

33 Providing Information about SUDEP

The Benefits and Challenges

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INTRODUCTION

Sudden unexpected death in people with epilepsy was recognized in the early twentieth century (Spratling 1904), along with identified risks and recommendations for its prevention; however, by the 1960s a myth had gained hold in the medical literature that epilepsy was not fatal. "Patients with epilepsy had moved from asylums into the community and there was much less opportunity for observation. Risks from epilepsy were minimized then denied; that seizures could not be fatal became 'common knowledge' despite evidence to the contrary" (Nashef 1995). The development of effective pharmacotherapies gave added reassurance to the community that the condition would be controlled. However, deaths continued to occur and it became clear that there was no medical explanation. In the United Kingdom, bereaved families, shocked by the lack of scientific knowledge, began working to provide mutual support and to seek answers. A charity was formed in 1995 (originally known as Epilepsy Bereaved and now known as SUDEP Action), and the phenomenon defined as sudden unexpected death in epilepsy (SUDEP) rapidly became a specific focus for research and action in clinical practice.

There is much about SUDEP that remains unknown, and this complicates the assessment of risk and communication of advice to patients. The goal of SUDEP information provision is to assist informed patient decision making and to reduce risk, while avoiding any negative impact on quality of life. However, opinions differ on how best to facilitate such outcomes. This chapter explores that challenge and considers options for the timing and method of providing information to patients and families.

SUDDEN UNEXPECTED DEATH IN EPILEPSY: OUT OF THE SHADOWS

The nature of the scientific uncertainty on SUDEP has changed. Although there is no known cause, research has made significant advances in the understanding of risk factors associated with SUDEP. As a result, SUDEP awareness campaigns have multiplied with international networking as the key to effective use of experience and resources.

The first ever international workshop on SUDEP in 1996 identified the need for comprehensive research (Nashef et al. 1997) and led to a government-funded nationwide audit of epilepsy-related deaths in the United Kingdom. This National Sentinel Clinical Audit of Epilepsy-Related Death (Hanna et al. 2002) found significant problems of access to specialist epilepsy services, reviews, and problems with medication use, concluding that there was significant potential for reduction of death if these risk factors were addressed. The audit motivated policy makers in the United Kingdom to develop government action plans on epilepsy (Department of Health 2003; Welsh Assembly Government 2009) and to develop national clinical guidelines that included SUDEP.

When SUDEP occurs in families who are unaware of the phenomenon, the distress to the bereaved and the wider community is multiplied. A qualitative study found that the bereaved reported positively when the medical team offered condolences, support, and information and negatively when contact was not offered, delayed, or was defensive in nature (Kennelly and Riesel 2002; Bellon et al. 2014). Families were grateful for the offer regardless of whether it was taken up. As a result of this evidence, epilepsy guidelines in the United Kingdom recommend that health professionals contact families to offer their condolences, invite them to discuss

the death, and offer referral to bereavement counseling and SUDEP support groups (Kennelly and Riesel 2002; Stokes et al. 2004; Cook 2005).

Community-based SUDEP support services are gradually appearing internationally. The first service was developed in the United Kingdom by the charity Epilepsy Bereaved (www.sudep.org) in 1995, which is known today as SUDEP Action. The service is not dissimilar to the sudden infant death syndrome (SIDS) program (Woodward et al. 1985) and offers bespoke support to the bereaved through a dedicated helpline; collaborative meetings of families, researchers, and clinicians; a biennial memorial service; and information, education, and research services.

In Australia, epilepsy agencies support and link SUDEP families, organize biennial memorial services, and develop resource materials. A SUDEP-related research fund has also been established (www.epilepsyaustralia.net/).

In 2005, Epilepsy Australia and SUDEP Action initiated an international global conversation on SUDEP (www.sudep-globalconversation.com) and disseminated an all-encompassing book filled with a combination of scientific research and family stories, which has been used by epilepsy agencies worldwide on SUDEP campaigns (Chapman et al. 2005). In 2010, SUDEP Aware from Canada (Jeffs and Elizabeth 2014) joined the collaboration for a second edition titled *SUDEP—Continuing the Global Conversation* (Chapman et al. 2011). SUDEP Aware provides support for bereaved families and funding for research (www.sudepaware.org/). SUDEP Aware's *Making Sense of SUDEP* information brochures are widely used by epilepsy agencies across North America and Canada.

