

Short Communication

AWARENESS OF SUDDEN UNEXPECTED DEATH IN EPILEPSY AMONG NEUROLOGISTS IN LATVIA

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Communicated by Aivars Lejnīks

Sudden unexpected death in epilepsy (SUDEP) is a common cause of mortality in patients with epilepsy, but it is unknown how neurologists disclose this risk when counselling patients. This study was aimed at examining SUDEP discussion practices of neurologists in Latvia, as well as the awareness of the syndrome. Two hundred questionnaires were distributed, and we received 84 responses. We found that the majority of Latvian neurologists (79.0%) do not inform their patients of SUDEP, which is opposite to the findings in other countries. Despite the existing practice, 93.1% of neurologists believed that patients should be informed about SUDEP. A partial explanation for not discussing the negative aspects of epilepsy is that 75.3% of caregivers believe that being informed about SUDEP would cause permanent anxiety in patients, whereas 69.4% believe that it would improve compliance. This study revealed average awareness of SUDEP risk factors and warrants further studies for in-depth analysis of existing counselling practice.

Key words: epilepsy, SUDEP, truth telling.

People with epilepsy have a two-to-threefold higher mortality rate than the general population, because of the epilepsy itself (epilepsy-related death) or the underlying cause of epilepsy (Mohanraj *et al.*, 2006). Sudden unexpected death in epilepsy (SUDEP) is defined as the sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning death of a person with epilepsy with or without a seizure, excluding documented status epilepticus, and in whom post-mortem examination does not reveal a structural or toxicological cause of death (Nashef, 1997). SUDEP is the most important (Tomson *et al.*, 2005) and the most common (Duncan and Brodie, 2011) direct epilepsy-related cause of death. A population-based incidence cohort of epilepsy (Ficker *et al.*, 1998) reported incidence of 0.35 cases per 1000 person-years, but the incidence can be as high as 9 cases per 1000 person-years in chronic refractory epilepsy patients who are candidates for epilepsy surgery (Dasheiff, 1991). While the physiological mechanisms underlying SUDEP are poorly understood, evidence from epidemiologic, observational, clinical, and pathological studies strongly suggests that in most cases, sudden unexpected death in epilepsy occurs after a seizure, usually a tonic-clonic seizure (Devinsky, 2011). While it appears that seizure control reduces the risk of SUDEP, there are no clear additional prevention strategies (Friedman *et al.*, 2014), although some measures, such as altered sleeping arrangements and seizure alarms, have been suggested to reduce

the risk (Hitiris *et al.*, 2007). Self- and family-management of epilepsy is critical to seizure control. Therefore, patients and families should be informed about SUDEP and the importance of seizure control in potentially preventing SUDEP (Miller *et al.*, 2014). There is controversy regarding whether to inform and counsel patients with epilepsy and their caregivers about SUDEP (Friedman *et al.*, 2014). Reasons in favour of informing the patient are based on the evidence that awareness of the increased risk may promote patient and parent cooperation with therapy (Vegni *et al.*, 2011).

Discussing SUDEP with patients and family members is recommended by epilepsy care guidelines (Anonymous, 2012), yet there are no clear recommendations in terms of how, or when, these discussions should take place. Information on SUDEP may not alter the outcome and, conversely, disclosure to patients with a low risk of SUDEP (e.g., well-controlled seizures) may lead to unnecessary distress. A survey of 383 UK neurologists found that 5% discussed SUDEP with all patients, 26% with a majority, 61% with a few, and 7.5% with none (Morton *et al.*, 2006). A study examining practices among the U.S. and Canadian epileptologists found that 6.8% discussed SUDEP with all of their patients, 14.0% discussed it most of the time, 33.4% of neurologists reported discussing SUDEP sometimes, and 30.0% rarely, while 11.6% reported never discussing

SUDEP with patients or their caregivers (Friedman *et al.*, 2014). The purpose of this study was to describe the practices and preferences of neurologists and epileptologists regarding discussing SUDEP with their patients in Latvia, as well as the awareness of caregivers about the syndrome.

