AMERICAN EPILEPSY SOCIETY

AES POSITION STATEMENT ON SUDEP COUNSELING

The American Epilepsy Society (AES) recognizes that the majority of patients with epilepsy, families, and caregivers want to be informed of Sudden Unexpected Death in Epilepsy (SUDEP) risk by their healthcare providers. AES encourages counseling and offers guidance for providers by proposing an evidence-based approach to counseling patients, families, and caregivers about the risk of SUDEP. SUDEP counseling should be individualized within the patient-provider relationship, with consideration of a patient's epilepsy type, preferences, and psychosocial circumstances.

The American Academy of Neurology (AAN)/American Epilepsy Society (AES) 2017 practice guideline summary on SUDEP is the basis for AES guidance on SUDEP counseling, and tools based on that guideline are available for providers and patients.[1]

The AES also recognizes that evidence and understanding of this devastating entity is rapidly evolving. As further data become available, counseling recommendations may change.

1. SUDEP Definition and Incidence

SUDEP refers to unexpected, witnessed or unwitnessed, non-traumatic, and non-drowning death that occurs in an individual with epilepsy with or without evidence for a seizure (excluding documented status epilepticus), in which post-mortem examination does not reveal a toxicologic or anatomic cause of death.[2]

The cause of SUDEP is unknown but appears to be multifactorial; proposed mechanisms include cardiac arrhythmia, respiratory dysfunction, dysregulation of systemic or cerebral circulation, and seizure-induced hormonal and metabolic changes during and after seizures.[3]

The 2017 AAN/AES SUDEP practice guideline summary describes the risk of SUDEP for adults with epilepsy as small.[1] In one year, SUDEP typically affects 1 in 1,000 adults with epilepsy; in other words, annually, 999 of 1,000 adults with epilepsy will ***not*** be affected by SUDEP. SUDEP in children with epilepsy is thought to be less common. In one year, SUDEP typically affects 1 in 4,500 children with epilepsy; in other words, annually 4,499 of 4,500 children with epilepsy will ***not*** be affected by SUDEP.[1] However, some recent literature suggests it is more difficult to ascertain SUDEP incidence in children than previously appreciated and thus the incidence may be higher and similar to adults.[4,5] Further, in some specific childhood epilepsy syndromes, such as Dravet syndrome, incidence is known to be much higher.[6,8] In one study, up to 49% of deaths of patients with Dravet syndrome were due to SUDEP.[6]

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2. Risk Factors for SUDEP

The strongest SUDEP risk factors are the presence of generalized tonic-clonic seizures, particularly at high frequency (3 or more convulsive seizures per year).[1] Absence of seizure freedom is a reported risk factor, as is not escalating treatment in medically refractory epilepsy patients.[1] Seizures during sleep may also increase risk of SUDEP.[1]

There are many other potential risk factors that have been suggested. Evidence is minimal or incomplete for these risk factors, but providers may consider discussing them with selected patients and families. These include: male sex; epilepsy onset before 16 years of age; disease duration longer than 15 years; patients under 16 years of age with intellectual disability; structural brain lesion, or abnormal neurological exam; adults with alcohol use; and psychiatric comorbidities, particularly in female patients.[1,6,7,8,9]

3. Scenarios in Which SUDEP Counseling Should be Considered

- **Convulsive seizures, particularly if frequent**: Presence of seizures is a risk factor for SUDEP, and risk may increase with greater seizure frequency (3 or more per year).[1]
- **Dravet syndrome**: In this syndrome SUDEP is the leading cause of death.[6]
- **Seizures during sleep**: There are data suggesting nocturnal seizures may increase the risk of SUDEP.[1,6]
- **Patients with a history of non-adherence to treatments**: Individuals who are not adherent to medication treatments are at greater risk for seizures and therefore SUDEP.[1,7]
- **Patients with concerns about the risks of dying from epilepsy**: Overall, the risk of SUDEP is quite low, and patient and family anxiety may be reduced by understanding the risk factors and incidence.[1]
- **New epilepsy diagnosis**: Many people with epilepsy and parents of children with epilepsy report that they would like to be informed of SUDEP at time of epilepsy diagnosis or shortly afterwards.[10]

