

Status Epilepticus and Childhood Seizures

Childhood Seizures

Neurology: Location, Location, Location (Primarily Anatomical)

Child Neurology: *Location, Location, Location*, Anatomy, Time, Space, DNA, situation

Seizure Semiology

Describe features: Don't just label "seizure". **Be specific:** Screams out, falls to ground, head turns to left, eyes deviate to left and up, drools, hand flexed, legs extended for 4 minutes with urinary incontinence, followed by confusion for 10 minutes, then slept for 30 minutes awakening with headache and soreness of body and tongue.

What is a Seizure? Excessive, synchronous discharge of neurons time locked to abnormal behavior, sensory or motor activity. Unpredictable as to when a seizure may happen. **May be a** single event or recurring episodic. Occur in 4-5% of children. Stereotypic with similar features whenever they occur. Usually brief and last only seconds to a few minutes.

Except for: Status Epilepticus which is prolonged seizure activity.

Seizures need First Aid but may not need long term treatment.

Seizure First Aid consist of 1). Position on side to open & drain airway, 2). Cushion or protect head and body from harm. 3). Do NOT put anything in the mouth 4). Helpful to time the seizure. 5). Initiate treatment if found seizing (don't know when it started) or seizures lasting longer than 5 minutes. Need a rescue plan and rescue therapy.

Approach to Childhood Seizures:

1. Status Epilepticus
2. Single Seizure or First Seizure: May be "Provoked" or "Unprovoked"
3. Recurring Seizures: Epilepsy
4. Special Seizure Situations: Neonatal Seizures, Infantile Spasms, and Febrile Seizures

Neonatal Seizures: Seizures in the first month of life

Clinical classification: Four types of Neonatal Seizures

1. Tonic: extension of arms and legs, decerebrate posturing
2. Subtle: sucking, rowing pedaling, abnormal eye movements
3. Clonic: focal or multi-focal, migratory
4. Myoclonic

Etiology: Hypoxic Ischemic Encephalopathy
Infection: GBS, HSV, E. coli, Listeria
Metabolic: Glucose, Na⁺, Ca⁺⁺, Mg⁺⁺
Drug: toxicity and withdrawal
Structural: hemorrhage, malformation
Genetic: B-6 Dependency and Familial Neonatal Convulsions

Evaluation of Neonatal Seizures:

History: Prenatal, Birth, Discharge, Feeding
Physical: HC, AF, sutures, fundi, skin
Dextrostix, Septic screen-W/U, CMP, NH₃,
Imaging: Cranial US vs. CT vs. MRI

Treatment: Treat and Reverse cause if possible

Pyridoxine (B-6) 100 mg IV
Phenobarbital 20 mg/kg IV, may repeat 1-3 times over 24 hrs
Fosphenytoin 20 mg/kg P.E., may repeat x 1

Infantile Spasms: Syndrome of clusters of myoclonic seizures in infancy (West Syndrome)

Onset 4-12 months, multiple causes

Progressively worsen: Number, duration

Common on first awakening

Developmental plateau and regression

Poor treatment options: Early Rx best

Long term impairments and epilepsy likely

Febrile Seizure: Seizures triggered by fever from 6 months to 5 years of age without CNS infection. Neurodevelopment not defined.

Simple Febrile Seizure: Short, generalized, non-recurring in 24 hours

Complex Febrile Seizure

Prolonged (> 15 minutes)

Focal

Multiple per 24 hours

Evaluation: Confirm is Febrile Seizure by history, exam and if needed testing.

Lumbar Puncture if: Any concern for meningitis or encephalitis, Less than 18 months old

Acute Treatment: Lorazepam or phenobarbital if multiple, load and observe

Diastat® for future prevention (>2 yrs, 10 kg)

Rectal diazepam (Diastat®) Used as “Rescue Therapy” and as alternative when I.V. not available. Used for prolonged, recurrent, and for history of prolonged or recurrent or Family History of complex febrile seizures. Dose based on age and weight. Diastat Acudial® 2.5 mg, 5-10 mg, and 12.5-20 mg strength.

