Epilepsy in Childhood

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Objectives

- Provide overview of epilepsy
- Identify treatment options (including managing status epilepticus)
- Outline care of the child during & following a seizure
- Address commonly asked questions
- Describe learning & psychosocial issues associated with seizures

Epilepsy & Seizures

Epilepsy

- a condition of recurrent (>2) unprovoked seizures

Seizures (symptoms or behaviors)

- produced by sustained synchronous (abnormal) firing of neurons within the cerebral cortex

Prevalence

Total Population

- 1 in 11, one spontaneous seizure
- 3 in 100 (3%), two or more seizures
- 5 to 10 cases p/1000 ongoing seizures

Child Population

- birth to 20 years - risk for epilepsy about 1%
- 20 - 25 % of children with epilepsy have continuing seizures refractory to AEDS

Hauser WA & Hesdorffer DC. Remission, intractability, mortality, and comorbidity of seizures. In Wyllie E, editor. The treatment of epilepsy: principles and practice. Philidelphia. PA: Lippincott Williams and Wilkins, 2001. pp. 139-145.

Etiology

- unknown etiology, 70%
- genetic factors (idiopathic/cryptogenic)
- metabolic (encephalopathy)
- infection (viral encephalitis, Rasmussen's)
- head trauma
- cerebrovascular (stroke)
- congenital brain abnormalities (cerebral dysgenesis, tumors, cystic lesions, etc.)
- neurological syndromes (? genetic factors)
  - progressive (progressive myoclonic epilepsy)
  - congenital (Tuberous Sclerosis)

Annegers JF. The epidemiology of epilepsy. In: Wyllie E, ed. Treatment of epilepsy: Principles and Practice. 2001:131-138

Classification of Epileptic Seizures (ILAE-1989)

- **Generalized (convulsive, non-convulsive)**
  - electrographic - simultaneous, synchronous epileptic discharges over entire cortex
  - clinical features- absence, myoclonic, clonic, tonic, tonic-clonic, atonic

- **Partial seizures (focal, local)**
  - electrographic - focal (localized) epileptiform activity
  - clinical features
    - A. simple partial, no impairment of consciousness
    - B. complex partial,
      - impaired consciousness at onset
      - simple partial f/by impaired consciousness
    - C. partial evolving to GTC seizures

- **Unclassified epileptic seizures** – e.g. reflexic seizures

Proposed Classification of Generalized Epilepsies & Epilepsy Syndromes

- **idiopathic**
  - neurologically normal
  - hereditary component

→

- childhood absence epilepsy (2-8%)
- juvenile absence epilepsy
- juvenile myoclonic epilepsy
- benign neonatal convulsions,
- benign familial neonatal convulsions

- **symptomatic**
  - CNS structural or metabolic abnormality

→

- infantile spasms (1 in 4000)
- Lennox-Gastaut syndrome
- myoclonic-astatic seizures
- other syndromes- e.g. lissencephaly

- **cryptogenic**
  - probably symptomatic
  - unidentified cause

Commission on Classification & Terminology of the ILAE, Proposal for Revised Classification of Epilepsies & Epilepsy Syndromes In: Wyllie E, ed. Treatment of epilepsy: Principles and Practice. 2001: 457-66

Proposed Classification of Localization Related Epilepsies & Epilepsy Syndromes

distinct, localized epileptogenic foci

- **Idiopathic**
  - may involve both hemispheres
  - hereditary predisposition
  - easily treated
  - often spontaneously remit

  examples:
    - **benign childhood epilepsy with centrotemporal spikes (BECTS)**
    - **childhood epilepsy with occipital paroxysms (BECOS)**
    - **primary reading epilepsy**

- **Symptomatic**
  - focus in one hemisphere
  - CNS lesions +/-
  - seizure semiology

  (related to anatomic location)
    - **temporal**
    - **frontal**
    - **parietal**
    - **occipital**