In 2013, SUDEP Action launched the first online registration of epilepsy deaths (www.epilepsydeathsregister.org), and it is currently working with partner organizations to support register development in other countries using this web platform. Cases can be registered by families and friends, health professionals, or coroners/medical examiners to provide a better understanding of why individuals die and how the deaths are investigated. It will also raise public awareness of SUDEP as well as strengthen the campaign for more effective clinical and coronial epilepsy services.

In France, families have been involved in the French Epilepsy Mortality Surveillance Network, a network of neurologists who systematically report deaths of people with epilepsy and case details, following family consent, to a central registry. This is in parallel with the Mortality in Epilepsy Monitoring Unit Study (MORTÉMUS), which collects detailed information about deaths in hospital video-EEG monitoring units across France and worldwide.

Following the first national workshop on SUDEP in the Netherlands during 2013, epilepsy agencies, including bereaved families, are collaborating on a series of initiatives aimed at developing SUDEP awareness and research.

In the United States, bereaved families have supported the development of a SUDEP-targeted research program through CURE (www.cureepilepsy.org/home.asp) and participated in the 2008 SUDEP symposium sponsored by the National

Institute of Health and the National Institute of Neurological Disorders and Stroke (NINDS). Today, in North America federal agencies are providing significant funding opportunities to gain a better understanding of risk factors, mechanisms, and management of individuals who die from SUDEP. Initiatives include the development of a Sudden Death in the Young Registry funded by the National Heart, Lung, and Blood Institute; NINDS; and Center for Disease Control (CDC) (<http://www.cdc.gov/sids/CaseRegistry.htm>). Recent efforts by organizations such as the Epilepsy Foundation's SUDEP Institute, events such as the Partners Against Mortality in Epilepsy educational conferences (www.aesnet.org/pame/epilepsy_currents), and data collection through the North American SUDEP Registry (www.sudep-registry.org) have resulted in increased knowledge and awareness of SUDEP.

RISK COMMUNICATION: CURRENT PRACTICE

A number of risk factors for SUDEP have been put forward, including suboptimal seizure control, young adult, early onset of epilepsy, absence of treatment or nonadherence to antiepileptic drugs (AEDs), polytherapy, sudden and frequent changes to AEDs, the prone position, being in the bedroom, sleeping, being alone, and being male (Monte et al. 2007; Tomson et al. 2008; Hughes 2009; Shankar et al. 2013). The effect of nocturnal supervision was examined in only one case-control study but was found protective (Langan et al. 2005). Overall, a high frequency of seizures (particularly generalized tonic-clonic seizures) is the strongest risk factor for SUDEP (Hesdorffer et al. 2011). A recent study of deaths in a 9-year study population in Cornwall found that the majority did not have an epilepsy specialist review in the year before death, mirroring the earlier national audit that a significant number of deaths may be in people not engaged with epilepsy services (Shankar et al. 2014). The UK National Sentinel audit identified some 45 individuals who were never referred to secondary care or who died waiting for referral (Hanna et al. 2002).

Despite some contradictory voices, the body of medical opinion has been building that some SUDEP deaths are potentially avoidable (Hanna et al. 2002; Opeskin and Berkovic 2003; So 2006; Hittiris et al. 2007; Monte et al. 2007; Faught et al. 2008; Tomson et al. 2008; Hughes 2009; Surges et al. 2009; So et al. 2009). A report of the American Epilepsy Society and the Epilepsy Foundation Joint Task Force on SUDEP supports the view that certain risk factors associated with SUDEP may be modifiable, namely, uncontrolled seizures, subtherapeutic drug levels, and number of AEDs used. The report supported optimization of seizure control as a treatment goal. It also noted the need to raise awareness of SUDEP in both medical and lay communities (So et al. 2009).

It has been suggested that the precautionary approach of the SIDS risk reduction campaign is a suitable model for SUDEP (So et al. 2009) (Table 33.1). SIDS is a rare event with no one cause identified, yet during the past two decades many

TABLE 33.1
Risk Factors and Preventative Strategies

Risk Factors	Strategies	Comment
Young age, early onset of epilepsy and IQ	Nonmodifiable	Individual risk assessment information provision
Seizure activity (especially GTCS)	Optimal diagnosis, treatment, and review	
Long duration of epilepsy	Consider surgery	
Polytherapy	Medication review	Patients should use the smallest number of AEDs to control seizures
Subtherapeutic levels AED	Medication review and assess adherence	
Nocturnal seizures	Nocturnal supervision but no recommendation about seizure detectors	Limited evidence on efficacy of seizure detectors

Source: So, EL et al., *Epilepsia*, 50(4), 917–22, 2009.

