PAROXYSMAL NON-EPILEPTIC EVENTS

Khoo Teik Beng
Paediatric Institute
Hospital Kuala Lumpur
Epileptic seizures

• Transient clinical events that result from the abnormal, excessive activity of a set of cerebral neurons.

• May give rise to a positive or negative phenomena

Aicardi’s  Epilepsy in Children 2004
Paroxysmal Non-epileptic Events

• Physiological or exaggerated physiological responses, parasomnias, movement disorders, and behavioural or psychiatric disturbances that may mimic epileptic seizures
Paroxysmal Non-epileptic Events

- Infancy
  - Jitteriness
  - Neonatal sleep myoclonus (video deleted)
  - Sandifer syndrome (video deleted)
  - Benign paroxysmal torticollis
  - Shuddering attacks (video deleted)
  - Hyperekplexia
Neonatal Sleep Myoclonus

• Rhythmical jerks of the limbs during sleep
• Generalized or focal
• Occur in brief or more prolonged bursts
• Trunk and face are unaffected
• Jerks immediately cease on awakening
• Usually disappears in a few weeks but it can persist up to several months
The infant with arches and jerks

- 13 months old boy

- Dystonic quadriplegic CP from severe perinatal asphyxia.
- Had recurrent prolonged episodes of severe tonic spasm of limbs with arching of the back

- Previously treated as seizures.

- Diagnosis: Sandifer syndrome
Sandifer Syndrome

• Sudden extension of the neck in an opisthotonotic position
• Associated with severe GORD or hiatus hernia
• Effective treatment of GORD or hiatus hernia leads to complete resolution of the problem
Infants with opisthotonus

Other Differential Diagnoses

- Tetanus
- Dystonic CP
- Neurotransmitter disorders (L AADC, Dopa –responsive dystonia)
- Lysosomal disorders (Krabbe, Neuropathic Gaucher’s disease)
Benign Paroxysmal Torticollis of Infancy

- Appears before 1 year of age
- Attacks often last several hours and up to 2-3 days
- Associated with repeated vomiting, discomfort, tilting of head to one side, and eye movements
- The affected side may change with successive attacks
- In some, typical migraine will follow in later childhood
Hyperekplexia

- Excessive startle response to auditory, visual and light touch (nose tapping)
- Infants may present with marked hypertonia from birth (stiff baby syndrome) which disappears during sleep
- Episodes of prolonged apnoea can be lethal
- AD / AR, mutation in glycine receptor
- Rx: Clonazepam and valproate
Paroxysmal Non-epileptic Events

• Childhood
  – Benign paroxysmal vertigo
  – Paroxysmal tonic upgaze (video deleted)
  – Breath holding spells (video deleted)
  – Tics and ritualistic movements (video deleted)
  – Self gratification (video deleted)
  – Paroxysmal choreoathetosis
  – Parasomnias
Paroxysmal Tonic Upgaze

- Attacks of tonic upgaze sometimes a/w forward flexion of head
- Worsen with tiredness and febrile illness
- Relieved by sleep
- Few may respond to L-dopa
- ½ have good outcome, ½ have ataxia, borderline cognitive impairment
Breath-holding Spells

- Occurs in 4% of all children below 5 years old
- Cyanotic and pallid types
- Attacks often provoked by fright, pain, anger, frustration or minor fall
- Cries vigorously then holds the breath in expiration
- Benign condition
- Reassurance
Benign Paroxysmal Vertigo

• Recurrent attacks of vertigo between 1 - 5 years old
• The attacks are brief, lasting from one to several minutes
• The child appears in distress, pale but conscious, may stagger or fall during the attacks. Nystagmus may be noted
• Frequency between one to few per month
• Most of them cease by 5-6 years old
Self Gratification

- Onset: 3 months to 3 years;
- Stereotyped episodes often with pressure on the perinium and posturing of the lower limbs
- No alteration of consciousness
- Facial flushing, grunting or sweating
- Cessation with distraction
- Normal examination and investigations
Paroxysmal Kinesiogenic Choreathetosis

• Unilateral or bilateral attacks of dystonic movements or chorea that are precipitated by sudden movement
• Attacks usually last one to a few minutes
• May occur up to 100 times daily
• Consciousness is preserved but bizarre, often writhing movements, may result in falls
• AD / Sporadic
• Rx: Carbamazepine / Phenytoin
Parasomnias

