

# Mortality and SUDEP in Dravet Syndrome: Information from Families for Doctors

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## RATIONALE:

Dravet syndrome is a devastating epileptic syndrome with onset in infancy. The mortality rate is described as very high (Roger, Bureau, Dravet et al). Sudden Unexpected Death in Epilepsy (SUDEP) is a recognised syndrome where a person with epilepsy dies suddenly and no other cause of death is revealed. In the general population of people with epilepsy the risk of SUDEP is of the order of 1:1000 per year. In children with Dravet syndrome the incidence of SUDEP appears much higher than this and the need for greater understanding about the reason for this is urgently needed. The high incidence of SUDEP in recent years on our Family Network, 10 cases since 2006, has further increased anxiety of parents and highlighted a desperate need for both research into SUDEP in Dravet syndrome and open discussion between doctors and families.

## METHODS:

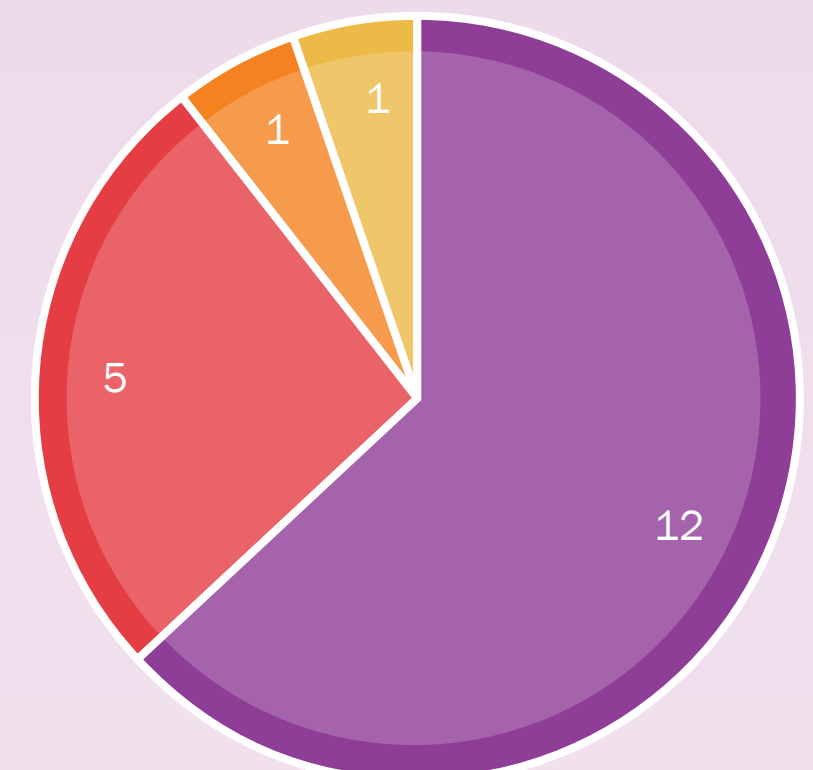
The Dravet syndrome Family Network, a support group for parents with affected children, is associated with the International Dravet syndrome Epilepsy Action League (IDEA League). In September 2009, 608 individual children from 40 different countries, were represented by their families on the support group. Since 2000, at least 19 children from the support group have died. Information about the children was collected from the parents through interviews and questionnaires.

## RESULTS:

All children who died were aged between 18 months and 8 years. Twelve children died from SUDEP (ages 1.5-8 years), five from status epilepticus (ages 5-7), one from keto acidosis (age 3) and one from a seizure-related accident (age 7). Detailed questionnaires were completed by 6 families. Four of the six children died from SUDEP at ages 2, 5, 6, and 8 years, and two from Status Epilepticus (SE) at ages 5 and 7 years. Five of the children had SCN1A mutations and the sixth child had died in 2000 prior to testing being available so their status is unknown. None of the children had medication changes in the 3 months prior to death and at the time of death, health and seizure control were described as good for the children, although one had a slight cold and one a slight temperature in the day of their death. One child was on just the ketogenic diet, one on valproate monotherapy and the remaining four were on polytherapy including valproate, two also including stiripentol and three also including clobazam. Three out of the six children had had limited cardiac testing in the past. One had shown evidence of a long QT interval a year before death but this had not been evident at follow up testing. Another had had a heart ultrasound at the mother's request, and this was normal, and one had had a normal EKG prior to oral surgery 3 months prior to death. In two cases of SUDEP that occurred during the day in 2007 and 2008, the children were observed 10 and 15 minutes prior to death, one awake and one asleep, and nothing untoward was observed. Other cases happened during the night. Parents often report little or no visible signs of a seizure or movement.

