NeuroTrace Academy Study Guide

Category: Domain III - Pattern Recognition
Topic: EEG Syndromes Classification & Diagnostic Yield Enhancement
Style: Syndrome-based, activation-focused, clinical correlation

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1. Core Principle

EEG Syndromes = Clinical + EEG Pattern + Age

Understanding EEG syndromes requires:

- **Clinical features** (seizure types, manifestations)
- **EEG patterns** (morphology, distribution, frequency)
- **Age group** (syndrome-specific age ranges)
- **Activation procedures** (how to increase diagnostic yield)

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2. Syndrome Classification

A. Idiopathic Generalized Epilepsies (IGE)

Characteristics:

- Normal neurological exam
- Normal imaging
- Genetic predisposition
- Generalized EEG patterns
- Good prognosis (usually)

Syndromes:

- Childhood Absence Epilepsy (CAE)
- Juvenile Absence Epilepsy (JAE)
- Juvenile Myoclonic Epilepsy (JME)
- Generalized Tonic-Clonic Seizures Alone (GTCA) - GTC upon awakening
- Eye Closure Sensitivity Epilepsy - seizures triggered by eye closure

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B. Idiopathic Focal Epilepsies

Characteristics:

- Normal neurological exam
- Normal imaging
- Age-related (childhood)
- Focal EEG patterns
- Excellent prognosis (remit by adolescence)

Syndromes:

- Benign Rolandic Epilepsy (BRE) / BECTS
- Panayiotopoulos Syndrome
- Other benign focal epilepsies

C. Developmental and Epileptic Encephalopathies

Characteristics:

- Abnormal neurological exam
- May have abnormal imaging
- Developmental delay/regression
- Severe EEG abnormalities
- Poor prognosis

Syndromes:

- Ohtahara Syndrome (EIEE) - earliest onset (first few days to 3 months)
- Early Myoclonic Encephalopathy (EME) - earliest onset (first few days to 3 months)
- Dravet Syndrome - first year of life (peak 6 months)
- West Syndrome (Infantile Spasms) - 3-12 months
- ESES/CSWS - 3-14 years (peak 5-7 years)
- Landau-Kleffner Syndrome - 3-8 years (peak 4-6 years)
- Lennox-Gastaut Syndrome (LGS) - 1-8 years
- Doose Syndrome (MAE)

D. Focal Epilepsies

Characteristics:

- May have abnormal neurological exam
- May have abnormal imaging
- Focal EEG patterns
- Variable prognosis

Syndromes:

- Temporal Lobe Epilepsy (TLE)
- Frontal Lobe Epilepsy (FLE)
- Parietal Lobe Epilepsy
- Occipital Lobe Epilepsy

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3. Syndrome-Specific Patterns & Diagnostic Yield

Idiopathic Generalized Epilepsy (IGE) - Overview

Classification: Idiopathic Generalized Epilepsy
Age: Childhood to young adulthood
EEG Patterns: Variable - 3 Hz spike-and-wave, polyspike-and-wave, slow spike-and-wave
Morphology:

- Generalized, symmetric, synchronous patterns
- High amplitude (200-500 μV)
- Normal background (characteristic)
- Patterns vary by specific syndrome

IGE Includes:

- **CAE** (Childhood Absence Epilepsy) - 3 Hz spike-and-wave
- **JAE** (Juvenile Absence Epilepsy) - 3-4 Hz spike-and-wave

- **JME** (Juvenile Myoclonic Epilepsy) - Polyspike-and-wave
- **Jeavons** (Absence with Eyelid Myoclonus) - 3-4 Hz spike-and-wave, photosensitive
- **GTCA** (Generalized Tonic-Clonic Seizures Alone)

Diagnostic Yield Enhancement (Variable by Syndrome):

1. **Hyperventilation** - 70-95% yield (most effective for CAE)
2. **Photic Stimulation** - 10-95% yield (most effective for Jeavons)
3. **Sleep Deprivation** - 40-95% yield (most effective for JME with awakening)
4. **Sleep** - 20-95% yield (essential for LGS/atypical absence)

Clinical Features:

- Multiple seizure types possible (absence, myoclonic, GTC)
- Normal neurological exam
- Normal imaging
- Genetic predisposition
- Age-dependent onset

Clinical Course:

- Onset: Childhood to young adulthood (varies by specific syndrome)
- Outcome: Variable - excellent for CAE, good for JAE/JME/Jeavons
- Treatment: Valproate, lamotrigine, ethosuximide, levetiracetam (varies by syndrome)
- Prognosis: Variable - excellent for typical absence, good for JME/Jeavons with treatment

🔑 **Exam Pearl:** IGE is an umbrella term - activation procedures vary by specific syndrome. Choose activation based on clinical suspicion.

🔑 **Exam Pearl:** All IGE syndromes show generalized, symmetric, synchronous patterns - distinguishing them from focal epilepsies.

🔑 **Exam Pearl:** Normal background and normal exam/imaging are characteristic of IGE - distinguishing from symptomatic generalized epilepsies.

