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Resumo	Dravet syndrome (DS) is a rare and severe epileptic syndrome of childhood with prevalence between 1/22,000 and 1/49,900 of live births. Approximately 80% of patients with this syndrome present SCN1A pathogenic variants, which encodes an alpha subunit of a neural voltage-dependent sodium channel. There is a correlation between PCDH19 pathogenic variants, encodes the protocadherin 19, and a similar disease to DS known as DS-like phenotype. The present review aims to clarify the differences between DS and DS-like phenotype according to the SCN1A and PCDH19 variants. A systematic review was conducted in PubMed and Virtual Health Library (VHL) databases, using “Dravet Syndrome” and “Severe Myoclonic Epilepsy in Infancy (SMEI)” search words, selecting cohort of studies published in journal with impact factor of two or greater. The systematic review was according to the Preferred Reporting Items for Systematic Review and Meta-Analysis recommendations. Nineteen studies were included in the present review, and a significant proportion of patients with DS-carrying SCN1A was greater than patients with DS-like phenotype-harboring PCDH19 variants (76.6% versus 23.4%). When clinical and genetic data were correlated, autism was predominantly observed in patients with DS-like-carrying PCDH19 variants compared to SCN1A variant carriers (62.5% versus 37.5%, respectively, P-value = 0.044, P-value corrected = 0.198). In addition, it was noticed a

	significant predisposition to hyperthermia during epilepsy crisis in individuals carrying PCDH19 variants (P-value = 0.003; P-value corrected = 0.027). The present review is the first to point out differences between the DS and DS-like phenotype according to the SCN1A and PCDH19 variants.
Fomento	