



# Epilepsy in Children & Adolescents: How to Evaluate, Medicate & Educate

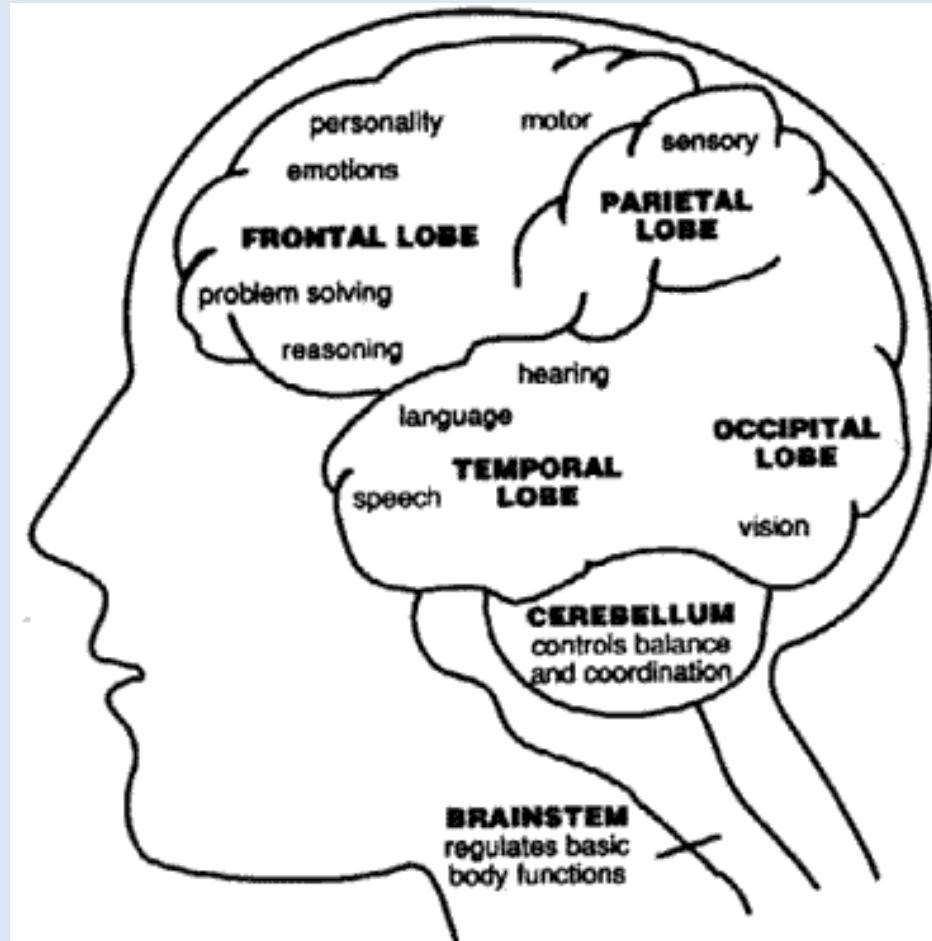
California Association of  
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# Seizures and Epilepsy

- Seizure:
  - Caused by abnormal brain electrical activity
  - Brain's electrical system malfunctions:
    - Uncontrolled electrical discharges
    - Brain cells keep firing causing abnormal surge of energy
- Seizure physical characteristics depend upon what part of the brain is involved

# The Brain



# Seizures and Epilepsy

- Seizure:
  - Most common paroxysmal disorder of infancy and childhood
- 25,000-40,000 children diagnosed with seizures yearly

# Seizures and Epilepsy

- Epilepsy: two or more *afebrile* seizures
- Status Epilepticus:
  - > 30 minutes of continuous seizing
  - Sequential seizures without recovery
  - Can be life threatening or permanently disabling
  - Represents 1/3 of initial epilepsy presentations
  - 20% of epilepsy patients experience status epilepticus within the first 5 years of diagnosis