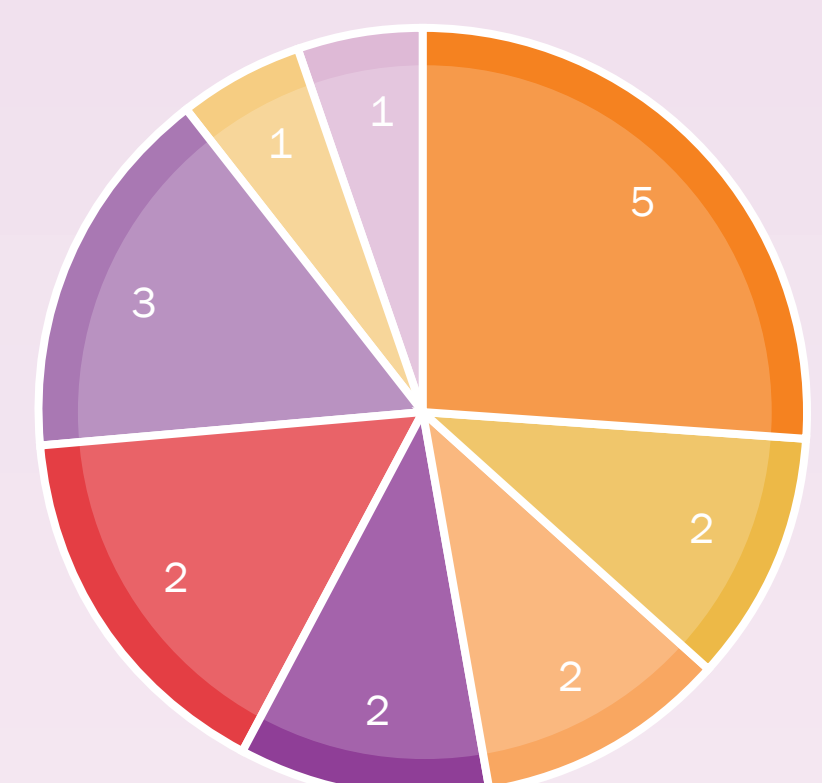
## CAUSE OF DEATH

■ SUDEP  
■ Status Epilepticus  
■ Accident  
■ Keto Acidosis



## AGE AT DEATH

■ 1 y, 0m to 1y, 11m  
■ 2y, 0m to 2y, 11m  
■ 3y, 0m to 3y, 11m  
■ 4y, 0m to 4y, 11m  
■ 5y, 0m to 5y, 11m  
■ 6y, 0m to 6y, 11m  
■ 7y, 0m to 7y, 11m  
■ 8y, 0m to 8y, 11m



## CHILD DEATHS ON THE IDEA LEAGUE FAMILY FORUM

Country	Gender	Year of Death	Age at Death	Cause of Death	
EC	Canada	Female	2000	7 years	Status Epilepticus
AM	South Africa	Male	2001	1 years	SUDEP
KR	England	Male	2003	5 years	Status Epilepticus
NT	Australia	Male	2004	6 years	Status Epilepticus
DG	England	Female	2004	6 years	SUDEP
AB	Australia	Female	2006	3 years	SUDEP
JL	USA	Female	2006	2 years	SUDEP
CB	Germany	Female	2007	7 years	Status Epilepticus
BW	Canada	Female	2007	7 years	Accident
ZW	USA	Male	2007	3 years	Keto Acidosis
GID	Argentina	Female	2007	2 years	SUDEP
CC	France	Female	2007	2 years	SUDEP
SC	USA	Male	2008	8 years	SUDEP
JL	England	Male	2008	2 years	SUDEP
AD	USA	Female	2008	4 years	SUDEP
RC	USA	Female	2009	5 years	SUDEP
MM	USA	Female	2009	2 years	SUDEP
TC	England	Male	2009	6 years	Status Epilepticus
HW	USA	Female	2009	4 years	SUDEP

## CONCLUSIONS:

The high incidence of SUDEP recently experienced on our Family Network, 10 cases since 2006, has increased anxiety of parents and highlighted a desperate need for research into SUDEP in Dravet syndrome. Preliminary results from our detailed questionnaires are showing commonalities in some areas. This study needs to continue to ascertain whether the common factors continue for a larger cohort and whether further issues are highlighted that warrant greater research and analysis in order to help identify possible causes of SUDEP and status epilepticus in children with Dravet syndrome.

## REFERENCES:

Roger J, Bureau M, Dravet Ch, Genton P, Tassinari CA, Wolf P, editors. Epileptic Syndromes in Infancy, Childhood and Adolescence, Third Edition. London: John Libbey Ltd, 2002.