*

- With JME 75 – 100% relapse = life-long Rx
  
  *(Delgado-Escueta et al. 1984; Penry et al. 1989)*
Predictors - successful discontinuation

- Generalised seizures
- Benign rolandic epilepsy
- Age at onset < 10 years
- Normal neurological status

(Dooley, 1996)

- (+/- resolution of interictal EEG spikes)

All good features = 90% success
How to wean??

☐ How long?
☐ How fast to taper?
☐ In polypharmacy – greater caution?
☐ Is there a major advantage in a prolonged (> 6 weeks) taper?

(Tennison, 1994; Todt, 1984)
Intractability

“Failure, for lack of seizure control, of > 2 first-line anti-epileptic drugs with an average of >1 seizure/month for 18 months and no more than 3 consecutive months seizure-free interval”

(Berg et al. 2001)

Risk factors point towards intractability but cannot be used for individual children to make management decisions.
Risk factors - \textit{intractability}

- Poor initial response to medication
- Remote symptomatic aetiology
- Status epilepticus (esp. neonatal)
- IGE – generally do well
  - 613 children; Connecticut
  \textit{(Berg and Shinnar et al. 2001)}

- Intractability tends to decrease with prolonged follow-up
  \textit{(Huttenlocker and Hapke; 1990)}
## Alternatives

### Table 1 Non-conventional antiepileptic drug (AED) treatment of epilepsy

<table>
<thead>
<tr>
<th>Non-AED medical treatment</th>
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<tbody>
<tr>
<td>Steroids (for example, ACTH [tetacosactide], prednisolone)</td>
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<tr>
<td>Intravenous immunoglobulins</td>
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<td>Vitamins (for example, pyridoxine, pyridoxal phosphate, biotin, folinic acid)</td>
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<tr>
<td>Melatonin</td>
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<tr>
<td>Dietary manipulation</td>
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<tr>
<td>Ketogenic diet</td>
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<tr>
<td>Classical ketogenic diet</td>
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<tr>
<td>MCT diet</td>
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<tr>
<td>Atkins diet</td>
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<tr>
<td>Oligoamnogenic diet</td>
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<tr>
<td>Epilepsy surgery techniques</td>
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<tr>
<td>Lesional surgery (for example, tumour, amygdalo-hippocampectomy, temporal lobectomy, extra-temporal resections, anatomical hemispherectomy or functional hemispherectomy, removal of cortical seizure foci)</td>
</tr>
<tr>
<td>Specific surgical techniques (for example, sub-pial transection for Landau-Kleffner syndrome)</td>
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<tr>
<td>Palliative surgery (for example, callosotomy or vagus nerve stimulator implantation)</td>
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</tbody>
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**ACTH**, adrenocorticotropic hormone; **MCT**, medium chain triglyceride.

### Table 2 Non-pharmacological treatment of epilepsy

<table>
<thead>
<tr>
<th>Lifestyle changes</th>
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</thead>
<tbody>
<tr>
<td>Exercise</td>
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<tr>
<td>Avoidance of sleep deprivation</td>
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<tr>
<td>Avoidance of excessive alcohol consumption</td>
</tr>
<tr>
<td>Psychological approaches</td>
</tr>
<tr>
<td>Techniques to abort seizures or reduce seizure frequency (for example, avoidance, relaxation, biofeedback, aversive therapy)</td>
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<tr>
<td>Promotion of emotional wellbeing (for example, yoga)</td>
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<tr>
<td>Reduction of psychiatric co-morbidity (for example, anxiety or depression)</td>
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<tr>
<td>Coping strategies for living with epilepsy (for example, CBT, counselling, psychotherapy, educational interventions)</td>
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<tr>
<td>Alternative therapy</td>
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<tr>
<td>Herbal medicine</td>
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<tr>
<td>Homeopathy</td>
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<tr>
<td>Others</td>
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<tr>
<td>Aromatherapy</td>
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<tr>
<td>Hypnosis</td>
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<tr>
<td>Acupuncture</td>
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<tr>
<td>Seizure alert dogs</td>
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</tbody>
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CBT, cognitive behaviour therapy.
Other practical aspects

- Seizure diaries
- Emergency care plans/Hospitalisation
- Follow up care
- Immunisations
Social grants

School placement

Sports, leisure

Respite
Outcome / prognosis

- **Psychosocial** (Camfield et al. 1993 – Normal I.Q.)
  - school failure (34%)
  - use of special education resources (34%)
  - mental health consultation (22%)
  - psychotropic medication (5%)
  - unemployment (20%)
  - social isolation (27%)
  - criminal conviction (2%)

- **Death** – SUDEP
  (Sudden Unexplained Death in Epilepsy)
Mortality

- Childhood death is still relatively rare
- Death rate is higher in those with serious neurological handicap
  
  *(Camfield, 2002; Callenbach, 2001; Lhatoo, 2001)*

- SUDEP – occurs in children with epilepsy, but very low risk *(Donner, 2001)*
Support groups and local resources

- Epilepsy SA
- Cape Mental Health
- Other support groups (CP association)
- Community Organisations
Goal

- Educate
- Support
- Nurture

- Make them the BEST they can be
The sky is the limit !!!

THANK YOU !!!