EPILEPSY PATIENTS AND THEIR FAMILIES SHOULD ROUTINELY BE TOLD ABOUT THE RISK OF SUDEP

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SUDEP has been defined as: sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning deaths in patients with epilepsy, with or without evidence of a seizure, and excluding documented status epilepticus, in which postmortem examination does not reveal a toxicologic or anatomic cause of death (Nashef 1997). When witnessed, SUDEP has usually occurring during or immediately after a tonic-clonic seizure. The frequency varies depending on the severity of the epilepsy, but overall the risk of sudden death is over 20 times higher among people with epilepsy than in the general population (Ficker et al . 1998). A recent study from Finland demonstrated that over a 40-year follow-up of subjects diagnosed with epilepsy in childhood, the risk of SUDEP was 7% (Sillanpää and Shinnar, 2010). Hence, SUDEP is clearly the leading epilepsy-related cause of death among people with epilepsy.

The risk of SUDEP varies almost 100-fold, depending on the type of epilepsy population. The lowest incidence rates, 0.09 to 0.35/1,000 person-years, have been reported from unselected cohorts of incident cases of epilepsy. Rates in general epilepsy prevalence populations have ranged between 0.9 to 2.3/1,000 person-years, in chronic refractory epilepsy between 1.1 to 5.9/1,000 person-years, and in epilepsy surgery candidates or patients who continue to have seizures after surgery, 6.3-9.3/1,000 person-years (Tomson et al 2008). In patients with chronic refractory epilepsy attending epilepsy referral centres, SUDEP has been the leading cause of premature death accounting for 10-50% of all deaths.

A pooled analysis of risk factors in case-control studies show that the higher the frequency of tonic-clonic seizures, the higher is the risk of SUDEP, and risk are also elevated in males, those with long duration epilepsy and those on antiepileptic polytherapy.

SUDEP usually occurs when the seizures are unwitnessed and often at night. There are probably a number of different mechanisms, but most research has focused on seizure-related respiratory depression, cardiac arrhythmia, cerebral depression and autonomic dysfunction.

Can SUDEP be prevented? Although controlled prospective studies are lacking, it is reasonable to assume that improved control of tonic-clonic seizures (by means of a more effective pharmacological or surgical treatment, enhanced compliance, and life style adjustments) could reduce SUDEP risk. Over-night supervision of high risk patients might also be protective.

Patients should generally be fully informed about the risks of any condition or its treatment, and it is odd if SUDEP should be an exception. Nevertheless a survey of British Neurologists showed that only 4.7% discussed SUDEP with all their epilepsy patients, 25.6% with a majority of patients, 61.2% with 'few of their patients', and 7.5% with none of their patients. Patients and relatives have the right to know even more so as it is likely that the SUDEP risk can be modified by different interventions. The relevant question is when and how, rather than if. This author's opinion is that the information is best provided early and as part of a comprehensive counseling concerning general risks and prevention.

REFERENCES

1. Nashef L. Sudden unexpected death in epilepsy: terminology and definitions. Epilepsia 1997; 38 (Suppl. 11), S6-S8.

2. Ficker DM, So EL, Shen WK, Annegers JF, O'Brien PC, Cascino GD, et al. Population-based study of the incidence of sudden unexplained death in epilepsy. Neurology. 1998 November 1, 1998;51(5):1270-4.

3. Sillanpaa M, Shinnar S. Long-term mortality in childhood-onset epilepsy. N Engl J Med. 2010 Dec 23;363(26):2522-9.

4. Tomson T, Nashef L, Ryvlin P. Sudden unexpected death in epilepsy: current knowledge and future directions. Lancet Neurol. 2008 Nov;7(11):1021-31.

5. Hesdorffer DC, Tomson T, Benn E, Sander JW, Nilsson L, Langan Y, et al. Combined analysis of risk factors for SUDEP. Epilepsia. 2011.

*6. Morton B, Richardson A, Duncan S. Sudden unexpected death in epilepsy (SUDEP): don't ask, don't tell? J Neurol Neurosurg Psychiatry 2006;**77**:199-202.*