Note: GTCS, generalized tonic-clonic seizure; IQ refers to Learning Disabilities.

countries around the world have launched successful campaigns informing the public about the risk factors for SIDS (Moon et al. 2007; Ryvlin et al. 2013). A precautionary approach for SUDEP would support prevention strategies based on optimization of seizure control, effective AED treatment, supervision in appropriate cases, and information to patients and caregivers. This precautionary approach is supported by a reasonable body of policy makers (Department of Health 2003; Stokes et al. 2004; Welsh Assembly Government 2009) and expert opinion in North America (Institute of Medicine 2012).

For more than 10 years, clinical guidelines in Scotland and those produced by the National Institute of Health and Clinical Excellence (NICE) for England and Wales have recommended discussion of SUDEP as part of general epilepsy information (Scottish Intercollegiate Guidelines Network 2003; Stokes et al. 2004). The NICE guideline sets out information to show why preventing seizures is important. It is based on the view that SUDEP risk can be minimized by optimizing seizure control and by families being aware of the potential consequences of nocturnal seizures. It also recommends tailored information on the individual's risk of SUDEP as part of any counseling checklist for people with epilepsy, and their families and/or caregivers. This advice is also given by the American Epilepsy Society, which recommends disclosure but, in keeping with the Scottish and English guidance, does not give specific advice about how and when SUDEP should be discussed and by whom (Vegni et al. 2011; Mendonça et al. 2011).

The first survey of clinical practice related to risk communication and SUDEP asked neurologists in the United Kingdom if they told patients about SUDEP (Morton et al. 2006). The neurologists who replied reported a wide variation in practice. Whereas 30.3% of respondents reported that they discussed SUDEP with the majority or all of their patients, 68.7% discussed the issue with very few or none of their patients. Interestingly, clinicians with a special interest in epilepsy were significantly more likely to discuss SUDEP. A more recent hospital audit in Scotland of documented SUDEP discussion found that routine discussion was noted in only 4% of cases (Waddell et al. 2013).

A survey of Italian epileptologists reported similar findings. The survey found that physicians reported a wide variation of practice, with 28% discussing SUDEP with all or the majority of their patients and 62% discussing SUDEP with only a few (Vegni et al. 2011). This lack of consensus has been explored in the United States using a focus group study of neurologists, epileptologists, and advanced practice nurses (Miller et al. 2014). The findings mirrored the U.K. experience. Disclosure was supported by all professional groups, driven by practical and moral accountability to ensure that patients and families had accurate information on which to form their decisions about epilepsy. However, there was also a moral concern not to cause fear in patients by giving this information and all groups expressed a wish for standardized information delivery.

Specialist nurses have been shown to be more likely than clinicians to discuss SUDEP. A survey in the United Kingdom found that 56% of specialist nurses discussed SUDEP with the majority of their patients. Most combined SUDEP with discussion of general and specific risks of epilepsy (Lewis et al. 2008). No specialist nurse supported nondisclosure.

Research with parents in Wales found that 52% were aware of SUDEP. Only 16% of patients had been informed by a healthcare professional, with 70% reporting that they found the information via alternative sources. It concluded that parents of children with epilepsy should receive tailored information about SUDEP as this could help minimize risk factors and help reassure the parents. It also concluded that providing parents with tailored information could prevent the use of less reliable information sources (Jones and Naude 2013).

ETHICAL AND LEGAL ARGUMENTS FOR DISCLOSURE

The main arguments for disclosure are that it supports patient autonomy, informs choices on adherence and self-management, reduces fear by managing natural anxieties, and avoids the harm of false assurance. Research repeatedly tells us that people with epilepsy feel they receive insufficient

information about their condition (Coultridge et al. 2001). A qualitative study found that patients felt a pressing need for information, including "... more concrete answers on SUDEP" to discuss treatment options and medication in detail (Prinjha et al. 2005). This suggests that clinicians need to check regularly with patients how much information they would like about their condition. The perspective of caregivers is also a consideration. Research shows that patient decision making involves an extended social context (Fraenkel and McGraw 2007); however, in any event caregivers may need accurate information in their own right. Families affected by SUDEP described a variety of roles associated with care such as keeping records of seizures and responding to seizures. Many considered that they needed more information on SUDEP as it pertains to these roles (Kennelly and Riesel 2002). One study found that parents expected this (Gayatri et al. 2010). Research in Canada that included parents of children with epilepsy and families who had lost children found that all participants wanted information about SUDEP and generally wanted this early at the time of diagnosis. Knowing the actual risk of SUDEP was relieving especially to the parents who overestimated the risk (Ramachandran Nair et al. 2013).

