EU version

LIVING WITH DRAVET SYNDROME A DEVASTATING TREATMENT-RESISTANT FORM OF EPILEPSY

What is Dravet syndrome?

Dravet syndrome is a rare, devastating and life-long form of epilepsy that begins in infancy, and is marked by severe, prolonged, and often treatment-resistant seizures; frequent hospitalisations; and significant cognitive, motor, and behavioural impairments.[1,2] Patients with Dravet syndrome have an increased risk of death (about 20% higher than other epilepsies), primarily from SUDEP (sudden unexpected death in epilepsy) or a medical emergency called status epilepticus.[1,3]

Seizures associated with Dravet syndrome[4,5]

- Begin within the first year of life
- Present in a developmentally normal infant
- Can be repetitive
- Can sometimes be prolonged
  lasting more than 5 minutes for tonic-clonic seizures (status epilepticus)

Can be triggered by common daily occurrences[4,6]

- Emotional stress or excitement
- Sunlight, or flashing lights or patterns
- Changes in body temperature

Who does Dravet syndrome impact?

Dravet syndrome affects approximately one in 20,000 to 40,000 people in Europe and can be devastating in nature, as patients often can not care for themselves.[1,4] Due to constant care and worry, the quality of life for caregivers and families is severely impacted. This commitment can result in physical, psychosocial, emotional, and financial burdens, such as continuous monitoring, the need to co-sleep, and fear of their Dravet syndrome family member's death.[1,2,7,8]

66%
of caregivers experience depression due to the devastating nature of Dravet syndrome[8]

2
of the most straining aspects of caregiving are lack of sleep and inability to stop worrying[9]

74%
of caregivers report concerns about the emotional impact on siblings of children with Dravet syndrome[8]

Dravet syndrome by the numbers

More than
90%
of children with Dravet syndrome live with persistent seizures[10]

~85%
of children diagnosed with Dravet syndrome carry a SCN1A gene mutation[6]

Dravet syndrome has a mortality rate of up to
15-20%
by adulthood[4]

>50%
of all deaths in children and young adults with Dravet syndrome are attributed to sudden unexpected death in epilepsy[11]

How is Dravet syndrome treated?

Dravet syndrome is highly resistant to treatment and less than 10% of patients experience seizure freedom with current treatments.[10,12,13] Most patients with Dravet syndrome are taking combinations of 3+ antiepileptic drugs (AEDs). Despite treatment with multiple AEDs, many patients continue to experience multiple seizures on a weekly or even daily basis.[2] Existing treatment options may not be able to provide the amount of seizure control that patients desire.[1]

Despite taking 3 or more AEDs, 45% of patients continue to experience 4 or more seizures per month[1]

Less than 10% of children with Dravet syndrome are able to achieve freedom from their persistent seizures, despite treatment with AEDs based on a recent study[10]

1 Aras LM, Isla J, Mingorance-Le Meur A. The European patient with Dravet syndrome: results from a parent-reported survey on antiepileptic drug use in the European population with Dravet syndrome. Epilepsy Behav. 2015;44:104-9.
2 Lagae L, Brambilla I, Mingorance A, et al. Quality of life and comorbidities associated with Dravet syndrome severity: a multinational cohort survey. Dev Med Child Neurol. 2018;60:63-72.
3 Gataullina S, Dulac O. From genotype to phenotype in Dravet disease. Seizure. 2017;44:58-64.
4 Sullivan J, Knupp K, Wirrel E. Rare disease database: Dravet syndrome. National Organization of Rare Diseases. Available at: https://rarediseases.org/rare-diseases/dravet-syndrome-spectrum/. Last updated: 2018. Last accessed: June 2020.
5 Dravet C. The core Dravet syndrome phenotype. Epilepsia. 2011;52:3-9.
6 Wirrell EC, Laux L, Donner E, et al. Optimizing the diagnosis and management of Dravet syndrome: Recommendations from a North American consensus panel. Pediatr Neurol. 2017;68:18-34.
7 Lagae L, Sullivan J, Knupp K, et al. Fenfluramine hydrochloride for the treatment of seizures in Dravet syndrome: a randomised, double-blind, placebo-controlled trial. Lancet. 2020;394:2243-2254.
8 Villas N, Meskis MA, Goodliffe S. Dravet syndrome: characteristics, comorbidities, and caregiver concerns. Epilepsy Behav. 2017;74:81-86.
9 Jensen MP, Liljenquist KS, Bocell F, et al. Life impact of caregiving for severe childhood epilepsy: results of expert panels and caregiver focus groups. Epilepsy Behav. 2017;74:135-143.
10 Lagae L, Brambilla I, Mingorance A, et al. DISCUSS Dravet – key socioeconomic findings from a large multinational survey of Dravet syndrome caregivers. Presented at the 12th European Paediatric Neurology Society Congress. 2017 June 20-24; Lyon, France
11 Cooper MS, Mcintosh A, Crompton DE, et al. Mortality in Dravet syndrome. Epilepsy Res. 2016;128:43-47.
12 Nabbout R, Mistry A, Zuberi S, et al. Fenfluramine for treatment-resistant seizures in patients with Dravet syndrome receiving stiripentol-inclusive regimens. JAMA Neurol. 2020;77:300-308.
13 Dravet C. Dravet syndrome history. Dev Med Child Neurol. 2011;53(suppl 2):1-6.

ZOGENIX

To learn more, please visit www.zogenix.com