



**DRAVET**  
Syndrome UK

*Hope for families with  
life-limiting epilepsy*

## **Dravet Syndrome Family Guide**



[www.dravet.org.uk](http://www.dravet.org.uk)



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## DRAVET SYNDROME PARENT'S GUIDE

Dravet Syndrome UK was founded in October 2008 and registered with the Charity Commission of England and Wales in January 2009. It was founded by three mums and one grandparent who had children diagnosed with the condition. They realised the lack of awareness, appropriate support and medical research opportunities within the United Kingdom. Since the charity was founded the membership has increased dramatically, rising from just 30 families to nearly 400. Dravet Syndrome UK now has a team of trustees and a world-renowned Professional Advisory Board.

The charity operates with three main aims:

- To fund medical research into Dravet Syndrome.
- To raise awareness and understanding of Dravet Syndrome within the professional community.
- To support families affected by Dravet Syndrome emotionally, practically and financially.

From very early on the Dravet Syndrome UK team recognised the lack of information available to families and the idea for this family guide started to take shape. The aim is that this guide will be a resource that you and your family can come back to time and time again when you face particular issues with your Dravet child/adult.

We hope that it addresses the questions that are regularly posted on our Dravet Forum and that we have been able to share the experiences of many families through the use of case studies.

For newly-diagnosed families, we hope that the guide will provide many of the answers that you are faced with when you have been given a diagnosis for such a little-known condition, as well as providing signposting to organisations that may be able to help and support.

As Dravet Syndrome is a spectrum disorder there may be parts of the book that are not relevant to your family, but we do sincerely hope that you and your family members find it a useful resource.

Thanks to the team at Edelman for all their input into the guide and to our professional advisors Professor Sanjay Sisodiya and Honorary Clinical Associate Professor Sameer Zuberi. Thanks also to the families who kindly contributed case studies.

With very best wishes  
Dravet Syndrome UK



Professor  
Sameer Zuberi



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## I. INTRODUCTION

### a) What is Dravet Syndrome?

Dravet Syndrome is a rare and severely disabling type of epilepsy presenting in infancy, sometimes called a catastrophic epilepsy. It was first described in 1978 by Charlotte Dravet, a paediatric epilepsy specialist working in Marseille. Other names that have been used for the condition include Severe Myoclonic Epilepsy of Infancy and Severe Polymorphous Epilepsy of Infancy.

A 'syndrome' is a group of signs and symptoms that, when considered together, suggest a particular medical condition. An epilepsy syndrome is diagnosed from the combination of age of onset of seizures, the different types of seizures, how they change over time, findings on EEG tests and other features such as behaviour and learning. As such, epilepsy is just one part of Dravet Syndrome; there are various other health conditions and symptoms, which are often found in children with the condition.

Dravet Syndrome is rare, affecting between one in 29,000 to one in 40,000 people. Normally, the first seizure takes place in the first year of life; three-quarters of these seizures accompany a high fever caused by an illness. Sometimes, if a child has a fever caused by immunisation, this might be associated with a seizure. At first, doctors may diagnose a one-off febrile convulsion (a 'fit' that a child might have associated with a very high fever). Febrile convulsions are fairly common and, while very alarming for parents, most children tend to have normal development and have only one or very few seizures restricted to early childhood. However, with Dravet Syndrome, other seizures follow, increasing in severity.

Seizures in the first year of Dravet Syndrome are often clonic (jerking movements), prolonged, and tend to affect one side of the body.

The child's development during the first year of life is usually normal; after this first year, a range of seizures may appear including:

- **Myoclonic seizures, single or multiple muscle jerks**, which may involve one part of the body or the whole body.
- **Atypical absences** (where the child will stare) with brief loss of awareness.
- **Focal seizures**, which may involve loss of awareness.
- **Non-convulsive status** where the child develops a groggy, poorly functional state.
- **Serial seizures or status epilepticus**, a prolonged seizure or a cluster of shorter seizures with little or no recovery in between.

Patients with Dravet Syndrome are particularly prone to status epilepticus, therefore swift medical intervention is essential to stop the seizure as soon as possible to reduce the risk of damage to the brain or breathing problems caused by a long seizure.

From the second year onwards, the child's development slows down or regresses, sometimes severely. Features of autism and attention deficit hyperactivity disorder (ADHD) are common.

Dravet patients often experience mobility issues, in particular with regards to an unsteady gait called ataxia. Some patients are affected severely, whilst in others the problem may be less severe.

*Dravet Syndrome  
is about much more  
than epilepsy*



## b) Why does my child have Dravet Syndrome?

About 80% of patients with Dravet Syndrome have a mutation (or change) in a gene called SCN1a. The SCN1a gene contains instructions (the genetic code) for the creation of a protein that controls how sodium ions move into the cells in our bodies. A change in this gene may lead to faulty functioning of this protein, called a sodium ion channel, in the brain. The movement of sodium ions in and out of cells help control electrical messages in the brain so a faulty ion channel can cause seizures. Not all mutations to this gene cause Dravet Syndrome. Other SCN1a mutations are associated with other less severe forms of epilepsy, such as Genetic Epilepsy with Febrile Seizures + (GEFS+).

In Dravet Syndrome, the gene mutation nearly always arises spontaneously and is not passed down (inherited) from parents. It is rare for mutations of the SCN1a gene to be passed from parent to child; however, some people with Dravet Syndrome may have some history of febrile seizures or epilepsy in their extended family. Much remains to be understood about the causes of Dravet Syndrome and research is ongoing. In children with Dravet Syndrome who do not have SCN1a mutations, other genetic mutations are being explored.

**Above all, the one thing that families need to understand is that it is not their fault. Gene mutations happen in everyone. It is simply random when they occur in an important gene like SCN1a.**

*In most cases Dravet Syndrome is caused by a gene mutation, which occurs randomly*

## c) Is there a cure for Dravet Syndrome?

Unfortunately there is no cure for Dravet Syndrome. Treatment focuses on controlling or minimising seizures in order to minimise their impact on development and reduce injuries due to seizures. Anti-epileptic drugs are used, but these are not always effective in people with Dravet Syndrome. Comprehensive testing and support is required for the multiple challenges that people with Dravet Syndrome and their families face. Many children with Dravet Syndrome have a good life expectancy, however children who develop severe disability may have problems which will affect their lifespan.

## d) Sudden Unexpected Death in Epilepsy Patients (SUDEP)

SUDEP is when a person with epilepsy dies suddenly and prematurely and no other cause of death is found. Children who have very long seizures are tragically at risk of death and patients with Dravet Syndrome are at a higher risk of SUDEP than many other types of epilepsy. This topic is something that is very difficult to discuss, yet many parents believe it is better to have the full information and feel empowered to do everything they can to ensure their child leads a safe and happy life.

Professionals believe that the best way to try and prevent death due to SUDEP and status epilepticus is to try and control the seizures as much as possible. Working with your medical team to find the best treatment plan for your child is the best way to try and achieve a good level of seizure control, whether this be through medication, dietary treatments or alternative treatments. Statistically by the age of seven years, five out of 88 (6%) children born in the UK between 2003 and 2007 with Dravet Syndrome had died due to SUDEP (3) or status epilepticus (2).

SUDEP Action provides information and support, which can be accessed online. Please see the resource section for contact details (page 61).

## 2. SEIZURES

Dravet Syndrome is initially characterised by prolonged recurrent seizures. Seizures usually start in the first year of life; over time other seizure types can occur:

### a) Initial Seizures

Recognising **febrile seizures** is important in the diagnosis of Dravet Syndrome. Febrile seizures are caused by a high or rapidly rising temperature, often due to illness or vaccination. These are usually tonic clonic or hemiclonic seizures.

**Tonic clonic seizures** involve a stiffening of the arms and legs (tonic phase), followed by jerking of the arms, legs, and head (clonic phase).

**Hemiclonic seizures** are similar, though only one side of the body convulses.

Febrile seizures are very common in childhood and usually are not associated with a form of epilepsy.

The first suggestion of Dravet Syndrome may be when the first febrile seizure happens in early infancy and is prolonged, lasting more than ten minutes and sometimes over half an hour (status epilepticus). These very long seizures may cause the child to be admitted to intensive care and have emergency treatment in A&E.

### b) Other Seizures

These seizure types may occur between ages one to four:

**Myoclonic seizures:** These can be described as 'jumps'. They are caused by rapid contraction and relaxation of the muscles. Small jerks in sleep are perfectly normal and can occur in people without epilepsy. In someone with epilepsy, myoclonic seizures cause abnormal movements on both sides of the body at the same time resulting in a jerk of the shoulders and arms or a drop of the head. Sometimes a rapid myoclonic seizure will cause a fall.

#### **Focal seizure with impairment of awareness:**

With this type of seizure the person may appear unresponsive and confused as their consciousness is impaired. Automatic movements such as smacking of the lips, wandering, or fumbling movements of the hand may be present.

**Atypical absence seizures:** Absence seizures are usually brief (usually less than 20 seconds), and have a sudden start and finish. They have two essential components:

- Clinically, the impairment of consciousness (absence) – they can be associated with head nodding and a fall may occur.
- Electroencephalography (EEG) shows generalised spike-and-slow wave discharges.

**Atonic seizures:** Also called drop seizures, akinetic seizures or drop attacks, atonic seizures are a type of seizure that comprises a brief lapse in muscle tone caused by temporary alterations in brain function. The seizures are brief – usually less than 15 seconds. The seizure itself causes no damage, but the loss of muscle control can result in indirect damage from falling. In some cases, a person may become temporarily paralysed in part of his or her body. This usually does not last longer than three minutes.

**Non-convulsive status:** Refers to less-than-full mental capacity in a patient, typically as a result of continuous seizure activity.



## c) Changes to Seizure Patterns

### Increase in seizure frequency, decrease in

**status episodes:** Generally as the child gets older, typically around four years of age, status seizures begin to settle. However, patients with Dravet Syndrome will remain prone to status seizures, particularly when experiencing illness and infection. Seizure frequency may increase around the time when the status seizures begin to settle, therefore patients may have more seizures but they may be shorter in duration and not always require emergency medication.

### Development of focal seizures with impairment

**of awareness:** Sometimes a person may get warning of a seizure called an aura and the patient may walk up to their parents/carers in this phase. Once consciousness is impaired, the person may display automatic behaviour such as lip smacking, chewing or swallowing. There may also be loss of memory (amnesia). The person may still be able to perform routine tasks such as walking, although such movements are not purposeful or planned. You may not recognise that anything is wrong. It can be very difficult to know if a patient is experiencing a complex partial seizure, particularly if they are non-verbal or have a severe developmental delay or learning disability.

**Development of myoclonic seizures:** These seizures may appear, generally between the ages of one and four, often building up to a generalised tonic clonic seizure.

### Development of nocturnal (night-time)

**seizures:** As children get older they may begin to experience their seizures at night, particularly as they transition through sleep phases. For some families this feels like a positive change as their child will not sustain any injuries from falling, but for others it may cause additional anxieties and fears (see section [d] opposite).

## d) Nocturnal (Night-time) Seizures

Night-time can be the most stressful time for parents due to the worry about missed seizures and the risk of SUDEP (sudden unexpected death in epilepsy).

There are a number of options to reduce worry and incidence, notably:

- Obtaining a specialist bed/cot for your child. These beds can have high sides to ensure the patient cannot fall out and injure themselves. Some cots have sides that are high enough to ensure the patient cannot climb out.
- Sleeping in a double bed with your child.
- Pulse oximeter machines that monitor a person's heart rate and oxygen levels. Typically when someone has a seizure their heart rate increases and their oxygen levels drop. If they are being monitored on a pulse oximeter machine during a seizure the monitor may sound an alarm, so that parents/carers can administer emergency treatment if necessary. The pulse oximeter is attached to the patient by a sticky probe, generally on the toe. These machines are not routinely available on the NHS, as so far there is no proof that they reduce the chance of SUDEP, however they can provide reassurance for many families. It is important for families to remember that machines can develop faults. This type of monitoring should never be a replacement for the observation of clinical signs.
- Epilepsy mattress monitors which alert to tonic clonic seizures and can also be set up to sound an 'out of bed' alarm to alert if the patient has got up.
- Good quality video camera (baby monitor) or CCTV.
- Having a single bed in the patient's bedroom for a parent or carer to use when the patient is unwell.
- Exploring respite options, for example the patient staying somewhere where there is waking night staff or accessing waking nights in your own home.

## The Dravet Syndrome UK Night-time Monitoring Programme

Since 2009 Dravet Syndrome UK (DSUK) has been funding Pulse Oximeters for our families as a way of monitoring Dravet children and adults throughout the night. At the beginning of 2012, we gave five families the opportunity to trial the Emfit Mattress Monitor. Following feedback, we have come to the conclusion that the monitoring system for each family is a very personal choice and what works for some families may not work for others. As a result, families can apply to DSUK for one of the following monitors: Pulse Oximeter Monitor, Emfit Epilepsy Alarm, Summer Baby Touch Digital Video Monitor.

Please go to <http://www.dravet.org.uk/family-support/monitors/> for more information.



### 3. SECONDARY DIAGNOSIS: OTHER CONDITIONS ASSOCIATED WITH DRAVET SYNDROME

Epilepsy and developmental delays are the main problems for children with Dravet Syndrome; however; they also have a number of other health problems. Some conditions are common, other less so, and all will vary in severity from child-to-child. Secondary conditions can include the following:

#### a) Autistic Spectrum Disorder

Autism affects how a person communicates with, and relates to, other people and how they make sense of the world around them. It is a spectrum condition, which means that, while all people with autism share certain difficulties, their condition will affect them in different ways. Some people with autism are able to live relatively independent lives but others may have accompanying learning disabilities and need a lifetime of specialist support. People with autism may also experience over- or under-sensitivity to sounds, touch, tastes, smells, light or colours.

It is not unusual for Dravet patients to receive a secondary diagnosis of autism. As autism and Dravet Syndrome are both spectrum disorders patients will vary considerably, with some children perhaps showing mild autistic traits through to others having a full diagnosis of profound autism. Dravet patients often display communication and social difficulties and it can be useful to be referred to an ASD specialist if you are at all concerned.

