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The Facts About Dravet Syndrome

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Dravet Syndrome is an increasingly recognized genetic cause of seizures in infancy. Most cases relate to mutations in the alpha-one subunit of the voltage-gated sodium channel (SCN1A). Infants with Dravet Syndrome appear to be developmentally normal until they develop their first seizure, often in the setting of a febrile illness, after which they develop variable degrees of developmental delay. Most experts believe that a subset, perhaps a substantial percent, of patients who develop seizures and fever in response to immunization in infancy with status epilepticus and subsequent developmental delay or regression suffer from Dravet Syndrome. A group of physicians writing from Australia lead by Dr. Kenneth Myers may have shined some light on this very murky area with a well-studied CASE REPORT. The authoring physicians note that Dravet Syndrome mortality is 15% over the first two decades of life and while half of these deaths occur during uncontrolled status epilepticus the other half occur after seizure control. They report that these unexplained deaths likely occur due to post epilepsy cerebral edema leading to herniation. It is not too difficult to speculate that those children who seize and survive may have cognitive injury because of the same but less severe process that leads to fatal cerebral edema in the non-seizure associated deaths.