

Case report

□ **A case of possible sudden unexpected death in epilepsy in a patient affected by septic shock**

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SUMMARY: INTRODUCTION. Sudden unexpected death in epilepsy is defined as a sudden, nontraumatic and nondrowning death in absence of documented status epilepticus or an identifiable anatomic or toxicological cause for death at autopsy.

CASE REPORT. We report a patient with a possible sudden unexpected death in epilepsy at her first seizure. Our patient was an aged woman (85 years old) who was admitted in Department of Emergency for fever and stupor. She presented generalized myoclonic jerks after about six hours of the admission; after the generalised seizure she died. Clinical symptoms of myoclonic jerks, generalized seizure and death were registered by EEG and ECG simultaneously.

DISCUSSION. The pathophysiology of SUDEP is poorly understood, but the autonomic nervous system is suspected to be involved. In our case a notable finding was a terminal cessation of EEG activity before any cardiac changes suggesting an irreversible "cerebral electrical shutdown" and, possibly, reflecting profound central inhibition and subsequent ictal asystole. However, whether ictal asystole can lead to SUDEP remains an open question because a unitary explanation of the mechanism of SUDEP remains elusive. More cases are needed as they can contribute to our understanding of SUDEP mechanisms. Finally, our case confirms that SUDEP is mainly, but not exclusively, an issue for patients with chronic uncontrolled epilepsy.

KEY WORDS: Clinical case, Death, Epilepsy, EEG.

□ INTRODUCTION

SUDEP is defined as "sudden, nontraumatic and nondrowning death in absence of documented status epilepticus or an identifiable anatomic or toxicological cause for death at autopsy"⁽⁷⁾.

It is generally assumed that SUDEP is a risk only for those with intractable epilepsy, ignoring the group of individuals experiencing a first epileptic seizure⁽³⁾.

Notably, few studies examined patients who experienced SUDEP following their first seizure confirming that SUDEP is mainly, but not exclusively, an issue for

patients with chronic uncontrolled epilepsy^(4,8). The pathomechanism underlying SUDEP is still largely unknown, but the MORTEMUS study suggested an early postictal neurovegetative breakdown with initial GTCS triggering a short period of normal or increased heart and respiratory rates then severe bradycardia and central apnoea with post-ictal EEG suppression that is fatal in a third of cases⁽⁶⁾.

Here we report a patient who experienced a possible SUDEP following her first seizure that showed abrupt cessation of cerebral electrical activity followed by terminal asystole, thus suggesting an irreversible "cer-

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LIST OF ACRONYMS AND ABBREVIATIONS: ECG = ElectroCardioGram; EEG = ElectroEncephaloGram; GTCS = Generalized tonic-clonic seizure; MORTEMUS = MORTality in Epilepsy Monitoring Unit Study; SUDEP = Sudden Unexpected Death in Epilepsy.

bral electrical shutdown” or a post-ictal neurovegetative breakdown associated with the seizure as a possible cause of SUDEP.

□ CASE REPORT

In our short report we describe an aged woman (85 years old) who was admitted in Department of Emergency for fever and stupor. An ovarian cancer affected her for unknown time, complicated by septic shock. She had no cardiologic antecedents. A TC scan of brain showed mild spread bilateral vascular chronic lesions in internal and external capsules and thalami and symmetrical cortical-subcortical atrophy. Urgent laboratory tests evidenced an increase of white blood cells (22,500 x mmc), and VES (66 mm/h), procalcitonin (88.90 ng/ml). The temperature was 40.5 °C. At admission chest X ray had evidenced small thickening of lung of right lower lobe and of the left subclavicular with no clear evidence of inflammatory infiltrates.

She presented generalized myoclonic jerks after about six hours of the admission. The EEG recording was immediately undertaken using a digital EbNeuro (BE-light model), jasper montage, 7 µV/mm amplitude, 30 HZ filter; 0.01 sec. time constant.

The ECG recording was obtained from monitor that registered all vital parameters (body temperature, arterial pressure, oxygen saturation, cardiac frequency). At the beginning of the EEG activity, myoclonic jerks were registered with recurrent spike waves (Figure 1 A), after 100 seconds the patient presented generalized seizure lasted about 80-90 seconds (Figure 1 B), monitored by EEG with continuous diffused spike waves. After the seizure the patient presented bradycardia registered on ECG.

At the same period EEG examination showed slow wave delta records that gradually reduced the amplitude and, after 8 minutes, EEG become flat (flat line) (Figure 1 C and 1 D). At the beginning of the EEG registration, during seizure, ECG evidenced tachycardia (110 cardiac frequency media value, Q-T interval: 407 m/sec) and after the generalised seizure ECG rhythm rapidly changed in slowing its frequency (Q-T interval: 397 m/sec); and finally became a flat line (Figure 2 and 3). So we registered the death of the patient. Summarizing the clinical neurological status

with ECG and EEG registrations, the patient presented an absence of electroencephalographic activity a few seconds before highlighting absent ECG activity.

