

Epilepsy 101

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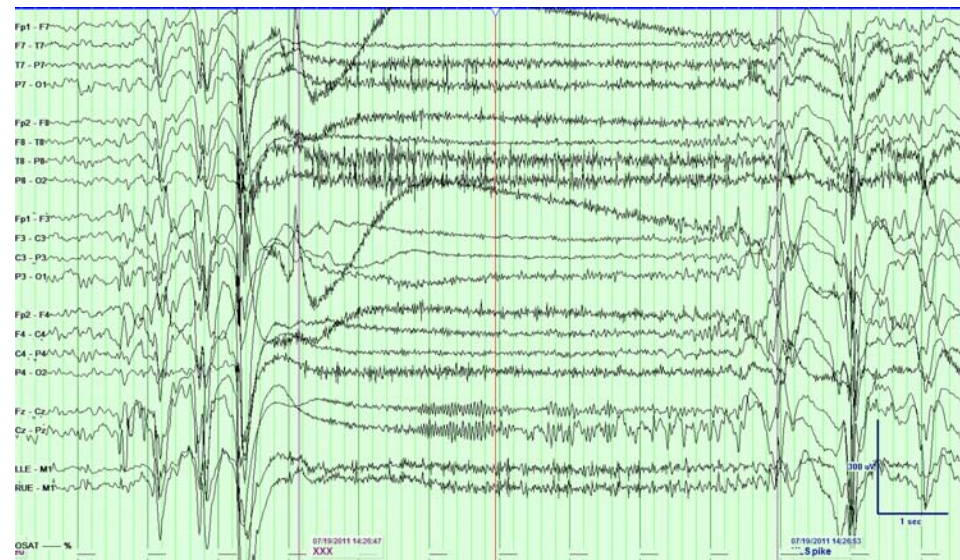
November 2011

Specific Aims

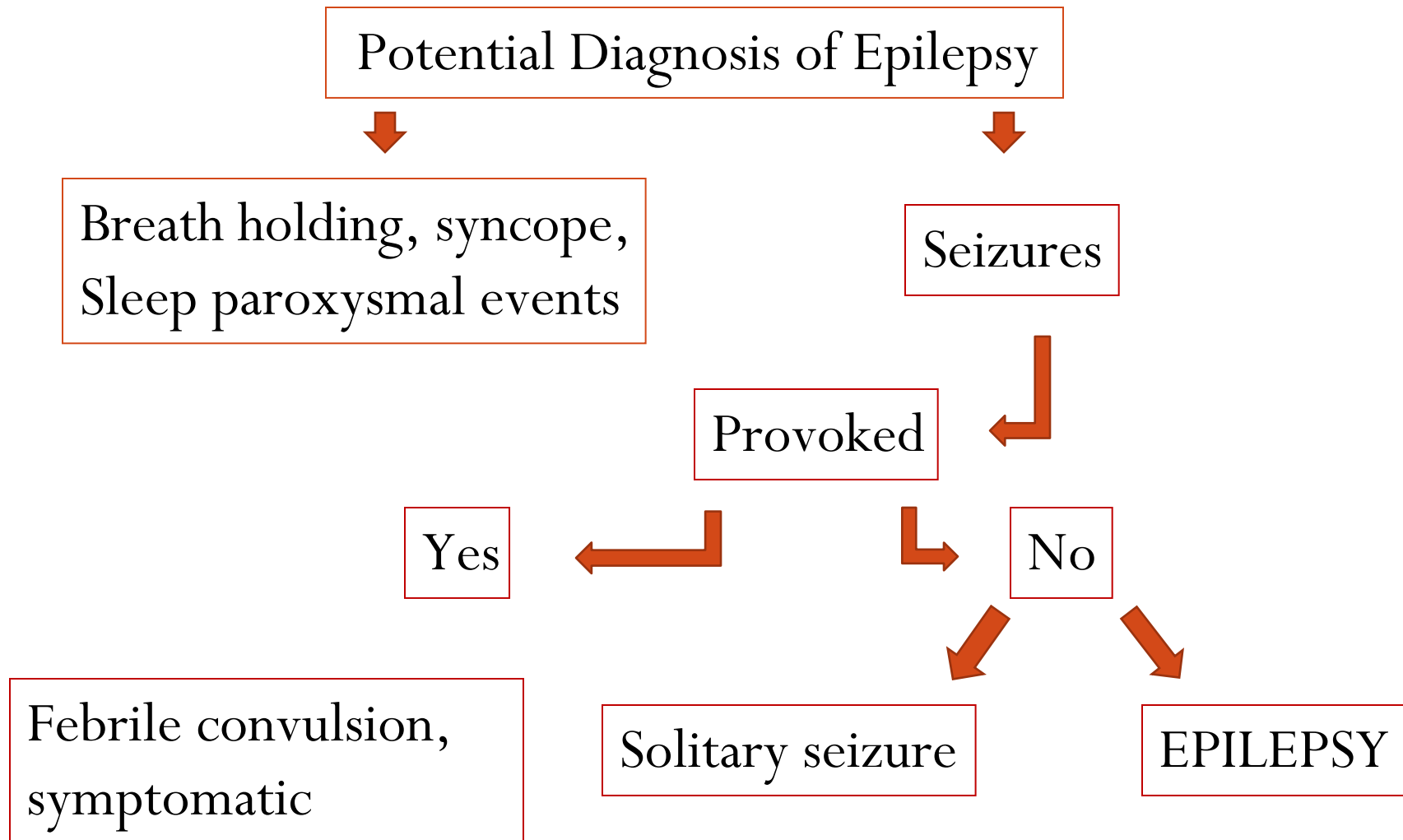
- How do we define epilepsy?
- Do seizures equal epilepsy?
 - What are seizures?
- Seizure medications, “does one size fit all?”
 - Does my son/daughter have to take medication?
- Why do we want to perform electroencephalograms (EEG), brain scans (Magnetic Resonance Imaging or MRI), and ask you so many questions when we see you?
- Is there more to epilepsy than just seizures?