# Old Terminology: Simple Partial Seizures

- Sometimes accompanied by an aura
- No alteration of consciousness
- Motor movements are unilateral
- Somatosensory symptoms: unilateral
- Autonomic changes:
  - Sweating, tastes, blood pressure, heart and breathing rates, body temperature, digestion

# Old Terminology: Complex Partial Seizures

- Alteration of Consciousness
- Involves one side of the body
- Unilateral motor movements
- Can spread to opposite side of the brain = **Secondarily generalization**
  - motor movements are bilateral
- **Simple → Complex → Secondarily Generalized**

# Old Classification Terminology: Generalized Seizures

- Absence: Very brief pauses
- Myoclonic: Jerking
- Tonic: Stiffening
- Atonic: Drop attacks
- Clonic: Rhythmic jerking or twitching
- Tonic-Clonic: Stiffening & jerking



# Old Terminology: Etiology of Seizures

- Provoked: A result of an acute condition
  - head trauma, central nervous system infection, tumor, hypoxia (lack of oxygen), metabolic imbalance
- Unprovoked
  - Symptomatic
    - Pre-existing brain problem: (malformation, prior injury or insult, etc.)
  - Cryptogenic (no known cause)
  - Idiopathic (genetic)
    - Healthy developmentally normal children, normal brains, inherited trait

# New Terminology (ILAE, 2010)

- Mode of Seizure Onset
- Classification of Seizures
- Syndrome or Non-Syndrome
- Underlying Cause (Etiology)
- ILAE Report & outline of changes:

[http://www.ilae.org/Visitors/Centre/ctf/documents/ClassificationReport\\_2010\\_000.pdf](http://www.ilae.org/Visitors/Centre/ctf/documents/ClassificationReport_2010_000.pdf)

[http://www.ilae.org/Visitors/Centre/ctf/documents/ILAEHandoutV10\\_000.pdf](http://www.ilae.org/Visitors/Centre/ctf/documents/ILAEHandoutV10_000.pdf)

