**SUDEP prevention action – EPEN Guidelines for professionals**

Sudden unexpected death in epilepsy (SUDEP) is a very small but recognisable 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.

**What is SUDEP?**

“The sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning death in patients with epilepsy, with or without evidence for a seizure and excluding convulsive status epilepticus in which post-mortem examination does not reveal a toxicological or anatomical cause for death.”¹

**How common is SUDEP in Children?**

Occurrence of SUDEP is rare in children under 17 years compared to adults. Recent evidence² suggests that incidence of SUDEP is 0.22 per 1000 patient years in children. This means that in 1 year, SUDEP typically affects 1 in 4,500 children with epilepsy; therefore, 4,499 of 4,500 children will not be affected. The risk of unexpected, unexplained death in children with idiopathic epilepsy is extremely small.³ On the other hand, every year 500 individuals with epilepsy (including adults) lose their life in UK due to SUDEP.⁴ 42% of these deaths are in people with inadequately addressed factors that are known to be associated with increased incidence of SUDEP.

**Pathophysiological mechanisms of SUDEP**

The pathophysiology of SUDEP is not clearly understood. There is ongoing research into SUDEP. It is believed to occur during or following a seizure. The underlying mechanism may be cardiac arrhythmia or apnoea.

**What are the risk factors?**

Risk of SUDEP is higher in the following situations.

MODERATE TO HIGH

Generalised tonic clonic seizures (presence and frequency)

 Nocturnal/sleep seizures especially with no supervision

 Lack of seizure freedom (1-5 years)

 Medically refractory epilepsy (when not being treated)

 Individuals with poor concordance

 Never treated with antiepileptic drugs

 Many drugs used (not polytherapy)

 Young adults who live alone especially males

 Associated significant learning difficulties

 LOW TO UNKNOWN

Lesional epilepsy

Polytherapy

 Sudden withdrawal of antiepileptic drugs

 Frequent and abrupt changes to medication

 Young adults addicted to alcohol and drugs

 Associated psychological comorbidity

 Duration of epilepsy

Epilepsy surgery

Age of onset

It is the type of the seizure, not necessarily the epilepsy syndrome that indicates the risk.⁵ However, some of the complex epilepsy syndromes carry additional risk, e.g. Dravet syndrome carries a 15% risk of SUDEP.

**How to minimise the risk of SUDEP?**

Optimising 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 centres 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 minimising 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)
* May recommend safety devices for seizures in sleep
	+ Anti-suffocation pillow
	+ Apnoea/seizure monitor

**Discussion about SUDEP**

What is the right time for 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 risk of SUDEP is minimal in children therefore introduction of the topic would cause undue stress/ worry and may break professional relationship with family. On the contrary it is easier to initiate discussion at the time of diagnosis as seizure freedom is still the goal of treatment.⁵ 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 realising the true risk may be less daunting than true experience. SUDEP discussion should done with family and young adult at some point during the journey if not made during 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 small but definite risk of SUDEP”⁶.

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 discuss⁷

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

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

With generalised tonic clonic seizures (GTCS)

With symptomatic epilepsy, (i.e. epilepsy in the context of a structural brain

 abnormality, metabolic disorder or neuro-degenerative 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 centro-temporal spikes (BECTS)

 Well controlled benign epilepsies

Who should discuss?

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.⁸ (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 the left to the choice of child, young person and their family. Parents should be made aware of rate of false alarms and disturbed sleep for the families may be highly problematic. As children get older, their right 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/)**⁹** 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 support team (01235 772852)

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**Appendix**

