Disclosures

None
Objectives

- Recognize and diagnose electroclinical and epilepsy syndromes in the adolescent and adult

- Summarize treatment strategies and prognostic implications in these epilepsy syndromes
**AGE OF ONSET**

**NEONATAL/INFANTILE**
- Self-limited neonatal and familial neonatal epilepsy
- Early myoclonic encephalopathy
- Ohtahara syndrome
- West Syndrome
- Dravet syndrome
- Myoclonic epilepsy in infancy
- Epilepsy of infancy with migrating focal seizures
- Myoclonic encephalopathy in non-progressive disorders

**ADOLESCENT/ADULT**
- Juvenile absence epilepsy
- Juvenile myoclonic epilepsy
- Epilepsy with generalized tonic-clonic seizures alone
- Autosomal dominant epilepsy with auditory features
- Other familial temporal lobe epilepsies

**CHILDHOOD**
- Epilepsy with myoclonic-atonic seizures
- Epilepsy with eyelid myoclonias
- Lennox-Gastaut syndrome
- Childhood absence epilepsy
- Epilepsy with myoclonic absences
- Panayiotopoulos syndrome
- Childhood occipital epilepsy (Gastaut type)
- Photosensitive occipital lobe epilepsy
- Childhood epilepsy with centrotemporal spikes
- Atypical childhood epilepsy with centrotemporal spikes
- Epileptic encephalopathy with continuous spike-and-wave during sleep
- Landau-Kleffner Syndrome
- Autosomal Dominant nocturnal frontal lobe epilepsy

**VARIABLE**
- Reflex epilepsies
- Progressive myoclonic epilepsies
- Familial focal epilepsy with variable foci
- Juvenile absence epilepsy (JAE)
- Juvenile myoclonic epilepsy (JME)
- Epilepsy with generalized tonic-clonic seizures alone
- Progressive myoclonic epilepsies
- Autosomal dominant epilepsy with auditory features (ADEAF)
- Other familial temporal lobe epilepsies
- Reflex epilepsy
Definitions

- **Generalized seizure** “engage or involve networks on both sides of the brain at the onset”

- **Epilepsy Syndrome** “cluster of features incorporating seizure types, EEG and imaging features that tend to occur together...often has age-dependent features...seizures triggers, diurnal variation, sometimes prognosis...distinctive comorbidities...It may have associated etiologic, prognostic, and treatment implications.”

- Never a formal classification of epilepsy syndromes by ILAE

CASE: 12-year-old healthy boy who presents with first convulsive seizure without aura. He has also had weekly staring episodes over the past year that were treated originally as ADHD.

JUVENILE ABSENCE EPILEPSY (JAE)
Primary Generalized Epilepsies (PGE)
- Idiopathic Generalized Epilepsies (IGE)
- Genetic Generalized Epilepsy (GGE)

- Childhood Absence Epilepsy
- Juvenile Absence Epilepsy
- Juvenile Myoclonic Epilepsy
- Generalized Tonic-Clonic Seizures Alone
Operational classification of seizure types by the International League Against Epilepsy: Position Paper of the ILAE Commission for Classification and Terminology

*Robert S. Fisher, ‡J. Helen Cross, §Jacqueline A. French, ¶Norimichi Higurashi, ¶Edouard Hirsch, #Floor E. Jansen, **Lieven Lagae, ††Solomon L. Moshe, ‡‡Jukka Peltola, §§Elaine Roulet Perez, †††Ingrid E. Scheffer, and ††***Sameer M. Zuberi

Epilepsia, 58(4):522–530, 2017
doi: 10.1111/epi.13670
Juvenile Absence Epilepsy

- Development: normal
- Onset: around puberty, 8 to 16 years (peak 10-12)
- Seizure Types:
  - Absence seizures predominate but not as frequent as CAE and not typically triggered by hyperventilation
  - GTC seizure
  - Rare myoclonic seizures
- Can be inherited (positive family history in 1/3)
JAE Electrical Features

Generalized spike wave complexes 3 to 4 Hz
JAE Electrical Features
Generalized spike wave complexes 3 to 4 Hz
CASE: 16-year-old healthy girl who presents with first convulsive seizure at school after staying up until 3am studying for an exam. On further history you find she has been having morning myoclonic jerks for a while and that she has to close her eyes for flashing lights as they make her feel “jumpy”.

