The impact of **developmental** and **epileptic encephalopathies** (DEEs)

Our ambition

UCB is at the forefront of epilepsy research, developing a portfolio of life-changing solutions for people living with a variety of seizure types and rare epileptic syndromes.

Our core focus is on finding differentiated solutions that can positively impact the lives of individuals, carers and families living with epilepsies.

Background on developmental and epileptic encephalopathies (DEEs)

An epilepsy associated with developmental impairment that may be due to either the underlying aetiology or the superimposed epileptic activity, or both.[1]

Exact worldwide prevalence of DEEs is not known, but regional data have shown cumulative incidence of 169/100,000 children under 16 years of age.[2]

Up to 50% of patients with a DEE remain undiagnosed.[3]

The impact of DEEs

- The impact of epilepsies, including DEEs, reaches far beyond seizures and clinical features.[4]
- There is a wide range of non-seizure clinical manifestations of DEEs, such as [5,6,7,8,9,10]

Risk of mortality (including SUDEP - Sudden Unexpected Death in Epilepsy)

Intellectual disability

Motor & Movement disorders

Sleep Issues

Psychiatric problems

Behavioral problems

- DEEs affect not only the individual but the whole family. Siblings of children with DEEs grow up in an environment permeated by **stress**, **anxiety** and **fear**. Families are encouraged to speak to a healthcare professional about ways to address the challenges siblings face when having a brother or sister with a DEE based on sibling voices survey.[10]

79% of young siblings aged 9-12 expressed fear that their sibling might die[10]

35% of adult siblings report a history of treatment for clinical depression[10]

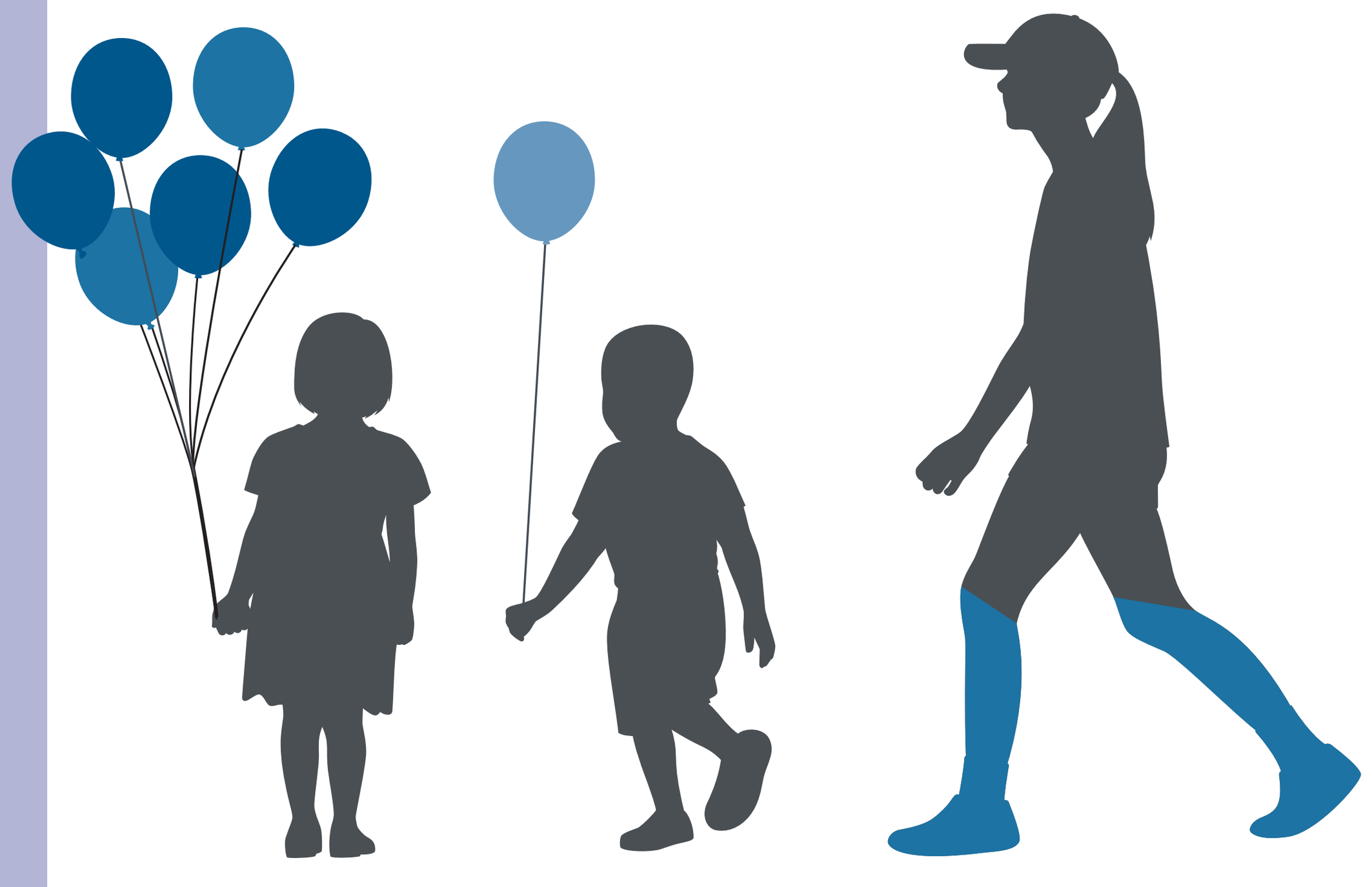
Dravet Syndrome

Incidence and prevalence

- Dravet syndrome is estimated to affect one in **20,000-40,000** people per live births in Europe, with a similar incidence in the US (about **1 in 15,700** live births)[11,12]