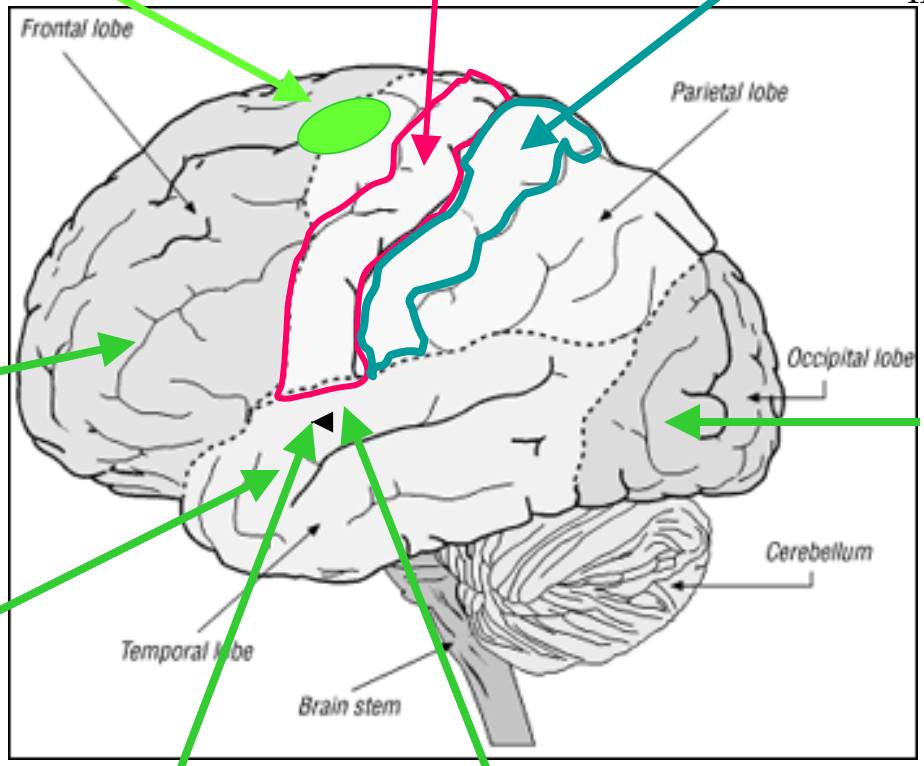
Commission on Classification & Terminology of the ILAE, Proposal for Revised Classification of Epilepsies & Epilepsy Syndreomes In: Wyllie E, ed. Treatment of epilepsy: Principles and Practice. 2001: 457-66

Seizure Characteristics

Frontal Pre-central
-tonic extension
-flexion of limbs,
-head/eye turning

Central or Motor
- involuntary clonic movements

Parietal (Post-central
-sensory phenomena
e.g. tingling, numbness in limbs or face