JUVENILE MYOCLONIC EPILEPSY (JME)
Juvenile Myoclonic Epilepsy

- Development: normal
- Onset: Later adolescence, 12 to 18 years (peak 15)
- Seizure Types:
  - Myoclonic seizures
    - Upper extremity prominence, typically extensors
    - Single or repetitive
  - GTC seizures
  - Absence in 1/3 but infrequent
- Can be inherited (positive family history in 1/3)
- Sensitivity to light, sleep deprivation, alcohol
- Often requires long term treatment

JME Electrical Features

Generalized spike wave complex, often polyspike components
JME Electrical Features

Generalized Polyspikes
JME Electrical Features

Photoparoxysmal Response
CASE: 28-year-old man presents for increasing frequency (now monthly) generalized tonic clonic seizures usually occurring in the morning before work. No myoclonic jerks, photosensitivity, or other seizure types. His paternal grandfather and one of his paternal cousins also has epilepsy.

EPILEPSY WITH GENERALIZED TONIC-CLONIC SEIZURES ALONE
GTC seizures alone

- Development: normal
- Onset: second decade: 5 to 50 years (peak 15)
- Worsens with age
- Seizure Types:
  - Predominantly GTC on awakening
  - Less frequent myoclonic or absence
- Sensitive to light, sleep deprivation, alcohol
# GGE Summary & Genetics

<table>
<thead>
<tr>
<th></th>
<th>Absence</th>
<th>Myoclonic</th>
<th>GTC</th>
<th>Onset</th>
<th>Identified Genes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CAE</strong></td>
<td>+++</td>
<td>-</td>
<td>-</td>
<td>Childhood 4 to 10</td>
<td>GABRG2, GABRA1, GABRB3, CACNA1H</td>
</tr>
<tr>
<td><strong>JAE</strong></td>
<td>++</td>
<td>+</td>
<td>++</td>
<td>Puberty 8 to 16</td>
<td>GRIK 1</td>
</tr>
<tr>
<td><strong>JME</strong></td>
<td>+/-</td>
<td>+++</td>
<td>+++</td>
<td>Late adolescence 12 to 18</td>
<td>EFHC1, GABRA1, BRD/RING3, Cx-36, CACNB4, GABRD, CASR, ME2, SCN1B, EFHC2, CLCN2, BRD2</td>
</tr>
<tr>
<td><strong>GTC only</strong></td>
<td>+/-</td>
<td>+/-</td>
<td>+++</td>
<td>Second decade 5 to 50</td>
<td>CLCN2</td>
</tr>
</tbody>
</table>

Risk to first degree relative of GGE patients ~ 7%, but remains a generic prediction.\(^1\)

Take home points GGE

- Genetic ≠ Inherited
- Normal development
- Typically good prognosis: Not progressive, can resolve, typically well-controlled, no neurological deficits
- Adult onset is not rare
  - 28% of GGE have onset after age 20\(^1\)

\(^1\)Marini C, et al. *J Neurol Neurosurg Psychiatry* 2003; 74:192-6
The SANAD study of effectiveness of valproate, lamotrigine, or topiramate for generalised and unclassifiable epilepsy: an unblinded randomised controlled trial

Anthony G Marson, Asya M Al-Kharusi, Muna Alwaigh, Richard Appleton, Gus A Baker, David W Chadwick, Celia Cramp, Oliver C Cockerell, Paul N Cooper, Julie Doughty, Barbara Eaton, Carrol Gamble, Peter J Goulding, Stephen J L Howell, Adrian Hughes, Margaret Jackson, Ann Jacoby, Mark Kellett, Geoffrey R Lawson, John Paul Leach, Paola Nicolaides, Richard Roberts, Phil Shackley, Jing Shen, David F Smith, Philip F M Smith, Catrin Tudur Smith, Alessandra Vanoli, Paula R Williamson, on behalf of the SANAD Study group

Lancet 2007; 369: 1016-26

✓ Valproate and topiramate have better seizure control than lamotrigine

✓ Valproate better tolerated than topiramate

“valproate should remain the drug of first choice for many patients with generalized and unclassified epilepsies. However, because of known potential adverse effects of valproate during pregnancy, the benefits for seizure control in women of childbearing years should be considered.”
Ask the Experts

Special Communication

Epilepsy treatment in adults and adolescents: Expert opinion, 2016

Jerry J. Shih a,*, Julia B. Whitlock a, Nicole Chimato b, Emily Vargas b, Steven C. Karceski c, Ryan D. Frank b

a Department of Neurology, Mayo Clinic, Jacksonville, FL, United States
b Department of Health Sciences and Research, Mayo Clinic, Jacksonville, FL, United States
c Department of Neurology, Weill Cornell Medical Center, New York, NY, United States

