



DAVET Syndrome UK

Hope for families with
life-limiting epilepsy

Supporting those with Dravet Syndrome: Guidance for Care Providers

Dravet Syndrome is an unpredictable, complex and little known condition.

Families describe the demands of caring for a child or dependent adult who has complex epilepsy and a wide range of other challenging symptoms as immense. Amongst current providers of respite or support, a significant lack of knowledge of the condition characterised early contact with families; and the complexities of the condition were not always understood or catered for. This guide seeks to address these gaps.

- As a guide for those responsible for awarding or providing short breaks (respite), care or support to children or dependent adults with Dravet Syndrome and their parents and families*
- Provides some background information on the condition based on informed research
- Provides feedback and information from families and providers who participated in a comprehensive research and engagement project completed Spring 2013
- Builds on emerging national policy development around integrating health and social care; transforming the quality of care; and ensuring that the right support is available, regardless of who commissions it



What is Dravet Syndrome?

Dravet Syndrome is a severely disabling type of life-limiting epilepsy. When it was first described in 1978 by Charlotte Dravet, it was known as Severe Myoclonic Epilepsy of Infancy. Dravet Syndrome is rare, affecting between 1 in 20,000 to 1 in 40,000 births. Normally, the first seizure takes place in the first year of life, often accompanied by a high fever.

Dravet Syndrome is typically characterised by multiple, frequent, resistant and sometimes rare type of seizures. Seizures in the first year are often **tonic**

clonic or **clonic** (jerking movements), prolonged, and may affect one side of the body. But following this a range of seizures appear which may include:

- **myoclonic jerks, 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
- **partial seizures**, which may involve loss of awareness
- **non-convulsive status** where the child develops a prolonged groggy, poorly functional state
- **status epilepticus**, a prolonged seizure or a cluster of shorter seizures with little or no recovery in between

Individuals with Dravet Syndrome are particularly prone to **status epilepticus** (potentially damaging prolonged seizures); and face a higher incidence of **SUDEP (Sudden Unexpected Death in Epilepsy)** than the general epilepsy population.

Every child or adult with Dravet Syndrome is different, with some affected more severely than others or with different emphasis of symptoms. But for all, the condition is complex, symptoms can be interrelated and seizures can be highly unpredictable.



Epilepsy is just one part of Dravet Syndrome.

After the first year of life, the child's development typically slows down or regresses, sometimes severely. A wide range of secondary conditions ('comorbidities') and issues may commonly include:

- **learning difficulties**, often severe, frequently including communication/speech problems and incontinence
- **features of autism and/or attention deficit hyperactivity disorder**, alongside inappropriate or obsessive behaviours, lack of awareness of danger or sensory issues
- **orthopaedic or mobility issues** such as gait disturbance and deterioration, ataxia (unsteadiness), poor co-ordination/motor skills and muscle tone issues
- **sleep disturbances**, often severe and persistent; and often alongside disruptive nocturnal seizures
- **eating, growth or nutritional concerns**, potentially requiring a gastrostomy

Other reported health issues amongst some include dysautonomia, cardiovascular problems, infections and immune dysregulation and dental health problems.

Why do families seek respite?

'Dravet Syndrome leaves no aspect of family life untouched.'

Epilegia 2011



High quality respite or support is essential for families of patients with Dravet Syndrome, but not always easy to come by. Our engagement with families told us that:

Parents seek respite to provide them with essential breaks from coping with the complexities of the condition; and to ensure that the needs of the whole family are met, particularly those of siblings, for whom the impact can be enormous.

'I need time out with my other children without the stress of seizures and bad behaviour.'

Typically families are already exhausted by the time they seek support; and some have faced lengthy, difficult and sometimes demoralising assessment and decision processes.

'We are so painfully tired the majority of the time.'

Some have reservations about admitting the need for help or anxiety about handing over care; whilst others have faced refusals for support or have not been aware of what is available.

'How could anyone cope with my child as well as me?'

Implications for Care

'The main goal in caring is to search for the attainment of the best quality of life for the patients and their families'

T Granata. Comprehensive care of children with Dravet Syndrome. *Epilepsia* 2011 Apr; 52(2): 90–4

Proven benefits

'Respite is the light at the end of the tunnel.'

Parent

Our engagement with families and providers told us that good quality respite can provide the whole family with a range of vital benefits:

Benefits for the child with Dravet Syndrome:

they can bond with carers; gain independence; access play activities and social interaction; and be kept safe and well.

'If your child is happy, you are happy'

Benefits for parents: they can gain rest and sleep; gain a vital break from the complexities of caring; have support at critical times; have time and space to do other things from relaxing to housework to working; address marriage stress; and crucially have time to support the wider family.

'We are able to benefit from time to rest from the physical demands'

Benefits for siblings and the whole family:

siblings – who often feel neglected – can access vital uninterrupted parental time; access normal activities that they so frequently miss out on; or they may gain support from the provider themselves.

