Benign and Malignant Rolandic and Occipital Spikes

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Objectives

- To review the differences between benign and malignant rolandic and occipital spikes
- To review the electroclinical features of idiopathic and symptomatic rolandic and occipital epilepsies
- To discuss differential diagnosis
Disclosures

- None
History of Benign Rolandic Epilepsy

- EEG pattern - Gastaut in 1952
- Clinical pattern - Nayrac and Beaussart 1958
- EEG abnormality may occur without clinical seizures - Gibbs et al. 1954
- Genetic factors - Bray and Wiser 1964
Interictal Discharge
Benign Rolandic Epilepsy

- Most common partial epilepsy
- Population study: 6.2-10.7 per 100,000
- Onset 3-13 years
- M:F 1.5:1
- 10-13% have a single seizure
- 20% have frequent seizures
- 65% nocturnal; 15% nocturnal or diurnal; 10-20% in waking state only
Description of Seizures

- Somatosensory onset with unilateral parasthesiae of tongue, lips, gums or inner cheeks
- Unilateral tonic, clonic or tonic-clonic activity in face, lips, tongue, pharyngeal and laryngeal muscles
- Speech arrest or dysarthria
- Drooling
- Preservation of consciousness
Description of Seizures

- Simple partial hemifacial seizure with somatosensory aura
- Often associated loss of awareness
- Secondarily generalized seizure - onset not witnessed
Video of Benign Rolandic Seizure
Rolandic Spike Dipoles

- Very common
- Only 9% of children with rolandic spike dipoles develop epilepsy
- Rolandic spike dipoles may occur in symptomatic rolandic epilepsy
High Voltage Diffuse Rolandic Spikes
Sensitivity 50μV/mm
Ictal Dipole Discharges

F4 - A2

C4 - A2

T4 - A2
Ictal EEG in Benign Rolandic Epilepsy
Neuropsychological Function in BREC

- 17 patients with BREC
  - 7 - 14 years
  - 12 on medication

- 17 controls
  - age, sex and estimated intelligence

- Assessments
  - Neuropsychological testing
  - Parent and Teacher rating

*Croona et al Dev Med Child Neurol 1999;41:813*
Neuropsychological Function in BREC

- Significant differences in
  - Auditory-verbal learning
  - Memory
  - Executive function
- Parent and Teacher Rating
  - Parents - distractibility, impulsivity,
  - Teachers - reading comprehension

Croona et al Dev Med Child Neurol 1999;41:813
Evolution of Benign Rolandic Epilepsy

35 children

- 6 monthly assessments
  - clinical
  - EEG
  - neuropsychology

- First seizure to recovery

Massa et al. Neurology 2001;57:1071-1079
Prospective Study of BREC

- Decline mainly involved
  - Performance IQ
  - Sustained attention
  - Working memory
  - Executive functioning.
# EEG Features Predictive of Poor Prognosis

<table>
<thead>
<tr>
<th>EEG Abnormality</th>
<th>P value</th>
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</thead>
<tbody>
<tr>
<td>Intermittent slow wave focus</td>
<td>&lt;0.001</td>
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<tr>
<td><strong>Asynchronous bilateral spike wave foci</strong></td>
<td>&lt;0.001</td>
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<tr>
<td>Rhythmic clusters of spike-wave</td>
<td>&lt;0.001</td>
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<tr>
<td>Generalized 3-4 Hz spike waves</td>
<td>&lt;0.05</td>
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<tr>
<td>Atonia, myoclonia correlates with SW</td>
<td>&lt;0.05</td>
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3 of 5 features in 10 children for at least 6 months
Simon

- Antenatal ultrasound - right pachygyria
- Mild left hemiplegia
- 5 X GTC with fever - 18 months to 5 years
- Intermittent L facial and speech difficulty for up to 30 mins at 5 years
Simon’s EEG
Effect of AEDs on Interictal Spikes in Children

![Bar chart showing the clearance rate of PHB, CBZ, and VPA for focal and generalized spikes.]

