Paroxysmal Non-Epileptic Disorders

- What makes a fit, a faint or funny turn an epileptic seizures?
- There are a host of paroxysmal conditions that can simulate seizures in children
- Over diagnosis of epilepsy is common - 20 – 30% of patients sent to a specialised epileptic clinic do not have epileptic seizures

Pseudo-epileptic seizures

How does one differentiate between - true seizures and pseudo-seizures?

Pseudo-seizures

- Episodes usually preceded by dizziness, funny feelings, numbness
- The movements are not true clonus but a spectrum of movements – from quivering to flailing of the limbs – and usually coordinated and bilaterally symmetrical

Pseudo-seizures

- Onset is usually not sudden – but gradually builds up to a paroxysm
- Activity may be “theatrical” with loud screaming and shouting
- Post-ictal state is very uncommon
- Never occur during sleep and unlikely when alone
- Beware of Munchausen by proxy syndrome

Paroxysmal Non-Epileptic Disorders

1. Anoxic – ischaemic
2. Migraine
3. Psychological/psychogenic
4. Sleep disorders
5. Movement disorders

Paroxysmal Non-Epileptic Disorders

- Most diagnostic errors can be avoided by a good and careful HISTORY:
  - Subjective sensations before (light-headedness)
  - Character of the seizure “mimic”
  - Videos & cell phones – great help
  - Post-ictal phenomena
- It is preferable to defer the diagnosis of epilepsy until the evidence is sufficiently clear

Paroxysmal Non-Epileptic Disorders

1. Anoxic – ischaemic
   - Syncope
   - Breath holding
   - Apnoeic spells
1. Anoxic/ischaemic

**Syncopes** (“faints & funny turns”)

- Syncopes - defined as a sudden loss of consciousness & postural tone due to transient cerebral hypoperfusion (< 30ml/100g brain)
- Cardiovascular, neuro-cardiogenic (vasovagal), convulsive, reflex, situational, drug induced, psychogenic, orthostatic hypotension
- A brief tonic, clonic or myoclonic seizure may accompany syncope – these are not indicative of an epileptic seizure

2. Situational syncope

- Cough, sneeze, swallowing, micturition, defecation, hair brushing, diving, weight lifting
- Common denominator is that most triggers are accompanied by a Valsalva maneuver

3. Reflex anoxic seizures

- Non-epileptic events resulting from cardiac asystole of vagal origin
- Often provoked by pain & fright (surprise) – blow to the head, veni-puncture (?) Prolonged pallid breath holding
- Loss of muscle tone → tonic posturing, may see a few myoclonic jerks at beginning and end of event

2. Breath holding – Cyanotic & Pallid

**Cyanotic**

- Typically between 6-18 months
- Precipitated by anger, frustration
- Cries vigorously – becomes still – holds breath in expiration → cyanosis
- May be associated with limpness, opisthotonus or clonic jerks
- Pathophysiology is complex – autonomic dysregulation – sympathetic & parasympathetic excess
- Check for iron deficiency

**Pallid breath holding spells**

- Induced by minor trauma (usually to the head), may be a very brief cry, followed by loss of postural tone and loss of consciousness - pallor
- Pulse may be slow and “thready” during episode
- Regain consciousness within a minute, but may sleep for a few hours thereafter
- Occasionally clonic movements and incontinence of urine
- Thought to be due to excessive vagal tone → cerebral hypoperfusion
- Ocular compression test
- ECG → Long QT syndrome
- Rx = Atropine 0.01mg/Kg BD/TDS
  = Piracetam (nootropil)
2. Psychological / psychogenic / psychiatric

- Conversion reaction
- Malingering
- "Panic attack" – Often accompanied by episodic flushing, sweating, breathing difficulties, tachycardia, palpitations, feelings of fear and doom. Epigastric or laryngeal sensations of pressure.
- Fugues – dissociative reactions (childhood neuroses)
- Confusional states – hypoglycaemia
- Hyperventilation syndrome – adolescent girls

2. Psychological/psychogenic

- Self gratification syndrome
  - Repetitive stereotypes
  - Body rocking, head banging, thighs tightly adducted or crossed
  - Pelvic thrusting, contractions of the gluteal muscles
  - Sweating, flushing, of face
  - May terminate in fatigue, exhaustion or sleep
  - Incredulous parents
  - Rx = reassurance

3. Migraine vs. epilepsy

Complicated and controversial

1. Alice in Wonderland – Distortions of perception, change in size or shape of body parts, micropsia, distortions of surroundings.
   Confused with hallucinations as seen in TLE
   Throbbing headache & normal consciousness favour migraine