Epilepsy: Many different conditions with recurring seizures.

Symptomatic Epilepsy: Condition with recurring seizures and evidence of preceding brain insult by history, exam or imaging (*physically broken*)

Epileptic Syndrome: Condition with recurring seizures with characteristic features (Clinical and EEG) that develop and occur over a specific age range. Several are of Genetically based with known DNA mutations.

–Childhood Absence Epilepsy

–Benign Rolandic Epilepsy

–Benign Familial Neonatal Epilepsy

–GEFS+

–Panayiotopoulos Syndrome

–Doose syndrome

–Dravet’s Syndrome

–West Syndrome

–Lennox Gastaux Syndrome

Work Up: Performed at time of presentation and includes labs, EEG, MRI. Not every subsequent seizure needs repeat testing. May repeat testing if unexpected change in course, or failure to respond to therapy or follow expected natural history.

Treatment is based on Seizure type or epileptic syndrome. With goal: of 1). No Seizures 2). No Side Effects and 3). Having a normal life. Unfortunately, often can not achieve this and have to reach a compromise based on “Risk vs. Benefit” for drug, age, side effects. Alternatives to medications for REFRACTORY Epilepsy include: Surgery, VNS, and Ketogenic Diet.

Childhood Seizures

1st Single Seizure (Unprovoked): 2-4% Children have an event worrisome for Seizure. Must exclude: Syncope, Breath Holding, tantrums, Vertigo, night terror. In children with a first seizure, 40% recur consistent with Epilepsy

Evaluation for 1st seizure: Full history including, Family History, Review of Systems. Details of preceding 48+ hours may be needed to determine unprovoked. History of any jerks, stares, drops? Physical and Neurological Exam: Normal or abnormal (? provoked)

Labs: Consider (CRP, CMP, Mg⁺⁺, UDS). CT Scan in ER is practical and generally performed for presentation for first and often recurrent seizures. EEG can be helpful to evaluate for focus or epileptic syndrome but can be done at a later date, especially if given anticonvulsant medication. Does not need long Anti-convulsant Medication. Family does need Seizure First Aid information and consider Diastat.

1st Provoked Seizure: Seizure triggered by an acute medical, metabolic, toxic, traumatic or infectious disturbance of the brain. Needs more evaluation. Consider any child in hospital for non seizure diagnosis that has a seizure in the hospital has a proved seizure. Evaluation: H&P, Labs, CT, LP. Treat cause of disturbance and consider “Brain Salve” to be used for variable duration. Fosphenytoin 20 mg/kg P.E. IV, Phenobarbital 20 mg/kg IV, Lorazepam 0.1 mg/kg (max 4 mg) IV, or Levetiracetam 20 mg/kg IV

Status Epilepticus: 1993 Classical Definition: 30 minutes or longer of:
 Continuous seizure activity
 Repeated seizures without regaining consciousness

Status Epilepticus: Practical Definition: 10 minutes or longer of:
 Continuous or repeated seizures
 Seizures on arrival in E.R. or when seen
 Seizure recurring in E.R. or hospital

Convulsive Status Epilepticus: Tonic Clonic Sz → Tonic Clonic Status: **Medical Emergency**
 Non-convulsive Status Epilepticus: Absence or Partial Seizures needs appropriate treatment.
 Refractory Status Epilepticus is continued seizure after 60 minutes of appropriate therapy.
 More than 50% of people who develop status epilepticus have no past history of epilepsy
 Incidence: 50-250,000/year an onset varies by age (More frequent in younger children)

Causes of Status Epilepticus:

	<u>Children</u>	<u>Adults</u>
Idiopathic	30%	33%
Acute symptomatic	35%	50%
Fever	25%	0%
Remote Symptomatic	15%	10%
Progressive CNS Dz	5%	10%

Precipitants of Status Epilepticus

	<u>Children</u>	<u>Adults</u>
Cerebrovascular	3.3%	25.2%
Med change	19.8%	18.9%
Anoxia	5.3%	10.7%
Fever	35.7%	4.6%
CNS Infection	4.8%	1.8%
Congenital	7.0%	0.8%

