

# Epilepsy Management: Treatments, emergencies, SUDEP

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# Disclosures

1. RO1 NS076577 (Benke)

NIH/NINDS

Molecular mechanisms linking early life seizures, autism and intellectual disability

Role: PI.

2. Questcor Pharmaceuticals (Benke)

Whole-exome sequencing and ACTH responsiveness in Infantile Spasms

Role: PI.

3. 1U10NS077277(Vollmer)

NIH/NINDS

Rocky Mountain Network for Neuroscience Clinical Studies (NeuroNext Clinical Site)

Role: Co-I.

4. U54 HD061222 (Percy)

NICHD

Rett syndrome, MECP2 Duplication Disorder, and Rett-related Disorders Natural History.

Role: Site Director, Co-I.

5. Rett Clinic at Children's Hospital Colorado (Benke)

Rocky Mountain Rett Association

Role: PI/Medical Director

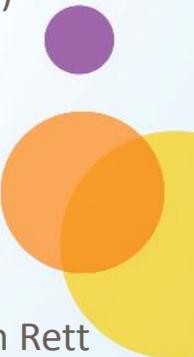
6. CDKL5 Center of Excellence (Benke)

International Foundation for CDKL5 Research

Role: PI

7. Neuren: Study of trofinetide, also known as NNZ-2566, for females with Rett Syndrome

Role: site PI



# Topics today



- Key items to discuss with your neurologist about seizures and epilepsy
- Question and answer time

# Key items to discuss with your Neurologist



- Review current treatments
- Create seizure plans for home and for school
- When to call your neurologist
- When to see your neurologist
- Procedures/therapies/referrals that should be done or discussed for your child



# Review current treatments

Suggested questions for your neurologist:

What is the treatment plan for this medicine?

Q: What is the starting dose?

A: usually in mg/kg/d

Q: When should it be increased or decreased (how soon should I call you)?

A: usually depends on the half-life of the drug

Q: what are the side effects to watch for? Any that need additional testing? Can this medicine be used for an extended period of time?

A: Rash. Sedation. Irritability? Blood or liver toxicity? Other toxicity?

Q: How will we know if the medicine is working? Do we need another EEG to follow this? When? How do you want me to keep a diary?

A: EEGs are more accurate for reporting seizure frequency than parental or self report. Keeping a diary is the next best thing. There is an app for that: [seizuretracker.com](http://seizuretracker.com)



# Create Seizure Action Plans

What do I do?

- Safe place, don't panic
- Tummy or side, nothing in mouth
- Time it
- Video it

What do I tell others (family/friends/care-givers)?

What are rescue medications? When are they given? What are the side-effects?

When do I call 911? Go to ER?

When do I call the neurologist?



# Rescue medicines

IR Valium (“Diastat”)

IN midazolam (“Versed”)

- Key side effects: sedation and respiratory depression
- Which is better?
- How do I use this? Can you show me? (practice)
- When to use? (discuss with neurologist)
- When to repeat?
- When to call 911?
  - First use ever?
  - Repeated use?



This student is being treated for a seizure disorder. The information below should assist you if a seizure occurs during school hours.

Student's Name _____	Date of Birth _____	
Parent/Guardian _____	Phone _____	Cell _____
Other Emergency Contact _____	Phone _____	Cell _____
Treating Physician _____	Phone _____	
Significant Medical History _____		

### Seizure Information

Seizure Type	Length	Frequency	Description

Seizure triggers or warning signs: \_\_\_\_\_

Student's response after a seizure: \_\_\_\_\_

### Basic First Aid: Care & Comfort

Please describe basic first aid procedures: \_\_\_\_\_

Does student need to leave the classroom after a seizure?  Yes  No  
 If YES, describe process for returning student to classroom: \_\_\_\_\_

### Basic Seizure First Aid

- Stay calm & track time
  - Keep child safe
  - Do not restrain
  - Do not put anything in mouth
  - Stay with child until fully conscious
  - Record seizure in log
- For tonic-clonic seizure:**
- Protect head
  - Keep airway open/watch breathing
  - Turn child on side

