

DRAVET SYNDROME

Dravet syndrome is a rare, severe, lifelong form of epilepsy that typically begins in the first year of life with frequent and/or prolonged seizures. Previously known as severe myoclonic epilepsy in infancy (SMEI), it affects between 1 in 20,000 to 1 in 40,000 people (over 5,400 people under the age of 20 in the U.S.).^{1,2} About 80 percent of people with this syndrome have a gene mutation that causes problems in the way the brain works.³

Children with Dravet syndrome can develop many different seizure types and approximately 15 percent die within 10 years of diagnosis due to issues such as SUDEP (sudden unexpected death in epilepsy), prolonged seizures, or seizure-related accidents such as drowning and infections.⁴

RELATED CONDITIONS

Most children with Dravet syndrome develop some level of developmental disability and have other conditions that are associated with the syndrome, including movement and balance issues, orthopedic conditions, delayed language and speech issues, growth and nutrition issues, sleeping difficulties, sensory integration disorders and disruptions of the autonomic nervous system.³

It is recommended that children, with the support of their families or caregivers, avoid seizure triggers, which may include immediate changes to surroundings or body

temperature, illness, stress, or flashing lights.³ Some children may experience a prolonged seizure event, known as status epilepticus, that often requires medical intervention to bring the seizure to an end.³ These long seizures are very dangerous and increase the chance of death.⁵

The constant supervision required for many of these patients can be emotionally draining, highly stressful and financially overwhelming for families.⁶

TREATMENT OPTIONS

Unfortunately, there is no cure for Dravet syndrome. Families often work with a multidisciplinary team of neurologists, dietitians and pediatricians to help manage the disease.⁷

Treatment is focused on obtaining the best seizure control with the fewest side effects. Most patients will require two or more seizure medications.⁸ However, all seizure types in Dravet syndrome are resistant to anti-epileptic drugs (AEDs).¹ The U.S. Food and Drug Administration recently approved the first prescription medicine for the treatment of Dravet syndrome, the first in a new class of AEDs and the first plant-derived cannabinoid product. A ketogenic (low-carbohydrate) diet and vagus nerve stimulation therapy may also be helpful for patients with Dravet syndrome.⁸

Greenwich Biosciences, Inc. is the U.S. subsidiary of GW Pharmaceuticals plc.

¹ Dravet C. The core Dravet syndrome phenotype. *Epilepsia*. 2011;52(Suppl. 2):3-9.

² Dravet C, Bureau M, Oguni H, Cokar O, Guerrini R. Dravet syndrome (severe myoclonic epilepsy in infancy). In: Bureau M, Genton P, Dravet C, et al., eds. *Epileptic Syndromes in Infancy, Childhood and Adolescence*. Montrouge, France: John Libbey Eurotext Ltd.; 2012:112-156.

³ Dravet Syndrome Foundation. What is Dravet Syndrome? Available at <https://www.dravetfoundation.org/what-is-dravet-syndrome/>. Accessed May 15, 2018.

⁴ Cooper MS, McIntosh A, Crompton DE, et al. Mortality in Dravet syndrome. *Epilepsy Res*. 2016;128:43-47.

⁵ Epilepsy Foundation. What is Status Epilepticus? Available at <http://www.epilepsy.com/learn/impact/seizure-emergencies/status-epilepticus>. Accessed May 15, 2018.

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 e13.

⁷ Wirrell EC. Treatment of Dravet syndrome. *Can J Neurol Sci*. 2016;43 Suppl 3:S13-18.

⁸ Epilepsy Foundation. Dravet syndrome. Available at <https://www.epilepsy.com/learn/types-epilepsy-syndromes/dravet-syndrome>. Accessed May 15, 2018.