

Developmental and Epileptic Encephalopathy (DEE) - *What is it and Why is it so Hard to Treat?*

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Disclosures: None

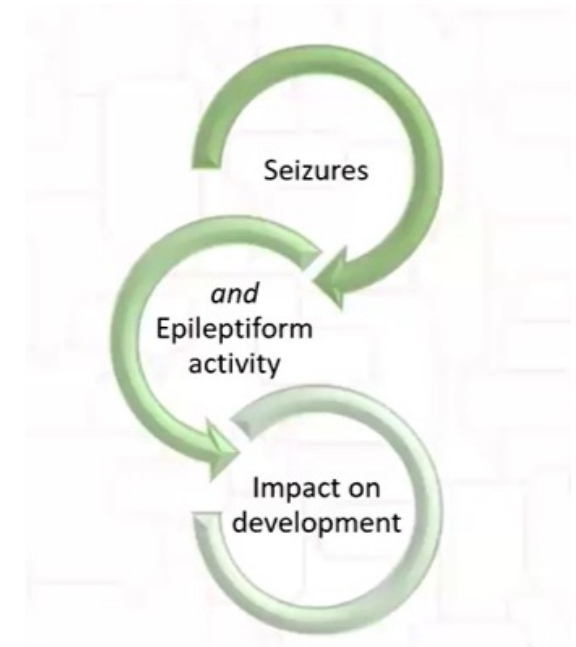
Learning Objectives

-  **What is a DEE?**
-  **Why do DEEs happen?**
-  **Why are seizures so hard to control?**
-  **Why is treating seizures simply not enough?**
-  **What is in store for a hopeful future?**

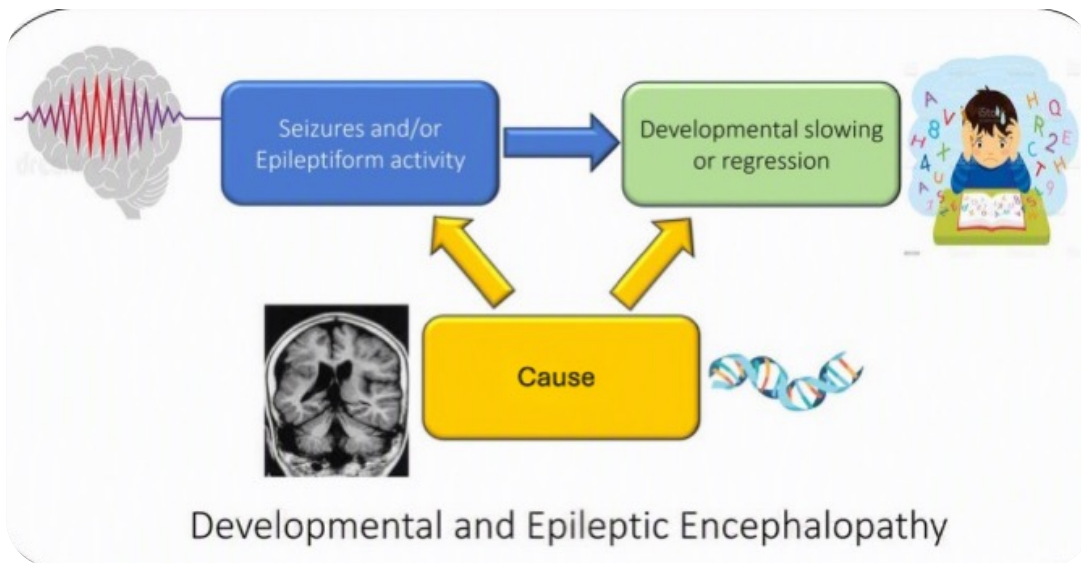
Defining the Concepts

Epileptic Encephalopathy

- Normal brain development before epilepsy
- Epilepsy slows brain development
- Early seizure treatment salvages development



What is a DEE?