*More than half of people with Dravet Syndrome report some characteristics of autism*

#### b) ADHD/Behavioural Difficulties

Attention deficit-hyperactivity disorder (ADHD) is common, affecting up to 10% of all school-age children, particularly boys. It normally emerges before seven years of age. Children with ADHD act without thinking; they are hyperactive, impulsive and have trouble focusing. They may understand what is expected of them but struggle to complete a task because they cannot sit still, pay attention, or focus to details. All children act like this sometimes, but with ADHD these symptoms are present more often. They impair a child's ability to function socially, academically and at home. The positive news is that with good treatment, children with ADHD can manage their symptoms.

#### c) Dysautonomia

Dysautonomia, or problems with autonomic body functions, are present in more than 60% of people with Dravet Syndrome. Issues include problems with temperature regulation, decreased sweating, fast heart rate (tachycardia), and sluggish digestion and blood circulation.

#### d) Growth and Nutrition Concerns

These include so-called 'failure to thrive' (underweight, slow growth, small stature), osteopenia (poor bone density), scoliosis (curvature of the spine), and problems with eating, appetite or with the absorption of nutrients. Each child is unlikely to get all of these issues, but about 60% of children with Dravet Syndrome show some growth and nutrition issues of varying extent.

### **e) Coordination and Orthopaedic Disorders**

Up to 80% of families of people with Dravet Syndrome report symptoms including gait (walking) disturbances, such as lack of coordination (ataxia) and crouch-gait. Flat feet and turned-out toes are common along with poor muscle tone (hypertonia), tight muscles (spasticity) or hypermobile (over-flexible) joints.

### **f) Infections and Immune Problems**

Frequent ear, nose, throat, lung and digestive infections can be a problem for some people with Dravet Syndrome. At this point in time it is unknown whether this is because of the overall disability or a more specific relationship to Dravet Syndrome.

### **g) Sleep Problems**

Sleep disturbances that are severe, persistent and difficult to treat are also common in children with Dravet Syndrome (see page 25 for advice on managing sleep problems).

*Sleep disturbance is very common in Dravet patients and can have a huge impact on quality of life for both the patient and the family. Managing sleep issues is an important aspect in the care of your child*



## 4. EMERGENCY MEDICATIONS

It may sometimes be necessary for you to give your child emergency medication to control their seizures. Below is a list of emergency medication options. Medication varies from child-to-child and the following are not placed in any order of importance.

### a) Emergency Medication for Home Use

- **Diazepam:** First marketed as Valium, diazepam is a benzodiazepine drug and commonly used for treating anxiety, panic attacks, insomnia, seizures including status epilepticus (state of continuous seizure) and muscle spasms. It offers a rapid onset of action and has high efficacy rates. Home use diazepam is administered rectally.
- **Midazolam:** Midazolam for home use is administered to the side of the cheek (buccal) or as drops into the nose (nasal) and is available as a licenced pre-filled syringe. Using a pre-filled syringe makes administration easier and as there is no need to measure out the correct dose, it means the administration is less prone to error. Two preparations are available with different concentrations of the medicine.
- **Paraldehyde:** Used to treat status epilepticus and, unlike diazepam and other benzodiazepines, does not suppress breathing at therapeutic doses and so is safer when no resuscitation facilities exist or when breathing is already compromised. This makes it a useful emergency medication for children with epilepsy. It is not an easy medication to use nor is it easy to obtain. Families should also be aware that it may come with a very short shelf life. Home-use paraldehyde is administered rectally. Paraldehyde has a very strong smell and the odour can stay on the breath for about 24 hours, so don't be alarmed.

### b) Emergency Medication Used in Hospital

- **Lorazepam:** A high potency benzodiazepine drug used for the short-term treatment of acute seizures. Lorazepam is generally given intravenously when in hospital.
- **IV (intravenous) phenytoin:** IV phenytoin is highly effective at stopping seizures. The intravenous treatment route is preferred because of the delay in absorption of phenytoin when administered into the muscle. Although phenytoin is not recommended as a daily maintenance medication for Dravet patients, it can be particularly effective at stopping status seizures when used intravenously. However, some children can have more seizures when given phenytoin.
- **IV phenobarbital:** IV phenobarbital depresses brain function and produces drowsiness, sedation, and hypnosis. For parental administration, direct IV administration is recommended.

*Using emergency medication at home can be a very scary and stressful experience. Try to keep calm and take things one step at a time, following your child's individual protocol*

## c) Ventilation

Ventilation is not a medication in itself. The anaesthetic agents are the medication and then ventilation becomes essential as natural breathing rhythms are lost.

- **Why ventilate?** There are two main reasons why your child may need to be ventilated, either because they are unable to breathe effectively for themselves or because they are unconscious and are unable to protect their airway sufficiently. The first occurs when their own breathing is poor; ineffective or absent, and is most commonly due to infections (locally in the lungs or more widespread sepsis), obstruction or tiredness. Problems with breathing can also be caused by some chronic conditions. The second reason, when they are unable to 'protect their own airway', is normally due to fluctuating or very low conscious levels – this typically happens when a child is seizing or is post ictal (following a seizure). An unconscious child is in danger because they are at risk of the contents of their stomachs passing into their lungs and causing obstruction or infection (aspiration), but also they may not be able to breathe effectively enough to take in the oxygen required by all the other cells and tissues in their bodies.
- **What happens when your child is ventilated?** Your child will be given some medication to help them relax; even if they appear unconscious. An endotracheal tube will be placed into the mouth and fed down the trachea (wind pipe) under direct vision. It sits just above a cartilage ring in the main bronchus (called the carina) and is where the trachea splits into two so that ventilation is effective to both lungs. This is called intubation. The tube is then connected to a ventilator; sometimes called a life support machine.

The machine then takes over or assists your child to breathe until they are well enough to do without it. This time can range from a couple of hours to a few days, and occasionally weeks. There are several different ways of using these machines, and each child will need something different, but they all deliver air/oxygen into the lungs and take carbon dioxide away. During this time your child will be monitored extremely closely, particularly their heart rate and their saturations, and will be given sedation to ensure they are comfortable and sleepy.

- **Will we be consulted?** If your child does require ventilation this will be discussed with you by a paediatrician and an anaesthetist. They will try and discuss the 'pros & cons' with you before they proceed, but it is sometimes an emergency procedure and so they may have to explain the procedure afterwards. It is up to the individual doctors and nurses whether you are able to be present during the intubation, but they will always keep you fully informed. This is a difficult time as your child will be in an intensive care unit with several tubes and wires around them. If you do not have a paediatric intensive care unit in your local hospital your child will need to be transferred to the nearest centre.



### d) The Practicalities of Using Emergency Medications

It does not matter how many times you have to administer emergency medication to your child, it will always be a scary experience. Over time parents get used to having to do it but the fears and anxieties never go away. It is a stressful situation: you may find that as well as administering the medication, you are dialling 999 and trying to calm other children. Take a deep breath and try to remain as calm as possible. What you are doing for your child is amazing and potentially life-saving. Many parents say that at the time of administering the medication they remain calm, it's afterwards when their child has come round that the panic and upset sets in. This is completely normal and is the body's way of coping in times of stress.

### e) Emergency Protocols

As Dravet Syndrome is a rare condition it would be unrealistic to expect all medical professionals to know how it presents and how it should be treated, both on a day-to-day basis and in an emergency situation. For this reason we strongly recommend pro-actively working with your medical team to produce some protocols. The three we would recommend are:

1. **Emergency seizure protocol:** Detailing the medications to be used when the patient is seizing, the order in which to use them and the appropriate doses (mg/kg).
2. **Ambulance protocol:** Detailing the medications which can be used en route to the hospital and information about the measures needed to be taken upon arrival, for example, a resuscitation call being put out. You will need to work with your local consultant who will liaise with your local ambulance service.

3. **General hospital protocol:** There will be times when you just know that something is not right with your child, yet you can't quite put your finger on it. A general hospital protocol would provide some brief information on Dravet Syndrome, any other diagnosis your child may have and what tests should be carried out, e.g. blood tests, swabs, checking ears, throat etc.

The important thing with protocols is to work WITH your medical team to ensure they are accurate and the medical staff can work with them. They should be on hospital letterhead and signed off by your consultant. Once you have your protocols, we advise you keep a copy in your child's bag at all times, at their school/college/residence, in the A&E department of your local hospital, at your GP surgery, with your ambulance station and on your local children's ward.

All protocols should include your child's information, i.e. date of birth, address, current medications, allergies and contact numbers for your medical team. We have provided some protocol examples as appendices at the back of the book.

*Having emergency protocols that are readily available and accessible can make treating your child in an emergency situation a lot easier for everyone concerned and ensure that wherever you are the same protocol is being followed*

## 5. DAILY MAINTENANCE MEDICATION

It may be necessary for you to give your child daily medications. Dravet Syndrome patients typically need to be taking a combination of medications to help control or reduce seizures.

### a) Types of Medication

It is always important to remember that medication regimes remain very individual to the patient.

- **Sodium valproate:** Also known as valproate sodium, is the sodium salt of valproic acid and is an anticonvulsant used in the treatment of epilepsy. The intravenous formulations (i.e. directly into a vein) are used when the person is unable to take the medication through the mouth.
- **Clobazam:** A benzodiazepine drug approved for use with other treatments, for patients with epilepsy who have not responded to or who are resistant to first-line drugs. It is not recommended for use in children between the ages of six months and three years.
- **Stiripentol:** Is added to sodium valproate and clobazam for treating people with Dravet Syndrome whose seizures are not adequately controlled with valproate and clobazam alone. In addition, it may be used to treat refractory childhood epilepsy in conjunction with carbamazepine. Stiripentol is currently only licenced for use in Dravet Syndrome.
- **Topiramate:** An anticonvulsant drug used to treat epilepsy in children and adults.
- **Levetiracetam:** An anticonvulsant medication used to treat epilepsy; it has been approved for treatment as a single medication in the case of partial seizures, or as an add on medication for partial, myoclonic and tonic clonic seizures.
- **Bromides:** This is one of the older anti-epileptic medications. It is not prescribed much in the UK but is often the first-line medication for Dravet Syndrome in Germany and Japan. It is used by some UK Dravet Syndrome specialists.

**Medications which can increase or prolong seizures in some patients with Dravet Syndrome include:** Carbamazepine, lamotrigine, phenytoin (although phenytoin can increase seizures if used as a daily maintenance medication, it can be useful when used as an IV as an emergency treatment).

Researchers are also investigating new treatment options for Dravet Syndrome. With two potential new medicines currently undergoing clinical trials, we anticipate exciting times ahead for the future treatment of Dravet Syndrome.

### b) A Ketogenic Diet

The ketogenic diet is a medically prescribed diet for those patients with drug resistant epilepsy and can work well for Dravet Syndrome. It is worth considering early if appropriate medications for Dravet have failed. All types of ketogenic diet have restricted carbohydrate and are high in fat. Protein is always given in an adequate amount depending on which version of ketogenic dietary therapy you are using.

These diets work by replacing fuel sources for the brain to use. In a normal diet our energy comes from carbohydrates such as rice, breads, pasta, vegetables, fruit and sugar which is why we eat it so abundantly and when eaten it changes into glucose and is used by the brain for fuel. Glucose can only be stored in the body for approximately 24 to 36 hours, and if all the glucose is not used then it is converted into fat. However, if you take this fuel source away, such as when you are not well and not eating, the brain still needs fuel to function so our body's metabolism will switch over from burning glucose to the 'back up' system of burning fat for its fuel. When we burn fat, we produce ketones. It is being in this 'ketotic' state that, for some reason, can have a positive effect on seizure control and quality of life. The reasons why are still unclear; researchers are continuing to investigate this.



There are four different types of ketogenic dietary therapy available and each diet is tailored to individual needs. It is not something that should be tried alone and you should be monitored by a dietician who has experience in ketogenic dietary therapies, as well as being supported by your neurologist. Blood tests are required whilst on the diet every few months and you should fill out weekly reports for your dietician, especially at the start of the diet to make sure that adequate ketone levels are being achieved.

*Avoid: high-carbohydrate foods*

*Consume: foods high in fat*

The diet is started with no other changes to treatment being made. Usually, it is possible to tell within three months whether a diet is helping a patient or not. If the diet is proving beneficial, the neurologist should then look at possibly reducing medication. Some patients need a little medication with the ketogenic diet, whilst others can be totally weaned off all medication. It is just a question of finding the correct levels of both diet and medication that can achieve the best possible quality of life; this is why the diet needs to be medically managed by an experienced team.

All the diets need vitamin and mineral supplementation as they are not nutritionally complete. These diets are usually started at home, although some centres prefer to admit their patients. There is no fasting necessary to start a diet and patients are not fluid restricted. If the diet is beneficial, it is usually recommended that the patient stays on it for two years before slowly being weaned off. In most cases the benefits of the diet remain even after being weaned off the diet and patients can go on to a normal diet.

For more information on all types of ketogenic dietary therapy then please contact the Matthew's Friends ketogenic diet charity (see resources section, page 61).

### **c) Fever Control**

Fever management is key to managing Dravet Syndrome as a fever often triggers a seizure. Keep a close eye on your child and manage the symptoms of a fever quickly. It may be necessary to use rectal fever treatments if your child is ill.

Your child may be reluctant to take oral medication when unwell, or perhaps they are vomiting or have a stomach bug. In cases such as these it can be easier to use rectal paracetamol suppositories. These can be put on prescription by your GP and are handy to have at home just in case the oral medication is not tolerated at any time. As with all medications seek advice from your GP before commencing anything new and never exceed the dose given to your child. Rectal fever management is there as a replacement for oral fever management, not in addition.

It is useful to know your child's 'normal' baseline temperature. Parents of Dravet patients often report that this is significantly lower than the accepted norm of 37.7. At this moment in time this is anecdotal as there are no studies into this. Sometimes patients may take extra anti-epileptic medication, such as clobazam, during a febrile illness. This is something that must be discussed with your medical team.

### **d) The Importance of Compliance to Medications**

As with all medications, it is very important to comply with all the instructions provided when administering medications to your child, especially dosing and course completion. It is suggested you speak to your child's doctor if you have any concerns over their medication – it is better to seek guidance than to alter their medication yourself.

