

# What is a DEE?

**DEE =**

**Developmental and**

**Epileptic**

**Encephalopathy** <sup>1-4</sup>

## In short...

- **DEE** stands for **D**evelopmental and **E**pileptic **E**ncephalopathy
- This is a term used to describe conditions in which developmental delay or impairment is caused by an underlying condition (such as a change in the SCN2A gene or damage to the brain) **and** by frequent epileptic seizures
- DEEs usually begin early in life, often soon after birth

**D**

### Developmental

Developmental delays related to the underlying cause to the epilepsy

**E**

### Epileptic

Epilepsy that contributes to developmental delays

**E**

### Encephalopathy

A condition that affects the structure or function of the brain.

## DEE

A condition in which developmental impairment is related to both the underlying cause of the condition and the effects of the epilepsy on the brain

## DEE = Developmental and Epileptic Encephalopathy <sup>1-4</sup>

- DEE is a term used to describe conditions in which developmental delay or impairment is caused by the underlying condition **and** the effects of frequent seizures.
- DEEs usually begin early in life, often starting soon after birth.
- Individuals with DEE typically have frequent seizures as well as developmental delay / intellectual disability, which continue throughout their lives.

## Causes of DEE <sup>1,4-6</sup>

- DEEs often develop due to a change in a single gene, eg *SCN2A*. Other genes that cause DEEs include *SCN1A*, *SCN8A*, *CDKL5* and *KCNQ2*.
- Other causes include abnormal brain development, head injuries, and metabolic factors.

## Seizures <sup>3</sup>

- Seizures may be frequent and severe.
- Many different types of seizures may be seen, such as epileptic spasms, and tonic, atonic or myoclonic seizures.

## Epilepsy syndromes <sup>6</sup>

- Some individuals with DEE are considered to have a specific epilepsy syndrome, such as:
  - o Dravet syndrome
  - o Infantile spasms or West syndrome
  - o Lennox-Gastaut syndrome
  - o Ohtahara syndrome
  - o Early myoclonic encephalopathy
  - o Doose syndrome.

## Other challenges <sup>7</sup>

- In addition to seizures and developmental delay / intellectual disability, individuals with DEE may also have:
  - o Movement disorders e.g. difficulty walking, tremors, unusual walking styles (gait abnormalities)
  - o Lower or higher muscle tone than usual
  - o Behavioural problems
  - o Autism spectrum disorder
  - o Feeding difficulties.

## More information

- Epilepsy Foundation:
- Developmental and epileptic encephalopathy
- Causes of DEE

## References

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7. Zuberi SM, et al. ILAE classification and definition of epilepsy syndromes with onset in neonates and infants: Position statement by the ILAE Task Force on Nosology and Definitions. *Epilepsia* 2022;63:1349-1397. [PubMed]