



# SUDDEN UNEXPECTED DEATH IN EPILEPSY (SUDEP) A CASE REPORT

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## ABSTRACT

Sudden unexpected death in epilepsy (SUDEP) is a fatal complication of epilepsy. It is defined as the sudden and unexpected, non-traumatic and non-drowning death of a person with epilepsy, without toxicological or anatomical cause of death detected during the post-mortem examination. we report a 50 years old lady presented with ruptured PICA aneurysm underwent clipping successfully, had sudden death in POD 8 diagnosed as Possible SUDEP.

**Keywords:** Sudden unexpected death in epilepsy (SUDEP), Posterior inferior cerebellar artery (PICA) aneurysm, Clipping

## 1. INTRODUCTION

Epilepsy is a common neurological disorder bearing the rare possibility of devastating results. Sudden unexpected death in epilepsy (SUDEP) is one of the most important but least known of these possible worrying results. The first formal definition of SUDEP has been made in 1997 by Nashef et al. as “sudden, unexpected, witnessed or unwitnessed, nontraumatic and non-drowning death in patients with epilepsy, with or without evidence for a seizure and excluding documented status epilepticus, in which post-mortem examination does not reveal a toxicologic or anatomic cause of death”(1)

## 2. CASE REPORT

A 50 years old female brought with low GCS with history of headache. CT brain was taken showed widespread subarachnoid haemorrhage with intraventricular and subtentorial haemorrhages, haematoma at the level of cervicomedullary junction mildly compressing the spinal cord and hydrocephalus suggestive of ?ruptured PICA/vertebral artery aneurysm. CT angiogram of brain showed a saccular bilobed aneurysm measuring 5.2 x 4.5mm seen arising from the right proximal PICA. Neck of the aneurysm measures 2.2mm. Focal narrowing of the PICA seen proximal to the aneurysm likely due to spasm - Narrow neck right posterior inferior cerebellar artery aneurysm. Patient underwent left frontal EVD followed by right sub occipital craniotomy, clipping of the PICA aneurysm without any complications. CT brain POD 1 -status post suboccipital craniotomy and clipping of ruptured right PICA aneurysm with EVD in-situ, residual subarachnoid, intraventricular and subtentorial haemorrhages and haematoma at the level of cervicomedullary junction around clipping. On 5th POD EVD tube was removed, patient was mobilised and started on normal diet. On 8th POD afternoon patient had sudden drop in GCS, developed apnea, ? seizure and became unresponsive, patient was

intubated and resuscitated. Patient was started on inotropes and emergency CT brain was done showed Post op status with evolving acute /subacute ischemic changes in the left watershed territory (MCA-PCA) and diffuse cerebral edema. Cardiac evaluation was done which was normal and other possible causes were ruled out, but inspite of all resuscitative measures patient expired, hence diagnosed as probable/possible SUDEP.

### 3. DISCUSSION

Sudden unexpected death in epilepsy (SUDEP) is a fatal complication of epilepsy. It is defined as the sudden and unexpected, non-traumatic and non-drowning death of a person with epilepsy, without a toxicological or anatomical cause of death detected during the post-mortem examination.(2)(3) While the mechanisms underlying SUDEP are still poorly understood, it is possibly the most common cause of death as a result of complications from epilepsy, accounting for between 7.5 and 17% of all epilepsy-related deaths and 50% of all deaths in refractory epilepsy.(4)(5) The causes of SUDEP seem to be multifactorial and include respiratory, cardiac, and cerebral factors as well as the severity of epilepsy and seizures. Proposed pathophysiological mechanisms include seizure-induced cardiac and respiratory arrests. Among epileptics, SUDEP occurs in about 1 in 1,000 adults and 1 in 4,500 children annually. Rates of death as a result of prolonged seizures (status epilepticus) are not classified as SUDEP. The overarching term SUDEP can be subdivided into four different categories: Definite, Probably, Possible, and Unlikely.(6)

- A. Definite SUDEP: a non-traumatic and non-drowning death in an individual with epilepsy, without a cause of death after post-mortem examination.
  - a. Definite SUDEP Plus: includes the presence of a concomitant condition other than epilepsy, where death may be due to the combined effects of both epilepsy and the other condition.
- B. Probably SUDEP: all the same criteria for Definite SUDEP are met, but no post-mortem examination is performed.
- C. Possible SUDEP: insufficient information is available regarding the death, with no post-mortem examination.
- D. Unlikely SUDEP: an alternate cause of death has been determined, ruling out the possibility of SUDEP being the cause.