Definition of Epilepsy

- Epilepsy is defined as two or more unprovoked events (seizures) that are produced by abnormal synchronization of cortical neurons that result in a change in perception or behavior.



Definition of Epilepsy

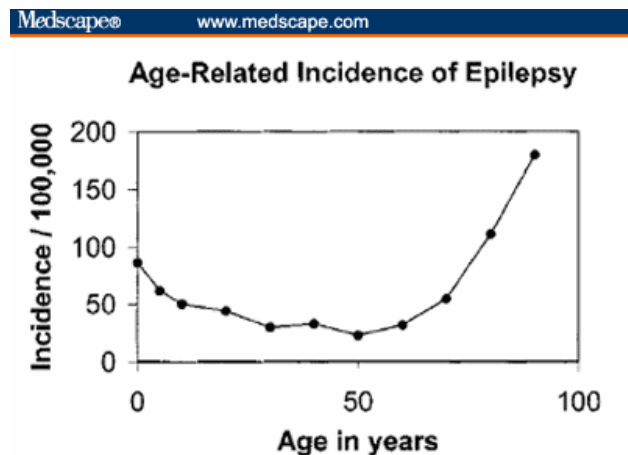


Seizures

- Seizures are repetitive events that are produced by abnormal synchronized cortical discharges.
 - Most patients have a limited repertoire of events.
- Distinct from:
 - Tics
 - Involuntary movements (i.e. dystonia)
 - Behaviors (i.e. day dreaming)
 - Sleep paroxysmal events (i.e. sleep walking)
 - Tremors
 - Typical migraine headaches