✓ **Valproate** for all GGE (except women of child-bearing age)

✓ **Lamotrigine** or **levetiracetam** women of child-bearing age with GGE

✓ **Ethosuximide** in patient with absence only

✓ **Levetiracetam** in patients with GGE with GTC & myoclonic
### Monotherapy in adults and children with new-onset GE or unclassified GTC seizures

<table>
<thead>
<tr>
<th>Level</th>
<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>No Recommendation</td>
<td>Evidence is insufficient to compare efficacy of LTG and TPM with that of valproic acid (VPA) in children and adults with new-onset or relapsing GE (1 Class III study).</td>
</tr>
</tbody>
</table>

### For adult and pediatric patients with TR generalized epilepsy (GE), are these AEDs effective in reducing seizure frequency when used as adjunctive therapy (compared with no adjunctive therapy)?

<table>
<thead>
<tr>
<th>Level</th>
<th>Recommendation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Level B</td>
<td>For add-on therapy for GE, immediate-release and extended-release lamotrigine use should be considered as add-on therapy to decrease seizure frequency in treating adults with TR generalized tonic-clonic (GTC) seizures secondary to GE.</td>
</tr>
<tr>
<td>Level B</td>
<td>Levetiracetam (LEV) use should be considered to decrease seizure frequency as add-on therapy for TR GTC seizures and for TR juvenile myoclonic epilepsy.</td>
</tr>
</tbody>
</table>
Generalized Epilepsy Treatment

- First Line: **Valproate** (consider teratogenicity)

- Second Line: **Topiramate, Lamotrigine**
  - Lamotrigine could exacerbate myoclonus *level F*

- Third Line:
  - Levetiracetam (good for myoclonus) *class B*
  - Zonisamide *case series*

- Possibly Effective: Benzodiazepines, Primidone, Phenobarbital

- **Avoid:** (aggravate seizures) *class IV*\(^2\)
  - Carbamazepine
  - Oxcarbazepine
  - Vigabatrin
  - Gabapentin
  - Tiagabine
  - Phenytoin

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\(^2\)Guerrini et al., 1998; Genton, 2000; Somerville, 2009
## Long Term Outcome of JME

<table>
<thead>
<tr>
<th>First Author</th>
<th>Pub. Year</th>
<th>n</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delgado</td>
<td>1984</td>
<td>43</td>
<td>28%(12) relapse with medication withdrawal after 2 yr seizure free</td>
</tr>
<tr>
<td>Camfield</td>
<td>2009</td>
<td>23</td>
<td>17% seizure free at 25 yrs f/u</td>
</tr>
<tr>
<td>Seneviratne</td>
<td>2012</td>
<td>review</td>
<td>&lt; 20% remain in remission without treatment</td>
</tr>
<tr>
<td>Geithner</td>
<td>2012</td>
<td>31</td>
<td>68% seizure free at 25-63 yr f/u</td>
</tr>
<tr>
<td>Senf</td>
<td>2013</td>
<td>66</td>
<td>59% seizure free &gt; 5 years at 45 yr f/u</td>
</tr>
<tr>
<td>Hofler</td>
<td>2014</td>
<td>175</td>
<td>8% seizure free &gt; 10 yr</td>
</tr>
<tr>
<td>Vonderwulbecke</td>
<td>2017</td>
<td>176</td>
<td>60% &gt; 5 years seizure free at 43 yr f/u</td>
</tr>
</tbody>
</table>
Prognosis of GGE

- Response to treatment overall good (but not complete)
- Previous reports long term remission rates higher in CAE than JAE/JME

Long-term outcome in adolescent-onset generalized genetic epilepsies

Bernd J. Vorderwülbecke, Alexander B. Kowski, Andrea Kirschbaum, Hannah Merkle, Philine Senf, *Dieter Janz, and Martin Holtkamp

Epilepsia, 58(7):1244–1250, 2017
doi: 10.1111/epi.13761

- Long term seizure outcome hardly differed between the three subsyndromes
- 60% seizure free 5 years
- 14% > 10 years seizure free with 5 years off medication
13-year-old developmentally normal teenage boy:
- First generalized tonic clonic seizure at age 9
- Myoclonic jerks started age 12
- Absence seizures started at age 8 (not having currently)
- Older brother with similar history controlled on medications

