Understanding seizures with Dravet syndrome

Harrison, at age 2
Living with
Dravet syndrome

Jazz Pharmaceuticals

Dravet Syndrome Foundation

Seizures, Dravet syndrome, and the journey ahead

YOU ARE NOT ALONE

DRAVET SYNDROME AFFECTS
1 IN 15,700 PEOPLE

DRAVET SYNDROME AFFECTS
6% OF CHILDREN
WHO HAVE EPILEPSY AND ARE YOUNGER THAN 3 YEARS OLD

Dravet syndrome. Two words that you may have never heard before your loved one received a diagnosis, and since then it's all you seem to think about.

Ever since your loved one was diagnosed, you've probably had a million thoughts running through your mind.

It's perfectly normal to experience a range of emotions, especially with your child having seizures and being unsure of what lies ahead. Just know that you can take valuable action by learning how to work with your family and care team to prepare for seizures, identify them, and even reduce their occurrence.

We hope you'll find comfort in knowing there is a great deal of research, support, and guidance available to you.

Katie, at age 2
Living with
Dravet syndrome

What is Dravet syndrome?

Dravet syndrome (noun): drä-vā, sin-drōm—a rare, severe, and life-long form of epilepsy that typically begins in the first year of life. It is marked by frequent, often prolonged seizures, developmental delays, speech impairments, and other possible symptoms.

Dominic, at age 4
Living with
Dravet syndrome

Possible signs and symptoms of Dravet syndrome

No two people living with Dravet syndrome are exactly alike and symptoms vary, including the types of seizures your loved one experiences. The table below contains some of the symptoms that may occur in Dravet syndrome. *Your loved one may not experience all of these.*

Common signs and symptoms of Dravet syndrome:

SEIZURES

- Seizures triggered by certain events, including overheating, rapid temperature changes, lighting, and visual patterns
- Prolonged seizures in an otherwise normally developing infant

MOTOR SYMPTOMS

- Instability and difficulty walking
- Intention tremor (a tremor produced with purposeful movement toward a target)

BEHAVIORAL DISORDERS

- Attention deficits/hyperactivity
- Oppositional disorders (defiant, disobedient behavior)
- Personality disorders (unstable mood, behavior, and relationships)
- Autistic-like traits

OTHER

- Developmental delays after initially normal development
- Sleep disturbances

Understanding seizures with Dravet syndrome

Seizures are a defining sign of Dravet syndrome. A seizure is a sudden, uncontrolled electrical disturbance in the brain that may cause changes in your loved one's behavior, movements, and levels of awareness.

In Dravet syndrome, seizures generally start during the first year of life, and they are often triggered by an illness or fever.

The information provided is not intended to replace a doctor's medical guidance.

Graeme, at age 12
Living with Dravet syndrome

MYOCLONIC SEIZURES START BETWEEN AGES 1 TO 5 IN

85% OF CHILDREN WITH DRAVET SYNDROME

SEIZURES IN DRAVET SYNDROME OFTEN LAST LONGER THAN **5** MINUTES

IN INFANTS AND CHILDREN, THESE

LONG-LASTING SEIZURES

TYPICALLY HAPPEN EVERY FEW WEEKS

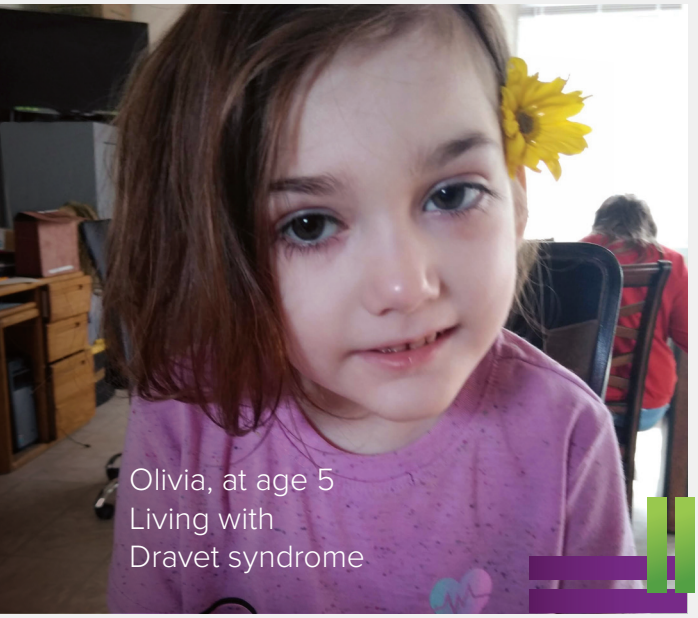
Types of seizures most common with Dravet syndrome

