Seizure and Paroxysmal Disorders: Fits and Faints

Max Wiznitzer, M.D.
Rainbow Babies & Childrens Hospital
Cleveland, Ohio

Neonatal Events

- Seizures
- Apnea
- Jitteriness/tremor
- Benign sleep myoclonus

Neonatal Seizures

Types

- Focal clonic
- Multifocal clonic
- Myoclonic
- Tonic
- Subtle

Causes

- Hypoxic-ischemic encephalopathy
- Brain malformation
- Perinatal stroke
- Meningitis/encephalitis
- Metabolic disorders
- Benign neonatal seizures (5th day seizures)
- Familial neonatal convulsions

Neonatal Seizures

- Image of neonatal monitoring equipment
Apnea

- Definition: Cessation of respiratory function
- Occurs in premature>term newborns
- Rare as sole seizure manifestation
- Apnea and depressed consciousness can be due to:
  - Increased intracranial pressure
  - Infection
  - Metabolic disturbance

Jitteriness/Tremor

- Rhythmic and usually rapid repetitive extremity>trunk movement
- Can have exaggerated response to stimulation
- Association with:
  - Hypoxic-ischemic encephalopathy
  - Infection
  - Drug withdrawal
  - Metabolic disturbance
  - Idiopathic

Neonatal Tremors

- Manifested by extremity jerking in sleep
- Movements stop with arousal
- Awake and sleep EEG is normal
- Gradually fades over weeks to months

Benign Sleep Myoclonus

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Benign Sleep Myoclonus

Paroxysmal Events in Infancy

- Seizures
- Benign myoclonus of infancy
- Gastroesophageal reflux
- Benign paroxysmal torticollis
- Hyperekplexia
- Spasmus nutans
Seizures in Infancy

- Febrile
- Infantile spasms
- Severe myoclonic epilepsy of infancy
- Partial
- Generalized tonic-clonic

Febrile Seizures

- Occur from 6 months to 6 years old
- Prevalence 3-4%
- Types
  - Simple
  - Complex
- 1/3 have second febrile seizure
- Need to differentiate from CNS infection and epilepsy

Febrile Seizures

- Treatment
  - Usually treat underlying condition and fever
  - Can use oral or rectal diazepam with illness
  - Consider prophylaxis for recurrent or prolonged seizures

Infantile Spasms

- Incidence: 25/100,000
- Onset: usually 4-7 months
- Type
  - Flexor
  - Extensor
  - Mixed
- Seizures occur in clusters and frequently after arousal

Infantile Spasms

- EEG shows hypsarrhythmia
- Treatment
  - ACTH, prednisone
  - Vigabatrin
  - Valproate
  - Lamotrigine
  - Topiramate
  - Clonazepam
  - Surgery
- Outcome: most have developmental impairment
Severe Myoclonic Epilepsy of Infancy
- Initial seizures usually febrile, prolonged, focal
- Myoclonus after age 1 year
- Later partial complex and/or generalized seizures
- Seizure control is difficult
- Outcome: Cognitive impairment, some with autism

Benign Myoclonus of Infancy
- Recurrent flexor or extensor spasms
- Normal EEG, MRI, development
- Wax-wane over months
- Disappear without treatment

Gastroesophageal Reflux
- Abnormal neck, trunk, extremity posturing
- Associated with GE reflux/hiatal hernia
- Treatment of GE reflux leads to cessation
- Differentiate from other causes of dystonia

Benign Paroxysmal Torticollis
- Features
  - Intermittent lateral posturing of head/neck
  - No change in consciousness
  - Lasts minutes to days
- Onset 2-8 months
- Resolution 2-3 years
- Variable response to medication

Hyperekplexia
- Excessive stimulus sensitive startle response with fixed body posturing
- Autosomal dominant genetic disorder of glycine receptor
- Can cause feeding problems, apnea, death
- Responds to clonazepam or other benzodiazepine

Spasmus Nutans
- Features
  - Monocular nystagmus
  - Head nodding
  - Abnormal head positioning
  - No change in consciousness
- Onset 6-12 months
- Spontaneous resolution
- Differentiate from effects of intracranial lesion
Paroxysmal Events in the Preschool/School Age Years

- Seizures
- Syncope
- Sleep disturbances
- Shuddering attacks
- Migraine headache
- Stereotypies
- Rhythmic movements
- Extrapyramidal movements
- Cyclic vomiting

Seizures

- Generalized absence
- Complex partial
- Partial with secondary generalization
- Myoclonic
- Epileptic spasm

Seizures

Partial to Generalized Seizure

Absence Seizure

Myoclonic Seizure
Frontal Seizure

Why Treat Epilepsy?