Physiologic Consequence of Status Epilepticus

- Hypoxia, Hyperthermia, Acidosis
- Increased catecholamine release
- Leukocytosis
- Increased BP and Pulmonary vascular pressure
- Pulmonary trans-vascular fluid flux
- Increased Cerebral Blood Flow (CBF)
- Increased cerebral metabolic activity
- ? Apoptosis - programmed cell death

Sequelae of Convulsive Status Epilepticus

None: complete recovery

Residual Neurological Disability

Mental Handicap

Motor Deficit

Epilepsy

Death ! Children 2-4%, Adults up to 25%

Status Epilepticus: Mortality: Less likely in children (except progressive CNS disease), More favorable outcome with febrile and idiopathic status, much higher if over 1 hour in duration. (Childhood death 1-2 % < 1hour but 4-5 % > 1 hour.)Outcome Determined By: Cause of the Seizures, Age of Patient, Duration of status, and Treatment. **Can influence only the treatment.**

Treatment and Evaluation of Status Epilepticus requires recognition that you are dealing with Status Epilepticus (identify features of seizure) and following a standard protocol. Have a plan, Treat intravenously, Therapeutic endpoint is cessation of convulsions, **Be prepared to ventilate, use adequate doses of anti-seizure medications, Treat all status patients the same, and Intensive Care is needed.**

Status Epilepticus Treatment Outline:

0 to 5 Minutes

Diagnosis: Identify features of the seizure, Get History: Baseline? Illness? Epilepsy?

Stabilize Airway: Head & Body Position, Dextrostix, Pulse Ox, Oxygen

5 to 10 minutes

Start IV Line, Dextrose

Draw Lab work (Na+, Glucose, Ca++, Mg++, AED Levels, Toxicology screen, CBC, CRP, Blood Cultures)

Dose #1 of IV Benzodiazepine (BZD)

10-20 minutes

Dose #2 of IV Benzodiazepine

Anti-Convulsant (AED) #1 Load: Phenobarbital 20mg/kg (<2 yo) or Fosphenytoin 20 mg/kg P.E.

Consider and start antibiotics and antivirals

30-60 minutes

Intubation: (Sedation treatment or still seizing), ventilation, BP support in ICU

Anti-Convulsant #2 Load: Phenobarbital 20mg/kg or Fosphenytoin 20 mg/kg P.E.

Pharmacologic Coma, (EEG monitoring to titrate dose not to start)

CT scan or LP when stable if needed

Refractory Status Epilepticus: Continuous status after **1 hour** of aggressive therapy

BARBITUATES: High dose Phenobarbital (levels 50-60 +) or Pentobarbital

BENZODIAZEPINES: IV drip of diazepam, lorazepam, or midazolam

Anesthetic Agents Propofol

Neuroimaging: CT: Evaluate for blood, calcification, trauma, or shift from mass.

Lumbar Puncture: Fever with first seizure, Fever with Seizure less than 18 months of age, or History or Symptoms suggestive of Meningitis/encephalitis.

Status Epilepticus: Easily Missed Diagnoses:

Meningitis and encephalitis

Abuse or Unsuspected trauma: SHAKEN BABY SYNDROME

Retinal hemorrhages are an important finding on exam, subdural on CT.

Drug Ingestion

Contraindications to LP

Cardiovascular Instability or Respiratory compromise

Intra-cranial mass with increased ICP, Impending herniation, papilledema, coma

Infection of skin at LP site

Bleeding disorder

IF LP is contraindicated: 1).Treat Patient with Status protocol 2). Draw Blood Cultures 3). Start anti-infective therapy (Duration can be re-evaluated when patient is stable)

The decision to start long term therapy should be individualized. Patients and families should know there are options and uncertainties. Patients at low risk of seizure recurrence should not be treated long term Patients at high risk of seizure recurrence should be treated long term

Long Term Therapy Indicated: Recurrent Status or previous seizures, Structural Brain Abnormality, Progressive Neurological Disease, Certain Epileptic Syndromes.