□ DISCUSSION

We report a case of possible SUDEP associated with a first GTCS in a patient affected by septic shock.

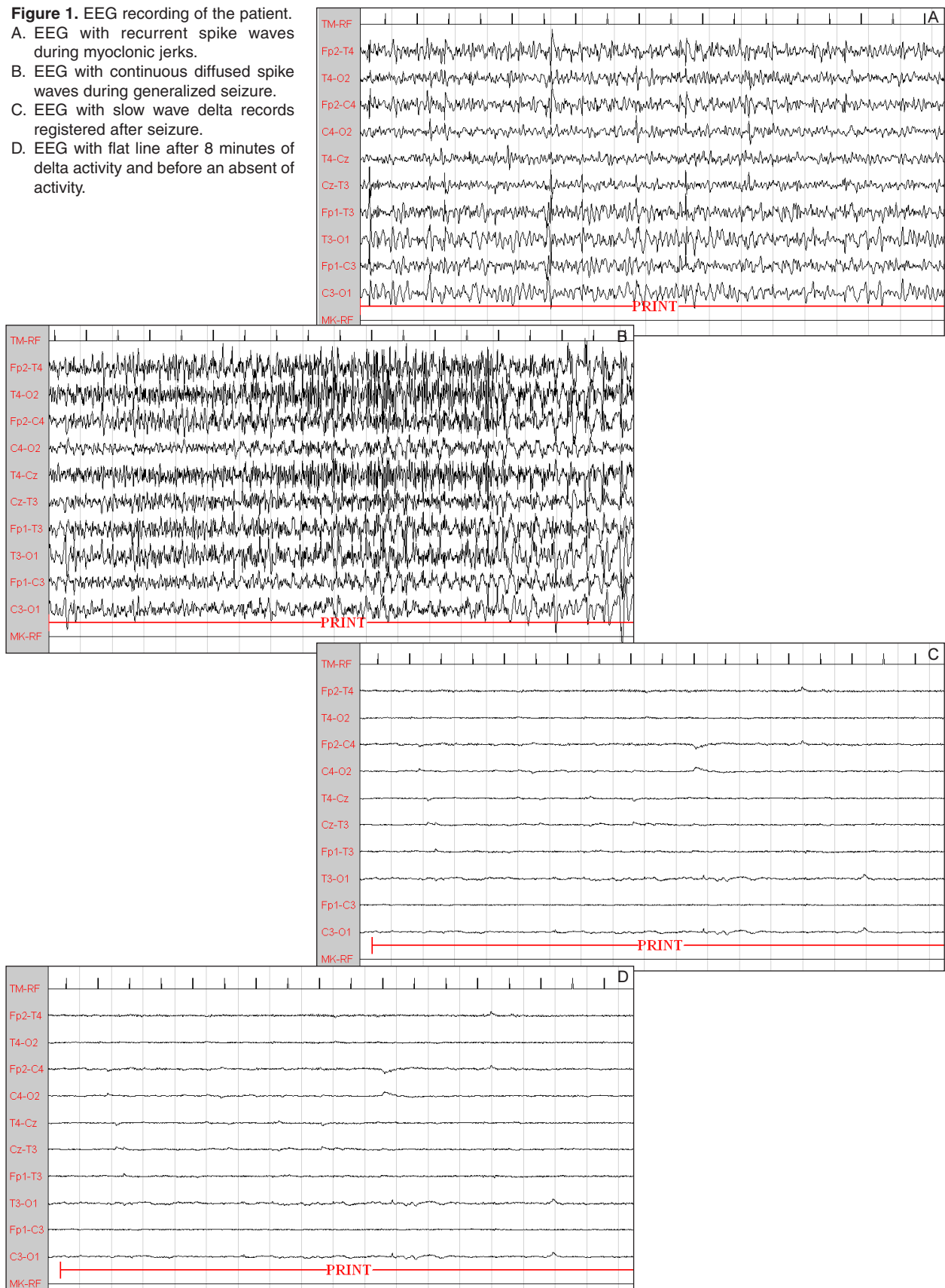
The pathophysiology of SUDEP is poorly understood, but the autonomic nervous system is suspected to be involved. Recently Freitas J. et al. have described that GTCS is the commonest seizure type associated with SUDEP⁽¹⁾. The GTCS type is relatively stereotyped, but the post-critical characteristics has both clinical and electroencephalographic heterogeneous.

