

# Child Health Reviews - UK

Clinical Outcome Review Programme

**Coordinating Epilepsy Care: a UK-wide review of healthcare in cases of mortality and prolonged seizures in children and young people with epilepsies**

**Report  
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Royal College of Paediatrics and Child Health



## Report prepared by

Dr Peter Sidebotham, Associate Professor of Child Health, University of Warwick

Lindsey Hunter, Researcher, Royal College of Paediatrics and Child Health

Dr Alison Elderfield (PhD), Programme Manager, Royal College of Paediatrics and Child Health

Dr Colin Dunkley, Epilepsy12 Project Lead and Consultant Paediatrician

Dr Jyotsna Vohra (PhD), Science and Research Coordinator, Royal College of Paediatrics and Child Health

Joanne Morgan, Data Collection Assistant, Royal College of Paediatrics and Child Health

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## Programme Board membership

Professor Neena Modi (Chair), Royal College of Paediatrics and Child Health Vice-President, Science and Research

Dr Alison Elderfield, Programme Manager, Royal College of Paediatrics and Child Health

Laura Gibbon, Former Programme Manager, Royal College of Paediatrics and Child Health

Professor Ruth Gilbert, Professor of Clinical Epidemiology, Centre for Paediatric Epidemiology and Biostatistics, Institute of Child Health, University College London

Professor Jenny Kurinczuk, Professor of Perinatal Epidemiology, National Perinatal Epidemiology Unit

Alan McMahon, Head of Information Systems, Royal College of Paediatrics and Child Health

Jenny Mooney, Clinical Outcome Review Programme Business Manager, Healthcare Quality Improvement Partnership

Dr Angela Moore, Consultant Paediatrician, The Royal Wolverhampton NHS Trust

Dr Peter Sidebotham, Associate Professor of Child Health, University of Warwick (Review Lead)

John Thain, Senior Lecturer in Children's Nursing, University of Wolverhampton

Dr Jyotsna Vohra, Science and Research Coordinator, Royal College of Paediatrics and Child Health

## Topic Expert Group

Dr Richard Appleton, Consultant Paediatric Neurologist, Honorary Senior Clinical Lecturer in Paediatrics and Child Health, Alder Hey Children's NHS Foundation Trust

Lesley Alexander, Community Epilepsy Nurse Specialist, The Northern Health and Social Care Trust, Northern Ireland

Dr Jonathan Cardwell, Consultant Community Paediatrician, Northumberland NHS Care Trust

Donna Cox, Lay Member

Dr Colin Dunkley, Epilepsy12 Clinical Lead, Consultant Paediatrician, Sherwood Forest Hospitals NHS Foundation Trust

Emma Carter, Clinical Nurse Specialist, Great Ormond Street Hospital for Children NHS Foundation Trust

Dr Alex Hamilton, Consultant Paediatrician, Huddersfield NHS Trust

Dr Ailsa McLellan, Consultant Paediatric Neurologist, NHS Lothian

Zoe Picton-Howell, Lay Member

Dr Alison Shefler, Consultant Paediatric Intensivist, Oxford University Hospitals NHS Trust

Dr Henry Smithson, NICE fellow and Academic GP, University of Sheffield

Dr Johann te Water Naude, Consultant Paediatric Neurologist, Cardiff and Vale University Health Board

Dr Michael Yoong, Specialist Trainee, General Paediatrics

## Case assessors

Dr Rachel Atkinson, Community Paediatric Registrar (ST7), Guy's and St Thomas' NHS Foundation Trust

Dr Janice Bothwell, Consultant Community Paediatrician, Belfast Health and Social Care Trust

Alexandra Cook, Paediatric Epilepsy Nurse Specialist, Western Sussex Hospitals NHS Foundation Trust

Lucinda Dawson, Paediatric Nurse (bank), Great Ormond Street Hospital for Children NHS Foundation Trust

Dr Wendy Forbes, Paediatric Registrar (ST7), Yeovil District Hospital NHS Foundation Trust

Alison Gormley, Complex Case Manager for Children, NHS Oldham Clinical Commissioning Group

Dr Sarah Gough, Specialist Registrar Community Paediatrics, Nottingham University Hospitals NHS Trust

Dr Stephanie Govendon, Specialty Trainee Registrar (ST6), Mid Yorkshire Hospitals NHS Trust

Dr Carl Harvey, Consultant Paediatrician (Paediatric Neurodisability), City Hospitals Sunderland NHS Foundation Trust

David Hyde, Specialist Paediatric Epilepsy Nurse, South Tees Hospitals NHS Foundation Trust

Eleanor Jeans, Paediatric Nurse Advisor, Independent Sector

Dr Helen Lewis, Consultant Community Paediatrician, Pennine Care NHS Foundation Trust

Mary McDonald, Early Education & Childcare Lecturer, City of Glasgow College

Dr Maria Moran, Paediatric Registrar, Nottingham University Hospitals NHS Trust

Dr Stephen Nirmal, Consultant Paediatrician with a special interest in Epilepsy, James Paget University Hospitals NHS Foundation Trust

Dr Anas Olabi, Consultant Paediatrician (Neurodisability), University Hospitals of Morecambe Bay NHS Foundation Trust

Dr Chinwe Osuchukwu, Consultant Community Paediatrician, Central and North West London NHS Foundation Trust

Dr Chaniyil Ramesh, Consultant Paediatrician with Special Interest in Paediatric Epilepsy and Neurology, West Hertfordshire NHS Trust

Nicola Rooke, Senior Staff Nurse, Cardiff and Vale University Health Board

Dr David Scott, Consultant Paediatrician and Clinical Lead for Paediatrics, East Sussex Healthcare NHS Trust

Keita Thomas, Paediatric Epilepsy Nurse Specialist, Western Sussex Hospitals NHS Foundation Trust

Dr John Trounce, Consultant Paediatrician, Brighton and Sussex University Hospitals NHS Trust

Georgina Ward, Paediatric Epilepsy Specialist Nurse, East Sussex Healthcare NHS Trust

Margaret Wilson, Paediatric Epilepsy Nurse Consultant, NHS Greater Glasgow and Clyde

## Stakeholders

Association of Neurophysiological Scientists  
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Epilepsy Action  
Epilepsy Connections  
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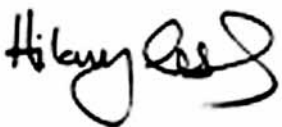
# Foreword

It is with great pleasure that I write the foreword for this report which presents a detailed understanding of children with epilepsies and their journey and experiences through the care pathway.

The United Kingdom has the worst levels of child mortality in Western Europe. Compared to Sweden, which is the best performer, it has been estimated that there are almost 2000 excess child deaths a year; that's an average of five a day.<sup>A</sup> Of course, not all these excess deaths are from healthcare amenable conditions, so it is essential that we gain a more detailed understanding of the heterogeneous factors underlying our relatively poor performance. International comparators give impetus, not answers, as evidenced by the fact that this worrying statistic is beginning to gain traction with Ministers and health officials.<sup>B</sup> To make sense of the data we need much more detailed information which allows us to really focus on the improvements that could be within our grasp, and to formulate recommendations for changes to services and training.

This report, from the Child Health Reviews - UK (CHR-UK) team, drills down into the detail of the epilepsies and highlights where services are doing well and where improvements are required. Because children with epilepsies can test every part of the healthcare system from diagnosis right through to end of life care, this report has implications for all professionals involved in the care of children, whether in hospitals, schools or other community settings. The ability to provide safe, equitable, integrated care for all children with epilepsies, in particular those with difficult to control epilepsies and multiple co-morbidities, is a true measure of the competence of our healthcare provision. This report sets out the essential improvements needed to move us towards that goal. Most importantly, it clearly shows the need for a partnership of care between those providing health services, and the children and families they serve.

The Royal College of Paediatrics and Child Health is committed to gathering and updating the evidence surrounding UK child mortality. The recommendations from this report will be taken into account by the RCPCH Mortality Task and Finish Group and, together with the other evidence gathered, will form part of a series of wider recommendations to improve health outcomes for children in the UK with the aim of reducing the number of avoidable deaths.



Dr Hilary Cass  
President, Royal College of Paediatrics and Child Health

<sup>A</sup> Health services for children in western Europe  
The Lancet, Volume 381, Issue 9873, Pages 1224-1234  
Ingrid Wolfe, Matthew Thompson, Peter Gill, Giorgio Tamburlini, Mitch Blair, Ann van den Bruel, Jochen Ehrich, Massimo Pettoello-Mantovani, Staffan Janson, Marina Karanikolos

<sup>B</sup> [http://www.local.gov.uk/c/document\\_library/get\\_file?uuid=fb487980-4306-4bee-8601-8538a30a2df3&groupId=10171](http://www.local.gov.uk/c/document_library/get_file?uuid=fb487980-4306-4bee-8601-8538a30a2df3&groupId=10171)



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# Executive Summary

The Clinical Outcome Review Programme: Child Health Reviews - UK (CHR-UK) was a UK-wide programme of work aiming to inform clinical practice and improve the healthcare provided to children and young people in England, Scotland, Wales and Northern Ireland and the Channel Islands and the Isle of Man by systematically examining mortality and morbidity in children and young people from age one to 17 years inclusive. Reviews of this nature provide the opportunity for reflection on the specialist nature of the child healthcare system and the provisions that are made.

This report covers a themed case review of mortality and serious morbidity in children and young people with epilepsy at all stages of the care pathway, including primary and emergency care. Through this review CHR-UK, for the first time, included the concept of serious morbidity as well as mortality. The review aimed to evaluate the standards of care provided to the children concerned against existing evidence based guidelines.

This is the first national review of serious morbidity in children and young people with epilepsies, and the first review of epilepsy-related deaths since the 2002 National Sentinel Clinical Audit of Epilepsy-Related Death. Therefore, this review provides a valuable contribution to the evidence base in the area of epilepsy, with the report recommendations having the potential to improve clinical practice.

## Key Review Questions

1. What demographic, clinical and system-related (organisational and management) factors are associated with deaths, intensive care and high dependency care admissions in children and young people with epilepsy in the UK?
2. To what extent are these adverse clinical outcomes (deaths and prolonged seizures requiring intensive or high-dependency care) associated with divergence from established best practice in clinical care for children and young people with epilepsy in the UK?
3. To what extent do demographic, clinical and system-related factors differ between those children and young people with epilepsy who die and those children and young people with epilepsy receiving intensive and high dependency care as a result of prolonged seizures?
4. What can be learnt through different approaches to reviewing cases of death or serious morbidity in children and young people with epilepsy (including child death overview panel reviews and serious untoward incident reviews)?

## Methodology

- The review was based on children with epilepsies aged one to 17 years inclusive from across the UK who died (of any cause) or who received intensive or high dependency care for prolonged seizures.
- The Programme Board oversaw the project, providing expert guidance and governance. In addition, a Topic Expert Group (TEG) supported the CHR-UK team in developing the data collection tools and defining the standards that formed the basis of the review. The TEG included hospital and community based paediatricians, paediatric neurologists, paediatricians with expertise in epilepsy, specialist nurses, general practitioners, and lay representation including parents of children and young people with epilepsy to ensure their perspectives were included.
- Case notification data were requested from RCPCH consultant members across the UK over a 10 month period. For each reported case meeting the inclusion criteria, the reporting clinician was provided with access to the RCPCH secure electronic web portal (EWP) to complete a clinical questionnaire.
- Cases were purposively selected for a detailed review to ensure that both deaths and cases of serious morbidity were included. A case assessment tool structured around six phases of care based on national clinical standards and structured implicit review was developed. Case notes were obtained from relevant healthcare settings and assessors worked in pairs to complete assessments at the Royal College of Paediatrics and Child Health (RCPCH) located in London and in local hospitals.
- A mixed methods approach to analysis was used, combining both quantitative and qualitative data.

Sixty-one cases of child mortality and morbidity were reviewed by assessors in order to inform and improve clinical practice and healthcare provided to children and young people in the UK. Case assessments were undertaken on 33 children who died, along with 17 children receiving intensive care and 19 receiving high dependency care for prolonged seizures, of whom eight subsequently died. The findings from the case notes review have been set in the context of demographic and clinical information obtained from the clinical questionnaires submitted for 162 children.

## Key findings

- **Key finding 1:** In spite of the severity of these children's health needs, this review has emphasised that the care provided by parents and professionals working together provides the best possible quality of care.
- **Key finding 2:** The findings emphasise the importance of rigour in the diagnostic process, While taking into account the often evolving nature of the diagnosis in epilepsy. There is a particular need for careful consideration of epilepsy syndromes, and this is likely to be aided by the early involvement of a paediatric neurologist. The findings from this review support the recommendations set out by the NICE and SIGN guidelines that the diagnosis of epilepsy should be made by a paediatric neurologist or paediatrician with expertise in childhood epilepsy.
- **Key finding 3:** The review has highlighted the importance of taking into account all information around the diagnosis and classification of epilepsy before beginning anti-epileptic drug treatment. Although treatment should follow NICE or SIGN guidelines and the BNFC, there may be reasons and thought processes for diverging from these. This review has highlighted the importance of clear documentation of such decisions. Consistency in following guidelines and clarity around the reasons for any divergence could be improved by processes of peer review in clinical teams.
- **Key finding 4:** The complexity of the children's epilepsies and wider health needs, presented in this review, means that there are often multiple professionals working with the child and family. The review has highlighted the importance of clear communication between professionals, and the need for one professional to clearly take a lead in the overall coordination of care. Where an epilepsy nurse specialist is involved, this must be documented in the clinical notes, together with any decision making, communication with parents or changes to management made by them. Such information needs to be communicated to all members of the clinical team. This supports and reinforces the recommendations made by the NICE and SIGN guidelines that a named clinician should have overall responsibility for coordination of care.
- **Key finding 5:** Early, thorough and ongoing discussions with children and young people and their parents or carers are crucially important. This is clearly emphasised in both NICE and SIGN guidelines. However, although such discussions may be taking place, there was a paucity of documentation around these.
- **Key finding 6:** Many of the children in this review experienced repeated hospital admissions for prolonged seizures. This along with the multiple co-morbidities, a lack of forward planning and appropriate care plans being in place highlighted the potential danger of clinicians focusing on the management of individual acute episodes, and the failure of anyone to step back and consider the wider ongoing long term needs of the child. In such situations, it is vital to ensure each child receives regular coordinated reviews of their epilepsy management.
- **Key finding 7:** This review has highlighted the importance of clear and comprehensive care plans for parents, schools and others caring for children with epilepsies, and providing them with information on how to respond to prolonged seizures, including training in resuscitation and the use of rescue medication. This is important for all children with epilepsies, but particularly where the child is known



to have suffered or be at high risk of prolonged seizures. Such care plans could be included in an 'epilepsy passport' as highlighted in **Recommendation 3**. This finding supports the recommendations on emergency care plans as set out in the NICE and SIGN guidelines.

- **Key finding 8:** The different formulations of buccal midazolam currently in use give rise to potential medication errors because of different dilutions (5mg/ml or 10mg/ml). This can give rise to either under or over-dosing, particularly when children are changed from one formulation to another.
- **Key finding 9:** Evidence was found of good initial assessment and response by ambulance staff, but some concerns were highlighted around the timing and administration of appropriate rescue medication. The findings from this review support the recent updated guidelines from the Joint Royal Colleges Ambulance Liaison Committee (JRCALC). However, these guidelines could be further strengthened, with updates of all local guidelines, to ensure all ambulance crews are trained and equipped to be able to administer buccal midazolam to children experiencing prolonged seizures.
- **Key finding 10:** There was good evidence from this review that both the emergency department and high dependency or intensive care provided to children presenting with prolonged seizures was, on the whole, meeting high standards of care. Nevertheless, all emergency departments must ensure that their clinical staff are able to apply current NICE and APLS prolonged convulsion guidance, as well as ensuring availability and competency with buccal midazolam, IV lorazepam and IV phenytoin administration. Deviation from these standards may, in some cases, be appropriate but this information should be clearly documented, as stated in **Recommendation 1**.
- **Key finding 11:** Admission to intensive or high dependency care provides an opportunity for reviewing the child's overall care, and making appropriate adjustments to their management and follow up, as well as reflecting on the care provided and learning lessons locally.
- **Key finding 12:** Children with epilepsies who die do so from a variety of causes, with over half of the children in this review dying of causes other than their epilepsy. This can lead to multiple professionals being involved in the child's care, allowing it to become fragmented. This further highlights the need for a single clinician taking charge of the coordination of the child's care.
- **Key finding 13:** Many of the children's deaths were anticipated deaths in the context of known life-limiting illnesses. A careful review of all the clinical findings and the circumstances of death are important for accuracy in classifying and registering the death. This should include discussion with the coroner or procurator fiscal and a joint agency 'rapid response' when a child with epilepsy dies unexpectedly, including those cases that appear to meet criteria for SUDEP and those as a result of status epilepticus. This supports the approach being taken by English Child Death Overview Panels, and the all-Wales Child Death Review Programme to reviewing all children's deaths and of a rapid response for unexpected deaths.
- **Key finding 14:** For many of the children with known life-limiting conditions there was evidence in this review of good supportive and anticipatory planning for the children and their families, in keeping with guidelines from the Association for Children's Palliative Care/Together for Short Lives (ACT). This should be a standard adhered to for all such children.

- **Key finding 15:** Overall, in eight of the deaths reviewed the case assessors identified contributory factors which, by means of locally or nationally achievable interventions, could be modified to reduce the risk of future child deaths. This emphasised the value of a thorough approach to reviewing each child's death in order to identify lessons at a local and wider level as highlighted in **key finding 13** and **Recommendation 8**.
- **Key finding 16:** When a child with epilepsy or other neuro-developmental impairments dies, he or she does not cease to be a part of their family. It is important that clinicians recognise this and ensure that the family receive appropriate support, advice and information.
- **Key finding 17:** The review findings showed that there were potentially modifiable factors leading to children's deaths in relation to the communication with parents. This highlights the need for clear information and advice to parents and carers, in a manner they can understand, on the signs indicating when a child is unwell. Furthermore the clinician responsible for the care of the child should ensure there are clear and careful discussions around the risks of seizures and SUDEP, as set out in the recommendations in the NICE guidelines. This would help empower parents and carers to recognise and respond promptly in such situations.
- **Key finding 18:** This review identified some concerns with recognition of and response to status epilepticus in hospitals, as highlighted in **key finding 10**. Ensuring clear channels of communication between different hospital teams are established will help to ensure that acute episodes are managed effectively.

## Recommendations

### **Recommendation 1**

Clinicians looking after children and young people with epilepsies should follow NICE and SIGN guidelines for all aspects of care, and document the reasons for any deviations from these standard treatment guidelines.

### **Recommendation 2**

Clinical teams looking after children and young people with epilepsies should consider establishing a process of peer review as a means of monitoring and improving practice.

### **Recommendation 3**

Clinical teams looking after children and young people with epilepsies should consider introducing an 'epilepsy passport' for all children as a means of improving communication and clarity around ongoing management.

### **Recommendation 4**

Whenever a child is admitted to hospital with a prolonged seizure, the consultant responsible for the admission should notify the clinician in charge of the child's overall care. The clinician with overall responsibility should then review the child's epilepsy management in the light of that admission.

**Recommendation 5**

When prescribing buccal midazolam for rescue medication in prolonged seizures, prescribing clinicians must clearly state the formulation being used and the dose to be given in both mg and ml. The consultant in overall charge of the child's epilepsy care should ensure that the parents and all other carers have an up to date emergency treatment plan that clearly outlines the dose to be given and the circumstances in which to give the rescue medication.

**Recommendation 6**

Ambulance Trusts should consider updating their protocols for seizure management in children and young people to recommend the use of buccal midazolam as the first line treatment for prolonged seizures. This should be backed up with appropriate training of all ambulance crews in the use of buccal midazolam and provision of buccal midazolam to all ambulance crews.

**Recommendation 7**

Emergency departments should ensure that children and young people presenting with prolonged seizures are treated according to current NICE and APLS guidance through appropriate departmental guidelines, training of staff and audit.

**Recommendation 8**

Child Death Overview Panels in England and the all-Wales Child Death Review Programme should ensure that the case of each child with epilepsy who dies is subject to a child death review, including, where appropriate, a multi-agency rapid response to investigate the death and provide support to the family. NHS Scotland, HSC Northern Ireland, Public Health Jersey, Public Health Guernsey and Department of Health Isle of Man should consider how such reviews could be built into any plans for development of child death review in their devolved nations.

**Recommendation 9**

The consultant responsible for the care of any child with epilepsy who dies should ensure that all subsequent actions after death, including registration of the death, referrals to the coroner or procurator fiscal, and follow up of the family together with child death review are documented in the child's notes and shared with other members of the clinical team.

# 1. Introduction

## 1.1 Mortality and morbidity in UK children

Each year over 5,000 children and young people die in the UK. Of the 4,061 child death reviews completed in England in 2010/11, 800 (20%) had factors that, if modified, would help in preventing future deaths.<sup>1</sup> This mirrors findings from other studies suggesting that as many as a quarter of all child deaths in high income countries are potentially avoidable.<sup>2,4</sup> In spite of significant reductions in child mortality over the past decades, mortality rates for children and young people in the UK are still higher than many western European countries. It has been argued that the care provided in the UK is not to the standard of our European counterparts.<sup>5</sup> and that this is compounded by regional variation in care and socio-economic inequalities.<sup>6,8</sup> The majority of these deaths are from natural or medical causes including perinatal and congenital conditions, as well as a range of acquired natural causes in particular neurological, respiratory and cardiovascular disorders, infections and cancers.

A large proportion of childhood deaths occur in children and young people with known chronic diseases or life-limiting conditions. This is discussed in the retrospective epidemiological review of all-cause mortality in children conducted as a component of the CHR-UK programme; Module A: "*Overview of Child Deaths in Four UK Countries*". These children, young people and their families will require the input of a range of health and social services throughout their lives. This will include supportive care in the community, hospital care for acute illnesses (or complications of their conditions) and anticipatory planning with appropriate end of life care. For those who die in childhood there needs to be support for them, and additionally to their family both prior to and following their death. Many disabled children and those with chronic conditions require frequent hospital admissions, often in different hospitals, and those with more complex conditions frequently have care provided by a range of carers and professionals. This increases potential for fragmentation of care across different settings and between professionals within the same healthcare setting.

The past decade has seen changes in the provision of emergency care in the UK. A recent study of English Hospital Episode Statistics (HES) showed a 28% increase in emergency admission rates from 63 to 81 per 1,000 children, equating to over 700,000 emergency admissions each year from 1999-2010.<sup>9</sup> The greatest increase has been in short stay (<1 day) admissions for acute conditions. Additionally, although overall admission rates for chronic conditions, including asthma, diabetes and epilepsy, have fallen, admissions for less than one day have increased. This suggests that many of these children are admitted for short periods of treatment or observation following acute events. Gill *et al.* (2013) suggested that these trends may indicate, among other factors, systematic failures in the assessment of children with acute illness that could be managed in the community.<sup>9</sup>

Following the Darzi Report *High Quality Care for All*,<sup>10</sup> where the importance of measuring patient reported outcomes and experiences was identified, there has been growing interest in the concept of using the Patient Reported Experience/Outcome Measures (PREMs and PROMs) as indicators of the quality/standard of healthcare received by patients. The Darzi Report stated that by measuring the patient experience within the healthcare system, a route would be provided for improving the quality of care. Following the Darzi Report, the NHS Outcomes Framework (2010)<sup>11</sup> has further emphasised the need to measure the quality of care from the patient's perspective and to develop appropriate feedback systems to 'understand and improve the experience of patients'. More recently

the Kennedy report,<sup>12</sup> Department of Health guidance<sup>13</sup> and ‘You’re Welcome’ quality criteria<sup>14</sup> have highlighted the importance of developing tools that are suitable for children to use.