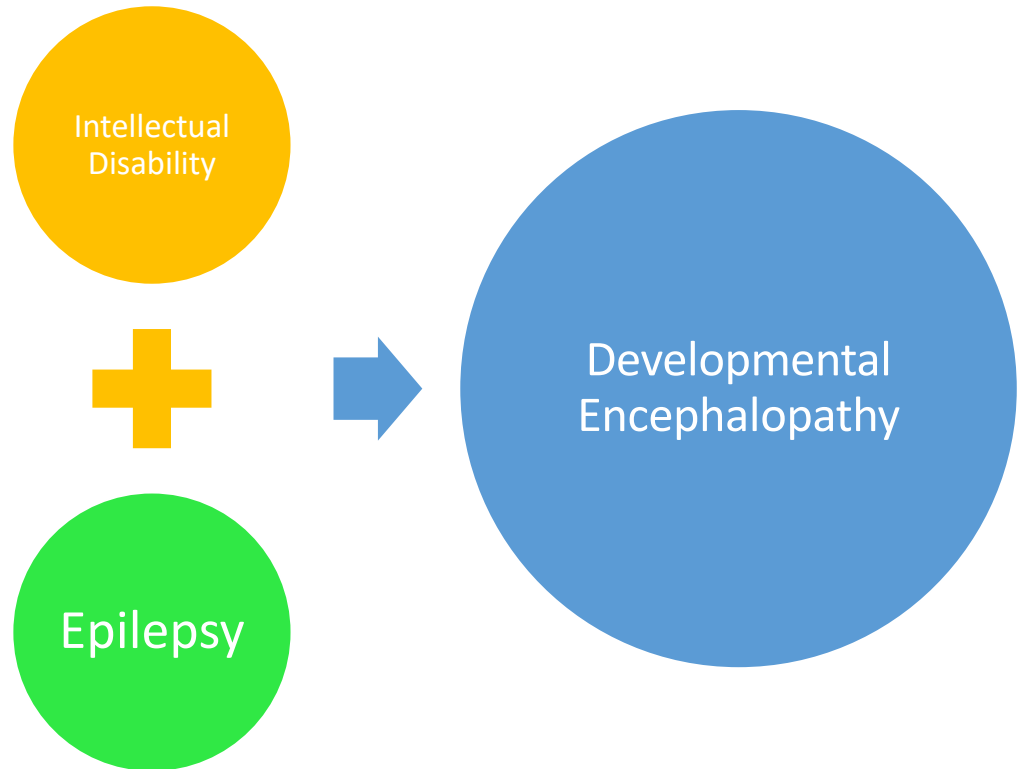


1. Frequent seizures and/or abnormal EEG activity (“spikes” or “sharps”)
2. Slowed development or loss skills
3. The underlying cause also contributes to developmental challenges