Study questionnaire. We performed an electronic, web-based survey to assess awareness and experience with SUDEP among Latvian neurologists, conducted in the period January–March 2015. We sent a questionnaire to all practising neurologists in Latvia listed in the Society of Latvian Neurologists database and to all current residents in neurology. The paper version of the questionnaire was also distributed at a conference of the Society of Latvian Neurologists.

The survey questionnaire contained demographic information about the respondents, including their gender, age, and practice (adult or child neurology; Inpatient Clinic and/or Outpatients Department; practicing in the capital city Rīga or in a regional medical institution).

Respondents were asked to choose the correct definition of SUDEP from the following given answers: “Sudden unexpected death in epilepsy in otherwise healthy individuals”; “Sudden unexpected death in epilepsy due to cardiac pathology”; “Sudden unexpected death in epilepsy due to trauma, drowning, choking”; “Sudden unexpected death in epilepsy due to status epilepticus”). Then they were asked to rate their subjective evaluation of knowledge about SUDEP with the answers: “good”, “minimal and insufficient”, and “none”). Respondents were also asked whether they discuss SUDEP with patients (if yes, then whether with all patients or particular groups of patients), and when should SUDEP be discussed (answers: “when diagnosis of epilepsy is confirmed”, “only if the patient has known risk factors for SUDEP”, and “never”). The awareness of known clinical factors and lifestyle factors that increase risk for SUDEP was assessed by the question “Which groups of patients should be informed about SUDEP?”, asking to choose from a list of items that included:

- eight previously described risk factors — seizures at night (Langan *et al.*, 2005; Lamberts *et al.*, 2012), three antiepileptic drugs (AEDs) (Nilsson *et al.*, 1999; 2001; Walczak *et al.*, 2001), subtherapeutic medication doses (Opeskin *et al.*, 2000), non-compliance with medication (Langan, 2000; Lear-Kaul *et al.*, 2005; Williams *et al.*, 2006), drug-resistant epilepsy (Annegers and Coan, 1999; Hesdorffer *et al.*, 2011), age 20–40 years (Langan, 2000; Tomson *et al.*, 2005), alcohol consumption (McGugan, 1999; Opeskin *et al.*, 2000), generalized tonic-clonic seizures (Langan, 2000; Walczak *et al.*, 2001; Ryvlin *et al.*, 2013);
- 16 items with no known influence on the risk of SUDEP, or with no increased risk (e.g. atrial fibrillation, hypertension, smoking, well-controlled epilepsy); and

- two factors for which all answers are correct — gender (Walczak *et al.*, 2001; Lear-Kaul *et al.*, 2005), and different seizure frequencies (Nilsson *et al.*, 1999; Walczak *et al.*, 2001).

Subjective feeling of influence on quality of life and compliance was also assessed, by the questions: “Do you think being informed about SUDEP may cause permanent anxiety in epilepsy patients?”, “Do you think awareness of SUDEP may have negative influence on the quality of life by causing fundamental lifestyle changes in patients?”, “Please, rate your beliefs of influence of being informed about SUDEP in the following aspects: improvement of compliance; effectiveness of epilepsy treatment; quality of life changes.” The question “should patients decide how much information on SUDEP to receive” was also addressed.

Descriptive statistics were used for general, professional data, and for closed questions. For the purpose of statistical analysis, we divided all respondents into two categories: a) respondents who rated their knowledge about SUDEP as good; b) respondents who rated their knowledge as minimal and insufficient or had no information about the syndrome.

The Chi-square test was used to compare distributions of demographic and professional data between the groups with good and poor knowledge about SUDEP. The Mann–Whitney rank-sum test was used to compare mean age between the groups.

Summary of findings. Of the 200 questionnaires distributed, we received 84 responses (42% response rate). There were 12 partially completed questionnaires, which was taken into account during data analysis. Of the 84 responders 19 were males and 65 were females. Mean age of respondents was 41.9 years (23 to 79 years).

The majority of respondents (55.9%, $n = 47/84$) identified themselves as adult neurologists, 18 were child neurologists (21.4%) and 19 were postgraduate trainees in neurology (22.6%). 33.3% of respondents were working in an Inpatient Clinic, 34.5% in an Outpatients Department, and 32.1% in both. Most of the respondents (77.4%) were practicing in the capital city Rīga and 22.6% in a regional medical institution.