‘Treatment and care should take into account people’s needs and preferences. People with epilepsy should have the opportunity to make informed decisions about their care and treatment, in partnership with their healthcare professionals. If people do not have the capacity to make decisions, healthcare professionals should follow [appropriate advice from DH/Welsh Government]’ (NICE)

In view of this, it is essential that the quality of healthcare provided to children, young people, and their families is examined from a variety of perspectives. This includes individual supervision, reflective practice, local audits and patient and carer experiences, together with national audits and reviews. Children and young people are important, and therefore deserve the same standards of care as any adult. Given their inherent vulnerability and dependence, they require care measures to be appropriate and tailored to their specific needs. Therefore, there is an ongoing need for critical scrutiny of the care provided to children and young people across services in relation to the whole range of conditions and circumstances that children may face.

## 1.2 Clinical Outcomes Review Programmes and CHR-UK

The Clinical Outcomes Review Programmes are a series of national programmes designed to assess the quality of healthcare and stimulate improvement in safety and effectiveness by systematically enabling clinicians, managers and policy makers to learn from adverse events and other relevant data. These programmes replaced the previous confidential enquiry programmes, and the CHR-UK programme was established with two distinct strands in order to explore the quality of healthcare for children and provide data to enable learning and improvements. Module A provided a retrospective epidemiological study of child deaths and hospital admissions in the UK provided information on the overall patterns and trends of both mortality and morbidity that have implications for children’s healthcare. A second strand, involving a themed review of child deaths and serious morbidity across the UK, allowed an in-depth exploration of factors affecting quality of care in children aged one-17 years. A comprehensive consultation process identified mortality and morbidity in children and young people with epilepsies as an important topic for review. This has broader implications for more general aspects of children’s healthcare across the UK.

## 1.3 Childhood epilepsies

Epilepsies are some of the most common chronic neurological conditions of childhood with approximately one in every 200 children affected. The child with epilepsy is by definition at risk of epileptic seizures, but may also have a number of associated neurological, educational or psychosocial problems relating to the cause of their epilepsy or associated co-morbidities. Therefore, the population of children with epilepsies places significant demands on many different types of health service. This includes emergency departments, paediatric wards, paediatric intensive care, neurodisability, and community paediatric services. Children with epilepsies also place demands on non-health services, both in terms of providing care for a child at risk of seizures and in ensuring

that the child can fulfil their potential. HES data indicates that in 2010-11 there were 10,624 hospital admissions of children aged 0 to 14 with a primary diagnosis of epilepsy, and 1,257 with a diagnosis of status epilepticus.<sup>15</sup> The National Report of the Paediatric Intensive Care Audit Network (PICANet), January 2008 to December 2010, listed 1,053 admissions to intensive care units with status epilepticus.<sup>16</sup> Five per cent of emergency department and outpatient paediatric attendances are for children with seizure-related problems.<sup>17;18</sup>

Epilepsy-related deaths can be from a range of causes including the seizures themselves (for example, through accidents or aspiration related events), sudden unexpected death in epilepsy (SUDEP) or related to an associated underlying neurological problem. During 2008-10, the average annual number of registered deaths in the UK for children and young people (1 to 17 years inclusive) with any mention of epilepsy on the death certificate was 107. Epilepsy was recorded as the underlying cause of death in 52.<sup>19</sup>

As each child is different and epilepsy is a heterogeneous condition, a care plan suitable for one child may differ greatly from that of another. In recent years there has been a focus on improving care for people with epilepsies, partly in response to high-profile cases of epilepsy-related deaths and continuing concern regarding high rates of misdiagnosis and mismanagement.<sup>20,26</sup> A number of initiatives have been introduced in response to these concerns and have largely centred on the implementation of National Institute of Clinical Excellence (NICE)<sup>27;28</sup> and Scottish Intercollegiate Guideline Network (SIGN)<sup>29</sup> recommendations. For example, the British Paediatric Neurology Association (BPNA) has introduced a national programme of Paediatric Epilepsy Training (PET) courses aiming to establish pragmatic training of all relevant health professionals. Many regions have developed clinical epilepsy networks to support collaborative working and service development.

In 2002, the National Sentinel Audit<sup>22</sup> investigated epilepsy-related deaths for the first time, concluding that 59% of child deaths were potentially avoidable. Despite the subsequent focus on epilepsy care, there has not been any further national review of clinical care in children and young people with epilepsies.

In general the effective management of a child with epilepsy includes:

- Reducing the risk of seizures by choosing appropriate anti-epileptic drugs while avoiding or minimising side effects
- Reducing the impact of seizures
- Optimising the likelihood of early cessation of prolonged convulsive seizures
- Identifying and treating causative or associated neurological problems
- Maximising educational and quality of life outcomes
- Undertaking individualised risk assessment to balance risk of harm against appropriate participation and quality of life
- Identifying children who may benefit from non-pharmacological treatments such as a ketogenic diet, vagal nerve stimulation, or epilepsy surgery

**Impact of epilepsy on children and their families**

*As mum to a young boy with very difficult to control epilepsy, who has experienced care around the UK, I have witnessed the very best and the very worst of paediatric epilepsy care. I asked my child, a bright 13 year old, what are the worst and the best things about his epilepsy care?*

*The worst were 1) being left alone in a room seizing, which he described as really scary; and 2) doctors who don't talk to him but talk about him, assuming he cannot understand.*

*The best things were 1) staff who talk to him and ask him about himself and his care; 2) getting good care quickly when needed and not being left getting worse and worse; and 3) when things are just right, so that he can get into school and enjoy life.*

*I fully echo what my child says. Good epilepsy care has been due to the expertise of several wonderful and caring neurologists, but good communication between patients, families and health professionals has always been crucial. Staff who ask questions, of their specialist colleagues and of my child and the family, have always been the ones who have given good care. Treating my child as an individual and treating the person, rather than a "condition" or a patient "type", has always been crucial.*

*Education of all health professionals caring for children with epilepsy, not just the specialists; to recognise seizures; to recognise when something other than seizures is the problem; to know when specialist advice is needed; to recognise when prompt action is needed; to correctly administer emergency medication; to understand the potential risks of severe seizures and to recognise that children with severe epilepsy come in all shapes and sizes, with all manner of abilities and are just like other children, much loved and needing the best the NHS can offer, would be a wonderful outcome from this review.*

Zoe Picton-Howell, parent of Adam aged 13

## 1.4 Standards of care

Comprehensive national recommendations for childhood epilepsies were first published by NICE and SIGN in 2004 and 2005 respectively,<sup>27,29</sup> with NICE publishing their revised epilepsy guidelines in 2012.<sup>28</sup> In 2011 the Royal College of Paediatrics and Child Health (RCPCH) led Epilepsy12 National Audit of Childhood Epilepsies published a comprehensive report outlining quality of care for children across the UK.<sup>30</sup> The audit systematically assessed children presenting with seizures to paediatric services and retrospectively reviewed their first year of care against 12 performance indicators derived from NICE and SIGN guidelines. The audit examined how services for children with epilepsies are resourced and whether certain standards of care were delivered. It has revealed significant gaps between national recommendations and care delivered, highlighting variation between providers. For example, the audit showed that 956/1775 (54%) of children diagnosed with epilepsy had no evidence of input from an epilepsy specialist nurse up to 12 months after their first paediatric assessment. Some units were achieving this type of input for all relevant children, while

some units were for a proportion of children, and for other units no input was being received. The report made 12 key recommendations and invited action plans from each paediatric provider.<sup>30</sup> The Epilepsy12 audit did not include in its scope the specific analysis of those children dying with epilepsy or the care of those children experiencing prolonged seizures.

## 1.5 Aims and objectives

The aim of the themed reviews component of the Child Health Reviews - UK programme was to conduct case reviews of deaths and serious morbidity in children and young people<sup>A</sup> aged between one and 17 years (inclusive) with epilepsies<sup>B</sup>, to identify and learn from any clinical, organisational, management or personal issues that may have contributed to the adverse outcomes in order to inform clinical practice and improve clinical services for children with epilepsies across the UK.

The following questions were addressed:

1. What demographic, clinical and system-related (organisational and management) factors are associated with deaths, intensive care and high dependency care admissions in children and young people with epilepsy in the UK?
2. To what extent are these adverse clinical outcomes (deaths and prolonged seizures requiring intensive or high-dependency care) associated with divergence from established best practice in clinical care for children and young people with epilepsy in the UK?
3. To what extent do demographic, clinical and system-related factors differ between those children and young people with epilepsy who die and those children and young people with epilepsy receiving intensive and high dependency care as a result of prolonged seizures?
4. What can be learnt through different approaches to reviewing cases of death or serious morbidity in children and young people with epilepsy (including child death overview panel reviews and serious untoward incident reviews)?

<sup>A</sup> This review included children and young people across the age range of one-17 years inclusive. For simplicity, they are referred to as 'children' throughout the report to encompass both children and young people; where findings or comments relate specifically to teenagers they are referred to separately as young people.

<sup>B</sup> Throughout the report, references are made to children with epilepsies, recognising that the epilepsies are a heterogeneous group of conditions; individual cases are referred to as a child with epilepsy or with an epilepsy.



## 2.0 Review methodology

### 2.1 Developing the methodology

The review aimed to learn from previous confidential enquiries and incorporate aspects into the methodology which have been previously lacking.<sup>31</sup> A mixed-methods approach, combining both quantitative and qualitative data was used that incorporated the collection of questionnaire-based demographic and clinical data, and for selected cases an explicit criterion-based assessment of clinical care and a more in-depth holistic review.<sup>32</sup> This mixed approach allowed an exploration of both evidence-based standards of care and more subtle nuances of care. In addition, the mixed approach also enabled lessons to be learnt within the context of the case. The project was able to link with the ongoing Epilepsy12 audit which is also hosted at the RCPCH,<sup>30</sup> which focuses on quality of care in relation to initial diagnosis and management of children with epilepsies. This enabled the CHR-UK team to set the adverse outcomes explored through CHR-UK in the broader context of the general management and care of all children with epilepsies.

The review has drawn on the principles of systems methodology to go beyond a stance of identifying individual failings, seeking to understand the deeper underlying organisational and cultural issues that allow errors to be made, or for adverse outcomes to arise in spite of good clinical care.<sup>33; 34</sup>

Previous work has emphasised the importance of involving clinicians in case review.<sup>3;32</sup> Such involvement allows cases to be assessed from a clinical perspective and for important factors to be identified and interpreted appropriately. In order to ensure a comprehensive review of cases, CHR-UK used pairs of assessors (a doctor and a nurse) to review each selected case, as well as a structured framework to evaluate the clinical information. A pre-defined explicit criterion-based review was used to measure care against best practice and provide objectivity to assessments of care. Additionally, a structured framework for implicit review was used to allow a greater depth of understanding of each case, and for the identification of factors that may not have been anticipated or captured by the criterion-based review. This framework draws on the National Framework for Reporting and Learning from Serious Incidents Requiring Investigation;<sup>35</sup> in particular, their tool for Root Cause Analysis investigation.

The results of the case assessments were combined with clinical information provided directly by the involved clinicians. The CHR-UK team analysed all these data using appropriate quantitative and qualitative methods of analysis, informed by a Topic Expert Group (TEG) bringing broad expertise. This approach brought methodological rigour to the research, ensuring that the analysis was based on the case data and informed by an evidence base of literature and clinical context.

Questions for the clinical questionnaire, criterion-based assessment and holistic frameworks were designed through advice from the TEG. The TEG met periodically throughout the project to review progress and advise on any developments. In the later stages, the TEG contributed to interpretation of the results and their translation into learning and clinical implementation (see Appendix 1 for more information on topic selection and the TEG).

A detailed protocol was developed by the CHR-UK team at the RCPCH in consultation with relevant methodological and clinical experts. The draft protocol was sent to other experts for independent peer review and their comments were then taken into account in developing the protocol. This process ensured both academic rigour in the methods applied and clinical relevance of the approach. In addition, key stakeholders, including relevant groups of patients and carers were consulted and were given opportunity to comment on the draft protocol.

## 2.2 Case definition and inclusion criteria

Once the topic of epilepsy was selected, the CHR-UK team and TEG wanted to ensure that the review covered the entire patient pathway, including primary and emergency care, together with the inclusion of serious morbidity as well as mortality. The CHR-UK team recognised that many of the deaths in children and young people with epilepsy would be from causes other than their epilepsy, and that there was potential for misdiagnosis and inappropriate classification of deaths in epilepsy. In order to explore these issues, ensure that no relevant cases were excluded, and to look at broader aspects of care for these children, the scope of the review was expanded to include all deaths in children and young people with epilepsy, rather than just seizure-related deaths.

The concept of serious morbidity is complex, and it was recognised that the full scope of this could not be covered within this programme. For epilepsy, children and young people receiving intensive or high dependency care for prolonged seizures was used as a measure of serious morbidity, which was considered achievable and clear to define.

Case definitions and inclusion criteria were drawn up in consultation with the TEG and taking account of internationally agreed definitions (Box 2.1; glossary).

### Box 2.1: Inclusion criteria

**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 18<sup>th</sup> birthdays at the time of incident

**AND**

- d. Prior to the incident the child or young person had a diagnosis of epilepsy (based on two or more epileptic seizures more than 24 hours apart that were not acute symptomatic seizures or febrile seizures).

The definitions used are provided in the glossary

## 2.3 Case notification

An electronic reporting system, modelled on that used by the British Paediatric Surveillance Unit (BPSU), was used to collect notifications of children and young people who met the inclusion criteria for the review. Case notifications were collected over a 10 month period from 01 June 2012 to 31 March 2013. Due to the length of time required to obtain various approvals and developments, together with the limitations of a two year contract period, the timescales for data collection had to be compressed to a 10 month timescale. An active surveillance model was used with monthly emails sent to all RCPCH-registered consultant paediatricians requesting they respond, whether or not they had seen a case. A reminder email was sent to all consultants who had not responded to the monthly email after two weeks.

When reporting a case the consultant was re-directed to an electronic reporting card on the RCPCH website. This enabled the consultant to record the type of case they had seen (death, intensive care or high dependency care case) and their own contact details to enable the CHR-UK team to monitor respondents. Death certificates were not included as a notifier due to time delays in obtaining these data. No patient identifiable details of the case were collected at this stage of data collection.

Consultants unlikely to come into contact with children with epilepsy were given the option of opting out of the review; those doing so were sent no further notification emails. If a consultant opted out of the review, and later found they did have a case to report, they were able to report the case to the CHR-UK team and add themselves to the mailing list.

In addition to all consultant paediatricians registered with the RCPCH, other non-paediatric consultants were able to notify cases, and information about the review was disseminated to relevant groups of professionals, including PICANet. A data sharing agreement was set up between the CHR-UK team and PICANet, whereby PICANet, on behalf of the CHR-UK review, contacted units who had admitted children and young people following a prolonged seizure. PICANet wrote to each unit asking the consultant overseeing the care of the child to notify the case to CHR-UK.

Details of case ascertainment and validation are covered in Chapter 7 with comments on case ascertainment of mortalities using data from ONS, GRO, NISRA and PICANet for the morbidity group.

## 2.4 Clinical questionnaire

For each case reported to the review, the reporting consultant was sent a link to the RCPCH secure electronic web portal (EWP) and asked to complete a clinical questionnaire (Appendix 2). In the event the reporting consultant did not feel they were able to complete the questionnaire they were asked to identify a clinician who could complete the questionnaire. Clinicians were required to register their details onto the EWP and, once the CHR-UK team had verified and approved the clinician's registration, the clinician was sent a username and password to log in. For each questionnaire the EWP generated a unique CHR-UK project identifier allowing the CHR-UK team to refer to a case without using patient identifiable information.

The clinical questionnaire provided a descriptive clinical dataset of all cases notified to the CHR-UK project. It included a minimum number of patient and hospital identifiers needed to ensure that questionnaire data on a single case submitted by two or more clinicians could be merged and questionnaire data could be linked to case notes. The clinical dataset was used to guide case selection for detailed case review and to provide background demographic and clinical information on the reported cases (Box 2.2).

#### **Box 2.2: Clinical questionnaire domains**

1. Inclusion/exclusion criteria (any case not meeting the inclusion criteria were filtered out at this stage and no further information will be collected).
2. Case identifying details, including patient name, date of birth, date of incident, death or admission to intensive or high dependency care, postcode, NHS number, reporting paediatrician and contact details, hospital, other professionals involved together with whether any other investigation has taken place into the incident. These data were stored separately from the other sections of the clinical questionnaire.
3. Generic questionnaire with core questions on the circumstances of the incident, factors intrinsic to the child (e.g. co-morbidities, education), factors in the family and environment, parenting and care and general service provision.
4. Pre-incident care questionnaire (for all cases), relating to initial diagnosis and ongoing management prior to the incident.
5. Emergency department management and pre-hospital care (for all cases).
6. Intensive and high dependency care management (for all cases which received intensive or high dependency care, including those children dying following admission to intensive or high dependency care).
7. Management following the death, covering general principles of the clinical response, investigation of the death and support for the family (for all deaths).

For each section, the reporting clinician was asked to complete as much of the questionnaire as they were able or, if they were unable to complete it, to identify which clinician would be able to complete it. Completed questionnaires were then checked by the CHR-UK team for completeness and accuracy.

## **2.5 Case selection for detailed case review**

Cases were selected across both the mortality and morbidity groups for more detailed case review. The CHR-UK review aimed to include each death while recognising that 100% recruitment may not be achieved. In relation to the intensive care and high dependency care admissions, a purposive sample of cases was used to ensure inclusion of the following:

- Boys and girls
- Three age groups: under five years old, five to 12 years old and 13 to 17 years old
- Epilepsy as sole diagnosis, and epilepsy with concurrent learning difficulties or other disabilities ('epilepsy plus')
- White Caucasian and ethnic minorities
- Intensive care and high dependency care
- All UK devolved nations

Intensive and high dependency care cases were recruited sequentially. Each month, the cases selected were reviewed according to the purposive sampling criteria, and groups that were underrepresented (particularly young people aged 13-17 years and those from the UK devolved nations) were preferentially selected.

## 2.6 Case notes review

### 2.6.1 Case assessment tool

A case assessment tool was developed by the CHR-UK team in conjunction with the TEG. This was structured around six phases of care in 61 cases of child mortality and morbidity:

- initial diagnosis and management (all cases);
- ongoing management from initial diagnosis to reported incident (all cases);
- pre-hospital care (all cases);
- emergency department care (all cases);
- intensive or high dependency care (prolonged seizure group only); and
- care of the child and family around and following the death (for children who died).

The case assessment tool combined a criterion-based assessment, based on recognised clinical standards (Box 2.3), and a structured implicit review for each phase of care.

#### Box 2.3: Guidance used to develop standards

- The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care NICE 2012<sup>28</sup>
- Diagnosis and management of epilepsies in children and young people SIGN 2005<sup>29</sup>
- Paediatric advanced life support Resuscitation Council (UK) 2010<sup>36</sup>
- Integrated Multi-Agency Care Pathways for Children and Young People with Life-Threatening or Life-Limiting Conditions and their Families ACT 2004<sup>37</sup>
- Working Together to Safeguard Children, Chapter 7, Child Death Review Processes Department for Education (2010)<sup>38</sup>

Forty-two key clinical questions were developed for the criterion-based review. Supplementary factual questions were also included in the case assessment tool (Appendix 2) to enable the case assessors to answer these key clinical questions.

The framework for the structured implicit review was developed by the CHR-UK team in consultation with the TEG and guided by the literature available on epilepsy management. The structured implicit review allowed the case assessors to identify key learning points from the case and to make judgments about the presence of avoidable or remediable factors, together with identifying elements of good practice and care. For each of the six phases of care, the case assessors, having completed the criterion-based assessment, would discuss the case as a pair and grade the quality of care in that phase according to a six point grading system, adapted from a comparison of case note review methods<sup>32</sup> (Box 2.4). For each case where a child died, the case assessors were required to classify the cause of death according to a six category system (Appendix 4), developed by the CHR-UK team in conjunction with the TEG. For quality assurance purposes, the RCPCH lead reviewed the case notes and classified the death for each child undergoing case note review for comparison with that of the case assessors.

#### **Box 2.4: Grading of care (implicit review)**

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient
4. This was satisfactory care, falling short of current best practice in two or more minor areas
5. This was good care, which fell short of current best practice in only one or two minor areas
6. This was excellent care and met current best practice

### **2.6.2 Obtaining case notes**

For each case selected for detailed case review, the case notes were requested from the child's first seizure through to the notified incident. Case notes were requested from all relevant healthcare settings, including hospitals, community care, tertiary care units, primary care, hospice and respite care.

All settings were contacted by letter and follow-up phone calls requesting copies of case notes to be sent to RCPCH in two sealed envelopes via special delivery. The CHR-UK unique project identifier and one patient identifier (NHS/CHI number) were included on initial correspondence with healthcare settings, so the healthcare setting could identify the child. The CHR-UK project identifier was used in further correspondence with the healthcare setting to refer to the case. The Caldicott Guardian of each relevant trust was informed by letter that data were being collected.

The CHR-UK team monitored the arrival of case notes and chased up healthcare personnel as necessary. Follow up of case notes ceased for a case when either all case notes had been received by the RCPCH or the agreed four week timeframe had passed. All case notes received at the RCPCH were anonymised and all patient identifiable details removed, while staff and hospital details were not. The notes were labelled with the unique CHR-UK project identifier and stored in the RCPCH secure storage facility until case assessments took place, after which they were destroyed.

### 2.6.3 Case assessments

Case assessors were recruited from paediatric consultants and ST6-8 level trainees, all with a special interest in epilepsy or neurodisability using an open recruitment process with advertisement on the RCPCH website, e-bulletins and e-portal. Paediatric nurses were recruited through the Royal College of Nursing, Epilepsy Specialist Nurse Association and Epilepsy Action nurse networks. All case assessors attended one day training at the RCPCH. This included specific training in methods of case note review, topic-specific training on epilepsy care, the specific use of the case assessment tools developed for this project, the HQIP *cause for concerns policy*<sup>39</sup>, together with data protection and security legislation. Additionally, all case assessors had completed PET1 epilepsy training or equivalent.

Case assessments were carried out at the RCPCH and on-site in hospitals. Where hospitals in England and Wales were willing to accommodate the case assessors arrangements were then made for the assessors to attend the hospital for a day to complete the case assessment.

Case assessors worked in pairs comprising of one paediatrician and one paediatric nurse to carry out the case note review. Assessors were allocated to cases outside their own trusts and regional areas and were asked to notify the CHR-UK team if there was any conflict of interest while reviewing the notes. When completing the assessment tool if the assessors could not agree on a response they were asked to try to reach a consensus. However, if a consensus could not be reached the assessors were able to record their responses separately in the case assessment tool.

### 2.6.4 RCPCH assessments

The majority of case assessments were carried out at the RCPCH; this included all cases from Scotland and Northern Ireland. Pairs of assessors were given access to all case notes held at the RCPCH, and entered data directly onto a laptop. All assessors were provided with a case assessor handbook, British National Formulary for Children (BNFC) and access to ICD-10 codes to aid them in completing the assessment tool. Assessors did not have access to any patient identifiable data. Assessors were informed that no hospital or staff data were to be recorded in the case assessment tool. The CHR-UK team were on hand to support case assessors when carrying out case assessments at the RCPCH, helping with any technical issues and answering any questions regarding the assessment tool. Members of the TEG and the RCPCH lead of the project attended a proportion of RCPCH-based assessments to provide support and guidance on clinical issues. Upon completion of each case assessment the assessors were given the opportunity to discuss the assessment with the CHR-UK team and raise any concerns regarding the care provided. Following completion of the case assessment, all case notes and completed case assessment tools were collected by the CHR-UK team.