- Generalized tonic-clonic
- Hemi-clonic seizures, along with other focal aware or impaired awareness seizures
- Myoclonic
- Atypical absence
- Tonic
- Atonic

In Dravet syndrome, it is common for many of these seizure types to last longer than 5 minutes or occur very close together without breaks. This is known as status epilepticus, which requires emergency medical intervention. Seizure clusters can also occur when seizures happen close together with breaks in between them.

See pages 10-11 for a list of seizure types and tips on how to identify them.

The importance of early recognition, diagnosis, and treatment



Olivia, at age 5
Living with
Dravet syndrome

By catching seizures early on, your loved one's doctor can develop the right treatment plan.

Unrecognized and/or uncontrolled seizures are linked to developmental disabilities, learning and behavioral disorders, and other negative long-term outcomes.

Since some of these impairments may not be obvious until seizures persist for years, it is essential to recognize uncontrolled seizures and talk to your loved one's doctor right away.

Uncontrolled or undetected seizures can **increase** the severity of

Learning and/or cognitive impairments

Developmental disabilities

A few tips to help:

Talk with your doctor as soon as you notice a seizure in your loved one. See pages 10-11 for a list of seizure types and how to identify them.

According to the Epilepsy Foundation, **seek emergency medical attention** if a seizure lasts for more than 5 minutes or there is no recovery in between seizures.

While it may be difficult in the moment, **take a video** to show to your doctor if you suspect your loved one is having a seizure.

The information provided is not intended to replace a doctor's medical guidance.

Tips on how to identify a seizure

Seizures in Dravet syndrome can vary from momentary disruptions of the senses, to short periods of unconsciousness or staring spells, or even convulsions. Typically, seizures are short lived; however they can become long-lasting, leading to status epilepticus. Any seizure type listed below, including non-convulsive types, can lead to status epilepticus.

Your loved one may experience many different types of seizures. On the next page is information to help you identify seizures most common with Dravet syndrome—so you can talk with their doctor right away. The information provided is not intended to replace a doctor's medical guidance.

Some common seizure triggers include

- Hyperthermia (overheating or above-normal body temperature)
- Illness and/or fever
- Lighting
- Visual patterns
- Hot baths
- Emotional stress or excitement
- Exertion or physical activity

Types of seizures common with Dravet syndrome and how to identify them:

HEMI-CLONIC

- A type of focal seizure commonly seen with Dravet syndrome
- Rhythmic jerking movements on one side of the body
- Usually prolonged in people with Dravet syndrome

GENERALIZED TONIC-CLONIC

- Formerly known as “grand mal” seizures and is what most people think of when they hear the word seizure
- Person loses consciousness or awareness, muscles extend and become rigid, and then muscles jerk rhythmically on both sides of the body

MYOCLONIC

- Sudden, brief shock-like muscle movements or jerks that usually don’t last more than a second or two
- Person is alert
- May occur in clusters and may be more pronounced upon wakening, or when concentrating on a task

ATYPICAL ABSENCE

- Blank staring, with eye blinking, chewing movements, or lip smacking
- Could include finger or hand rubbing, or other small hand movements
- May begin and end gradually, usually lasting for about 5-30 seconds
- May be difficult to distinguish between this seizure type and typical behavior in those with cognitive impairment

ATONIC

- May be referred to as a “drop attack” or “drop seizure”
- Person has a sudden loss of muscle tone and goes limp
- Mild seizures look like a head nod or drop, while severe seizures can cause a person to fall to the ground
- People with these types of seizures may wear helmets to protect from injuries