'We get some brief normality'

Providing aggressive management of epilepsy

Given that patients with Dravet Syndrome are particularly prone to status seizures or clusters, **swift medical intervention** is essential to stop the seizures as soon as possible and reduce the risk of brain damage. Carers must be trained in **aggressive seizure management** including seizure avoidance and identification; temperature management; administration of anti-epilepsy drugs and/or specialist diets; emergency procedures including administration of emergency medication; and postictal care.

Given the frequent presence of nocturnal seizures and sleep disturbances; and the higher risk of SUDEP, **effective observation and monitoring** is vital for all overnight care. Depending on the child's needs, carers may need **training in the use of vital equipment**, such as pulse oximeters; video monitors; oxygen; or VNS (vagus nerve stimulation).

A comprehensive, proactive and consistent care plan

should be agreed between professionals and family and strictly adhered to.

Addressing comorbidities and providing interdisciplinary care

Patients with Dravet Syndrome may present with multiple problems beyond the intractable epilepsy, such as those listed overleaf. Those responsible for assessment and awards should ensure the family have **adequate inter-**

'Identification of specific comorbidities will facilitate a distinct and multidisciplinary approach to management, addressing seizure control, behaviour problems, cognitive difficulties, and motor impairment.'

Comorbidities and predictors of health-related quality of life in Dravet Syndrome: A Brunklaus, L Dorris, and S M Zuberi 2011

disciplinary professional support

to assess and address the complex range of symptoms.

Those involved in direct care should familiarise themselves with the specific complexities of the child's condition and ensure they work in partnership with the family to **provide skilled care for the whole person**. Additional training may be required.

Risk assessments, behavioural strategies, psychological, speech, occupational and physio therapies may accompany care plans and should be familiarised by care givers.



'Treatment demands prompt and focused therapy, as prevention of status epilepticus with regular medication and emergency protocols is important and may influence developmental outcome'

Prognostic, clinical and demographic features in ... Dravet Syndrome:
A. Brunklaus et al. *Brain* June 19, 2012

What good quality respite looks like

Good short break or respite provision is evidenced by peace of mind and confidence felt by parents when they leave their child in a provider's care, to deal with unpredictable medical needs and complex wider symptoms. It has the following features:

- **High calibre and consistent carers.** Well loved by the child and friends of the family; well trained, reliable and responsive to change
- **Flexibility in the overall care package.** Ensuring care is given when and where it is most needed, given the unpredictable, complex and changing nature of the condition; and in accordance with the preferences of families
- **A well informed and proactive social worker.** Providing assessment, support and co-ordination from an early age
- **Meeting the needs of the whole family.** Benefiting child, parents and siblings alike
- **Enabling the child to engage in social, fun or independent activities.** Ensuring sufficient resources are in place for provision the child might otherwise not access
- **Providing support overnight, either at home or away.** Considering carefully how provision can address parent's exhaustion and, if appropriate, provide for the child's complex night-time needs
- **High level of knowledge about Dravet Syndrome.** Ensuring carers are regularly trained; parents' expertise is honoured; and sufficient time is given to get to know the child's specific symptoms, needs and routines
- **Underpinned by up to date planning and procedures.** Clear and robust emergency protocols; risk assessments; comprehensive care plans; and input from multi agency professionals
- **Consistency and continuity of care.** Families become dependent on familiar and consistent support that benefits the child and builds trust for the families



Advice from providers or carers

'We take the lead from the parents and aim to work in partnership, valuing each other'

Hospice Manager

- **Fully understand the complexities** of the condition
- **Value the input of parents** and adopt an approach of joint working
- **Plan well** by building up with regular visits; ensuring sufficient resources, protocols and assessments are in place, relevant to the setting; and matching carers to child/family well
- **Have carers that can meet the child's needs** and are confident in dealing with seizures and other associated disabilities – access regular specialist training if needed
- **Ensure good medical protocols** are in place; and build links with health professionals or local hospitals
- **Keep an open mind**, regardless of prior experience and knowledge
- **Value the child**, meet their needs and try to keep to their routines – learn from them
- **Support the whole family** – see your provision as part of a multi-disciplinary team approach
- **Provide as much continuity of care** as possible
- **Commit to the long term** – it takes a long time to fully understand the child and gain confidence

'I had never cared for anyone with such a complex and unpredictable condition. I needed to think and work in a new way.'

Carer

'Parents need to be safe in the knowledge that they can have a break from their caring responsibilities knowing their child is safe.'

Direct Payments Support Worker



www.dravet.org.uk

Dravet Syndrome UK is 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.

With grateful thanks to the families and providers who contributed to the research and engagement project on care and management of Dravet Syndrome, November 2012 – May 2013

*Certain terms are used for simplicity throughout this guide which may have a wider meaning: **'Child'** or **'Patient'** refers to the child or dependent adult who has Dravet Syndrome or a related condition. **'Parent'** refers to the adult(s) with primary responsibility for caring for the child/adult with Dravet Syndrome. **'Respite'** refers to short break provision or regular care that may take the form of day and evening care; play schemes or activities; or overnight care. It may be offered in the child's own home; at the provider's premises or may involve trips out. Awards may be made by Social Services, Health Authorities or other statutory services; or they may be made independently by charities, hospices and the like. **'Provider'** refers to the agency organisation, individual carer or centre involved with delivering the care to the child.