Prevention of Sudden Death in Patients With Epilepsy

To the Editor: Sudden unexpected death in epilepsy (SUDEP) occurs sometimes in people with ictal disorders.[1] The diagnosis does not require evidence of a seizure and excludes status epilepticus as a cause of death. The postmortem examination must not reveal any toxic or anatomic pathology, and death must not be due to trauma or drowning.

The prevalence of epilepsy in the United States is approximately 2.3 million in adults and 480,000 among children.[1,2] One of 26 people in the United States and 1 of 103 people in Great Britain have epilepsy in their lifetime. The incidence of SUDEP is approximately 1–2 per 1,000 person years in patients with chronic epilepsy and 3–9 per 1,000 person years in severe, refractory seizures. There are more than 500 SUDEP deaths every year in Great Britain among the 600,000 individuals reported to have epilepsy.[2] Postmortem studies indicate that up to 60% of all epilepsy-related deaths can be accounted for by SUDEP, making it the most common epilepsy-related cause of mortality.[3]

The risk of dying suddenly and unexpectedly for patients with epilepsy has been estimated to be more than 20 times greater than for the general population.[1,2] The cause in two-thirds of the cases is unidentified. The most common risk factors for SUDEP are generalized tonic-clonic seizures, long-term epilepsy, male gender, polypharmacy, poor anticonvulsant drug compliance, and sleep deprivation.[4]

Case report. A 5-year-old girl, was treated in the pediatric neurology clinic. The diagnosis was severe, refractory seizures with a documented, recurrent myoclonic component of possible Dravet syndrome, according to *ICD-9* diagnostic criteria. She had exhibited this convulsive disorder since the age of 3 years. Her epilepsy was managed with divalproex, temazepam, and a ketogenic diet. Despite pharmacotherapy, the child was never seizure free.

One morning, the child was found dead in her bed. The family reported no new illnesses, change in ictal status, trauma, or other concerns recently. A postmortem examination revealed no toxic or anatomic pathology. The cause of death was considered to be SUDEP, noting a higher frequency of such occurrences in children with Dravet syndrome.[5]

Integration of different mechanisms is proposed to explain SUDEP: cardiac arrythmias, respiratory depression, and cerebral or autonomic dysfunction.[6] Neuronal networks activated during a seizure inhibit the respiratory centers in the medulla, leading to hypoventilation with an acid-base imbalance, which can induce bradycardia, asystole, and death.

Cardiorespiratory control by the brainstem is regulated via neurotransmitters like 5-hydroxytryptamine and adenosine.[6] There is a rise in adenosine levels during convulsive activity that causes postictal hypoventilation, apnea, and arrhythmias. Serotonin decreases postictal hypoxemia, and reportedly in an animal experiment, selective serotonin reuptake inhibitor medications attenuated this influence.[6]

Table 1. Prevention of Sudden Unexpected Death in Epilepsy[1–7]

Engage in regular physical activity
Diminish stress and anxiety
Decrease alcohol intake
Comply with antiepileptic drugs
Counsel the patient and family about sudden unexpected death in epilepsy
Provide nighttime seizure activity monitoring
Train family in cardiopulmonary resuscitation
Consider a vagus nerve stimulator for seizure suppression[5]

Talking about SUDEP with patients and family at the initial onset of seizures is controversial; rather, it is advised that it be discussed after patients are more established in the clinic.[7] Utilizing a SUDEP safety checklist can help clinicians in assessing the risk of sudden death in patients with epilepsy.[2] Discussion is for higher-risk individuals. A preventative plan is provided to patients and family, hopefully to improve patient compliance with medications and facilitate changes in lifestyle.[7] Such a plan targets diminishing prominent risk factors (Table 1).

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