# New Terminology: Mode of Onset

## New

- Focal seizure
- Generalized seizure
- Unknown if focal or generalized seizure

## Old

- Partial or localization related
- Same
- No previous category

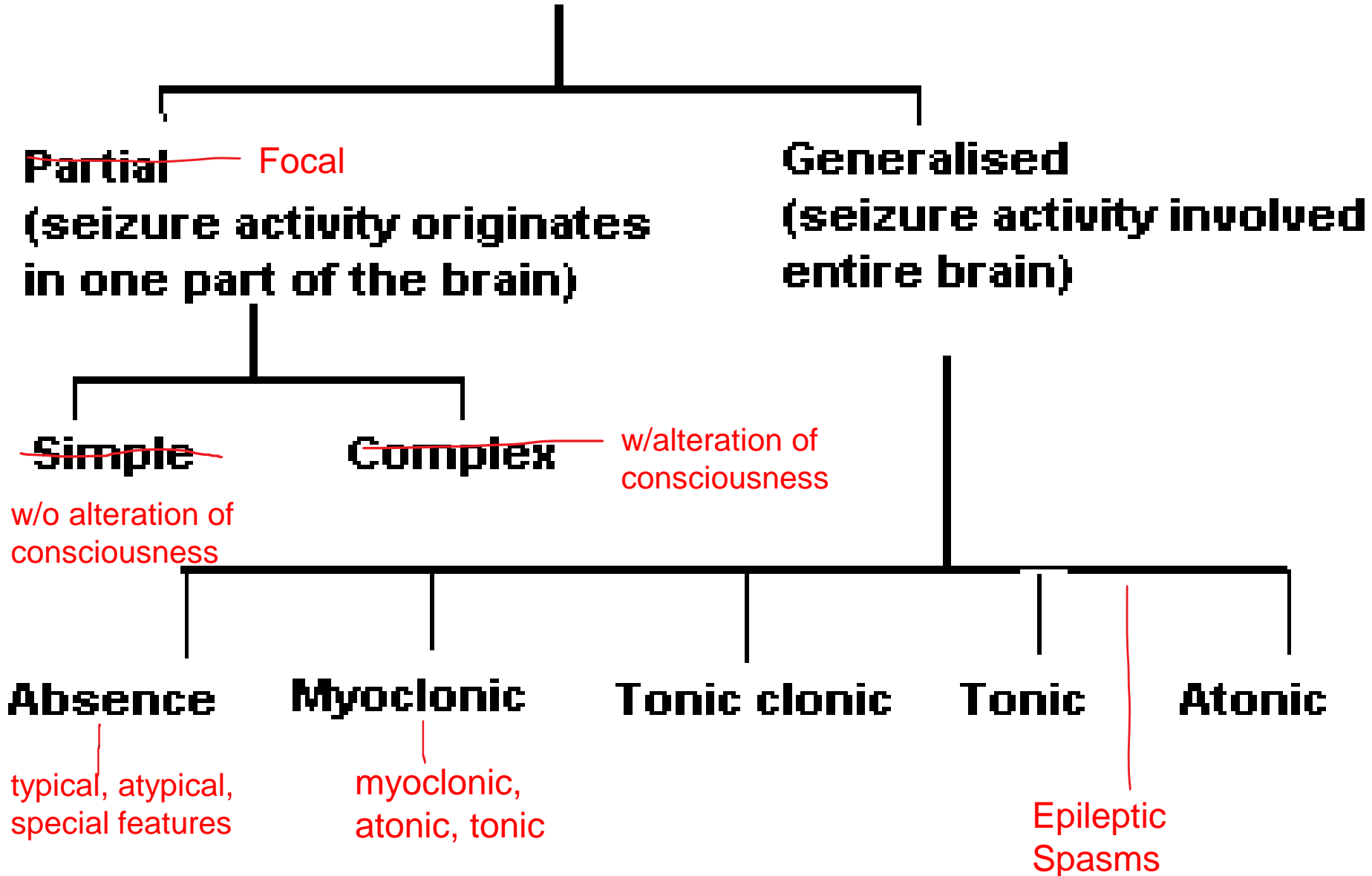
# New Terminology: Focal Seizure Classifications

- Based upon describing characteristics:
  - Aura +/-
  - Motor movements unilateral
  - Autonomic features
- Unaltered or altered consciousness
- May evolve to bilateral brain involvement → with bilateral motor movement

# New Terminology: Generalized Seizure Classification

- Tonic
- Clonic
- Tonic-Clonic
- Absence
  - Typical
  - Atypical
  - Special features:
    - Myoclonic Absence
    - Eyelid Myoclonia
- Myoclonic
  - Myoclonic
  - Myoclonic atonic
  - Myoclonic tonic
- Atonic
- Epileptic Spasms

# Seizure Classification



# New Terminology: Underlying Cause (Etiology)

## Old Term

- Idiopathic
- Symptomatic
- Cryptogenic

## New Term

- Genetic or presumed
- Structural/Metabolic
- Epilepsy of unknown cause
- Infectious
- Immune

# Description of Symptoms and History of the Seizure

- Possible seizure triggers:
  - Illness (URI, fever or other)
  - Sleep deprivation
  - Travel
  - Compliance issues with medication doses
  - Hormonal changes (puberty)
  - Weight gain (outgrew dosage)
  - Other



# Treatment Goals

- Prevent seizures or keep them short
- Prevent injury
- Improve learning by reducing time spent having & recovering from seizures
- Increase time spent in school
- Improve quality of life

# Treatment Options

- Anti-Epileptic Medications (AED's)
- Epilepsy Surgery (focal resection)
- Implantation of Vagus Nerve Stimulator (VNS)
- Ketogenic Diet Therapy
- Alternative treatments: None proven

# Medication and Treatments

- Anti-Epileptic medications (AED's)
  - Maintenance:
    - Monotherapy is ideal
    - Polytherapy is often the reality
  - Rescue:
    - Oral Benzodiazepines
      - Diazepam (Valium®), Lorazepam (Ativan®)  
Clonazepam (Klonopin®)
    - Rectal Diazepam (Valium®)
    - Nasal Midazolam (Versed®)

