Paediatric Epilepsy Update 2018

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Epilepsy Service CUH

- ~550 children
- New diagnosis-education, support, clinic follow up
- Epilepsy phoneline service
Aims of Paediatric Neurology service

- Accurate diagnosis
- As well controlled epilepsy as possible
- No side effects
- Well managed co-morbidities
- Good quality of life for the child and family
- Reach maximum developmental potential
- Become well adjusted adults
What is epilepsy

- A chronic condition in which a person has a tendency to have recurrent, unprovoked seizures; normally diagnosed after 2 or more seizures >24 hours apart.
- An epileptic seizure is an intermittent, stereotyped disturbance of behaviour, emotion, motor function or sensation resulting from abnormal cortical neuronal discharges.
- Epilepsy is the tendency to have recurrent epileptic seizures.
Seizure type

- Two patterns of epileptic discharges recognised:
  - **Generalised epilepsy** – discharges arising simultaneously from both hemispheres
  - **Focal epilepsy** – discharges arising from a focal cortical disturbance- these can evolve to generalised seizure
Terms no longer in use

- Grand Mal, Petit Mal
- Complex partial
- Simple partial
- Partial
- Dyscognitive
- Secondarily generalized tonic-clonic
ILAE 2017 Classification of Seizure Types Basic Version

- **Focal Onset**
  - Aware
  - Impaired Awareness
  - Motor Onset
  - Nonmotor Onset
  - Focal to bilateral tonic–clonic

- **Generalized Onset**
  - Motor
    - Tonic–clonic
    - Other motor
  - Nonmotor (Absence)

- **Unknown Onset**
  - Motor
    - Tonic–clonic
    - Other motor
  - Nonmotor
  - Unclassified

1. ILAE classification
2. Unclassified
Causes of Epilepsy

Causes:

- Neurological Infections
- Stroke
- Head Injuries
- Genetic Basis
- Defects in Brain Development
- Brain Tumours
- Brain Damage from Prenatal or Perinatal Causes
- Unknown in Many Cases

Contagious in Many Cases

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https://www.facebook.com/wfneurology
Diagnosis

- Clinical history.. **Eyewitness account**
  An incorrect diagnosis of epilepsy is made in 20-30% of cases; & up to 40% of children referred to a tertiary clinic do not have epilepsy (JEC, 2011).
- NICE Guidelines 2014
- video
- EEG
- ECG
- MRI
- Bloods
Diagnostic difficulties

- Children are prone to non epileptic paroxysmal events causing diagnostic confusion
- Every epileptic seizure type has a non epileptic differential
- 30% of individuals referred to a specialist clinic did not have epilepsy
- Concordance among neurologists for epileptic events is high but there is a tendency to overcall as epileptic non epileptic events
Why treat epilepsy?

- Improve Quality of Life
- Reduce morbidity/mortality
- To maximise academic potential
Status epilepticus

- One of the most common neurological emergencies affecting children worldwide
- Regarded as medical emergencies requiring immediate seizure control measures
- Untreated seizures can progress to status epilepticus (SE)
- SE carries increased risk of morbidity and mortality (case fatality in children, 2.7–8%)
- Early treatment is imperative
Treatment options

- Antiepileptic Medication
- Ketogenic Diet
- VNS therapy
- Resective surgery
<table>
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<tr>
<th>Topic</th>
<th>Description</th>
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<tr>
<td>Focal versus generalised specific medication</td>
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<td>Goal of treatment- no seizures with no/least side effects possible.</td>
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<td>Up to 70% of people with epilepsy respond to 1&lt;sup&gt;st&lt;/sup&gt; or 2&lt;sup&gt;nd&lt;/sup&gt; AED.</td>
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<td>15-35% of all children will have medically intractable epilepsy.</td>
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<td>Monotherapy preferred; side effects.</td>
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<td>Practical issues.</td>
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Traditional versus new antiepileptic medication

- New generation vigabatrin, lamotrigine, pregabalin,
- Side effect profile improving with newer AED
- Genetic epilepsies- drug specific AED therapy

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<th>Traditional AEDs</th>
<th>New AEDs</th>
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<tr>
<td>phenytoin</td>
<td>Lacosamide</td>
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<td>phenobarb</td>
<td>Zonisamide</td>
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<tr>
<td>Sodium Valproate</td>
<td>Lamotrigine</td>
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<td>Benzodiazepines</td>
<td>Levetiracetam</td>
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Ketogenic Diet

- Ketogenic diet-high fat, low carbohydrate; considered in those where AEDs fail.
- GLUT-1 deficiency- first line treatment
- Changes the metabolism in the body from burning glucose for energy to burning fat.
- Dietician/expert led, requires monitoring blood ketones and sugars
Case study

- Patient CO’B- 6 year old girl, hx infantile spasms & tuberous sclerosis, refractory epilepsy, 10 AEDs,
- Encephalopathic EEG 2014
- Commenced Ketogenic Diet Jan ‘17, 2 provoked seizures since c/o diet.
- Making developmental progress
- EEG June 2017 - much improved background, still focal discharges.
- No hospital admissions since c/o diet
Tuberous sclerosis
Vagal Nerve Stimulator

- **Vagal Nerve Stimulator** - used in those resistant to AEDs, for whom surgery is not an option.
- Device implanted below collar bone; connected to vagus nerve, sends inhibitory impulses to desynchronise seizure activity in the brain.
- Devices are externally programmed- dedicated VNS clinic- patients seen every 3 months
- VNS therapy takes time, may take up to 2 years to see the full benefits
VNS
Case Study

- TR - 17 year old boy, seizures since 8/12 old; developmental regression, tried ketogenic diet with little response; 9 AEDs, profound intellectual disability, multiple seizure types
- VNS 2005, battery replaced 2009, 2014,
- Parents report reduction in drop attacks, GTCs, improvement in behaviour, seizures persist but improved
Advances in VNS therapy

- **AspireSR** detects seizures by detecting pre-ictal tachycardia and provides automatic stimulation to stop or shorten a seizure and improve post-ictal recovery time
- Earlier activation reduces seizure duration and seizure severity
- Detects seizures in sleep
Resective Surgery

- **Surgery** - if AEDs fail; & focus of seizure can be localised, consideration for surgery. Extensive work up for surgery, need to be sure that removal of the offending area will not cause further problems; quality of life will improve for the child, acceptable risk: benefit ratio.
MD 15 yr old boy
Hx infantile spasm @ 7/12 old, TLE 2007;
Video telemetry 03/12-sharp waves left temporal lobe
MRI confirmed temporal lobe gliosis
Refractory epilepsy
Resection of left temporal ganglioma march 2015
Seizure free since 2015