Impact of Dravet Syndrome

- Less than **10%** of people with Dravet syndrome are able to achieve freedom from their persistent seizures.[14]



- People with Dravet syndrome experience SUDEP at rates **6 x higher** than persons with other forms of epilepsy[5]
- There is a **15-20%** mortality rate by adulthood [5,12,13]

66% of caregivers experience depression due to the devastating nature of Dravet syndrome.[15]

74% report concerns about the emotional impact on siblings of children with Dravet syndrome based on a survey conducted by Dravet Syndrome Foundation.[15]

Treatment

- Newer treatment options are available; Healthcare professionals can provide patients and carers with information about the risks associated with their condition to help them make informed decisions about their treatment options and care.[7,20]

Lennox-Gastaut Syndrome

Incidence and prevalence

- Lennox-Gastaut syndrome affects an estimated **2 in 10,000** people in European Union.[16]
- There are **48,000** Lennox-Gastaut syndrome patients in the United States, and around **1 million** worldwide.[18]
- LGS usually develops between 3-8 years of age. LGS may evolve from West syndrome or infantile spasms in about **20%** of patients.[19,20]

This condition represents 1%-10% of childhood epilepsies[19]

1-2% of all epilepsies [19]

Seizures tend to persist on a daily or weekly basis in more than two-thirds of the patients.[20b]

	SUN	MON	TUE	WED	THU	FRI	SAT
1	1	2	3	4	5	6	7
2	8	9	10	11	12	13	14
3	15	16	17	18	19	20	21
4	22	23	24	25	26	27	28
5	29	30	31				

Impact of Lennox-Gastaut Syndrome

- LGS is characterized by tonic seizures with at least one other seizure type, such as atonic, generalized tonic-clonic, atypical absence, and myoclonic. These may result in frequent falls and injuries.[17,19,20]

Treatment

- Newer treatment options are available; an individualised approach is particularly important in LGS due to the complexity and range of underlying causes.[7,22]

1 Wirrell E, et al. Epilepsia. 2022;63:1333–48.
2 Poke G, Stanley J, Scheffer I, et al. Neurology. 2023;100:e1363-75.
3 Aledo-Serrano A, Sánchez-Alcudia R, Toledano R, et al. J Transl Genet Genom. 2021;5:443-55.
4 Nabbout R, et al. Dev Med Child Neurol. 2019;61:1229–36.
5 Cross JH, Galer BS, Gil-Nagel A, et al. Seizure. 2021;93:154-59.
6 Sullivan J, et al. Epilepsy Behav. 2022;130:108661.
7 Strzelczyk A and Schubert-Bast S. CNS Drugs. 2022;35:217-37.
8 Palmer E, Howell K and Scheffer I. Neurotherapeutics. 2021;18:1432-44.
9 Scheffer E, et al. Epilepsia. 2017;58(4):512-21.
10 Bailey LD, et al. Epilepsy Behav. 2020;112:107377.
11 Wu YW, Sullivan J, McDaniel SS, et al. Pediatrics. 2015;136(5):e1310-15.
12 Aras LM, Isla J, Mingorance-Le Meur A. Epilepsy Behav. 2015;44:104-9.
13 National Organization for Rare Disorders. Dravet Syndrome. https://rarediseases.org/rare-diseases/dravet-syndrome-spectrum/#disease-overview-main Last accessed May 2023.
14 Lagae L, Brambilla I, Mingorance A, et al. Dev Med Child Neurol. 2018;60(1):63-72.
15 Villas N, Meskis MA, Goodliffe S. Epilepsy Behav. 2017;74:81-86.
16 European medicine agency EU/3/17/1855. https://www.ema.europa.eu/en/medicines/human/orphan-designations/eu3171855. Last accessed May 2023.
17 Specchio N, et al. Epilepsia. 2022;63:1398-42.
18 LGS Foundation. How Many People Have LGS? https://www.lgsfoundation.org/about-lgs-2/how-many-people-have-lgs/ Last accessed May 2023.
19 Asadi-Pooya AA. Neurol Sci. 2018;39:403–14.
20 Gallop K, Wild D, Nixon A, et al. Seizure. 2009;18:554-58.
21 Skluzacek JV, et al. Epilepsia. 2011;52(Suppl.2):95-101.
22 Strzelczyk A and Schubert-Bast S. CNS Drugs. 2022;36:1079–111.
23 Oguni H, et al. Epilepsia. 1996;37:44-47.

a. Epidemiological study of children in Wellington region of New Zealand.
b. Based on a study in 72 patients followed up for >10years.[23]

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Intended for Healthcare professional

Inspired by **patients.**
Driven by **science.**