

## Toby James O'Grady

DOB: 22<sup>nd</sup> November 2013

45 West Side Rise, Olney, Bucks, MK46 5HP

NHS number 652-861-3745

MRN- MK 797808

MRN- Oxford 10278648

'Red Box' open access to PAU/Ward 5 Milton Keynes General Hospital

## Emergency Care Plan – Drugs

1. **5mg** Buccal Midazolam 3-5 mins into seizure.
2. **12ml** of Paraldehyde 5mins after dose of Midazolam
3. 2.5-5mg Buccal Midazolam 10 mins after step 2 or if in hospital move to step 4.
4. IV Lorazepam (A&E)
5. Admission to HDU for IV Phenobarbitone

Toby is NOT to be given **IV PHENYTOIN, CARBAMAZEPINE OR OXCARBAZEPINE** as these can worsen seizures in Dravet patients.

As per Dr McShane, Neurologist, Oxford John Radcliffe, July 2014

Revised 21<sup>st</sup> April 2016

**Weight – 14.3kg**

### Current Medications:

- Sodium Valproate (200mg/5ml) – 160mg/4ml bd
- Clobazam (5mg/5ml liquid) – 2.5mg/2.5ml bd
- Stiripentol (250mg sachet) – 450mg bd
- Midazolam – 2.5-5mg mg as required
- Paraldehyde – 0.8ml per Kg as required

### Ketogenic Diet:

- Toby is on the **KETOGENIC DIET – Only Ketocal** to be given as feed on a 3:1:5 ratio
- Do not include dextrose in IV fluid due to impact on ketones.

### Medical History:

- Seizures started at 8 weeks, **Dravet Syndrome** diagnosed at 6months of age
- Inoculations all up to date, **No Known Allergies**
- Pregnancy was all fine with no complications, delivered by emergency C-Section 10 days after due date.
- Large doses of **electrolytes** can **trigger seizures**.
- Other tests completed which all have been clear including ECG, EEGs, MRI and full blood work.
- During **intubation** (May 2015) administered: Thiopentone, Ketamine, Rocoronium, morphine & Midazolam for sedation, **Note:** Once sedated it took Toby 24+hrs to start to come round after the treatment had stopped.
- Atracurium and Fentanyl successfully administered as GA.

### Typical seizures

- Partial seizures involving twitching and jerks and absence seizures, Generalised Tonic Clonic range from 1-70mins, **Status epilepticus** is common.
- **Apnea** during seizures is common, often requiring rescue breaths and oxygen.
- **Hemiplegia** is common post seizure.
- Often had 2 doses of Midazolam and 1 dose of Paraldehyde with no visible impact.
- Viral infection, Temperature, Stress + Excitement seem to trigger prolonged seizures

### Family:

- Tim O'Grady – 17/05/1975 – 07970 263638  
tim.ograde@hotmail.co.uk
- Kim O'Grady – 29/04/1977 – 07713064432  
kim.ograde@hotmail.co.uk
- No limitations on visitors on the Ward
- No Siblings, lives at home with parents

### Grandparents:

- Kay O'Grady – 07901 748930
- Gill Burfitt – 07800 522751

### Medical Personnel

- **GP** – Dr Jon Walter, Cobbs Garden Surgery, Olney
- **Paediatrician** - Dr Oommen, Milton Keynes
- **Neurologist** – Dr Kate Lamb, John Radcliffe, Oxford
- **Dietician** – Phillipa Thomas, Bristol Children's Hospital, Phillipa.Thomas@UHBristol.nhs.uk 0117 3428802
- **Complex Needs Nurse** – Anne Sharples, anne.sharples@mkchs.nhs.uk 07909527087
- **Social Worker** – Sue Bateman, susan.bateman@milton-keynes.gov.uk 01908253977

### Historical AED's tried

- Topiramate, Keppra, Clonazepam, Phenol barbitone.
- Pyridoxine & Biotin (vitamins)

### Dravet Syndrome

- Severe life limiting epilepsy.
- Also known as severe myoclonic epilepsy of infancy (SMEI), is a rare genetic disorder, occurs one in every 30,000 births.
- It is a progressive disorder characterized by multiple seizure types, often including life-threatening status epilepticus and is resistant to all AED's.
- The course of the condition is variable from patient to patient and earmarks of the syndrome include multiple seizure types that are resistant to treatment, developmental delays, lowered immunity, orthopedic concerns, and hyperactivity.
- Individuals with DS also face a higher incidence of SUDEP.



## Dravet Syndrome

Dravet syndrome, also known as severe myoclonic epilepsy of infancy (SMEI), is a rare genetic disorder which occurs in roughly one in every 30,000 births. It is a progressive disorder characterized by multiple seizure types, often including life-threatening status epilepticus (prolonged seizures that require emergency care.) The course of the condition is variable from patient to patient and earmarks of the syndrome include multiple seizure types that are resistant to treatment, developmental delays, lowered immunity, orthopedic concerns, and hyperactivity. A significant number of patients have a family history of febrile seizures or seizure disorders.

During the second year of life the seizures become more frequent and persistent, and are often more obviously focal (also called partial) involving one part of the body. They may happen with or without a fever, and at any time of day and night. In addition to the tonic-clonic seizures, myoclonic seizures ('myo' meaning muscle, and 'clonus' meaning jerk) and focal seizures become common. Often the children are photosensitive (have seizures brought on by flashing or flickering lights). Seizures may also sometimes be brought on by hot environments or hot showers or baths.

Children with Dravet syndrome (DS) have poor language development and impaired motor skills. Early development is normal before seizures start, but in the second year of life development typically slows. Later, regression becomes evident and is often accompanied by hyperactivity and mental retardation. As children with DS get older their decline in cognitive function stabilizes and seizures have a tendency to improve. However, most teenagers with DS are dependent on caregivers and will require life-long care. Individuals with DS also face a higher incidence of SUDEP (sudden unexplained death in epilepsy) and have associated co-morbid conditions which also need to be properly managed.

### References

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

[www.dravetfoundation.org](http://www.dravetfoundation.org)

[www.epilepsy.org.uk/info/syndromes/severe-myoclonic-epilepsy-in-infancy](http://www.epilepsy.org.uk/info/syndromes/severe-myoclonic-epilepsy-in-infancy)