Some patients quickly recover, while other have varying periods of postictal stupor. Some have brief periods of focal generalized EEG slowing, while others have prolonged post-ictal generalized EEG suppression and slowly recovering. Whether this prolonged post-ictal generalized EEG suppression may be independent risk factor for SUDEP is debated. Poh et al. demonstrated that GTCS with prolonged post-ictal generalized EEG suppression (> 20 seconds) had significantly higher sympathetic activation and greater vagal reduction than GTCS with short post-ictal generalized EEG suppression suggesting a strong correlation between the degree of autonomic dysregulation and duration of post-ictal generalized EEG suppression after GTCS for both sympathetic and parasympathetic nervous system⁽⁵⁾. In our case a notable finding was a terminal cessation of EEG activity before any cardiac changes suggesting an irreversible “cerebral electrical shutdown” and, possibly, reflecting profound central inhibition and subsequent ictal asystole⁽²⁾.

However, whether ictal asystole can lead to SUDEP remains an open question because an unitary explanation of the mechanism of SUDEP remains elusive.

In the end, in our case it may be the sum of a number of unfortunate concurrences that comprise a generalized tonic-clonic seizure in a senile brain, genetically or otherwise predisposed to unknown neurotransmitter-driven dysregulation of postictal arousal mechanisms, with cardiorespiratory homeostasis compromised by the septic shock and a subsequent cycle of cerebral and cardiorespiratory dysfunction that ends in a possible SUDEP.

Figure 1. EEG recording of the patient.
 A. EEG with recurrent spike waves during myoclonic jerks.
 B. EEG with continuous diffused spike waves during generalized seizure.
 C. EEG with slow wave delta records registered after seizure.
 D. EEG with flat line after 8 minutes of delta activity and before an absent of activity.



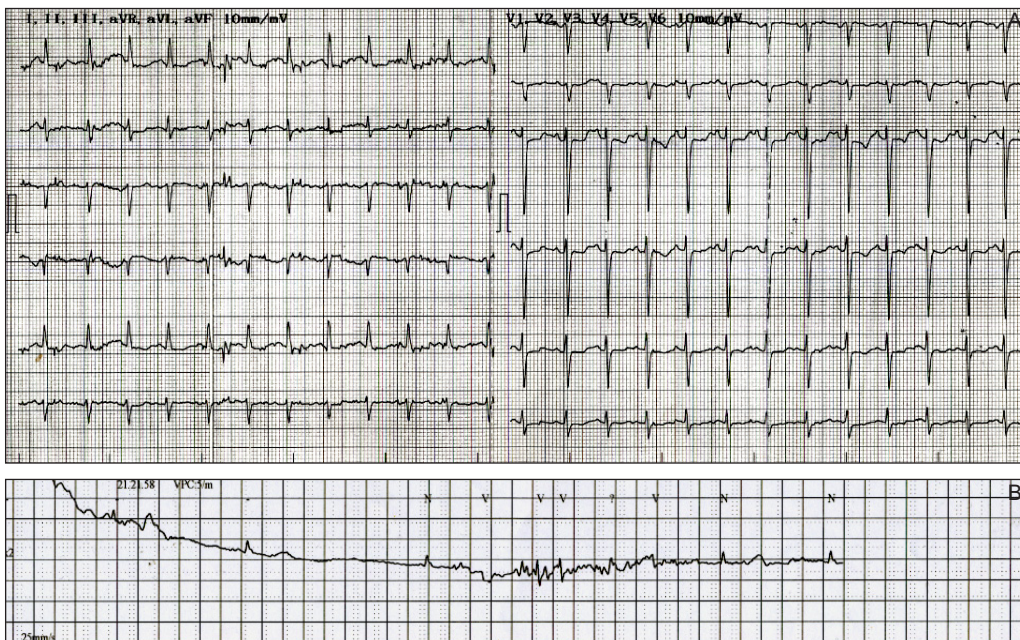


Figure 2. A. ECG with tachycardia during seizure. B. ECG slowing its frequency at the end of seizure.

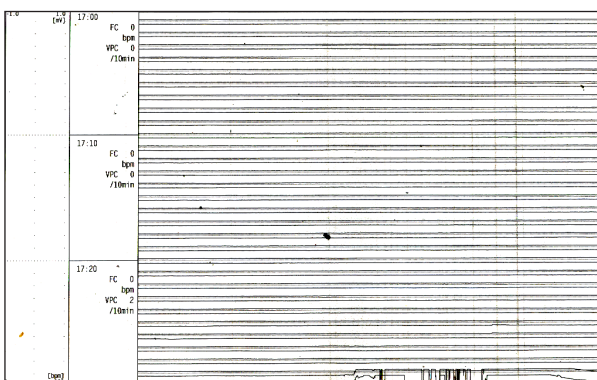


Figure 3. ECG as a flat line.

More cases are needed as they can contribute to our understanding of SUDEP mechanisms.

Finally, our case confirms that SUDEP is mainly, but not exclusively, an issue for patients with chronic uncontrolled epilepsy

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DISCLOSURE. The Authors declare no conflicts of interest.