

What is a DEE?

Developmental and epileptic encephalopathy, in plain language

“**DEE**” stands for **developmental and epileptic encephalopathy**. It describes a group of rare, serious epilepsies that usually start in babies and young children. **Epilepsy** means a tendency to have **seizures** (sudden bursts of electrical activity in the brain). **Encephalopathy** means the brain is not working as it should. In a DEE, two things affect a child's development: the underlying cause itself, and the frequent seizure activity.¹ Many children also have differences in learning, movement, communication, sleep and feeding.

THE KEY POINTS

- DEEs are rare and usually start in the first months or years of life.¹
- Seizures are often hard to control, so several medicines may be tried.
- Development can be delayed, and some children lose skills they had, called **regression**.
- Most DEEs have a genetic cause; others come from differences in brain structure, metabolic conditions or injury.¹
- A faster, accurate diagnosis helps target the right treatment sooner.

How doctors describe a DEE

Doctors name a DEE in two ways at once: the **syndrome** (the pattern of seizures, the age they start, and findings on an **EEG**, a painless test of the brain's electrical activity) and the **cause** (for example a specific gene). Under the current international classification (ILAE 2022), some older names have changed:^{1,2}

- **Early-infantile DEE (EIDEE)** starts in the first three months. It now includes what used to be called Ohtahara syndrome and early myoclonic encephalopathy.¹
- **Infantile epileptic spasms syndrome (IESS)** means clusters of brief stiffening movements (“spasms”), often with developmental slowing. It includes what was called West syndrome.¹
- **Dravet syndrome** (often caused by changes in the *SCN1A* gene) and **Lennox-Gastaut syndrome** (many seizure types, including sudden “drop” seizures, usually hard to control) are other examples.²

Changes in the **SCN2A** gene are one genetic cause of DEE. The gene a child has can shape both their seizures and which treatments are tried.³

Treatment

Anti-seizure medicines (ASMs) are the first step, sometimes in combination. Seizure freedom is not always possible, so goals are set for each child. Other options can include the **ketogenic diet** (a strict medical diet), **vagus nerve stimulation** (a small device that helps reduce seizures), surgery in selected cases, and increasingly **gene-targeted (precision) treatments**.

Ask your neurologist about **SUDEP** (sudden unexpected death in epilepsy) and how to lower the risk. It is a hard topic, and an important one.

YOU ARE NOT ALONE

Caring for a child with a DEE is a lot. Support is available.

- **SCN2A Australia** — connect with other families
- **NDIS** — 1800 800 110
- **Lifeline** — 13 11 14 (any time)
- **Suicide Call Back Service** — 1300 659 467

General information, not medical advice. Always discuss your child's diagnosis, testing and treatment with your treating team. Genetics knowledge changes quickly; this reflects current understanding at the date shown.

SOURCES

1. Zuberi SM, et al. ILAE classification and definition of epilepsy syndromes with onset in neonates and infants. *Epilepsia*. 2022;63(6):1349–1397.
2. Specchio N, et al. ILAE classification and definition of epilepsy syndromes with onset in childhood. *Epilepsia*. 2022;63(6):1398–1442.
3. Wolff M, et al. Phenotypic spectrum and genetics of SCN2A-related disorders. *Epilepsia*. 2019;60(S3):S59–S67.

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