It is essential that you speak to your medical team before altering any medications. Patients with Dravet Syndrome can be very susceptible to the slightest change so all medication changes must be done in consultation with a medical professional.

### e) Your Rights: Prescription of Generic versus Branded Medications

Generic names are the names given to the main ingredient of the medication (for example Sodium Valproate); branded names are those given by the drug company making the medication (for example Epilim) and refer to specific formulations made by a particular company. The important thing to remember is that you stick with the brand that was initially prescribed, assuming it is working for your child. So, for example, if you have always been prescribed Epilim it is best to stick with that rather than a different form of Sodium Valproate as there is a chance that a different version of your child's epilepsy medicine would have a different effect. Patients have described an increase in side effects and a change in seizure patterns as a result of using a different brand or version.

The National Institute for Health and Care Excellence (NICE) recommends that people with epilepsy continue to get the same version of their epilepsy medicine wherever possible. If the person who prescribes your epilepsy medicine wants to change your child's version, they should discuss this with you first.

*If, when collecting your child's medication, it looks at all different to what you are used to make sure you query it with the pharmacist.*

*Your child has the right to stay on the medication, whether generic or branded, that they are established on. If you are at all concerned speak to your GP*

### f) Taking a Proactive Role with Your Healthcare Professional

You know your child better than anyone, so it is important to monitor and report any positive and negative effects, which may be associated with your child's medication. Medicating a child with Dravet Syndrome is an art as well as a science; it will vary from patient-to-patient and as your child grows and changes. So don't be afraid to take an active role in your child's management, and discuss all questions, suggestions and concerns with your healthcare professional. Go prepared to all meetings so you can get the answers you need.

Many families report that keeping a seizure diary, or tracking seizures, is very useful. Not only is it helpful for families to keep an eye on any seizure patterns, it can prove incredibly helpful for your medical team to see a possible correlation between seizures and medications. A simple diary can be filled in by hand, noting things such as seizures, medications, child's general health and well-being, or there are a number of Apps or websites that can be accessed to fill in online.

*Do research and ask lots of questions. It is likely you will be in the driving seat of your child's care*



## CASE STUDY: THE IMPORTANCE OF DIAGNOSIS AT ANY AGE

### Late Dravet Syndrome diagnosis



**Interview with Dr Sanjay Sisodiya, Professor of Neurology at UCL Institute of Neurology and Honorary Consultant Neurologist at the National Hospital for Neurology and Neurosurgery.**

Patient X first saw Professor Sisodiya when he was in his early 50's. His case history prior to this was that he had been completely well as an infant but started to have seizures after receiving the whooping cough vaccination. From then on, his development began to slow down and his parents started to notice how he was starting to lag behind his younger siblings.

Over the next few years, learning and behavioural difficulties emerged and the medicines that were available at the time did not successfully control his seizures. Over the years, he tried various experimental drugs and new epilepsy drugs as they became available, but as he experienced many side effects, the family eventually decided to stop trying new medications.

Later in life, when patient X eventually saw Professor Sisodiya, he was still experiencing prolonged seizures and went for long periods of being unresponsive. They cautiously tried some new medication and initially patient X went downhill losing what little ability he had to speak, and also his ability to swallow. As a result he needed to have a feeding tube fitted.

However, the family and Professor Sisodiya persevered and tried more medications. They remained concerned about patient X as his ability to communicate became less and less.

At this point, Dravet Syndrome was not an obvious diagnosis to the neurologists that patient X was seeing, as they were specialists in adult neurological problems. However, in 2006 a paper was published about cases of alleged brain damage following vaccination. A significant proportion of these patients had a history consistent with Dravet and the SCN1a mutation.

The adult neurologists took notice of this paper and started re-examining their patients.

Professor Sisodiya performed a genetic test on patient X and he tested positive for the SCN1a mutation. A Dravet diagnosis was confirmed. This allowed Professor Sisodiya to understand the underlying processes causing the seizures at a molecular and cellular level, and select medications appropriate to Dravet Syndrome. The diagnosis also confirmed why patient X had so many problems with medications he was given in the past.

Since his treatment was adjusted, patient X has experienced less frequent seizures, interacts more and can now even speak in sentences. He has an improved memory, is able to express his own views and has recovered some of his personality that had been lost before. He is now in his early 60's.

## 6. ADDITIONAL THERAPIES

It has probably been a long road to your child's diagnosis and now, at last you can start to consider how to treat and manage your child's condition so their health and quality of life is the best it can be. It might be a shock to find out that there are often other, associated health issues with Dravet Syndrome.

### a) Managing Your Child's Health with Additional Therapies

While you will need to manage seizures as a priority (see pages 9–10), it is also important to manage the other health and developmental issues that your child may have (see pages 12–13). It will be necessary to organise developmental assessment with a range of therapists and specialists. It's best to coordinate these referrals as early as possible, even if your child isn't showing any signs of delay or difficulty. They will help you understand and manage your child's overall health needs relating to Dravet Syndrome. You will need repeat assessments at regular intervals but once you are 'in the system' you should find that these are scheduled at regular intervals by the healthcare professionals who support your child. **Don't be afraid to ask questions, or persist in getting a full range of therapies for your child.**

Children with Dravet Syndrome often need physical, occupational, speech, and social/play therapies. They benefit from a full and stimulating environment; they may also benefit from alternative therapies.

Many therapies are available on the NHS, and some parents choose to supplement NHS care with private therapy, especially if a particular alternative therapy is not available on the NHS.

The additional therapists your child needs should be coordinated by your local health service, clinical commissioning group (CCG) or social worker. In theory they should coordinate all the extra NHS services and therapies your child needs so you don't have to approach each one separately.

You may need to contact your local disability team to request an assessment and to be allocated a social worker. Social workers are there to help you and your family so do not be afraid to request one. They are often the key to accessing many services, which will be beneficial to you, your child and your family. Having a social worker should never be seen as a sign of failure. It is a sign of how proactive you are in helping your child and your family to reach their fullest potential.

### b) Additional Therapies with NHS-funding

**Speech and language therapy:** Speech and language therapists can assess and treat your child's speech, language and communication to enable them to communicate to the best of their ability. They can also support children with eating and swallowing problems and advise on alternative forms of communication that may benefit your child including:

- **PECS** – the Picture Exchange Communications System was developed in 1985 and uses pictures to help children communicate, gradually building up speaking capacity.
- **Makaton** – a type of sign language which uses signs (gestures) and symbols (written/visual) alongside regular speech to aid communication.
- **Augmentative communication devices** – includes a range of electronic devices and software to support communication.

**Finding a speech therapist:** Your GP or healthcare professional can, and should, refer you for additional therapies. However, in most areas of the UK you can also 'self-refer' i.e. contact your local service directly. You can also find an independent (private) SLP via the independent practice website [www.helpwithtalking.com](http://www.helpwithtalking.com).



**Occupational therapy:** Paediatric Occupational Therapists help children develop skills around self-care, school and play. They will assess whether your child has functional difficulties (sensory and motor skills) and help your child overcome these. For example, they might help your child learn to use the toilet and exert bladder control. They will work closely with your family to enable your child to discover just how much they can do for themselves to achieve maximum independence. Your GP or consultant can refer you.

**Physiotherapy:** Physiotherapy can help your child to achieve his/her maximum level of functional independence. It is also used to prevent and minimise recognised complications that can develop. For example, your child's mobility might reduce after a seizure or as gait difficulties progress, so physiotherapy is often ongoing. Your GP or consultant can refer you.

**Social/play therapy:** Guided play therapy helps children learn and make sense of their world or come to terms with difficult issues and emotions. This can include one-to-one activities or group play with other children. Research suggests that play therapy is effective, particularly when the parents/main carers are involved in the sessions. Your local health authority (CCG) or local authority might employ play therapists. Find out what services are available locally and ensure your GP or main health contact refers you to local services. See [www.bapt.info](http://www.bapt.info) for more information on play therapy.

### **c) Additional Therapies Less Likely to be NHS-funded**

These types of therapies are generally not funded by the NHS as there is unlikely to be any evidence based research proving efficacy. However, some families may believe them to be beneficial.

**Cranial osteopathy:** An osteopath applies very specific, skilled, light pressure onto a child's head to help assist the natural ability of the body to release stresses and tensions. It might be a comfort to the child and many therapists believe it supports and restores the central nervous system after seizures.

**Homeopathy:** Homeopathy claims to prevent and treat diseases using diluted forms of plants, herbs and minerals.

**Chiropractic therapy:** Chiropractors use their hands to treat disorders of the bones, muscles and joints, generally by manipulating the spine (similar to osteopathy). See [www.nhs.uk/chq/Pages/1098.aspx?CategoryID=68](http://www.nhs.uk/chq/Pages/1098.aspx?CategoryID=68) for more information.

**Developmental therapy:** These are extremely child-oriented therapies, which means that there is no one-size-fits-all approach. In addition, they demand a great deal from parents. They are useful for children with autism. Centred around a child's own interests or actions, the therapy slowly builds engagement, interaction, communication, affection, reasoning and symbolic thinking. It is first recommended for very young children, but the techniques are applicable through adulthood.

*It can sometimes feel as if your life is one big therapy appointment but accessing therapies can be extremely beneficial and early intervention can be key in maintaining or improving skills and abilities*

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## 7. LIVING WITH DRAVET SYNDROME

### a) Sleeping

There is an inherent relationship between sleep and epilepsy as sleep activates the electrical charges in the brain that result in seizures, which generally mirror the sleep-wake cycle. Sleeping difficulties in children suffering from Dravet Syndrome are particularly common.

- **Sleep disorders and disturbances:** Problems with sleep vary between children with Dravet Syndrome but can include insomnia (20%), premature awakening (28%), nocturnal seizures (50%) and sleep apnea (52%). These can be attributed to a number of factors including side effects from medication and seizure activity during the night. With lack of sleep being cited as a key trigger for seizures, it is something that can cause many parents a restless night.
- **Nocturnal (night-time) seizures:** Seizures are common when the brain is undergoing transitions (e.g. as your child is falling asleep or shortly before waking).

We all know that not getting a good night's sleep can affect how we function during the day, leading to fatigue and an inability to function. Nocturnal seizures are no different as they significantly reduce the time spent in deep sleep and can be dangerous for children whose parents are unable to monitor them throughout the night.

- **Lack of sleep as a cause of seizures:** Lack of sleep is often cited as a key trigger for seizures and can cause a vicious cycle, in which a child that becomes sleep deprived has more seizures, and the increase of seizure activity causes a child to become even more sleep deprived. It is therefore vital that you try to ensure your child sticks to a healthy sleeping pattern, where possible. Sleep disturbance could be a phase that your child will come out of once their bodies are used to the medication they are taking.

*If you are the parent of a child with Dravet Syndrome, it is very important that you make healthy sleep a priority for yourself too, which as we all know is often easier said than done*

- **Importance of healthy sleep for parents:** If you are the parent of a child with Dravet Syndrome, it is very important that you make healthy sleep a priority for yourself too, so that you have the energy you need to function throughout the day and look after your child. Knowing your child is being monitored during the night may give you the peace of mind to have a good night's rest. Respite, which is discussed in a later section, may be one way of ensuring you get some better quality sleep, while knowing your child is safe and cared for.



## b) Eating

It is very common for patients with Dravet Syndrome to have feeding difficulties. These can vary from child-to-child and can be caused by a number of things including side effects from medication, seizure activity and autism spectrum disorders (ASDs).

One of the first steps, if you are concerned about your child's diet or eating patterns, should be to create a food diary. A food diary could shed some light as to why your child is over or under eating. By regularly recording your child's eating habits you will be provided with useful information about their eating pattern and the potential triggers for their lack of appetite or their inclination to over-indulge at certain times.

*Keep a food diary: this could offer valuable insights into the reasons behind your child's feeding difficulties*

It is important to identify if possible, from the food diary, whether it is the volume or type of food being eaten which is causing your child's dietary issue. There are a number of factors that could be contributing to this, including:

- **Physical:** your child may be over-eating as a result of lack of control and the ability to know when they are full.
- **Obsession:** one feature of ASDs can be obsessive behaviours, so food and the consumption of it may have turned into an obsession or area of special interest, possibly leading to over-eating and weight gain.
- **Coping strategy:** We all have different ways of dealing with stress and difficult periods in our lives. Your child may see food as a source of comfort, or they may be under eating as a way of dealing with stress.
- **Sensory:** Your child may have a particular craving for a specific food, because it may be providing them with sensory satisfaction.
- **Medication side effects:** You may have noticed a change in your child's eating habits since altering their medications, even if the alteration is very slight.

If your child is having problems with eating too much or not enough we encourage you to get a referral to a dietician. They will be able to advise you on techniques to ensure your child is getting the nutrients they need. Your GP should be able to make that referral.

## c) Gastrostomy/Tube Feeding

When eating problems become very severe having a gastrostomy may be an option. A gastrostomy is a surgical procedure for inserting a tube through the abdomen wall and into the stomach. The tube is then used for feeding. Having a gastrostomy can be a really difficult decision to make, as it is very hard psychologically to further medicalise your child. For some patients, it can be incredibly beneficial and impact, in a positive way, on all aspects of life. The procedure must be discussed with the patient's medical team, including the consultant and dietician.

## CASE STUDY: GASTROSTOMY

### Galia Wilson – mum to Arlo

Arlo Wilson is a seven year old boy who was diagnosed with Dravet Syndrome when he was three years of age. He didn't wean well and didn't seem to take to food at all. At nine months old the family saw a Psychologist who thought that all the intrusive procedures he'd had in hospital had traumatised him

Over time he began to eat, but would select only one or two things at a time, and would become fixated on one type of food. Each of these phases would last approximately three months. He would sometimes eat fruit and has always tolerated milk well, so as far as the dieticians were concerned he was having a fairly balanced diet and because he maintained a good weight they weren't concerned.



