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Sudden unexpected death in epilepsy: Knowledge and experience among U.S. and Canadian neurologists

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ABSTRACT

Importance: Sudden unexpected death in epilepsy (SUDEP) is a common cause of mortality in patients with the disease, but it is unknown how neurologists disclose this risk when counseling patients.
Objective: This study aimed at examining SUDEP discussion practices of neurologists in the U.S. and Canada.
Design: An electronic, web-based survey was sent to 17,558 neurologists in the U.S. and Canada. Survey questions included frequency of SUDEP discussion, reasons for discussing/not discussing SUDEP, timing of SUDEP discussions, and perceived patient reactions. We examined factors that influence the frequency of SUDEP discussion and perceived patient response using multivariate logistic regression.
Participants: The participants of this study were neurologists who completed postgraduate training and devoted >5% of their time to patient care.
Results: There was a response rate of 9.3%; 1200 respondents met eligibility criteria and completed surveys. Only 6.8% of the respondents discussed SUDEP with nearly all (>90% of the time) of their patients with epilepsy/caregivers, while 11.6% never discussed it. Factors that independently predicted whether SUDEP was discussed nearly all of the time were the following: number of patients with epilepsy seen annually (OR = 2.01, 95% CI = 1.20–3.37, $p < 0.01$) and if the respondent had a SUDEP case in the past 24 months (OR = 2.27, 95% CI = 1.37–3.66, $p < 0.01$). A majority of respondents (59.5%) reported that negative reactions were the most common response to a discussion of SUDEP. Having additional epilepsy/neurophysiology training was associated with an increased risk of a perceived negative response (OR = 1.36, 95% CI = 1.02–1.82, $p = 0.038$), while years in practice (OR = 0.85, 95% CI = 0.77–0.95, $p < 0.005$) and seeing both adults and children were associated with a decreased likelihood of negative response (OR = 0.15, 95% CI = 0.032–0.74, $p = 0.02$).
Conclusions: U.S. and Canadian neurologists rarely discuss SUDEP with all patients with epilepsy/caregivers though discussions are more likely among neurologists who frequently see patients with epilepsy or had a recent SUDEP in their practice. Perceived negative reactions to SUDEP discussions are common but not universal; more experienced neurologists may be less likely to encounter negative reactions, suggesting that there may be ways to frame the discussion that minimizes patient/caregiver distress.

1. Introduction

Sudden unexpected death in epilepsy (SUDEP) is the leading disease-related cause of mortality among people with seizure disorders [1]. Previously called sudden unexplained death, SUDEP is defined as a sudden and unexpected nontraumatic or nondrowning-related death in a person with epilepsy which may or may not associated with a recent seizure [2]. The incidence of SUDEP varies by several orders of magnitude depending on the population studied; it ranges from 0.09 to 1.2/1000 person-years in the general population with epilepsy, from 1.1 to 5.9/1000 person-years in patients with medically refractory epilepsy, and from 6.3 to 9.3/1000 person-years in patients who are epilepsy surgery candidates [1,3]. Sudden unexpected death in epilepsy rates are low in children with epilepsy, with reported rates of 0.1–0.4 per 1000 person-years [3]. The mechanisms of SUDEP remain uncertain though frequent seizures, especially generalized tonic–clonic seizures (GTCSs), are the greatest risk factor [4]. While it appears that seizure control reduces the risk of SUDEP, there are no clear additional prevention strategies.

There is controversy whether or not to inform and counsel patients with epilepsy and their caregivers about SUDEP, a typically rare outcome. Some organizations such as the National Institute for Health

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and Clinical Excellence (NICE) in the United Kingdom [5] and the Scottish Intercollegiate Guidelines Network (SIGN) [6], advocate that all patients with epilepsy be counseled about SUDEP as part of essential education about their disorder. Other authors suggest a more individualized approach such as waiting to discuss SUDEP when patients are ready to receive the information or when discussing antiepileptic drug (AED) compliance or epilepsy surgery [7,8] since disclosure to patients with a low risk of SUDEP (e.g., well-controlled seizures) or low-risk syndromes (e.g. childhood absence epilepsy) may lead to unnecessary distress. Surveys of physicians in the UK [9,10] and Italy [11] suggest that most physicians do not discuss SUDEP with all of their patients. A survey of 383 UK neurologists found that only 4.7% discussed SUDEP with all of their patients with epilepsy, 61.2% discussed SUDEP with few of their patients, and 7.5% never discussed SUDEP [9]. One-third of the respondents felt that the discussion leads to anxiety though, interestingly, neurologists with a special interest in epilepsy had less negative reactions to the discussion. A more recent survey of 46 pediatric neurologists in the UK found that only 20% provided SUDEP information to all of their patients/caregivers [10]. The same study found that most parents (91%) wanted to know about SUDEP often at diagnosis.

In the US and Canada, there are no national guidelines regarding SUDEP discussion. A recent statement by the American Epilepsy Society/Epilepsy Foundation joint task force on SUDEP [8] as well as the recent Institute of Medicine report on epilepsy [12] provide some guidance on discussing SUDEP, stating that SUDEP should be discussed in the context of comprehensive education about epilepsy. However, there is no accepted policy or consensus among the general neurology community in the US and Canada regarding this matter. Neurologists provide the majority of epilepsy care, and their understanding of SUDEP and current practices of SUDEP are unknown. Furthermore, reasons why neurologists do and do not discuss SUDEP are not understood. Therefore, we undertook a survey of neurologists in the US and Canada to assess knowledge and experience with SUDEP. We examined the frequency of SUDEP discussion, reasons for discussing and not discussing SUDEP, as well as the respondents' understanding SUDEP risk factors.