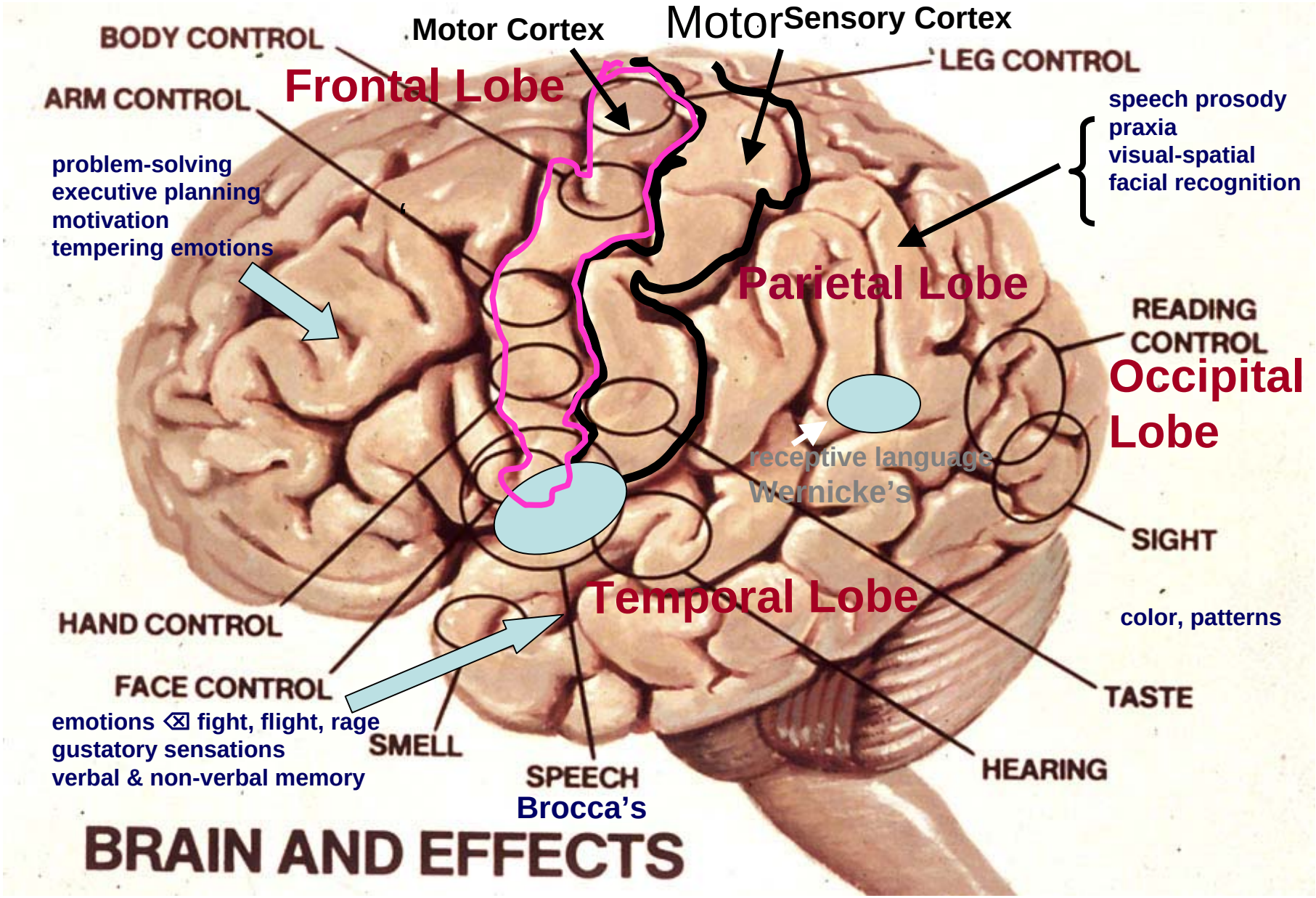
Pre-frontal
-bicycling,
-other unusual movements,
-screaming, vocalizations

Occipital
-visual disturbances,
e.gs. stars, bright lights, microposia, macropsia

Temporal
-auditory or visual hallucinations
-staring, automatisms, ↓awareness

Autonomic symptoms
-vomiting, pallor, flushing, piloerection, pupil dilataion, etc.

Mesial Structures (limbic region) -Temporal
-smells, tastes, gustatory sensations,
-fear, irritability, aggression
-dysmnesic symptoms


BODY CONTROL
Motor Cortex
Motor
Sensory Cortex
LEG CONTROL
ARM CONTROL
Frontal Lobe
speech prosody
praxia
visual-spatial
facial recognition
problem-solving
executive planning
motivation
tempering emotions
Parietal Lobe
READING CONTROL
Occipital Lobe
receptive language
Wernicke's
SIGHT
Temporal Lobe
color, patterns
HAND CONTROL
FACE CONTROL
TASTE
emotions ⌫ fight, flight, rage
gustatory sensations
verbal & non-verbal memory
SMELL
SPEECH
Brocca's
HEARING
BRAIN AND EFFECTS

Reflex seizures

- Visually triggered
  - photosensitivity, self-induced flicker, pattern sensitive, eye closure

- Non-Visually induced

thinking	proprioceptive
praxis	touch
reading	hot water
language	audiogenic (startles)
musicogenic	somatosensory (tapping/rubbing)
eating	

Antiepileptic Drugs

Partial Seizures

- Carbamazepine (Tegretol)
- Clobazam (Frisium)
- Clonazepam (Rivotril)
- Gabapentin (Neurontin)
- Lamotrigine (Lamictal)
- Nitrazepam (Mogadon)
- Phenytoin (Dilantin)
- Phenobarbital
- Topiramate (Topamax)

Infantile Spasms

- Vigabatrin & ACTH

Generalized Seizures

- Valproic Acid (Depakene, Epival)
- Ethosuximide (Zarontin)