The ethical principle of patient autonomy involves the patient's right to know about their own medical condition and prognosis. The American Epilepsy Society and the Epilepsy Foundation Joint Task Force on SUDEP considered discussion of SUDEP consistent with the ethical principles of patient autonomy and consistent with the need to accept that some persons with epilepsy have increased risks of morbidity and death (So et al. 2009). Examples of professional guidance based on the autonomy principle fully support the patient's right to information about his or her condition. The amount of information given on a condition should depend on the patient's wishes and need for information. This should be determined by discussion with a patient and not be based on assumptions of what patients require (General Medical Council 2008). Provision of information is also important to the relationship of trust between doctor and patient. Where information on risks is withheld and risks later materialize, it is natural that those affected by the consequences will seek to understand why information was not shared. They may experience the harm of false assurance. There is some limited evidence on the negative impact of false assurance in the context of medical screening, including public confidence and legal action (Petticrew et al. 2001); however, the impact of most seizures being presented as benign (apart from accidents and status epilepticus) has not been researched. Relatives bereaved through SUDEP frequently report, however, that their grief is exacerbated because epilepsy was presented in this way and it proved to be false (Kennelly and Riesel 2002). Further, the relationship of trust between the bereaved and the medical professional is more likely to be maintained where information withholding is patient centered and withstands scrutiny.

A common assumption is that discussing the risk of SUDEP will create anxieties. The only research on epilepsy

and death anxiety comprises studies looking at the death anxiety associated with epilepsy generally. A study of 373 epilepsy patients found that approximately two-thirds harbored fears of death from their next seizure (Mittan 1986). In a more recent cross-sectional study of 92 patients having epilepsy for a minimum of 5 years, 56.52% of patients had either moderate or high death anxiety (Otoom et al. 2007). Not surprisingly, death anxiety was likely to be higher in patients with generalized epilepsy. Otoom et al. (2007) emphasize the importance of counseling patients to reduce anxiety. Anecdotal reports from clinicians who regularly talk about SUDEP mention the potential for reduction of fear by putting fears into perspective. Indeed, there is some evidence to support this logic. A case-control study on the benefits of a weekend educational program (SEF, known then as the Sepulveda Epilepsy Education program but known more recently as Seizures and Epilepsy Education), which included discussion of risk of mortality, found subsequent decrease in anxiety (Helgeson et al. 1990). The experience of this program was that lack of discussion of mortality led to adverse suppression of natural anxiety. Fear was also associated with overprotection and overcontrol of the person with epilepsy (Mittan 2005).

SUDEP is now in the public domain, and use of the Internet to access health information is common (Fox and Duggan 2013). Not all websites provide clear and accurate information, and there is the potential for anxiety founded on inappropriate advice (Jones and Naude 2013; Tonsaker et al. 2014). In addition, patients who discover SUDEP themselves may be disappointed or angry that their doctor did not discuss it. Trust between patient and physician may be so damaged that any future attempts to put the issue in context may no longer be well received or followed. Personal discussion with their own doctor is the best way for people to hear about, and appraise, their own unique epilepsy-related risk. Frank discussion can build trust in the therapeutic relationship.

A legal duty to discuss a risk *prima facie* arises where there is a significant risk that would affect the judgment of the reasonable patient (*Bolitho v. City and Hackney Health Authority* 1998; *Pearce v. United Bristol Healthcare NHS Trust* 1999). Risks that are only remote possibilities can be regarded as material if the severity of the risk materializing is very serious. In the context of a SUDEP, it could be argued that there is a material risk to be addressed where discussion might influence behavior, improve compliance, or be relevant to issues of supervision or where optimum treatment is not in place (Beran et al. 2004). The courts in many countries will test the medical evidence offered by parties in litigation to reach their own conclusions on the magnitude and severity of risk, and the case by which the risk might be avoided (*Videto v. Kennedy* 1981; *Rogers v. Whitaker* 1992; *Bolitho v. City and Hackney Health Authority* 1998). The existence of guidelines, although not predictive of negligence, would be a relevant consideration in examining the rationality of withholding information. It has been argued that a claim would fail in a SUDEP case because of the need to prove causation between the actions of the physician and the death (Beran 2006). However, the more recent authority

of *Chester v. Afshar* (2005) has extended the law of causation to include the scenario in which risk is inherent in a condition and not caused by medical intervention. This would be a helpful legal authority in a SUDEP case. There are also two Scottish judicial determinations (Taylor 2002; Duff 2011) as well as recorded cases of out-of-court settlements and related judicial proceedings in the United States, all of which support the need for disclosure (Wannamaker 2011).