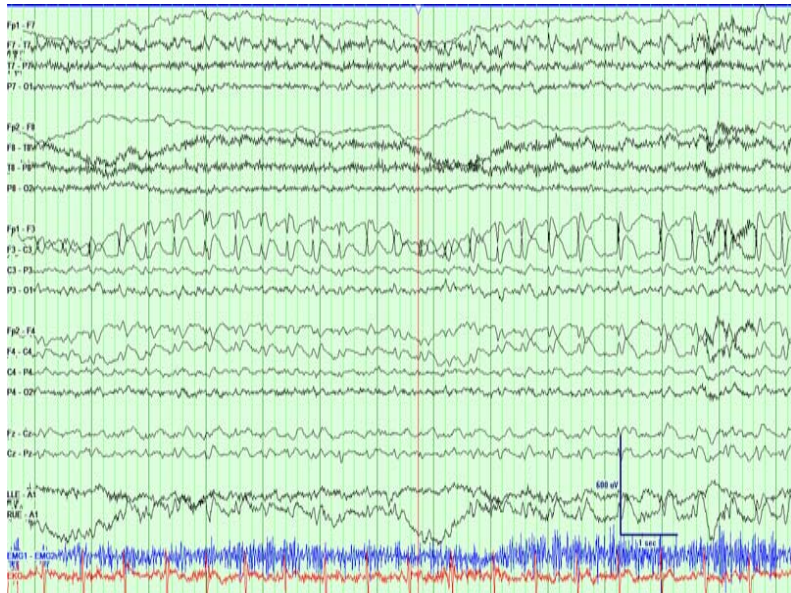
Epidemiology

- If you live long enough, approximately 3% of the population will have epilepsy (Cumulative Incidence).
- The highest frequency of afebrile seizures is during the first years of life and at the end of life.



Seizures

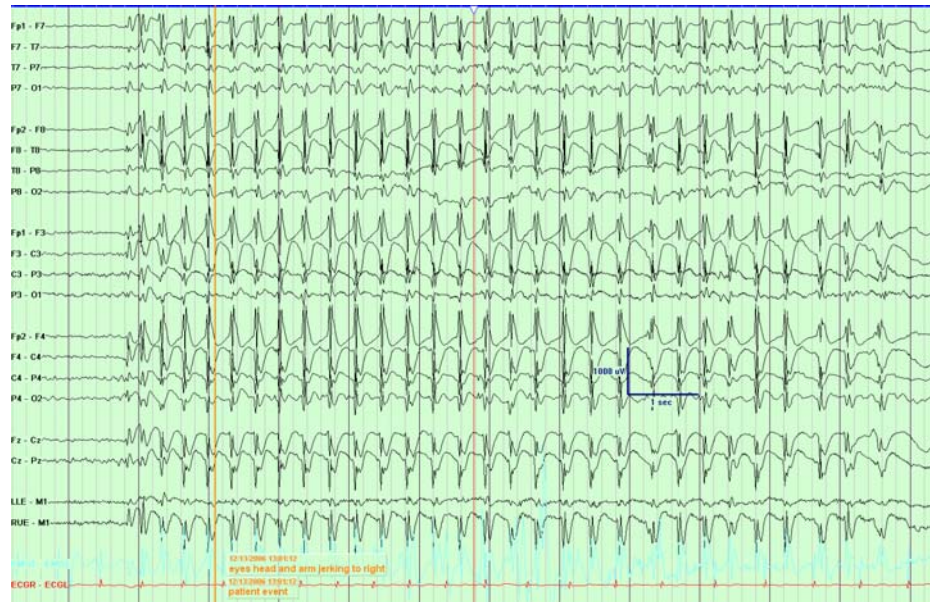
- Seizures can begin:
 - In one part of the brain-Focal onset or focal seizures (partial seizures)
 - EEG demonstrate a focal onset.
 - The patient stopped and stared: partial complex seizure.



Patient, suddenly stopped and stared.

Seizures

- Seizures can begin from multiple sites in the brain.
- When seizures begin on both sides of the brain at the same time, we call these generalized seizures.
 - The EEG demonstrates a generalized onset.
 - The patient stopped and stared-a generalized seizure.



Doctor, she just stopped
And stared.

Seizures

- Did you pick up how the two seizures were described on the EEG and by the parent?
 - The EEG showed a focal seizure and generalized seizure, yet these were described the same.
 - Stopped their behavior and stared.
 - Does the difference matter?



Medications

- What are the decisions to be considered before starting medication therapy?
 - Risks of further seizures outweigh the risks of treatment.
 - From seizures themselves
 - Injury to self
 - Injury to others
 - Psychosocial consequences
 - The effectiveness of medications to prevent recurrences.
 - Almost no child should be treated with medication after the first seizures.
 - The overall recurrence rate within 2 years is about 40% - 50%.
 - After the second seizure, the rate increases to 80% - 90%.

Medications

- So, when we think we have to treat seizures:
 - What are our choices?
 - Why do we make the choices we do?
 - What happens when everything fails?
- What is your response to what we use to treat your child?
 - Does it matter to us?
 - How do we need to work together to get things right?

