



Dr Kate Riney

SUDEP – seeing the full picture

Dr Damian Clark, Dr Kathryn Urankar and Dr Kate Riney

Introduction

Sudden unexpected death in epilepsy (SUDEP) is the main reason why people with epilepsy have an increased risk of death compared to the general population. Raising awareness of and working towards preventing SUDEP has been the common goal of doctors caring for people with epilepsy, and those who suffer from this condition. When tabling SUDEP with governments and health service leads, hard data on numbers of SUDEP deaths can be powerful information to help drive change towards reducing SUDEP. However, accurate determination of the number of SUDEP deaths is commonly hampered by a number of factors.

Background

Sudden death is a phenomenon that has been long recognized as occurring in people with epilepsy, with reference to this in medical literature from as early as the late nineteenth century. The term SUDEP was first coined to describe this phenomenon by Doctors Lina Nashef and Stephen Brown in 1996, at the first International Workshop on Epilepsy and Sudden Death in London,¹ with a definition for SUDEP proposed at that time: “the sudden, unexpected, witnessed or unwitnessed, nontraumatic and nondrowning death in an individual with epilepsy, with or without evidence for a seizure and excluding documented status epilepticus where postmortem examination does not reveal a cause for death”.

Despite SUDEP being clearly defined, low rates of use of the term SUDEP on post-mortem reports have been noted in a number of studies.²⁻⁴ In a national survey of coroners and pathologists in the United States, with over 500 respondents, 84% of US pathologists who responded acknowledged that they considered SUDEP a valid diagnosis if no cause of death were found at autopsy, but only 23% of these actually used the term SUDEP in more than

half of cases where criteria for SUDEP were met.² Whilst the definition for SUDEP provides clarity around the circumstances that must be met in order for SUDEP to be diagnosed, the area of ambiguity that often arises is around the concept of the absence of an identifiable cause for death being revealed at post-mortem examination. SUDEP is not always a phenomenon that is unexplained, but rather unexpected. When the postmortem examination shows evidence of a recent seizure (such as tongue biting) or when there is evidence of asphyxia or suffocation (usually arising in the context of a seizure due to failure to correct body position in response to difficulties in breathing during or after the seizure), the death may certainly be explained as likely due to the adverse consequences of a seizure. Leading SUDEP authors argue that, although findings of asphyxia or suffocation or seizure may be noted at post-mortem, these deaths should still be considered as SUDEP deaths as the definition of SUDEP encompasses unexpected death in a seizure (which may be related to cardiac or respiratory factors in the peri-seizure phase) and that it is unhelpful to separate out deaths with post-mortem findings of seizure or asphyxia or suffocation (not least because the absence of these findings at post-mortem does not exclude these as having happened and caused death).⁵ It is for this reason that the term SUDEP includes the term ‘unexpected’ rather than ‘unexplained’, as it is likely that the major cause of SUDEP may be explained as being the consequence(s) of a seizure (especially of an unwitnessed and therefore an unattended nocturnal seizure). Nonetheless, it is this ambiguity that may lead to under-use of the term SUDEP as a post-mortem diagnosis.

Other factors that impact on understanding the incidence of SUDEP include consistency of terminology in recording epilepsy related deaths on death certificates,⁴ subsequent

Authors affiliation:

Dr Damian Clark is a Neurology Fellow at the Royal Children’s Hospital in Melbourne.

Dr Kathryn Urankar is a Forensic Pathologist at The John Tonge Centre, Forensic & Scientific Services Queensland and Neuropathologist at Queensland Pathology, Royal Brisbane Hospital.

Dr Kate Riney is a Paediatric Neurologist and Epileptologist at the Mater Children’s Hospital in Brisbane, co-chairing the Queensland Paediatric Epilepsy Network.

For more information on the Queensland Paediatric Epilepsy Network (QPEN) and activities currently being undertaken to improve health outcomes for young people with epilepsy in Queensland, please contact QPEN Network Manager Terry Lack, PO Box 128, Royal Brisbane & Women’s Hospital Herston, QLD 4029.

coding of cause of death by national statistics offices, practice in reporting of deaths to the coroner, actions of the coroner in deciding further examinations, completeness of post-mortem investigation, access to these sources of information retrospectively by those researching SUDEP and the availability of prospective data capture systems. In addition, variations in the laws of different countries may impact on the approach to investigation of sudden deaths in people with epilepsy. For example, in Queensland, mandatory reporting of sudden death per se is not required under the Coroners Act, 2003. However, in England and Wales, there is a particular emphasis in law on the coroner's legal duty to hold an inquest in the event of death being sudden and of unknown cause (United Kingdom Coroners Act, 1988 section 8(1)).

