

Rescue Medications for Children with Severe Developmental and Epileptic Encephalopathies (DEEs)

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

Overview

- What differences are there in rescue medications use in the DEEs?
- How do you choose the right rescue med?
- How often should/can rescue meds be used?
- Tips to optimize your Seizure Action Plan

General Recommendations for Rescue Medication Use

- Convulsive seizure > 5 minutes
- Seizure cluster – definition of a cluster is not clear, but for many, consider ≥ 3 convulsive seizures in an hour, each lasting >1 minute
- These situations represent a **clear change** in the persons usual seizure pattern, and typically do not occur very frequently!

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?

Convulsive Seizures

- In some cases, we know that children who start having a convulsive seizure typically progress to status epilepticus – i.e. young child with Dravet syndrome
 - Give RM at ONSET of convulsive seizure instead of waiting 3-5 minutes
- Clusters of convulsive seizures
 - Individualize!
 - i.e. if Danny has 3 briefer convulsive seizures in an hour, there is a high likelihood of a long convulsive seizure in the next few hours

Non-convulsive Seizures

- No clear consensus on what to do amongst clinicians
- Must individualize: Families appreciate patterns
 - Many children with DEEs have frequent nonconvulsive seizures - do not give if this is the regular pattern – i.e. David has recurrent brief myoclonic jerks with absences 6-10 x per hour
 - But consider if this is indicative of a concerning pattern: i.e. Beth has recurrent periods of nonconvulsive status epilepticus, lasting up to 8-10 hours 1-2 x per month. These always start with back to back myoclonic jerks

How to find the best RM option

- Diazepam rectal gel (Diastat)
- Diazepam or lorazepam intensol
- Clonazepam ODT
- Midazolam nasal (nayzilam or other)
- Diazepam nasal (Valtoco)

Diastat rectal gel

- FDA indicated for children age 2 years and older
- 0.2-0.5 mg/kg depending on age
- Doses of 2.5-20 mg (2.5 mg increments)



Diazepam or Lorazepam intensol

- Not FDA indicated for seizure rescue but often used
- Diazepam 0.1-0.2 mg/kg (max 5 mg)
- Lorazepam 0.05 mg/kg (max 2 mg) – needs to be refrigerated
- Give buccally



Clonazepam ODT

- Rapidly dissolving tablet which can be placed buccally
- Dose ranges from 0.125-2 mg
- Not FDA approved as rescue – recommended for prophylactic treatment of seizures

Valtoco (nasal diazepam)

- FDA approved for seizure rescue in children >6 yrs of age



Nayzilam (nasal midazolam)

FDA approved for
persons >12 years as
rescue for seizures



Considerations

- Effectiveness
- Side effects
- Ease of administration and ability to administer
- Social embarrassment (Diastat)
- Insurance coverage and cost (Nayzilam ≥ 12 yrs, Valtoco ≥ 6 yrs)

Who Can Give Rescue Meds?

- Diastat – package insert states that caregiver administering this should review steps with the prescribing MD
- Clonazepam ODT, Valtoco and Nayzilam are easy to give
- More concern if you need to draw up a certain amount – greater room for error
 - Diazepam and lorazepam intensol, nasal midazolam using IV vial
 - Non-medical professionals may be hesitant to administer these products

Videos on how to give rescue meds exist online



How Often Can RMs be Used?

- FDA guidance often not consistent with what is done in the real world
 - Diastat – no more than 5x/month and no more than once every 5 days
 - Valtoco – no more than 2 doses – 4 hours apart. No more than 5x/month and no more than once every 5 days
 - Nayzilam – no more than 2 doses – 10 minutes apart. No more than 5x/month and no more than once every 3 days

- Real world is very different!

Risk of Overuse

- Giving too high of a dose:
 - Respiratory depression
 - Excessive sedation

- Giving too often:
 - Decreased likelihood of efficacy of rescue
 - Develop tolerance and risk of increased seizures if you try to reduce the frequency

What if you are needing it more often?

- Discuss seizure control with your neurology provider
- Can prophylactic therapies be optimized?
- Is your child a candidate for other therapies? dietary, palliative surgical options

What if your rescue medication does not seem to be effective?

- Is it the right dose – most children with DEE have some tolerance to meds, so often need the higher end of the dose range to be effective
- If one rescue med has not worked for several successive treatments, consider switching to a different agent
 - But little data to know how often that is successful

How to Optimize your Seizure Action Plan

- Pick the best rescue option for you:
 - If other caregivers involved, try to find one that they are comfortable giving quickly
- Clear guidance:
 - Pre-rescue – can you do anything before you get to needing a rescue
 - What should and should not be rescued
 - Can you repeat the dose – if so when, and how much
 - When to seek emergency care/call 911 if rescue meds do not work

How to Optimize your Seizure Action Plan

- When in the ER, what should be done
 - Most peds hospitals have protocols for status epilepticus, but these are generic
 - Children with DEEs should have an individualized plan – worked out with parent(s) and HCP – what has worked or not worked in past
 - Written out on letterhead
 - Option to include in chart as Care Plan
 - If smaller local hospital, can you house in the ER
 - Carry in your purse/wallet

Learn More & Develop/Update Your SAP

Additional Rescue Series Webinars

all dates to be confirmed

July 1st – A Clinicians Perspective & Advice

July 15th – The Lived Rescue Experiences of DEE Families

August 1st - Hands on, Peer-led Workshop to Develop/
Update Seizure Action Plans

Questions?

