Seizure 30 (2015) 113–119

Contents lists available at ScienceDirect

Seizure

journal homepage: www.elsevier.com/locate/yseiz

Deaths in children with epilepsies: A UK-wide study

Peter Sidebotham [a,*], Lindsey Hunter [b], Richard Appleton [c], Colin Dunkley [d]

[a] Warwick Medical School, University of Warwick, Coventry, UK
[b] Royal College of Paediatrics and Child Health, London, UK
[c] Alder Hey Children's Hospital, Liverpool, UK
[d] Kings Mills Hospital, Mansfield, UK

ARTICLE INFO

Article history:
Received 3 December 2014
Received in revised form 16 March 2015
Accepted 6 May 2015

Keywords:
Epilepsy
Mortality
Case review
National audit

ABSTRACT

Purpose: This UK-wide review of deaths in children with epilepsies was undertaken to ascertain any demographic, clinical, organisational, or management factors associated with the deaths, and to determine the extent to which any of these may have deviated from nationally agreed best practice.
Method: Paediatricians across the UK were asked to notify any deaths in children with epilepsies over a 10-month period. Hospital and community case notes were reviewed by pairs of case assessors using a structured assessment tool combining holistic and criterion-based approaches.
Results: Of 46 deaths notified, case notes were obtained on 33. The majority of children had associated developmental impairments. The majority (24), died of an associated co-morbidity rather than of epilepsy. Seven died of convulsive status epilepticus and seven as sudden unexpected deaths in epilepsy. Twenty four percent of deaths were judged to be preventable; potentially modifiable factors included fragmentation of care, support for families, and recognition of and response to acute illness in the child, including the appropriate management of prolonged seizures.
Conclusions: Although this audit has demonstrated significant improvements in quality of care when compared with the last national audit of epilepsy deaths in 2002, further improvement is still required.

1. Introduction

Epilepsies are some of the most common chronic neurological conditions of childhood, with an estimated prevalence of 4.3 per 1000 children [1,2]. This equates to an estimated 65,000 children and young people with active epilepsy living in the UK. An average 57 children and young people aged <18 years die each year in the UK with epilepsy recorded as the underlying cause of death, and a further 55 registered deaths cite epilepsy as an underlying condition on the death certificate (Ruth Gilbert, Pia Hardelid, personal communication). Epilepsy-related deaths may occur as a direct result of the seizure (including convulsive status epilepticus), as a consequence of treatment given for the epilepsy, as an accident (including drowning), as sudden unexpected death in epilepsy (SUDEP), or may be related to an associated underlying neurological problem or arise from an unrelated cause.

Some epilepsy-related deaths are likely to be unavoidable, but others may relate to deficiencies in the care provided to children and their families. A previous national audit in 2002 concluded that 59% of epilepsy-related child deaths were potentially avoidable [3]. This was particularly related to limited access to specialist paediatric neurology expertise and the use of potentially inappropriate anti-epileptic medication. Since the publication of this National Sentinel Audit, a number of initiatives have focussed on the improvement of care of people of all ages with epilepsies [4], including the publication of national guidelines for the management of epilepsies [5–7]. A recent review of services for children with epilepsies [4] identified a number of improvements in overall care of these children, findings echoed in the 2012 national 'Epilepsy 12' audit [8]. However, there has been no more recent national review of epilepsy deaths, and recent high-profile cases suggest that poor quality of care may still be contributing to the deaths of some children with epilepsies [9].

As part of a national review of the quality of healthcare for children and young people with epilepsies, we sought to evaluate the case records of all reported deaths in children and young people with epilepsies across the UK between June 2012 and March 2013 [10]. The aim of the review was to ascertain any demographic, clinical, organisational or management factors associated with these deaths, and to determine the extent to which any of these deaths may have been associated with divergence from nationally agreed best practice. It was anticipated

* Corresponding author. Tel.: +44 24 7657 4878.
E-mail address: p.sidebotham@warwick.ac.uk (P. Sidebotham).

http://dx.doi.org/10.1016/j.seizure.2015.05.003
1059-1311/

that any identified deficiencies in care, as well as examples of good practice, could be used to promote good quality care and to make recommendations for the provision of services to children and young people with epilepsies.

2. Methods

The review used a mixed-methods approach, incorporating questionnaire-based demographic and clinical data, and a case notes review combining an explicit criterion-based assessment of clinical care with a more in-depth holistic review [11]. The review included any child or young person aged from 1 year and up to their eighteenth birthday with a prior diagnosis of epilepsy who died of any cause (Box 1). The time period for the review was 1 June 2012 to 31 March 2013. The time period for notification was limited by the requirements of the commissioning bodies.