- **PHB**: Lower clearance rate for focal spikes compared to generalized spikes.
- **CBZ**: Higher clearance rate for generalized spikes than focal spikes.
- **VPA**: Consistent clearance rate for both focal and generalized spikes.
Sulthiame in Benign Rolandic Epilepsy

- Sulthiame (31) seizure free: 80
- Placebo (35) seizure free: 20
Sulthiame in Benign Rolandic Epilepsy

Rating et al. Epilepsia 2000;41:1284
Drugs which Suppress Interictal Discharges

- Sulthiame
- Diazepam
- Valproic acid
- Lamotrigine
- Corticosteroids
  - Landau-Kleffner
  - Continuous Spike-Wave in Slow Sleep
Malignant Rolandic Spikes

- Christine developmentally normal
- Referred at 9 years of age
- At 7.5 years gradual deterioration in speech
- No clinical seizures
- Initially diagnosed with “Selective mutism”
Landau-Kleffner Syndrome

- Difficulty understanding speech and sounds
- Gradual deterioration in ability to speak
- More distractible in school
- Behavior problems
Christine: Treatment

- Prednisone 2mg/kg
- Sulthiame added 3 months post diagnosis
  - Sulfonamide derivative
  - Mechanism of action
    - Carbonic anhydrase inhibition
    - Blocks sodium channel
Christine - Evolution

- Dramatic improvement clinically
- 10 months following diagnosis speaking well
- Presently – 6.5 years since diagnosis
  - Mild comprehension difficulty in a noisy environment
  - EEG normal
- Now off medication for 5 years
Elizabeth

- 3.5 year old girl
- Onset of seizures at 3 years
  - Staring, eyes and head to left
  - Unresponsive, limp, pale
  - Vomiting
  - Duration 12-20 minutes
- Normal development and exam
- Maternal grandmother epilepsy
Panayiotopoulos Syndrome

- 3-5 years (range 1-14 years)
- Nocturnal seizures in 2/3
- Tonic eye deviation, vomiting
- Visual symptoms rarely reported
- Prominent autonomic features
Panayiotopoulos Syndrome

- Seizures prolonged, status in 1/3
- Infrequent seizures
- Prognosis excellent
- Seizures rare after 13 years
- Children may develop rolandic epilepsy
- EEG: multifocal posterior quadrant epileptiform discharges
Gastaut Syndrome

- Brief seizures characterized by visual hallucinations or ictal blindness
- Children 4-16 years
- 5% had symptoms in adulthood
EEG Features

- Normal background
- High amplitude spike-wave (80%) or sharp waves (20%) over the occipital and or posterior temporal area
- Discharges occur rhythmically
- May disappear on eye-opening in 94%
- 38%: generalized spike-wave or centrotemporal spikes
Clinical Features

- Amaurosis in 52%
- Phosphenes in 45%
- Complex visual hallucinations in 14%
- Visual illusions in 14%
  - micropsia
  - palinopsia
Clinical Features

- Hemiclonic seizures in 43%
- Complex partial seizures in 14%
- Generalized tonic-clonic seizures 13%
- Other features in 25%
  - dysphasia
  - dysesthesiae
Clinical Features

- Post-ictal headache in 33%
- Nausea in 17%
- No clear precipitating factors
- Features may be difficult to differentiate from migraine
Prognosis of Gastaut Syndrome

- Complete seizure control in 60%
- Remission in late adolescence although up to 5% of adults may continue to have seizures
- Diagnosis of this condition is difficult
Idiopathic OLE with Photosensitivity

- **Guerrini et al.** *(Epilepsia 1995;36:883-891)*
- 5-17 years of age
- Seizures induced by light, visual hallucinations, tonic head and eye deviation, nausea, headache, may be aware
Symptomatic Occipital Epilepsy: Ictal onset

Asleep. No clinical signs.
Evolution of Seizure
4 MINUTES INTO SEIZURE: EYES OPEN & TO THE RIGHT
Challenges in Occipital Lobe Epilepsy

- 66 children with OLE (BC Children’s Hospital series - Schrader et al. submitted)
- 21 Symptomatic
- 12 Probable symptomatic
- 33 idiopathic
  - Panayiotopoulos syndrome (n=9)
  - Gastaut syndrome (n=12)
  - Overlap (n=11)
  - Idiopathic OLE with photosensitive epilepsy (n=1)
Predictors of abnormal MRI

- Early age of seizure onset
- Abnormal neurological examination
- No difference in clinical semiology between idiopathic and symptomatic group
Key points

- Benign rolandic epilepsy is most common partial epilepsy in children
- Rolandic spikes may occur in children without epilepsy
- Rolandic spikes may occur in symptomatic epilepsies
- Cognitive and behaviour changes may occur with interictal spikes
Key Points

- 3 variants of idiopathic epilepsy with occipital or posterior epileptiform discharges
- Panayiotopoulos syndrome
  - is under recognized and now categorized as an autonomic epilepsy
  - rolandic spikes/ epilepsy may develop
- Clinical features do not differentiate symptomatic and idiopathic occipital lobe epilepsy