Psychogenic Non-epileptic Seizures

Dr Katharina Junejo

Consultant Child and Adolescent Psychiatrist

Aim

- Overview of basic principles in childhood epilepsy
- PNES

Diagnosis of epilepsy

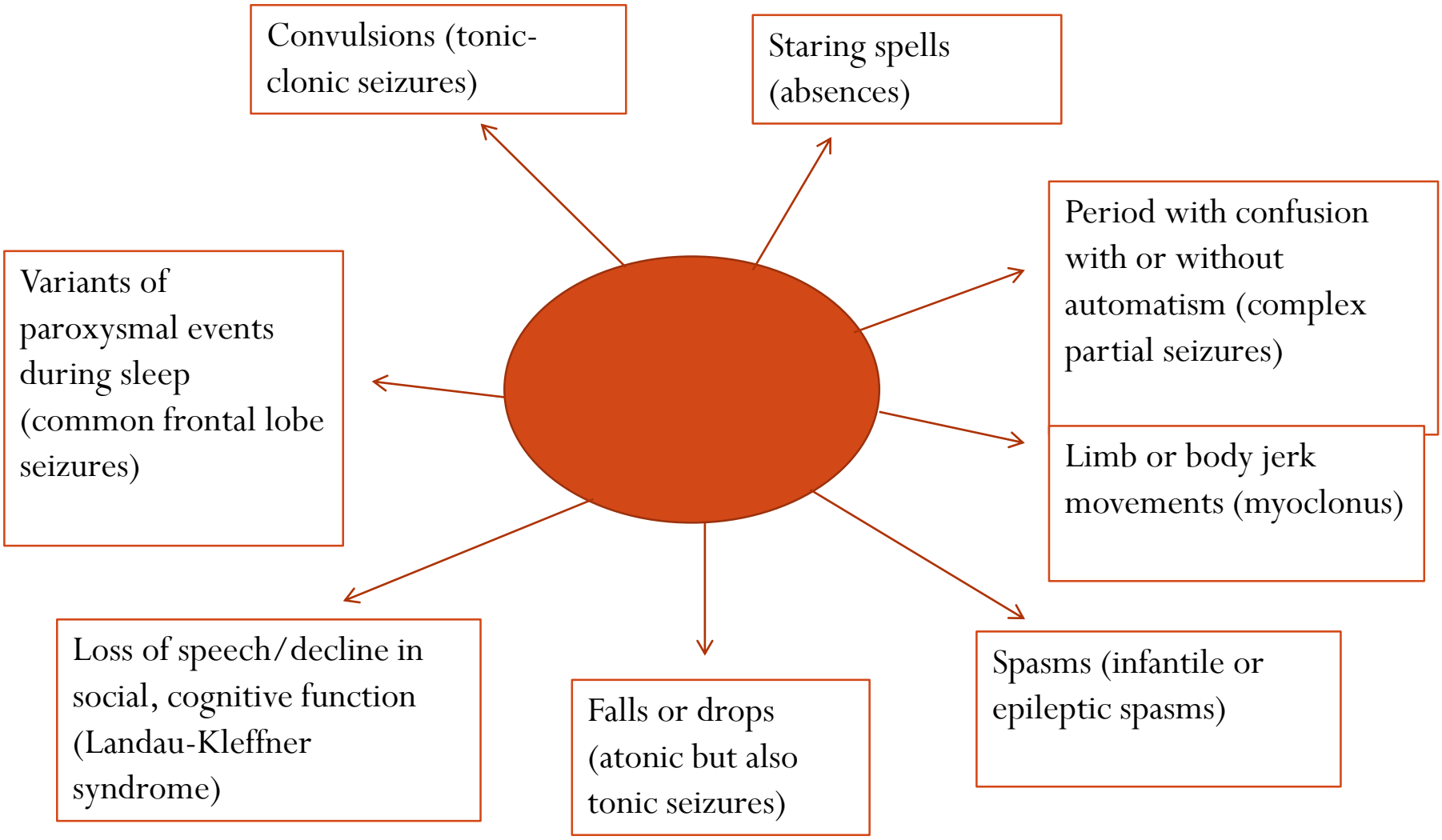
- Every child's brain is capable of generating seizures with certain pro-convulsive drugs, acute metabolic disturbance, CNS infection, acute head trauma

- Risk of epilepsy after acute and provoked seizure is 3-5%

- Emotional stress is not considered provoking factor

- Henri Gastaut and colleagues proposed a classification of seizures: “all attempts at classification of seizures are hampered by our limited knowledge of the underlying pathological processes within the brain and that any classification must of necessity be a tentative one and will be subject to change with every advance in scientific understanding of epilepsy” (Gastaut et al 1964).