Childhood Absence Epilepsy (CAE) - Typical Absence

Age: 4-10 years
EEG Pattern: 3 Hz generalized spike-and-wave
Morphology:

- Generalized, symmetric, synchronous
- High amplitude (200-500 μV)
- Abrupt onset/offset
- Normal background
- **Subtype:** Typical absence - no motor features

Diagnostic Yield Enhancement:

1. **Hyperventilation** - **90-95% yield** (MOST EFFECTIVE)
    - Protocol: 3-5 minutes deep breathing
    - Notes: Most effective activation procedure for CAE
2. **Sleep** - 30-40% yield
    - May show different morphology (faster frequency)
3. **Sleep Deprivation** - 40-50% yield
    - Increases likelihood of capturing events
4. **Recording Duration** - Minimum 20-30 minutes awake, include sleep

5. **Montage** - Referential (average reference) better shows generalized distribution

Clinical Features:

- Brief staring spells (5-30 seconds)
- Abrupt onset/offset
- Impaired awareness
- **NO eyelid myoclonus** (distinguishes from Jeavons)
- **NO myoclonic jerks** (distinguishes from JME)
- Multiple episodes per day (10-100+)
- Normal exam and imaging

Clinical Course:

- Onset: 4-10 years
- Outcome: 60-70% remit by adolescence
- Treatment: Ethosuximide, valproate, lamotrigine
- Prognosis: Excellent if treated early

🔑 **Exam Pearl:** Hyperventilation is the most effective activation procedure for CAE (90-95% yield).

🔑 **Exam Pearl:** Typical absence has NO motor features - distinguishes from Jeavons (eyelid myoclonus) and JME (myoclonus).

Absence with Eyelid Myoclonus (Jeavons Syndrome)

Age: 2-14 years (peak 6-8 years)
EEG Pattern: 3-4 Hz generalized spike-and-wave or polyspike-and-wave
Morphology:

- Generalized, symmetric, synchronous
- 3-4 Hz (may be faster than typical absence)
- High amplitude (200-500 μV)
- Brief duration (3-6 seconds, shorter than typical absence)
- Normal background
- **Subtype:** Absence with eyelid myoclonus - photosensitive
- **Special Feature:** Eyelid myoclonus (rhythmic jerking of eyelids)

Diagnostic Yield Enhancement:

1. **Photic Stimulation** - **90-95% yield** (MOST EFFECTIVE)
    - Protocol: Standard IPS protocol, especially 10-20 Hz
    - Notes: MOST EFFECTIVE for Jeavons. Highly photosensitive. Eye closure during IPS may trigger events.
2. **Eye Closure** - **85-90% yield** (CRITICAL)
    - Protocol: Repeated eye closure and opening
    - Notes: Eye closure often triggers eyelid myoclonus and absence. Critical activation procedure.
3. **Hyperventilation** - 50-60% yield
    - Less effective than photic stimulation for Jeavons
4. **Sleep** - 20-30% yield
    - Less effective than photic stimulation
5. **Recording Duration** - Minimum 20-30 minutes awake, include photic stimulation and eye closure
6. **Montage** - Referential (average reference). May need EOG channels to see eyelid myoclonus.
7. **Special Note:** Photic stimulation and eye closure are CRITICAL - Jeavons is highly photosensitive

Clinical Features:

- **Eyelid myoclonus (rhythmic jerking of eyelids) - MOST CHARACTERISTIC**
- Brief absence seizures (3-6 seconds, shorter than typical absence)
- Often triggered by eye closure
- Often triggered by photic stimulation (highly photosensitive)
- Upward deviation of eyes during eyelid myoclonus
- Impaired awareness during absence
- May have brief myoclonic jerks of upper extremities
- Normal exam and imaging

Clinical Course:

- Onset: 2-14 years (peak 6-8 years)
- Outcome: Variable - may remit or persist
- Treatment: Valproate (first line), lamotrigine, levetiracetam, ethosuximide. Avoid photosensitivity triggers.
- Prognosis: Good with treatment, but photosensitivity may persist

🔑 **Exam Pearl:** Photic stimulation is the most effective activation procedure for Jeavons (90-95% yield) - highly photosensitive.

🔑 **Exam Pearl:** Eye closure often triggers eyelid myoclonus and absence - repeated eye closure testing is critical.

🔑 **Exam Pearl:** Eyelid myoclonus (rhythmic jerking of eyelids) is the most characteristic feature - distinguishes from typical absence.

🔑 **Exam Pearl:** Jeavons has shorter absence duration (3-6 seconds) compared to typical absence (5-30 seconds).

Atypical Absence Seizures

Age: 1-8 years (typically in LGS context)
EEG Pattern: Slow spike-and-wave (1.5-2.5 Hz), GPFA
Morphology:

- Interictal: Slow spike-and-wave (1.5-2.5 Hz), GPFA, abnormal background
- Ictal: Slow spike-and-wave (1.5-2.5 Hz), gradual onset/offset, may be asymmetric
- High amplitude (200-500 μV)
- Longer duration (10-60+ seconds, longer than typical absence)
- **Subtype:** Atypical absence - gradual onset/offset, slower frequency
- **Special Feature:** Gradual onset and offset (unlike abrupt typical absence), abnormal background

Diagnostic Yield Enhancement:

1. **Sleep** - **95-100% yield** (MOST EFFECTIVE)
    - Protocol: Natural sleep, especially NREM sleep
    - Notes: Slow spike-and-wave and GPFA are most prominent during sleep. Essential to record sleep.
2. **Awake** - 80-90% yield
    - Slow spike-and-wave present but may be less frequent
3. **Hyperventilation** - 20-30% yield
    - Less effective than in typical absence
4. **Photic Stimulation** - 10-20% yield
    - Less effective than in typical absence
5. **Recording Duration** - Minimum 1-2 hours, must include sleep
6. **Montage** - Referential (average reference) better shows generalized distribution
7. **Special Note:** Sleep recording is CRITICAL - slow spike-and-wave and GPFA are most prominent during sleep