# Old generation Anti-Epileptic Drugs

- Phenytoin (Dilantin®)
- Phenobarbital
- Carbamazepine (Tegretol®)
- Valproic Acid (Depakote®)
- Ethosuximide (Zarontin®)

# Old generation Anti-Epileptic Drugs

- Multi-system effects of old AED's:
  - Liver, bone marrow, pancreas
    - Follow drug levels, CBC, ALT, AST, lipase
- Many interact with other AED's and affect efficacy or cause toxicity
  - Valproic Acid (Depakote®) increases other drugs
  - Some AED's decrease other drug levels

# Newer Generation Anti-Epileptic Drugs

- Lamotrigine (Lamictal®)
- Topiramate (Topamax®)
- Levetiracetam (Keppra®)
- Zonisamide (Zonegran®)
- Oxcarbazepine (Trileptal®)
- Rufinamide (Banzel®)
- Lacosamide (Vimpat®)
- Clobazam (Onfi®)

# Advantages of Newer Generation Anti-Epileptic Drugs

- Typically, less routine blood work
  - Optional drug levels
  - Carbon Dioxide
    - Topiramate (Topamax®)
    - Zonisamide (Zonegran®)
  - No significant drug interactions
    - Except Lamotrigine (Lamictal®)

# What Guides Choice of AED?

- Seizure Type
  - Focal onset
  - Generalized onset
- Syndrome
- EEG pattern
  - Focal onset
  - Generalized onset

- Co-morbidities
  - Psychiatric: Labile mood
  - Weight: Overweight vs. underweight
  - Other medical problems



# Narrow Spectrum AEDs

## Focal Epilepsy

- Oxcarbazepine (Trileptal®)
- Carbamazepine (Tegretol®)
- Phenytoin (Dilantin®)
- Gabapentin (Neurontin®)
- Lacosamide (Vimpat®)

## Generalized Epilepsy

- Ethosuximide (Zarontin®) absence only
- Rufinamide (Banzel®) Lennox-Gastaut

# Broad Spectrum AED for Generalized and Focal Epilepsy

- Lamotrigine (Lamictal®)
- Zonisamide (Zonegran®)
- Topiramate (Topamax®)
- Levetiracetam (Keppra®)
- Valproic Acid (Depakote® and siblings)
- Clobazam (Onfi®)

# Anti-Epileptic Drug Side Effects

- Temporary and are dose dependent
- Sedation, grogginess
- Difficulty thinking – cognitive slowing
- Off balance, “dizzy,” ataxic
- Behavior changes (good or bad)
- Stomach irritation
- Folic Acid deficiency

# Anti-Epileptic Drug Unique Side Effects

- Weight gain:
  - Valproic Acid (Depakote®)
  - Oxcarbazepine (Trileptal®), maybe
- Weight loss:
  - Zonisamide (Zonegran®)
  - Topiramate (Topamax®)
- Weight neutral: Most other AEDs

# Anti-Epileptic Drug Unique Side Effects

- Tremor:
  - Valproic Acid (Depakote®)
  - Lamotrigine (Lamictal®)
- Osteoporosis:
  - Phenytoin (Dilantin®)
  - Phenobarbital
- Acidosis:
  - Zonisamide (Zonegran®)
  - Topiramate (Topamax®)

# Anti-Epileptic Drug Unique Side Effects

- Rash:
  - Any AED
  - Steven Johnson Syndrome
    - Lamotrigine (Lamictal®)
    - Clobazam (Onfi®)
- Hypohydrosis, hyperthermia and kidney stones:
  - Zonisamide (Zonegran®)
  - Topiramate (Topamax®)

# Oxcarbazepine (Trileptal®)

*FDA approval adjunct:  $\geq 2$  yr. ; monotherapy:  $\geq 4$  yrs.*

Genetic testing: HLA-B\*1502 variant (Asian descent)