Epilepsy has variable clinical presentations



ILAE Revised Terminology for Organization of Seizures and Epilepsies 2011 - 2013

- Classification of seizures: generalised and focal seizures
- Electroclinical syndromes and other epilepsies grouped by specificity of diagnosis (arranged by typical age of onset)
- Major conceptual changes: using aetiological classification of epilepsy-genetic, metabolic, immune, infectious and unknown causes
- Changes in terminology

- Operational diagnosis of epilepsy: occurrence of 2 or more unprovoked seizures, irrespective of seizure type (as recurrence risk is 80-90%)

- Careful history is only diagnostic test as many paroxysmal disorders that mimic epileptic seizures (most common vasovagal syncope or reflex anoxic seizure)

- Limitations of EEG: approx. 3-5% of normal children will show spike-wave or spike activity, approx. 20-25% of children with definite epilepsy will show no epileptic discharge on routine EEG.

Role of EEG

- Help and identify specific epilepsy syndrome
- Identify areas of focal brain dysfunction to assist in interpretation of brain imaging studies
- Confirmation or exclusion of non-convulsive status epilepticus

Epilepsy syndrome

- Age at onset of seizure
- Seizure type
- Child's neuro-developmental profile
- EEG (ideally ictal and inter-ictal recording)
- Future: specific biochemical or genetic markers

Staring spells

- Absence seizures: occur many times a day, last from 10-30 seconds
- Differential: day dreaming, engrossed in a task (watching TV), nausea, hyperventilation, pre-syncopal
- Non-epileptic seizures can be interrupted with stimulation such as tickling

Co-morbidities

Co-morbidity	Approx. prevalence
Cognitive delay	33%
ADHD	33%
Behaviour problems	29 or 58% if brain injury
Depression	25%
Anxiety	33%
Psychosis	rare
Sleep disturbance	33-50%

Distinguishing features for syncope and general tonic-clonic seizure

	Syncope	Gen tonic-clonic seizure
Aware of onset	Yes: light headedness and gradual visual loss	No: unless preceded by aura
Sudden collapse	Sometimes, but may have time to sit down	yes
Muscle tone	Limp, then may stiffen	Stiffening of arms/legs, clenching teeth
Jerking	Four or five jerks	Continuous jerking
Usual duration of jerking	Few seconds	Continuous , fast first then slowing, 30 sec-2 min
Eye rolling	sometimes	yes
Cyanosis	sometimes	often
Post-episode tiredness	yes	yes
Incontinence	sometimes	frequent

Table 1: Classification of NES based on system

System	Condition
Cardiac	Long QT syndrome Postural Orthostatic Tachycardia Syndrome (POTS) Brugada syndrome Ventricular tachy arrhythmias Heart blocks Congenital heart disease with paroxysmal pulmonary hypertension Reflex anoxic seizures
Vascular	Orthostatic syncope Vaso-vagal syncope
Respiratory	Breath holding attacks Prolonged expiratory apnoeas
Neurological	Tics Hyperekplexia Episodic ataxias Paroxysmal dyskinesias Alternating hemiplegia of children Cataplexy Chiari type 1 malformation Raised intracranial pressure Tetany Encephalitis/Encephalopathies
Psychological	Day dreams Gratification Stereotypies Out of body experience Panic/anxiety Conversion disorder / Psychogenic pseudo seizures (NEAD)
Gastrointestinal	Gastro-oesophageal reflux Sandifer syndrome Vaso-vagal syncope Familial rectal pain syndrome
Sleep related	Arousal disorders Night terrors Nightmares Sleep-wake transition disorders Benign neonatal sleep myoclonus Sleep starts Restless leg syndrome Narcolepsy
Channelopathies	Benign paroxysmal torticollis in infancy (BPTI) Episodic ataxia (EA) types 1 and 2 Familial hemiplegic migraine Benign paroxysmal vertigo of childhood (BPVC) Cyclical vomiting Benign paroxysmal tonic up gaze of childhood Hyper/hypokalemic periodic paralysis Paroxysmal dyskinesia
Unclassifiable or involving more than one system	Vaso-vagal syncope Hyperventilation syncope Benign myoclonus of early infancy (BMEI) Benign infantile spasms Fabricated illness Non-epileptic head drops Functional blinking Jitteriness Shudder Tetany

