Rescue Medications in Severe Developmental and Epileptic Encephalopathies (DEEs) Series

Part 2: Rescue Medications… A Clinicians Perspective & Advice

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RESCUE MEDICATION PLAN Part 2:

What Does This Mean to My Individual Child's Condition?

RECAP of Part 1 from Rescue Medication Series:

LGS FOUNDATION
LENNOX-GASTAUT SYNDROME

DEE-P CONNECTIONS

RESCUE MEDS SERIES: PART 1

Rescue Medications in Severe DEEs , including LGS

JUNE 12TH. 2020

10am PT I 11am Mountain
12pm CT I 1pm EST

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Learn about administering rescue meds and the value of seizure action plans (SAP).

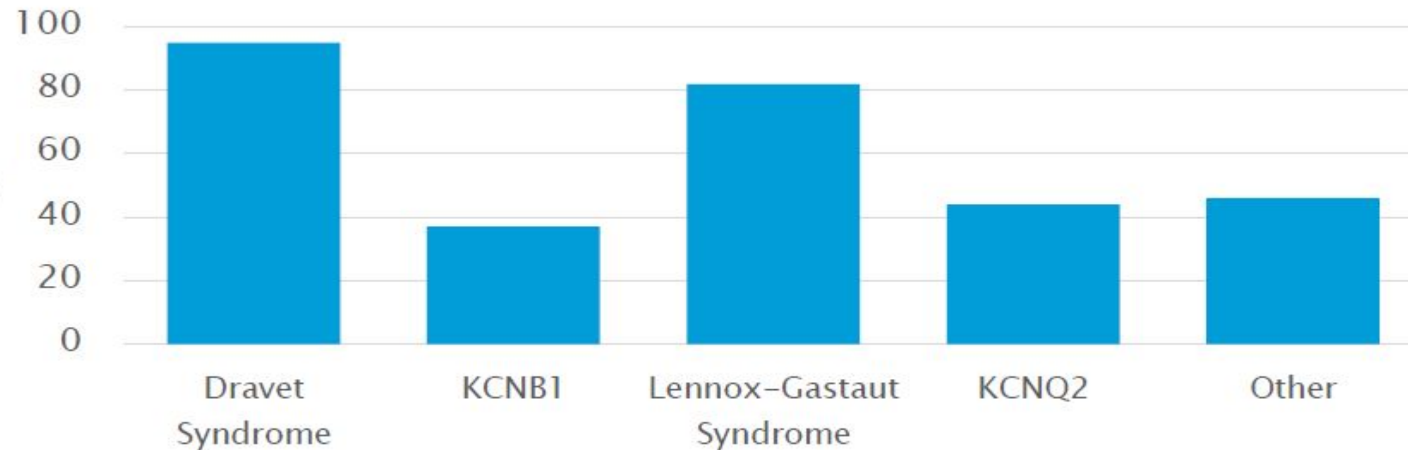
- DEEs, including the DEE known as LGS, cases have increased seizure frequency and increase need for rescue medications
- Many available rescue medications and routes of admission
- 40-50% of DEE families lack a Rescue Plan
- May have different seizure types and seizure patterns requiring rescue medication

DEE cases have increased seizure frequency and increase need for Rescue Medications

Seizure Emergencies

- 65% ever have had a seizure or seizure cluster >10 minutes

Prolonged seizure by type of epilepsy



Rescue Medication (RM) Use in DEEs

- Convulsive Seizures: focal or generalized **clonic or tonic clonic**
- Non-Convulsive Events: **Not Tonic clonic** can be myoclonic, tonic , atonic
- Absence or atypical absence
- Partial Seizures with impaired consciousness or sensory seizures

Differences in RM use in DEEs

- Children often having very frequent seizures. How does one define important worsening?
- RM is often used more frequently
- AND used differently:
  - For prolonged convulsive seizures – but often may not wait 3-5 minutes!
  - For clusters of convulsive seizures
  - For clusters of nonconvulsive seizures?

Many Rescue Medication Choices Exist

- Diastat Rectal gel
- Nasal Sprays (Valtoco and Nayzilam)
- Dissolvable Strip Sympazam
- Oral dissolvable tablets (ODT) such as clonazepam, ativan, etc
- **Only Diastat, Valtoco, and Nayzilam are FDA approved for Rescue Seizure Clusters, others may be used offline**

Case Examples: Patients with DEEs

- Case Example: Child with Phelan-McDermid Syndrome and has frequent status epilepticus. Was already on clobazam at 60 mg per day: Needed 3-4 doses of Diastat totaling 40-60 mg/night several nights per week to stop status epilepticus
- Case Example: Child with Dravet Syndrome with any fever goes into status epilepticus > 40 minutes often requiring intubation, she weighed 21 KG, was already on Clobazam and Epidiolex valproic acid and levetiracetam. The Mom was told she could not give more than 10mg diastat however if given 20 mg seizures usually stopped within 5-10 minutes no longer requiring ER visits.
- Case Example: Child with LGS of unknown cause, was on onfi and other medications. This child required 60 mg of diastat to stop seizures at weight 60 kg as 20 mg always failed to stop the status.

Case Examples: Patients with DEEs

- Case Example: Child who suffered lack of oxygen at birth (hypoxic injury) and later developed LGS, mainly has seizures with fever and told give diastat or dose of valium every 6 hrs if fever over 100.5 degrees for first 24 hrs of their illness along with Tylenol or ibuprofen to prevent seizure clusters or status, however family almost always still goes to ER after calling 911 despite teaching for years that they do not need to do that if they were comfortable giving rescue meds to prevent emergency.