Review of SUDEP deaths in young people in Queensland

As part of the ongoing work of the Queensland Paediatric Epilepsy Network (QPEN) to improve health care outcomes for young people with epilepsy in Queensland, a retrospective review of epilepsy related deaths in young people (<18 years) was undertaken over a 5 year period to help understand the incidence and risk factors for SUDEP in the younger Queensland population. In Queensland, the Queensland Child Death Register (QCDR) was established under the Commission for Children and Young People and Child Guardian Act, 2000. The QCDR allows for the centralized collection and collating of mortality data from both coronial and non-coronial deaths of young people under the age of 18 years registered in Queensland. Through this process, data was captured on all deaths in young people under the age of 18 years in Queensland where epilepsy or seizure or a similar term was listed on the death certificate and/or post-mortem report.

In the 5-year period of this review, a search of the QCDR data identified over sixty young people with epilepsy who died, with reference to epilepsy or a similar term (e.g. seizures, convulsions etc) on the death certificate (two-thirds of deaths) or having the death referred to the coroner (one third of deaths). There was considerable inconsistency in the terminology used to record deaths from epilepsy on death certificates. Terms

used included 'epilepsy', 'seizure', 'convulsion' and a range of other terms that might refer to the seizure, the epilepsy type, the epilepsy syndrome or the underlying aetiology for the epilepsy. Whether epilepsy (or related term) was considered as the leading cause of death on the death certificate was also noted to be variable. Epilepsy (or related term) was listed as a causal factor in the death in 58% cases, and as a contributing factor to the death in 42% of cases. In a number of cases, epilepsy (or similar term) was listed several times on the death certificate e.g. 1(a) convulsion 1(b) epilepsy 1(c) epilepsy syndrome. Whereas in other cases 'epilepsy' was simply listed at 1(a) with no other entry made. In some instances, every condition the person had was listed randomly on the death certificate whether it had any relationship to the death or not. The epilepsy was determined to be the leading cause of death in approximately half of all deaths in young people with epilepsy in Queensland in the five year period of this review.

Although around a third of all deaths in young people with epilepsy were sudden, the term SUDEP was only used as the final post-mortem diagnosis in around a third of sudden deaths that had post-mortem diagnosis available. In the majority of sudden deaths, the circumstances of death would have met the criteria for SUDEP or 'possible SUDEP'⁶ (where post-mortem investigations were not completed to allow criteria for SUDEP to be met, but all other factors were consistent). Instead of SUDEP, other diagnoses such as 'epilepsy', 'aspiration due to epilepsy' 'seizure' and 'respiratory failure due to epilepsy' were commonly found where SUDEP or 'possible SUDEP' would have been appropriate.

Conclusions

Data from this retrospective review indicates that we are a long way from being able to be clear that we can ascertain the number of SUDEP deaths currently occurring in Queensland (and by association in Australia), either from death certificate records or from post-mortem diagnosis records. In order to be able to identify SUDEP deaths, agreed standards for certification of deaths in people with epilepsy are essential as are standardized approaches

to reporting sudden deaths to coroners, standardized approaches to the post-mortem investigations of these cases and improved use of the term SUDEP on post-mortem reports when criteria for SUDEP are met. Central to this is collaboration amongst health professionals to agree such standards of care, education of the wider health community, and support for ongoing prospective data capture systems through funding resources. Accurate ascertainment of SUDEP deaths is essential to progress our understanding of SUDEP as without this it will always be more difficult to fully understand and study the risk factors that may predispose certain patients with epilepsy to SUDEP and ways in which SUDEP might be prevented.

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References

1. Nashef L. Sudden unexpected death in epilepsy: terminology and definitions. *Epilepsia* 1997; 38:S6–8.
2. Schraeder PL, Delin Kristen, McClelland RL, So EL. Coroner and medical examiner documentation of sudden unexplained deaths in epilepsy. *Epilepsy Res* 2006; 68:137–43.
3. Schraeder PL, Delin K, McClelland RL, So, EL. A nationwide survey of the extent of autopsy examinations in sudden unexplained death in epilepsy. *J Forens Med Pathol* 2009; 30(2):123–126.
4. Epilepsy – death in the shadows. National Sentinel Audit of Epilepsy Related Death Report 2002. Available from www.sudep.org.uk or via the following direct link: <http://www.archive2.official-documents.co.uk/document/reps/nscaerd/nscaerd.pdf>
5. Nashef L and Ryvlin P. Sudden Unexpected Death in Epilepsy (SUDEP): Update and Reflections. *Neurol Clin* 2009; 27:1063–1074.
6. Annegers JF, Coan SP. SUDEP: overview of definitions and review of incidence data. *Seizure* 1999; 8(6):347–52.