Clinical Features:

- **Gradual onset and offset** (unlike abrupt typical absence)

- Longer duration (10-60+ seconds, longer than typical absence)
- Impaired awareness (may be incomplete)
- May have automatisms
- May have atonic components (head drop, loss of tone)
- May have myoclonic components
- Often occurs in context of LGS
- **Cognitive impairment** (unlike typical absence)

Clinical Course:

- Onset: 1-8 years (typically in LGS context)
- Outcome: Poor - resistant to treatment, persistent seizures
- Treatment: Valproate, lamotrigine, rufinamide, felbamate, ketogenic diet, VNS (same as LGS)
- Prognosis: Poor - part of LGS syndrome, persistent seizures, cognitive impairment

🔑 **Exam Pearl:** Atypical absence has gradual onset/offset (unlike abrupt typical absence) - key distinguishing feature.

🔑 **Exam Pearl:** Atypical absence has slower frequency (1.5-2.5 Hz) compared to typical absence (3 Hz).

🔑 **Exam Pearl:** Atypical absence occurs in context of LGS with cognitive impairment - distinguishing it from typical absence.

Juvenile Myoclonic Epilepsy (JME)

Age: 12-18 years
EEG Pattern: Polyspike-and-wave, 4-6 Hz spike-and-wave
Morphology:

- Generalized, symmetric, synchronous
- Polyspike-and-wave (more common than pure 3 Hz)
- 4-6 Hz (faster than CAE)
- High amplitude (200-500 μV)
- Normal background

Diagnostic Yield Enhancement:

1. **Sleep Deprivation + Awakening** - **90-95% yield** (MOST EFFECTIVE)
    - Protocol: 4-6 hours sleep deprivation, record upon awakening
    - Notes: Myoclonus often occurs upon awakening - CRITICAL
2. **Sleep** - 70-80% yield
    - Especially upon awakening from sleep
3. **Photic Stimulation** - 30-40% yield
    - Especially 15-20 Hz in photosensitive patients
4. **Recording Duration** - Include sleep and awakening, minimum 30 minutes
5. **Montage** - Referential (average reference) better shows generalized distribution
6. **Special Note:** Record upon awakening - myoclonus is most common at this time

Clinical Features:

- Myoclonic jerks upon awakening (most characteristic)
- Bilateral, symmetric jerks
- Upper extremities more common
- Generalized tonic-clonic seizures (often upon awakening)
- Normal exam and imaging

Clinical Course:

- Onset: 12-18 years
- Outcome: Lifelong condition, rarely remits
- Treatment: Valproate (first line), lamotrigine, levetiracetam
- Prognosis: Good with treatment, but requires lifelong medication

🔑 **Exam Pearl:** Sleep deprivation with recording upon awakening is the most effective activation procedure for JME (90-95% yield).

Ohtahara Syndrome (Early Infantile Epileptic Encephalopathy - EIEE)

Age: First few days to 3 months (peak: first 2 weeks)
EEG Pattern: Burst-suppression pattern
Morphology:

- Interictal: Burst-suppression pattern - alternating bursts and suppression
- Ictal: Burst-suppression pattern, tonic seizures show bursts or electrodecremental pattern
- Very high amplitude bursts (200-1000+ μV)
- Suppression periods near isoelectric
- Generalized, may be asymmetric
- **CRITICAL:** Pattern is CONTINUOUS (wake and sleep)

Diagnostic Yield Enhancement:

1. **Awake** - **95-100% yield** (CRITICAL)
    - Protocol: Awake recording
    - Notes: Burst-suppression pattern is continuous and present in both wake and sleep - CRITICAL for diagnosis
2. **Sleep** - **95-100% yield** (CRITICAL)
    - Protocol: Natural sleep
    - Notes: Burst-suppression pattern persists in sleep (unlike normal trace alternant which is sleep-only)
3. **Ictal Recording** - Critical for diagnosis
    - Protocol: Video-EEG with clinical correlation
    - Notes: Tonic seizures correlate with bursts or electrodecremental pattern
4. **Recording Duration** - Minimum 1-2 hours, include both wake and sleep
5. **Montage** - Referential (average reference) better shows generalized distribution
6. **Video Recording** - Essential - must correlate EEG with clinical tonic seizures
7. **Special Note:** Burst-suppression pattern is CONTINUOUS (wake and sleep) - distinguishing it from normal neonatal trace alternant (sleep only)

Clinical Features:

- Tonic seizures (most characteristic) - brief, frequent
- Onset in first few days to 3 months (peak: first 2 weeks)
- Severe developmental delay
- Poor prognosis
- May evolve to West Syndrome (hypsarrhythmia) or LGS

Clinical Course:

- Onset: Neonatal period (first few days to 3 months, peak: first 2 weeks)
- Progression: May evolve to West Syndrome around 3-6 months, or to LGS
- Outcome: Very poor - severe developmental delay, high mortality
- Treatment: ACTH, vigabatrin, ketogenic diet, antiepileptic drugs
- Prognosis: Very poor - severe developmental delay, high mortality, persistent seizures

🔑 **Exam Pearl:** Ohtahara syndrome shows burst-suppression pattern that is CONTINUOUS (present in both wake and sleep) - distinguishing it from normal neonatal trace alternant (sleep only).

🔑 **Exam Pearl:** Onset in first few days to 3 months (peak: first 2 weeks) - earliest onset of epileptic encephalopathies.