When Arlo turned four and half he suddenly went off the one food he was eating at the time and could only tolerate milk. Every time he tried to eat something he would retch and often vomit. It was becoming very apparent that Arlo had major sensory issues around food. Sometimes even the smell of food would make him sick. Arlo's dietician prescribed fortified milk, which he tolerated and resulted in him having more energy. After a while though, this milk started making him retch every time he tried to drink it. This was when the doctors first started talking about a gastrostomy. Galia and Oli, Arlo's parents, did not want to go down this route.

His weight was ok and his eating seemed to come in phases so would hopefully come back. Eventually Arlo stopped taking his medication, and this persuaded Galia and Oli that maybe a gastrostomy was the way forwards. He was referred and placed on the waiting list.

While being on the waiting list, Arlo requested a pizza one night by taking it out of the freezer and handing it to his Dad. To Oli's surprise he ate the whole thing. This was a new food phase and he would tolerate the pizza with absolutely anything on it. He started taking his meds orally for the first time ever, rather than in milk as he had done previously. He had so much energy and seemed so well that, when the date for his surgery came through, the family cancelled it.

This phase lasted for nearly three months and then Arlo just stopped eating again. At this point Galia and Oli decided there was no choice but to proceed with the operation. Unfortunately the waiting list was quite long and Arlo didn't eat anything for nearly two months, existing just on cow's milk. He had no energy, looked pale and became very withdrawn. He didn't want to do anything because he didn't have any energy; he became even more wobbly than before and would struggle to walk up the stairs. He also lost a lot of the speech he had, and became lethargic and unengaged.

Eventually Arlo had the gastrostomy as an emergency procedure. Galia said: "we still had mixed feelings about the surgery, but a week has passed and he is tolerating his feeds well. He is a different child. He is happy, giggly and is so excited to do things, like go to the park, which he hasn't wanted to do for ages. It is easy to give him his feeds and especially his medicine." Galia and Oli firmly believe that the gastrostomy has been one of the best decisions they've made for Arlo with regards to his care.



## d) Gait

Gait refers to how a person walks; abnormalities relating to gait are typically associated with a physical disease, condition, or deformity. Any child with a condition that affects their motor and sensory development can develop gait problems. These problems are not necessarily specific to Dravet Syndrome, as they have also been seen in other epilepsy syndromes, but they are strongly associated with it. For example, over time, a child with Dravet Syndrome maybe unable to straighten their knees properly and this will affect how they stand, walk or run.

*Following an individualised physiotherapy programme can be very beneficial to maintaining your child's mobility, however simple things such as walking on different surfaces (e.g. pavement, pebbles, sand), climbing up and down steps and bending and stretching can be just as useful for helping to build strength*

A number of studies have shown that children with Dravet Syndrome typically exhibit progressive deterioration of gait after the age of ten, which can have a significant impact on their mobility and independence. As a rule, children usually start walking at a normal age but develop an unsteady gait as they become older:

There are a number of things you can do to try to manage and improve your child's gait, including specifically designed footwear and strengthening muscle exercises. Speak to your healthcare professional for advice and request referrals to:

- A physiotherapist.
- An orthotics clinic.
- A specialist gait clinic.

## e) Toileting

It is not unusual for patients with Dravet Syndrome to toilet train later than an average child, and depending on the level of learning disability some patients may remain incontinent. If you have access to an occupational therapist they may be able to give you ideas on toilet training, alternatively you should have access to an incontinence service who will also be able to offer advice and experience. The incontinence service also provide and supply nappies and pads for children and adults who are incontinent. You can get referred by your health visitor, community nurse or GP. At the time of going to print the incontinence service provides free nappies and pads to all children and adults aged over 4 years of age.

## 8. SCHOOLING

Every child with Dravet Syndrome will have different educational needs and it is important for you to understand the options available to support your child's learning. This will help you to ensure that you make the right decision for you and your child and that their special educational needs (SEN) are met. This may be in a mainstream school or nursery with extra support, or in a special school.

### a) Your Child's Rights to Additional Support

If you are considering sending your child to a mainstream nursery or school, make sure you are aware of the additional support that you can receive. This will vary from school-to-school so it is important that you research your local schools and arrange a visit to understand what extra support is available at each. All state-funded schools (including nurseries and playgroups) should follow the Special Educational Needs Code of Practice Guidelines [https://www.gov.uk/government/uploads/system/uploads/attachment\\_data/file/398815/SEND\\_Code\\_of\\_Practice\\_January\\_2015.pdf](https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/398815/SEND_Code_of_Practice_January_2015.pdf) which provides information on how schools should assess and meet SEN.

Your child should be allocated a Key Worker or Special Educational Needs Co-ordinator (SENCO) who can help you to work out what additional needs your child might require. This could be a teaching assistant with them in the classroom, a specific method of teaching or additional help outside the nursery or school, such as a speech and language therapist.

### b) Responsibilities of Mainstream Schools

It is important to remember that the Government's education policy is focused on helping all children achieve their potential and have equal opportunities within their learning environment. It is also illegal for any child to be discriminated against due to a disability or special educational needs.

This is reinforced by a law known as The Equality Act 2010 and The Special Educational Needs Code of Practice which plays an important role in ensuring that schools are aware of their role in ensuring this is realised.

The focus of The Special Educational Needs Code of Practice is ensuring that children's special educational needs are identified as quickly as possible and that early action is taken to meet those needs. It also places emphasis on monitoring the progress of children with special educational needs towards identified goals.

### c) Early Years Action and Early Years Action Plus

The Early Years Action plan is offered to children at an early age with additional educational needs. This may be due to them not progressing as expected or due to behavioural, sensory or physical problems they are experiencing.

A Key Worker or Special Educational Needs Co-ordinator (SENCO) will carry out an assessment of your child's needs to decide what support they require and develop an **Individual Education Plan (IEP)**. This will highlight what special help will be provided at school/nursery, what you can do at home, and include targets for your child to achieve.

You should meet regularly with your Key Worker or SENCO to discuss your child's progress and if your child is not making enough progress through the Early Years Action programme, they may be able to receive additional support. This is referred to as Early Years Action Plus.

*Familiarise yourself with the educational guidelines and codes of practice to get the most out of the system for your child*



## d) Education, Health and Care Plan (EHC Plan)

You or your nursery/school can ask the local authority for a formal assessment of your child's special educational needs to help ensure they receive additional support. Based on your application, the local authority will decide whether or not to assess your child or issue an Education Health and Care (EHC) plan, which brings your child's education, health and social care needs into a single, legal document (this document replaces Statements of SEN and Learning Difficulties Assessments for children and young people with special educational needs).

The assessment will be based on conversations between the local authority and a number of people including you and your child, their school or nursery, doctor and an educational psychologist. Once the authority has collected all the information it needs for the assessment, it will decide whether or not an EHC plan is necessary (if they decide not to proceed with a plan, they must inform you within 16 weeks of the initial request).

When drafting the EHC plan, your local authority should work closely with you and your child to make sure the plan takes full account of your views, wishes and feelings. Once the plan has been written, a draft will be sent to you. You will be given 15 days to comment on the draft and you can ask for a meeting to discuss it if you want one. At this stage, you are able to request a specific school, or other setting, that you want your child to attend. Your local authority has 20 weeks from the request for the EHC needs assessment to issue the final plan to you. Once an EHC plan has been finalised, your local authority has to ensure that the special educational support in section F of the plan is provided, and the health service has to ensure the health support in section G is provided. This should help to enable your child to meet the outcomes that you have jointly identified and agreed.

Your local authority has to review your child's EHC plan at least every 12 months. That review has to include working with you and your child and asking you what you think and what you want to happen, and a meeting which you must be invited to. It is also an opportunity to update the plan to assist your child's development and ensure it is relevant for their progression. It is important that you provide an honest account of how you feel your child is progressing and discuss any concerns you have. You can also actively suggest ways you feel this can be improved or any further support you feel would benefit your child.

If your child has an EHC plan you may be entitled to a personal budget, which allows you to have a say in how to spend the money on support for your child. You can use your personal budget in three ways (or as a combination of two or more of these options):

- direct payments made into your account – you buy and manage services yourself
- an arrangement with your local authority or school where they hold the money for you but you still decide how to spend it (sometimes called 'notional arrangements')
- third-party arrangements – you choose someone else to manage the money for you

You have the right to appeal if you disagree with your child's final EHC plan (or if you disagree with the local authority's decision not to provide an EHC plan if this should be the case). Your local Parent Partnership Scheme will be able to guide and support you through the process.

## e) Tips to Help You through Your Child's Educational Journey

- Keep track of your child's learning and progress.
- Understand and follow their schoolwork and encourage additional learning at home.
- Get involved with your child's school and arrange regular meetings to discuss your child's progress.
- Let the school know what's going on at home, improvements or areas you feel need developing.
- Don't be afraid to ask for additional support or make suggestions to improve your child's experience.
- Talk to your child about how they are enjoying school and where possible involve them in decisions.
- Research local support services available through your local authority.
- Discuss any concerns you have with a doctor or health visitor.
- Talk to other parents who have children with learning disabilities.
- Ask the school to provide a home/school diary so that daily information can be shared and continuity of care achieved.

## f) Additional Information

More information on schooling and education can be found through the following organisations:

- Your local authority:  
**<http://local.direct.gov.uk>**
- Directgov: **[www.gov.uk](http://www.gov.uk)**
- Department for Education:  
**[www.education.gov.uk](http://www.education.gov.uk)**
- National Association of Family Information Services:  
**[www.daycaretrust.org.uk/nafis](http://www.daycaretrust.org.uk/nafis)**
- National Parent Partnership Network:  
**[www.parentpartnership.org.uk](http://www.parentpartnership.org.uk)**



## 9. RESPITE

### a) Respite Care

Caring for a child with Dravet Syndrome is a full time job, one that demands a lot of time and energy and can have a huge impact on your life. As such it is vital that you take care of your own health – you won't be any use to your child if you are exhausted, rundown or ill.

A certain degree of stigma often surrounds the term 'Respite Care' but all it really means is that your child is looked after elsewhere, for as little as a couple of hours or as long as overnight – to give you some alone time, to rest, relax and recharge your batteries. This might sound like a small undertaking, but it can have a huge impact on both your wellbeing and the wellbeing of those around you. Respite will give you the opportunity to do the things that other people take for granted, like spending time with other children, catching up on sleep or even just having time to stop and think.

*A break is not a luxury but essential to maintain the caring role*

### b) Residential Respite Care

Anyone who undertakes a caring role may be eligible for respite care services, such as day respite, which can be provided to offer short periods of help in a range of different ways, either in the home or in day centres. We really encourage all parents of children with Dravet Syndrome to explore the respite options available to them, it may well be that you don't feel it would work for your particular situation, but it is definitely worth at least thinking about – we all need a little help from time-to-time!

### c) Emergency Respite Care

While residential respite care is usually planned ahead of time, emergency respite care is arranged as and when the need arises. This could be because you just feel as though you need a break, or because your situation has changed and you need to be somewhere or need to be there for someone else. Emergency respite could offer the short notice support you need. Contact your local authority for details on respite care in your area and remember that asking for help is not failure.

### d) Hospice Care

It can be very difficult for families to accept that a hospice may be the right place to provide respite, particularly as often hospices are associated with end of life care. Although hospices do provide end of life care, they also provide regular respite and can help with day care and respite within the home. It is worth seeing if you have a hospice local to you and going along for a visit. You will find that children's hospices in particular are places where children have lots of fun and are given many opportunities. See the resources section for the Together for Short Lives website details (page 62).

### e) Accessing Respite

To access respite you can either self-refer to your local disability team or be referred by a professional. Initially you will have to be assessed, this can seem very intrusive but remember social services are there to help you maintain your role as a parent/carer. The outcome may be services being provided directly through social services or you may be offered direct payments, whereby you can employ your own respite carers. You are also entitled to undertake a carer's assessment for yourself, which may help the social worker build up a better picture of your family circumstances. If you are at all unsure how to access any support or respite care speak to your GP, health visitor or community nurse; all of whom should be able to refer you.

## CASE STUDY: RESPITE

### *Rita Mills – mum to Ruby*

Ruby is now 13 years old and was diagnosed with Dravet Syndrome when she was nine months old. Ruby's behaviour was difficult to manage and, coupled with her severe learning difficulties and the arrival of a new baby when Ruby was three years old, Ruby's condition has had a huge and challenging impact on family life.

When Ruby was three years old, her mother Rita felt like she was at breaking point. With a new baby and her husband, Justin, working overseas three to four months of the year, Social Services awarded her 19 nights respite. A respite centre was available locally where a child can stay overnight. However, Rita and Justin were uncomfortable with that arrangement, as Ruby was still very young. Instead, Social Services found a 'link-family' that would provide respite for 19 nights a year.

They met with four families, each of which required three to six months of build-up meetings, gradually leading up to the first overnight. The first two families were not able to cope with Ruby, and unfortunately the third and fourth families retired from providing respite after 12 months each. Ruby's family found the whole experience disjointed and inconsistent.

In the meantime, the Shooting Star Hospice awarded the family two weeks of respite a year. This was a wonderful, safe environment which Ruby enjoyed and a place the family could stay if they felt the need to.

As the 'link-family' programme had also concluded, the family was put on their local councils' Direct Payments' scheme. They have now been on the scheme for seven years and are currently receiving 12 hours respite per week.

With this arrangement, they are able to choose their own carers and pay for them directly using the money from the council.

They also have enough money left over for other activities such as Ruby's weekly horse riding for the disabled, a gym membership for her and a carer, music lessons, trips out and occasionally for a piece of equipment, like a bike. They also use the respite money to pay for family activities that are specially adapted so that Ruby can join in, such as Surfing for the Disabled and Abled in Cornwall.

Direct Payments have also allowed the family to have a carer to look after Ruby when they need time-out to do 'ordinary things', such as taking their youngest child out with her friends, or having a peaceful meal without worrying about Ruby's behaviour or the possibility of a seizure. Rita said: "the Direct Payments respite programme works very well for our family, as it gives us control and allows us to spend the money in ways that best help Ruby."



## 10. RESIDENTIAL CARE

### a) Understanding Residential Care

Residential Care is a living arrangement, in which your child resides outside of the family home, in a facility specially equipped to meet their needs and requirements. This could be in a care home, a nursing home, a residential school/college or a hospice. It is quite common for many parents to initially dismiss the idea of placing their child in a home; however, it is important to realise that residential care centres have changed a lot over the years and truly could offer your child the support they need on a daily basis.