FOCAL

FOCAL AWARE

- Used to be called simple partial seizures
- Person is fully alert and able to interact
- Experiences can include
  - — Involuntary motor movements on one side of the body
  - — Intense sensory or emotional episodes, such as déjà vu or feeling unexplained emotions

FOCAL IMPAIRED AWARENESS

- Used to be called complex partial seizures
- Person loses awareness, may not respond, and has no memory of the seizure
- Can vary between people, but may start with a blank stare, followed by chewing/lip smacking
- Can also include random activity like picking at the air or clothes, attempting to pick up objects, remove clothing, repeat words or phrases, etc

Your loved one may not experience all of these seizure types.

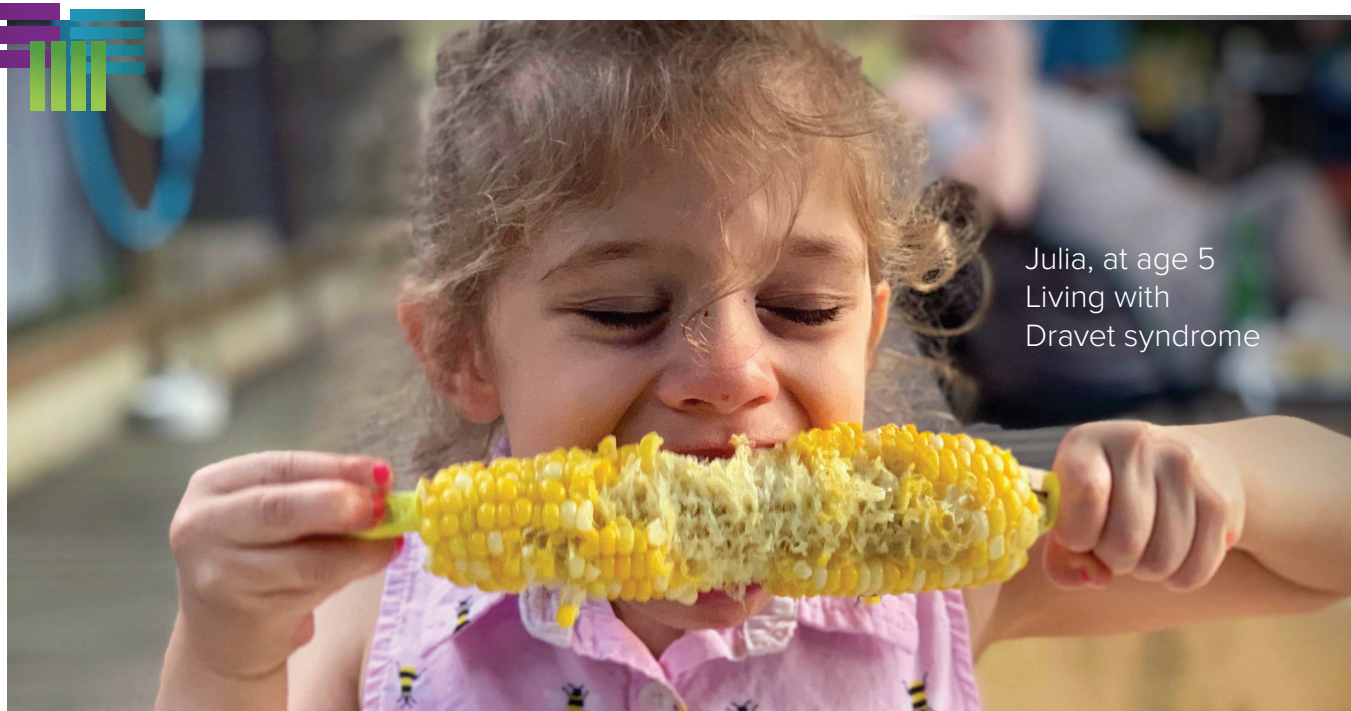
Living with Dravet syndrome

Seizures can change over time

Although seizures usually start in infancy, this is an ever-evolving condition—seizure types and severity of seizures can change over time.

This section contains information about the changes in seizures you may witness in your loved one from birth to adulthood.