### 2.6.5 Hospital assessments

Hospital-based case assessments were only carried out in England and Wales, and if the hospital was willing to accommodate the case assessors. For each hospital-based assessment the clinical audit department was contacted to make arrangements for the case assessors to visit the hospital for a day. Hospital clinical audit staff members were asked to make all case notes and other relevant records available to the case assessors for their visit, provide assessors with a room to carry out the assessment and collect the case notes with the assessment tool once it had been completed.

The assessments were carried out in the hospital where the child was located at the time of the reported incident. Due to case assessors only having access to the hospital notes, the assessors attended an additional assessment day at the RCPCH to complete the assessment on case notes and records from other healthcare settings.

### 2.6.6 Completing the assessment tool

For case note review, pairs of case assessors jointly reviewed the available clinical notes and completed the case assessment tool. The case assessors recorded their assessments onto electronic versions of the tool or, in the case of hospital-based assessments, onto paper versions of the tool which were later transcribed onto a database by the CHR-UK team. Where case assessors did not agree, both views were taken into consideration in the analysis and interpretation of the results.

The case assessors recorded their reasoning behind their grading of the case. They were encouraged to comment on the care received during each phase, as well as commenting on any notable aspects of care. Each individual case assessor was able to record their grading of the case. Where individual members of a pair differed in their assessment, they were asked to record the reasons for disagreement. After completing their assessment of all relevant phases of care, assessors provided an overall judgement of the care received by the child and family together with the quality of the case notes reviewed.

To ensure reliability and consistency across case assessments, case assessor pairs rotated, so that the same individuals were not always placed together. The first five case assessments carried out were double assessed, allowing the CHR-UK team to compare assessor responses to ensure understanding of how to complete the assessment tool and to extract information so lessons could be learnt. The CHR-UK team made any necessary amendments to the assessment tool and clarified any language or phrases used based upon the information they provided. Double assessments continued through the course of the data collection period for quality control; a total of 11/61 cases (18%) were double assessed.



### 2.6.7 Cause for concern

Where any case assessors identified concerns that indicated a practitioner, team or Trust might place future patients at risk, assessors notified the CHR-UK team. If the CHR-UK team found there to be a significant cause for concern and there was no evidence that this had been investigated or addressed locally, the incident was reported to HQIP in line with their “cause for concerns” policy.<sup>39</sup>

## 2.7 Northern Ireland data collection processes

Data collection in Northern Ireland was coordinated by the Northern Ireland Maternal and Child Health (NIMACH) office. Once notified, the CHR-UK team passed on the contact details of the reporting clinician with the unique project identifier to the NIMACH team for follow up. Northern Ireland has different legislation in place to the rest of the UK in that no patient identifiable data are able to leave Northern Ireland. Thus Northern Ireland based consultants were unable to complete the clinical questionnaire on the EWP. NIMACH contacted consultants directly asking them to complete a paper-based clinical questionnaire on notified cases.

For all morbidity cases Northern Ireland consultants sought consent from the parents, carers or patients before any data were collected. Consent was not required for mortality cases. The reporting clinician completed the clinical questionnaire with patient identifiable information and returned to NIMACH, who entered the clinical questionnaire data minus the patient identifiable information onto the EWP. The questionnaire data in the EWP were then used to select cases for assessment.

Once the CHR-UK team selected a Northern Ireland case to undergo detailed case assessment NIMACH collated and anonymised all case notes from all relevant healthcare settings before sending them to the RCPCH London office.

## 2.8 Service description questionnaire

For each case reviewed, the CHR-UK team collected data on the services available for the unit in which the child or young person was treated. The questionnaire was pre-populated with data previously captured in the Epilepsy12 audit<sup>30</sup> before being sent out to the Epilepsy12 audit leads to review and amend as necessary (See Appendix 5).

## 2.9 Ethics and data security

### 2.9.1 Ethics and permissions

Ethical advice was sought from the National Research Ethics Service, who confirmed that the remit of this work would not require ethics approval.

Patient identifiable data were collected at the clinical questionnaire and case assessment stage for the purposes of linking clinical questionnaires completed on the same case by more than one clinician and linking the clinical questionnaire to case notes.

CHR-UK was granted National Information Governance Board 251 approval and Scotland Caldicott Guardian approval to collect patient identifiable data without consent. The Northern Ireland Privacy Advisory Committee granted permission for the collection of patient identifiable data without consent for cases of children who died. For cases of serious morbidity, consent was required from the child's parent or carer. The project received permissions to send notification emails to Jersey, but permission was not received to send to Guernsey.

### 2.9.2 Data security

The clinical questionnaire was held on the EWP which enabled the CHR-UK project to collect and store data securely. Fry, an independent company providing Information Technology services, was contracted to develop the EWP using the open source data management system software CKAN, which powers the UK government data.gov.uk open data portal.

The EWP specification included a module enabling system administrators to identify which data fields are to be removed and replaced by an encryption key. All questionnaires created were assigned a unique identifier, specifically for the purposes of identifying the case without reference to patient identifiable information. Only CHR-UK team members had system administrator rights and were able to decrypt and access patient identifiable data. Minimal patient identifiable data were kept on the EWP throughout the data collection period to ensure data and records could be linked, and to ensure data completeness. Once the data collection period ended and all data were linked, the dataset was fully anonymised and all hard copy case notes and assessment tools were destroyed.

All staff involved with the project were required to sign a confidentiality clause requiring staff to comply with data protection legislation and not to disclose any information considered confidential by the RCPCH, either during or following termination of employment. Case assessors recruited to work on the project were required to sign the confidentiality clause and to declare any conflicts of interest.

All case notes received at the RCPCH were matched to data stored on the EWP for that case, using patient-identifiable information. All patient identifiers were removed from case notes and labelled with the unique project number assigned to the case at the point of the questionnaire being created. Anonymised case notes were stored in the secure storage facility in accordance with the RCPCH data security policy.

## 2.10 Analysis and interpretation

Clinical questionnaire data were analysed using the Statistical Package for the Social Sciences (SPSS). Descriptive statistics were used to provide a profile for the entire group of reported cases. There were a limited number of textual comments included in some clinical questionnaires. These were analysed as qualitative data, through direct reading of the comments to identify important points. These data were used to supplement the qualitative data obtained through the case notes review.

Quantitative data from the case assessment tools were transferred to an SPSS database and linked to the clinical questionnaire data for those cases, using the unique project identifier. Each case was checked for the level of agreement or disagreement between case assessors; for those cases that had been assessed by more than one pair of assessors, both sets of assessment were compared. For the purposes of quantitative analysis, only the first assessor's responses were used. However, where the first assessor had recorded an answer as 'unclear' or had left a response blank, and a subsequent assessor had provided a clear answer to the question, the response was amended. Where there were disagreements between assessors, the RCPCH project lead reviewed all responses and accompanying comments, and amended the response if there was a clear indication to do so.

Descriptive data were produced on each phase of the care pathway, using the case assessors' responses. For an initial profile of the children, and for consideration of the initial diagnosis and management, all children were included, regardless of outcome. For children who presented with prolonged seizures, data on their pre-hospital care, emergency department care and intensive or high dependency care were analysed. This included those children who presented with prolonged seizures and subsequently died. For the children who died (including those initially receiving intensive or high dependency care), data on the circumstances and response to the child's death were analysed.

For each phase of care, consideration of the quality of care provided to the child and family was ascertained from the case assessors' grading of care using the six point grading tool (implicit review; Box 2.4) and from a compilation of the case assessors' responses to the key clinical questions in the criterion-based review, this led to a four point grading (Box 2.5).

### Box 2.5: Quality of care (Criterion based review)

1. On the basis of the available recorded information, the care provided to the child and family in this phase met current agreed standards of management
2. On the basis of the available recorded information, the care provided to the child and family in this phase did not meet one or more current agreed standards of management
3. On the basis of the available recorded information, the care provided to the child and family in this phase did not meet any of the current agreed standards of management
4. There is insufficient available recorded information to judge the quality of care provided to the child and family in this phase

Qualitative analysis was carried out using a framework approach based on that developed by Ritchie and Spencer for applied policy research.<sup>40</sup> The CHR-UK team prepared a case summary for each

case based on the information recorded by the case assessors and the clinical questionnaire data. Qualitative data on each phase of care were entered into an access database. Since the assessment tool was highly structured within the six phases of care (see section 2.6.1), this provided the initial framework for analysis. All comments by the case assessors within each phase were carefully read and re-read to identify emerging themes.

The data were discussed within the CHR-UK team to clarify and modify the themes. In a series of meetings, members of the TEG were presented with both quantitative and qualitative data from the case assessments, and asked, in small groups, to identify the themes that they felt were emerging from these data. These themes were then compared with those identified by the CHR-UK team, allowing modification and clarification of the core themes and further review of the source data. A secondary review of the data was carried out in the light of these themes, to identify consistencies and discrepancies in the data and to triangulate the qualitative and quantitative data before compiling the final report.

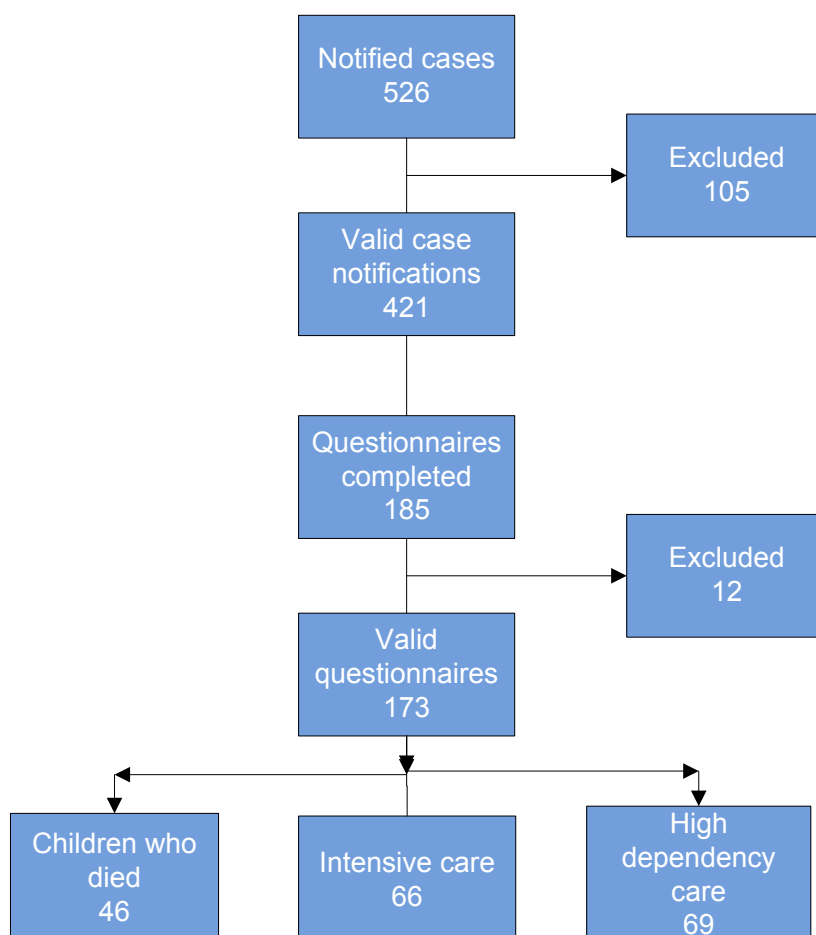
For cases that reviewed by a pair of assessors the intra-rater reliability was calculated. This measured the degree of difference between two assessors reviewing the same case. Intra-class Correlations (ICC) was calculated in SPSS for all cases assessed by a pair of assessors, with missing values and single assessments removed for purposes of this analysis.

## 3.0 The children and young people in the review

### 3.1 Case notification

Case notification took place over a 10 month period, from 01 June 2012 to 31 March 2013. During this period notification emails were sent to all RCPCH registered consultant paediatricians. From June to August emails were sent to England and Wales only, from August onwards Scotland and the Isle of Man were also included, and Northern Ireland and Jersey from September. The response rate for consultants over the 10 month period was 39% (further details are provided in Chapter 8). In total, 526 cases were notified, with the majority of these being from England (433). There were 105 invalid reports; 11 due to duplicate notifications, 59 cases not meeting the inclusion criteria, 20 clinicians being unable or unwilling to complete the questionnaire and 15 in Northern Ireland, from an inability to obtain informed consent within the time span of the review, therefore leaving 421 valid case notifications (Figure 3.1).

**Figure 3.1: Case notifications and questionnaire completion**



Completed clinical questionnaires were received for 185 cases, of which nine were found to be duplicate notifications and three did not fit the inclusion criteria. This left 173 confirmed cases in 162 children (i.e. some children had multiple admissions) with clinical questionnaire data (41% of all valid notified cases). Eight children who died had been admitted with prolonged seizures prior to

their death. The majority of cases were reported from England with numbers from Scotland, Wales and Northern Ireland reflective of their smaller populations. There were no cases notified from the Channel Isles or Isle of Man (Table 3.1).

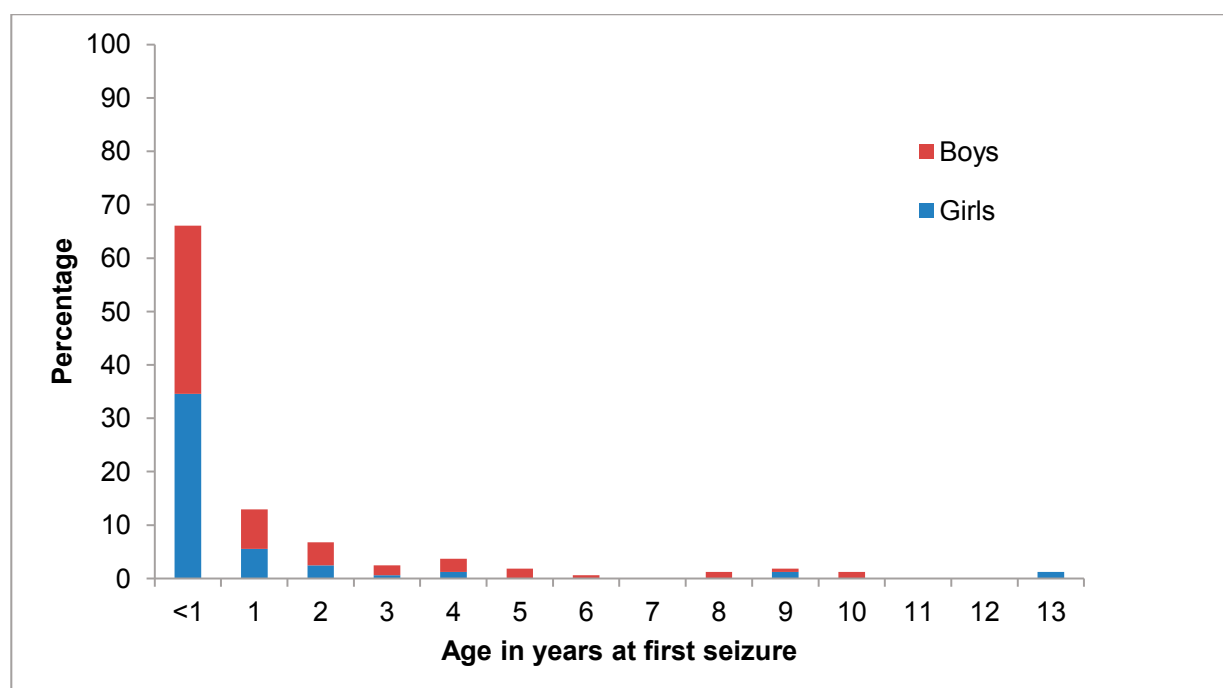
**Table 3.1: Total cases with completed questionnaires from each devolved nation**

Devolved nation	Children who died	Episodes of intensive care	Episodes of high dependency care	Total number of children (excluding duplicate or repeated admissions) %
England	33	64	62	141 (87%)
Scotland	5	1	3	9 (6%)
Wales	5	1	4	9 (6%)
Northern Ireland	3	0	0	3 (1%)
<b>Total</b>	<b>46</b>	<b>66</b>	<b>69</b>	<b>162 (100%)</b>

### 3.2 Case characteristics

The age and gender of the 162 children in the review are presented in Figure 3.2 and Table 3.2. There was a slight preponderance of boys (53%), and a skewed distribution with more young children. The age of the children at the time of their first seizure ranged from 0 months to 13 years 10 months (median age four months); their age at the time of the reported incident ranged from 12 months to 17 years and six months (median 65 months). In total, 42 (26%) children were of non-white ethnicity (Table 3.3). It is not clear whether this represents a true bias for children notified in this review.

**Figure 3.2: Age (in years) at time of first seizure**



**Table 3.2: Age and gender at time of incident**

Age group	Girls	Boys	Total (%)
1-4	36	33	69 (43%)
5-9	16	27	43 (27%)
10-14	18	19	37 (23%)
15-17	6	7	13 (8%)
<b>Total</b>	<b>76 (47%)</b>	<b>86 (53%)</b>	<b>162 (100%)</b>

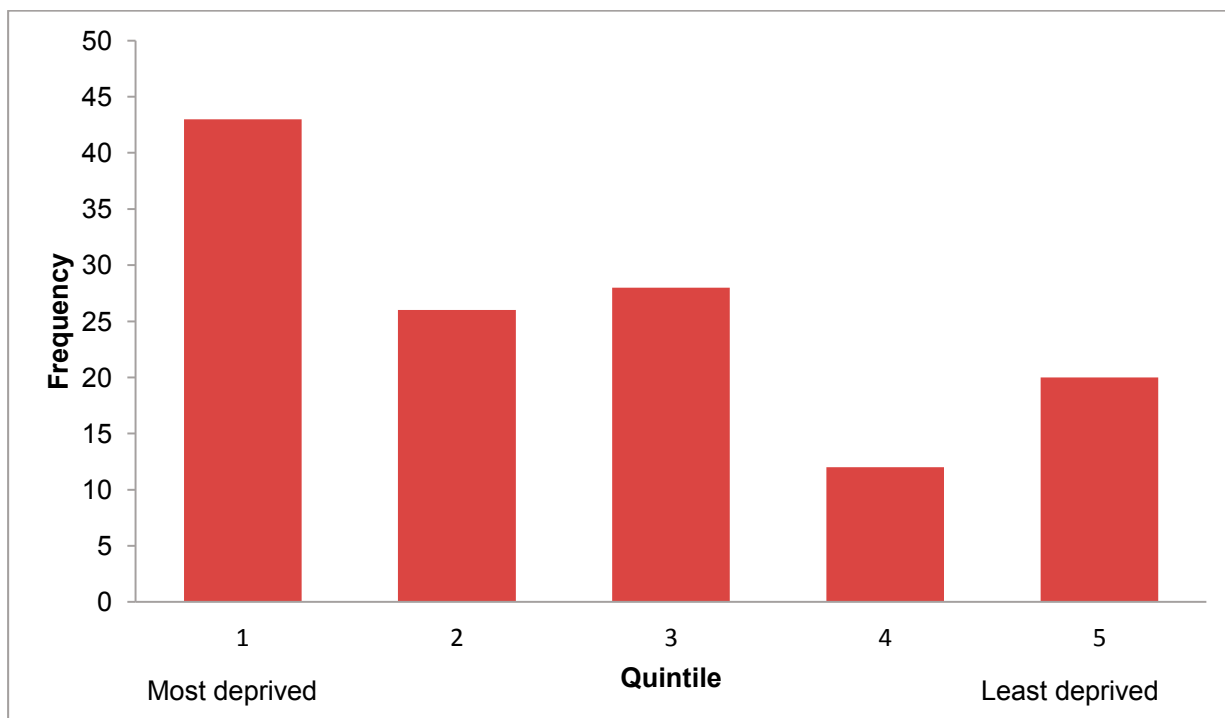
**Table 3.3: Ethnicity**

Ethnicity	Total (%)
Asian or Asian British	15 (9%)
Black/African/Caribbean/Black British	11 (7%)
Mixed/multiple ethnic groups	11 (7%)
Other ethnic group	5 (3%)
White	114 (70%)
Unknown	6 (4%)
<b>Total</b>	<b>162 (100%)</b>

### 3.3 Socio-economic status

Using the postcode of the child’s usual residence, the lower layer super output area of each child was found and matched to the Index of Multiple Deprivation.<sup>41, 44</sup> There was a tendency for cases to come from more deprived areas, with 43% of children living within the most deprived 20% of areas (Figure 3.3). It is not clear whether this represents a true bias towards more children from more deprived areas, or whether such cases were more likely to be reported to the review.

**Figure 3.3: Numbers of children living in each quintile of the Index of Multiple Deprivation**



### 3.4 The nature of the epilepsies

An epilepsy syndrome was identified in 31 (19%) children, of which the most common were West syndrome (eight cases) and Dravet syndrome (seven cases). A cause of epilepsy was identified for 100 children (62%) (Table 3.4), however for children where the reporting clinician did not provide a cause, 11 (7%) had been previously diagnosed or had an identified specific epilepsy syndrome. Therefore, 111 (69%) children had either an underlying cause or epilepsy syndrome identified. Known developmental impairments were reported in 137 children (85%); this included developmental delay or learning difficulties (75; 55%) ranging from mild to severe, cerebral palsy (19; 14%), autism spectrum disorders (nine; 7%), and regressive neurological conditions (six; 4%).