- Used for focal seizures
- Chemically similar to Carbamazepine (Tegretol®)
  - Fewer side effects: blood and liver issues
  - Less sedation, dizziness, or balance problems
  - Can be used in infants (2 wks. of age) -> elderly
  - No routine labs needed
  - Oxcarbazepine metabolite level is checked
  - Rare hyponatremia seen in infants

# Carbamazepine (Tegretol®)

*FDA approval Pediatric*

Genetic testing: HLA-B\*1502 variant (Asian descent)

- Used for focal seizures
- TID dosing vs. longer acting
  - Tegretol, Tegretol XR, Carbatrol
- Monitor CBC, LFT's (ALT/AST), blood levels
- Can cause hyponatremia
- Drug interaction:
  - Erythromycin and other antibiotics
  - INH
  - Grapefruit



# Levetiracetam (Keppra®)

*FDA approval adjunct: tonic-clonic, primary generalized  $\geq 6$  yr. oral;  $\geq 16$  yr. IV; myoclonic  $\geq 12$  yr. oral;  $\geq 16$  yr. IV; focal  $\geq 1$  month oral;  $\geq 16$  yr. IV, ER*

- Effective for focal and generalized epilepsy
- Can be used in all ages: Infants -> elderly
- Behavioral side effects:
  - Aggression, labile mood, can rarely lead to psychotic behavior
  - Higher risk: History of behavior problems or intellectual disability
  - Lower risk: Developmentally normal

# Levetiracetam (Keppra®)

- Good choice for children:
  - On multiple medications
  - With multi-organ failure
    - No medication interactions
    - Not metabolized or excreted by kidneys
- No routine labs or drug levels needed
- Now available as an IV preparation

# Zonisamide (Zonegran®)

*FDA approval adjunct: focal  $\geq$  16 yr.*

- Used from neonatal period -> elderly
- Broad spectrum with once a day dosing
- Weight loss +
- Some behavior problems
- Risk of kidney stones, hyperthermia and hypohydrosis
- Used with caution with sulfa drug allergies
- Drug levels optional

# Phenytoin (Dilantin®)

*FDA approval adjunct: generalized tonic clonic, focal; pediatric*

- Focal, secondarily generalized, generalized tonic-clonic, and status epilepticus
- QD or BID dosing
- Monitor CBC, LFT's (ALT/AST), AED levels
- Concentration can vary, so shake
- Side Effects:
  - Hirsutism, gum hyperplasia

# Gabapentin (Neurontin®)

*FDA approval adjunct: focal 3-12 yr.*

- Focal seizures
- Few drug interactions
- Renal metabolism
- Easy to titrate and wean
- Many other uses
  - Psychiatry, neuropathic pain, migraine
  - Drug levels not tested

# Lamotrigine (Lamictal®)

*FDA approval adjunct: oral IR Lennox-Gastaut Syndrome,  $\geq 2$  yr.; adjunct or monotherapy: focal  $\geq 2$  yr. ; oral ER  $\geq 13$  yr.; adjunct: tonic-clonic, primary generalized,  $\geq 2$  yr. ; oral ER  $\geq 13$  yr.*

- Broad spectrum & best side effect profile:
  - Least sedating and cognitive slowing
  - Good mood stabilizer
- Blood levels optional
- Negative:
  - “Black Box Warning”
  - Rash → Steven Johnson's Syndrome (titrated too fast, used with Valproic Acid (Depakote®), children < 2 yrs.)