### A seizure is generally considered an emergency when:

- Convulsive (tonic-clonic) seizure lasts longer than 5 minutes
- Student has repeated seizures without regaining consciousness
- Student is injured or has diabetes
- Student has a first-time seizure
- Student has breathing difficulties
- Student has a seizure in water

### Emergency Response

A "seizure emergency" for this student is defined as:

#### Seizure Emergency Protocol (Check all that apply and clarify below)

- Contact school nurse at \_\_\_\_\_
- Call 911 for transport to \_\_\_\_\_
- Notify parent or emergency contact
- Administer emergency medications as indicated below
- Notify doctor
- Other \_\_\_\_\_

### Treatment Protocol During School Hours (include daily and emergency medications)

Emerg. Med. <input checked="" type="checkbox"/>	Medication	Dosage & Time of Day Given	Common Side Effects & Special Instructions

Does student have a Vagus Nerve Stimulator?  Yes  No If YES, describe magnet use: \_\_\_\_\_

### Special Considerations and Precautions (regarding school activities, sports, trips, etc.)

Describe any special considerations or precautions: \_\_\_\_\_

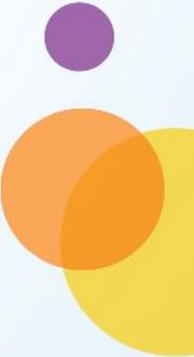
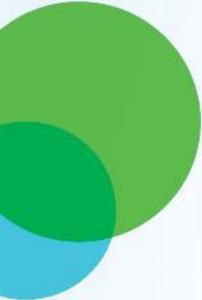
Physician Signature \_\_\_\_\_ Date \_\_\_\_\_

Parent/Guardian Signature \_\_\_\_\_ Date \_\_\_\_\_

## Seizure Action Plan For School/daycare

### Goals:

- Seizure first aid
- When to use rescues
- When to call 911





# SUDEP: Sudden unexplained death in epilepsy. Is my child at risk?



*">20% of patients with childhood-onset epilepsy who fail to achieve long-term seizure freedom will die of SUDEP within 40 years of follow-up (Sillanpaa & Shinnar, 2010)."*

## Risk factors

- Medically refractory pediatric epilepsy
- Generalized tonic-clonic seizures
- Anticonvulsant withdrawal/changes?
- Post seizure? Nocturnal seizures?
- Prone position while sleeping?
- Arrhythmias?
- Link to anticonvulsants/polytherapy controversial but unsupported

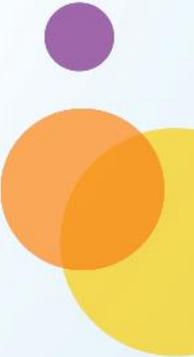
## Prevention

- Unknown
- After seizure: Repositioning? Supplemental oxygen?
- EKG? Diagnostics?
- Lattice pillows?
- Monitoring?

## Research

- Do we need a registry? Harvard Brain Bank/Alica Goldman/Baylor/Houston

*Rylvin, 2013*





# When to call your neurologist

- When you are concerned!
- When you see something unexpected!
- Worsening seizure duration or frequency. Ask them to clarify.
- Worsening side effects of medications. Ask them to clarify.

## ➤ Emergencies

Status epilepticus: seizures longer than 5/10/15 minutes, or clusters without regaining consciousness over 30 minutes, not responding to rescue medications: call 911.

Ask neurologist to clarify.



# What to tell your neurologist

- 1) The medicines currently taken and the doses you are giving.
- 2) Side effects you are concerned about
- 3) Spell characterization.
  - How many types of spells?
  - Time/action of day? (*before sleep*)
  - What happens first? (*rhythmic movements, scream, 20 s*)
  - Then what? (*extension of arms, flexion of legs, eyes open and turned to right, 30s*)
  - Then what? (*cluster of 8 spasms, 5 min*)
  - Then what? (*stops, sleeps for 1 hour*)
  - Total duration. Color change? Emesis?
  - How often (*three times daily for last 3 weeks, was monthly, here is my calendar...*)



# When to see your neurologist

If seizures are controlled: every 6-12 months, depending on medication monitoring

If seizures are not controlled: every 0.5-4 months, depending on medication monitoring

When you go:

- Take your list of questions (keep a binder)
- Take your seizure calendar
- Take your medicine bottles
- Take your cell phone with spell videos

Before you leave the office:

- Get a written after-visit summary of instructions that answers your questions
- Request copy of visit note sent to you (to keep in your binder)
- Request copy of EEG or other results sent to you (to keep in your binder)

# When to see your neurologist (cont'd)



How about an EEG?