🔑 **Exam Pearl:** Tonic seizures are most characteristic - brief, frequent, correlate with bursts or electrodecremental pattern.

🔑 **Exam Pearl:** May evolve to West Syndrome (hypsarrhythmia) around 3-6 months, or to LGS - part of epileptic encephalopathy spectrum.

Early Myoclonic Encephalopathy (EME)

Age: First few days to 3 months (peak: first week)
EEG Pattern: Burst-suppression pattern
Morphology:

- Interictal: Burst-suppression pattern - alternating bursts and suppression
- Ictal: Burst-suppression pattern, myoclonic seizures may show bursts or spikes
- Very high amplitude bursts (200-1000+ μV)
- Suppression periods near isoelectric
- Generalized, may be asymmetric or multifocal
- **CRITICAL:** Pattern is CONTINUOUS (wake and sleep)

Diagnostic Yield Enhancement:

1. **Awake** - **95-100% yield** (CRITICAL)
    - Protocol: Awake recording
    - Notes: Burst-suppression pattern is continuous and present in both wake and sleep - CRITICAL for diagnosis
2. **Sleep** - **95-100% yield** (CRITICAL)
    - Protocol: Natural sleep
    - Notes: Burst-suppression pattern persists in sleep
3. **Ictal Recording** - Critical for diagnosis
    - Protocol: Video-EEG with clinical correlation
    - Notes: Myoclonic seizures correlate with bursts or spikes. Must correlate with clinical myoclonus.
4. **Recording Duration** - Minimum 1-2 hours, include both wake and sleep
5. **Montage** - Referential (average reference). May need EMG channels to see myoclonus.
6. **Video Recording** - Essential - must correlate EEG bursts/spikes with clinical myoclonic jerks
7. **Special Note:** Burst-suppression pattern is CONTINUOUS (wake and sleep). Myoclonic seizures are most characteristic. Often associated with metabolic disorders.

Clinical Features:

- Myoclonic seizures (most characteristic) - fragmentary, erratic, multifocal
- Onset in first few days to 3 months (peak: first week)
- Severe developmental delay
- Poor prognosis
- Often associated with metabolic disorders (non-ketotic hyperglycinemia, etc.)
- May have erratic eye movements

Clinical Course:

- Onset: Neonatal period (first few days to 3 months, peak: first week)
- Progression: May persist or evolve to other patterns, rarely evolves to West Syndrome
- Outcome: Very poor - severe developmental delay, high mortality
- Treatment: Address underlying metabolic disorder if present, antiepileptic drugs, ketogenic diet

- Prognosis: Very poor - severe developmental delay, high mortality, persistent seizures

🔑 **Exam Pearl:** EME shows burst-suppression pattern with MYOCLONIC SEIZURES (most characteristic) - distinguishing it from Ohtahara (tonic seizures).

🔑 **Exam Pearl:** Myoclonic seizures are fragmentary, erratic, and multifocal - characteristic of EME.

🔑 **Exam Pearl:** Often associated with metabolic disorders (non-ketotic hyperglycinemia) - check metabolic workup.

🔑 **Exam Pearl:** EME vs Ohtahara: EME has myoclonic seizures; Ohtahara has tonic seizures. Both have burst-suppression pattern.

Dravet Syndrome (Severe Myoclonic Epilepsy of Infancy - SMEI)

Age: First year of life (peak 6 months), persists into childhood
EEG Pattern: Variable - generalized spike-and-wave, polyspike-and-wave, multifocal spikes
Morphology:

- Early: may be normal
- Later: generalized spike-and-wave, polyspike-and-wave, multifocal spikes
- Photosensitive (pattern triggered by photic stimulation)
- Variable frequency (2-5 Hz)
- High amplitude (200-500 μV)
- Generalized, may be asymmetric, may be multifocal

Diagnostic Yield Enhancement:

1. **Photic Stimulation** - **60-80% yield** (HIGH)
    - Protocol: Standard IPS protocol
    - Notes: Dravet is photosensitive - photic stimulation may trigger seizures or EEG abnormalities
2. **Sleep** - 70-80% yield
    - Protocol: Natural sleep
    - Notes: May show generalized spike-and-wave, polyspike-and-wave during sleep
3. **Awake** - 50-70% yield
    - May show generalized or multifocal spikes
4. **Ictal Recording** - Critical for diagnosis
    - Protocol: Video-EEG with clinical correlation
    - Notes: Must correlate EEG with clinical seizures (febrile, myoclonic, focal, GTC)
5. **Recording Duration** - Minimum 1-2 hours, include sleep
6. **Montage** - Referential (average reference) better shows generalized distribution
7. **Video Recording** - Essential - must correlate EEG with multiple seizure types
8. **Special Note:** Dravet is photosensitive and fever-sensitive. Photic stimulation may trigger seizures. Early EEG may be normal, pattern evolves over time.

Clinical Features:

- Febrile seizures in first year (most characteristic early feature)
- Myoclonic seizures (develop later)
- Focal seizures (hemiclonic, unilateral)
- Generalized tonic-clonic seizures
- Status epilepticus (common, may be prolonged)
- Developmental delay and regression
- Ataxia, hypotonia
- Photosensitive (seizures triggered by lights, patterns)

- Fever-sensitive (seizures triggered by fever, illness)

Clinical Course:

- Onset: First year of life (peak 6 months) - febrile seizures
- Progression: Chronic, persistent, may worsen over time
- Outcome: Poor - resistant to treatment, persistent seizures, developmental delay
- Treatment: Valproate, clobazam, stiripentol, topiramate, ketogenic diet, fenfluramine, cannabidiol. Avoid lamotrigine (may worsen).
- Prognosis: Poor - persistent seizures, developmental delay, cognitive impairment, high mortality risk

🔑 **Exam Pearl:** Dravet syndrome begins with febrile seizures in first year of life (peak 6 months) - this is the most characteristic early feature.