What is the best diagnosis?
Continuum of GGE

CAE  JAE  JME

GTCs Only

AGE (years)

ABSENCE  MYOCLONIC  GTC

4  10  18
Same patient returns at age 15 refractory to levetiracetam and zonisamide. He develops unsteady gait and tremor, as well as worsening myoclonus that now occurs for hours and with startle. He begins to have decline in school performance, becoming more withdrawn. What is the diagnosis?
PROGRESSIVE MYOCLONIC EPILEPSY (PME)

PME

1. Seizure
2. Delay
3. Ataxia
4. Myoclonus
Progressive Myoclonic Epilepsy

- Heterogenous conditions that manifest clinically in a similar way
- Rare
- Genetic
- Progressive, neurodegenerative with poor outcome
- Childhood or adolescence
- EEG: Slow, generalized spikes and wave 2.5 to 6 Hz, polyspikes, focal sharp waves
- Photosensitivity (lower frequency than GGE)
# Progressive Myoclonic Epilepsy (PME)

<table>
<thead>
<tr>
<th>Autosomal Recessive</th>
<th>Autosomal Dominant</th>
<th>Mitochondrial</th>
</tr>
</thead>
</table>
| Unverricht-Lundborg (ULD)  
CSTB (EPM1) | DRPLA  
DRPLA | Myoclonic epilepsy with ragged red fibers (MERRF)  
MTTK |
| Lafora Disease  
EPM2A, NHLRC1 | Adult onset NCL  
(Kuff’s) | |
| Sialidoses  
NEU1 | | |
| Neuronal Ceroid Lipofuscinos (NCL)  
TPP1 CLN3, CLN5, CLN6 | | |

Shahwan et al. Lancet Neurol 2005  
Girard JM et al. Handb Clin Neurol 2013
Unverricht-Lundborg Disease (ULD)

- Onset: 6 to 18 years, stabilize by 40, can live to 60s
- Seizures: Stimulus sensitive myoclonus + GTC/absence
- Other Features:
  - Progressive ataxia, dysarthria, intention tremor
  - Mild dementia (late feature)
- Genetics: autosomal recessive, mutation in cystatin B gene (CSTB, previously EPM1)
- Treatment tips: Phenytoin dramatically worsens ataxia
- EEG: generalized spikes and polyspikes with photosensitivity
- MRI: mild cerebellar / brainstem and less often cerebral atrophy
Lafora Bodies

Periodic-acid-Schiff (PAS) positive intracellular polyglucosan inclusion body in neurons, heart, skeletal muscle, liver, sweat gland ducts


Lafora Disease

- **Onset:** 6 to 19 years, death < 10 years
- **Seizures:** GTC, clonic, myoclonic, astatic, atonic, absence, focal, and *occipital seizures*; often deteriorate to status
- **Other Features:**
  - Normal development until onset, isolated febrile or nonfebrile seizures in childhood
  - Progressive cognitive decline
  - Blindness
  - Ataxia
- **Genetics:** autosomal recessive, generalized polyglucosan storage disorder
  - EPM2A gene Laforin
  - EPM2B (*NHLRC1*) Malin
- **Diagnosis:** skin biopsy, lafora bodies, genetics
- **EEG:** low frequency photoparoxysmal response
Neuronal Ceroid Lipofuscinosis (NCL)

- Onset: varies with 5 subtypes, rapid progression, early death
- Seizures: + myoclonus, variable
- Other Features:
  - Progressive blindness characterized by retinal degeneration and optic atrophy
  - Progressive cognitive decline
- Genetics: autosomal recessive (except adult subtype can be dominant) accumulation of lipopigments in lysosomes
  - *PPT1, CLN1, TPP1, CLN3, CLN5, CLN6, MFSD8, CLN8, CTSD, DNAJC5, CTSF, ATP13A2, GRN, KCTD7*
- Diagnosis: intracellular inclusions (eccrine, conjunctival, muscle biopsy) or genetics
- EEG: Low frequency photic stimulation may induce occipital spikes, giant VEPs
- MRI: global atrophy, white matter T2 hyperintensities
NCL Subtypes

- **Type 2 Late infantile (Jansky-Bielschowsky)**
  - Onset 2.5 to 4 years, death in 5 years
  - **TPP1**