Choosing between residential care and independent living can be very hard, which is why we would encourage you to talk about your available options and the impact of making this decision with people who have been in the same situation. Scope and Carers UK both provide internet forums, offering peer-to-peer support as well as information services about local support networks.

Visiting different types of residential homes and talking to their residents can really help to dispel any preconceptions you may have and many offer trial periods so you can see what it's like before you commit to anything.

### b) Funding

Each Local Authority funds respite and residential care differently. You may be asked to fund respite care yourself through your child's Support Package or you may have to make a separate application.

If your Local Authority is unable to fund your care you may still be able to apply for help from a charity or fund. Look online for organisations that could meet your needs or ask your social worker, GP, health visitor or local carers support group to give you more information. Some respite and residential care can also be arranged privately.

NHS Choices has clear information about different types of care:

**<http://www.nhs.uk/carersdirect/Pages/CarersDirectHome.aspx>**

Care Home is a recommendation site with information about different homes in your area with testimonials from residents: **[http://www.carehome.co.uk/care\\_search.cfm](http://www.carehome.co.uk/care_search.cfm)**

As part of Young Epilepsy, St Piers College is a warm, friendly and unique residential and day college for students aged 19–25 with epilepsy, autism, and other neurological conditions or special needs.

A number of young people with Dravet Syndrome have attended St Piers College during recent years. Find out more at :

**<http://college.youngepilepsy.org.uk/about-st-piers-college/>**

## CASE STUDY: RESIDENTIAL CARE

### Angie Lay – mum to Katie Lay

Katie Lay is 33 years old and received a diagnosis of Dravet Syndrome when she was 30. When Katie was younger life at home had become very difficult for the Lay family, putting a strain on their relationships and making it difficult to give their son, who had his own health issues, the attention he needed. As neither Angie nor Dave drove, going out with Katie proved difficult. Accessing information was hard as internet, Facebook and emails weren't around and the family were not aware of any support groups. The Lay family felt very isolated at times and struggled to lead a 'normal' life.

Making the decision to send Katie to a special needs residential school was the hardest decision that her parents ever had to make. There was a lot of soul searching. Were they making the right decision? Was it in Katie's best interests? How were they going to tell people and what would they think of them? But once they'd made that decision – and knew that family, friends and healthcare professionals were supportive – it helped them enormously.

When the possibility of residential schooling was first discussed, the family's Social Worker arranged for Katie to go to visit a special needs respite home for a three week stay, to see what difference it would make at home. Fortunately, Katie really enjoyed her time there. Although the family missed her terribly, life was a lot easier at home: they had more time to spend with their son and their relationship improved; they found they weren't stressed and tired all the time, and they felt less trapped by their situation. At the end of Katie's respite time everyone agreed that this could work for both Katie and the entire family.

The family visited three schools before deciding on which one was right for Katie. Numerous reports were gathered together to enable a decision to be made by the local authority. Once Katie started her residential school, life at home for the Lay family changed dramatically. Initially, there was an adjustment period, where the family experienced feelings of guilt. It also took Katie around six months to fully settle into her new home and school. It helped that Katie was surrounded by her familiar possessions, toys, family photographs; also she bonded with a couple of her carers really well. In the meantime, Angie passed her driving test so the family could visit at weekends.



When Katie turned 18, another transition process began, and the Lay family started to look for a suitable residential home for adults. Again the Social Worker helped with searching for a home and compiling reports. Regular phone calls and letters to the all relevant organisations kept communication flowing and ensured that everyone knew what was going on. Katie was granted funding for full-time care at an adult hospice.

Katie has been at the adult hospice for 12 years now. The family admits that there have been ups and downs: there have been occasions when they were unhappy about certain things, but they have always found ways to address concerns with staff and get any problems resolved.

Angie said: "we can look back on it now and know that it was the right decision for all of us. Katie has a life and a home for life. Importantly, it fulfils her medical needs with 24/7 nursing staff on-site; she is happy and settled. Knowing that she is safe and well-cared for means we can also live our life – after all you do only get one life."



## 11. BENEFITS

### a) Disability-related Financial Support

There is a range of disability-related financial support available from the Government, including benefits, tax credits, payments, grants and concessions. The system can be very complicated and confusing, even to experts; however you are strongly encouraged to persevere and claim what you are entitled to.

**You should not feel embarrassed or ashamed about asking for financial help. Caring for someone with Dravet Syndrome incurs extra costs; you and your family are entitled to receive as much of this support as possible.**

The main benefits which you need to be aware of are as follows:

- Disability Living Allowance or Personal Independence Payment.
- Carer's Allowance.
- Employment and Support Allowance.
- Disabled Facilities grants.

Help to navigate the benefits system is available through

- Carers UK AdviceLine (0808 808 7777)
- Your local Citizens Advice Bureau (CAB)
- At **[www.gov.uk/benefits-adviser](http://www.gov.uk/benefits-adviser)**

### b) Disability Living Allowance (DLA) or Personal Independence Payment (PIP)

#### What is it?

Disability Living Allowance (DLA) was replaced in 2013 by a Personal Independence Payment (PIP). PIP is a weekly, tax-free benefit for people under 65, who have a disability and need help with the extra costs caused by a disability. It is made up of a care component and mobility component.

#### Am I entitled?

PIP is paid at different rates depending on the level of help you need. You will need to complete an assessment to find out how much support you are entitled to. PIP is not means tested.

If you already have an existing Disability Living Allowance (DLA) claim, you can use an online PIP checker (**<https://www.gov.uk/pip-checker>**) to find out how your DLA is affected by PIP.

Note that other benefits you receive may increase if you start receiving a PIP.

#### How do I apply?

Call the Department for Work and Pensions (DWP) to make a new Personal Independence Payment (PIP) claim if you're in Great Britain. (The process is different for claims in Northern Ireland).

DWP – Personal Independence Payment claims  
Telephone: 0800 917 2222  
Textphone: 0800 917 7777  
Monday to Friday, 8am to 6pm

You'll be sent a 'How your condition affects you' form. It comes with notes to help you fill it in. Return the form to DWP – the address is on the form. You must use the PIP application form DWP send you – don't use any Disability Living Allowance (DLA) forms you may have. You'll need an assessment to complete your application.

*The DLA has been replaced by a Personal Independence Payment (PIP) for people who are aged 16 to 64*

## c) Carer's Allowance

### What is it?

A weekly Carer's Allowance is available to support someone looking after a person with substantial caring needs

Carer's Allowance is taxable and can either increase or decrease other benefits you receive:

- If you receive Housing Benefit or Council Tax Benefit you may get extra towards these (called a 'carer premium').
- If you receive the following benefits you will still receive Carer's Allowance, but these benefits are then reduced by that amount:
  - Income Support
  - Income-based Jobseeker's Allowance
  - Pension Credit
  - Universal Credit.

### Am I entitled?

The carer needs to be aged 16 or over and spend at least 35 hours a week caring for a person; however, they do not have to be related to, or live with, the person they care for. If someone else looks after the same person, only one of you can receive Carer's Allowance.

The person being cared for must receive one of these benefits:

- Attendance Allowance.
- Disability Living Allowance (DLA) – the middle or highest care rate
- Personal Independence Payment (PIP) daily living component
- Constant Attendance Allowance at or above the normal maximum rate.

You may not get Carer's Allowance if:

- You are in full-time education, and studying for 21 hours a week or more (this includes supervised study and things like coursework and experiments, not just time spent with a tutor).
- You earn more than £110 a week (after tax and not including a pension).
- You receive another benefit from a pre-defined list (available at [www.gov.uk/carers-allowance/eligibility](http://www.gov.uk/carers-allowance/eligibility)).

### How do I apply?

Go to [www.gov.uk/carers-allowance/how-to-claim](http://www.gov.uk/carers-allowance/how-to-claim) to either apply online or download a DS700 form (or DS700SP form if you get a State Pension) and submit via post.

*If you were entitled to Carer's Allowance before claiming it, you can ask for it to be backdated for up to three months*



## d) Employment and Support Allowance (ESA)

### What is it?

Employment and Support Allowance (ESA) offers people financial support if they are unable to work, or personalised help so that they can work if they are able to.

ESA can be applied for if you are employed, self-employed, unemployed or a student receiving Personal Independence Payments. How much you receive depends on your circumstances (e.g. savings and previous National Insurance contributions).

### Am I entitled?

You may get Employment and Support Allowance (ESA) if your illness or disability affects your ability to work and if you are:

- Under State Pension age.
- Not receiving Statutory Sick Pay or Statutory Maternity Pay and you have not gone back to work.
- Not receiving Jobseeker's Allowance.

You may still get ESA if you have lived or work abroad and paid enough National Insurance (UK or equivalent).

### How do I apply?

The quickest way to apply for ESA is by phone (call 0800 055 6688). Alternatively, you can also send an ESA1 form to your local Jobcentre Plus office.

## e) Disabled Facilities Grants

### What is it?

A person with disabilities is entitled to a Disabled Facilities Grant from the council if they need to make changes to their home, e.g. improving wheelchair access, installing a stair lift or downstairs bathroom.

How much you receive depends on your household income and any savings over £6,000, and can range from £25,000–£36,000 depending on the region of the UK that you live in.

### Am I entitled?

In order to receive the grant, you or someone living in your property must be registered as disabled. Either you or the person you are applying for must:

- Own the property or be a tenant.
- Intend to live in the property during the grant period (which is currently five years).

The council needs to be happy that the work is necessary and appropriate to meet the disabled person's needs, as well as achievable in the property you are living in (e.g. age and condition of the property).

The grants are different for disabled children under 18, so you will need to speak to your local council to find out exactly what financial support you can receive.

### I've heard about the 'benefits cap'. How might this affect our family?

The benefits cap is a limit on the total amount of benefit that most people aged 16 to 64 can receive. It may apply to your household if you receive certain benefits, such as Carer's Allowance, Child Tax Credit, Housing Benefit and Income Support. However, some people are exempt from the Benefit Cap. This means their benefit isn't capped, even if their benefit income is above the limit of the cap.

**Importantly, people receiving DLA/PIP or Attendance Allowance will not be affected.**

You can find out further information at [www.gov.uk/benefit-cap](http://www.gov.uk/benefit-cap).

### How do I apply?

You will need to find out what changes are needed to your property and how much it will cost (you will need to provide two estimates). An occupational therapist can look at your circumstances and recommend the type of changes needed.

To get an application form, contact the housing or environment health department of your local council.

*You might not get any grant if you start work on your property before the council approves your application. Applications can take up to six months to approve*



## 12) TRANSITION INTO ADULT SERVICES

### a) Preparing for Transition

Transitioning through to adult services when a young person with Dravet Syndrome reaches the ages of 16–18 can be a very unsettling process for many families, as it brings significant medical, financial and legal changes in the organisation of care for your child. All these changes can make transitioning into adult services feel very uneasy and uncomfortable. There is currently limited help and guidance available to support families during this period – and it is not always easy to access and find. Being prepared, and arming yourself with as much information as possible, will help you to navigate through this challenging time.

*Importantly, parents and guardians need to apply to the Court of Protection for an official Deputyship in order to continue to be recognised as the legal guardian and primary decision-maker for the care for their young adult with Dravet Syndrome*

You can apply to become someone's deputy if they 'lack mental capacity'. As a deputy, you'll be authorised by the Court of Protection to make decisions on their behalf, including decisions about their personal welfare, e.g. medical treatment, how they are looked after; what schools and educational centres they attend, etc.

If deputyship is not obtained, legal responsibility for the young person with Dravet Syndrome will revert to the local authority when they legally transition into adulthood at the age of 18. This does not mean that, as a parent/guardian, you will no longer be expected to care for your child. BUT it does mean that you will no longer have sole responsibility for decisions regarding their personal welfare.

Transition also ushers in significant changes to the medical care of the young person with Dravet Syndrome. For example, after having years of working with the same medical team you they will move onto a new, adult-services team, who perhaps have less knowledge of Dravet Syndrome than the paediatric team you were used to working with. Hospital appointments or A&E visits can become more challenging as they will be based in an adult hospital or unit that may not have the facilities that are appropriate to your young adult's developmental stage. Hospital stays also become problematic as there is nowhere for a parent or carer to stay overnight on an adult ward.

The following two case studies provide insights and advice based on two very different and equally challenging experiences of coping with transition.

## CASE STUDY: TRANSITION INTO ADULTHOOD

### Teresa – mum to Amy

For Teresa, the changes that came as her daughter Amy turned 18 were a complete shock. "A week before Amy's 18th birthday we took her for some blood tests," said Teresa. "When I called for the results a week later, I was told I couldn't receive them without Amy's permission." This was Teresa's first indication that everything was about to change, bringing a whole new set of challenges into their family life. "For all of Amy's life I had made every single decision for her, whether I wanted to or not," she continued. "Every aspect of her care was down to me. And then suddenly I was told that I couldn't make those decisions by myself any longer. On the other hand I was still expected to care for Amy because she needs supervision 24 hours a day. It was such a horrible, strange feeling."

Dealing with Amy's transition into adulthood was one of the toughest times that the family experienced. "Nobody tells you what to expect," added Teresa. "We had the least amount of help, when we needed it most. Emotionally, transition was very stressful and also very hard to get our heads around because in reality Amy's condition had not changed. She was still the child we had cared for all her life, yet in the eyes of the law she became an adult."

Two years later on, Amy is 20 and doing well. She's living at home and four days a week she goes to an independent living group for 16–5 year olds, called 'Fair Play', which is paid for using Amy's Direct Payments. "Amy loves it," said Teresa. "It's a fantastic, amazing place – the only place where I've felt 100% happy to leave her." Yet caring for an adult with Dravet Syndrome continues to be challenging. Teresa has not yet applied for deputyship and shares responsibility for caring for Amy with her medical consultant and a social worker.



"It's still really difficult to know who is responsible for making decisions," admitted Teresa. For example, if Amy needed an operation, a 'best interests meeting' would be held with Teresa, Amy's doctor and social worker. Teresa's opinion would be listened to but the consultant or social worker could override it.

