

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 I in 20,000 to I in 40,000 people (over 5,400 people under the age of 20 in the U.S.). About 80 percent of people with this syndrome have a gene mutation that causes problems in the way the brain works.²

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, seizure-related accidents such as drowning and infections.^{3,4}

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.⁵

TREATMENT OPTIONS

Unfortunately, there is no cure for Dravet syndrome and there is currently no therapy approved by the U.S. Food and Drug Administration (FDA) for treatment. Because of this, families often work with a multidisciplinary team of neurologists, dieticians and pediatricians to help manage the disease.

Treatment is focused on obtaining the best seizure control with the fewest side effects, although available medications are not always able to fully control seizures in patients with this condition. Most patients will require two or more seizure medications.⁶ More than 90 percent of people with the condition are considered treatment-resistant, having failed two or more anti-epileptic drugs (AEDs).⁷ A ketogenic (low-carbohydrate) diet and vagus nerve stimulation therapy may also be helpful for patients with Dravet syndrome.⁸

A pharmaceutical formulation of cannabidiol (CBD), the first in a new class of AEDs, is currently in Phase 3 clinical trials for the treatment of Dravet syndrome and, if approved by the FDA, would be the first therapy approved for the disease.

GREENWICH Biosciences, Inc. is the U.S. operating unit of GW Pharmaceuticals.

² http://www.epilepsy.com/learn/types-epilepsy-syndromes/dravet-syndrome

¹ Forsgren, L. Incidence and prevalence. in:Wallace SJ, Farrell K (Eds.) Epilepsy in children. 2nd edn. Arnold, London; 2004: 21–25

³ https://www.dravetfoundation.org/what-is-dravet-syndrome/

⁴ Skluzacek J, Watts KP, Parsy O, Wical B, Camfield P. Dravet syndrome and parent associations: The IDEA League experience with comorbid conditions, mortality, management, adaptation, and grief. Epilepsia. 2011;52(Suppl. 2):95–101

⁵ https://www.dravetfoundation.org/what-is-dravet-syndrome/

⁶ http://www.epilepsy.com/learn/types-epilepsy-syndromes/dravet-syndrome

⁷ Dravet C.The core Dravet syndrome phenotype. Epilepsia. 2011;52(Suppl. 2):3-9

⁸ http://www.epilepsy.com/learn/types-epilepsy-syndromes/dravet-syndrome