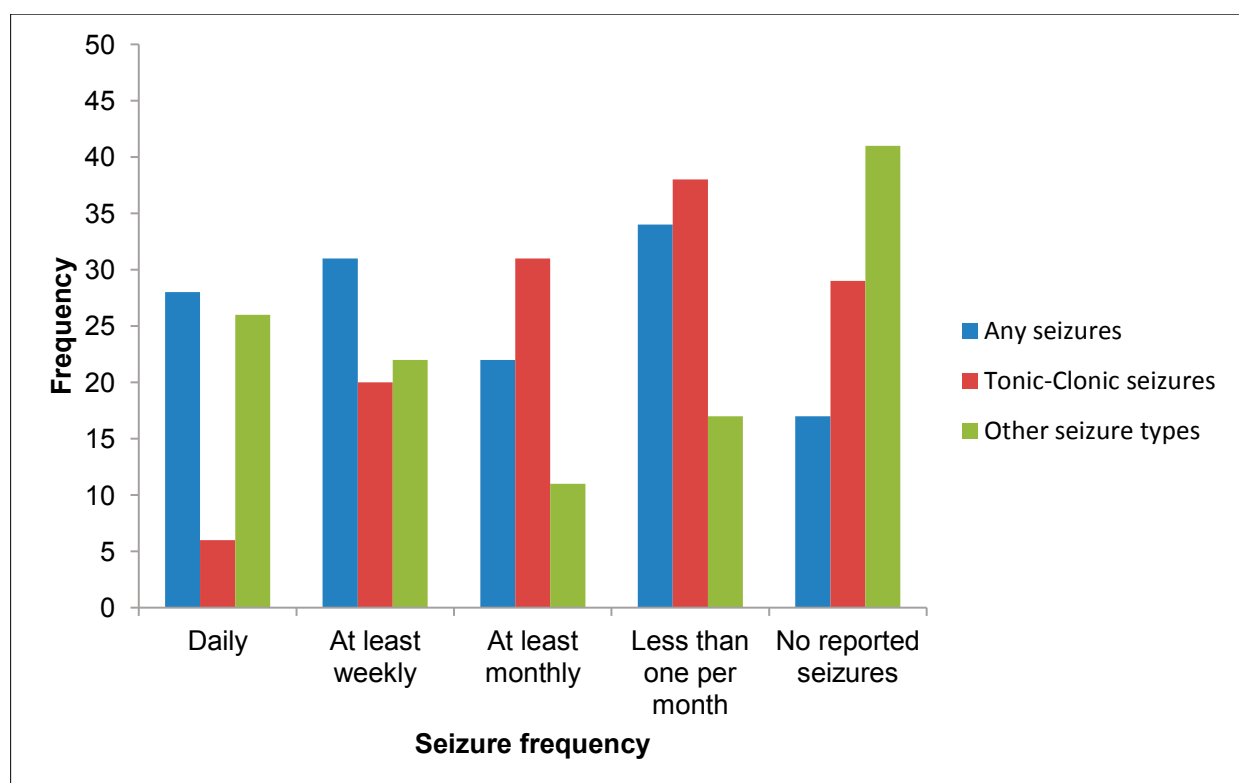


**Table 3.4: Identified causes for the epilepsy**

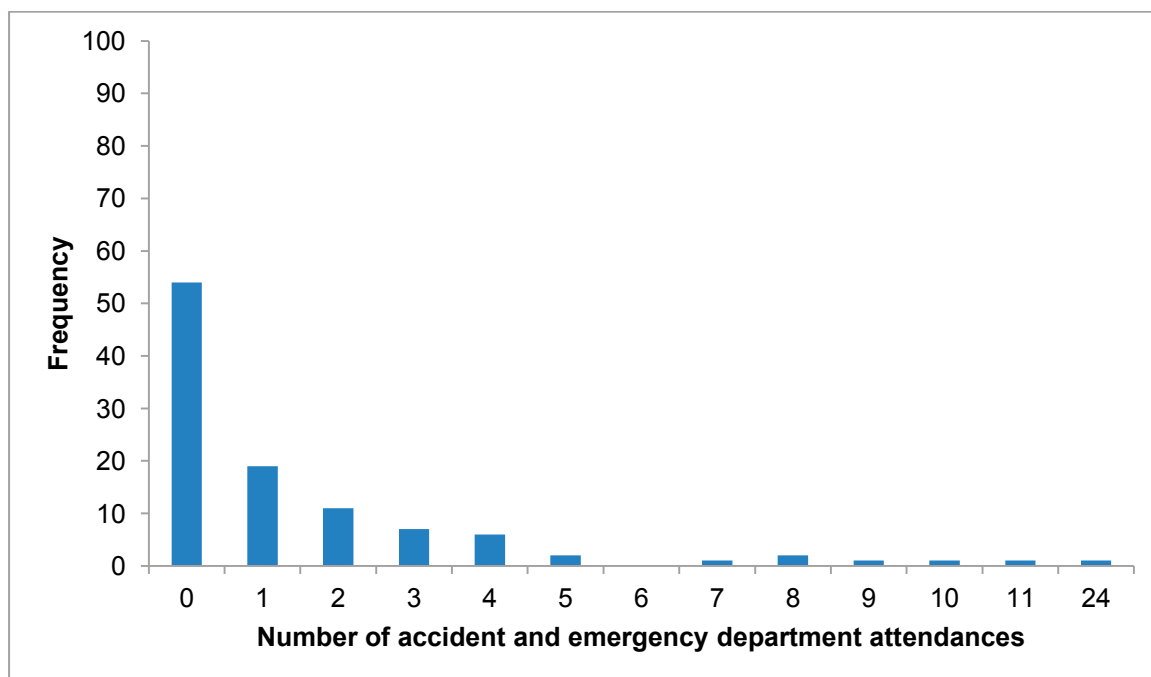
Identified cause	Number (%)
Identified genetic disorder	32 (20 %)
Hypoxic-ischaemic neonatal brain injury	29 (18%)
Cerebral malformation	21 (13 %)
Other structural/metabolic	18 (11 %)
Unknown	62 (38 %)
<b>Total</b>	<b>162 (100%)</b>

The most common seizure types were recorded as generalised tonic clonic seizures (102; 63% of children), followed by focal or secondary generalised (70; 37%), absence seizures (46; 28%), myoclonic (28; 17%) and infantile spasms (21; 13%). More than one seizure type was reported in 81 children (50%), with 40 (25%) having at least three seizure types.

Data on the frequency of seizures in the six months prior to the reported incident were available for 131 (81%) children. Of these, 114 (87%) children had previously experienced seizures with 28 (17%) experiencing seizures on a daily basis and a further 31 (19%) at least weekly (Figure 3.4). In the 12 months prior to the incident, 52 (49%) children had attended an accident and emergency department due to prolonged seizures with seven (14%) having had at least six attendances in that time (Figure 3.5). Sixty-seven (41%) children had, at some stage, previously been admitted to intensive or high dependency care.

**Figure 3.4: Frequency of reported seizures in six months prior to the incident (n=131)**

**Figure 3.5: Frequency of accident and emergency attendances with prolonged seizures in 12 months prior to the incident (n=106)**



### 3.5 Factors in relation to the child, their environment or the care they received

In 141 (82%) of the 173 reported incidents the child was being looked after by their parents at the time of the incident, often with other carers or healthcare professionals also present (Table 3.5). On most occasions (74%) the child was at their home at the time of incident. Two children were in residential care, one in a residential facility for children with complex disabilities and the other in a residential mental health facility. One child was reported to be subject to a child protection plan and was residing with other family members.

**Table 3.5: Who was present at the time of the incident**

Present at the time of the incident	Number (%)	
Parents	130	(75%)
Parents & other healthcare professionals/carers	11	(6%)
Other Carers	7	(4%)
Healthcare professionals	13	(8%)
School	7	(4%)
None	1	(1%)
Not known	4	(2%)
<b>Total</b>	<b>173</b>	<b>(100%)</b>

The majority of reporting clinicians portrayed pictures of children with complex health needs, including difficult to control epilepsy or multiple co-morbidities, with devoted parents who were working with professionals to provide the best care possible for their child.

*This child enjoyed as much love and care as any child could from their devoted family. The family engaged with professionals and followed all advice. When it became clear how extensive the brain damage was and that the child, despite their seizures coming under control, was not making any developmental progress at all and slept most of the time, an emergency health care plan was agreed with all involved professionals and parents to allow a natural death when the time came. The child received palliative care throughout an episode of bronchiolitis at home and survived a number of weeks after this. The child died peacefully at home with their family after a further short respiratory illness. (Case assessor comment)*

In nine cases the reporting clinician raised concerns regarding family engagement including one where the clinician had concerns about possible 'pseudoseizures'. The remaining eight cases involved missed appointments. In seven cases the clinician reported concerns with adherence to treatment.

### 3.6 Profile of cases reviewed

A total of 69 incidents in 61 children were subject to case note review. The background characteristics of the children are given in Table 3.6, with corresponding data from the clinical questionnaires. Forty-six questionnaires were received for children who had died, however due to a delay in questionnaire submission only 33 (72%) cases were subject to case note review. Seventeen (28% of all cases reviewed) were admitted to intensive care, of whom six subsequently died; and 19 (31% of all cases reviewed) were admitted to high dependency care, of whom two died. Case notes for the 61 children were received from hospitals (91%), community care (8%), GP (43%), hospice (7%), and epilepsy nurses (5%). In accordance with the selection criteria, notes were requested on all cases where a child died and a purposive sample of those with prolonged seizures. A mix of gender, age group and ethnicity for the children admitted with prolonged seizures was achieved. No case records were provided for intensive or high dependency care cases from Scotland or Northern Ireland due to delays in questionnaire submission and gaining parental consent. Otherwise, the selection represented the constituent countries.

**Table 3.6: Background characteristics of the children**

Characteristics		Children who died N=33		Children receiving high dependency/ intensive care for prolonged seizures N=36 (8 of whom died)		All children (clinical questionnaire data) N=162	
<b>Gender</b>	Boys	19	(58%)	20	(54%)	86	(53%)
	Girls	14	(42%)	16	(46%)	76	(47%)
<b>Age group</b>	1-4	15	(45%)	15	(43%)	69	(42%)
	5-9	8	(24%)	6	(16%)	43	(27%)
	10-14	7	(21%)	11	(30%)	37	(23%)
	15-17	3	(9%)	4	(11%)	13	(8%)
<b>Ethnicity</b>	White	23	(70%)	26	(72%)	114	(70%)
	Non-White	10	(30%)	10	(28%)	42	(26%)
<b>Country of Origin</b>	England	25	(76%)	32	(89%)	141	(90%)
	Scotland	3	(9%)	0	(0%)	9	(5%)
	Wales	3	(9%)	4	(11%)	9	(5%)
	Northern Ireland	2	(6%)	0	(0%)	3	(2%)
<b>Concurrent disability</b>		31	(94%)	17	(47%) <sup>1</sup>	137	(85%)

### 3.7 Services provided by units

For each case undergoing review, data were gathered on the services provided in each unit where the child presented for the notified incident. Fifty-three service description questionnaires were sent to Epilepsy12 Audit leads which covered all 61 cases that underwent case note review. Forty-five (85%) questionnaires were completed and returned.

Epilepsy12 unit leads reported an average of 11.1 whole time equivalent (WTE) general paediatric consultants employed and 1.6 WTE general paediatric consultants with 'expertise in epilepsy'. Over half (67%) of all audit units had at least one epilepsy specialist nurse. The average number of consultant (or associated specialist) led secondary level 'epilepsy clinics' per week for children or young people was 1.1. Thirty-seven (82%) of the units held at least one consultant led epilepsy clinic per week. Twenty-two (49%) units reported a database or register for children with epilepsies. Thirty-eight (84%) hosted a paediatric neurology outpatient service.

## **3.8 Summary**

This Chapter presents data on the characteristics of 173 incidents (deaths and prolonged seizures) that occurred in 162 children notified to the review. These children tended to be under 10 years old (70%), with 63% presenting with their first seizure before the age of four. A high proportion of the reported children were from ethnic minorities and the more deprived socio-economic areas. On the whole, the reported children with epilepsies were found to have a high prevalence of associated co-morbidities. Overall, just over two thirds of children had an identified epilepsy syndrome or cause of their epilepsy.

The cases of 61 children (with 69 incidents) were subject to more detailed case note review. This included all the children who died, where case notes were available, and a sample of the children receiving intensive or high dependency care. The findings from this more detailed review are presented in the subsequent chapters.

## 4. The initial diagnosis and ongoing management of children with epilepsies

*The child was just over two years old when they were diagnosed with epilepsy by a consultant paediatrician following MRI, EEG and genetic involvement. The consultant responsible for making the diagnosis had been involved in the child's long term care. Initially there was a vague history for seizures which was reassessed as carers witnessed episodes. Seizure types were reviewed regularly and there were good descriptions documented. Her seizure types changed over time and she had several types as time progressed. Treatment for the epilepsy was initiated by a consultant paediatrician who, around a year after diagnosis, sought advice when seizures became problematic and difficult to control. The child was started on carbamazepine which gradually increased over a few weeks. There was clear evidence a multi-professional team were working together on the care package for the child, with a community nurse and lifetime nurse package of care also being evaluated and set up. (Case assessor comment)*

The 61 children whose notes were subjected to case note review were aged between 0 and 151 months (0-12 years) at their first seizure, with a median age of 11 months; 34 (56%) were aged less than one year at their first seizure. The median recorded time from first seizure to the diagnosis of epilepsy was two months, with a range from 0-36 months. Within three months of presentation of first seizure 40 (65%) children had received a diagnosis; 48 (79%) within six months and 55 (91%) within 12 months.

There were seven (12%) children in whom the time from first seizure to diagnosis of epilepsy was at least 12 months. In two of these seven children the delay in diagnosis was attributed to the complexity of the presentation together with the time taken the work through relevant investigations; or to the infrequency of seizures. There was just one case where a delay in diagnosis was attributed to a failure to identify episodic behaviour in a young infant with a degenerative neurological condition as possible epilepsy, and instead attributed to gastro-oesophageal reflux. There were no other children where the assessors had identified a failure to recognise seizures, or missed or inappropriate diagnoses.

### 4.1 Establishing the diagnosis

In 54 (89%) children, it was possible to determine the expertise of the clinician responsible for confirming the diagnosis, either from the case note review, or from the clinical questionnaire. Forty-two children (68%) had their diagnosis established by a paediatric neurologist or paediatrician with expertise in epilepsy. Often the initial diagnosis was a staged process, whereby the child might be seen first by a junior doctor, or a non-specialist, but then referred onto or discussed with more experienced professionals.

The diagnosis of epilepsy in children and young people should be established by a specialist paediatrician with training and expertise in epilepsy. (NICE 3)

The diagnosis of epilepsy should be made by a paediatric neurologist or paediatrician with expertise in childhood epilepsy. (SIGN)

In the majority of children, there was good evidence from the case reviews that a thorough approach had been taken to establishing the diagnosis. For 41 children (67%) there was evidence that seizure type had been considered in establishing the diagnosis; in 48 (79%) that epilepsy syndrome or aetiology had been considered; and in 44 (72%) that co-morbidities had been considered. However, 46 children (75%) had no identified epilepsy syndrome. This may reflect difficulties in assigning a syndrome, with many children considered to have idiopathic or symptomatic epilepsies.

Epileptic seizures and epilepsy syndromes in children, young people and adults should be classified using a multi-axial diagnostic scheme. The axes that should be considered are: description of seizure (ictal phenomenology); seizure type; syndrome and aetiology. (NICE 46)

The seizure type(s) and epilepsy syndrome, aetiology, and co-morbidity should be determined, because failure to classify the epilepsy syndrome correctly can lead to inappropriate treatment and persistence of seizures. (NICE 47)

The case reviews suggested that establishing the diagnosis is often an evolving process as further clinical information emerges, new events occur or results of investigations are received. This was reflected in the child's notes and letters, particularly with reference to the involvement of different specialists, or staged investigations.

Case assessors particularly highlighted good practice in relation to clear thought processes around establishing the diagnosis, the consideration of aetiology, seizure type and syndrome, flexibility, together with a willingness to review, update or revise the diagnosis and classification of the epilepsy (Table 4.1).

**Table 4.1: Examples of good practice around establishing the diagnosis**

Examples of good practice
When the child presented with apnoeic episodes, seizure presentation was one of the differential diagnoses. When a diagnosis of Tuberous Sclerosis was confirmed on MRI/ECHO, there was discussion regarding epilepsy with parents. When the child had tonic seizure, epilepsy was confirmed and treatment started. Seizures evolved over six months with the EEG changing towards a picture of West syndrome.
At initial diagnosis of epilepsy the underlying aetiology was felt to be a structural lesion, secondary to a previous ischaemic insult. This underlying aetiology was subsequently revised when a genetic diagnosis was made.

Case assessors considered practice to be unsatisfactory when appropriate investigations or expertise were not sought or recorded. It was also unsatisfactory when there appeared to have been some misinterpretation of results, or a failure to take account of all relevant information relating to the child.

These findings highlight the importance of taking a thorough and systematic approach to diagnosis, reviewing and documenting all results, and avoiding assumptions about diagnosis without considering all possibilities. They emphasise the need to review information as it emerges and to take a flexible and evolving approach to diagnosis and classification of the epilepsies.

The main difficulty for case assessors in establishing whether standards around diagnosis were met related to the lack of documentation. Similarly, although diagnoses were written in the notes, there was not always documentation of the thought processes and decision making surrounding the approach to establishing the diagnosis and treatment plans. Clear documentation of this information would help inform clinical staff treating the child.

## 4.2 Initiating treatment

While the majority of children had evidence of involvement of an appropriately qualified clinician in the establishment of the diagnosis, this did not always occur in relation to starting anti-epileptic treatment. In accordance with NICE guidance the assessors found evidence that in 19 (31%) children anti-epileptic drugs had been administered by a paediatric neurologist or paediatrician with expertise in epilepsy, and in a further 27 (44%) children by a paediatrician. However, it was not always clear whether they had expertise in epilepsy. Overall, case assessors judged that 18 children (30%) had anti-epileptic drugs started by someone other than a paediatric neurologist or paediatrician with expertise in epilepsy (two of these had first presented prior to the introduction of the NICE guidelines in 2004). This information was not always clear in the notes. However, in some cases, the treatment was initiated after discussion with a paediatric neurologist. For other children, treatment was started by a general paediatrician, but later modified or confirmed by a paediatric neurologist. While the NICE guidelines stipulate that anti-epileptic drug therapy should be initiated in an acute situation by a specialist, it would be appropriate for a non-specialist to start treatment. This would avoid the delay of waiting for a specialist.

AED therapy in children and young people should be initiated by a specialist. (NICE 60)

In 10 children case assessors judged that anti-epileptic drugs had not been appropriately initiated according to seizure type, epilepsy syndrome, co-medication and co-morbidities. In seven of these children, the assessors felt that the child had been started on a drug that may have been inappropriate for the seizure type or syndrome. For example, giving sodium valproate rather than carbamazepine to children presenting with focal seizures, or giving sodium valproate to infants presenting with infantile spasms. In two children, the drug was changed following discussion with a paediatric neurologist, or in light of emerging information, again potentially representing good practice, and by not delaying treatment, but subsequently seeking specialist advice. In one child, the drug used was felt to be appropriate, but the starting dose was judged to be too low.

The AED treatment strategy should be individualised according to the seizure type, epilepsy syndrome, co-medication and co-morbidity, the child, young person or adult's lifestyle, and the preferences of the person and their family and/or carers as appropriate. (NICE 49)



When possible, choose which AED to offer on the basis of the presenting epilepsy syndrome. If the epilepsy syndrome is not clear at presentation, base the decision on the presenting seizure type(s). (NICE 80)

The decision to commence anti-epileptic drug treatment should be reached jointly by the epilepsy specialist and the family. It should be informed by a knowledge and understanding of the epilepsy syndrome, including an assessment of recurrence risk and the likelihood of long term remission. (SIGN)

These findings highlight the importance of taking into account all information when considering the initiation of anti-epileptic treatment. As recommended by both NICE and SIGN guidelines, treatment should be started by a paediatric neurologist or paediatrician with training and expertise in epilepsy. However treatment should not be delayed if such input is not immediately available. Additionally treatment should follow the NICE or SIGN recommendations and guidelines in the BNFC. However, it is recognised that there may be cogent reasons for diverging from these guidelines. In such cases, the reasoning behind any decision making needs to be clearly documented in the child's notes and communicated to the parents or carers as well as other members of the clinical team.

### 4.3 Communicating with families

Children, young people and adults with epilepsy should be given information about their seizure type(s) and epilepsy syndrome, and the likely prognosis. (NICE 48)

For 32 children (53%) there was evidence that the family had been given information about the diagnosis and prognosis within six months of diagnosis. This was highlighted as good practice with the importance of appropriate, thorough and timely communication with children and young people and their families. This is emphasised throughout the NICE and SIGN guidelines on epilepsy, in numerous research reports, and by families themselves. For some children this was considered to be an evolving situation, with discussions regarding prognosis taking place more appropriately at a later date. Where there is an evolving understanding of the child's condition, it may not be possible straight away to give clear information on diagnosis or prognosis. For many children, discussions about their epilepsy care, including acute management, need to take place within the context of wider discussions about the overall care of the child, including any co-morbidities. The good practice examples showed that, even in such situations, a thorough approach can be achieved, while maintaining flexibility (Table 4.2).

**Table 4.2: Examples of good practice in communicating with families**

Examples of good practice
Consultant paediatrician noted on at least two occasions long discussions with mother regarding difficult epilepsy, spastic quadriplegia, chest infections and feeding issues.
Clinic letters evidence good discussion regarding diagnosis, investigations and lifestyle issues, appropriate to her age and stage of development.
Discussions recorded around acute management and seizures with rescue medication and the longer term anti-epileptic medication. Referred to neurology to discuss appropriate medications and underlying diagnosis.
Correspondence indicates information/discussions about the diagnosis and necessary investigations took place within clinic appointments and on the telephone with the family. In-depth discussion documented by geneticist

Case assessors highlighted a lack of evidence of any documented discussions with children and families around their diagnosis, support services and key information. In some children it is possible that this reflects a paucity of documentation about information provided to the family, rather than any actual failure to provide the information (Table 4.3).

**Table 4.3: Examples of unsatisfactory practice in communicating with families**

Examples of unsatisfactory practice
There is no evidence that parents were spoken to about the nature of the child’s difficulties and no mention of prognosis in the letter or notes. There was also no mention of safety issues and local support services or leaflets given although parents were clearly involved with the acute admissions and were kept informed at the time.
Documentation states the epilepsy specialist nurse would be in touch with the family but no record of what information was given. No record from clinic letter as to information given regarding prognosis.

There was only one case where the assessors judged that a failure to communicate adequately with the parents may have had a direct impact on the child’s subsequent wellbeing.

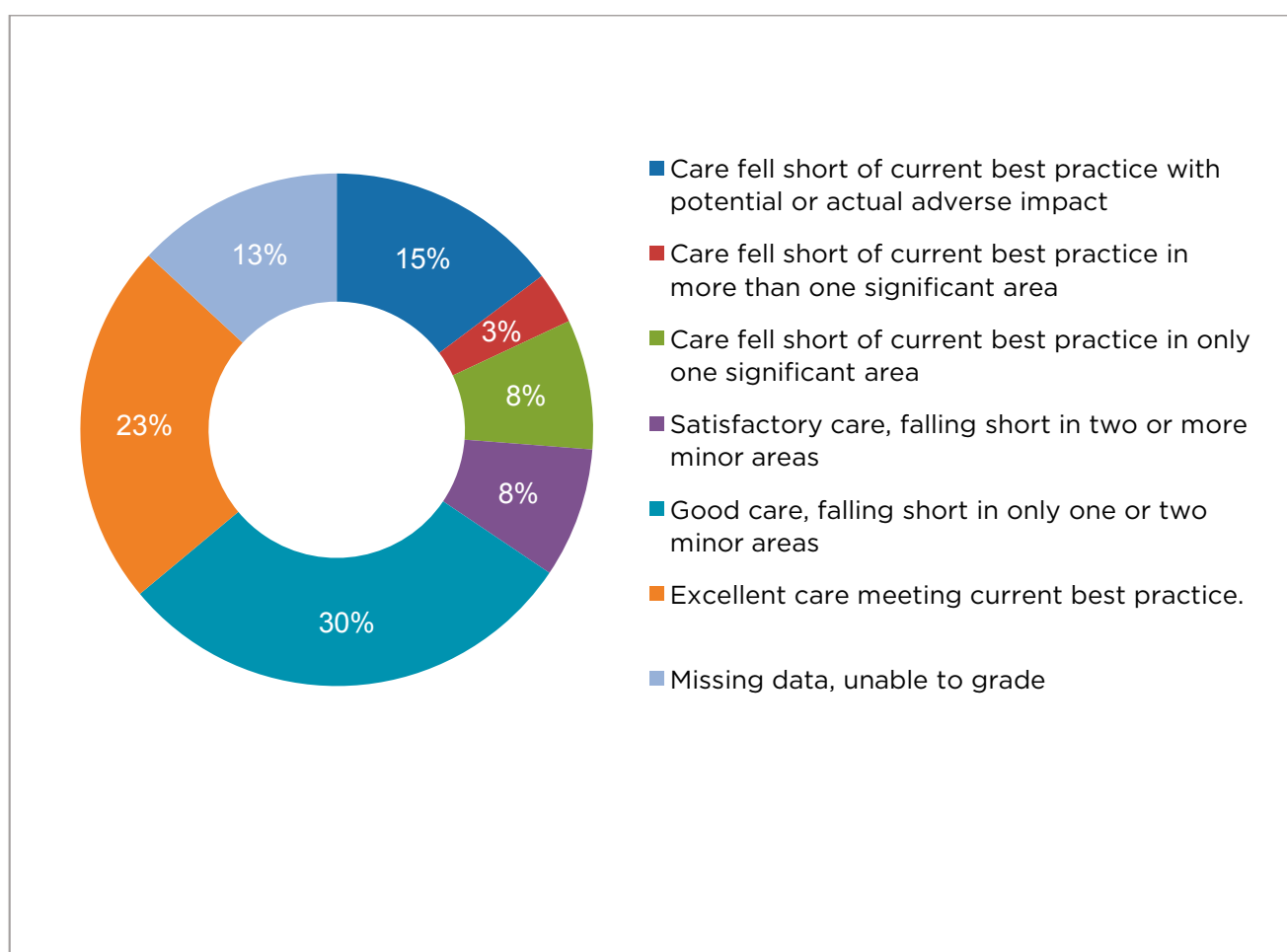
*Child had two seizures and a plan was made to start medication but the parents were not seen in clinic to discuss the medication and so they did not start it... There is evidence that the epilepsy specialist nurse was involved from the beginning of the diagnosis of epilepsy but no record as to the information given to parents around the time of diagnosis. The main failing is that the paediatrician did not discuss the introduction of anti-epileptic medication with parents and instead heard of the second fit and requested that the GP prescribe medication for the child. Because the parents did not start the medication, it suggests they did not fully understand the implications of the diagnosis, the child then went on to have further fits which may have been inevitable or may not. (Case assessor comment)*

This review has highlighted the importance of clear documentation of discussions with children and families, including the value of writing to the family following any consultation to clarify discussions that have taken place. Such documentation and correspondence are important not only for the families concerned, but also for all other professionals involved in caring for the child.