The financial impact of transition into adult services is another issue that the family continues to struggle with. "As a family with responsibility for looking after Amy, we only had one full-time salary, so we were entitled to Child Tax Credits with a disability element," explained Teresa.



Child Tax Credit can be claimed if the child is in full time education up until the age of 20. "Nobody told us if Amy was entitled to any benefit in her own right as an adult, but we later found out that she is entitled to Employment Allowance", Teresa continued. "These benefits were not nearly as much as Child Tax Credits, yet the expenditure on Amy's care remained just as high."

Due to these changes in income, Teresa's only option was to go back to work. But since leaving college, Amy is at home more than she has ever been before. "We are currently fighting for Direct Payments to be increased to cover Amy's care whilst I'm working," she added. "Yet even the increased Direct Payments don't cover things like the cost of fuel for taking Amy to 'Fair Play' or the cost of activities at the centre."

Looking to the future, Teresa plans to apply for deputyship, but knows it could be a long road ahead. She believes there should be more awareness and information about the impact of transition. "I would say to any parent of a child with Dravet Syndrome – start looking into transition as soon as possible, around the age of 14, look into every aspect of transition and start looking at deputyship. Be as prepared as you can possibly be."

## CASE STUDY: TRANSITION INTO ADULTHOOD

### Emma – mum to Matthew



Emma is mum to Matthew, a young adult with Dravet Syndrome, and Alice. Matthew is now 21 years old, but due to the brain damage associated with many years of his early years with seizures and ineffective medication, he has a developmental age of around 12 to 15 months. Emma started planning for Matthew's transition into adult services when he reached the age of 16. Although Emma eventually achieved Matthew's plan in full, the transition process was anything but easy, and she needed to fight every step of the way.

Matthew's transition plan including four main goals: remain living at home with his family, deputyship for Emma, a suitable day centre and special needs college that provided all the therapies Matthew needed to keep him healthy, happy and well-cared for, and extra care within the home via Direct Payments. Achieving each of these goals brought different challenges.

"Getting agreement for Matthew to remain living at home with his family was relatively straightforward," said Emma. "Everyone involved agreed that this was in his best interests. It is also the most cost effective option for the local authority as equivalent full-time residential care would work out at approximately £3,000 per week."

Becoming Matthew's Deputy was more complicated. Emma needed to apply to the Court of Protection and prove that she was the best person to advocate for Matthew and represent his interest. "The forms are unbelievable and a minefield to work through," she added. "You can employ solicitors to do it for you but it's really expensive. I did it myself and it was a lot of work – downloading and completing forms, copying papers, sending questions and answers to and from the court. However; it is absolutely worth it for peace of mind, the thoughts of responsibility for my son being with the local authority and not me once he turned 18 was just too hideous for me to even contemplate."

The next challenge was to find suitable day placements for Matthew which included a special needs college for term time and a day centre for the holidays. Emma was unhappy with the choices of special needs colleges recommended by social services and all local day centres were shutting down. She investigated options herself, including a special needs day centre that was outside her county but only a 45-minute drive from home. "We needed a college and a day centre that could keep Matthew healthy and mobile as well as cared for; stimulated and happy," she explained. "The places I found were not on the recommended lists by social services, although they were known to them. I stuck to my guns and listed out all the reasons why these were the most suitable option, such as the positive benefits to Matthew's quality of life, which is recognised by social services as an important consideration."



Last but not least, Emma needed to secure extra care within the home through Direct Payments, which meant working out a Personal Budget that would allow her to manage and pay for the extra care. "Never underestimate what you need in terms of extra care at home as the budget you apply for will be cut", she advised. "When you list out the hours that you need, include going to work (it is a parent's human right to go to work once a child reaches 18), holidays, sick days and overnights. I included two full nights' sleep per week plus at least one weekend per month and six weeks full weeks of cover per year to allow for holidays and my work commitments which take me away from home."

In putting Matthew's transition plan in place, Emma and the family went through a number of assessments, including Health Care, Social Care, Carers' and Young Carer's assessments, in order to determine the level of support needed and which part of the local authority should fund this. "Be prepared to be asked the same things time and time again", she said. "There's no getting around it. Again, doing your research helps. Read their pamphlets and quote these back at them, emphasizing that all decisions need to be in the best interests of the young adult."

Eventually the family was assigned an excellent Social Worker who was very helpful. "We went to the panel with our plan of what we needed", continued Emma. "We made our case and got everything we asked for, with payments funded by Social Care."

Every family's experience of transition will be different, because every young adult with Dravet Syndrome is different. For parents who may be about to go through transition, Emma noted, "The best advice I can give is to invest plenty of time in your research. Talk to other parents and professionals, but don't rely on advice – find out for yourself and make your own plan. Make a list of what you need and cost everything out. The cost of residential care is a useful comparator because this is much more expensive than home care. You'll need to be prepared to have a lot of arguments on the way but stick to your plan. If you know your stuff you can get what you and your family needs."

## b) Guidance and Further Information on Transition

Whatever happens, you won't be the first family to ever face these challenges. Parents who have been through transition are often an excellent source of advice and support. In addition, there are a number also useful websites that provide detailed guidance on what to expect during transition, as well as links for downloading relevant forms and information for a deputyship application.

- Gov.uk site: <https://www.gov.uk/become-deputy/overview>
- Court of protection application forms and guidance: [http://hmctsformfinder.justice.gov.uk/HMCTS/GetForms.do?court\\_forms\\_category=court\\_of\\_protection](http://hmctsformfinder.justice.gov.uk/HMCTS/GetForms.do?court_forms_category=court_of_protection)
- Working Families website guide to transition: <http://www.workingfamilies.org.uk/publications/from-child-to-adult-a-guide-to-disability-transition-and-family-financesupdated-2014/>
- Mencap website – factsheets on transition: [https://www.mencap.org.uk/search/apachesolr\\_search/transition](https://www.mencap.org.uk/search/apachesolr_search/transition)
- Southampton Children's Hospital Ready Steady Go programme – a useful example of transition of care from children to adult services which could be applied in other hospitals in the UK: <http://www.uhs.nhs.uk/OurServices/Childhealth/TransitiontoadultcareReadySteadyGo/Transitiontoadultcare.aspx>

## c) When to Start Preparing for Transition

To ensure that you are as prepared as possible, it's a good idea to start researching transition and talking with your care well before your child turns 18. When they are 14–15 years old consider raising the topic at your annual EHC plan review and with your medical team. If you wish to apply for deputyship, start looking into this when your child is 15–16 years old, as may be a time-consuming and complicated process.

## d) 16-Plus Assistance Fund from Dravet Syndrome UK

### What is it?

Intended to fill the gap in support for adults with Dravet Syndrome, the 16-Plus Assistance Fund is available from Dravet Syndrome UK to pay for anything that will improve the life of an adult with Dravet. The limit on each application is £1,000 per family per year.

### Am I entitled?

The fund is aimed at families who include a person with Dravet aged 16 and over. Each individual case is assessed for whether the needs are being met elsewhere. The applications are not means tested, but a letter of diagnosis is required as proof of the condition.

Any items of equipment or assistance that will improve the life of the adult with Dravet will be considered. Holidays will not be funded, but contributions towards the cost of a holiday may be considered if the trip can be shown to benefit the life of the applicant.

### How do I apply?

An application form and guidelines can be downloaded from [www.dravet.org.uk/family-support/](http://www.dravet.org.uk/family-support/).

*Sadly many funding opportunities cease when children transition into adult services, yet the equipment and assistance required remain the same. This is where the Dravet Syndrome UK 16-Plus Assistance Fund can step in to help*



## CASE STUDY: 16-PLUS ASSISTANCE FUND

### *Amanda McLeish – mum to Amanda*



Amanda will be 24 years old this year: When she left school her parents learnt that there was very little charitable help around for toys, equipment or holidays for young adults with Dravet Syndrome.

Dravet Syndrome UK set up the 16-Plus Assistance Fund in 2012 and to provide financial support for those 16 years and over with Dravet Syndrome.

When Amanda's parents first saw an iPad they knew it would be a great educational and fun tool for Amanda, helping her to learn new skills and maintain the ones she already had. Amanda's parent's applied to Dravet Syndrome UK for an iPad for Amanda. The application was very straight forward; the main element was to be able to show confirmation of a Dravet Syndrome diagnosis. The form has a few questions about what you need, what benefits it will bring for the person and an estimated cost. Her mum said: "I was very impressed by the whole process as it only took a few weeks from start to finish, and Amanda is really benefitting from the new technology. It continues to be the best way for her to learn with the benefits of increased concentration and hand eye co-ordination."

Amanda's parents used to have to take all her toys with them in a big bag when they went out, but it is so much easier with her iPad as it accommodates everything she enjoys through the vast amount of apps and videos geared towards her learning ability.

"We are very grateful to Dravet Syndrome UK for taking on-board one of the many problems faced by parents when our children grow-up and become adults", added Amanda's mum. "Through the 16-Plus Assistance Fund my daughter has been given the opportunity to experience up-to-date play and learning in the same way as others."

### 13) FAMILY SURVIVAL

This section aims to provide a more 'hands on, personal' approach to how the family survives the ups and downs of living with a Dravet Syndrome diagnosis. Having a family member with Dravet Syndrome does affect the entire family and can have a huge impact on many areas of life that most families take for granted. This final section of the book will provide you with some tips, ideas and hopefully help you realise you are not alone on your journey.

#### a) What is survival?

*“Survival is becoming an expert in putting a face on for all – saying all is fine when it isn’t, smiling when you feel like crying. Biting your lip when well-meaning people tell you all will be fine and your child will grow out of it! Survival is not being afraid to question or challenge any professional in social, health, education who thinks they know your child better than you. Survival is surrounding yourself with advocates, true friends and family – basically people who really understand the daily stresses, challenges and battles you are up against, and who can help you when you need it most.”*  
(Mum to a seven year old Dravet patient)

#### b) Relationships

When any child comes along relationships are bound to change but when you are dealing with seizures, alongside any of the other issues Dravet Syndrome presents, relationships can alter dramatically.

##### Relationships between Mum and Dad

All of a sudden you are not just Mum and Dad, you are care givers, nurses, teachers and advocates, among many other things. You may find you and your partner just slip into these roles, agreeing about the path you wish to take with regards to treatment. However, you may find that you disagree, perhaps one partner takes longer to come to terms with the diagnosis than the other; or you disagree about medications.

The important thing to remember is there is no right or wrong. Having a child with Dravet Syndrome can be stressful and it can cause tension. Try and have some time away – some planned respite so you can go and be yourselves again. It is important to remember the couple that you are.

*“It’s been hard staying together; we are more like two friends living in the same house, but respite is starting to help us find our marriage again.”*  
(Mum to a six year old Dravet patient)

You may, however, find your relationship becomes stronger:

*“No-one expects to have a severely disabled child when they get pregnant, and it can be very hard on the parents’ relationships especially when faced with a lifetime of care-giving. We found that ours has got stronger and we have learned more about each other and built more trust together through the extreme situation in which we found ourselves.”*  
(Mum to a five year old Dravet patient)

Being the parents of a child/adult with Dravet Syndrome presents many challenges and in times of stress and tension, it is very easy to take it out on those closest. Try to be as open and honest and you can with one another: share your fears and feelings as you will probably find you are thinking along the same lines. Some people find talking to counsellors helpful as a way of sharing how they feel. Speak to your GP or social worker about being referred to a counsellor.



## Relationships with Grandparents

Being a grandparent of a child with Dravet Syndrome can present some very different challenges. Not only are grandparents concerned for their grandchild, they also see their own child suffering – and perhaps struggling. Quite often there is not an awful lot they can do to help. Grandparents can feel helpless, particularly if they do not feel confident enough to look after their grandchild, and it is uncommon for there to be support available to grandparents for them to share their feelings and concerns with others in similar situations.

***“I expected my parents to be there for me throughout, after all they are there to look after me, protect me and make everything better. I never once considered how they might be suffering and I wish I had taken the time to realise that rather than being angry that they weren’t there for me.”***  
(Mum to an eight year old Dravet patient)

It is good to get grandparents involved. Maybe they could accompany you to an appointment to get a better understanding of Dravet Syndrome. If they feel uncomfortable looking after their grandchild alone maybe they could come round to play with your child whilst you go upstairs for a bath, or a much needed nap. This way if anything happens you are there; their confidence will grow and the relationship between your child and their grandparent will also strengthen.

## Relationships between friends

Having a child diagnosed with Dravet Syndrome makes you realise very quickly who your real friends are, the friends who will be there for you no matter what, no matter how many times you cancel meeting up, no matter how often your child demonstrates a challenging behaviour or seizures in their company. Many friends will disappear off the radar as time moves on, which can feel very upsetting and hurtful.

***“I now have 2 types of friends; my ‘normal friends’- people I’ve known for many years who I catch up with and enjoy spending time with and my ‘3am friends’, these are the people who I can call in a crisis at 3am and they will understand, be there for me and drop everything to help. That type of friendship is invaluable and to be treasured.”***  
(Mum to an eight year old Dravet patient)

## c) Siblings

As much as we never want it to happen it can be very easy to sometimes overlook the siblings of a Dravet Syndrome patient. There are times when our Dravet child just has to take priority.

***“I have lost count of the number of times plans have been cancelled due to seizures or illness. One year a family holiday had to be cancelled as our daughter had pneumonia and ended up in hospital on IVs. Her older sister just took it in her stride as always, despite the fact that it just wasn’t fair.”***  
(Mum to an eight year old with Dravet Syndrome)

Siblings of a Dravet patient have to experience many things that you would never wish your child to see: prolonged seizures, ambulance trips, hospital intervention and challenging behaviour; to name just a few.

It would be very easy for siblings to become resentful, but we have found that many families report how siblings take on a more caring role and become very protective towards their brothers and sisters.

***“I’ve never seen such a strong bond between brother and sister; he is like a third parent.”***  
(Mum to a seven year old with Dravet Syndrome)

Siblings tend to understand a great deal more than we initially expect them to. Honesty, at a level appropriate to them, is often the best way forwards. Try to involve the siblings where you can so that they do not become actual carers, but they have some level of involvement. Perhaps let them come along to the occasional appointment, help their sibling with physio exercises or help with bath time.