# Topiramate (Topamax®)

*FDA approval adjunct: Lennox-Gastaut Syndrome, focal, tonic-clonic, primary generalized  $\geq 2$  yr. XR  $\geq 6$  yr.; monotherapy: focal, tonic-clonic, primary generalized  $\geq 2$  yr. XR  $\geq 10$  yr.*

- Used neonatal period - elderly
- May cause weight loss
- Used for migraine headache prophylaxis
- Can cause cognitive slowing
- No routine labs
- Can be associated with kidney stones, hypohydrosis, hyperthermia and paresthesia

# Valproic Acid (Depakote®)

*FDA approval: absence, focal,  $\geq 10$  yr.*

- Positive:
  - Effective when other anti-epileptic drugs fail
  - Good mood stabilizer (psychiatric uses)
  - Good for migraine headache prophylaxis



# Valproic Acid (Depakote®)

- Negative:
  - Significant side effect profile:
    - Liver, pancreas, and bone marrow
    - Weight gain, polycystic ovaries, hair loss, and tremor
    - **Fulminate liver failure in children < 2 yrs.**
  - Interacts with other drugs, and increases levels
- Monitor CBC, LFT's (ALT/AST), blood levels

# Ethosuximide (Zarontin®)

*FDA Approval absence  $\geq$  3 yr.*

- Narrow spectrum old generation drug
- Used for primary generalized epilepsy with absence seizures
- Monitor drug levels, CBC and LFT's (ALT/AST)
- Take with food to avoid GI upset

# Newer Anti-Epileptic Drugs

- **Lacosamide (Vimpat®)** *FDA approval: no pediatric; adult focal*
  - Baseline and interval ECG
    - Known conduction problems
    - PR interval increasing drugs
    - Cardiac disease
- **Rufinamide (Banzel®)** *FDA approval adjunct: Lennox-Gastaut Syndrome  $\geq 4$  yr.*
  - Baseline and interval ECG
    - Can shorten QT interval

# Other Anti-Epileptic Drugs

- **Phenobarbital** *FDA approval pediatric*
- **Primidone + Phenobarbital = (Mysoline®)** *FDA approval adjunct or monotherapy: epilepsy pediatric*
  - Focal, secondarily generalized, generalized tonic clonic, status epilepticus
  - First choice for infants
  - QD or BID dosing
  - Monitor CBC, LFT's (ALT/AST), blood levels
  - Possible effect on development

# Other Anti-Epileptic Drugs: Benzodiazepines

- Clobazam (Onif®) *FDA approval Lennox-Gastaut Syndrome  $\geq$  2 yr.*
  - Adjunct therapy
  - Associated with Steven Johnson Syndrome
- Clonazepam (Klonopin®)
  - Adjunct therapy
  - Rescue medication
  - Can habituate which alters efficacy over time
  - Increases oral secretions

# Rarely used Anti-Epileptic Drugs

- **Felbamate (Felbatol®)** *FDA approval adjunct: Lennox-Gastaut Syndrome, focal and generalized 2-14 yr.*
  - Focal seizures, Lennox Gastaut Syndrome
  - Released 1993, FDA warning 1994
  - Associated with aplastic anemia, liver toxicity
- **Vigabatrin (Sabril®)** *FDA approval adjunct: focal  $\geq 10$  yrs.*
  - Available in US under special program
  - Infantile spasms, partial seizures
  - Significant side effect profile: risk of blindness

# Management: Rescue Anti-Epileptic Drugs (AEDs)

## ■ Indications:

- Prolonged seizures
- Seizure flurries
- Emergency medical services (EMS) unavailable
  - Travel
  - Rural areas

## ■ Delivery:

- Oral
- Rectal
- Nasal

# Management: Rescue Anti-Epileptic Drugs (AEDs)

- Oral Benzodiazepines
  - Diazepam (Valium®)
    - *FDA approved*
      - *Oral  $\geq$  6 months*
      - *Injectable > 30 days*
  - Lorazepam (Ativan®) buccal, sublingual, injectable
    - *Injectable only FDA approved adults status epilepticus*
  - Clonazepam (Klonopin®) *FDA approved seizures pediatric*



# Management: Rescue Anti-Epileptic Drugs (AEDs)

- Rectal Benzodiazepine: Diazepam
  - Diastat®, AcuDial®
    - *FDA approved 2 years and older*