#### 4.4 Quality of care in relation to initial diagnosis and management

Case assessors judged the overall quality of care in relation to initial diagnosis and management to be excellent and meeting current best practice, or falling short of current best practice in only minor areas in 37 children (61%). Care was judged to have fallen short of current best practice in one or more significant areas in 16 cases (26%), in nine (15%) of which the care was deemed to have resulted in the potential for or actual adverse impact on the child. For eight (13%) children no judgement could be made, which was often due to case assessors feeling unable to judge because documentation was lacking (Figure 4.1).

**Figure 4.1: Quality of care in relation to initial diagnosis and management as judged by the case assessors**



Based on key questions from the criterion-based review, an assessment of the quality of care showed that there was just one case where the care fell short of the defined standards in all areas;

eight (13%) where care met the standards in all areas; and 38 (62%) where it fell short of defined standards in at least some areas. Table 4.4 shows the percentage of children for which there was evidence that the key standards were met in relation to quality of care. These percentages reflect the number of children meeting these standards where there was documented evidence in the child's notes. Therefore, the proportion of children for whom these standards were met may have been higher. However, a lack of documentation meant that for some children the assessors were unable to confirm whether the standard had been met.

**Table 4.4: Standards around initial diagnosis and management**

Standard	Number (%) with evidence that this standard had been met
Was the diagnosis established by a paediatric neurologist or specialist paediatrician with training and expertise in epilepsy?	42/61 (69%)
Was there evidence that seizure type was considered in establishing the diagnosis?	41/61 (67%)
Was there evidence that an epilepsy syndrome was considered in establishing the diagnosis?	25/61 (41%)
Was there evidence that aetiology was considered in establishing the diagnosis?	48/61 (79%)
Was there evidence that co-morbidities were considered in establishing the diagnosis?	44/61 (72%)
Was anti-epileptic drug treatment initiated by a paediatric neurologist or specialist paediatrician with training and expertise in epilepsy?	19/61 (31%)
Were appropriate anti-epileptic drugs administered according to the seizure type, epilepsy syndrome, co-medication and co-morbidity?	42/61 (69%)
Was there evidence that the child and family were given information about their diagnosis and prognosis within six months of the diagnosis?	32/61 (53%)

## 4.5 Ongoing management

*This child was appropriately managed between the community paediatrician and tertiary services. Neurology was involved at an early stage given developmental issues. Referred back when epilepsy started and re-referred when management was difficult. Developed good joint working with six monthly clinic - paediatrician/neurology/epilepsy nurse in attendance. Medication reviewed regularly with updated emergency care plan. Liaison well documented between acute and community services. (Case assessor comment)*

Overall the children had a wide range of different health professionals recorded as being directly involved in their ongoing care (Table 4.5), with 28 (46%) children having at least three different health professionals. A large number of the children had evidence of multidisciplinary involvement, with health visitors, school nurses, different allied health professionals and members of child development teams. Tertiary specialists were involved in the ongoing care of 28 (46%) children, including geneticists, surgeons, orthopaedic surgeons, neurosurgeons, and ophthalmologists. In the children who died 13 (39%) had documented involvement of hospice, palliative care or community children's nursing teams. Only five (8%) children had any recorded involvement of Child and Adolescent Mental Health Service (CAMHS), although 15 (25%) were recorded as having psychological and/or psychiatric co-morbidity.

**Table 4.5: Health professionals recorded as being involved in the child's ongoing care**

Professionals involved	Number (%)
Paediatrician	51 (84%)
Neurologist	36 (59%)
Epilepsy specialist nurse	29 (48%)
GP	32 (53%)
Other professionals	45 (74%)

For 43 (71%) children there was evidence that the child and family had access to a named individual to contact for advice and support. For 52 (85%) a named clinician was responsible for the ongoing management of the epilepsy. Overall, fewer than half the children in this review had an epilepsy specialist nurse involved in their care. This is despite 67% (37/55) of the units where the child was treated for the sentinel incident reporting that they had epilepsy nurse specialists. For some children, this may have been because their epilepsy was well-controlled and not considered to be a central part of their ongoing care. Nevertheless, this does suggest that there are gaps in epilepsy specialist nurse provision, in spite of this being highlighted by NICE as a component of good care for children with epilepsies.

The child, young person or adult with epilepsy and their family and/or carers as appropriate should know how to contact a named individual when information is needed. This named individual should be a member of the healthcare team and be responsible for ensuring that the information needs of the child, young person or adult and/or their family and/or carers are met. (NICE 187)

Epilepsy specialist nurses (ESNs) should be an integral part of the network of care of children, young people and adults with epilepsy. The key roles of the ESNs are to support both epilepsy specialists and generalists, to ensure access to community and multi-agency services and to provide information, training and support to the child, young person or adult, families, carers and, in the case of children, others involved in the child’s education, welfare and well-being. Each epilepsy team should include paediatric epilepsy nurse specialists. (NICE 270)

The NICE guidelines indicate that tertiary services should be involved if a child’s seizures are not controlled and/or there is diagnostic uncertainty or treatment failure. They list eight criteria which should prompt consideration of a referral to tertiary services (Table 4.6). All but two of the children had at least one of these criteria, with 39 (66%) having at least three of the criteria recorded. Of those with one or more of these criteria recorded, 49 (83%) had evidence of referral to a tertiary specialist. This highlights again the complex nature of these children’s epilepsies. It was encouraging to note that the majority had been referred on to tertiary specialists.

**Table 4.6: Criteria indicating need for referral to tertiary services**

Criterion	Number (%) with this criterion
The epilepsy is not controlled with medication within two years	36 (59%)
Management is unsuccessful after two drugs	38 (62%)
Aged under two years at diagnosis	38 (62%)
Unacceptable side effects from medication	16 (26%)
Unilateral structural lesion	4 (7%)
Psychological and/or psychiatric co-morbidity	15 (25%)
Diagnostic doubt as to the nature of the seizures and/or seizure syndrome.	16 (26%)
Behavioural or developmental regression	24 (39%)

If seizures are not controlled and/or there is diagnostic uncertainty or treatment failure, children, young people and adults should be referred to tertiary services for further assessment. Referral should be considered when one or more of the following criteria are present:

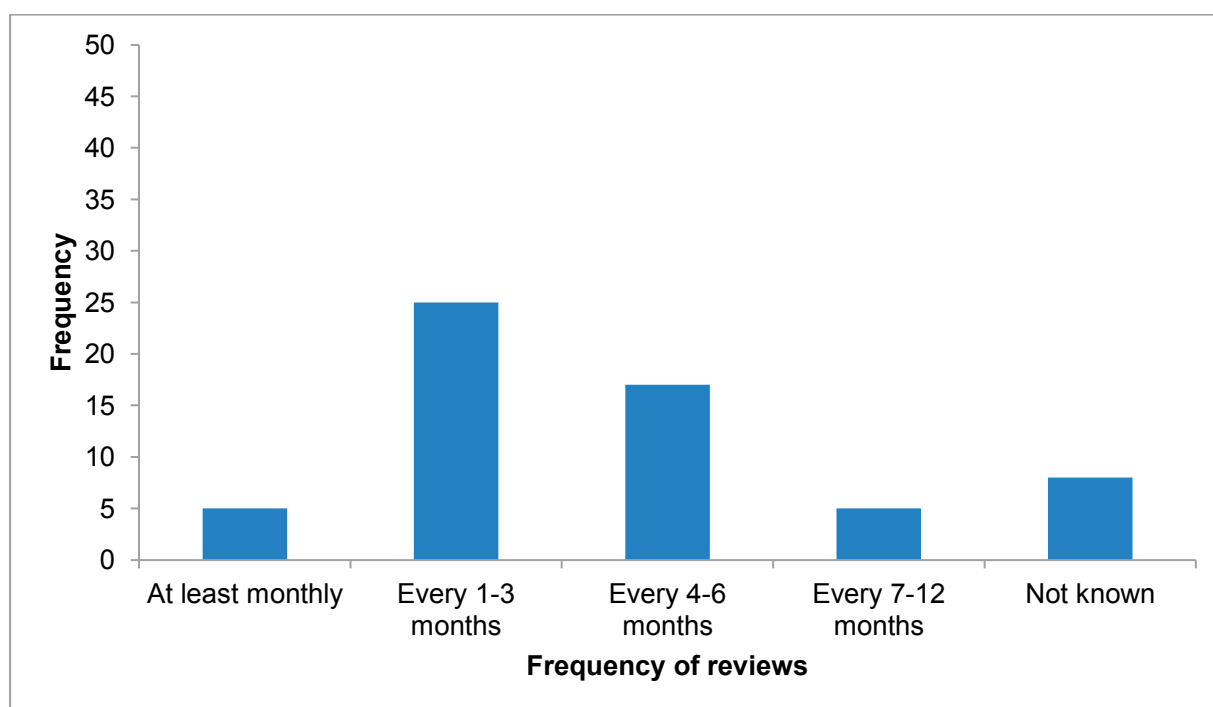
- the epilepsy is not controlled with medication within 2 years
- management is unsuccessful after two drugs
- the child is aged under 2 years
- a child, young person or adult experiences, or is at risk of, unacceptable side effects from medication
- there is a unilateral structural lesion
- there is psychological and/or psychiatric co-morbidity
- there is diagnostic doubt as to the nature of the seizures and/or seizure syndrome. (NICE 169)

Referral to tertiary specialist care should be considered if a child fails to respond to two AEDs appropriate to the epilepsy in adequate dosages over a period of six months. (SIGN)

Behavioural or developmental regression or inability to identify the epilepsy syndrome in a child, young person or adult should result in immediate referral to tertiary services. (NICE 171)

In this review, 52 (85%) of the children had their epilepsy reviewed at least annually, and typically more frequently, reflecting the age of the child and complexity of the care required (Figure 4.2).

**Figure 4.2: Frequency of epilepsy review**



For children and young people, the maximum interval between reviews should be 1 year, but the frequency of reviews should be determined by the child or young person's epilepsy and their wishes and those of the family and/or carers. The interval between reviews should be agreed between the child or young person, their family and/or carers as appropriate, and the specialist, but is likely to be between 3 and 12 months. (NICE 276)

Annual review is suggested as a minimum, even for children with well controlled epilepsy, to identify potential problems, ensure discussion on issues such as withdrawal of treatment, and minimise the possibility of becoming lost to follow up. (SIGN)

One issue emphasised by the case assessors was the potential for, or actual, fragmentation of care related to the complexity of these children's epilepsies and other co-morbidities. These were usually children in who a large number of professionals were involved both in hospital and in the community. Some had frequent hospital admissions which could interfere with routine reviews. In addition the focus of care shifted to the management of the acute episodes during these hospital admissions, without any professional taking a clear lead in the child's overall care.

*There was suboptimal care. The child was repeatedly attending A&E and nobody seemed to be taking an overview with a clear plan for medications, non-drug options, family support and discussion, liaising with school. An epilepsy nurse support would have helped address much of this. The child probably needed to be reviewed more often in consultant clinic as the child's epilepsy was unstable. It is not clear if the child was seeing a consultant with an interest in epilepsy. They needed to see a tertiary paediatric neurologist and possibly discuss surgical options. (Case assessor comment)*

#### 4.5.1 Management of anti-epileptic drugs

*Drug doses were not fully optimized and small/tentative increases in dose were made when they were increased. The child was on an appropriate choice of drugs; carbamazepine and levetiracetam. Carbamazepine was started in infancy and was not effective in controlling seizures at maximum dose (30mg/kg/day) so could potentially have been weaned rather than continued at moderate dose. There was mention of rescue buccal midazolam in clinic letters but no explicit emergency care plan found in the notes, despite several admissions for prolonged seizures. (Case assessor comment)*

For those children where data were available, 23 (41%) were on monotherapy at the time of the incident, 14 (25%) on two anti-epileptic drugs and 17 (30%) on three or more. Two children (4%) were not receiving any anti-epileptic drugs. There were seven children (12%) for whom the case assessors judged that, at their most recent review, the anti-epileptic drugs prescribed were not appropriate for the child, or were administered at an inappropriate dose, as highlighted in Table 4.7. In most cases this was related to the child having been prescribed anti-epileptic drugs either below or above the recommended doses. This may reflect difficulties in managing seizures leading to increasing doses being used; or, prescribing clinicians exercising a degree of caution leading to lower doses than recommended. In one case, a young person had stopped her anti-epileptic drugs due to perceived side effects. Poor compliance may be an issue in young people with epilepsies, emphasising the importance of providing accessible support and follow up to this group.

The AED treatment strategy should be individualised according to the seizure type, epilepsy syndrome, co-medication and co-morbidity, the child, young person or adult's lifestyle, and the preferences of the person and their family and/or carers as appropriate. (NICE 49)

It is recommended that children, young people and adults should be treated with a single AED (monotherapy) wherever possible. If the initial treatment is unsuccessful, then monotherapy using another drug can be tried. Caution is needed during the changeover period. (NICE 51)

The decision to commence anti-epileptic drug treatment should be reached jointly by the epilepsy specialist and the family. It should be informed by a knowledge and understanding of the epilepsy syndrome, including an assessment of recurrence risk and the likelihood of long term remission. (SIGN)



**Table 4.7: Examples of children where drug administration was deemed to be inappropriate at most recent review**

Reason	Case examples
<b>Sub-optimal dose of anti-epileptic drugs</b>	The child had suffered a prolonged seizure and two further seizures prior to the last scheduled review and, at the time, was on a sub-therapeutic dose of sodium valproate. Dose of sodium valproate was increased to the minimum recommended dose following the last scheduled review. This was not adjusted after the attendance at the Emergency Department following a second prolonged seizure.
<b>Inappropriate anti-epileptic drugs used</b>	The child was experiencing daily seizures (including GTC), however appropriate AEDs were not administered; child wasn't on rufinamide despite intractable seizures, and dose of clonazepam is lower than recommended, the dose of phenobarbitone is higher than recommended, the plan was to withdraw lamotrigine but there is no evidence this was initiated.
<b>Lack of documentation of reasons for divergence from guidelines</b>	There were frequent changes to medication in his first few years, with some anti-epileptics tried at moderate dose but not necessarily maximum dose before being withdrawn. The rationale behind some decisions was sometimes unclear, for example ACTH was used in infancy although there was no clear description of infantile spasms or investigation for that.
<b>Lack of a named clinician coordinating care</b>	In the six months prior to this incident sodium valproate was discontinued. The child had been on a dose at the lower end of the therapeutic range. The decision to wean AEDs was taken by a junior doctor following parental request as the child had been seizure-free for more than 2years. It was unclear who was in overall charge of the decisions regarding AEDs as the child was under two consultant paediatricians and two consultant paediatric neurologists.

In just one child, there was concern that underdosing may have been a contributory factor in the incident. The child was deemed to be on a sub-therapeutic dose of anti-epileptic drugs and had attended hospital with prolonged seizures twice in the 12 months prior to the incident. On neither occasion had the dose been increased, although it had been increased at the last scheduled review, six weeks before the incident. Shortly after the last attendance at hospital, this child was found dead in bed. The death was classified as a sudden unexpected death in epilepsy (SUDEP).

In the six months prior to the incident, 11 (18%) children had anti-epileptic drugs withdrawn. For seven (64%) of these, the case assessors judged that this withdrawal had been carried out as recommended, slowly, over at least two to three months, with a clear plan and under the guidance of a specialist.

When AED treatment is being discontinued in a child, young person or adult who has been seizure free, it should be carried out slowly (at least 2–3 months) and one drug should be withdrawn at a time. (NICE 76)

There should be a failsafe plan agreed with children, young people and adults and their families and/or carers as appropriate, whereby if seizures recur, the last dose reduction is reversed and medical advice is sought. (NICE 78)

Withdrawal of AEDs must be managed by, or be under the guidance of, the specialist. (NICE 79)

#### 4.5.2 Communication with families

All children, young people and adults with epilepsy should have a comprehensive care plan that is agreed between the person, family and/or carers where appropriate, and primary care and secondary care providers. This should include lifestyle issues as well as medical issues. (NICE 272)

Each child should have an individual management plan agreed with the family and primary care team. (SIGN)

An individual treatment pathway should be formulated for children, young people and adults who have recurrent convulsive status epilepticus. (NICE 153)

A healthcare plan was documented in the case notes for 39 (64%) children. This included 30 (77%) children with an emergency care plan for the management of seizures; 10 (26%) children with a school healthcare plan; and nine (23%) children with an end of life care plan.

There was evidence of discussion of different aspects of medical care in the majority of the children's notes (Table 4.8). However, there were some notable gaps in recording of discussions, particularly in relation to the side effects of medication, information about support, the child's academic progress, and risks and hazards including SUDEP. The absence of any documented evidence of such discussions could reflect a failure to consider these issues in the clinical reviews, or that any such discussions were not recorded in the clinical notes or clinic letters.

**Table 4.8: Issues discussed in most recent or previous clinical reviews**

Issues discussed	Number (%) with evidence of discussion in the notes
Discussions around treatment plans and the management of seizures	50 (82%)
Discussions around medication, including concordance and adherence, effectiveness, and side effects	53 (87%)
Discussions around support and academic progress	37 (61%)
Discussions around risks and hazards, including SUDEP	35 (57%)

Treatment should be reviewed at regular intervals to ensure that children, young people and adults with epilepsy are not maintained for long periods on treatment that is ineffective or poorly tolerated and that concordance with prescribed medication is maintained. (NICE 71)

Annual review should include an enquiry about side effects and a discussion of the treatment plan to ensure concordance and adherence to medication. (NICE 70)

At the review, children, young people and adults should have access to: written and visual information; counselling services; information about voluntary organisations; epilepsy specialist nurses; timely and appropriate investigations; referral to tertiary services including surgery, where appropriate. (NICE 277)

All children with epilepsy should have their behavioural and academic progress reviewed on a regular basis by the epilepsy team. Children with academic or behavioural difficulties should have appropriate educational and/or psychological assessment and intervention. (SIGN)

Information on SUDEP should be included in literature on epilepsy to show why preventing seizures is important. Tailored information on the person's relative risk of SUDEP should be part of the counselling checklist for children, young people and adults with epilepsy and their families and/or carers. (NICE 193)

Families should be advised if the child has an increased risk of SUDEP. They can be reassured if the risk is considered to be low. (SIGN)

All children, young people and adults with epilepsy and learning disabilities should have a risk assessment including:

- bathing and showering
- preparing food
- using electrical equipment
- managing prolonged or serial seizures
- the impact of epilepsy in social settings
- SUDEP
- the suitability of independent living, where the rights of the child, young person or adult are balanced with the role of the carer. (NICE 253)

There was very limited evidence that young people themselves had been involved in discussions around their own medication and lifestyle issues, only two cases having such discussions documented. In many cases, the case assessors felt this discussion was not appropriate due to the age of the child, the severity of any associated learning difficulties and other factors. There were eight young people for whom the case assessors felt it would have been appropriate to have involved them in such discussions, but could find no evidence that this had happened. This may have been either because the young people were not involved in such discussions, or that their involvement had not been documented. It is concerning that, even in this group of children and young people with recognised complex needs, there were only two cases where case assessors found direct evidence that the young person had been involved in discussions around their care.

Healthcare professionals should adopt a consulting style that allows the young person with epilepsy to participate as a partner in the consultation. (NICE 255)

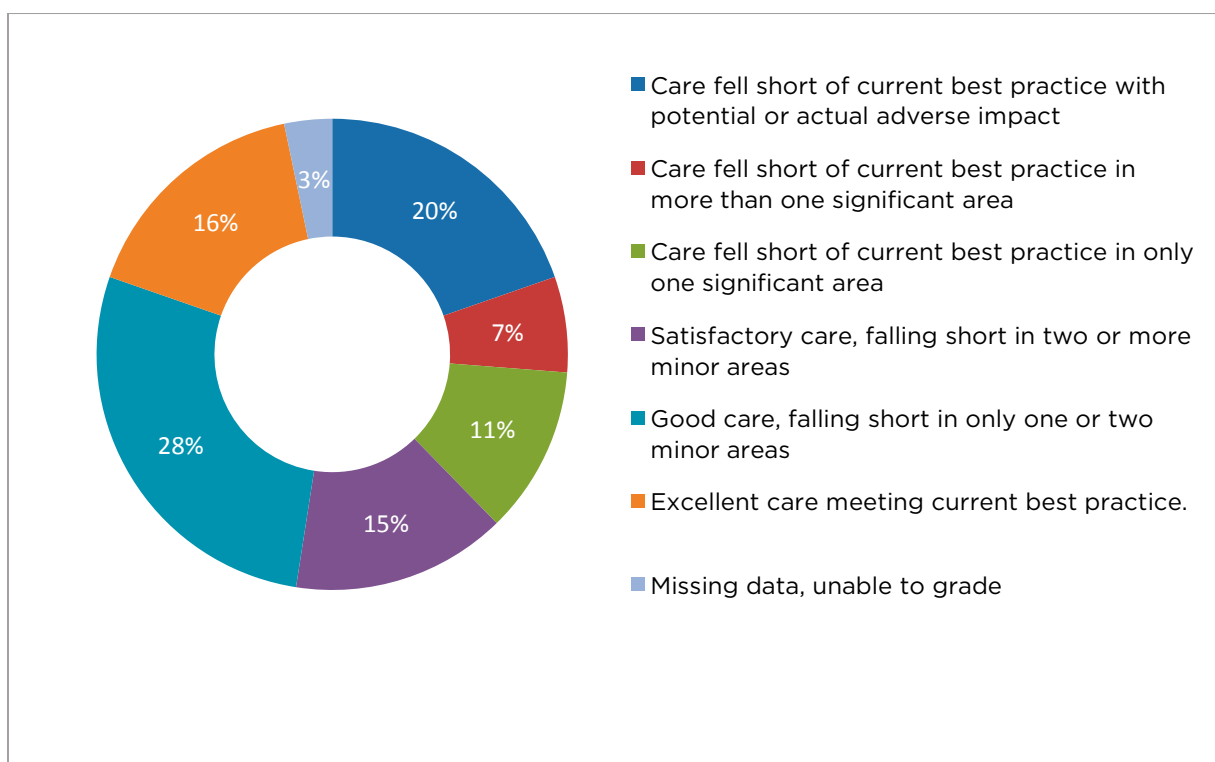
Decisions about medication and lifestyle issues should draw on both the expertise of the healthcare professional and the experiences, beliefs and wishes of the young person with epilepsy as well as their family and/or carers. (NICE 256)

The information given to young people should cover epilepsy in general and its diagnosis and treatment, the impact of seizures and adequate seizure control, treatment options including side effects and risks, and the risks of injury. Other important issues to be covered are the possible consequences of epilepsy on lifestyle and future career opportunities and decisions, driving and insurance issues, social security and welfare benefit issues, sudden death and the importance of adherence to medication regimes. Information on lifestyle issues should cover recreational drugs, sexual activity and sleep deprivation. (NICE 260)

### 4.5.3 Quality of care in relation to ongoing management

Case assessors judged the overall quality of care in relation to ongoing management to have fallen short of current best practice in one or more significant areas for 23 children (38%), in 12 (20%) of whom this was deemed to have resulted in the potential for, or actual, adverse impact on the child. They judged the care to be excellent and meeting current best practice, or falling short of current best practice in only minor areas, in 36 children (59%; Figure 4.3). For two children no judgement could be made due to lack of documentation.