If you think your child's sibling is struggling there are sibling groups available that you may be able to access. Many local authorities run sibling groups, where they can get together with other children in a similar situation. Although the focus is not on being a sibling of a special needs or disabled child, there are opportunities to partake in activities that may help the siblings understand their situation and share their fears or worries. Many hospices also run sibling activities or have play therapists who are able to help siblings organise their thoughts and feelings. If you feel this is something that would be beneficial speak to your social worker or GP who will be able to refer you to young carers' organisations.

Overall, through speaking with families and through our forum, we have found that siblings tend to see their Dravet sibling just like any other child sees their sibling. They do not notice the differences between them and other children because that is their brother or sister; they are who they are and that is that. Children are very resilient, way more resilient than adults, and tend to just take things in their stride.

The upcoming case study by Mark Lay, an adult sibling of a Dravet patient, is about growing up with a sister with Dravet Syndrome.

As Mark points out in his case study the siblings of disabled children just have a different 'normal', which has the potential to shape their lives. Openness and honesty within a supportive environment can only be a positive experience for the entire family.

The organisation 'Sibs' provides useful information and support for siblings of disabled children and for parents too. See the resource section for contact details (page 61).

*Siblings may be reluctant to talk about their feelings or join in with sibling groups initially but these services are there to help and can be hugely beneficial in helping siblings to understand their feelings and the knowledge that they are not alone*



## CASE STUDY: SIBLING

### *Rachel Spray – mum to Jessie Spray*

The Spray family found that having Jessie, who started having seizures at 10 months of age, was taking up a lot of time and energy and they found it hard to give their son, Thomas, the time and attention he needed; even for simple things like taking him to clubs, teaching him to swim or even ride a bike.

Tom was five years old when his sister Jessie started presenting with seizures. He adapted well to the situation, sometimes waiting at the door for an ambulance or fetching medications for Jessie. As he got older Rachel and Dan found him to be of even more help, and Rachel describes him as 'our hero'. However, the family came to a point where he no longer wanted to go to friend's houses for sleepovers and became very anxious about leaving his parents and Jessie. The family knew they had to do something to build Tom's confidence so Rachel spoke to their social worker, who referred Tom onto Young Carers.



He was accepted straight away, but unfortunately the group had just lost their funding. Rachel decided to go and speak to Tom's head teacher at school. She told Rachel about the Multi Agency Team (MAT) and after a very easy assessment Tom was accepted. Tom has a youth worker and he goes out on day trips in the school holidays – bowling, to the cinema and other fun activities. Rachel said: "Tom is still nervous but I always notice he is full of confidence and buzzing when he gets home."

The Spray family have also accessed counselling for Tom. He unfortunately started having trouble sleeping and experienced a few panic attacks. Tom was referred through the MAT once again. Rachel and Dan firmly believe that talking would really help Tom to overcome his worries, but whenever they try to speak with him he says that he knows they have Jessie to worry about and he does not want to worry them any further. Through Tom's youth worker the family are about to start on a new kind of counselling called 'irelate'. This is an online service accessed from home. He is also starting a scheme called 'sports and arts mentoring'.

This is where a young counsellor will come and spend time with Tom doing his favourite things and getting him out and about.

Rachel said it makes them sad that they can't always help him themselves and it's heart-breaking that Tom feels the way he does.

But they are immensely proud of him and know that he loves his sister with all his heart.

## CASE STUDY: ADULT SIBLING

### Mark Lay – brother to Katie Lay

Being asked to reflect on Katie and the effect it had on me as a child seems a straightforward and normal question if you're looking from the outside of our family unit, but for me it was slightly strange. Why would I be asked to reflect on what was to me 'normal'? One of the most wonderful things of being a child is the ability to accept life as it is. I recall hearing a story of a frustrated mother who could never find socks in her family home, so decided to create a sock drawer in the kitchen next to the washing machine; her children grew up thinking this was a normal and acceptable situation and that every family had a sock drawer in the kitchen. My partner grew up in a home with no carpets upstairs because of financial restrictions, and to her this again was a normal and acceptable situation. Similarly for me, growing up in a home with a sister with learning difficulties was a 'normal' and acceptable situation.



I have no memories of any specific things that I would say have affected me negatively; only things that have helped mould me into a person that I hope my parents would be proud of. It's only natural that growing up with Katie, both at home and when she went into full time care, life had to be adjusted to accommodate her needs. I do feel that this gave me certain values, which I have carried into my adult life. I feel I can attribute the successes in my life to these values, such as structure, routine and discipline. Also growing up with Katie has aided the development of my emotional side: it has made me into what I hope is a sensitive, loving and caring person.

The only thing I can recall that ever upset me and made me cry as a child was when Katie had seizures. I would hear her moan, then there were the 'standard' three bangs on the floor if I was upstairs with her; and mum and

dad would be up within seconds. For me those few seconds felt like forever! I was always told to keep her safe but not restrict; I can recall holding what limb she was chucking about firmly but allowing her to move safely. The one thing I couldn't be there for was when she had rectal Valium. Even in her seizure she would try and push it away; I couldn't handle that. I think this may have been when she was slightly older.

I can remember the day Katie went into her residential school. We went in a social workers car and the journey seemed to take forever. I can recall sitting in the back of the car with mum, and she was crying as we left. I can still recall the pain in her tears, but as I previously wrote, this was normal for me and I couldn't understand the emotion.

As I got older I think I began to realise who Katie was and what my parents had gone through. I can't remember what age I was, but I can recall having the same dream every night. Katie was running and laughing because she was running away from me (something she often did), but in my dream she was running towards a cliff. I tried to chase her but something was stopping me running. I tried running backwards and sideways but couldn't. I always woke before she got to the edge.

On reflection, I feel that although my childhood had to be adapted, and at some point I may have had to give up things, it has given me so much more back, growing up with Katie and my parent's strong ability to cope. I've taken these influences into my adult life; they have helped inform the upbringing of my son and my three step-children. Too often we assume people with learning difficulties have nothing to contribute to life, but in fact this couldn't be further from the truth.



## d) Recreation

Being the parent of a Dravet patient can become all encompassing. It is very easy to become so involved, and at times overwhelmed, that you simply forget about you and making time for yourself. It is important to think back to life before Dravet and the things you enjoyed doing then. Is there anything stopping you from doing them now? Of course there are times when it's just not possible to get out, or perhaps you are just too tired – but it is essential that you keep sight of who you are. A trip to the cinema or a meal out once a month can make a huge difference to your personal wellbeing. This in turn will reflect on how you are able to cope with the caring responsibilities.

*“My netball and my Zumba classes have kept me sane! If I didn't have somewhere to escape to where I can be me and not carer mummy, and where I can switch off completely just for a short time I would crack up completely. I do find myself feeling down if I miss it for a few weeks. It doesn't matter that I may have been up all night dealing with seizures and am so shattered that the thought of exercise is the last thing I want. To be around adult conversation and be with my friends can give me the lift I need when I am feeling down and it helps me to forget all my responsibilities for just an hour. That is therapy and a half, and survival for me.”*

*(Mum to an eight year old Dravet patient)*

Of course it's not always so easy to make this time for yourself. If you don't already access respite care of some description, maybe now is a good time to consider it. Even getting out for a couple of hours to do something that YOU enjoy will be worthwhile. Initially you may not feel confident and comfortable enough to go out as a couple and you may prefer to go out with friends while your partner stays home. Remember there are no right or wrong rules; just go at your own pace, but do find time for yourself and the things you enjoy.

## e) Holidays

When you are the parent of someone with Dravet Syndrome, holidays can easily become something that you just don't consider; maybe it's too much like hard work!

You are concerned about travelling abroad or that your child may not adapt to change well. You worry that they may not be well enough to travel or you don't feel confident to be too far away from your local hospital and the people who understand and know your child...the list could go on!! As with all the other topics in this section, holidays are a very personal thing and there is no right or wrong answer; your family has to do what is right for them.

*“We abandoned our plans for our first holiday to the Isle of Wight when we found ourselves having a conversation about putting the air ambulance on standby to lift our son to the mainland in case of a status seizure. Our wings have been substantially clipped with holidays, but we still try to get away somewhere within the UK. We endeavour to take a carer with us, ensure proximity to adequate medical facilities and ensure that we're in facilities that are accessible for our son.”*

*(Mum to a five year old Dravet patient)*

Another issue with holidays is the expense. Many families are down to one wage and a holiday is a big expense that not everyone can afford. There are a number of charities that may be able to help with holidays, such as 'React' (who own a number of caravans) – please see resource page for more information (page 62).

Dravet Syndrome UK also organise the annual, family weekend away, which you may wish to consider.

*“We take holidays when we can, though that's not often, so when we go with the charity to Centerparcs it's a godsend. Holidays are about creating special memories so that's what we try to do.”*

*(Mum to an 18 year old Dravet patient)*

For information on the Dravet Weekend away please see our website, [www.dravet.org.uk/family-support/annual-centre-parcs](http://www.dravet.org.uk/family-support/annual-centre-parcs).

If you do decide to go abroad it is very important to ensure you have adequate insurance cover. The Genetic Alliance UK provides some useful information on insurance; please see the resources section for contact details (page 62).

## f) Work

Having a child with Dravet Syndrome has an enormous impact on the working life of the parents. As with all of the topics in this family survival section there is no right or wrong answer when it comes to working. Families have to do what works best for them, as well as what is necessary financially. Some families choose for one parent to give up work and stay home, others decide they have to carry on working either for financial reasons or their own sanity. You need to consider your own needs, as well as those of the rest of the family.

***“When my daughter received her diagnosis it made my husband and I rethink our priorities. It suddenly became more important to stay at home with her than go out to work. To keep my mind occupied I continued studying for my degree. Money was tight at times but it was the right decision for us.”***

*(Mum to an eight year old Dravet patient)*

The important thing to remember if you are considering going back to work is to have good contingency plans and good quality care for your child.

***“This is going to sound ridiculous but I go to work for a break!”***

*(Mum to a seven year old Dravet patient)*

Being at home all day with a child with Dravet Syndrome can be very difficult and stressful. Not only are you dealing with seizures but you may be dealing with behavioural issues, isolation and struggling to manage things yourself. It can become very easy to feel resentful towards your partner who goes out every day, speaks to other adults, has an actual lunch break and gets to drink coffee when it's still hot! On the other hand, your partner would probably give anything to spend some extra time at home.

If you are considering working but unsure of whether it is right for your family you can get advice and information from the Carer's Trust. See resources for contact information.

Alternatively you may decide that giving up work is what will work best for your family.

This can be a very big life change, particularly if you enjoyed your work. You may find being at home is fulfilling, but you may find you miss the challenge and responsibility of work, along with the adult interaction of course. If you do choose to give up work then it will be important to have some structure and routine to your days, find out about local children's groups and play groups. Your children's centre or social worker should be able to signpost you to appropriate groups where you can meet others, helping to reduce the isolation that can be felt with such a big life change.

## g) Support

Support comes in a variety of ways. Some families need face-to-face support: someone to come to their house to talk with, confide in and share their thoughts, feelings and fears with. For others, support can come through groups or forums: meeting up with or talking online to other families whose children have the same diagnosis, or attending events that provide information as well as support. Four parents of children and young adults with Dravet Syndrome have kindly shared their experiences of support networks and mechanisms that have worked for them.

No matter what support mechanism you use, or combination of mechanisms, the important thing is to know you are not alone. There are a number of ways to access support and reduce the isolation that so many families feel. It is important to remember that your need for support may vary at different times in your life. Sometimes we need to surround ourselves in a fully supportive environment. Other times we just need to work things through ourselves. There is no right or wrong way. You must always do what you feel comfortable with, but know that these resources are available to you and your family as and when you need them.

See the resources section for details on the Dravet Syndrome UK online private forum and the support group 'Hope' (page 61).



## CASE STUDY: ONLINE SUPPORT

### *Sarah Johnson – mum to Ollie*

In August 2011 Ollie started having seizures; he was just four months old. The seizures continued on a regular basis, ranging from eight minutes to over an hour. He had regular hospital appointments and various tests including Lumber, puncture, MRI and EEGs, all of which came back fine. In February 2012 Ollie had a genetic test carried out, and just 2 weeks before his first birthday the results came back – positive for a SCN1a missense mutation.

The family had already done a lot of research into Dravet Syndrome, so once they received the diagnosis, they realised the extent of Ollie's condition. However, their own GP hadn't heard of Dravet Syndrome, ambulance crews hadn't heard of it and neither had most of the staff at the hospital.

The family felt very alone in the beginning with so many questions to ask, but no one to answer them. Sarah found the Dravet Syndrome UK website and sent an email to enquire about alarms to alert them to Ollie's seizures, as many of these were nocturnal. She spoke to DSUK, who arranged for the charity to fund an oxygen saturation monitor for Ollie. DSUK also told Sarah about a private parents/family online forum. Sarah soon joined up and couldn't believe how many other people were going through the same as them. They were no longer alone!

Sarah said: "it has opened a new world to us. It is full of amazing people, all knowing exactly how you feel and what our children go through. The forum is great! If you have a question about a different seizure type, or something relating to medications, side



effects, anything really. It is such a nice relief to have other people to talk to about Dravet Syndrome, especially as you can chat at any time of the day or night (many of our wonderful children have trouble sleeping, so there always seems to be someone awake). Even if you can't answer the question asked, it's lovely to be able to offer support to each other and it really is like one big extended family. I know I would be lost without it now. Although we haven't all met, I feel like we have gained lots of new friends."

*Sadly Ollie lost his life to Dravet Syndrome on 27th December 2013. DSUK would like to thank his family for allowing us to continue to share his story.*

## CASE STUDY: MEETING OTHER FAMILIES

### Sue Perry – mum to Tom

Tom was born in December 1993 and had his first seizure when he was five months old. It lasted 40 minutes and he was unconscious for nine hours after it. From that point on Tom had seizures every few weeks; these all needed emergency medication to bring them to an end. Tom's parents were told that he had epilepsy and he was put on medication to control it.