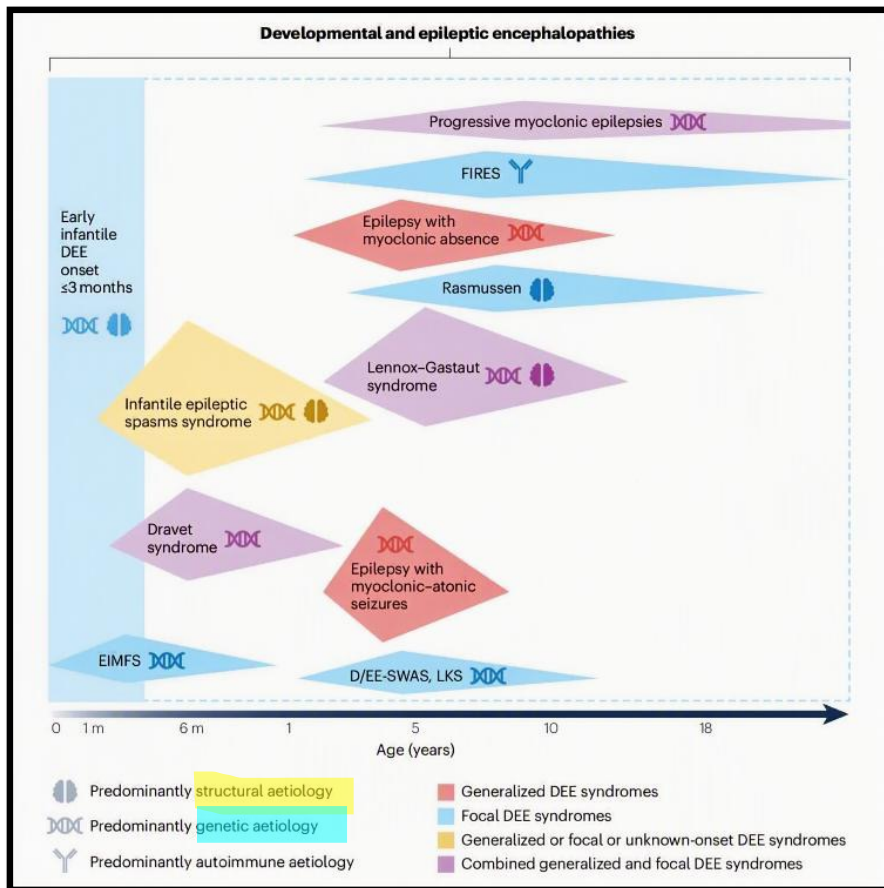
Defining the Concepts

Developmental Encephalopathy

- Brain developed “differently” from early life
- Developmental problems are not caused by seizures
- Seizure control alone does not improve development



Why do DEEs happen?



DEEs result from

- ❖ Genetic changes
 - > 900 genes and counting!
- ❖ Structural brain differences/malformations
- ❖ Metabolic disorders
- ❖ Unknown cause

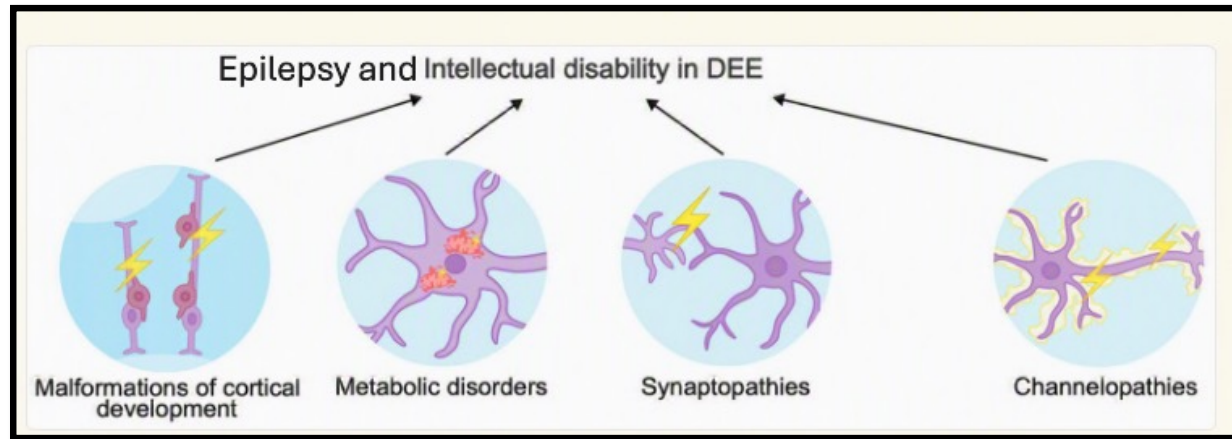
Why are seizures so hard to control in DEEs?

The underlying problem remains!

- Most ASMs:
 - Simply reduce seizures
 - Do not correct the underlying cause



Why are seizures so hard to control in DEEs?