The information provided is not intended to replace a doctor's medical guidance.



Julia, at age 5
Living with
Dravet syndrome

During infancy

The most common initial seizure types include

- Unusually long hemi-clonic seizures
- Generalized tonic-clonic seizures
- Infants may also experience recurrent seizures that last more than 5 minutes or seizures that occur close together without recovery periods in between

By approximately 2 years of age

Your loved one may also experience

- Myoclonic seizures
- Focal impaired awareness seizures
- Shorter generalized tonic-clonic and hemi-clonic seizures

Obtundation status may occur after 2 years of age, a type of long-lasting, non-convulsive seizure marked by less alertness.

During early childhood

Seizure types may include

- Myoclonic seizures
- Focal impaired awareness seizures
- Atypical absence seizures

During adolescence/adulthood

People may continue to have uncontrolled seizures: focal, generalized tonic-clonic, atypical absence, myoclonic, and tonic seizures. Nighttime generalized tonic-clonic seizures become more common, and seizures are less frequently triggered by hyperthermia.

Alyssa, at age 10
Living with Dravet syndrome

Recognizing and tracking seizures

How to help your doctor identify the seizure type your loved one is experiencing

1. As difficult as it may seem in the moment, try to take a video to show your doctor if you suspect your loved one is having a seizure. This will help the doctor better identify the type of seizure and how to proceed with treatment.

2. Keep a diary of seizure activity in a dedicated notebook or by downloading a seizure tracking app. This can help you track to the best of your ability how often seizures happen and their possible triggers.

The information provided is not intended to replace a doctor's medical guidance.

> Follow the seizure action plan you have created with your doctor to determine when to use rescue medication and when to visit the emergency department.

How your loved one's doctor may approach monitoring and treating seizures

There are many monitoring and treatment options for recurring seizures:

Electroencephalogram (EEG)
A test used to determine if there is abnormal brain activity that is causing seizures.

Magnetic resonance imaging (MRI)
An imaging technique used to look for abnormalities in the brain that could be causing the seizures.

Antiseizure medications
Therapies used to treat seizures. Certain rescue medications may be used for seizures that last longer than 5 minutes, or occur very close together without breaks.

Dietary approaches
Your loved one's doctor may prescribe a high-fat, low-carb medical diet that needs to be carefully monitored by a dietitian.

Surgical therapies
For some people, the use of implanted devices or surgical methods may be recommended.

Some people with Dravet syndrome may not respond to these treatment approaches. Talk with your loved one's doctor for more information about what may work for your loved one.

Seizures may be worsened by certain antiseizure medications that target sodium channels. Ask your loved one's doctor for a list of these therapies to potentially avoid.

It may be necessary to try more than one antiseizure medication at a time to effectively reduce the number of seizures because these drugs may work through different targets in the body. These therapies may take a while to start working, and it is not uncommon for people to be on multiple therapies at a time.

Where can I find support?

Finding out that your loved one has Dravet syndrome can be overwhelming, but there is hope. No two experiences with Dravet syndrome are exactly alike, and you and your loved one are not alone. There's a community of support and resources to help you and your loved one navigate life with Dravet syndrome. If your loved one is newly diagnosed, we encourage you to connect with the Dravet syndrome community of caregivers and experts who openly share their stories and advice.

The Dravet Syndrome Foundation (DSF)

Founded in 2009 by a group of parents, the DSF was created to support and advance research in order to find a cure and treatment options for their children. Since then, the DSF has advanced research with more than $4.6 million in funding. The DSF is here to help you and your family as you work through your fears and concerns. The foundation offers many programs and services to help you cope, keep up with the latest in research, and educate yourself to assure your loved one is receiving the best quality of care.

Learn more about the Dravet community by visiting

DravetFoundation.org

@dravetsyndromefoundation

@DravetSyndromeFoundation

@curedravet

Jazz Pharmaceuticals

At Jazz Pharmaceuticals, we are dedicated to offering innovative solutions that go beyond medicine. Our mission is to transform the lives of children, adults, and families who live with rare neurological diseases. Families are at the center of everything we do.

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