🔑 **Exam Pearl:** Dravet is photosensitive - photic stimulation may trigger seizures or EEG abnormalities (60-80% yield).

🔑 **Exam Pearl:** Dravet is fever-sensitive - seizures triggered by fever, hot baths, illness. Avoid fever triggers.

🔑 **Exam Pearl:** Early EEG may be normal - pattern evolves over time to show generalized spike-and-wave, polyspike-and-wave, multifocal spikes.

🔑 **Exam Pearl:** Status epilepticus is common in Dravet - may be prolonged and difficult to control.

West Syndrome (Infantile Spasms)

Age: 3-12 months (peak 4-6 months)
EEG Pattern: Hypsarrhythmia
Morphology:

- Interictal: Chaotic high-amplitude slow waves with multifocal spikes
- Ictal: Electrodecremental pattern (generalized attenuation)
- Very high amplitude (200-1000+ μV)
- Generalized, asymmetric, multifocal
- Brief (0.5-2 seconds for spasms)

Diagnostic Yield Enhancement:

1. **Sleep** - **95-100% yield** (MOST EFFECTIVE)
    - Protocol: Natural sleep, especially NREM sleep
    - Notes: Hypsarrhythmia is most prominent during sleep - ESSENTIAL
2. **Awake** - 80-90% yield
    - Hypsarrhythmia present but may be less prominent
3. **Ictal Recording** - Critical for diagnosis
    - Protocol: Video-EEG with clinical correlation
    - Notes: Electrodecremental pattern during spasms
4. **Recording Duration** - Minimum 1 hour, must include sleep
5. **Montage** - Referential (average reference) better shows multifocal distribution
6. **Video Recording** - Essential - must correlate EEG with clinical spasms
7. **Special Note:** Sleep recording is CRITICAL - hypsarrhythmia is most prominent during sleep

Clinical Features:

- Clusters of epileptic spasms
- Flexor, extensor, or mixed
- Brief (0.5-2 seconds)
- Occur in clusters (5-50+ per cluster)
- Often upon awakening

- Developmental delay or regression

Clinical Course:

- Onset: 3-12 months (peak 4-6 months)
- Outcome: Variable - depends on underlying cause
- Treatment: ACTH, vigabatrin, prednisolone, ketogenic diet
- Prognosis: Variable - poor if symptomatic, better if cryptogenic

🔑 **Exam Pearl:** Hypsarrhythmia is most prominent during sleep - sleep recording is CRITICAL for diagnosis.

🔑 **Exam Pearl:** West Syndrome = Infantile Spasms + Hypsarrhythmia + Developmental Delay/Regression (triad).

Lennox-Gastaut Syndrome (LGS)

Age: 1-8 years (peak 3-5 years)
EEG Pattern: Slow spike-and-wave (1.5-2.5 Hz), GPFA
Morphology:

- Interictal: Slow spike-and-wave (1.5-2.5 Hz), generalized paroxysmal fast activity (GPFA)
- Ictal: Slow spike-and-wave, tonic seizures show GPFA or electrodecremental pattern
- High amplitude (200-500 μV)
- Generalized, may be asymmetric
- Variable duration

Diagnostic Yield Enhancement:

1. **Sleep** - **95-100% yield** (MOST EFFECTIVE)
    - Protocol: Natural sleep, especially NREM sleep
    - Notes: Slow spike-and-wave and GPFA are most prominent during sleep - ESSENTIAL
2. **Awake** - 80-90% yield
    - Slow spike-and-wave present but may be less frequent
3. **Ictal Recording** - Critical for diagnosis
    - Protocol: Video-EEG with clinical correlation
    - Notes: GPFA or electrodecremental pattern during tonic seizures
4. **Recording Duration** - Minimum 1-2 hours, must include sleep
5. **Montage** - Referential (average reference) better shows generalized distribution
6. **Video Recording** - Essential - must correlate EEG with multiple seizure types
7. **Special Note:** Sleep recording is CRITICAL - slow spike-and-wave and GPFA are most prominent during sleep

Clinical Features:

- Multiple seizure types (characteristic)
- Tonic seizures (most characteristic) - brief, often during sleep
- Atypical absence - gradual onset/offset, longer than typical absence
- Atonic seizures - drop attacks (sudden loss of tone, falls)
- Status epilepticus - may occur (non-convulsive or convulsive)
- Cognitive impairment
- Resistant to treatment

Clinical Course:

- Onset: 1-8 years (peak 3-5 years)
- Outcome: Poor - resistant to treatment, persistent seizures
- Treatment: Valproate, lamotrigine, rufinamide, felbamate, ketogenic diet, VNS

- Prognosis: Poor - persistent seizures, cognitive impairment

🔑 **Exam Pearl:** Slow spike-and-wave (1.5-2.5 Hz) is characteristic of LGS - slower than typical 3 Hz spike-and-wave.

🔑 **Exam Pearl:** GPFA (generalized paroxysmal fast activity) during sleep is highly characteristic of LGS.

🔑 **Exam Pearl:** Multiple seizure types + slow spike-and-wave + cognitive impairment = LGS triad.

🔑 **Exam Pearl:** Drop attacks (atonic seizures) are common in LGS - sudden loss of tone causing falls.