**Figure 4.3: Quality of care in relation to ongoing management as judged by the case assessors**



An assessment of the quality of care based on key questions from the criterion-based review showed that there were no cases where the care fell short of the defined standards in all areas; four (7%) where care met the standards in all areas; and 53 (87%) where it fell short of defined standards in at least some areas. Table 4.9 shows the percentage of children for which there was evidence that the key standards in relation to quality of care were met.

The highest levels of standards being met were around issues in relation to medical management, including having a named clinician, appropriate referrals to tertiary specialists, regular reviews and both the prescribing and review of anti-epileptic medication. In all of these areas, there was documented evidence in the notes that these standards had been met in at least 44 (72%) children. There was, however, a lack of clarity over the process of withdrawal of anti-epileptic drugs where this was the case. The evidence of standards being met is less clear in relation to access to epilepsy specialist nurses and the provision of care plans or treatment pathways. It is also unclear as to wider discussions with the child and family regarding support, academic progress, risks, hazards and involvement of the child as an active player in the management of his or her epilepsy.

**Table 4.9: Standards around ongoing management**

	<b>Standard</b>	<b>Number (%) with evidence that this standard had been met</b>
1	The child and family had access to a named individual to contact for advice and support	43/61 (71%)
2	The child and family have access to an epilepsy specialist nurse	31/61 (51%)
3	A named clinician was responsible for the ongoing management of the child/young person's epilepsy	52/61 (85%)
4	Where appropriate, the child/young person had been referred to a tertiary specialist	49/59 (83%)
5	The child's epilepsy was reviewed at least annually (for uncomplicated epilepsy) or more frequently (for epilepsy that was difficult to control, or where other factors such as co-morbidities, compliance indicated)	54/61 (89%)
6	Appropriate anti-epileptic drugs were administered according to the seizure type, epilepsy syndrome, co-medication and co-morbidity	48/61 (79%)
7	For each anti-epileptic drugs, the child/young person was on an appropriate dose for his/her age and weight	45/61 (74%)
8	Where a child had an anti-epileptic drugs withdrawn, the treatment was carried out slowly over at least two-three months with a clear plan, and under the guidance of a specialist	7/11 (64%)
9	The child/young person had an appropriate individual care plan/treatment pathway	25/61 (41%)
10	There had been discussions around treatment plans and the management of seizures	50/61 (82%)
11	There had been discussions around medication, including concordance and adherence, effectiveness, and side effects	53/61 (87%)
12	There had been discussions around support and academic progress	37/61 (61%)
13	There had been discussions around risks and hazards, including SUDEP	35/61 (57%)
14	Where appropriate, the young person been involved in discussions about medication and lifestyle issues	2/10 (20%)

## 4.6 Summary

This chapter examined the quality of care in all 61 children whose notes were subject to a detailed case review and focused on the initial diagnosis and ongoing management, particularly in the 12 months prior to the reported incident (death or prolonged seizure requiring intensive or high dependency care). The case assessors made judgements on the quality of care on the basis of their overall impressions of the care provided in these two phases (implicit and holistic review), and by evidence in the notes of compliance with key standards from the NICE and SIGN guidelines.<sup>27,29</sup> In looking at the quality of care around initial diagnosis and treatment, it is important to recognise

that some children would have first presented with epilepsy prior to the implementation of these guidelines. In total, 10 children (16%) presented with their first seizure prior to 2004.

Overall, the evidence demonstrated reasonable standards of care, although with some notable gaps. Case assessors judged the quality of care in relation to initial diagnosis and treatment to be good (excellent and meeting current best practice, or falling short of current best practice in only minor areas) in 37 children (61%). Evidence of compliance with specific standards varied from 31% to 79%. For the majority of children there was evidence of both the involvement of a clinician with appropriate expertise in the diagnostic process, and that seizure type, aetiology and co-morbidities had been considered. There was also evidence of appropriate administration of anti-epileptic drugs. However, it was less clear whether the appropriate specialists had been involved in initiating anti-epileptic medication, and there were potential gaps in the timely provision of information to children and families about their diagnosis and prognosis.

Case assessors judged the quality of care in ongoing management to be good (excellent and meeting current best practice, or falling short of current best practice in only minor areas) in 36 children (59%). Evidence of compliance with specific standards varied from 20% to 89%. There was documentation of appropriate follow up relating to these children including access to a named clinician, and involvement of tertiary specialists. There was also evidence of the appropriate administration of anti-epileptic drug treatment. However, there were two specific areas where there was inferior evidence of compliance with standards: the provision of an individual treatment plan, and the involvement of young people in discussions about their medication and lifestyle.

These findings emphasise the importance of rigour in the diagnostic process, taking into account the often evolving nature of diagnosis in the epilepsies. There is a particular need for careful consideration of epilepsy syndromes, and this is likely to be aided by the early involvement of a paediatric neurologist. Classification of the epilepsies is an evolving field, with changing terminology, and ongoing developments in genetic understanding, thus classification by epilepsy syndrome may continue to prove challenging. The Epilepsy12 audit<sup>30</sup> has highlighted the importance of an appropriate multi-axial epilepsy classification, and this review has provided evidence that this could be improved. Where the diagnosis is evolving, the steps taken to reach a classification should be clearly documented and updated as appropriate when more information comes to light.

While there was limited evidence of the involvement of a paediatric neurologist or specialist paediatrician in the initiation of anti-epileptic drugs, the case assessors judged that, in the majority of cases, treatment had been appropriate. It is possible that, when a child presents acutely with seizures, the treating clinician is appropriately administering anti-epileptic treatment according to local protocols. Such an approach avoids delays in treatment. However, this must be carefully documented, and the treatment reviewed by a specialist as soon as possible. A small number of cases were found where inappropriate anti-epileptic drugs had been used, or where appropriate drugs had been given in inadequate doses. Such errors could be minimised by following national and local guidelines and by early discussion with, and review by, a neurologist or paediatrician with epilepsy expertise.

There was evidence of some very good care during initial diagnosis and ongoing management of these children's epilepsies. However, the complexity of their needs in relation to both difficult to control epilepsy and associated developmental problems, gave rise to the potential for disjointed care and inadvertently

additional stress for the children and families concerned. The findings here endorse the recommendations set out by NICE and SIGN, as well as the importance of a clinician taking ownership of the coordination of the child's care, as part of a larger multidisciplinary clinical team. Greater coordination could be achieved through the input of epilepsy specialist nurses. The Epilepsy12 Audit has recommended that all children diagnosed with epilepsy should have specialist nurse input in accordance with the NICE and SIGN guidelines. This review showed that only half the children included had this access.

Where children are presenting with difficult to control seizures, it is essential to involve tertiary specialists, to ensure that the children are reviewed regularly, and that there is someone who takes responsibility for the overview of their care. This is particularly important where the children may be having frequent hospital admissions and the focus can shift to management of the acute episodes rather than longer term planning. It is vital that any changes to medication and dosages are carefully documented, along with the decision making behind that, particularly if deviating from standard protocols for drug management. Many of the case notes reviewed did not have clearly documented individual treatment plans. This potentially can lead to difficulties in fragmentation of care, and uncertainty about appropriate management of acute events, a topic that will be covered in more detail in the chapter 5. The use of an 'Epilepsy Passport' has been highlighted as a possible approach to facilitating care.<sup>60</sup> This passport would hold information about each child or young person's epilepsy, such as up-to-date care and treatment plans as well as other important information. Such a document, if carried by the parents or the young person, would provide the opportunity for greater participation with children and families, and would improve coordination of care and clarity around emergency management.

Early, thorough and ongoing discussions with children and young people and their parents or carers is crucial. While such discussions may have been present more often than is indicated from the assessment of standards, it is essential that discussions are clearly documented in the clinical notes and backed up with written information for the family. Clear communication can be facilitated by writing directly to parents and young people following any clinic appointments or hospital attendances. Clinic letters need to include clear information about the underlying diagnosis, any co-morbidities and current seizure control. These should also contain details of current medication and any changes to medication, side-effects of medication, current plans for emergency management, together with documentation of any discussions around wider issues of risks and hazards, support, participation and lifestyle issues.



## 4.6.1 Key findings and recommendations

**Key finding 1:** In spite of the severity of these children's health needs, this review has emphasised that the care provided by parents and professionals working together provides the best possible quality of care.

**Key finding 2:** The findings emphasise the importance of rigour in the diagnostic process, while taking into account the often evolving nature of the diagnosis in epilepsy. There is a particular need for careful consideration of epilepsy syndromes, and this is likely to be aided by the early involvement of a paediatric neurologist. This review's findings support the recommendations set out by the NICE and SIGN guidelines that the diagnosis of epilepsy should be made by a paediatric neurologist or paediatrician with expertise in childhood epilepsy.

**Key finding 3:** The review has highlighted the importance of taking into account all information around the diagnosis and classification of epilepsy before beginning anti-epileptic drug treatment. Although treatment should follow NICE or SIGN guidelines and the BNFC, there may be reasons and thought processes for diverging from these. This review has highlighted the importance of clear documentation of such decisions. Consistency in following guidelines and clarity around the reasons for any divergence could be improved by processes of peer review in clinical teams.

### **Recommendation 1**

Clinicians looking after children and young people with epilepsies should follow NICE and SIGN guidelines for all aspects of care, and document the reasons for any deviations from these standard treatment guidelines.

### **Recommendation 2**

Clinical teams looking after children and young people with epilepsies should consider establishing a process of peer review as a means of monitoring and improving practice.

**Key finding 4:** The complexity of the children's epilepsies and wider health needs, presented in this review, means that there are often multiple professionals working with the child and family. The review has highlighted the importance of clear communication between professionals, and the need for one professional to clearly take a lead in the overall coordination of care. Where an epilepsy nurse specialist is involved, this must be documented in the clinical notes, together with any decision making, communication with parents or changes to management made by them. Such information needs to be communicated to all members of the clinical team. This supports and reinforces the recommendations made by the NICE and SIGN guidelines that a named clinician should have overall responsibility for coordination of care.

**Key finding 5:** Early, thorough and ongoing discussions with children and young people and their parents or carers are crucially important. This is clearly emphasised in both NICE and SIGN guidelines. However, although such discussions may be taking place, there was a paucity of documentation around these.

### **Recommendation 3**

Clinical teams looking after children and young people with epilepsies should consider introducing an 'epilepsy passport' for all children as a means of improving communication and clarity around ongoing management.

## 5.0 The care of children and young people with prolonged seizures

This chapter covers the care of 36 children who had presented with prolonged seizures, drawing on the findings from the review of case notes. Eight of these children died during or following their admission. The care received by other children who died during this review will be reported separately in Chapter 6. The analysis in this chapter is based on all 36 presenting with prolonged seizure, regardless of the outcome.

Twenty eight of the children (78%) had known developmental impairments. In the 12 months prior to the notified incident, 16 children (44%) had been admitted to hospital with prolonged seizures; 17 (47%) had at some stage previously received intensive or high dependency care for prolonged seizures. However, only four children were reported to be having frequent tonic-clonic seizures (daily or weekly), and 17 were reported to be having tonic-clonic seizures less than once a month, or not at all. Two children were not taking any anti-epileptic drugs; 17 received monotherapy; and 14 children took two or more anti-epileptic drugs.

### 5.1 Pre-hospital care

*The child had been in hospital with seizures and following recovery had been discharged home without a plan. On the same evening whilst at home and under the care of his parents, the child was discovered in status epilepticus. It was unclear as to whether the family had been given any training in emergency treatment and there was no documented emergency plan. The ambulance crew treated the child with IV diazepam at half the recommended dose, and transferred the child to hospital. (Case assessor comment)*

The majority of children (28; 78%) were at their home of residence at the time of the prolonged seizure, and four (11%) were already in hospital (Table 5.1). The analysis of pre-hospital and emergency department care is limited to those children presenting with prolonged seizures in the community (n=32).

**Table 5.1: Location of the child at the time of the incident**

Location at the time of the incident	Number (%)
Home of normal residence	28 (78%)
Acute Hospital	4 (11%)
School	3 (18%)
Hospice	0 (0%)
Other (including mental health inpatient unit, respite care, residential care)	1 (3%)
Not known	0 (0%)
Total	36 (100%)

Treatment should be administered by trained clinical personnel or, if specified by an individually agreed protocol drawn up with specialist, by family members or carers with appropriate training. (NICE 149)

For 17 children (53%) the case assessors judged that an appropriately trained person, able to administer first aid or emergency treatment was present at the time of the incident. In the majority of the remaining children it wasn't clear from the records whether those present with the child had been trained or not. In three cases the assessors judged that no-one was present with the child who had been trained to administer first aid or emergency treatment; all three were at home in the care of their parents. In two of these children, it was unclear whether there was an emergency care plan, and for the third child the emergency care plan did not involve the parents giving emergency medication as the child had always previously had short seizures. These cases highlight the need for adequate training for parents in the administration of rescue medication, clear documentation and regular review of care plans in light of children's evolving epilepsies.

### 5.1.1 Emergency care plans

*This child had been admitted shortly before the incident, and discharged with evidence of not being 100% better. One would have expected clear advice to be given to mum on what signs/symptoms to look for which would be her guide in contacting the epilepsy nurse or ward/A&E/ambulance. But this information was lacking in evidence. Also there was mention of a care plan but we did not find a written copy of what mother was asked to do or of the care plan. (Case assessor comment)*

There was evidence in 15 (47%) children's notes of an emergency care plan for the management of prolonged seizures, however in only seven was there was evidence that this plan had been followed. In all but three of the remaining cases it was not clear whether an emergency care plan was present or had been followed. For the three children, the case assessors determined that there was a plan but it had not been followed, raising concerns relating to failure or delay in administering adequate rescue medication. One child was not given rescue medication by the family as the buccal midazolam they had was out of date. In this case and one other, the children's parents had received training and an emergency care plan for rescue medications but failed to administer the medication to the child seizing. The remaining child had an emergency care plan with the mother trained in rescue medication administration, but only half the dose was given. These three cases highlight the need to clarify and make care plans more explicit, while ensuring parents are fully trained and feel comfortable administering rescue medication. Overall, for 13 (41%) children there was evidence in the notes that some rescue medication had been given prior to the arrival of an ambulance; nine of these involved buccal or nasal midazolam, three rectal diazepam, and one rectal paraldehyde.

There was one child where the case assessors identified that on a previous admission the parents had administered an inappropriate dose of buccal midazolam. This error had occurred as a result of the GP prescribing Buccolam (midazolam 5mg in 1ml) instead of the unlicensed product Epistatus (midazolam 10mg in 1ml), which was usually prescribed. The parents had given the same volume as previously prescribed, not realising that this resulted in a lower dose. This was felt to be a significant error, which had contributed to the child's need for admission, possible delays in terminating the seizure, and the potential for more serious adverse effects through excessive administration of

benzodiazepines. The error had been investigated by the hospital responsible as a serious untoward incident. The potential for further errors to occur was highlighted by the case assessors as an important learning point, and supports the need for clinicians to ensure parents are fully trained and feel comfortable administering medication.

An individual treatment pathway should be formulated for children, young people and adults who have recurrent convulsive status epilepticus. (NICE 153)

Give immediate emergency care and treatment to children, young people and adults who have prolonged (lasting 5 minutes or more) or repeated (three or more in an hour) convulsive seizures in the community. (NICE 154)

## 5.1.2 Ambulance management

Depending on response to treatment, the person's situation and any personalised care plan, call an ambulance, particularly if:

- the seizure is continuing 5 minutes after the emergency medication has been administered
- the person has a history of frequent episodes of serial or has convulsive status epilepticus, or this is the first episode requiring emergency treatment or
- there are concerns or difficulties monitoring the person's airway, breathing, circulation or other vital signs. (NICE 157)

Care must be taken to secure the child, young person or adult's airway and assess his or her respiratory and cardiac function. (NICE 148)

Administer buccal midazolam (in line with normal emergency care) as first-line treatment in children, young people and adults with prolonged or repeated seizures in the community. Administer rectal diazepam if preferred or if buccal midazolam is not available. If intravenous access is already established and resuscitation facilities are available, administer intravenous lorazepam. (NICE 156)

Prolonged or serial seizures should be treated with either nasal or buccal midazolam or rectal diazepam. (SIGN)

For most children where information was available, it appeared that an ambulance had been called promptly; typically within 10 minutes of the start of the seizure. However, there were four children where there was evidence of a delay of at least 30 minutes before calling an ambulance. Two of these incidents occurred while the child was at school, and for both there was some indication that the school staff did not have a clear plan to follow and had not recognised the incidents as seizures. It was unclear in the other two children why there had been a delay in calling the ambulance; although for one child the parents did seem to be attempting to manage the child's seizure activity at home.

In two cases, a parent took the child directly to hospital, rather than calling an ambulance. In 17 (57%) of the remaining children, the case assessors found evidence that the ambulance crews had taken appropriate steps to assess the situation, secure the airway/breathing/circulation, and administer appropriate emergency treatment, taking account of any treatment already administered. In most of the other children, the documentation was insufficient for the assessors to determine what actions had been taken by the ambulance crews. In three children (9%), it was deemed that the ambulance crews had not implemented appropriate management, this related to inadequate or delayed administration of benzodiazepines (Table 5.2).

**Table 5.2: Cases where ambulance crews had not instituted appropriate emergency management**

<b>Inadequate dosage</b>	The ambulance was promptly called and arrived 12 minutes later and they administered rectal diazepam, however the dosage was too low. The correct dose was 3-4 times what was given.
<b>Delays in giving rescue medication</b>	The ambulance crew assessed ABC's but there was a 20 minute delay in giving any medication (39 minutes after the start of the seizure). The paramedics gave Intravenous (IV) diazepam. Unclear why they did not give another preparation (not IV) of benzodiazepine which could possibly have been administered more rapidly.
<b>Rescue medication not being administered</b>	An ambulance was called after five minutes of continued seizing, and arrived 20 minutes after the call. There were no further benzodiazepines administered until the child arrived at A&E 40 minutes after the seizing started (these parents had given one dose of buccal midazolam, but had been advised not to give any more doses due to previous respiratory problems).

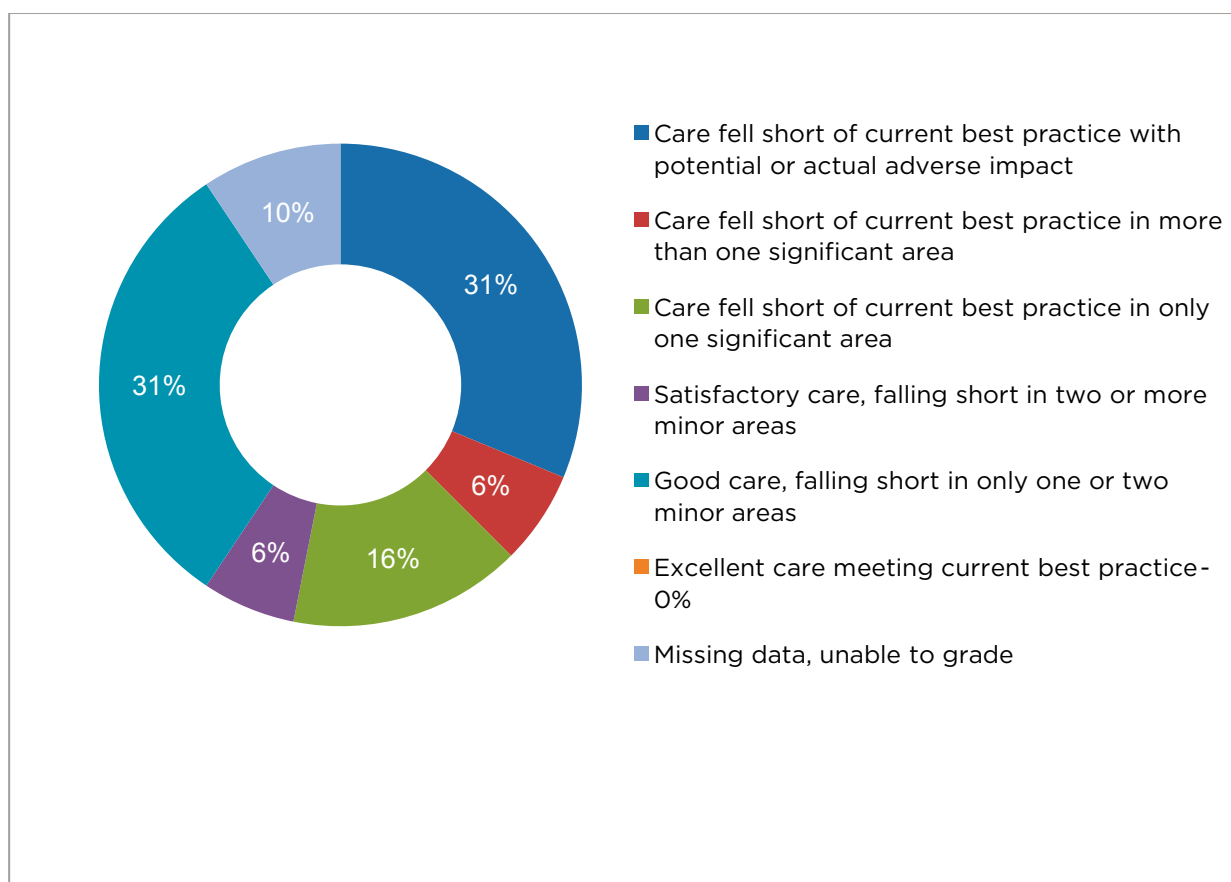
There was only one child where an ambulance crew administered buccal midazolam as emergency treatment. This was in contrast to eight children (28%) where rectal diazepam was given and four (14%) where IV diazepam had been administered. While buccal midazolam is now the treatment of choice for community management of prolonged seizures; it is not currently being used by ambulance crews. The cases highlighted above (Table 5.2) illustrate how this can potentially lead to delays in administering appropriate medication, or inadequate doses being given. The 2006 Joint Royal Colleges Ambulance Liaison Committee Pre-Hospital Guidelines<sup>45</sup> recommend the use of diazepam for children with seizures. These have recently been updated and the new 2013 JRCALC guidelines<sup>46</sup> state that ambulance crews are able to administer the patient's own midazolam provided they are aware of what indications to look out for and are competent and familiar with the use of midazolam. If the patient does not have access to midazolam, or if ambulance crews are unfamiliar with its use, then either IV or rectal diazepam should be used. However, the findings from this review do support the use of buccal midazolam as the drug of choice for ambulance crews.

