

Dravet Syndrome

Dravet [syndrome](#), previously known as severe myoclonic epilepsy of infancy (SMEI), is a type of [epilepsy](#) with [seizures](#) that are often triggered by hot temperatures or [fever](#).

Dravet and Bureau in [1981](#) described “benign myoclonic epilepsy in infancy” in 7 normal [children](#) with onset of myoclonic seizures in the first 3 years of life ¹⁾. The [syndrome](#) was defined as including myoclonic seizures only, except rare simple febrile seizures, with good prognosis regarding response to therapy and cognitive functions.

Dravet Syndrome (DS) is a severe epileptic [encephalopathy](#) of childhood involving intractable seizures, recurrent status epilepticus and cognitive decline. Because DS is a rare disease, available data is limited and [evidence](#)-based treatment [guidelines](#) are lacking.

Both [VNS](#) and [corpus callosotomy](#) (CC) can be effective at reducing seizure frequency. Patients with DS may benefit from earlier and more aggressive surgical intervention. Studies using larger patient cohorts will help clarify the role that surgery may play in the multidisciplinary approach to controlling seizures in DS. Further studies will help determine the appropriate timing of and type of surgical intervention ²⁾.

Pathogenesis

Loss of function in the [Scn1a gene](#) leads to Dravet syndrome (DS). Reduced excitability in cortical inhibitory neurons is thought to be the major cause of DS seizures.

Gong et al., investigated whether SCN1B and SCN2B variants are commonly happened in DS patients without SCN1A variants. A total of 22 DS patients without SCN1A variants and 100 healthy controls were enrolled in this genetic study. DNA from DS patients was sequenced by Sanger method in whole exons of SCN1B and SCN2B genes. We identified two exon variants (c.351C>T, p.G117G and c.467C>T, p.T156M), which were present both in 1000 egenomes database and in healthy controls with a frequency of 0.54% and 4%, 0.06% and 0%, respectively. Additionally, eight intron or 3 prime UTR variants showing benign clinical significance have also been identified. The results suggest that variants of SCN1B and SCN2B may not be common causes of DS according to this data. Further large sample-size cohort studies are needed to confirm our conclusion ³⁾.

Ritter-Makinson et al., showed enhanced excitability in thalamic inhibitory neurons that promotes the non-convulsive seizures that are a prominent yet poorly understood feature of DS. In a mouse model of DS with a loss of function in Scn1a, reticular thalamic cells exhibited abnormally long bursts of firing caused by the downregulation of calcium-activated potassium [SK channels](#). The study supports a mechanism in which loss of SK activity causes the reticular thalamic neurons to become hyperexcitable and promote non-convulsive seizures in DS. They propose that reduced excitability of inhibitory neurons is not global in DS and that non-GABAergic mechanisms such as SK channels may be important targets for treatment ⁴⁾.

Treatment

[Dravet Syndrome Treatment.](#)

Unclassified articles

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