4. Recommended Approach and Content for Counseling

The extent and frequency of SUDEP counseling should be individually tailored to each patient and family as appropriate to the specific epilepsy type, patient preference, caregiver availability/engagement, and living environment. Survey data demonstrate that the majority of patients with epilepsy and their families prefer to be informed about SUDEP risk by their providers.[11] The following potential messages and considerations may be helpful to providers when individualizing SUDEP counseling.[7,10]

- **What is SUDEP?** *SUDEP is the sudden, unexpected, non-traumatic, non-accidental death of a patient with epilepsy that cannot be explained by another cause of death. SUDEP most often occurs at night and may or may not be in the context of a known seizure. The mechanisms of SUDEP are not fully understood but are considered to be multifactorial*.[1]

- **Who is at highest risk?** *SUDEP is uncommon. In adults, 1 in 1000 adults will be affected per year; in other words, 99.9% of adults will* ***not*** *be affected annually. In children, 1 in 4,500 children will be affected per year; 99.99% of children will* ***not*** *be affected annually.[1] However, some recent studies suggest that the incidence in children may be higher and similar to that in adults. The greatest overall risk factor for SUDEP among people with epilepsy is frequent (3 or more per year) generalized tonic-clonic seizures. Seizures during sleep may also increase risk*.[1,6] *In certain syndromes, such as Dravet syndrome, SUDEP risk rises dramatically.[6]*

  The chief goal of risk counseling about SUDEP is to educate, in order to support and encourage possible risk reduction for those at high risk, and to alleviate potentially inappropriate fear of SUDEP for those with low risk. This requires a personalized approach for each patient.

- **What can be done?** *Seizure freedom decreases the risk of SUDEP. The most effective measures to decrease SUDEP risk are to prevent generalized tonic-clonic seizures and/or attain seizure freedom. Therefore, patients with epilepsy should take their antiseizure medication as prescribed and avoid any recognized seizure triggers.*

  Providers should be aggressive in the treatment of medically refractory epilepsy, including referral to a specialized epilepsy center when appropriate.

  Despite limited evidence, patients with epilepsy and their families may wish to consider nighttime supervision or use of seizure detection devices, if appropriate and feasible. [1,7,12,13] *The value of such devices (e.g. seizure detection wearables) and anti-suffocation pillows remains unproven. Even emergent medical intervention does not always preclude the occurrence of SUDEP*.[12]

  Counseling should attempt to match patient risk with the potential benefit of selected interventions, balanced against the emotional or practical impact of implementation on the patient or their family. For example, the limited potential benefit of co-sleeping with a grown child for some families is outweighed by the psychological need for independence, but for other families results in anxiety reduction that is a significant emotional benefit. The lack of evidence to support SUDEP benefit for any particular intervention underscores the need for highly individualized counseling.

- **What is the future?** *Much research is underway to understand the cause of SUDEP and to identify effective methods of SUDEP prevention. Recommendations and counseling will evolve as new data become available*.

5. Methods for Counseling and Available Resources

Most patients and families would like to be informed about SUDEP at the time of epilepsy diagnosis or shortly afterwards.[1] The optimal timing and setting of counseling should be determined on a case-by-case basis. While the most effective method of counseling has not been established, patients and caregivers have endorsed a preference for conversation with their providers followed by provision of written materials (for example, *AAN/AES Summary of Practice Guideline for Patients and their Families*. https://www.aan.com/Guidelines/home/GetGuidelineContent/851).[1,10]

Many patient advocacy groups recommend counseling about SUDEP and offer formal statements and individual support through their websites.

This Position Statement was developed by a work group comprised of AES Practice Management Committee Chair, Gabriel Martz, MD; Committee Members Susan Duberstein, MD, Chloe Hill, MD, and Marianna Spanaki-Varelas, MD; and patient advocate representative Tom Stanton, Executive Director, Danny Did Foundation. The statement was approved by the AES Practice Management Committee on September 16, 2019; approved by the AES Council on Clinical Activities on September 16, 2019; and approved by the AES Board of Directors on October 8, 2019.

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