Medications: Choices

- ACTH*
- Carbamazepine*
- Clobazam
- Ethosuximide*
- Felbamate
- Gabapentin
- Lamotrigine
- Lacosamide
- Levetiracetam
- Oxcarbazepine
- Phenobarbital*
- Pregabalin
- Primidone*
- Rufinamide
- Stiripentol
- Topiramate
- Valproic Acid*
- Vigabatrin
- Zonisamide

Questions to you: Why?

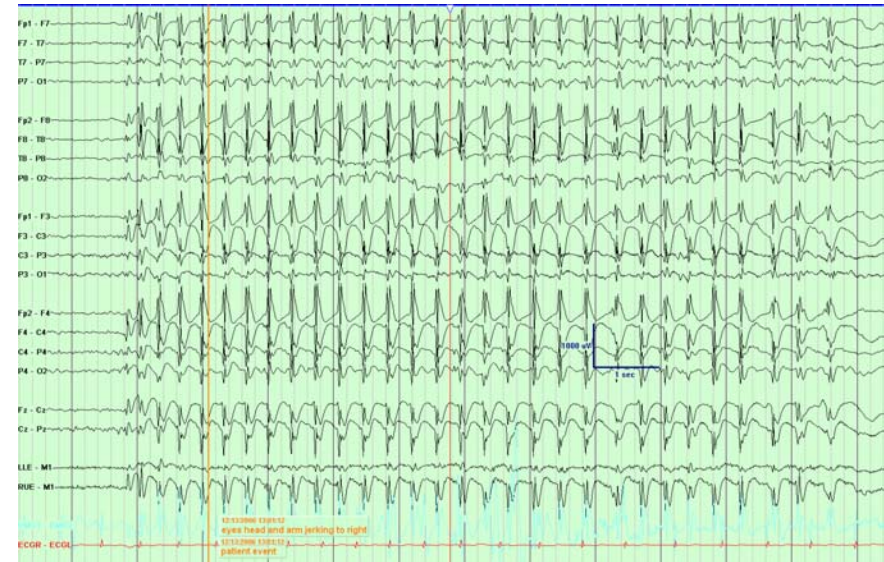
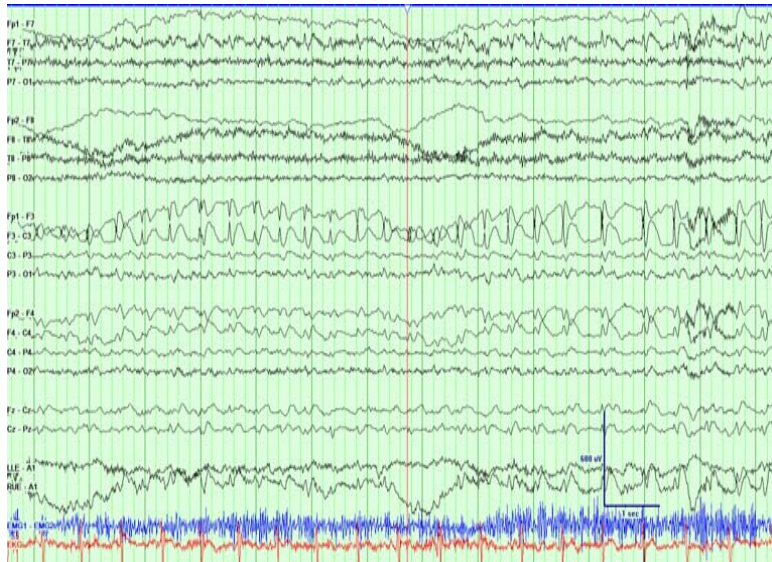
- The truth: “History”-why we ask such funny questions.
 - We do not see your sons/daughter’s seizures.
 - We rely on how you describe the seizures.
 - Look for clues to see if we can determine if it begin focally or generalized.
 - How did the seizure look when it first began?
 - How long did it last?
 - What did your son/daughter do during the event?
 - One side moved, both sides, rhythmically or just stiffening.
 - We think it makes a difference how we begin treatment.
 - Gives us options of which medications.

Questions to you: Why?

- Family history:
 - Your son/daughter's seizures may be due to genetics.
 - We are finding that many epilepsies are due to genes.
 - If we find a gene at fault it might give us direction for treatment.
- For example: Dravet syndrome
 - Due to a mutation in the sodium channel.
 - We know this produces a very difficult seizure to control with medications.
 - Valproic acid, Clobazam, Stiripentol
 - We can tell you the development problems to look for and what the future may hold for your son/daughter.