2. Acute confusional migraine – acute confusion, blindness, paraesthesias, hemiplegia and amnesia

3. Basilar artery migraine – intense dizziness, vertigo, visual disturbances, ataxia and diplopia
   May see brief loss of consciousness, prolonged confusion, transient amnesia, tinnitus, N & V, visual field defects, hearing loss, dysarthria, weakness

4. Hemiplegic migraine

5. Periodic syndromes

There is a higher incidence of epilepsy in migraine sufferers

EEG commonly shows slow waves during or straight after a migraine attack

Not every attack of migraine is followed by nausea or vomiting and headache

4. Sleep disorders

Night terrors
- 1-5 years
  - Occur in first few hours of sleep
  - Intense arousal from deep slow wave sleep

Nightmares
- REM sleep
  Both may be confused with complex partial seizures

Somnambulism
The above are disorders of arousal from deep sleep – and often associated with autonomic & muscular activation

4. Sleep disorders

Narcolepsy

1. Irresistible sleep attacks (chronic daytime sleepiness)
2. Cataplexy (mimic drop attacks - sudden loss of skeletal muscle tone in response to emotional triggers)
3. Sleep paralysis
4. Hypnagogic hallucinations (Vivid dreams at onset of sleep)
5. Fragmented night sleep (may be accompanied by periodic limb movement activity)

Strong association HL-A groups DR2 or DQw1
5. Movement disorders

- Sandifer’s syndrome
- Benign neonatal sleep myoclonus
- Benign myoclonus of early infancy (3 - 8 months)
- Spasmus Nutans
- **Hyperekplexia** — (excessive jerking/jumping triggered by visual, auditory or movement)
- Paroxysmal dystonia
- Benign paroxysmal vertigo
- Shuddering attacks

Paroxysmal Non-Epileptic Disorders

**Spasmus Nutans**

- Triad:
  - asymmetrical ocular oscillations - (nystagmus)
  - head nodding
  - anomalous head position in infants
- Self limiting

- May not always be benign?
- Do CT/MRI
  - Chiasmatic
  - Ependymomas
  - Empty sellar
  - Opsoclonus/myoclonus
  - Retinal disorders
- Long term follow up

**Shuddering Attacks**

- Stiffening; adduction of knees & arms; flexion of head, elbow, trunk & knees
- No LOC
- Last a few seconds
- Ppt. by anger, fear, frustration
- Most improve by 10 years

**Hyperekplexia** (Excessive startle disease)

- Exaggerated startle response associated with generalised muscle stiffness & loss of postural control
- “Stiff-baby” syndrome
- Triggered by visual, auditory or movement or blowing on face)
- No LOC
- Familial – AD
- Rx rivotril/valproate

**Paroxysmal Non-Epileptic Disorders**

- Hyperekplexia
- Paroxysmal dystonia
  - paroxysmal dystonic nonkinesigenic choreoathetosis
  - paroxysmal dystonic kinesigenic choreoathetosis
  - paroxysmal nocturnal dystonia
  - supplementary sensorimotor seizures
- Dopa-responsive dystonia (Segawa syndrome)
- Benign paroxysmal tonic upgaze
- Benign paroxysmal torticollis

- Benign paroxysmal vertigo

  - Stereotyped
  - Usually occur in young children (1-5 years)
  - Child suddenly appears frightened, falls or holds onto to something to maintain balance
  - Usually last less than 1 minute
  - No loss of consciousness – recovers rapidly
  - Older children may complain of the vertigo and nausea
  - Frequency range from several daily to one or two per month
  - Resolves spontaneously
  - Diagnosis = preserved consciousness & memory of the event
  - May find a positive family history of migraine
  - Rx = Diphenhydramine
What about the EEG?

- Remember epilepsy is a clinical diagnosis
- Video telemetry is very helpful
- Can have a normal EEG in an epileptic
- Abnormal EEG does not always mean epilepsy – even with the occasional spikes!!
- Only clear EEG abnormalities should be regarded as significant in young children
- Remember epilepsy is a clinical diagnosis

"Only the simultaneous occurrence of a clinical seizure and characteristic EEG phenomena establishes the diagnosis."
Jean Arcardi

Video telemetry is very helpful

Other laboratory tests are occasionally useful in the differential diagnosis of epilepsy. Endocrine changes follow epileptic seizures whether generalized or partial (92), whereas they are absent following pseudoseizures. Thus, the dosage of prolactin within an hour of a seizure may help to establish its true nature (756) whether the seizures are generalized or partial (228,191,2197) as prolactin release is related to the epileptic discharge itself. Creatine-kinase on the other hand is elevated only after generalized seizures (2222) as it is related to muscle activity.