- **Type 3 Juvenile (Spielmeyer-Vogt-Sjogren or Batten)**
  - Onset: 4 to 10 years
  - Prominent feature is loss of vision, seizures less prominent
  - **CLN3**

- **Type 4 Adult (Kuf's or Parry)**
  - Onset: adolescence or adulthood
  - no eye features
  - Can be autosomal recessive or dominant

- **Type 5 Finnish variant late infantile**
  - **CLN5**

- **Type 6 Late infantile variant**
  - **CLN6**
Myoclonic Epilepsy with Ragged Red Fibers

Gomori trichrome with ragged red fibers

Myoclonic Epilepsy with Ragged Red Fibers (MERRF)

- Onset: childhood to young adult, insidiously or with metabolic crisis
- Genetics: Mitochondrial (maternal) or autosomal or sporadic tRNA Lys gene (MTTK) mutation
- Diagnosis: Ragged red fibers on muscle biopsy, genetics
- MRI: atrophy and calcificatins of basal ganglia, T2 grey matter intensity changes
- Treatment tips: Caution with valproate, sometimes empirically treated antioxidant vitamins, cofactors (coenzyme Q, L-carnitine)

<table>
<thead>
<tr>
<th>Clinical Features:</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Epilepsy (myoclonus and GTCs)</td>
</tr>
<tr>
<td>• Ataxia</td>
</tr>
<tr>
<td>• Myopathy, neuropathy</td>
</tr>
<tr>
<td>• Deafness</td>
</tr>
<tr>
<td>• Optic atrophy</td>
</tr>
<tr>
<td>• Dementia</td>
</tr>
<tr>
<td>• Short stature</td>
</tr>
<tr>
<td>• Less common: diabetes mellitus, cardiomyopathy, retinopathy, ophthalmoparesis, multiple lipomas, pyramidal signs</td>
</tr>
</tbody>
</table>
Sialidosis

Fundus with Cherry – Red Spot

Sialidosis I

- Onset: Juvenile
- Seizures: severe action/intention myoclonus, later GTC
- Other Features:
  - Cherry red spot, progressive visual failure
  - Gradual ataxia, spasticity
  - Painful sensory peripheral neuropathy
  - Dysmorphism: coarse faces, corneal clouding, skeletal dysplasia
  - Much less cognitive issue
- Genetics: autosomal recessive, neuroaminidase A, Nue1 gene
- Diagnosis: Vacuolate Kupffer cells, genetics

- TYPE II: similar but more severe, as early as neonatal onset, with hepatosplenomegaly and mental deterioration
Dentato-rubral-pallido-luysian atrophy (DRPLA)

- Onset: PME phenotype only if onset < age 20
- Seizures: myoclonic, GTC
- Other Features:
  - Extrapyramidal symptoms
  - Dementia
- Genetics: autosomal dominant with anticipation, unstable expansion of CAG trinucleotide repeats of DRPLA
- MRI: parasagittal atrophy esp. in pons and cerebellum
## Summary of common PME

<table>
<thead>
<tr>
<th>Disease</th>
<th>Age of onset</th>
<th>Seizures</th>
<th>Dementia</th>
<th>Ataxia</th>
<th>Distinctive features</th>
</tr>
</thead>
<tbody>
<tr>
<td>ULD</td>
<td>6 to 18</td>
<td>Myoclonic ++++ GTC/absence</td>
<td>Minimal</td>
<td>Late</td>
<td>Milder, live to 60s No PHT!</td>
</tr>
<tr>
<td>Lafora</td>
<td>6 to 19</td>
<td>Myoclonic, GTC, atonic, focal +++ occipital (status)</td>
<td>Early and severe</td>
<td>Early</td>
<td>Death &lt; 10 yr</td>
</tr>
<tr>
<td>NCL</td>
<td>Vary</td>
<td>Myoclonic ++, Variable</td>
<td>Progressive</td>
<td>Variable</td>
<td>Blindness, Visual abnormalities, early death</td>
</tr>
<tr>
<td>MERRF</td>
<td>Child to young adult</td>
<td>Myoclonus +, GTC</td>
<td>Variable, typically present</td>
<td>Variable, typically present</td>
<td>Ragged red fibers, can present in metabolic crisis, deafness, DM, myopathy, lipomas No VPA!</td>
</tr>
</tbody>
</table>
| Sialidoses | Vary, juvenile
II: neonate | Myoclonus ++++, later GTC | I – absent
II – learning disability | Gradual | Cherry red spot on fundus, progressive visual failure, dysmorphic |
| DRPLA   | < age 20 (for PME) | Myoclonic +, GTC | Variable, but present | variable | |