Currently in the UK there is no national registry of epilepsy, although a voluntary epilepsy deaths register was established in 2013 by SUDEP Action (www.sudep.org/article/epilepsy-deaths-register). However, national guidance specifies that 'the diagnosis of epilepsy in children and young people should be established by a specialist paediatrician with training and expertise in epilepsy' [5]. While there may be small numbers of older young people managed solely by general practitioners or adult neurology services, in practice nearly all, and certainly all complex cases are managed by paediatricians. All consultant paediatricians across the UK were e-mailed on a monthly basis and asked to notify the team of any cases they had seen in the previous month. For each reported case, the consultant was asked to complete a secure web-based questionnaire providing demographic and clinical details. In order to maximise case ascertainment the study was advertised widely so others could notify cases, including intensive care units and child death overview panels, and a data-sharing agreement was set up with PICANet, a national paediatric intensive care audit network.

Box 1. Inclusion criteria and definitions.

Inclusion criteria
a. A child or young person with epilepsy who has died, of any cause
OR
b. A child or young person that has received intensive care or high dependency care following a prolonged seizure
AND
c. The child or young person was aged between their first and 18th birthdays at the time of the incident
AND
d. Prior to the incident the child or young person had a diagnosis of epilepsy.

Definitions
A pragmatic definition for epilepsy was used in this review: 'two or more epileptic seizures more than 24 h apart that are not acute symptomatic seizures or febrile seizures'. For the purposes of this review, any child or young person was included for whom the reporting clinician considered there to have been a previous diagnosis of epilepsy based on that definition.
Prolonged seizures were defined as 'any tonic–clonic seizure lasting longer than 5 min, or serial, repeated seizures continuing over a period of more than 30 min'.
Intensive care was defined as any child or young person requiring admittance to an intensive care unit, or receiving an equivalent level of care.
High dependency care was defined as requiring 'on-going close intervention or monitoring because of neurological or cardio-respiratory compromise'. This will include any child or young person receiving care requiring a nurse to patient ratio of 0.5:1 (1:1 in cubicle), and any child or young person requiring at least hourly neurological or cardio-respiratory observations.

For each notified case, the case notes were requested from the child's first seizure through to the death, incorporating records from primary, secondary and tertiary care settings. Each set of case notes was reviewed by a pair of case assessors comprised of a paediatric nurse and a paediatrician who were actively involved in the care of children with epilepsies, and had undergone training in epilepsy care. None of the case assessors had been involved with the care of the children prior to, or following their death. The case assessors were trained in case notes review methodology and were supported by the research team, with regular briefing meetings and the opportunity to discuss any queries that arose from their review. The case assessment tool was structured around six phases of care (initial diagnosis and management; ongoing management; pre-hospital care; emergency department care; intensive or high dependency care; and care of the child and family around and following the death). The tool included a criterion-based assessment using recognised clinical standards [5,7,12,13], and a structured implicit review in which the case assessors were asked to rate the quality of care in each phase and to comment on any learning points and identified avoidable or remediable factors; they were also tasked with the identification of elements of good clinical practice and care. Details of the case assessment tools and their development are provided in the full CHR-UK report and in Appendix 1 [10]. Case assessors were required to classify the cause of death, according to a structured proforma (Appendix 2) and each case was reviewed by the lead researcher (PS) to confirm the classification of the death.

Ethical advice on the review was sought from the National Research Ethics Service. As a national service evaluation, the review was granted National Information Governance Board 251 approval to collect patient identifiable data without consent, along with equivalent approvals from the Scotland Caldicott Guardian and the Northern Ireland Privacy Advisory Committee. All patient-identifiable data were removed from the database prior to analysis by the research team.

3. Results

A total of 46 deaths in children with epilepsies were notified to the study. Hospital and community case notes were obtained for detailed review on 33 children. In spite of repeated attempts, case notes for the remaining 13 children were not provided by the hospital or community health providers. Details of the 46 children are shown in Table 1. The majority (94%) were known to have associated co-morbid conditions, including developmental impairments, most of which were severe and required multi-disciplinary care. Thirty-one children (67%) had a recognised cause for their epilepsy, of which the most common were identified genetic disorders [10], hypoxic-ischaemic neonatal brain injury [7], and cerebral malformations [6]. Eight children (17%) had an identified epilepsy syndrome, including West, Dravet, a progressive myoclonic epilepsy, Lennox Gastaut, epilepsy with generalised tonic–clonic seizures on awakening, and epilepsy with myoclonic absences.

The majority of children (29/46, 63%) were at home when they died or at the start of the incident that led to their death; 23 of these were transferred to hospital, including seven who received intensive or high dependency care for a prolonged seizure prior to their death. Five children were already in a hospice at the time of their death and a further four were transferred to a hospice for end of life care. Sixteen children (35%) were experiencing at least weekly seizures prior to their death, although these rarely necessitated a hospital attendance. Seven children (15%) had been admitted to hospital with a prolonged seizure (usually tonic–clonic) in the previous 12 months.