Most of the respondents (76.8%, $n = 63/82$) rated their subjective feeling about knowledge of SUDEP as minimal and insufficient, 13.4% ($n = 11$) as good, and 9.8% ($n = 8$) did not have any knowledge about the syndrome. Two respondents did not rate their level of knowledge. Sixty-one of 81 respondents chose the correct definition of SUDEP from the given samples. The majority of neurologists (79.0%, $n = 64/81$) did not inform their patients of SUDEP, 3.7% ($n = 3$) of neurologists informed all epilepsy patients, and 17.3% ($n = 14$) informed individual groups of patients.

To analyse the opinion of the respondents on how their patients should be counselled about SUDEP, we excluded responses of the neurologists ($n = 8$) who were not aware of the syndrome. Of 72 neurologists who responded to the

question “When should SUDEP be discussed?”, 41.7% (n = 30) considered that discussion should occur when the diagnosis of epilepsy was confirmed, 51.4% (n = 37) thought that patients should only be informed if there were known risk factors for SUDEP, and 6.9% (n = 5) considered that SUDEP should not be discussed. 93.1% (n = 67) of neurologists believed that SUDEP should be discussed with patients at some point during the course of the disease or under specific circumstances. Only the respondents who considered that SUDEP should be discussed (n = 67) were included in the analysis of risk factors of SUDEP. When asked which groups of patients should be informed about SUDEP, there were confirming answers for the following previously described risk factors: age 20–40 years (56.7%, n = 38), alcohol consumption (59.7%, n = 40), generalized tonic-clonic seizures (62.7%, n = 42), number of seizures 1–2/month (7.5%, n = 5) number of seizures 2–4/month (19.4%, n = 13), number of seizures 4/month (55.2%, n = 37), seizures at night (47.8%, n = 32/67), male gender (50.7%, n = 34/67), female gender (29.9%, n = 20), three AEDs used (44.8%, n = 30), subtherapeutic medication doses (28.4%, n = 19), non-compliance with medication (55.2%, n = 37), and drug-resistant epilepsy (50.7%, n = 34). The majority of respondents (91.0%, n = 61/67) identified at least one risk factor.

Respondents (n = 72) rated their beliefs of impact on the course of epilepsy and quality of life after being informed about SUDEP. Most neurologists (69.4%) thought that compliance would improve, 11.2% thought that compliance would not improve, and 19.4% did not know. Rating effectiveness of epilepsy treatment, 47.2% believed that it would improve, 22.2% that it would not improve and 30.5% did not know. Regarding quality of life, 47.2% of doctors believed that there is an influence on quality of life, 13.8% did not think so, and 38.8% did not know. The majority of 73 respondents (75.3%) thought that being informed about SUDEP might cause permanent anxiety in epilepsy patients, and the others (24.7%) believed that awareness would cause no anxiety. Almost half of the 72 respondents (48.6%) believed that awareness of SUDEP might have a negative influence on the quality of life by causing fundamental lifestyle changes in patients. Only 18.3% of 71 neurologists believed that the negative influence on quality of life is a sufficient reason for not informing a patient about SUDEP. The majority of 74 respondents (72.9%) agreed that a patient has the right to choose the amount of information they receive about SUDEP.

To determine which demographical or professional factors influenced the level of knowledge about SUDEP, we compared data from respondents with good knowledge about the syndrome, with those who rated their knowledge as minimal and insufficient or had no information about SUDEP. There was no statistically significant difference in distribution of occupation (neurologist, child neurologist or post-graduate student) between the group of respondents with good knowledge and the poor knowledge group ($p > 0.05$). There was no significant difference ($p > 0.05$) in

self-rated level of knowledge between respondents who were practicing in the capital city Riga and in regional medical institutions. The proportion of respondents practicing at outpatient clinic only was not statistically different ($p > 0.05$) between both groups. Mean age in the group of respondents with good knowledge was 46.1 years, and 40.6 years in the group with poor self-rated level of knowledge ($p > 0.05$).