- Case Example: Child with LGS with recurrent clusters of seizures around her period with her tonic seizures more frequent, so started using phone app for menses and give extra onfi two days before starts and two days after.

Individual Rescue Medication Plan: Seizure Rescue Plan (SRP, may also be called Seizure Action Plan)

- IEP: Just like you work with schools to make an individual education plan that marks out problems and goals each patient with DEEs, including LGS, also needs individual planning for Seizure Management
- Individual Seizure Rescue Plan (I-SRP): Recognize each patient for their individual Seizure Types and Individual Seizure Patterns
- The SRP is only a part of overall Seizure Management Plan

Seizure Rescue Plan (SRP): Things to Know

- Caregiver needs to know essential basic facts to make a seizure plan work and to make the I-SRP work, you need overall seizure plan to make rescue plan make sense
- Gaps in care commonly noted: Caregivers do not often know name or dose of current daily Anti-Seizure Medication (ASM) which is first step
- Know times and doses of medication that are regularly to the child
- This is TEAM JOB. Needs patient caregiver and physician together, neither can do alone
- Need to have good relationship with primary physician to discuss these issues working together to establish these goals

Seizure Rescue Plan (SRP): Things to Know

- Know Which Medications are aimed at which type of seizure you are trying to treat.
- Focal vs convulsive generalized vs non-convulsive seizures may have different medications. Treating different types of seizures in individuals with more than 1 type of seizure which is most DEE, is always difficult but is doable.
- Know individual patient issues and sensory or oral motor issues that may make certain routes for medication more likely to work and be absorbed.

Seizure Rescue Plan (SRP): Things to Know

- Know where medications are at home and when traveling and how to access them. Don’t forget to take medications when go to ER or hospital.

- Know basic Seizure Rescue First Aid: How to position patient and how not to worsen risks and when to call for help. Learn what to watch for during a seizure, like breathing changes and preventing injury.

- Know baseline Seizure frequency and triggers: example know if miss dose and know if fevers increase risks for more seizures, clusters, or status.

SRP: 10 Seizure Rescue Plan Steps

- Step 1: Know your loved one's diagnosis and if possible when last EEG and MRI were done, if they have a genetic condition, if they have any allergic reactions, and medications they take now.

- Step 2: Know the frequency and types of seizures that occur daily, weekly, monthly in your loved one.

- Step 3: Know when your loved one is having seizure and when they stop. You may want to use the lack of difference as your signal for when to give a rescue medication (for example; ie Defining emergencies clearly)

SRP: 10 Seizure Rescue Plan Steps

- Step 4: Know your baseline daily maintenance medications, their names, doses, time of day given, and most recent drug monitoring levels if available.
    - I recommend having written list stored on phone or online or on paper copy listing all medications and times and amount of doses.

- Step 5: Know if is there any medication your loved one is on that may affect rescue medication efficacy

- Step 6: mark any prior bad reactions to treatments and know any allergies (behavioral issues on chronic or high rescue type medication is different than allergic rash or airway closure)

SRP: 10 Seizure Rescue Plan Steps

- Step 7: Know any contraindications: Examples if Dravet Syndrome do not typically do well with IV Phenytoin(Dilantin) or Lacosamide (Vimpat) IV or if on chronic benzodiazepine class of medications like clobazam or clonazepam then they may need higher doses of this type of rescue medication. Also if on Ketogenic Diet avoid IV glucose solutions if possible

- Step 8: If on Hard to Get Medication or experimental medications then have travel bag to take with on ambulance or to Emergency room in case prolonged stay or admission have a written medication sheet online or on phone or printed version as part of rescue kit

SRP: 10 Seizure Rescue Plan Steps

- Step 9: **STAY CALM** and be courteous and make yourself part of rescue team by providing good information, being knowledgeable about your child and offering clear communication with others helping your child EMT or Triage nurses school nurses and physicians as may be the case

- Step 10: Try to be proactive in rescue as it is easier to treat earlier than later in most cases

Four Questions for your Neurologist

Ask if they do not discuss first:

1. How comfortable are you working with my child(family member) condition?

2. Can you help me understand or tell me what the type(s) of seizures we are dealing with?

3. Define your style and plan for treatment, how do you communicate with families, and who and how do we call for help in am emergency? What constitutes an emergency in your medical opinion?

4. How will the seizures affect us: Safety; Activities; Developmental Concerns; Behavioral Concerns; and chance of SUDEP among ?

Conclusions

- Seizure Rescue Plan (SRP, also sometimes called Seizure Action Plan) is part of overall seizure management plan

- Establish and Find a physician care team that you can discuss these issues with comfortably. If physician says things and you think of questions later write these down to ask next time so you don’t forget. There are no stupid questions

- Knowledge is Power improves outcomes if you know your loved one’s seizure types, baseline medications, most recent EEG or MRI testing dates, last time they had drug levels if available, and any contraindications for your individual child. An informed SRP makes emergencies less stressful also

Conclusions

- Communication in clear calm manner when in an emergency situation

- Plan Ahead and have paperwork of your SRP available and remember Treating Earlier usually better than treating later

- If something worked before mention it early to EMT or physicians assisting you if need to involve emergency services before wasting time on things that may not have worked before; also know contraindications