New AEDS

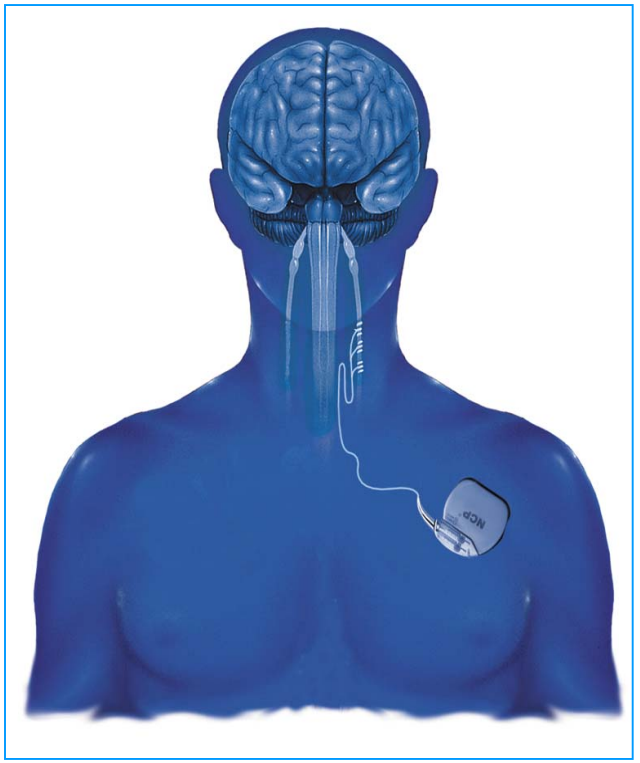
- Tiagabine (Gabitril)
- Levetiracetam (Keppra)
- Oxcarbazepine (Trileptal
- Zonisamide

Other

- Steroids (IVG, Prednisone)

Vagus Nerve Stimulation

- Vagus = “wanderer”
- Vagus Nerve = major communications link between body and brain
- Vagus nerve stimulation = mild electrical signals which are applied to the vagus nerve in the neck for transmission to the brain



Cyberonics

Ketogenic Diet

- a special diet used to treat seizures
- initially 1920's to treat intractable seizures
- AEDs replaced diet - now resurging interest
- diet high in fat, low in carbohydrate & protein, results in ketosis
- fluids limited
- ketosis exerts antiepileptic effect, though precise mechanism of action not known

Epilepsy Surgery

- Corpus Callostomy
- Temporal/Extratemporal Resection
- Hemispherectomy

Care of the Child During a Seizure

- Remain CALM!!
- Protect from injury
- Loosen clothing
- Roll child on his/her side?
- Time & observe seizure behaviours
- Give PRN medications as ordered

- Do not restrain the child
- Do not put anything in the child's mouth
- Do not give mouth-to-mouth resuscitation
- Stimulation (verbal or physical) will not bring a child out of a seizure

Use of O_2 During a Seizure

- No evidence O_2 by mask is effective during short seizure
- Contraction of airway inhibits O_2 intake by mask
- Short periods of apnea (up to 45 seconds) during the tonic phase is common
- Blood flow reduced at the periphery and increased to the vital organs (↑200% to brain)
- O_2 is rarely used at home for seizures

Care of Child After a Seizure

- Allow a period of rest if needed
- Do not give anything by mouth until child is fully alert
- Record seizure symptoms and post-ictal state
- Re-orient the child if necessary

Describing Seizures

- Level of consciousness
- Motor activity - where and what
- General behaviours (scared look, automatisms)
- Autonomic features (pupils, facial flushing)
- Vocalizations
  - Patient reported symptoms

Helping Families Cope with Ongoing Uncertainty

- Clarify parent's current understanding of diagnosis and treatment
- Provide information about Epilepsy
- Provide information about related tests
- Acknowledge and normalize concerns and
- Encourage connection with local Epilepsy Association
- Identify sign posts in the adaptation process
- Explore resource needs

Questions commonly asked by Parents

- Can my child die from a seizure?
- Do seizures cause brain damage?
- Are seizures painful?
- How will I know if my child has a seizure at night?
- Will my child outgrow his/her seizures?
- When should I call the doctor or 911?
- Is there anything I can do to prevent a seizure?

Can my child die from a seizure?

