

SUDEP prevention action – EPEN Guidelines for professionals

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**Introduction**

Sudden unexpected death in epilepsy (SUDEP) is a very small but recognizable risk in children with epilepsy. Health care professionals dealing with families of children with epilepsy have professional responsibility to inform about this risk at some appropriate time during diagnosis and treatment. This document helps professionals guiding appropriateness of initiation and content of these discussions with families.

**Definition**

"The sudden, unexpected, witnessed or unwitnessed, nontraumatic, and non-drowning death in patients with epilepsy with or without evidence of a seizure, and excluding documented status epilepticus ≥30 minutes in duration, in which postmortem examination does not reveal a structural or toxicologic cause for death." **(1)**

**Incidence**

SUDEP is generally rare in children. One systematic review and meta-analysis by the American Academy of Neurology reported the incidence of SUDEP in children to be 0.22 per 1,000 patient-years (95% CI 0.16–0.31). This means in 1 year, SUDEP typically affects 1 in 4,500 children with epilepsy. The numbers were higher in adults (1.2 per 1000 person-years, 95% CI 0.64-2.32) **(2)**

Although, a later Swedish population-based cohort study that included > 57,000 epilepsy patients reported a higher number in children as 1.11 per 1,000 patient-years (95% CI 0.45–2.29). This means in 1 year, SUDEP typically affects 1 in 900 children with epilepsy. **(3)**

Pathophysiology

The pathophysiology of SUDEP is not clearly understood. It is believed to occur during or following a seizure. It has been suggested to be due to cardiac arrhythmia, respiratory dysfunction, cerebral shutdown, genetic, drug, metabolic, and environmental factors. **(4)**

Risk factors

Data from a recent Swedish population-based case-control study suggest risk factors related to seizures character and frequency, treatment, comorbidity, demographic characters, and living conditions. **(5)**

**Moderate to high risk**

* Generalised tonic clonic seizures (presence and frequency)
* Nocturnal/sleep seizures especially with no supervision
* Medically refractory epilepsy (when not being treated)
* Individuals with poor concordance
* Never treated with antiepileptic drugs
* Young adults who live alone especially males
* Sharing household but not bedroom
* Combination of having at least one GTCS and not sharing a bedroom with someone
* Previous diagnosis of substance abuse or alcohol dependence (two-fold risk)

**Low to unknown risk**

* Mental health disorders were not associated with increased SUDEP risk
* Lesional epilepsy
* Sudden withdrawal of antiepileptic drugs
* Frequent and abrupt changes to medication
* Duration of epilepsy
* Age of onset

**Reduced risk**

* Both monotherapy and polytherapy were associated with a reduced risk.
* Epilepsy surgery was not associated with increased or reduced risk.
* Vagus nerve stimulation was associated with a 59% reduced risk.

How to minimize the risk of SUDEP?

Optimizing management of the epilepsy and striving for seizure freedom, where possible, should remain at the forefront of preventive care. This includes accurate diagnosis, using appropriate AEDs and referring for tertiary centers at an early stage in case of refractory epilepsy are the main stays of risk reduction. The following steps are helpful for professionals involved in care of children and young persons with epilepsy.

* Nocturnal supervision (age appropriate and consider privacy)
* Raising the awareness of risks with epilepsy including SUDEP
* Identifying and minimizing triggers for seizures
* Encouraging compliance / adherence
* Regular reviews
* Monitoring including self-monitoring by young adults using technology
* In refractory epilepsy consider earlier involvement of tertiary centres and referral for ketogenic diet or epilepsy surgery when appropriate
* Use of SUDEP safety checklist ([www.sudep.org](http://www.sudep.org))
* May recommend safety devices for seizures in sleep
	+ Anti-suffocation pillow
	+ Apnoea/seizure monitor