Adapted portions from Shahwan A, 2005.
Other Rare PME

- Gaucher III (glucocerebrosidase deficiency)
- SSPE (Subacute Sclerosing Panencephalitis)
- Juvenile GM2 gangliosidoses (Taysachs, Sandhoff, Hexosaminidase A&B deficiency)
- Pantothenate kinase-associated neurodegeneration (Hallervorden-Spatz)
- Ataxia-PME disease (PRICKLE1)
- Action myoclonus renal failure syndrome (SCARB2)
- SCARB2 without renal failure (ULD like)
- Neuroserpin mutations
## Management of PME

### Work-up
- MRI
- EEG
- Ophthalmology consultation
- Genetic testing

### Treatment
- **Seizure Management**
  - No VPA in MERRF
  - No PHT in ULD
  - LEV for myoclonus
  - Try ZNS, VPA, PHB, benzodiazepines
  - Avoid: LTG, PHT, CBZ, GBP, VGB, TGB
- **Rehabilitation**
- **Symptomatic, Palliation**
- **Genetic counseling**
Autosomal Dominant Partial Epilepsy with Auditory Features

- Onset: 2 to 60 years (peak teens)\(^1\)
- Isolated auditory auras or many evolve to loss of consciousness or generalized tonic clonic
- Seizure frequency low, can be reflexic
- Typically nonlesional MRI
- EEG often normal (vs temporal spikes)
- Responsive to treatment (96%)
- Genetics: RELN, LGI1 mutations\(^2\)

\(^1\)Bisulli et al, Brain 2004
\(^2\)Michelucci et al, Epilepsy & Behavior, 2017
Other Familial Temporal Lobe Epilepsy

- Familial Mesial Temporal Lobe Epilepsy (FMTLE)
  - Heterogeneous
  - Onset: later childhood to early adult
  - Psychic, abdominal auras which can progress
  - Benign prognosis and good response to meds
  - Normal MRI or mesial temporal sclerosis
  - Varied genetics, autosomal dominant or recessive

Crompton et al, Brain 2010
Reflex Epilepsy

- Seizures not spontaneous but precipitated by stimulus
  - Should be objective and consistent
- ILAE in 1989: “epilepsies characterized by specific modes of seizure precipitation”
- Tasks/stimuli activate an area that may be hyperexcitable. If a large enough volume (or network) becomes activated it can reach “critical mass” and result in a seizure.
- Generalized or focal
Reflex Epilepsy Types

- **Language or Reading Induced**: Often cause jaw jerks or visual symptoms may progress to GTC, Inherited, onset late puberty, benign, typically no spontaneous seizures.

- **Startle**: (sudden unexpected arousal), typically symptomatic to large brain lesions, often injury to sensorimotor and premotor cortex, many delayed.

**Elementary**

- **Visual**: (most common), photosensitive, pattern sensitive, eye closure sensitivity idiopathic photosensitive occipital lobe epilepsy, occipital hyperexcitability.

- **Audiogenic**: stimulus very specific, can be musicogenic (association with right temporal?), often also have spontaneous seizures.

- **Proprioceptive/Touch**: can be active or passive movement, often involves sensorimotor of contralateral hemisphere.

**Situational**

- **Thinking**: spatial or sequential, memory/pattern recognition, typically partial.

- **Bathing**

- **Praxis**: generalized, more with challenging task.

- **Eating**: Often temporolimbic, extratemporal symptomatic epilepsy.

HIGH YIELD POINTS

- Young adult with myoclonus, GTC = JME
- VPA drug of choice in GGE (teratogen)
- LTG may worsen myoclonus, LEV may help
- In GGE, no narrow spectrum CBZ, OXC, VGB, GBP, TGB, PHT
- PME can look like GGE initially
- Myoclonus, seizure, neurological deterioration, ataxia = PME
- Occipital epilepsy + PME = Lafora
- Mitochondrial + PME = MERRF; no VPA
- Cherry-red-spot + PME = Sialidosis
- Mild PME that survives to adult = ULD; no PHT
- ADLTE = LG1, RELN
References

- International League Against Epilepsy Website, https://www.epilepsydiagnosis.org/syndrome/epilepsy-syndrome-groupoverview.html
Cleveland Clinic

Every life deserves world class care.