No Long Term Therapy for Acute CNS disturbance that resolves, i.e. Febrile Seizures, Drug intoxication or withdrawal, Electrolyte imbalance, or early seizures with uncomplicated meningitis.

Summary: Status Epilepticus

Time	Evaluation	Treatment
0-2 minutes	Diagnosis	ABCs
3-5 minutes	IV, Labs	BZD #1
10 minutes		BZD #2
15 Minutes		AED #1
20-60 minutes	Intubate	AED #2
60+ minutes	CT, LP	Pentobarbital

Summary: First Seizure

History: Full Hx, Family Hx, ROS. Details of preceding 48+ hours; any jerks, stares, drops?

Exam: Normal or abnormal (unprovoked v. provoked). Labs based on circumstance, CT, LP.

Treat cause of disturbance if provoked 1st seizure. Consider “Brain Salve” for variable duration if recurrent provoked. No treatment for 1st Unprovoked.

Emory Division of Pediatric Neurology Web site:

www.pediatrics.emory.edu/divisions/neurology

Handout for Acute Care Symposium handout:

www.pediatrics.emory.edu/divisions/neurology/acs2009neurology.pdf

Handout for Epilepsy by Dr. Larry Olson:

www.pediatrics.emory.edu/divisions/neurology/Epilepsyldo.pdf

Medications Used for treatment of seizures and status epilepticus:

Selected and incomplete information

LORAZEPAM: Loading Dose of 0.1 mg/kg (2 mg max) and repeat dose 0.1 mg/kg (2 mg max).

Expected onset in 5-10 minutes with a duration of 12 to 24 hours.

Major Side Effects: Depression of consciousness for hours. Infrequent respiratory and BP depression, 3rd or more doses not better than 2nd dose.

DIAZEPAM: Loading Dose of 0.15 to 0.25 mg/kg may repeat dose 0.15 to 0.25 mg/kg.

Expected onset in 1-3 minutes with duration or only 15-30 minutes

Major Side effect of depression of respiration and consciousness for 10-30 minutes.

PHENOBARBITAL: Loading Dose of 20 mg/kg with need to repeat dose of 20 mg/kg.

Expected onset in 10-30 minutes and duration of 12-48 hours.

Major Side Effects: Depression of Consciousness lasting hours to days, dose related respiratory depression (marked with diazepam, less so with lorazepam).

PENTOBARBITAL: Loading Dose of 5-12 mg/kg, then 5mg/kg q 20 min. to effect and

maintenance dose of 1-10 mg/kg/hr for 12-48 hours to ? Number of days

Need EEG monitoring to adjust dose based on "burst suppression"

FOSPHENYTOIN: Loading Dose of 15-20 mg PE /kg. Repeat dose X 1 of 10 mg PE /kg.

Expected onset in 5-10 minutes and duration +/- 24 hours.

Major Side Effects: Infrequent respiratory depression, Hypotension and Bradycardia. Has a pH=7.7, does not cause a chemical phlebitis and mixes with glucose.

PHENYTOIN: Loading Dose of 15-20 mg/kg. Repeat dose X1: 10 mg/kg

Expected onset in 5-10 minutes with duration of +/- 24 hours.

Major Side Effects; Hypotension and Bradycardia: if given > 1 mg/kg/min, Infrequent respiratory depression. Has a pH=11, causing phlebitis, and does not mix with glucose. Very caustic to skin with severe tissue injury if extravasates from vein. No longer needed on market with generic fosphenytoin available. Requested to be removed from formulary.

MIDAZOLAM: Loading Dose of 150-200 µg/kg maintenance dose of 1-10 µg/kg/minute. Mean dose (Rivera et. al. 1993) of 2.3 µg/kg/minute

PROPOFOL: Loading Dose of 3 - 5 mg/kg with maintenance dose 1-15 mg/kg/minute. Side effects respiratory arrest, rapid infusion, and abrupt dose decrease: "seizure-like" movements, acidosis. Egg Allergy.