🔑 **Exam Pearl:** Status epilepticus may occur in LGS - both non-convulsive (atypical absence status) and convulsive forms.

Benign Rolandic Epilepsy (BRE) / BECTS

Age: 3-13 years (peak 7-10 years)
EEG Pattern: Centrotemporal spikes
Morphology:

- Interictal: Centrotemporal spikes (C3, C4, T3, T4) - high amplitude, diphasic
- Ictal: Focal onset in centrotemporal region, may spread
- High amplitude (100-300 μV)
- Focal - centrotemporal (C3, C4, T3, T4), may be bilateral independent
- Brief (seconds to minutes)

Diagnostic Yield Enhancement:

1. **Sleep** - **95-100% yield** (MOST EFFECTIVE)
    - Protocol: Natural sleep, especially NREM sleep
    - Notes: Centrotemporal spikes are dramatically activated by sleep (10-100x increase) - ESSENTIAL
2. **Awake** - 40-60% yield
    - Spikes may be present but less frequent
3. **Sleep Deprivation** - 80-90% yield
    - Increases spike frequency
4. **Recording Duration** - Minimum 30 minutes, must include sleep
5. **Montage** - Bipolar (longitudinal and transverse) better localizes spikes
6. **Special Note:** Sleep recording is CRITICAL - centrotemporal spikes are dramatically activated by sleep (10-100x increase)

Clinical Features:

- Facial twitching (most characteristic)
- Speech arrest
- Drooling
- Sensory symptoms (tingling in face/tongue)
- Often occur during sleep (nocturnal)
- Normal exam and imaging

Clinical Course:

- Onset: 3-13 years (peak 7-10 years)
- Outcome: Excellent - remits by age 13-16 years
- Treatment: May not require treatment if infrequent
- Prognosis: Excellent - complete remission by adolescence

🔑 **Exam Pearl:** Centrotemporal spikes are dramatically activated by sleep (10-100x increase) - sleep recording is CRITICAL.

Temporal Lobe Epilepsy (TLE)

Age: Adolescence to adulthood
EEG Pattern: Temporal spikes/sharp waves, temporal slowing
Morphology:

- Interictal: Temporal spikes/sharp waves, temporal slowing
- Ictal: Focal onset in temporal region, rhythmic theta/delta evolving to faster frequencies
- Variable amplitude (50-200 μV)
- Focal - temporal (T3, T4, T5, T6, F7, F8), may be mesial or lateral
- Variable duration (30 seconds to minutes)

Diagnostic Yield Enhancement:

1. **Sleep** - 70-80% yield
    - Protocol: Natural sleep, especially NREM sleep
    - Notes: Temporal spikes are activated by sleep
2. **Awake** - 50-60% yield
    - Spikes may be present but less frequent
3. **Sleep Deprivation** - 70-80% yield
    - Increases spike frequency
4. **Ictal Recording** - Critical for localization
    - Protocol: Video-EEG with clinical correlation
    - Notes: Ictal onset pattern is critical for localization
5. **Recording Duration** - Minimum 1-2 hours, must include sleep
6. **Montage** - Bipolar (longitudinal and transverse), referential for field analysis
7. **Video Recording** - Essential for ictal recording

Clinical Features:

- Aura (epigastric, déjà vu, fear, olfactory)
- Staring, unresponsiveness
- Automatisms (lip smacking, fumbling, picking)
- May have dystonic posturing
- Postictal confusion
- May show hippocampal sclerosis on MRI

Clinical Course:

- Onset: Adolescence to adulthood
- Outcome: Variable - may be controlled with medication or require surgery
- Treatment: Carbamazepine, oxcarbazepine, levetiracetam, lamotrigine. Surgery if medication-resistant.
- Prognosis: Variable - good if medication-responsive, excellent if surgery-responsive

🔑 **Exam Pearl:** Temporal spikes are activated by sleep - sleep recording increases yield.

🔑 **Exam Pearl:** Ictal onset pattern (rhythmic theta/delta evolving to faster frequencies) is critical for localization.

Frontal Lobe Epilepsy (FLE)

Age: All ages
EEG Pattern: Frontal spikes/sharp waves, frontal slowing
Morphology:

- Interictal: Frontal spikes/sharp waves, frontal slowing
- Ictal: Focal onset in frontal region, rapid evolution, may be brief
- Variable amplitude (50-200 μV)
- Focal - frontal (F3, F4, Fz, F7, F8), may be mesial or lateral
- Often brief (seconds to <1 minute)

Diagnostic Yield Enhancement:

1. **Sleep** - 70-80% yield
    - Protocol: Natural sleep, especially NREM sleep
    - Notes: Frontal spikes are activated by sleep
2. **Awake** - 50-60% yield
    - Spikes may be present but less frequent
3. **Sleep Deprivation** - 70-80% yield
    - Increases spike frequency
4. **Ictal Recording** - Critical for localization
    - Protocol: Video-EEG with clinical correlation
    - Notes: Seizures may be brief and frequent
5. **Recording Duration** - Minimum 1-2 hours, must include sleep
6. **Montage** - Bipolar (longitudinal and transverse), referential for field analysis. May need additional frontal electrodes.
7. **Video Recording** - Essential - seizures may be brief

Clinical Features:

- Brief seizures (seconds to <1 minute)
- Frequent (multiple per day)
- Motor manifestations (tonic, clonic, hypermotor)
- May have vocalizations
- Often occur during sleep
- Rapid recovery, minimal postictal confusion

Clinical Course:

- Onset: All ages
- Outcome: Variable - depends on underlying cause
- Treatment: Carbamazepine, oxcarbazepine, levetiracetam, lamotrigine. Surgery if medication-resistant.
- Prognosis: Variable - good if medication-responsive, excellent if surgery-responsive

🔑 **Exam Pearl:** Frontal spikes are activated by sleep - sleep recording increases yield.