The first question Sue and Adrian asked the consultant that he saw at the local hospital was 'why?' The consultant replied that the family would probably never get any answers, and that they just had to accept it; the Perry family changed hospitals and consultant soon after that. So 14 years later, when Tom was tested for Dravet Syndrome they didn't think too much about it. It was simply another test! When the family finally received the result almost a year later, Sue cried – they finally had the answer to that original question of 'why'. It was genetic and was always going to happen; there was nothing that they could have done to prevent it.

Once the Perry family had the diagnosis, it opened up a whole new world to them. There was support available through Dravet Syndrome UK and suddenly Tom wasn't a 'one-off with lousy epilepsy', as his consultant described it. There was a whole group of families in the same situations and a Facebook page where everyone could stay in contact daily.



When Tom was approximately five and a half he went through a really bad patch for about 18 months. His medication stopped working and each new medication that was tried brought with it new problems. The family was doing emergency dashes up to Guys Hospital in London every few weeks, and then having

to stay there for weeks while they tried to stabilise Tom. Sue said: "It was a very lonely time; I stayed in hospital with Tom while my husband Adrian worked and then came to the hospital in the evenings. The difference that support group would have made! Being able to be in contact with other parents that really understand because either they are going through what you are going through, or have been there before."

Every year Dravet Syndrome UK hosts a family weekend at Center Parcs. Sue said: 'the first year we went was quite surreal as we found there were all these

children behaving the same way as Tom, and they even seemed to look similar. Adrian and I found it quite mind blowing, especially being able to talk face to face with parents who were all experiencing the same things. It has now become a yearly essential in the Perry family calendar. We are hopeful now that we have gone through the worst with Tom. He is now 19 and his seizures seem to have settled into a pattern of one to two a night, with a couple of clear nights a week. Tom is happy in himself and has a good quality of life.



## CASE STUDY: SUPPORT THROUGH THE DRAVET SYNDROME COMMUNITY

### *Kelvin – dad to Iwan*

Iwan is a seven year-old boy. He was diagnosed with Dravet Syndrome at the age of four. Prior to diagnosis, Iwan had all the issues associated with Dravet Syndrome, but his family did not know who to turn to for help and advice. Iwan had his first seizure at eight months, a 50-minute seizure that his dad, Kelvin, arrived home from work to discover. Kelvin explains their journey and the impact that Dravet Syndrome has had on the whole family.

“It is difficult to put into words turning the corner into your street after a long commute and not being able to get your car any further as there is an ambulance sitting in the middle of the road right outside our home. Iwan was having a fit and was not stopping. The feeling you have following an ambulance rushing a loved one to hospital is hard to explain. This was the start of a journey that had us and the medical profession guessing what the problem might be. Or at least my wife and the medical profession guessing, as looking back now I can see that I was in denial. I would not accept that there was anything the matter with Iwan. I was clinging to the diagnosis of febrile convulsions, then atypical febrile convulsion and then childhood epilepsy. I refused to see that there was something majorly wrong.

With Iwan having seizures day and night, Heather, my wife, was beside herself with worry. I would like to think that I was supportive, but in the true male fashion I bottled up or hid my own feelings for fear of making things worse. I was the man, I had to be strong.

My work was very good in giving me time off when needed, although again I do not believe they understood the full facts about what was going on, as I had not really told them. I tended to work harder outside office hours. I am lucky to have some great friends, although there were awkward moments in the pub. They would talk about having a difficult week because their child had a cold, or how well they were doing at school, while we were worrying about so many things – medications, training one-to-one to look after Iwan in nursery, finding a new childminder; the impact of his condition on siblings, just to name a few. But bottling things up increased my isolation.

After we requested and changed Paediatric consultant, we started to investigate and treat Iwan’s unknown condition by trying different anti-epilepsy drugs. Some that were absolutely not for Dravet and we really did see



the impact of that through increased seizure activity! Eventually Heather met a mum at an epilepsy group whose child had very similar symptoms to Iwan. We requested a genetic test and Iwan's consultant immediately agreed. In the two months waiting for the results, we researched Dravet Syndrome. When our consultant confirmed the diagnosis we immediately pushed for what we understood to be – and luckily were – the best medications for Iwan and things began to improve. I think once I had something to label 'it' as, I could accept it, although it was a shock to have my head pulled out of the sand.

Following confirmation of diagnosis, we were accepted with open arms into the Dravet Syndrome UK community and it really has made a huge difference. The Facebook community in the private group is a great source of information, advice and support, and also a place where victories large or small are celebrated. We have been to Center Parcs three times and made some great friends. The Dads' Night Out held there allowed me to go relax with others who know what life is like living with Dravet Syndrome, but not to actually discuss the problems – to be normal!

I have been on the Real Ale Train (between Manchester and Leeds) with a group of Dravet Dads and been involved in fundraising (and social) events including: walking up Ben Nevis (twice), Yorkshire Three Peaks and the National Three Peaks. The Dravet Conference was a great event, being informative and somewhere to meet both old and new friends. The Christmas Winter Wonderland party was a fabulous party for the families.

My journey has been one that follows the stages of grief: denial that there was an issue and isolation of myself through not communicating feelings and believing I needed to be strong; anger at the medical profession; depression at various stages; and finally, acceptance. I love my little boy, I would change everything if I could, but I am happy that I have come out of it a stronger and better person with great pre- and post-Dravet friends and family."



## CASE STUDY: SUPPORT GROUPS

### Scott Liddle – dad to Thomas

When Scott and Sarah Liddle's son Thomas was first diagnosed with epilepsy (a long time before his diagnosis for Dravet Syndrome), the couple felt bewildered, overwhelmed and fearful for the future. They had a lot of friends who had children at the same time Thomas was born who were developing 'normally' and unfortunately Thomas was unable to keep up with his peers! This left the Liddle family feeling very isolated and conscious of the fact that their son was different.

Being part of a support group helped to address the way they were feeling by meeting

other families going through similar experiences and emotions. Support groups are a great way to make friends and share information – whether this is medication, benefits, school or support services. Scott said: "actually talking to another person who can empathise with you can be incredibly up-lifting and make it easier to cope with the stress and worry of having a child with epilepsy."

Support groups are also a 'safe place' where families can relax and in the case of HOPE, the London based support group that Scott opened, children with epilepsy and their siblings can have some fun!



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## 14) RESOURCES

Please note that this is by no means an exhaustive list of resources, and that Dravet Syndrome UK do not endorse any of the services, publications or resources provided by any of the organisations listed below.

### Dravet Syndrome UK:

An independent UK charity (registration number: 1128289) dedicated to improving the lives of children and adults living with Dravet Syndrome and other related genetic sodium channel epilepsies through medical research, education and awareness and appropriate support.  
**[www.dravet.org.uk](http://www.dravet.org.uk)**

### a) Seizures and Practical Help

#### SUDEP Action

SUDEP Action cares for the bereaved and works with families and professionals to make change. Using research and awareness they strive to get the answers that will save lives:

**[www.sudep.org](http://www.sudep.org)**

#### Matthew's Friends

An independent UK charity to support all those involved with the ketogenic diet and other dietary treatments for epilepsy and to help further research and development into the use of Dietary Treatments for Epilepsy:

**[www.matthewsfriends.org](http://www.matthewsfriends.org)**

### b) Support

#### Dravet Syndrome UK Facebook Forum

A private online social networking forum set up and managed by Dravet Syndrome UK to support UK families affected by Dravet Syndrome and other related genetic sodium channel epilepsies. Members need to apply to join and will be verified by a moderator to ensure privacy:

**<https://www.facebook.com/dravetsyndromeuk>**

#### Hope London

London-based epilepsy support group, set up and run by a Dravet Dad, holding monthly activity sessions where children with epilepsy and their siblings can meet each other and have some fun:

**[www.hopeforepilepsylondon.org.uk](http://www.hopeforepilepsylondon.org.uk)**

#### Hope Tameside

Tameside-based epilepsy support group, set up and run by a paediatric epilepsy nurse and parents affected by epilepsy, holding monthly activity sessions where children with epilepsy and their siblings can meet each other and have some fun:

**[www.hopetameside.org.uk](http://www.hopetameside.org.uk)**

#### Sibs

The only UK charity representing the needs of siblings of disabled people:

**[www.sibs.org.uk](http://www.sibs.org.uk)**

### c) Financial Information

#### Carer's Trust

UK based charity with the vision of a world where the role and contribution of unpaid carers is recognised and they have access to the quality support and services they need to live their own lives. Providing information and advice on all aspects of being a carer:

**[www.carers.org](http://www.carers.org)**



## d) Holiday Grants & Insurance

### React

Rapid Effective Assistance for Children with Potentially Terminal illness is a dynamic charity working to improve the quality of life for children with life-limiting illnesses living in financially disadvantaged households throughout the UK:

**[www.reactcharity.org](http://www.reactcharity.org)**

### Genetic Alliance UK

The national charity of over 150 patient organisations supporting all those affected by genetic conditions. Provides an information service, including information on travel insurance:

**[www.geneticalliance.org.uk](http://www.geneticalliance.org.uk)**

## e) General Grants

### Cerebra

A UK charity set up to help improve the lives of children with brain related conditions through research, education and directly supporting the children and their carers:

**[www.cerebra.org.uk](http://www.cerebra.org.uk)**

### Roald Dahl's Marvellous Children's Charity

A UK based charity, focusing on children with neurological conditions, which gives direct assistance to families with grants to help pay for things like expensive equipment or activities, such as weekends away with other children and families experiencing the same illnesses:

**[www.roalddahlcharity.org](http://www.roalddahlcharity.org)**

### Family Fund

The UK's largest provider of grants to low-income families raising disabled and seriously ill children and young people. They can help with essential items such as washing machines, fridges and clothing but can also consider grants for sensory toys, computers and much needed family breaks together:

**[www.familyfund.org.uk](http://www.familyfund.org.uk)**

### Caudwell Children

Charity providing family support services, equipment, treatment and therapies for disabled children and their families across the UK

**[www.caudwellchildren.com](http://www.caudwellchildren.com)**

## f) General

### Joint Epilepsy Council

An umbrella charity providing the representative voice working for the benefit of people affected by epilepsy. The JEC provides a number of resources and publications, which families may find useful:

**[www.jointepilepsycouncil.org.uk](http://www.jointepilepsycouncil.org.uk)**

### Young Epilepsy

Young Epilepsy exists to improve the lives of children and young people with epilepsy to enable them to fulfil their potential and ensure they have the best quality of life. They run a school, further education college and produce publications and resources, which families may find useful:

**[www.youngepilepsy.org.uk](http://www.youngepilepsy.org.uk)**

### Together for Short Lives

The leading UK charity that speaks for all children with life-threatening and life-limiting conditions and all who love and care for them. They provide information, publications and resources, which families may find useful:

**[www.togetherforshortlives.org.uk](http://www.togetherforshortlives.org.uk)**

## DISCLAIMER

This booklet is not designed to, and does not provide, medical advice, professional diagnosis, opinion or treatment to you or any other individual. It contains general information about medical conditions and treatments. The medical information in this booklet is provided 'as is' without any representations or warranties, express or implied. Dravet Syndrome UK makes no representations or warranties in relation to the medical information in this booklet. Medical information changes constantly. Therefore the information in this booklet should not be considered current, complete or exhaustive.

Never rely on the information in this booklet in place of seeking professional medical advice. You should never delay seeking medical advice, disregard medical advice, or discontinue medical treatment because of information in this booklet. You should ask your doctor to assist you in interpreting any information in this booklet or in applying the information to your individual case.

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## EMERGENCY SEIZURE PROTOCOL

**FOR**

**WHO SUFFERS FROM DRAVET SYNDROME**

Name: \_\_\_\_\_ D.O.B \_\_\_\_\_ Weight: \_\_\_\_\_

Mother: \_\_\_\_\_ Father: \_\_\_\_\_

Address: \_\_\_\_\_

\_\_\_\_\_

Phone (home): \_\_\_\_\_ Mobile/Work: \_\_\_\_\_

Emergency Contact (*list contact details for any other person who can attend in an emergency*):

\_\_\_\_\_

\_\_\_\_\_

Paediatrician (*name, hospital and contact number/email*):

\_\_\_\_\_

\_\_\_\_\_

Neurologist (*name, hospital and contact number/email*):

\_\_\_\_\_

\_\_\_\_\_

Remember this may need regular updating



Current Medication *(list current doses [ideally in mgs and mls] and the times given):*

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Allergies: \_\_\_\_\_

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Seizure response:

- 1) Place \_\_\_\_\_ in the recovery position, if possible during the seizure to ensure safety. **DO NOT** place anything in his/her mouth and remove any heavy objects from around the immediate area.
- 2) Note the time the seizure began. If it continues for \_\_\_\_\_ minutes, administer \_\_\_\_\_

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*(Continue here with your child's individual protocol. If you do not have one, work with your medical team to produce a step by step protocol that can easily be followed.)*



## HOSPITAL PROTOCOL

### FOR

### WHO SUFFERS FROM DRAVET SYNDROME

Name: \_\_\_\_\_ D.O.B \_\_\_\_\_

Mother: \_\_\_\_\_ Father: \_\_\_\_\_

Address: \_\_\_\_\_

#### Primary Diagnosis:

Dravet Syndrome – a rare and life-limiting neurological condition causing severe seizures that are extremely resistant to anti-epileptic drugs and will often go into status epilepticus. Please see **www.dravet.org.uk** for more information on Dravet Syndrome.

Secondary Diagnosis (*include here anything else your child is diagnosed with, e.g. Autism, ADHD etc*):

Medications: (*list all medications here. You may wish to put the doses on but remember you will have to regularly update if you do*):

#### Seizure Protocol:

If admitted with a seizure please see the attached seizure protocol.

Remember this may need regular updating



**General Protocol:**

If for reasons other than a seizure please ensure the following is carried out *(Here you may wish to include things like checking ears and throat, full bloods taken etc.):*

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If in any doubt please speak to *(put the names of your local team here, along with your child's neurologist if they have one):*

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*Hope for families with  
life-limiting epilepsy*

**Dravet Syndrome UK**  
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**[www.dravet.org.uk](http://www.dravet.org.uk)**

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