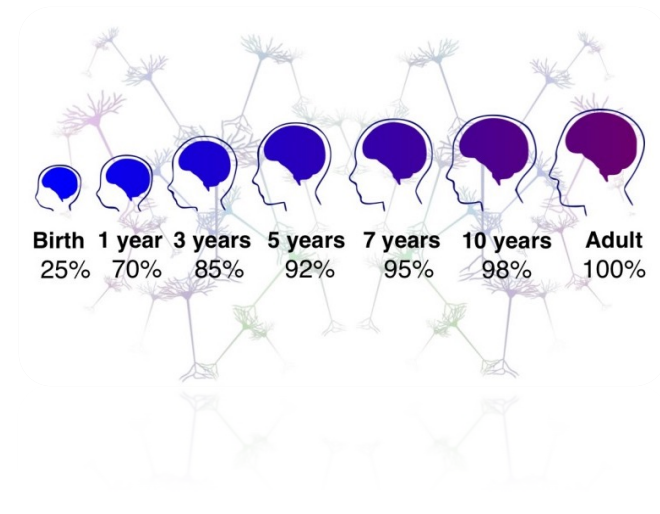
Many different biological mechanisms

- Brain development pathways
- Synapses
- Neurotransmitters
- Sodium or Potassium channels

Why are seizures so hard to control in DEEs?

Children's brains are constantly growing!

- Seizure types change
- Medication responses change

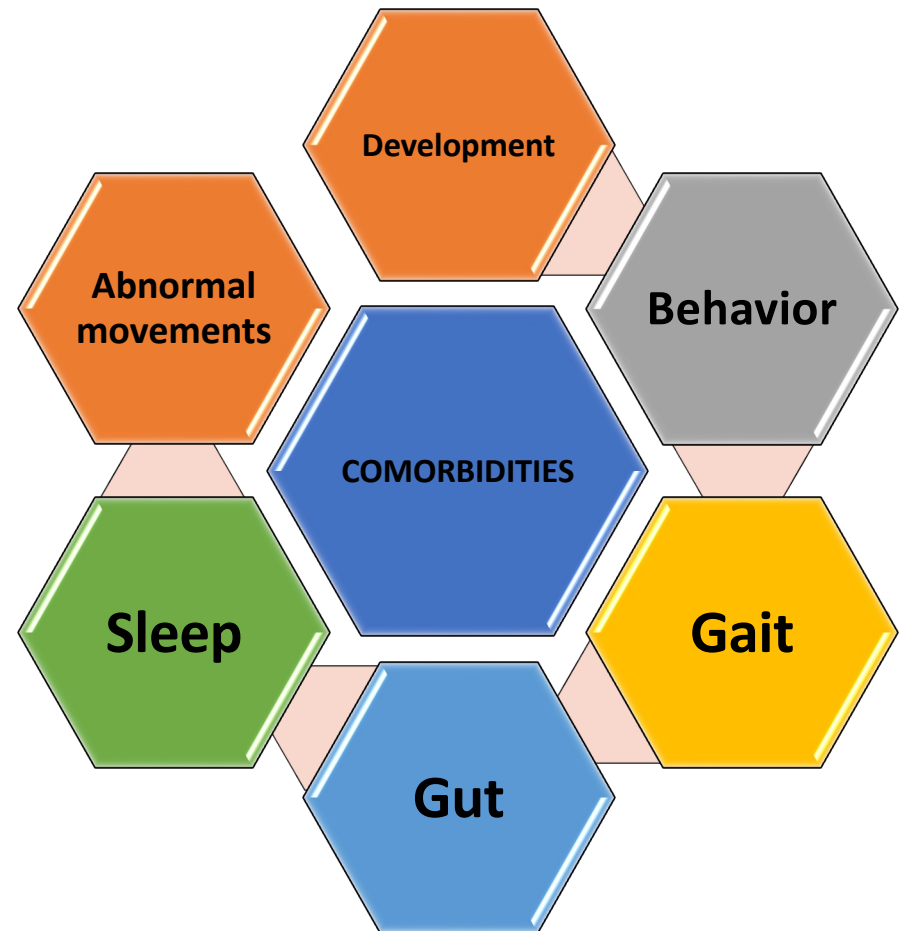
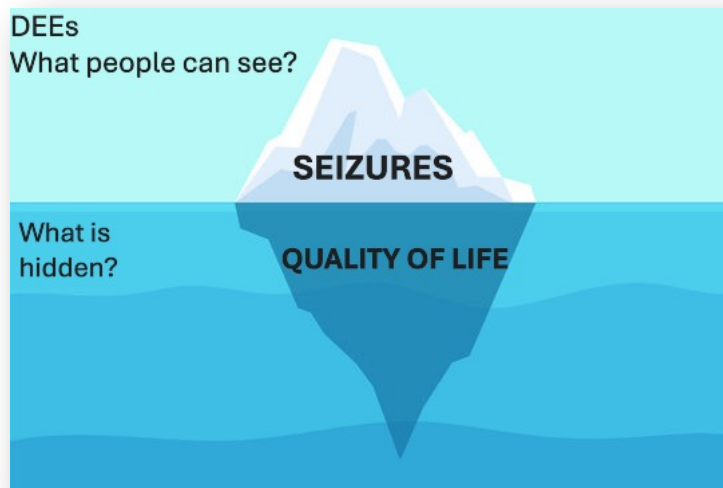