There are 221 registered adult neurologists in Latvia, and 46 child neurologists (11 with certification in both adult and child neurology) (Anonymous, 2013), and about 25 active postgraduate trainees in neurology. Our survey covered the opinions of 29.9% of all Latvian neurologists and trainees. This study suffers from many limitations, in particular due to the small study sample, and a relatively high proportion of postgraduate trainees in neurology and child neurologists, which does not represent the actual distribution among neurology specialists in Latvia. In contrast to the findings of surveys analysing SUDEP-discussion practices in the UK (Morton *et al.*, 2006), the U.S. and Canada (Friedman *et al.*, 2014), where only a small proportion of respondents (7.5% and 11.6%, respectively) never discuss SUDEP with their patients, only 21% of neurologists in Latvia inform their patients about the risk of SUDEP.

While the self-rated knowledge of respondents on the risk factors of SUDEP was low, and the majority of neurologists rated their knowledge as minimal and insufficient, most of respondents were aware of the SUDEP definition, and every risk factor included in the questionnaire was known to approximately half of the responders. Further studies are warranted to provide a rational basis for not discussing SUDEP with patients and family members, since despite the existing awareness and the opinion of almost all health-care providers (93.1%) that SUDEP should be discussed, it is not common practice in Latvia to inform patients about the risk. One explanation for not discussing the negative aspects of epilepsy is that most doctors (75.3%) believe such information would cause permanent anxiety, whereas 69.4% believe that it would improve compliance. This finding is consistent with the previously described reasons for not disclosing SUDEP risk (Vegni *et al.*, 2011; Beran *et al.*, 2004). Another aspect for not discussing SUDEP can be at least partially attributed to the local cultural and medical traditions of avoiding proactive addressing of negative information. We were able to demonstrate differences between practices of discussing SUDEP in Eastern Europe and in the Western European countries, the U.S. and Canada. The design of our questionnaire allowed determining groups of patients with increased risk of SUDEP, the majority of responders considered it was not important to discuss the risks with. For example, 1–2 seizures a month were regarded as a risk factor by 7.5% of respondents only, and female gender by 29.9% versus 50.7% in males. The results of our study show that there is a low proportion of respondents who discuss SUDEP with their patients, which together with average awareness of risk factors would be of importance when training about SUDEP is addressed. Statistical analysis did indicate differences in occupation (neu-

rologist, child neurologist or post-graduate student), localisation of practice (Riga or regional medical institution), type of practice (outpatient clinic only versus outpatient plus inpatient clinic) or age between the group of responders who rated their knowledge as good and the group who rated their knowledge as poor. Our results warrant further research.

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Received 28 August 2015

NEIROLOGU INFORMĒTĪBA PAR PĒKŠŅAS, NEGAIĀTAS NĀVES EPILEPSIJAS LAIKĀ SINDROMU LATVIJĀ

Pēkšņas, negaidītas nāves epilepsijas laikā sindroms (SUDEP) ir biežs mortalitātes iemesls epilepsijas pacientiem, tomēr nav zināms, kā neirologi, konsultējot pacientus, informē par šī sindroma risku. Šī pētījuma mērķis ir izvērtēt Latvijas neirologu informētību par SUDEP praksi. Tika izplatītas 200 aptaujas anketas un saņemtas 84 atbildes. Tika atklāts, ka vairums neirologu (79,0%) neinformē pacientus par SUDEP, un tas ir pretēji citu valstu ziņotajai praksei. Pretēji pastāvošajai informēšanas kārtībai, 93,1% aptaujāto Latvijas neirologu uzskata, ka pacienti par SUDEP ir jāinformē. Vismaz daļējs skaidrojums epilepsijas negatīvo aspektu neatklāšanai pacientiem ir tas, ka 75,3% ārstu uzskata, ka šāda informācija radītu pastāvīgu trauksmi, lai gan 69,4% uzskata, ka uzlabotos līdzestība. Šis pētījums atklāj viduvējas zināšanas par SUDEP riska faktoriem un ir pamatu iedrošināt veikt turpmākus pētījumus ar epilepsijas pacientu padziļinātu konsultēšanas prakses analīzi.