

Emergency medication and protocols

Emergency protocols

As Dravet Syndrome is a rare condition, it would be unrealistic to expect all medical professionals to immediately recognise it and know how it should be treated. Having emergency protocols that are readily available and accessible can make treating your child/adult in an emergency situation a lot easier for everyone concerned. Emergency protocols will also make sure that the same procedures are followed, wherever you are.

It's important to work proactively with your medical team to produce some protocols. We recommend preparing three emergency protocols:

- **Emergency seizure protocol:** Detailing the medications to be used when the person is seizing, the order in which to use them, and the appropriate doses in milligrams per kilogram (mg/kg).
- **Ambulance protocol:** Detailing the medications which can be used on the way to the hospital and information about the measures needed to be taken when you get there. You will need to work with your local consultant to write the protocol and they will liaise with your local ambulance service.
- **General hospital protocol:** There will be times when you just know that something is not right with your child/adult, yet you can't quite put your finger on it. A general hospital protocol would provide some brief information on Dravet Syndrome, any other diagnoses your child/adult has, and what tests should be carried out, for example, blood tests, swabs, checking ears and throat.

All protocols should include your child's/ adult's information – their date of birth, address, current medications, allergies and contact numbers for your medical team. They should be on hospital letter headed paper and signed off by your consultant. You may find it helpful to have a laminated version of your child's emergency protocols and an electronic copy of each plan on your phone.

Once you have your protocols, we advise you to keep a copy in the following places:

- Your child's/adult's bag at all times
- At their school/college/home
- With your ambulance station
- In the A&E department and on the ward of your local hospital
- At your GP surgery

Communication passports

A 'communication passport' provides a practical, person-centred approach to passing on key information about a person with complex communication difficulties. It's not a replacement for emergency protocol information, it should go alongside it. The passport is a useful tool for hospital staff to help them get to know a person with communication difficulties and interact with them. It can help the hospital staff to make sure your child/adult understands the situation they are in. A communication passport presents the person with communication difficulties positively, as an individual, not as a set of 'problems' or disabilities.



For more information, visit the *Family Guide* resources page of our website: www.dravet.org.uk/family-guide-resources



The practicalities of using emergency (rescue) medications

Rescue medication is the term used to describe medicine given in an emergency situation to relieve symptoms quickly. In epilepsy, if a seizure lasts more than five minutes, rescue medication is usually required.

Unfortunately, prolonged and 'cluster' seizures (where seizures occur close together without recovery time in-between) are all too common in Dravet Syndrome, particularly in early childhood. All children and adults with Dravet Syndrome will need to have an emergency protocol that has been developed with their neurologist, which includes details of their home rescue medication and instructions for administering it.

It's important to note that rescue medication varies from person to person. For some, oxygen is used at home, especially if there is 'respiratory depression' with seizures, which means the lungs fail to exchange carbon dioxide and oxygen efficiently. But this varies on a case-by-case basis and should always be discussed with your medical team.



For a list of emergency medications, see the Dravet Syndrome UK website:

www.dravet.org.uk/medications

It doesn't matter how many times you have to administer a rescue medication, it can still be a scary experience. Over time, parents/carers get used to having to do it, but the fears and anxieties never go away. It's a stressful situation – you may find that as well as administering the medication, you're dialling 999 and trying to calm others around you.

Take a deep breath and try to remain as calm as possible. What you're doing for

your child/adult is amazing and potentially life-saving. Many parents/carers say that at the time of administering the medication they remain calm, it's afterwards when their child/adult 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.

Initial emergency treatment once in hospital

- The medical team will want to check your child's/adult's basic airway, breathing and circulation, and are likely to set up an intravenous line (called a cannula), give oxygen and call other members of the team to help, depending on how unwell they are. They may take blood to check blood glucose levels and for infection, and to ensure that the kidneys and liver are working well.
- The first rescue medication given in hospital is usually an intravenous benzodiazepine, typically lorazepam, which can usually be repeated once.
- Your child's/adult's emergency protocol may indicate that they respond best to a particular emergency treatment and for this to be given, if possible, as the next step.
- In the past, intravenous phenytoin would be given as the next step. In current practice, intravenous levetiracetam is generally preferred. However, intravenous phenytoin can still be beneficial in some cases and may be given even if the person is already on regular levetiracetam.
- If there's any delay in inserting a cannula to administer intravenous medication, then diazepam or paraldehyde may be given rectally so it can get into the body quickly.

- In some extreme cases, when veins cannot be accessed by cannula, a procedure called Intraosseous Infusion (IO) is used. IO involves injecting medications, fluids, or blood products directly into the marrow of a bone. It can be very scary to witness but is an important, potentially life-saving procedure that enables vital medications and fluids to be quickly delivered directly into the vascular system when it's not possible to use other methods.
- If your child/adult doesn't respond to the initial intravenous medication, then an anaesthetist should be called, as further medication and the ongoing seizure may reduce the drive to breathe, meaning that help with breathing may be needed.

Ventilation

People may need ventilation if they have a condition that makes it hard for them to breathe properly or when they can't breathe on their own – for example if they are unconscious.

A medical ventilator is a machine that helps a person's lungs to work by pushing air in and out of their lungs so their body can get the oxygen it needs. They may wear a fitted mask to get oxygen from the ventilator into their lungs. Or, if the condition is more serious, a breathing tube may be inserted down the throat to supply the oxygen to the lungs.

Why ventilate?

There are two main reasons why your child/adult may need to be ventilated – either because they are unable to breathe effectively for themselves, or because they are unconscious and so can't protect their airway as they would usually.

The first reason is most commonly due to infections, either in the lungs or somewhere

else in the body, something obstructing their airway, 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 consciousness levels. This typically happens when a person is seizing or following a seizure. An unconscious person is in danger because there is a risk of their stomach contents entering their lungs, by inhaling vomit, which can cause an obstruction or infection. When someone is unconscious, it can result in them not being able to breathe effectively and take in the oxygen they need for their cells and tissues to function.

What happens when someone is ventilated?

If your child/adult is ventilated, they will be given some medication to help them relax, even if they appear unconscious. An 'endotracheal' tube will be put in their mouth and fed down the windpipe ('trachea'). This is called intubation. The tube is then connected to a ventilator, sometimes called a life support machine.

The machine then assists, or takes over, your child's/adult's breathing until they are well enough to breathe by themselves. It will take anything from a couple of hours to a few days, or occasionally, weeks. There are several different ways of using these machines. Each person will need something different. But all the machines deliver oxygen into the lungs and take carbon dioxide away.

During this time, your child/adult will be monitored extremely closely, particularly their heart rate and oxygen levels. They will be given sedation to make sure they are comfortable and sleepy.

Will I be consulted if my child/ adult needs ventilation?

For children: If your child needs ventilation, a doctor and an anaesthetist will try and discuss the pros and cons with you before they proceed. But in the case of emergencies, they may have to explain the procedure afterwards. It's up to the doctors and nurses whether you're able to be there during the intubation, but they will 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 don't have a paediatric intensive care unit in your local hospital, your child will need to be transferred to the nearest centre.

For adults: The process is slightly different for adults, where the medical staff will take what's known as a 'best interest decision'. As a parent/carer, you will be consulted, but it's ultimately the medical professional who has the final say on what will happen medically, or indeed in relation to any major decision affecting your adult son's or daughter's life. This can come as a shock if you're not prepared.





Hope for families with life-limiting epilepsy

Dedicated to improving the lives of people affected by Dravet Syndrome through support, education and research.

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