🔑 **Exam Pearl:** Frontal seizures are often brief and frequent - longer recordings may be needed.

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4. Diagnostic Yield Enhancement Summary Table

Syndrome	Most Effective Activation	Yield	Critical Notes
CAE (Typical Absence)	Hyperventilation	90-95%	Most effective for CAE
JAE (Typical Absence)	Hyperventilation	70-80%	Less effective than CAE
Jeavons (Absence + Eyelid Myoclonus)	Photic Stimulation	90-95%	Highly photosensitive, eye closure also critical (85-90%)

Atypical Absence (LGS)	Sleep	95-100%	Slow spike-and-wave most prominent during sleep
JME	Sleep Deprivation + Awakening	90-95%	Record upon awakening - critical
Ohtahara Syndrome	Awake + Sleep	95-100%	Burst-suppression pattern continuous (wake and sleep)
EME	Awake + Sleep	95-100%	Burst-suppression pattern continuous (wake and sleep)
Dravet Syndrome	Photic Stimulation	60-80%	Photosensitive - photic may trigger seizures
West Syndrome	Sleep	95-100%	Hypsarrhythmia most prominent during sleep
LGS	Sleep	95-100%	Slow spike-and-wave and GPFA most prominent during sleep
ESES/CSWS	Sleep	95-100%	ESES pattern ONLY in NREM sleep (≥85% of slow-wave sleep)
Landau-Kleffner	Sleep	95-100%	ESES pattern ONLY in NREM sleep (≥85% of slow-wave sleep)
BRE	Sleep	95-100%	Centrotemporal spikes dramatically activated (10-100x)
TLE	Sleep + Sleep Deprivation	70-80%	Sleep activates temporal spikes
FLE	Sleep + Sleep Deprivation	70-80%	Sleep activates frontal spikes

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5. Morphological Pattern Library Links

Generalized Patterns

- **3 Hz Spike-and-Wave** → `pattern_three_hz_gsw` → `/images/patterns/3-hz-generalized-spike-wave.png`
- **Slow Spike-and-Wave (1.5-2.5 Hz)** → `pattern_slow_spike_wave` → `/images/patterns/slow-spike-wave.png`
- **Polyspike-and-Wave** → `pattern_polyspike_wave` → `/images/patterns/polyspike-wave.png`
- **Hypsarrhythmia** → `pattern_hypsarrhythmia` → `/images/patterns/hypsarrhythmia.png`
- **GPFA** → `pattern_gpfa` → `/images/patterns/gpfa.png`

Focal Patterns

- **Centrotemporal Spikes** → `pattern_focal_spikes_temporal` → `/images/patterns/focal-spikes-temporal.png`
- **Temporal Spikes** → `pattern_focal_spikes_temporal` → `/images/patterns/focal-spikes-temporal.png`
- **Frontal Spikes** → `pattern_focal_spikes_frontal` → `/images/patterns/focal-spikes-frontal.png`
- **Temporal Slowing** → `pattern_focal_slowing` → `/images/patterns/focal-slowing.png`
- **Frontal Slowing** → `pattern_focal_slowing` → `/images/patterns/focal-slowing.png`

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6. Clinical Course & Manifestation Summary

Idiopathic Generalized Epilepsies

- **CAE:** Excellent prognosis, 60-70% remit by adolescence
- **JAE:** Good prognosis, may remit or persist
- **JME:** Good with treatment, but requires lifelong medication

Idiopathic Focal Epilepsies

- **BRE:** Excellent prognosis, complete remission by adolescence

Developmental and Epileptic Encephalopathies

- **West Syndrome:** Variable - poor if symptomatic, better if cryptogenic
- **LGS:** Poor - persistent seizures, cognitive impairment

Focal Epilepsies

- **TLE:** Variable - good if medication-responsive, excellent if surgery-responsive
- **FLE:** Variable - depends on underlying cause

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7. ABRET Exam Pearls

Typical Absence (CAE/JAE)

1. **Hyperventilation** is the most effective activation procedure for CAE (90-95% yield).
2. **Typical absence has NO motor features** - distinguishes from Jeavons (eyelid myoclonus) and JME (myoclonus).
3. **3 Hz spike-and-wave** is generalized, symmetric, and synchronous - distinguishing it from focal discharges.

Absence with Eyelid Myoclonus (Jeavons)

4. **Photic stimulation** is the most effective activation procedure for Jeavons (90-95% yield) - highly photosensitive.
5. **Eye closure** often triggers eyelid myoclonus and absence (85-90% yield) - repeated eye closure testing is critical.
6. **Eyelid myoclonus** (rhythmic jerking of eyelids) is the most characteristic feature - distinguishes from typical absence.
7. **Jeavons has shorter absence duration** (3-6 seconds) compared to typical absence (5-30 seconds).

Atypical Absence

8. **Atypical absence has gradual onset/offset** (unlike abrupt typical absence) - key distinguishing feature.
9. **Atypical absence has slower frequency** (1.5-2.5 Hz) compared to typical absence (3 Hz).
10. **Atypical absence occurs in context of LGS** with cognitive impairment - distinguishing it from typical absence.
11. **Sleep recording is CRITICAL** - slow spike-and-wave is most prominent during sleep.