Psychogenic nonepileptic seizures

- Sudden, involuntary seizure-like attacks that, unlike epileptic seizures, are not related to electrographic ictal discharges
- Changes in behaviour, motor activity, sensation, cognitive, and autonomic functions
- Accurate diagnosis is usually delayed by an average of 7 years & is confirmed via vEEG

Features

- Mimic a convulsive seizure
- Atypical features: thrashing movements, arching posturing of the back, responsive during event, eyes closed, long duration
- Mimic non convulsive seizures: Motionless starting, eyes closure

Causes

- PNES is heterogeneous condition
- Are not caused by abnormal brain activity
- Associated with Hx of abuse
- Reflect conscious/unconscious conflict
- Are not purposely produced by patient and individuals are not aware they have PNES

Triggers

- May happen spontaneously or be triggered by sensory experiences
- Can appear when people are relaxed as triggers may be below conscious awareness
- Protection of recurrent traumatic memories

Comorbidity

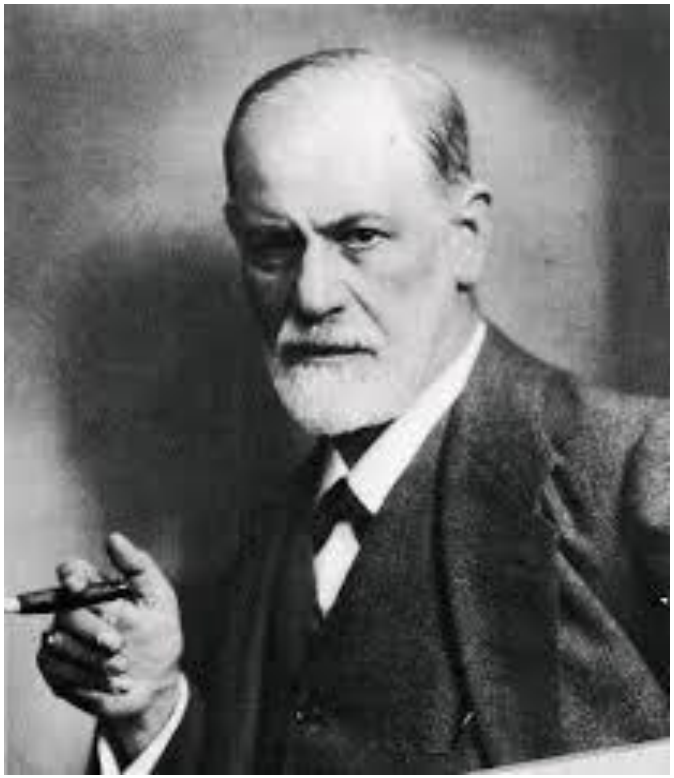
- 5-20 % have PNES and comorbid seizures
- More frequently in people with cognitive developmental delay
- Depression, anxiety
- PTSD
- Personality disorder
- 20% no current psychiatric comorbid condition

DSM IV conversion disorder->functional neurological disorder in DSM V

- **Conversion disorder with seizures or convulsions**
- Symptom affecting motor/sensory system suggesting neurological/medical condition
- Psychological factors associated because symptom is preceded by conflict or stressor
- Symptom is not intentionally produced
- Symptom cannot be explained by medical condition
- Symptom cannot be explained by a substance effect
- Symptom cannot be explained by cultural behaviour

Hysteria (1893)

- Non-epileptic convulsions were found to be related to psychical trauma
- Process of splitting and repression of memories
- Psychical trauma leads to a retention of the affect and hysterical symptoms arise as a result of excitation transposed from the sphere of the mental to the physical



Neurophysiological model

- Janet 1889-dissociation model: severe stress disrupts mental integration between ideas and sensory/motor function
- Innate defence response to danger-high arousal states activate defence system-collapsed or tonic immobility (freeze reaction)
- Loss of connectivity between different brain areas (prefrontal and subcortical)
- Proposed mechanisms involved in dissociative brain processes: catecholamine release, secretion of anaesthetic neurochemicals, network disruption (glial cell response to stress)

Epidemiology

Prevalence of epilepsy

- 4-5/1000 children in Europe/USA-**>up to 30% may have misdiagnosis**
- 10% of all medical services are provided for psychogenic symptoms

Frequency

- 20 to 30% of referrals to epilepsy centers are PNES
- 50 to 70% become seizure free after diagnosis
- 15% also have comorbid seizure disorder