Discussion about SUDEP

### What is the right time for the initiation of discussion?

It is still a contentious issue as to the initiation and right timing of discussion of SUDEP with families with children and young adults with epilepsy. Some professionals argue that the risk of SUDEP is minimal in children, therefore, the introduction of the topic would cause undue stress/ worry and may break professional relationships with the family. On the contrary, it is easier to initiate discussion at the time of diagnosis as seizure freedom is still the goal of treatment. Parents who have witnessed the first seizure carry a strong impression of life threat. Therefore, discussion about SUDEP during initial professional contact may be appropriate as realizing the true risk may be less daunting than true experience. SUDEP discussion should be done with family and young adults at some point during the journey if not made during the initial days. NICE (CG 137) recommends that “Tailored information and discussion between child, young person or adult with epilepsy, their family and/or carers (as appropriate) and health care professionals should take account of the small but definite risk of SUDEP”.

*EPEN recommends that discussion about all risks relating to seizures (including SUDEP when appropriate) and treatment should be made at an early stage, near to the time of diagnosis*

### Conditions where it is appropriate to discuss

EPEN recommends initiating the discussion about SUDEP under these broad guidelines which are not restrictive. Discussion should be sensitive and consider the child or young adult’s emotional maturity.

SUDEP **should always** be discussed with families of children:

* + With generalized tonic-clonic seizures (GTCS)
	+ With symptomatic epilepsy, (i.e. epilepsy in the context of a structural brain abnormality, metabolic disorder or neurodegenerative disorder)
	+ With drug-resistant epilepsy especially tonic-clonic seizures
	+ With recurrent episodes of status epilepticus (+/- admissions to PICU)
	+ With associated severe neurological impairment
	+ Who ask direct questions about death and epilepsy

SUDEP discussion should be **considered** in children:

* + With additional learning and physical difficulties
	+ Who are not complying with treatment
	+ Who are resisting treatment

SUDEP does **not** need to be routinely discussed with families of children with

* + Benign familial infantile seizures
	+ Childhood absence epilepsy
	+ Benign partial epilepsy with centrotemporal spikes (BECTS)
	+ Well-controlled benign epilepsies

### Who should discuss this?

Ideally, discussion regarding SUDEP should be done in the epilepsy clinic by a consultant, or a professional with special interest and/or expertise in paediatric epilepsy.

### Documentation of discussions

The content of the discussion should be clearly documented in the patient’s hospital notes and included in the correspondence to the GP that is copied to the parents and child. The family should also be given a generic information sheet on SUDEP. EPEN recommends providing the leaflet from young epilepsy (7) (see appendix).

### ****Advice on preventive devices****

There is insufficient evidence to recommend overnight monitoring as a means of preventing SUDEP. There is a wide range of monitors available in the market. Currently, they are not available under NHS prescription. Use of these monitors should be left to the choice of child, young person, and their family. Parents should be made aware of the rate of false alarms and disturbed sleep for the families may be highly problematic. As children get older, their rights and desire for privacy must also be taken into consideration.

### ****Support for families after SUDEP****

NICE guidance states that “Where families and/or carers have been affected by SUDEP, health care professionals should contact families and/or carers to offer their condolences, invite them to discuss the death, and offer referral to bereavement counselling and a SUDEP support group.” **(6)**

Charity [**SUDEP Action**](http://www.sudep.org/)**(8)** will give help in the event of a SUDEP. Their services include bereavement support, counselling, help with understanding the inquest process and in collaboration with UK research teams, the involvement of bereaved families and professionals in research through the Epilepsy Deaths Register. Families can be directed to the website and provide the contact number of the support team (01235 772850)

References

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6. National Institute for Health and Care Excellence [NICE]. (May 2021 update). Epilepsies: diagnosis and management [CG137]. <https://www.nice.org.uk/guidance/cg137/>
7. Patient information leaflet on SUDEP from Young Epilepsy website. Downloaded on 10th Aug 2021 from http://www.youngepilepsy.org.uk/about-epilepsy/living-with-epilepsy/sudep.html
8. SUDEP Action website: sudep.org

Appendix

PDF available from this link: https://www.youngepilepsy.org.uk/about-epilepsy/living-with-epilepsy/sudep.html