### 5.1.3 Quality of pre-hospital care

Case assessors judged the overall quality of care in relation to the pre-hospital care to have fallen short of current best practice in one or more significant areas in 17 (53%) children. In 10 children, assessors judged that this resulted in potential or actual adverse impact on the child. In contrast, they agreed the care to be meeting current best practice, or just falling short of current best practice, in only minor areas in 12 (38%) children (Figure 5.1). For three children the case assessors were unable to judge the quality of care, due to the lack of documentation of events in the child’s notes.

An assessment of the quality of care, based on the key questions from the criterion-based review, showed that for 26 children (81%) the care fell short of defined standards in at least some areas and in six children (19%) it was judged not to have met any of the defined standards (Table 5.3). In most cases, this reflected a lack of documentation regarding what took place prior to arrival at the hospital. Nonetheless, as highlighted above, there was, in some cases, a lack of clarity around emergency care plans for the management of prolonged seizures, including when in a school setting. This raises concerns that there may be children in the community at risk of prolonged seizures for whom those responsible for the child’s welfare may be unclear how or when to respond in the event of seizures. Furthermore, there is a concerning gap in the response to seizures by ambulance staff, with delays in the administration of emergency medication and, in some cases, inadequate doses of benzodiazepines being given.

**Figure 5.1: Quality of pre-hospital care as judged by the case assessors**



**Table 5.3: Standards around pre-hospital care**

	<b>Standard</b>	<b>Number (%) with evidence that this standard had been met</b>
1	Was an appropriate trained person available to administer first aid and emergency treatment?	17/32 (53%)
2	Did the child/young person have an emergency care plan, and if so, was this followed?	7/32 (22%)
3	Was an ambulance called at an appropriate time?	14/32 (43%)
4	Did the first responder/ambulance crew take appropriate steps to assess the situation, secure the airway/breathing/circulation, and administer appropriate emergency treatment, taking account of any treatment already given?	17/30 (57%)
5	Were buccal midazolam or other benzodiazepines given in an appropriate dose by the first responder/ambulance crew?	8/30 (27%)

## 5.2 Emergency department care

*The child arrived at A & E with continued seizure activity and was seen by a consultant in emergency medicine. He was treated with buccal midazolam, followed by other benzodiazepines (IV lorazepam, phenytoin, paraldehyde); the drugs were administered promptly and in appropriate doses. The child was then intubated and ventilated within 40 minutes of arrival to A&E due to ongoing seizures, however once the seizures were under control the child was transferred to PICU at another local children's hospital. Overall this was excellent care and the emergency guidelines were followed appropriately. (Case assessor comment)*

On arrival at the emergency department over half the children were continuing to experience a seizure (Table 5.4). Eight children were seen by a consultant, one by an associate specialist and nine by ST4-8 level trainees, with eight of these nine specialising in paediatrics. The remaining children were either seen by a lower grade trainee clinician or the clinician's grade was not recorded. Of those that recorded the speciality of the treating clinician, 10 children were seen by a paediatric specialist, five by an anaesthetist, three by an emergency care specialist and one by an intensivist. In 22 children (69%) there was evidence that appropriate tertiary expertise had been sought by the staff in the emergency department in a timely manner.

**Table 5.4: Condition on presentation to emergency department**

Condition	Number (%)
Fully alert, no longer seizing	1 (3%)
Presumed post-ictal	7 (22%)
Continued seizure activity	18 (56%)
Unconscious	2 (6%)
Not known	4 (13%)
<b>Total</b>	<b>32 (100%)</b>

There was evidence that appropriate steps were taken to assess and secure the airway, breathing and circulation in 24 (75%) children. In one child the assessors felt that the clinicians focused on trying to stop the seizure rather than securing the airway, and for the remaining children there was insufficient information recorded in the notes for assessors to be able to reach a judgement.

For children, young people and adults with ongoing generalised tonic-clonic seizures (convulsive status epilepticus) who are in hospital, immediately: secure airway give high-concentration oxygen assess cardiac and respiratory function check blood glucose levels and secure intravenous access in a large vein. (NICE 158)

Appropriate treatment was administered in 20 (63%) children, taking into account the treatment already given prior to arrival. Sixteen of these children were given rescue medication and four children, who were no longer seizing, were not given any further medication. For six (19%) children it was unclear whether appropriate medication was administered due to the lack of information in the case notes. In a further six (19%) children there was clear evidence that appropriate medication was not provided. In the majority of these children this was a result of the NICE or APLS guidelines not being strictly followed, and the reasoning for taking a different approach was not recorded. For one child the treating clinician had documented the reasons for deviating from the guidelines; this helped to keep all clinical staff involved with the child informed without making any assumptions of incorrect practices. This should be standard practice for all clinicians.

*Because buccal midazolam and PR [rectal] diazepam had been given prior to arrival, the child should have moved onto next level of medication, i.e. IV phenytoin. There was deviation from the status epilepticus guidelines in repeating the third dose of benzodiazepines and gave paraldehyde before IV phenytoin. However, the deviation and administering paraldehyde was because of previous experiences with side effects and was a good treatment option for this child. (Case assessor comment)*

There was one child where delays in appropriate management, failure to follow standard guidelines, and poor communication were felt to have possibly contributed to an adverse outcome.



Administer intravenous lorazepam as first-line treatment in hospital in children, young people and adults with ongoing generalised tonic-clonic seizures (convulsive status epilepticus). Administer intravenous diazepam if intravenous lorazepam is unavailable, or use buccal midazolam if unable to secure immediate intravenous access. Administer a maximum of two doses of the first-line treatment (including pre-hospital treatment). (NICE 159)

If seizures continue, administer intravenous phenobarbital or phenytoin as second-line treatment in hospital in children, young people and adults with ongoing generalised tonic-clonic seizures (convulsive status epilepticus). (NICE 160)

Administer intravenous midazolam or thiopental sodium to treat children and young people with refractory convulsive status epilepticus. Adequate monitoring, including blood levels of AEDs, and critical life systems support are required. (NICE 163)

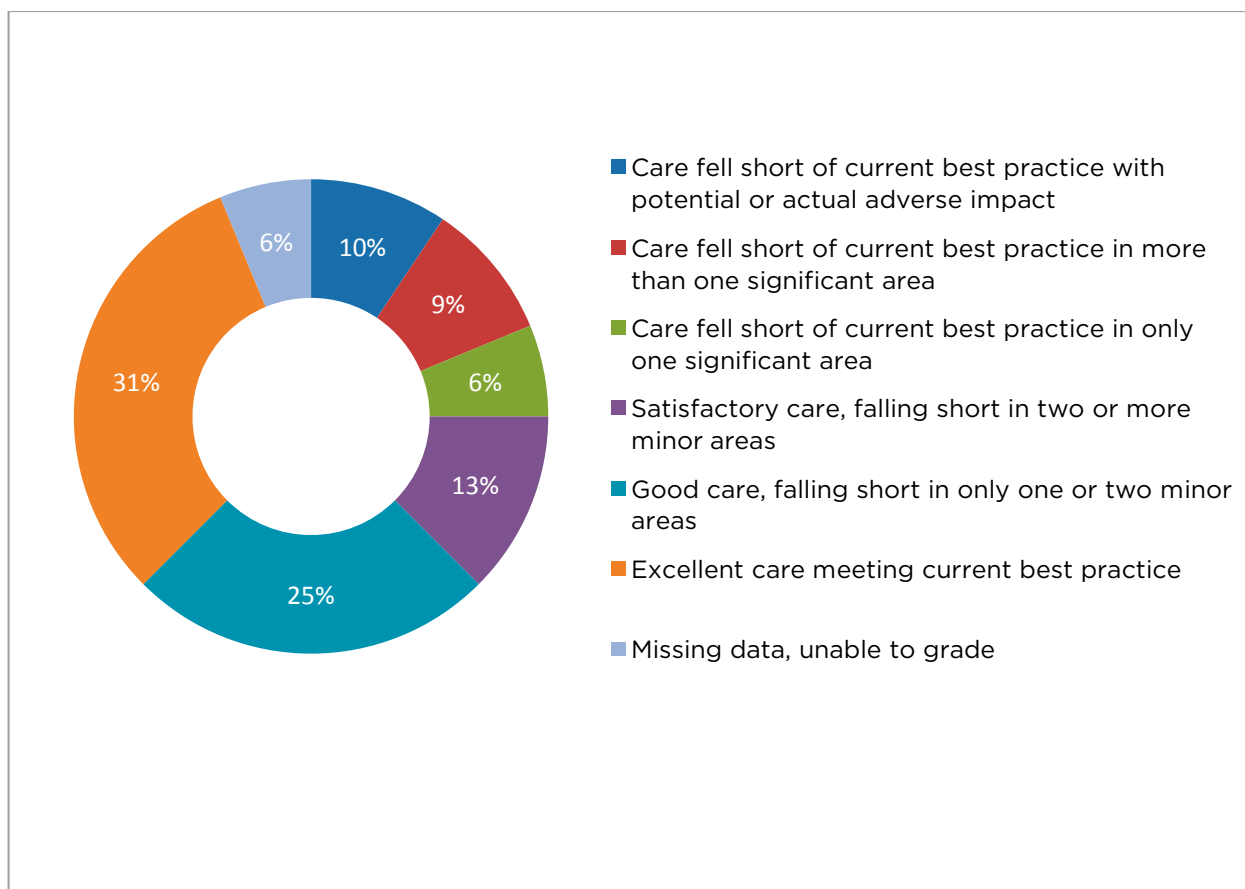
As the treatment pathway progresses, the expertise of an anaesthetist/intensivist should be sought. (NICE 151)

Following treatment in the emergency department, 16 of the children (50%) were admitted to high dependency care and 11 (34%) to intensive care and five (16%) were admitted to a general paediatric or other ward, where intensive or high dependency care was received.

### 5.2.1 Quality of emergency department care

Case assessors judged the overall quality of the emergency department care to have fallen short of current best practice in one or more significant areas in eight children (25%). In three of these children this was deemed to have resulted in the potential for or actual adverse impact on the child. They judged the care to be excellent and meeting current best practice, or falling short of current best practice in only minor areas, in 22 cases (69%) (Figure 5.2). For two children the case assessors felt unable to make a judgement of the quality of care.

**Figure 5.2: Quality of emergency department care as judged by the case assessors**



An assessment of the quality of care was made based on key questions from the criterion-based review which showed that 15 cases (47%) were found to meet all standards and eight (25%) met some of the standards. None of the cases were judged to have not met any of the defined standards (Table 5.5). For seven (22%) children there was a lack of documentation of events that took place in the emergency department therefore it was not possible to determine the quality of the care in relation to these standards. Overall, for most children, the care provided in the emergency department was well managed, with documented evidence of appropriate securing of airways, breathing and circulation, appropriate administration of treatment and appropriate tertiary expertise being sought.

**Table 5.5: Standards around emergency department management**

	Standard	Percentage meeting this standard
1	On arrival in the emergency department, were appropriate steps taken to assess and secure the airway, breathing and circulation?	24/32 (75%)
2	Was appropriate medication given, taking account of treatment already given before arrival?	20/32 (63%)
3	Was appropriate expertise (including an anaesthetist/intensivist) sought in a timely manner?	22/32 (69%)

### 5.3 Children who were already in hospital at the time of their seizure

*The child was on general paediatric ward when they presented a prolonged seizure. A grade ST4-5 trainee was on hand to respond to the child, and an anaesthetist was called in a timely manner, however arrived 1 hour 22 minutes after the seizure activity started. Appropriate steps were taken to control ABC, and midazolam, phenytoin, paraldehyde, oxygen, ventilator support were administered. The child was admitted to HDU initially pending transfer to PICU at another hospital. Overall the care was excellent with well documented information and discussions with parents about appropriateness of full intensive care and transfer. (Case assessor comment)*

Four children were already in hospital for other reasons when they had a prolonged seizure. In one case no information was available regarding the front line hospital treatment; however, the three children where information was available were located on general paediatric wards at the time of the prolonged seizure. On arrival of first line hospital staff, one child was found unconscious and the other two had continuing seizure activity.

Two of the three children where information was available had their airways secured, appropriate medications administered, and the expertise of an anaesthetist or intensivist sought in a timely manner. One of these children was admitted to high dependency care, and the other cared for by a specialist retrieval team. The case assessors rated the care of these two children as being excellent and meeting current best practice, and using the criterion-based assessment it was found to meet standards in all areas.

There was one child for whom the case assessors identified concerns in relation to the hospital management. In this case, there were delays in initiating appropriate treatment leading to a cardio-respiratory arrest. The criterion-based assessment showed the first line hospital management to have fallen short of current best practice in all areas.

*Management of status epilepticus was not according to NICE Guidance. This child had a cardio-respiratory arrest due to the prolonged seizure. The management was poor due to the non-availability of an anaesthetist and a delay in terminating the seizure. The child did not receive lorazepam, paraldehyde or thipentone induced anaesthesia. (Case assessor comment)*

## 5.4 High dependency care and intensive care management

*The child arrived at intensive care intubated and ventilated, with seizures under control. They were appropriately treated with a thiopentone infusion, acyclovir, cefotaxime and carbamazepine. There was adequate monitoring during the child's time on ICU. They were ventilated for a total of seven hours and eventually fully recovered to their pre-admission state. Overall the care was appropriate and following this episode an early outpatient department review was arranged and there was an increase in AED medication. (Case assessor comment)*

Thirty six children received high dependency (19; 53%) or intensive care (17; 47%) following a prolonged seizure. The majority of children were no longer having seizures on admission to high dependency or intensive care and were either post-ictal (11 children) or intubated with their seizures controlled (12 children); seven children were still having seizures and for six children, their state on admission was not known.

Administer intravenous midazolam or thiopental sodium to treat children and young people with refractory convulsive epilepticus. Adequate monitoring, including blood levels of AEDs, and critical life systems support are required. (NICE 163)

If either the whole protocol or intensive care is required the tertiary service should be consulted. (NICE 152)

Ten children (28%) were treated with intravenous midazolam or thiopental sodium and 10 (28%) with other anti-epileptic drugs. Eleven children (31%) did not receive any further anti-epileptic medication after admission to intensive or high dependency care. In all but one of these, the seizures had either stopped, or had been brought under control following intubation and ventilation. The exception was a child with an infantile-onset epileptic encephalopathy. This child had been brought directly to the paediatric ward for assessment due to an increased frequency of seizures. On the ward, the child was given repeated rectal doses of diazepam. The child continued to have seizure activity, and although a phenytoin infusion was considered, there was no clear plan and it was never administered. The case assessors rated this child's care as falling short of current best practice with the potential for or actual adverse impact on the patient.

*There was a plan in place regarding the management of further seizures but nothing documented regarding the timing of action. Also, there was no record as to whether contact had been made regarding anaesthetic staff if the condition deteriorated overnight. The child was transferred out of the resuscitation area on the ward to the main ward the following morning. The seizures were not completely settled for a further 72 hours with further administration of buccal midazolam on 3 separate occasions. The first documentation in the chart of a paediatric consultant reviewing the child in person was on day 3 on the ward round; the consultant may have been there before but this was not clearly documented. (Case assessor comment)*

A total of 15 (42%) children were ventilated either on admission or subsequent to their admission to intensive or high dependency care. The children were ventilated for anything from one hour to 70 days, with a median of 22 hours.

For 22 children (61%) the case assessors found evidence of adequate monitoring during their high dependency or intensive care stay. Four children (11%) were felt not to have had adequate monitoring, due to delays in starting observations or observations not being taken as often as case assessors thought appropriate. For the remaining 10 (28%) children it was difficult to determine the level of monitoring from the children's case notes. There was evidence that appropriate tertiary expertise had been sought for 24 (67%) children; in most cases this was from a paediatric neurologist.

*There was inadequate monitoring in the early phases as monitoring was hourly neuro obs and the assessors expected 15 mins to 30 mins neuro obs in the first few hours of admission/midazolam infusion. Also there was a 12 hour delay in doing any blood tests, which potentially may have been due to the child not being ventilated, but with altered neurology and receiving midazolam infusion simply recording that sats levels are ok wasn't thought to be enough. The child was on hourly observation and generally monitored but a little concern in that the conscious level was recorded as 'asleep'. Saturations were fine during this time. Over a seven hour period the child was not roused once. (Case assessor comment)*

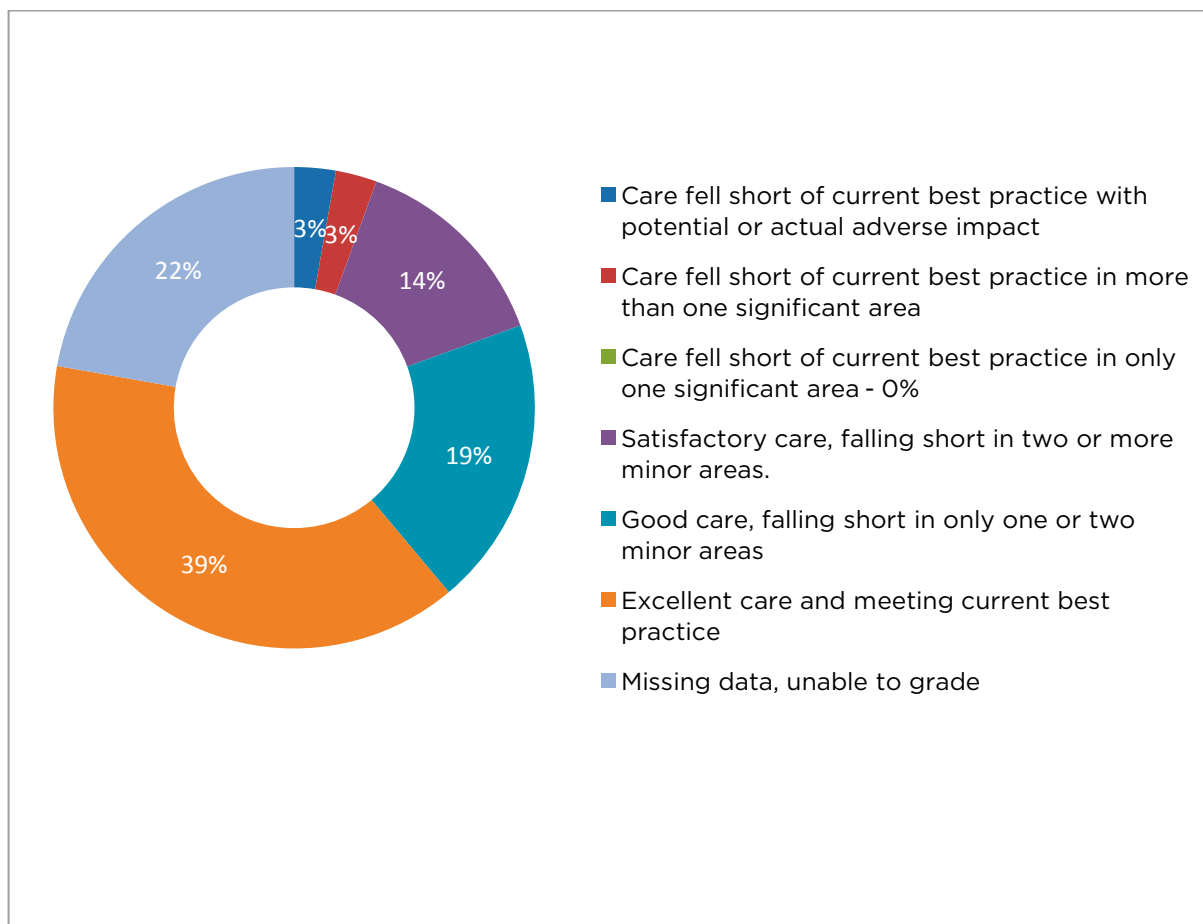
#### 5.4.1 Outcomes of the intensive/high dependency care

Twenty three children (64%) fully recovered to their pre-admission state, three children (8%) fully recovered but with residual new impairment, eight children (22%) died and information was not recorded for two children (6%). The eight children who died are considered in more detail in Chapter 6.

#### 5.4.2 Quality of intensive/high dependency care

Case assessors judged the overall quality of the intensive or high dependency care to have fallen short of current best practice in one or more significant areas in two children, in one of which this was deemed to have resulted in the potential for or actual adverse impact on the child. They judged the care to be excellent and meeting current best practice, or falling short of current best practice in only minor areas in 26 cases (72%; Figure 5.3). Case assessors felt unable to make a judgement of the quality of care in eight children due to lack of information provided in the case notes.

**Figure 5.3: Quality of intensive/high dependency care as judged by the case assessors**



An assessment of the quality of care based on key questions from the criterion-based review showed that 14 children (39%) were found to meet all standards; eight (22%) met some of the standards; and there were no children that were judged not to have met any of the defined standards (Table 5.6).

**Table 5.6: Standards around intensive and high dependency care**

	Standard	Percentage meeting this standard
1	If the child had ongoing seizure activity, was appropriate treatment with intravenous midazolam or thiopental sodium given?	3/7 (43%)
2	Was adequate monitoring of the child in place throughout the intensive care stay?	22/36 (61%)
3	Was appropriate tertiary expertise consulted?	24/36 (67%)

## 5.5 Summary

The expectation when carrying out this review was that the children requiring intensive or high dependency care for prolonged seizures would be those with difficult-to-control epilepsies, presenting with frequent seizures and multiple hospital admissions. This was not universally the case. Indeed, the majority of the children were having infrequent tonic-clonic seizures (less than once a week); 56% had had no hospital admissions in the previous 12 months; and 47% were only receiving one anti-epileptic drug, with two children not on any anti-epileptic drugs. Thus it is important that families and professionals recognise the risks associated with prolonged seizures, even in children whose epilepsy appears to be well-controlled. It is essential that all children with epilepsies should be regularly monitored, and should have a clear emergency care plan for the management of prolonged seizures, including the use of buccal midazolam, backed up by appropriate training for parents and carers. The majority of children requiring intensive or high dependency care had associated developmental impairments, and for this group of children it is even more important that their epilepsy is reviewed regularly with an emergency care plan forming part of their overall package of care.

This review identified some key concerns surrounding the care received by these children prior to arrival at hospital. In many children this seemed to reflect a lack of forward planning, with absent or unclear emergency care plans, leaving parents uncertain or untrained in how to respond to their child's seizures. Issues surrounding out of date or unclear emergency medication were also highlighted, together with the danger of inappropriate doses of rescue medication (primarily due to the different strengths of the two most commonly used preparations of buccal midazolam). This emphasises the importance of clear discussions with parents and carers, on a regular basis, around how to respond should the child have prolonged seizures. This is important for all children with epilepsies, but particularly where the child is known to have suffered or be at high risk of prolonged seizures. A clearly documented and up to date emergency care plan for parents and carers with copies in all relevant clinical notes was identified in some of the cases, and this is good practice that should be replicated for all children with epilepsies.

Furthermore, it should be recognised that schools need thorough, clear care plans and training in rescue medication in order to treat children who are having prolonged seizures. The findings showed that in some cases school staff and staff in residential care homes did not have a clear plan to follow, and did not recognise seizure activity in the child, resulting in delays in calling for an ambulance. Clearly documented and up to date emergency care plans for schools are required to maintain good practice.