The mechanisms underlying SUDEP are not well understood but probably involve several pathophysiological mechanisms and circumstances.(7)

- ❖ Cardiac factors: cardiac arrhythmias and other cardiac events are known to be involved in some cases of SUDEP. Such arrhythmias are defined as ictal arrhythmias and include the ictal asystole, which is a rare occurrence mostly in people that have temporal lobe epilepsy.
- ❖ Respiratory factors: impaired respiration and seizure-induced pulmonary dysfunction as well as central apnea as a result of brain-stem respiratory centres suppression is known to play a role in some cases of SUDEP
- ❖ Cerebral and autonomic nervous system dysregulation: cardiac arrhythmia and respiratory failure as a result of seizure-related changes to brain function and dysfunction of the autonomic nervous system have been described in cases of SUDEP. These include cases of post-ictal generalized EEG suppression described as cerebral shutdown, but its significance remains unclear.
- ❖ Genetic factors: mutations in several genes have been associated with an increased susceptibility to SUDEP. Over 33% of these are related to mutations which lead to increased susceptibility for arrhythmia. Genes involved include the hyperpolarization-activated cyclic nucleotide-gated channels genes (*HCN1*, *HCN2*, *HCN3* and *HCN4*).
- ❖ Anti epileptic drugs: most evidence suggests that antiepileptic drugs are not associated with an increased risk for SUDEP, but rather reduce its incidence. Some studies however indicate that some antiepileptic drugs such as lamotrigine and carbamazepine, may increase the risk of SUDEP in females and certain individuals
- ❖ Vagal nerve stimulation
- ❖ Currently, the most effective strategy to protect against SUDEP in childhood epilepsy is seizure control, but this approach is not completely effective and is particularly challenging in cases of intractable epilepsy. The lack of generally recognized clinical recommendations available are a reflection of the dearth of data on the effectiveness of any particular clinical strategy, but based on present evidence, the following may be relevant:

- ❖ Epileptic seizure control with the appropriate use of medication and lifestyle counseling is the focus of prevention.(8)
- ❖ Detection of seizures using wristbands which can alert carers in case the wearer has stopped breathing or has a heart problem.
- ❖ Reduction of stress, participation in physical exercises, and night supervision might minimize the risk of SUDEP.
- ❖ Knowledge of how to perform the appropriate first-aid responses to seizure by persons who live with epileptic people may prevent death.
- ❖ People with arrhythmias associated with seizures should be submitted to extensive cardiac investigation to determine the indication for on-demand cardiac pacing.
- ❖ Successful epilepsy surgery may reduce the risk of SUDEP, but this depends on the outcome in terms of seizure control.
- ❖ The use of anti-suffocation pillows has been advocated by some practitioners to improve respiration while sleeping, but their effects remain unproven because experimental studies are lacking.
- ❖ Providing information to individuals and relatives about SUDEP is beneficial.
- ❖ Night time supervision.

### 1. Pre op CT angiogram of brain



(a)



(b)

### 2. Pre op CT brain



(a)



(b)



(c)

### 3. Post op CT brain POD 1



(a)



(b)



(c)

### 4. Post op CT brain POD 8



(a)



(b)



(c)

1. Pre op CT angiogram of brain (a)(b) showing Right saccular bilobed PICA aneurysm
2. Pre op CT brain (a)(b)(c) showing widespread subarachnoid haemorrhage with intraventricular and subtentorial haemorrhages, haematoma at the level of cervicomedullary junction mildly compressing the spinal cord and hydrocephalus
3. Post op CT brain (a)(b)(c) showing status post craniotomy and clipping
4. Post op CT brain showing (a)(b)(c) showing Post op status with evolving acute /subacute ischemic changes in the left watershed territory (MCA-PCA) and diffuse cerebral edema.

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