- Prolonged (1-3 days) is better, if the goal is to capture a spell.
- When spells change in character or frequency
  - Are they really seizures? (Rett spells)
- Yearly, perhaps less if seizures are controlled
  - Has the background changed? Are there seizures during sleep that are being missed?
  - Unexplained regression?
  - Are we missing anything?

# When to see your neurologist (cont'd)



MRI?

Probably not, unless specifically indicated for other reasons.

Other tests?

- Spine films if scoliosis observed
- EKG? SUDEP prevention? We don't know: I suggest yearly.

What else?

- Have you seen a genetic counselor or geneticist? Did the neurologist/diagnosing physician answer all of your questions about the genetic basis of the diagnosis?
- Rehabilitation, ophthalmology, GI
- Therapy referrals
- Equipment needs
- Seizure action plan for school
- How much will my medicines cost? (GoodRX.com)



# Medication guidelines

Avoid multiple (>2-3) medications; 3 is ok if there is a plan to get rid of one.

Rett: valproate? avoid levetiracetam?

CDKL5: valproate? Valproate + clobazam?

“Only the right dose differentiates between a poison and a remedy”

Paracelsus, 1493-1541



ELSEVIER

Official Journal of the European Paediatric Neurology Society

**Table 1 – Responder rate of AED's after 3, 6 and 12 months.**

AED	n	Responder rate after 3 months	Responder rate after 6 months	Responder rate after 12 months
VPA	34	7 (21%)	6 (18%)	3 (9%)
LEV	31	5 (16%)	4 (13%)	0
TPM	31	5 (16%)	1 (3%)	1 (3%)
Steroids/ ACTH	26	5 (19%)	0	0
PB	26	2 (8%)	2 (8%)	2 (8%)
VGB	25	8 (32%)	2 (8%)	1 (4%)
LTG	23	5 (22%)	2 (9%)	2 (9%)
LTG	23	5 (22%)	2 (9%)	2 (9%)
CLB	17	4 (24%)	1 (6%)	0

## Original Article

## Retrospective evaluation of low long-term efficacy of antiepileptic drugs and ketogenic diet in 39 patients with CDKL5-related epilepsy



A. Müller<sup>a</sup>, I. Helbig<sup>b,c</sup>, C. Jansen<sup>a</sup>, T. Bast<sup>d</sup>, R. Guerrini<sup>e</sup>, J. Jähn<sup>b</sup>, H. Muhle<sup>b</sup>, S. Auvin<sup>f</sup>, G.C. Korenke<sup>g</sup>, S. Philip<sup>h</sup>, R. Keimer<sup>i</sup>, P. Striano<sup>j</sup>, N.I. Wolf<sup>k</sup>, B. Püschel<sup>l</sup>, Ch. Thiels<sup>m</sup>, A. Fogarasi<sup>n</sup>, S. Waltz<sup>o</sup>, G. Kurlemann<sup>p</sup>, T. Kovacevic-Preradovic<sup>q</sup>, B. Ceulemans<sup>r</sup>, B. Schmitt<sup>s</sup>, H. Philipp<sup>t</sup>, D. Tarquinio<sup>u</sup>, S. Buerki<sup>v</sup>, C. von Stülpnagel<sup>a,w,\*,1</sup>, G. Kluger<sup>a,w,\*,1</sup>

**Significance:** Considering both age-related and spontaneous fluctuation in seizure frequency and the unknown impact of many AEDs or KD on cognition, our data may help defining realistic treatment goals and avoiding overtreatment in patients with CDKL5 mutations. There is a strong need to develop new treatment strategies for patients with this rare mutation.