2. Methods

We performed an electronic, web-based survey to assess knowledge and experience with SUDEP among US and Canadian neurologists in October 2011. A copy of the survey is included in the Supplementary materials (Supplement 1). An invitation to participate in the survey was sent to 17,558 unique e-mail addresses of neurologists obtained through the Epilepsy Therapy Project. Subjects were instructed to complete the survey if they were neurologists who devoted >5% of their time to clinical care and had completed postgraduate training. A link to opt out of the survey was also included in the invitation. Two rounds of reminder e-mails were sent 2–3 weeks apart to subjects. The survey was performed using Limequery (limequery.org).

Survey questions included demographic information about the respondents including their practice (adult and/or child neurology; academic versus private practice), additional training in epilepsy or clinical neurophysiology, years in practice (0–5, 5–10, 10–15, or >15), and average number of patients with epilepsy followed per year (1–10, 11–20, 21–50, 51–100, or >100). To examine knowledge about SUDEP, respondents were asked to identify known SUDEP risk factors at the time of survey distribution from a list of seven items. A composite knowledge score was determined by subtracting the number of identified incorrect items (sleeping on two or more pillows; EKG showing QTc at the upper limit of normal; nocturnal complex partial seizures) from the number of identified correct items (treatment with 3 or more AEDs, lack of AED therapy, and recent GTCSs); the possible scores ranged from −3 to 4. This score is not validated, but the questions were determined based on the currently available literature.

Subjects who responded that they followed at least one patient with epilepsy per year were asked about their experience with SUDEP and discussing SUDEP with patients and caregivers. They were asked how many of their adult (>18 years old) and pediatric (<18 years old) patients had died from definite or probable SUDEP (using the definition of Nashef [2]) in the past 24 months. Respondents were asked how often they discussed SUDEP with patients or caregivers, and, if they discussed SUDEP, respondents were asked when they discussed it and what clinical factors and lifestyle factors influenced who they discuss SUDEP with. Respondents were also asked whether patient age factored in to their discussion of SUDEP and, if so, what patient age groups warranted a discussion of SUDEP (0–8, 9–16, 17–21, 22–54, or >55). Respondents who discussed SUDEP were also queried about their three most common reactions from patients and caregivers. Free text responses under the *other* response category were subsequently categorized into negative (e.g., anger, despair, and sadness), neutral (e.g., confusion, concern, and denial), and positive (e.g., appreciation, motivation to comply, inquisitiveness, and trust) responses for use in further analysis. Respondents were also asked about reasons for not discussing SUDEP with patients and if they felt knowledge of SUDEP would improve compliance with AEDs. We also inquired about tools that respondents would use to support and educate patients and caregivers around the issue of SUDEP.

In addition to the descriptive statistics of the responses, we examined which practitioner factors were independently associated with discussing SUDEP nearly all the time. Using multivariate logistic regression (SAS, Cary, NC), we examined whether having additional training in epilepsy/clinical neurophysiology, being in academic or private practice, number of years in practice, <100 versus >100 patients with epilepsy seen per year, having a patient die from SUDEP in the past 24 months, or being knowledgeable about SUDEP (defined as a SUDEP knowledge score >2) influenced whether respondents discussed SUDEP with nearly all of the their patients (≥90% of the time). We also examined whether the same factors and the additional factor of discussing SUDEP with most or nearly all of their patients (≥50% of the time) influenced whether the respondents identified the most common response to SUDEP discussion as a “negative” one. Responses classified as “negative” included choices such as anxiety, depression, anger, distress, fear, and similar responses. Independent factors which demonstrated a $p < 0.2$ of an odds ratio different from 1 were included in the multivariate model for further analysis. Additional analyses were performed using the chi-squared test for comparison of categorical data.

The study was approved by the institutional review board at the NYU School of Medicine.

3. Results

Of the 17,558 invitations sent, we received 1645 responses (9.3% response rate). Response rates among Canadian neurologists (8.6%) did not differ from the overall group. Of those, 425 opted out of the survey typically because they did not meet one of the inclusion criteria (active clinical practice or still in residency or fellowship training). There were 1200 completed surveys. Because we do not know the number of incorrect e-mail addresses and additional characteristics of all names on our mailing list, the true proportion of eligible subjects responding to the survey is unknown. The characteristics of the respondents are listed in Table 1. The majority (76.4%) identified themselves as adult neurologists, and 33.8% of the respondents had additional training in epilepsy or clinical neurophysiology. Most respondents (92.8%) were US-based neurologists, and 37.1% were in academic practice. A majority (54.1%) were in practice for 10 or more years, and 95.8% saw at least one patient with epilepsy annually; 43.0% saw >100 patients with epilepsy per year. Most respondents (82.8%) had incomplete knowledge of published SUDEP risk factors defined as SUDEP knowledge scores of ≤2.