# Management: Rescue Anti-Epileptic Drugs (AEDs)

## Nasal Midazolam (Versed®)



*No FDA approval for treating status epilepticus or seizures adults or children*

Retrieved from: <http://www.intranasal.net/Home/default.htm>

# Other Treatment Options

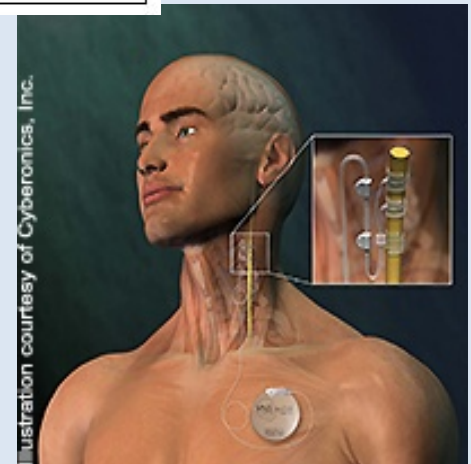
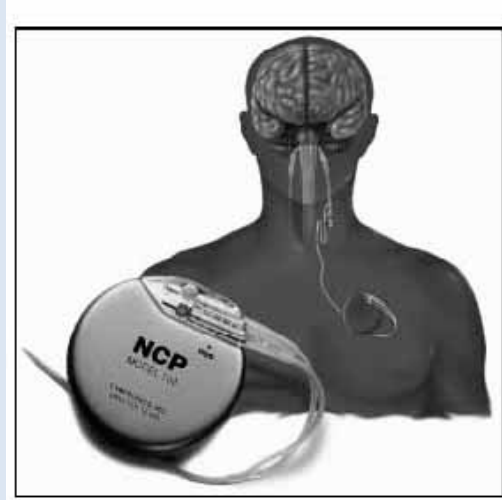
- Epilepsy surgery (resection if focal)
- Ketogenic diet therapy
- Vagus nerve stimulator
- Classic Ketogenic Diet
  - High fat, low CHO, low protein
  - Ketotic state enhances seizure control
  - Requires family commitment
- Modified Atkins Diet
  - Restricted CHO (10-15 gm)
  - Unrestricted fat & protein

# Vagus Nerve Stimulator

## Vagus Nerve Stimulation (VNS)

- Vagus nerve (in the neck) is stimulated with short bursts of electrical current
- Stimulation travels through the vagus nerve to the brain

- Implanted Device



# Interval History: Description of Symptoms

- Identify:
  - Epilepsy classification
  - Date of last seizure
  - Typical seizure presentation:
    - Onset, and associated factors
    - Frequency, timing, and duration
    - Level of consciousness
    - Body parts involved (unilateral or bilateral)
    - Recovery (post-ictal period)

# Interval History: Medication and Treatments

- Anti-epileptic and PRN rescue drugs:
  - Formulation (changes, brand-generic), dosage (adjustments), schedule (access)
  - Duration of Treatment and compliance
- Other medications/supplements
- Side effects, toxicity, drug interactions
- Toleration and efficacy of other treatments (VNS, ketogenic diet therapy)

# Interval History: Medical Update: Illness & Issues

- Age & weight
- Acute: Signs & symptoms of illness
- Chronic: Status of other medical or physical co-morbid problems or disabilities and environmental risk
- Recent laboratory or procedural testing
  - AED levels
  - LFTs , CBC, CO2
  - EEG, Imaging

# Action: Signs & Symptoms

## Medical Update

- Educate family:
  - Seizure etiology
  - Factors that can exacerbate seizures
    - Sleep deprivation
    - Illness
    - Missed medication doses (compliance)
    - Travel
    - Hormonal influences
  - Med side effects, and rescue meds



# Action: Signs & Symptoms

## Medical Update

- Educate the family:
  - Safety
  - Seizure first aid
  - Guidelines for activating EMS (911)
  - Guidelines for follow-up:
    - Seizures worsening in frequency, intensity or duration
    - Worsening co-morbid medical problems or disabilities