The Road to Seizure Control in DEEs

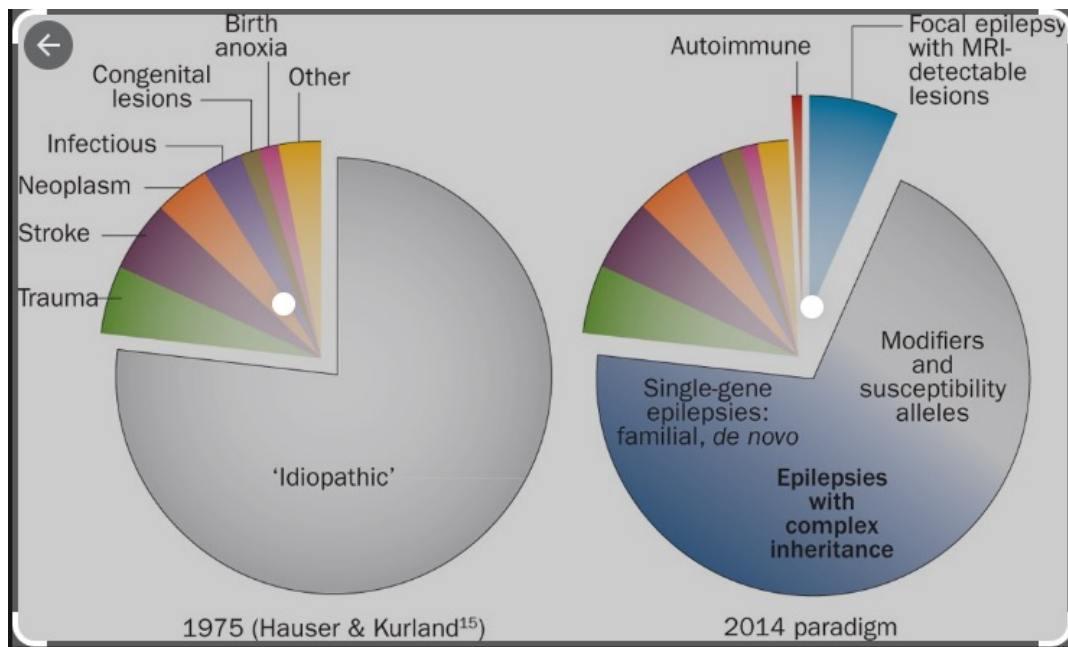
- “ The Power of the Right Diagnosis
- “ One at a Time: Less Chaos, More Clarity
- “ Start Low, Go Slow
- “ Less Is More
- “ Finding the Sweet Spot (Does not happen Overnight!)
- “ No Permanent Enemies



Why is treating seizures simply not enough?



What Gives Me Hope?



From Band-Aids to Breakthroughs

Targeted drugs:

Fenfluramine, Ganoxolone, Everolimus

Gene therapies & Antisense oligonucleotides:

Dravet syndrome

Together, We Create Hope

- ✦ Hope is NOT for every seizure to disappear BUT
- ✦ Hope is:
- ✦ Finding the right diagnosis
- ✦ Finding newer therapies
- ✦ Celebrating every gain, big or small
- ✦ Seeing a child more alert, comfortable & engaged



And So Is Their DEE