- Impact on quality of life
- Risk of physical injury
- Risk of sudden death
- Limitation of future educational or employment options
- Possibility of worsening CNS function or more difficult control with untreated seizures

Seizure Treatment Options

- Symptomatic
  - Control of precipitants
    - Sleep
    - Fever
    - Exertion
    - Stimuli
  - Oral/rectal/nasal benzodiazepine
    - Lorazepam
    - Diazepam
    - Midazolam

- Status epilepticus
  - ABCs
  - Metabolic disturbance
  - Benzodiazepine
  - Longer-acting AED
    - Phenytoin
    - Phenytoin
    - Valproate
    - Levetiracetam

Seizure Treatment Options

- Prophylactic (dependent on seizure type)
  - Generalized
    - Valproate
    - Felbamate
    - Lamotrigine
    - Topiramate
    - Zonisamide
    - Levetiracetam
    - Rufinamide
    - Ethosuximide (for absence)

- Partial
  - Phenobarbital
  - Phenytoin
  - Carbamazepine
  - Valproate
  - Oxcarbazepine
  - Gabapentin
  - Tiagabine
  - Lacosamide
  - Generalized AED's (except ethosuximide)
Primary Care Options
- Start low, go slow
- Increase every 4-7 days, dependent on AED
  - Lamotrigine increase per PI schedule to minimize risk of rash
- Increase to optimize seizure control or improvement in frequency or side effects
- Change medication or seek child neurology consultation if no response or worsening of seizure frequency or severity

AED Side Effects
- Laboratory testing
  - Carbamazepine, valproate, ethosuximide
    - CBC, LFT’s
  - Oxcarbazepine
    - Sodium at 4-6 weeks after initiation
  - Lamotrigine, levetiracetam
    - No routine lab recommended
  - Monitor blood levels for determination of “room to increase” and compliance

AED Metabolism
- Carbamazepine
  - CP3A4 induction
- Levetiracetam
  - No significant interactions
  - Urinary excretion
- Valproate
  - P450 inhibition

AED Side Effects
- Lethargy/fatigue
- Nausea/vomiting
- Dizziness
- Unsteadiness
- Tremor
- Rash (Steven-Johnson – carbamazepine, lamotrigine)
- Hepatopathy (valproate, carbamazepine)
- Bone marrow suppression (valproate, carbamazepine)
- Easy bleeding (valproate)
- Newer AED’s have less serious side effects

Failure of AED Effect
- Incorrect diagnosis
- Wrong choice for epilepsy type
- Inadequate dosing
- Early termination due to side effects
- Wrong prescription
- Noncompliance

Restrictions in Epilepsy
- Good night’s sleep (avoid fatigue)
- Ensure AED compliance
- Avoid activities with no safety net
  - No skydiving or mountain climbing
- Discuss sports participation
- Review driving rules (vary by state)
When is Medication Stopped?

- Seizure control in 70%
  - 35% of those with NDD
  - 50% response to 1st AED
- Inadequate control, especially after 2 AED’s, warrants further evaluation, including surgery consideration
- Consider wean if 2 years seizure free
  - 75% are successful
  - Certain epilepsies will recur after wean
  - Patient may choose to continue AED

Syncope

Breath holding spells

- Incidence 4-5%
- Involuntary and occur during expiration
- Types
  - Cyanotic
  - Pallid
- Onset 6-18 months
- Resolution by 7-8 years
- Treatment usually not indicated

Breathholding Spell

Syncope

Cardiovascular

- Associated with structural or conduction heart abnormality
  - Tet spell
  - Arrhythmia
- Can occur with exercise
- Requires cardiology evaluation

Syncope

Reflex “Anoxic” Seizure

- Occur with syncope
- Types
  - Brief tonic/opisthotonic posturing
  - Brief repetitive myoclonic jerks
- EEG - no epileptiform activity
- Identifiable triggers
- Treatment includes explanation of nonepileptic causation and, usually, no medication

Syncope

Munchhausen by Proxy

- Form of child abuse
- Deliberate airway obstruction with loss of consciousness
- Diagnosis
  - Evaluation of other potential causes
  - Occurrence only with 1 person
Sleep Disturbances

- Types
  - Sleep walking (somnambulism)
  - Sleep talking (somniloquy)
  - Night terrors (Pavor nocturnus)
  - Restless legs syndrome
- Treatment
  - Reassurance that they are not seizures
  - Medication usually not indicated

Night Terrors

- Onset 6 months-3 years
- Body stiffening with extremity adduction and quick “shiver”
- Lasts seconds
- May occur with upset or mealtime
- Resolve by 10 years old

Restless Legs Syndrome

- Episodes of cessation of activity, pallor, light/noise intolerance, whining/upset that may include head banging
- Periodic occurrence
- May be triggered by dietary component
- Treatment
  - Elimination diet/identification of triggers
  - Acute therapy
  - May need prophylactic treatment

Migraine Headaches

- Rhythmic and recurrent motor mannerisms
  - Fingerflicking
  - Object regard and spinning
  - Hand wringing
  - Tongue/mouth pulling
  - Hand flapping
Stereotypies

- Occur in:
  - Normal children
  - Autism
  - Rett's syndrome
  - Smith-Magenis syndrome
  - Sever/profound cognitive impairment
  - Children with visual impairment (blindisms)
  - Child neglect

Rhythmic Movements

- Types
  - Rocking
  - Head banging/rolling
  - Masturbation
  - Hand flapping with excitement
- Resolve spontaneously or by addressing underlying trigger
- More frequent with significant developmental impairment or visual impairment

Extrapyramidal Movements

- Recurrent episodes of nausea/vomiting lasting hours to days
- Onset in preschool years
- Related to migraine headaches
- Exclude organic causes
- Treatment
  - Supportive fluid administration
  - Antiemetics
  - Migraine therapy

Cyclic Vomiting

Paroxysmal Events

- May be seizures if:
  - Stereotypic
  - Nonsuppressible
  - Associated with change in consciousness
  - Not stimulus sensitive