MSA	3	0	0	0
FBM	3	3 (100%)	2 (67%)	1 (33%)
LCM	3	0	0	0
STP	3	0	0	0
PRM	2	0	0	0
Ketogenic diet	12	2 (17%)	2 (17%)	1 (8%)

LEV: levetiracetam; VPA: valproic acid; TPM: topiramate; PB: phenobarbital; VGB: vigabatrin; LTG: lamotrigin; CLB: clobazam; RUF: rufinamide; CBZ: carbamazepine; STM: sultiame; ZNS: zonisamide; PHT: phenytoine; ESM: ethosuximide; CLN: clonazepam; BR: Bromide; MSX: mesuximide; FBM: felbamate; LCM: lacosamide; STP: stiripentol; PRM: primidone.



# What about herbal/natural remedies? (See session later today!)



There is no data to support the use of CBD as an anticonvulsant. There is data that demonstrates CBD lowers IQ in the developing brain.

## Statement from the American Epilepsy Society:

The use of marijuana for various medical conditions, including epilepsy, has received increasing attention in the popular media. While there are some anecdotal reports of marijuana use in treating epilepsy, scientific evidence for the routine use of marijuana for this indication is lacking. The lack of information does not mean that marijuana is ineffective for epilepsy. It merely means that we do not know if marijuana is a safe and efficacious treatment for epilepsy. Healthcare professionals, patients, and caregivers are reminded that use of marijuana for epilepsy may not be advisable due to lack of information on safety and efficacy, and that, despite some states legalizing the use of medical marijuana, it is against Federal Law to possess or use marijuana. In addition, little is known about the long term effects of using marijuana in infants and children, and chronic exposure during adolescence has been shown to have lasting negative effects on cognition and mood. Such safety concerns coupled with a lack of evidence of efficacy in controlled studies result in a risk/benefit ratio that does not support use of marijuana for treatment of seizures at this time. **The American Epilepsy Society is supportive of well-designed research to determine the safety and efficacy of marijuana in the treatment of epilepsy.**

### References:

Gloss D, Vickrey B. Cannabinoids for Epilepsy. Cochrane Database of Systematic Reviews.2012; Issue 6. Art. No.: CD009270. doi: 10.1002/14651858.CD009270.pub.2

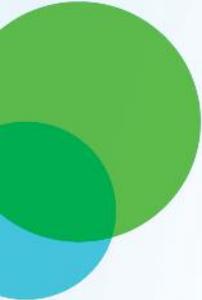
A commentary on the article is available at: [Epilepsy Currents, Vol. 13, No. 2 \(March/April\) 2013 pp. 81-82](#)

### Other references:

<http://www.epilepsy.com/>

<http://www.epilepsyfoundation.org/news/On-Medical-Marijuana-and-Epilepsy.cfm>

<http://www.miamiherald.com/2014/01/22/3886526/epilepsy-experts-call-for-more.html>

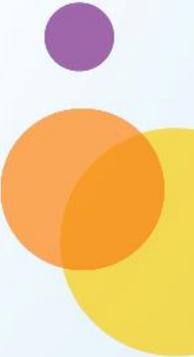


# Summary

## Medical advocacy with your neurologist

- Plan ahead
- Be prepared
- *Forewarned, forearmed; to be prepared is half the victory (Miguel de Cervantes)*

## Medical advocacy with your community

- Be supportive
  - *A community is like a ship; everyone ought to be prepared to take the helm. (Henrik Ibsen).*
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# Special Thanks!!!

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## Our patients and families

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### NHS-Natural History Study:

Eric Marsh, Alan Percy, Jeff Neul, Alex Paciorkowski, Laura Mamounas, Tim Roberts, Chuck Nelson, Sar Peters, Michela Fagiolini, Walter Kaufmann, etc

### **Rocky Mountain Rett Association**

Ponzo Family Chair in Neuroscience  
Research



questions