Next Steps

- Remember our two cases, both had a behavioral arrest and staring, but the EEG....
- This is why we usually ask you to get an EEG on your child.



Next Steps

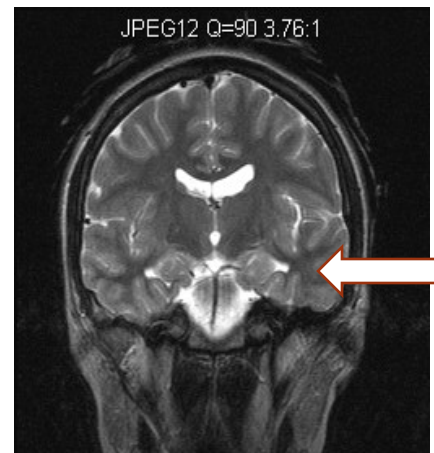
- What does the EEG tell us:
 - Focal versus Generalized
 - Possible epilepsy syndrome.
 - Why would knowing the epilepsy syndrome be important?
 - Just as in finding out possible genetics it tell us:
 - What drugs might work.
 - Allows us to give you a prognosis for the future.
 - Allows us to help you with other co-morbidities that are associated with that particular epilepsy syndrome.

Next Steps:

- We will probably ask you to get an MRI scan of your son/daughter. Why?
 - To look for structural causes for the seizures.
 - Again to help us define an epilepsy syndrome.
 - Gives us an expanded view for possible therapy.



DNET



MTLS

Medications

- So, what do we have at this point?
 - History
 - What the seizure looks like
 - EEG
 - MRI
 - Possibly some labs
 - If the history indicates that we might have:
 - Genetic
 - Metabolic

So what are the epilepsy syndromes?

- Neonatal period
 - Benign familial neonatal epilepsy
 - Early myoclonic encephalopathy
 - Ohtahara syndrome
- Infancy
 - Epilepsy of infancy with migrating focal seizures
 - West syndromes
 - Myoclonic epilepsy in infancy (MEI)
 - Benign infantile epilepsy
 - Dravet syndrome
 - Myoclonic encephalopathy in nonprogressive disorders

So what are the epilepsy syndromes?

- Childhood
 - Febrile seizures plus (FS+ can start in infancy)
 - Early onset benign childhood occipital epilepsy (Panayiotopoulos type)
 - Epilepsy with myoclonic atonic seizures
 - Benign childhood epilepsy with centrotemporal spikes
 - Autosomal dominant nocturnal frontal lobe epilepsy
 - Late onset childhood occipital epilepsy (Gastaut type)
 - Epilepsy with myoclonic absences
 - Lennox-Gastaut syndrome
 - Epileptic encephalopathy with continuous spike and wave during sleep
 - Landau Kleffner syndrome
 - Childhood absence epilepsy

So what are the epilepsy syndromes?

- Adolescence-Adult
 - Juvenile absence epilepsy
 - Juvenile myoclonic epilepsy
 - Progressive myoclonus epilepsies
 - Autosomal dominant epilepsy with auditory features
 - Other familial temporal lobe epilepsies
 - Epilepsy with generalized tonic-clonic seizures alone

Why is this important?

- Syndromes
 - Literature on:
 - Treatment
 - Prognosis
 - Co-morbidities
- Our aims are to see if your son/daughter fit into a syndrome so can more focussed on treatments and letting you know what lies ahead.

Reality

- Unfortunately, not everyone fits into as syndrome.
- What does this mean?
 - The literature tells us: If no syndrome is known:
 - First medication works about 45% of the time to stop seizures.
 - Second medication works about 25% of the time to stop seizures.
 - Third and beyond medications work about 3 – 5% of the time to stop seizures.
- What if medications do not work?

What Next?

- When medications do not work:
 - Vagus nerve stimulator
 - Ketogenic diet/Modified Atkins Diet
 - Epilepsy surgery

Why should we care?

- Emotional/Psychological aspects:
 - Learning/memory
 - Self-worth
 - Seizures in front of peers
 - I am “less” than my peers because I have seizures
 - Psychological aspects of not knowing when the next seizure will occur.
 - Development of “dependent” relationships
 - SUDEP

To be continued:

- Thanks for listening.
- Questions?