# Action: Seizure First Aid

- Most seizures < 2 minutes
- Goal: Protect the child
  - Keep calm and time the seizure
  - Turn gently to side
  - Do not hold down or restrict movement
  - Clear the area
  - Do not put anything in mouth or force it open
  - Reassure as awareness returns

# General Seizure Safety Tips

- **Water Safety:**
  - **NEVER** leave alone in a bathtub, hot tub or swimming pool
  - Encourage showers
  - Keep bathroom door unlocked
- Avoid pillows, blankets, or toys in bed
- Avoid heights and high risk sports activities

# General Health and Psycho-Social Issues

- Sleep:
  - Poor sleep patterns common
  - Sleep deprivation can worsen seizures
- Nutrition:
  - AED can increase or decrease appetite
  - AED can cause weight gain or loss
- Psychiatric co-morbidities are common
  - ADD/ADHD, anxiety, OCD, other

# General Health and Psycho-Social Issues

- Behavior
  - Temperament
  - New or worsening behaviors, and provoking factors
  - Emotional lability
- Effect of anti-epileptic drugs on behavior
  - Some AED's modulate behavior
  - Some AED's make behavior worse

# General Health and Psycho-Social Issues

- **Communication**
  - Language
  - Speech
  - Eye Contact
- **Social Skills**
- **Relationships**
  - Peers (teasing or bullying)
  - Family and sibling relationships

# Family Dynamics and Coping

- Identify and Describe:
  - Change in family dynamics or structure
  - Financial or health insurance issues
  - Parent/patient level of concern and understanding about epilepsy/seizures
  - Family support systems

## Action: Health, Psycho-Social, Family Dynamics and Coping

- Encourage family to follow-up for:
  - Medication side effects or interactions
  - Exacerbation of seizures
  - Behavioral or psychiatric issues
  - Changes in status of co-morbidities
  - Concurrent illness
- Update or consult child neurology provider as indicated



# Early Intervention, School and Therapy Programs

- Identify:
  - Grade level & classroom setting
  - Academic performance
  - Educational or ADD/ADHD testing
  - Learning disabilities
  - Services/Therapies (OT/PT, speech, adaptive PE)
  - Educational Interventions (504 Plan, IEP, Resource, tutoring)

# Early Intervention, School and Therapy Programs

- Identify:
  - Focusing/Attention issues
  - Cognitive skills
  - Language skills
  - Classroom behavior
  - Other identified co-morbidities
  - Social skills
  - Response to medication (ADHD, AED, other)

# Early Intervention, School and Therapy Programs

- Obtain/verify consent for exchange of information
- Contact the school nurse to help facilitate:
  - School seizure safety plan
    - Avoid water sports/PE unless full supervision
    - Rescue Medications:
      - Usually not necessary unless history of prolonged seizures or no access to EMS
      - CA Senate Bill 161: Controversial to have non-licensed personnel to administer
  - Educating school staff: seizure safety

# Early Intervention, School and Therapy Programs

- Facilitate appropriate evaluations:
  - Educational or ADD/ADHD
  - Speech, OT, PT or adaptive PE
- Provide input school management:
  - IEP (individualized educational plan)
  - 504 plan
  - Behavior plan
- Refer family to school liaison, advocacy sources, counseling as needed

# Epilepsy Resources

- Epilepsy Foundation of America: [www.efa.org](http://www.efa.org)
  - Books, Kits, Pamphlets and Videos
  - School:
    - Seizure Action Plan
    - Seizure Observation Record
    - Parent Questionnaire
  - Driving & Epilepsy: EFA link state by state laws: <http://www.epilepsyfoundation.org/resources/Driving-Laws-by-State.cfm>
- Association of Child Neurology Nurses:
  - Child Neurology Encounter Guides: [www.acnn.org/books](http://www.acnn.org/books)
- American Academy of Neurology: [www.aan.com](http://www.aan.com)
- American Epilepsy Society: [www.aesnet.org](http://www.aesnet.org)
- CDC Epilepsy website: <http://www.cdc.gov/Epilepsy/>



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