Neonatal/Infantile Encephalopathies

12. **Ohtahara syndrome** shows burst-suppression pattern that is CONTINUOUS (wake and sleep) - distinguishing it from normal neonatal trace alternant (sleep only).
13. **EME** shows burst-suppression pattern with MYOCLONIC SEIZURES - distinguishing it from Ohtahara (tonic seizures).
14. **Burst-suppression pattern** in Ohtahara/EME is continuous and pathologic - not just during sleep like normal trace alternant.
15. **Dravet syndrome** begins with febrile seizures in first year (peak 6 months) - photosensitive and fever-sensitive.
16. **Onset timing:** Ohtahara/EME (first few days to 3 months) → Dravet (first year) → West Syndrome (3-12 months) → LGS (1-8 years) - epileptic encephalopathy spectrum.

ESES/CSWS and Landau-Kleffner

17. **ESES pattern** is ONLY seen during NREM sleep (≥85% of slow-wave sleep) - sleep recording is CRITICAL for diagnosis.
18. **ESES pattern** disappears in REM sleep and wakefulness - must record full sleep cycle to see this pattern.
19. **Landau-Kleffner Syndrome** = ESES pattern + ACQUIRED APHASIA (language loss) - this is the defining clinical feature.
20. **Paradoxical in LKS:** Seizures may be infrequent or absent, but EEG shows continuous spike-and-wave during sleep (ESES).

Other Important Syndromes

16. **Sleep deprivation + awakening** is the most effective activation procedure for JME (90-95% yield).
17. **Sleep recording** is CRITICAL for West Syndrome, LGS, and BRE (95-100% yield).
18. **Centrotemporal spikes** are dramatically activated by sleep (10-100x increase) in BRE.
19. **Slow spike-and-wave (1.5-2.5 Hz)** is characteristic of LGS - slower than typical 3 Hz.
20. **GPFA during sleep** is highly characteristic of LGS.
21. **Multiple seizure types + slow spike-and-wave + cognitive impairment = LGS triad.**
22. **Drop attacks (atonic seizures)** are common in LGS - sudden loss of tone causing falls.
23. **Status epilepticus** may occur in LGS - both non-convulsive (atypical absence status) and convulsive forms.
24. **West Syndrome = Infantile Spasms + Hypsarrhythmia + Developmental Delay/Regression (triad).**
25. **GTCA (Epilepsy with GTC Alone)** = GTC seizures ONLY (no absence, no myoclonus) - often occur upon awakening.
26. **BMEI (Benign Myoclonic Epilepsy of Infancy)** = Myoclonic seizures ONLY, normal development, excellent prognosis - distinguishes from Dravet (severe, developmental delay).
27. **Doose Syndrome (MAE)** = Myoclonic-astatic seizures (myoclonic jerk followed by atonic drop) - variable prognosis, may remit or evolve to LGS.
28. **Eye Closure Sensitivity Epilepsy** = Seizures triggered by eye closure (85-95% yield with eye closure testing) - may be associated with Jeavons.
29. **Angelman Syndrome** = Genetic condition (15q11-13) with happy demeanor, ataxia, developmental delay, epilepsy - avoid carbamazepine.
30. **Rett Syndrome** = Genetic condition (MECP2) with hand wringing, developmental regression, epilepsy - avoid carbamazepine.
31. **Lissencephaly/Band Heterotopia** = Structural brain malformation (smooth brain or subcortical band) with epilepsy - structural imaging essential for diagnosis.

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8. Quick Reference: Syndrome Activation Priorities

Priority 1: Awake + Sleep Recording (CRITICAL)

- Ohtahara Syndrome (burst-suppression continuous)
- EME (burst-suppression continuous)

Priority 2: Sleep Recording (CRITICAL)

- West Syndrome
- LGS
- Atypical Absence (LGS)
- BRE
- ESES/CSWS (ESES pattern only in sleep)
- Landau-Kleffner Syndrome (ESES pattern only in sleep)

Priority 2: Hyperventilation

- CAE (Typical Absence)
- JAE (Typical Absence)

Priority 3: Photic Stimulation + Eye Closure (CRITICAL)

- Jeavons (Absence with Eyelid Myoclonus)

Priority 4: Sleep Deprivation + Awakening

- JME

Priority 5: Sleep + Sleep Deprivation

- TLE
- FLE

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9. Absence Subtypes Comparison Table

Feature	Typical Absence (CAE/JAE)	Absence with Eyelid Myoclonus (Jeavons)	Atypical Absence (LGS)
Age	4-17 years	2-14 years (peak 6-8)	1-8 years
Frequency	3-4 Hz	3-4 Hz	1.5-2.5 Hz (slower)
Onset/Offset	Abrupt	Abrupt	Gradual
Duration	5-30 seconds	3-6 seconds (shorter)	10-60+ seconds (longer)
Motor Features	None	Eyelid myoclonus	May have atonic/myoclonic
Photosensitivity	Low	Very high (90-95% yield)	Low
Best Activation	Hyperventilation	Photic stimulation + Eye closure	Sleep
Background	Normal	Normal	Abnormal
Cognition	Normal	Normal	Impaired (LGS)
Prognosis	Excellent/Good	Good	Poor (LGS)

Next Steps:

- Memorize syndrome-specific activation procedures and yields
- Understand morphological patterns for each syndrome
- Learn clinical features and course
- Practice correlating EEG patterns with clinical syndromes