DRAVET SYNDROME FACT SHEET

WHAT IS DRAVET SYNDROME?

Dravet syndrome, also known as Severe Myoclonic Epilepsy of Infancy (SMEI), is a rare and catastrophic form of epilepsy for which there is currently no cure. Seizures begin in the first year of life in an otherwise typically developing infant. Initial seizures are most often prolonged events (status epilepticus) and, in the second year of life, other seizure types emerge. All seizure types are remarkably resistant to medical therapy and the prognosis for Dravet syndrome is poor.

Individuals with Dravet syndrome face a higher incidence of SUDEP (sudden unexplained death in epilepsy) and have associated co-morbid conditions, which also need to be properly managed. Children with Dravet syndrome do not outgrow this condition and it affects every aspect of their daily lives.

HOW IS DRAVET SYNDROME DIFFERENT FROM OTHER TYPES OF EPILEPSY?

Many people living with epilepsy are able to control their seizures with medication and are able to live a relatively normal life. For those with Dravet syndrome (and other intractable forms of childhood epilepsy), current treatment options are extremely limited. Dravet syndrome patients require constant care to assure their safety due to the unpredictability of seizures as well as their inability to recognize real danger.

Unless a cure or better treatments for Dravet syndrome is found, individuals with this disorder face a diminished quality of life. Current treatment options are extremely limited. The constant care and supervision of an individual with such highly specialized needs is emotionally and financially draining on the family members/guardians who care for these individuals.

WHAT ARE SOME OF THE CO-MORBID CONDITIONS ASSOCIATED WITH DRAVET SYNDROME?

Co-morbid conditions associated with Dravet syndrome can include:

- Behavior issues and developmental delays
- Deficits in social and learning skils
- Movement and balance issues
- Growth and nutrition issues
- Sleeping difficulties
- Chronic upper respiratory infections
- Sensory integration disorders
- Disruptions of the autonomic nervous system (which regulates things such as body temperature, sweating, and toileting)

WHAT TREATMENTS ARE CURRENTLY AVAILABLE FOR DRAVET SYNDROME?

Unfortunately there is no one treatment protocol which works for every patient. Finding the medication(s) that work best for each individual is often a matter of trial and error. However, some medications have proven to be more effective in treating this syndrome, while others should be avoided due to their affect on the sodium ion channels.

Medications/treatments which have proven effective in some Dravet syndrome patients include:

- Stiripentol (Diacomit)
- Clobazam (Frisium)
- Depakote
- Topamax
- Ketogenic diet

Medications which may increase or prolong seizures in some patients with Dravet syndrome include:

- Dilantin (phenytoin)
- Lamictal
- Tegretol

HOW CAN SEIZURES BE PREVENTED?

Along with finding the medication or combination of medications that is most effective for the patient, it is important to recognize seizure triggers and avoid them when possible. These triggers will vary from patient to patient, but may include:

- Quick changes in environmental temperature (both hot or cold)
- Overexertion
- Overexcitement
- Overheating
- Stress
- Pattern sensitivity
- Light sensitivity
- Loud noises

HOW CAN WE PROVIDE THE SAFEST ENVIRONMENT FOR A CHILD WITH DRAVET SYNDROME?

First, it is important to realize that each child is an individual, not a diagnosis. A strong line of communication and support between parents/guardians and teachers is the best way to assure that a child with Dravet syndrome has a safe and appropriate school environment that will offer the best learning opportunities possible.

By speaking with parents/guardians, it will be easy to determine the seizure triggers that should be avoided. For instance, if light sensitivity is a trigger, fluorescent lighting may need to be replaced in the classroom. If the school is not air-conditioned, a window unit may be necessary in the classroom to prevent overheating. The majority of children with Dravet syndrome require a one-on-one aide in the classroom to assure their safety.

Due to the unpredictable nature of Dravet syndrome, teachers and school staff may have to respond to a seizure without any warning. It is important for the school staff, nurse, and local emergency medical personnel to understand that the treatment for seizures in a child with Dravet syndrome may vary from typical seizure protocol. **It is vital that the medical protocol established by the child's neurologist be followed at all times, particularly since some emergency intervention medications are contraindicated in patients with Dravet syndrome**. If there is a question about the protocol once it has been received by the school, the child's neurologist should be contacted immediately. **The time to question a protocol is not during a seizure**. It is not uncommon for a child with Dravet syndrome to experience multiple and/or prolonged seizure events, so each seizure must be treated quickly and aggressively.

TEACHING THE CHILD WITH DRAVET SYNDROME

There can be many challenges in teaching a child with Dravet syndrome. Aside from developmental delays, it is not uncommon for a patient with Dravet syndrome to miss multiple days of school due to seizures, illness, or medical appointments. A teacher who can adapt to the child's needs with a flexible teaching plan is the best option.

Due to seizure activity or medications, a child's behavior and demeanor may change frequently. For instance, during a medication change, a child may tire more easily and have a more difficult time concentrating on a task. Or prior to a seizure, behavior may change and a child may be more aggressive or argumentative. As the teacher and school staff learns more about the child as an individual they will learn what is typical and what is unusual behavior for the child and can establish a plan around their specific needs.

To learn more about Dravet syndrome and current research, we invite you to visit our website, www.dravetfoundation.org. You may direct specific questions or requests for more information to info@dravetfoundation.