- Night Terror
- Nightmares
- Sleep walking
- Restless leg syndrome

- Need to differentiate from nocturnal seizures especially Frontal Lobe Epilepsy
## Nocturnal FLE vs Parasomnias

<table>
<thead>
<tr>
<th></th>
<th>Nocturnal FLE</th>
<th>Parasomnias</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset</td>
<td>14 ± 10 yr</td>
<td>Usually &lt; 10 yr</td>
</tr>
<tr>
<td>Frequency</td>
<td>20 ± 11 / month 3 ± 3 / night</td>
<td>From &lt;1 to 4 / month 1 /night</td>
</tr>
<tr>
<td>Time of onset</td>
<td>Throughout the night</td>
<td>First third of night</td>
</tr>
<tr>
<td>Movement</td>
<td>Violent, stereotyped</td>
<td>Non-stereotyped</td>
</tr>
<tr>
<td>Duration</td>
<td>Brief</td>
<td>More Prolonged</td>
</tr>
<tr>
<td>EEG</td>
<td>Abnormal</td>
<td>Normal</td>
</tr>
</tbody>
</table>
Paroxysmal Non-epileptic Events

- Childhood and Adolescence
  - Vasovagal syncope (video deleted)
  - Migraine and related disorder
  - Narcolepsy (video deleted)
  - Panic attacks
  - Rage attacks
  - Non-epileptic attack disorders (video deleted)
  - (Pseudoseizures)
Vasovagal Syncope

• Stimulus / setting
• Usually upright
• Giddiness, pallor and sweating
• Gradual onset, ‘fall like a log’
• Injury rare
• May have clonic movements, incontinence
• Brief with no confusion afterwards
Cardiac Syncope

- Due to structural heart disease (AS) or arrhythmias (Long Q-T syndrome)
- Occurs during exercise or supine position
- Atonic, motionless then jerks
- Family history of sudden death
- Diagnosis – 12 leads ECG, Holter, Echo
<table>
<thead>
<tr>
<th>Clinical Feature</th>
<th>Epileptic seizures</th>
<th>Non-epileptic attack disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Sudden</td>
<td>Gradual</td>
</tr>
<tr>
<td>Incontinence</td>
<td>Infrequent</td>
<td>Rare</td>
</tr>
<tr>
<td>Biting</td>
<td>Tongue</td>
<td>Lips</td>
</tr>
<tr>
<td>Scream</td>
<td>At onset</td>
<td>During spell</td>
</tr>
<tr>
<td>Convulsion</td>
<td>Tonic / clonic</td>
<td>Bizarre, thrashing, pelvic thrust</td>
</tr>
<tr>
<td>Pupillary reflex</td>
<td>Slow, non-reactive</td>
<td>Normal</td>
</tr>
<tr>
<td>Effect of suggestion</td>
<td>No effect</td>
<td>Precipitate or terminate</td>
</tr>
<tr>
<td>Post-ictal Stupor</td>
<td>Frequent</td>
<td>Rare</td>
</tr>
<tr>
<td>Presence of others</td>
<td>Variable</td>
<td>Yes</td>
</tr>
<tr>
<td>During sleep</td>
<td>Yes</td>
<td>Rare</td>
</tr>
</tbody>
</table>
# Paroxysmal Non-epileptic Events (IPHKL, 2006)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Frequency</th>
<th>Description</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tics / Tourette</td>
<td>10</td>
<td>Shuddering attack</td>
<td>1</td>
</tr>
<tr>
<td>Syncopy (TRO Epilepsy)</td>
<td>6</td>
<td>Paroxysmal tonic upgaze</td>
<td>1</td>
</tr>
<tr>
<td>PKC / PKD</td>
<td>4</td>
<td>Head banging</td>
<td>1</td>
</tr>
<tr>
<td>Abdominal myoclonus</td>
<td>3</td>
<td>Sandifer syndrome</td>
<td>1</td>
</tr>
<tr>
<td>Self gratification</td>
<td>2</td>
<td>Non-epileptic attack disorder</td>
<td>1</td>
</tr>
</tbody>
</table>
Making a Diagnosis

- History from child
- Eye witness account
- Replication
- Home Video
Video Telemetry

- Video + simultaneous EEG and ECG
- Able to characterise clinical and electrical events
- Useful for ambiguous cases and paroxysmal neonatal events
Not all Fits, Faint and Funny Turns are Epileptic Seizures!

Thank You for your attention