- Rare for a child to die from a seizure
  - increased risk for Sudden Unexpected Death from Epilepsy call SUDEP in young adults
- Higher risk of death with prolonged status epilepticus
  - Seizures that go on for hours
- Children can die
  - unsafe situations (e.g., head injury, drowning, suffocation)

Donner EJ. Smith CR. Snead OC 3rd. Sudden unexplained death in children with epilepsy. Neurology. 57(3):430-4, 2001 Aug 14

Do Seizures Cause Brain Damage?

- **What we know!**
  - prolonged seizures clearly capable of injuring the brain (>30 min)
  - underlying brain pathology may cause global or selective deficits
  - poorly controlled seizures (& some AEDs) learning difficult

- **Area of continuing debate?**
  - single brief seizures do not cause brain damage
    however, growing evidence in animal models suggest
  - isolated, brief seizures are likely to cause some negative changes in brain function & possibly loss of specific brain cells
    - not true for all forms of epilepsy,
    - likely highly dependent upon type of seizure and etiology

- **More long-term studies of epilepsy are required**

Rovaniemi, Finland, June 2001 Workshop (35 scientist) aimed at addressing the question: "Do seizures damage the brain?" Details of this meeting have been published as Sutula T and Pitkänen A. Do seizures injure the brain. Progress in Brain Research 135. New York: Elsevier Science; 2002.

Are seizures painful?

- no evidence that seizures are painful
- frightening for a parent to watch but the child is not usually aware of what is happening
- child may experience discomfort after a seizure such as a headache, muscle aches or pain from an injury (e.g., bitten tongue)

Will my child outgrow seizures?

- Features predicting remission at time of diagnosis
  - Young age at onset, generalized-onset, normal neuro exam, & idiopathic or cryptogenic etiology
- **Positive prognostic predictors during epilepsy**
  - Normal MRI
  - Normal EEG between seizures
  - Seizures controlled on first or second AEDs during first year
- **If seizures remain uncontrolled >4years, then <10% remit**
- **Usually outgrow benign epilepsies of childhood**

  (e.g., BECTS, BECROS, typical absence)

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When should I call 911?

- If seizure > 5 minutes or there are repeated seizures that do not allow for recovery :
  - treat with sublingual Lorazepam (if ordered)
  - if seizure(s) continues - repeat Lorazepam & call 911

or

- Call 911
  - if Lorazepam not ordered or if previously used but not effective

- Inform doctor
  - increased seizure frequency, change in seizure type

How will I know if my child has a seizure at night?

- Use a baby monitor
- Attach a noisy device to the child (e.g. bell)
- Place child’s bed next to a wall adjacent to parent’s room
- Some parents sleep with their children
- Look for signs that child has had a nocturnal seizure (e.g., eneuresis, unusual morning fatigue)

Can I prevent a seizure?

- Ensure the child receives anticonvulsants on time and the amount prescribed
- Ensure adequate rest and nutrition
- Identify individual triggers and avoid or modify if possible

*Restraining the child does not stop a seizure

Cognitive Co-morbidities

Fixed

- **global deficits** - epilepsy syndromes, prolonged or repetitive seizures, hypoxia, infection, bleeding, congenital abnormalities

- **selected deficits** - repetitive focal seizures & localized lesions e.g. MTS, tumor, cortical dysplasia

Transient

- **seizures** - ↓ processing, consolidating & retrieval
- **interictal activity** - miss instruction & social cues, fail to respond
- **sleep disturbances** -↑fatigue,↓energy,↓availability to learn
- **antiepileptic drugs** - inhibitory (fatigue) or excitatory (inattention, impulsivity)

Psychosocial Co-morbidities

- **psychiatric disorders**
  - general child population 6%
  - children + chronic illness 11.6%
  - children + epilepsy 34.3% (*Rutter M, et al., Isle of Wight '70*)
- **behavior problems new onset epilepsy >25%** (*Austin, et al 2001*)
- **social relations** - risk for social maladaptation
  - feel separate, alienated, not belonging, different, not normal
  - excluded by peers
  - teased and bullied (>70%)* *(*Elliott, Lach, & Smith, 1999)*

Take Home Messages

- Improved understanding of:
  - what is epilepsy, etiologies, seizure types
  - treatment options,
  - facts about management of seizures
  - epilepsy as a complex disorder impacting on the cognitive & psychosocial development of children