The Scottish determinations were the outcome of Fatal Accident Inquiries. This type of inquiry is held in Scotland by a judicial officer called a sheriff where a death concerns public interest. The purpose is to make recommendations to prevent future deaths. In the case of Collette Findlay, the deceased's mother had died from epilepsy in 1988. The deceased presented with seizures in 1991. The family was reassured that she suffered from benign focal seizures of childhood and she was discharged by the specialist to general practice on AEDs. Beginning in 1991, she had four to five seizures yearly; varying in frequency and severity, but there was no annual review or rereferral for specialist care. The court found a catalog of failures to look after the deceased in a proper manner. These included a failure on the part of specialists to alert the general practitioner as to what circumstances required rereferral; a failure on the part of the general practice to prescribe appropriate levels of medication; a failure to rereferral the deceased when the seizures did not stop after 2 years; a failure to rereferral the deceased when the intensity, form, and duration of her seizures altered as the deceased matured; and a failure by the medical team to discuss with the deceased's family the diagnosis, the attendant risks, and how these risks might be properly managed. The sheriff stated that given the association between seizures and SUDEP and the potential for control it was a "short step" to the view that if the deceased had been referred for review she might not have died. He determined that the family ought to have been informed that the deceased was

suffering from epilepsy, the risks of SUDEP explained, and a discussion held on how her condition might be managed. The most important recommendation was considered to be the need for a personal care plan. The sheriff suggested that all the key issues would have been addressed if a care plan, "... shared or otherwise" had been produced, and "... it might have saved her life" (Taylor 2002). The sheriff was clear that for the purposes of the public inquiry it was not necessary for there to be any scientific certainty. Any legal judgment is determined by balance of probabilities, and in this case the concern was whether preventative measures "might" have saved a life. The sheriff was clear that information on SUDEP could be relevant to how proactive a family might be in probing decisions about treatment as well as informing discussions on how risks might be reduced. The sheriff accepted that the question of informing about SUDEP must be left to the discretion of the medical profession to form a view. In particular, he accepted that there might be people of "an extreme disposition" where discussion might cause harm. Nevertheless, he said, "I do, however, accept that in the vast majority of cases there should be such a discussion" (Taylor 2002).

Nine years later, a second Fatal Accident Inquiry was convened to investigate the deaths of two young women with epilepsy (Erin Casey and Christina Ilia), taking evidence from 29 witnesses over 30 days (Duff 2011). It concluded that SUDEP is normally associated with a seizure; the risk of SUDEP occurring is reduced if the frequency of seizures is reduced; the frequency and incidence of seizures can be reduced by a number of factors, some related to lifestyle but the most important related to adherence to antiepileptic medication; and if a seizure occurs intervention by another person might prevent SUDEP from taking place (Table 33.2).

The inquiry into the death of Erin Casey focused on the relationship between communication of risk and medicines management. Erin had been considered at low risk of

TABLE 33.2
Recommendations from the Fatal Accident Inquiry Scotland 2011

Clinicians	<p>The vast majority of patients with epilepsy, or their parents or carers where appropriate, should be advised on SUDEP on first diagnosis or shortly thereafter.</p> <p>A decision may be made not to inform or to delay informing where there is an assessment that there is a risk of serious harm to a particular patient from the information or that the person lacks capacity. The decision should be recorded.</p> <p>Consideration should be given of support needs of patients on diagnosis and in managing medicines especially where the medication regime is complex. Consideration should be given to monitoring the uptake of repeat prescriptions.</p> <p>A letter should be sent to the patient and any referring general physician summarizing any consultation and treatment decisions.</p> <p>Written information should be reviewed to check if it is adequate.</p>
Health services	<p>Consider the adequacy of existing guidelines including clarifying the status of guidelines.</p> <p>Consider training needs to include communication with particular emphasis on supporting teenagers or young adults moving toward independence, engaging in risky activities, or beginning to take responsibility for them.</p>
Investigation of death	<p>Review practice in relation to the scene of death of a sudden unexpected death and the practice of, <i>ab initio</i>, describing it as a "crime scene."</p>

Source: Duff, AJM, Determination of Sheriff Alistair Duff, Sheriff of Tayside Central and Fife at Dundee. Inquiry held under the Fatal Accidents and Sudden Death Inquiry (Scotland) Act 1976 into the deaths of Erin Casey and Christina Ilia, at <http://www.scotcourts.gov.uk/opinions/2011/FA140.html>, 2011.