Gender

- Women 70% of diagnosed PNES

Age

- Typically beginning in young adulthood
- Can occur in children and elderly

Suggestive PNES presentation

- Resistance to antiepileptic drugs
- Presence of specific triggers/life events
- Unusual circumstances when event occurs
- Other triggers like pain, sounds, specific movement/lights
- Usually do not occur during sleep
- Characteristics of event are inconsistent with epilepsy such as side-to-side head shaking, bicycling, weeping, stuttering and arching of the back
- Comorbid diagnosis of chronic pain, chronic fatigue
- Lack of concern (la belle indifference)
- Hx of trauma

Predictable difference

Epileptic seizure

- Abrupt onset
- Loss of awareness
- Eye opening/widening
- Movements are rhythmic and synchronous
- Tongue biting, ictal cry specific to gen tonic-clonic seizure

Psychogenic seizure

- Preserved awareness
- Eye flutter and resistance if attempts made to open eyes
- Episodes intensified or alleviated by observers
- Able to be provoked by an induction techniques

Medical work up

Laboratory studies

- Metabolic/toxic causes
- Prolactin and CK rise after gen tonic-clonic seizure

Imaging studies

- Imaging studies are normal
- Incidental findings should not confound diagnosis of PNES

EEG

- Routine EEG has low sensitivity but repeated normal results with repeated attacks and resistance to medication is a RED FLAG
- EEG video monitoring
- Analysis of video is as important as EEG as it shows behaviour incompatible with seizures
- Useful sign is preserved awareness during bilateral motor activity as a specific indicator of PNES

Vulnerability factors

- Dissociative tendencies
- Alexithymia
- Cognitive rigidity
- Hypervigilance
- Adverse childhood experiences

Discussion of diagnosis

- MDT presentation, both neurologist and psychiatrist present
- Discussion of findings on vEEG, ruling out epilepsy
- Individualised hypothetical explanation considering patient's vulnerability traits
- Introducing concept of psychotherapeutic interventions
- Treating psychiatric co-morbidities
- Involving neurologist post diagnosis

Treatment

- Prior to 2000 no clinical trial of specific interventions designed for PNES-"orphan disorder"

- Obstacles to conduct large trials: patients present in crisis but reject long term support, emotional lability, approach-avoidance behavioural pattern

Treatment studies in adult population

CBT (Goldstein et al, 2010)

1. Process: Engagement->reinforcement of independence->distraction/refocusing during event->graded exposure to avoided situation->cognitive restructuring->relapse prevention
2. Outcome: post treatment (12 sessions)->significantly lower event rate, 6 month FU: no statistical difference (compared to SMC)

Psychodynamic psychotherapy (Mayor et al, 2010)

1. Process: Symptoms are linked to difficulties within relationships influenced by interpersonal patterns developed earlier in life->identifying unhelpful patterns->more effective emotions processing
2. Outcome: post treatment (20 sessions), 3.5 years->25% event free, further 40% event reduction, median frequency of events decreased from 6 at baseline to one at FU

- **Group psychotherapy** (Barry et al, 2008)

1. Process: Conscious and verbal expression of emotional distress, reduce the need for somatic display of stress, developing assertive coping strategies
2. Outcome: overall event decrease, however would require replication study

- **Psychopharmacological intervention-**>lack of consistent neurobiological model

1. **Sertraline vs CBT vs Sertraline and CBT vs TAU** (LaFrance et al, 2014)
2. Outcome: Sertraline and CBT + CBT only showed reduction in event frequency and improvement in secondary outcome measures but not TAU or SSRI on its own

Summary

- Epilepsy in childhood has variable clinical presentations and needs careful diagnostic workup to prevent unnecessary medication treatment and potential restriction to the child's life
- Seizure type events need equally careful history taking and awareness of potential differential diagnosis
- Developing treatments for PNES is at an early stage and studies need further replication of results

References

- Childhood epilepsy. Management from diagnosis to remission, Ed. R Appleton, P. Camfield. 2011
- On the Psychical Mechanism of Hysterical Phenomena, S. Freud, 1893
- Psychogenic nonepileptic seizures: a treatment review. What have we learned since the beginning of the millenium?, Baslet, G, Neuropsychiatric Disease and Treatment, 2012
- Psychogenic non-epileptic seizures in children and adolescents: Part II – explanations to families, treatment, and group outcomes, K Kozlowska et al, Clinical Child Psychology and Psychiatry 2018, Vol. 23(1) 160–176