There was evidence of good initial assessment and response by ambulance staff attending, but some concerns regarding the administration of appropriate rescue medication. The use of outdated modes of treatment (rectal or IV diazepam) or inadequate doses appeared to have contributed to delays in stopping seizures in the children. Clear guidelines backed up by appropriate training and availability of buccal midazolam could help to ameliorate this risk. The current national guidelines (JRCALC, 2013)<sup>46</sup> go some way towards this, in allowing ambulance crews to administer the child's own midazolam provided they are both competent and familiar with the use of midazolam and indications to look out for. However, given its proven efficacy, all ambulance crews should be equipped with buccal midazolam and ambulance staff trained in its use. The local guidelines should be updated to recommend its use as the first line treatment in children with prolonged seizures.

There was good evidence that the emergency, high dependency and intensive care provided to children presenting with prolonged seizures was meeting high standards of care. However, in some children the management deviated from standard APLS<sup>47</sup> and NICE guidelines.<sup>28</sup> Emergency departments should ensure that all medical and nursing staff are able to apply current NICE/APLS prolonged convulsion guidance. This includes ensuring availability of and competency with buccal midazolam, IV lorazepam and IV phenytoin administration. Deviation from the standards may have been appropriate for some children, representing a responsive approach to the child's needs; however, if the reasons behind decision making are not clearly documented, it is difficult to judge whether such departures from established guidelines are appropriate.

In some cases evidence was found of good practice following an intensive or high dependency care admission for some children, including the clinical team reviewing the child's overall care, and making appropriate adjustments to their management and follow up. A careful review of the circumstances around the care provided during any intensive or high dependency care admission for prolonged seizures that will involve the family and professionals providing ongoing care to the child, will help to ensure that appropriate plans are put in place for ongoing management and for responding to future incidents.

### 5.5.1 Key findings and recommendations

**Key finding 6:** Many of the children in this review experienced repeated hospital admissions for prolonged seizures. This along with the multiple co-morbidities, a lack of forward planning and appropriate care plans being in place highlighted the potential danger of clinicians focusing on the management of individual acute episodes, and the failure of anyone to step back and consider the wider ongoing long term needs of the child. In such situations, it is vital to ensure each child receives regular coordinated reviews of their epilepsy management.

#### **Recommendation 4**

Whenever a child is admitted to hospital with a prolonged seizure, the consultant responsible for the admission should notify the clinician in charge of the child's overall care. The clinician with overall responsibility should then review the child's epilepsy management in the light of that admission.

**Key finding 7:** This review has highlighted the importance of clear and comprehensive care plans for parents, schools and others caring for children with epilepsies, and providing them with information on how to respond to prolonged seizures, including training in resuscitation and the use of rescue medication. This is important for all children with epilepsies, but particularly where the child is known to have suffered or be at high risk of prolonged seizures. Such care plans could be included in an 'epilepsy passport' as highlighted in **Recommendation 3** (Chapter 4). This finding supports the recommendations on emergency care plans as set out in the NICE and SIGN guidelines.

**Key finding 8:** The different formulations of buccal midazolam currently in use give rise to potential medication errors because of different dilutions (5mg/ml or 10mg/ml). This can give rise to either under or over-dosing, particularly when children are changed from one formulation to another.



**Recommendation 5**

When prescribing buccal midazolam for rescue medication in prolonged seizures, prescribing clinicians must clearly state the formulation being used and the dose to be given in both mg and ml. The consultant in overall charge of the child's epilepsy care should ensure that the parents and all other carers have an up to date emergency treatment plan that clearly outlines the dose to be given and circumstances in which to give the rescue medication.

**Key finding 9:** Evidence was found of good initial assessment and response by ambulance staff, but some concerns were highlighted around the timing and administration of appropriate rescue medication. The findings from this review support the recent updated guidelines from the Joint Royal Colleges Ambulance Liaison Committee (JRCALC). However, these guidelines could be further strengthened, with updates of all local guidelines, to ensure all ambulance crews are trained and equipped to be able to administer buccal midazolam to children experiencing prolonged seizures.

**Recommendation 6**

Ambulance Trusts should consider updating their protocols for seizure management in children and young people, to recommend the use of buccal midazolam as the first line treatment for prolonged seizures. This should be backed up with appropriate training of all ambulance crews in the use of buccal midazolam, and provision of buccal midazolam to all ambulance crews.

**Key finding 10:** There was good evidence from this review that both the emergency department and high dependency or intensive care provided to children presenting with prolonged seizures was, on the whole, meeting high standards of care. Nevertheless all emergency departments must ensure that their clinical staff are able to apply current NICE and APLS prolonged convulsion guidance, as well as ensuring availability and competency with buccal midazolam, IV lorazepam and IV phenytoin administration. Deviation from these standards may, in some cases, be appropriate but this information should be clearly documented, as stated in **Recommendation 1** (Chapter 4).

**Recommendation 7**

Emergency departments should ensure that children and young people presenting with prolonged seizures are treated according to current NICE and APLS guidance through appropriate departmental guidelines, training of staff and audit.

**Key finding 11:** Admission to intensive or high dependency care provides an opportunity for reviewing the child's overall care, and making appropriate adjustments to their management and follow up, as well as reflecting on the care provided and learning lessons locally.

## 6. The care of children with epilepsies who died and their families

*The child had deteriorated over several years to a severely disabled and completely dependent state. Seizures were an ongoing problem and worsened in the terminal phase. However the family received full palliative care support in the home, hospital and hospice. This child was still admitted via open access to a children's assessment unit with serial seizures (approx. 30 secs in duration, 22 seizures in last 24 hours) and was thus admitted to high dependency care for close monitoring. The child later died of a complex neurodevelopmental degenerative disorder, which was known to be a life-limiting condition. (Case assessor comment)*

During this review a total of 46 questionnaires were submitted relating to deaths in children with epilepsies. Of these, nine (20%) had received intensive or high dependency care for a prolonged seizure prior to their death. The majority of children (29; 63%) were at home when they died, or at the start of the incident that led to their death. The majority of these were then transferred to hospital where most deaths were confirmed (Table 6.1). 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.

**Table 6.1: Location of children at time of terminal incident and when death was confirmed**

Location	At the time of the incident	When death was confirmed
Home of normal residence	29 (63%)	6 (13%)
Hospital	6 (13%)	31 (67%)
Hospice	5 (11%)	9 (20%)
Other residence (residential care, respite)	3 (7%)	0 (0%)
School	1 (2%)	0 (0%)
Not known	2 (4%)	0 (0%)
<b>Total</b>	<b>46 (100%)</b>	<b>46 (100%)</b>

The 46 children who died varied in their reported characteristics. The majority of children (43; 94%) were noted to have associated developmental impairments and co-morbidities, many of which were severe and required considerable ongoing care. Thirty-three children (72%) had either an identified cause for their epilepsy or epilepsy syndrome.

Sixteen children (35%) were having seizures at least weekly, although these rarely resulted in hospital attendances; only three children had attended hospital for prolonged seizures in the previous 12 months. This indicated that parents were generally managing the seizures at home. Nevertheless, this suggests that for a proportion of these children seizures were a significant part of their overall morbidity. For the remaining children, the epilepsy itself did not seem to be a major component of their care needs, with infrequent seizures and rare hospital attendances for seizures.

## **6.1 Cause of death**

For the purposes of this review, a classification system was developed based on the likely major categories of death, and linked to existing ICD-10 codes for registering the cause of death (Appendix 4). For 33 cases which underwent case note review, the assessors were asked to determine the category of death based on information from the death certificate where available and supplemented by their review of the clinical notes. For the remaining 13 cases the classification of death was based on information from the clinical questionnaire. In addition, the project clinical lead reviewed the case notes and clinical questionnaires of all children who had died to clarify the categorisation of the death using the classification of death table.

Over half (26; 57%) of these children died of causes other than their epilepsy. The largest group (52%) were children who died as a consequence of a co-morbidity associated with their epilepsy. This proportion was followed by definite or probable SUDEP (15%), and deaths secondary to status epilepticus (15%). There were two children who died from causes unrelated to the epilepsy and one child's death may have been related to treatment administered for the epilepsy.

**Table 6.2: Classification of death**

	<b>Classification</b>	<b>Number (%)</b>	<b>Notes</b>
A	Death from a cause unrelated to the epilepsy	2 (4%)	Two children died of conditions unrelated to their epilepsy. Both children were also reported to have associated developmental impairments, but these were not related to the cause of death.
B	Death from a co-morbidity associated with the epilepsy	24 (52%)	Mostly these were children with complex co-morbidities who died of respiratory infections/respiratory failure in association with their co-morbidities.
C	Death as a consequence of treatment given for epilepsy	1 (2%)	One child died of a cause secondary to low platelets, for which anti-epileptic drugs are a recognised side effect. The death was therefore classified as a possible consequence of a treatment given for epilepsy.
D	Trauma associated with a seizure	0 (0%)	There were no children in whom the clinical questionnaire or review of the clinical records indicated that the child had died as a consequence of trauma associated with a seizure.
E	Death secondary to status epilepticus	7 (15%)	This category was used for all children in whom there was documented evidence of convulsive status epilepticus (continuous, convulsive seizures lasting at least 30 minutes, or two or more seizures during which the child does not return to baseline consciousness). Cases were excluded if the seizure had led to trauma as the direct cause of death, or if the death was directly caused by treatment given to stop the seizure.
F	SUDEP 1. Definite SUDEP 2. Probable SUDEP 3. Possible SUDEP	7 (15%)	This classification was used if the case met the criteria of a sudden, unexpected, witnessed or unwitnessed, non-traumatic and non-drowning death, occurring in benign circumstances, in an individual with epilepsy, with or without evidence of a seizure and excluding documented status epilepticus (49). Cases were labelled definite SUDEP if a post-mortem examination had not revealed a cause of death, and probable SUDEP if there had been no post-mortem examination, or if the results were not available to the reviewers. Possible SUDEP was used if a possible competing cause of death was found, but not proven.  There were two cases that satisfied the criteria for Definite SUDEP and five for Probable SUDEP.
	Unknown	5 (11%)	In five cases there was insufficient information in the case notes and clinical questionnaire to be able to classify the death.
	<b>Total</b>	<b>46 (100%)</b>	

### 6.1.1 Deaths from causes other than the epilepsy

*This young person initially presented with seizures at five years of age and was diagnosed with a severe and life-limiting neuro-degenerative disorder. The child was looked after by a multidisciplinary team, including a palliative care team, and the family seemed to be well supported through a variety of professionals. The child had been cared for at home by the family and had not required any hospital admissions during the previous 12 months. In the six months prior to death there had been an increase in opioids for pain relief and antibiotics for a chest infection. There was a documented care plan for management of prolonged seizures, including rescue medication and also an end of life care plan which was updated regularly to reflect changes with the child's ongoing needs. The parents were involved in discussions around all aspects of care including end of life plans. The child had gradually deteriorated over several years to a severely disabled and completely dependent state. Seizures were an ongoing problem and worsened in the terminal phase. The child was receiving palliative care in a hospice at the time of their death. (Category B: death from a co-morbidity associated with the epilepsy) (Case assessor comment)*

The majority of these deaths were in children with complex co-morbidities and reported associated developmental impairments who died of respiratory infections or failure. The commonest associated condition was severe cerebral palsy secondary to prematurity. For many of these children the death was anticipated and an end of life care plan or other palliative care measures had been put in place. The two children who died of unrelated causes also had associated developmental impairments. For all of these children, epilepsy was one component of their overall care. For some, there was evidence that seizure control was an issue that can cause additional stress for the children and their carers.

### 6.1.2 Deaths secondary to status epilepticus

*This child with Dravet syndrome was admitted to a paediatric intensive care unit following a prolonged seizure which started at home. In the previous 12 months the child experienced numerous admissions to hospital with prolonged seizures. There was an emergency and school care plan in place. Although the parents gave buccal midazolam and called an ambulance promptly, there were no further benzodiazepines administered until the child arrived at A&E, and it was over an hour before the seizures were brought under control. Once on the intensive care unit, there was appropriate medical management and monitoring, but the child deteriorated with hypoxic ischaemic encephalopathy, multi-organ failure and brainstem death. A decision was made therefore to withdraw care. (Category E: death secondary to status epilepticus). (Case assessor comment)*

Seven children, aged one to 14 years, died in the context of status epilepticus. Where there was further clinical information available, it indicated that the child had suffered a cardiac or respiratory arrest during the seizure and either could not be resuscitated, or went on to develop multi-organ failure on the intensive care unit. In two cases, the cardiac or respiratory arrest had occurred during treatment for the status, but in neither case was the death considered a direct consequence of the treatment. There were two cases where the clinical questionnaire suggested the child died from aspiration secondary to a seizure. However, this could not be confirmed as the cause of death in either case. There were no other cases where the clinical information suggested that the child had died as a result of trauma associated with a seizure.

Of these seven children, five had associated developmental impairments. Three of the children were experiencing frequent seizures (daily or weekly) in the six months prior to the admission, two of whom had been previously admitted for prolonged seizures. One of these three children was on one anti-epileptic drug, in spite of daily seizures and multiple hospital admissions. There were four children whose epilepsy was considered to be well controlled before the prolonged seizure that resulted in their death, with none having seizures more frequently than once a month, or requiring admission to hospital in the previous 12 months. For these four children, the prolonged seizure was therefore unexpected.

### 6.1.3 Sudden Unexpected Death in Epilepsy (SUDEP)

*A child with West syndrome and associated developmental impairments was found unresponsive in bed early in the morning. Given the complexity of the condition, the child had involvement from an extensive multidisciplinary team. The child was having daily seizures and was kept under regular review. The child was on multiple anti-epileptic drugs, which had recently been increased to maximum doses. The child experienced previous admissions with prolonged seizures in the 12 months prior to the death. An ambulance was called and the child was quickly transferred to the emergency department, but did not respond to ongoing cardio-pulmonary resuscitation. (Category F: Sudden Unexpected Death in Epilepsy). (Case assessor comment)*

Seven children aged between three and 16 years died suddenly and unexpectedly with no specific cause of death being identified. All seven were found in their beds by their parent or carers. In two cases, details of negative autopsy findings were recorded in the notes and these were therefore classified as definite SUDEP; one of these had a known chromosomal abnormality in association with the epilepsy. For the remaining five, autopsies may have taken place, however, the information was not recorded in the clinical notes and they were classified as probable SUDEP. Four of these had documented associated developmental impairments.

Five of the seven children appeared to have difficult-to-control epilepsy with daily seizures and previous hospital admissions for seizures within the last 12 months. All five were on multiple anti-epileptic drugs and several had required adjustments to their medication in the previous six months. There were two children whose epilepsy did not appear to be difficult to manage, and neither were reported to be having frequent seizures or had been admitted to hospital in the 12 months prior to their death. Both were managed on a single anti-epileptic drug.

There was one further child for whom the clinical questionnaire listed SUDEP as the cause of death. This was a child with cerebral palsy and associated developmental impairments who died in a hospice. This particular child's notes were not available for review and it was not therefore possible to confirm whether SUDEP was the correct classification for the child's death.

## 6.2 Factors contributing to the deaths

Of the 46 children who died, notes were received for 33 (72%) cases enabling a more in-depth review. Further analysis in this chapter is based on these case assessments, supplemented where appropriate by data from the clinical questionnaires.

In keeping with the approach used by Child Death Overview Panels (CDOPs), the case assessors were asked to identify any factors, in the family and environment, the parenting capacity, or in relation to service provision which may have contributed to the death. For most cases, there were identified factors intrinsic to the child. In particular, the severity of their co-morbidities with associated feeding difficulties and respiratory problems together with the complexity of their underlying epilepsy with poorly controlled seizures. The case assessors identified a small number of issues in relation to the family, environment or parenting capacity. This included some issues with family engagement or adherence with management and factors affecting the parents' ability to cope, or their response to the child's illness (Table 6.3). There were non-English speaking families for whom communication was considered to have been an issue. In two cases child protection concerns were identified. However, in neither case was this considered to be directly linked to the child's death. There were a minority cases where issues relating to service provision were identified (Table 6.4).

**Table 6.3: Factors in the family and environment contributing to the child's death**

Factors in the family and environment, including parenting capacity	Examples
Family engagement/adherence	Previous evidence of care concerns stopping AEDs, missing appointments.
Parental response to illness	Child had been unwell on the evening prior to admission - mother had not sought advice but phoned the ward the next day. Had not appeared to recognize that her child was very unwell. Took child to hospital on a bus rather than call ambulance.
Parental ability to cope	Parents found it difficult to cope with his frequent seizures and difficult behaviour. Two weeks prior to death there had been a child in need meeting because of the child's sleep problems and concern at the mother's ability to cope.
Child protection concerns	<p>In GP records there is a mention of child protection issues including possible domestic violence - this is not mentioned at all in the hospital notes. There was a multidisciplinary risk assessment conference carried out when the child was six weeks old.</p> <p>Records indicate that severe brain injury in early infancy was most likely non-accidental, but child had been returned in infancy to biological family. Child protection status at the time of death was not recorded but there was no evidence of neglect.</p>
Communication needs	The family is from overseas and have some limited English. It seems they are a hard to reach family. There were missed appointments over a long period of time.
Residential care	Child was in residential care with limited nursing care and inadequate monitoring.

**Table 6.4: Service provision factors relating to the child's death**

Factors in relation to service provision	Examples
Fragmentation of care	Child seen with a respiratory infection a week before death following a prolonged seizure. No review of AED undertaken.
Lack of tertiary input	No documented input from paediatric epilepsy specialist or tertiary neurology input.
Response to acute presentation	Failure to recognise the seriousness of the child's condition at an earlier stage.
Resource issues	Lack of appropriate beds and nursing care for an adolescent with severe behaviour problems and learning difficulties.



### 6.3 Identifying potentially modifiable factors

Having reviewed the case records, the case assessors judged eight cases (24%) to be preventable according to the Department for Education definition:

*'A death is considered preventable if the assessor has identified one or more factors, in any domain, which may have contributed to the death of the child and which, by means of locally or nationally achievable interventions, could be modified to reduce the risk of future child deaths'* Department for Education<sup>38</sup>

Sixteen deaths (49%) were judged not to be preventable, and for nine (27%) there was insufficient information. In the eight deaths which were judged to be preventable, the identified factors were particularly around the following:

- fragmentation of care
- support for families in responding to emergencies
- hospital responses to the acutely unwell child, including those presenting with status epilepticus

*Although not immediately life-limiting, discussion with the family around hospital care and acute illness would have been entirely appropriate. In relation to service provision, there was no paediatrician in charge of overall care. No evidence of a hospital care plan, no plan for chest deterioration at home i.e. with use of antibiotics. No epilepsy review for 18 months before death despite medication changes. Multiple professionals involved but no coordinator of care. (Case assessor comment).*

Fragmentation of care is a particular issue where children have epilepsy in conjunction with other complex co-morbidities. Often there will be multiple professionals involved, and the child may have frequent hospital attendances. It is important in such instances that one professional takes an overview and that the family should have a clear point of contact. For these children who may have recurrent acute episodes with a background of multiple health needs, families need clear advice and support in responding to emergencies. This should include written emergency care plans for the management of seizures and other acute events, which are comprehensive, accessible and easy to follow. These care plans should be discussed with parents and carers, along with risks and management, and supported in writing. If the child is in residential care there should be appropriate training and procedures in place for the care of the children. There are particular issues where English was not the family's first language, or where there were background social factors which may affect a parent's ability to cope.

The review identified a number of locally or nationally achievable interventions in relation to coordination of care, which could be modified to reduce the risk of future child deaths:

- Ensure that children with epilepsies or other complex needs who are looked after have a clear care plan for the management of their seizures, other acute events and aspects of their care
- Ensure each child has a named professional with overall coordination of their care
- Regular epilepsy review, particularly where there are medication changes
- Ensure that staff looking after children in residential care have appropriate training and procedures in place for the care of the children

*This child was unwell for a few days prior to the admission and was given antibiotics by the GP for a presumed chest infection. The child became increasingly unwell the night before. The following morning, the mother contacted the ward for advice. She was advised to call an ambulance but instead brought the child to hospital via a bus. There were background social issues, increasing the vulnerability of this child and the family. The case assessors felt there could have been better parent education around recognition of the sick child. Having an emergency care plan in place with advice for the mother on when medical attention should be sought, particularly for likely medical emergencies as this was a vulnerable child and likely to become unwell from respiratory or seizure-related causes. (Case assessor comment).*

If a child presents to a hospital or community services, including primary care, and is deemed well enough to be cared for at home, this needs to be supported with clear, documented advice to the parents or carers. This should provide them with knowledge of what signs to look for that might indicate any further deterioration, and empower them to seek further help promptly.

Possible interventions in relation to communication with families, which could be modified to reduce the risk of future child deaths included:

- Clear advice to parents and carers on what signs to look for that may indicate their child is acutely unwell, and what steps to take in response to this
- Clear information given to families in a manner they can understand, with use of interpreters and translated written information as appropriate
- Careful discussion of risks and management with families, backed up in writing

*There were identified delays in appropriate management of this child's status epilepticus both in the community and in the emergency department. There were long delays before the child received any benzodiazepines and before the child was seen by an anaesthetist and definitive treatment started to bring the seizures under control. During this time the child had been hypoxic and hypotensive. A focus on controlling the seizures meant that the airway, breathing and circulation had not been adequately secured. Issues were identified in the team working on the child, the communication between paediatric and anaesthetics and staff skills in managing status epilepticus and priorities of ABC. These issues had been identified in a hospital Root Cause Analysis and led to recommendations to review staff training in management of status epilepticus and team working skills between departments. (Case assessor comment).*

There were three cases where the case assessors identified concerns around the hospital response to an acute episode, with issues in relation to recognition of severity of illness, together with the promptness and appropriateness of responses to status epilepticus. Emergency departments should ensure they have and follow clear guidelines, for the management of status epilepticus, in keeping with NICE guidelines. Hospital-based training should be provided. Furthermore, all ward staff should be trained in paediatric advanced life support and take note of all aspects of the child's presentation, not just vital signs. Communication and team work were also flagged up as contributory factors in one of these cases, highlighting the need for appropriate channels of communication between the different clinical teams in hospitals.

Based on these findings, a number of interventions could be suggested around hospital care of children with epilepsies, which could reduce the risks of further child deaths:

- Ensure emergency departments have and follow clear guidelines for the management of status epilepticus, in keeping with NICE guidelines
- Ensure there are appropriate acute channels of communication between different clinical teams in the hospital
- Hospital-based training in the management of status epilepticus
- Ensure ward staff are all trained in paediatric advanced life support and management of prolonged seizures, and take note of all aspects of the child's presentation, not just the vital signs

It was notable that even in children with recognised complex co-morbidities and life-limiting conditions, the case assessors identified potentially modifiable factors in their care. These children deserve the same degree of care as any other child, and therefore, regardless of any underlying life-limiting condition, all steps should be taken to optimise their seizure control, minimise symptoms and respond to any acute deterioration. Careful coordination of their care, with consideration of all aspects of their condition; a named professional with overall responsibility for the care; close partnership working with the parents and carers; and the provision of both emergency care plans and anticipatory end of life care plans are important components of these children's care.

Of the seven children who died as a result of status epilepticus, there were two in whom the case assessors identified potentially modifiable factors in their care. In both, there were considerable delays in administering benzodiazepines and other definitive treatment to stop the seizures. In addition, there were issues identified in relation to perceived failure to follow NICE guidelines for the management of prolonged seizures, and in relation to communication between paediatric and anaesthetic teams in hospitals. As highlighted in section 6.1.2, three of these children had apparently difficult-to-control epilepsy with frequent seizures, previous hospital admissions and changes to their medication in the months prior to their death. It is essential that there is a coordinated approach to the care of these children's epilepsy, with a single consultant with relevant expertise taking overall control of the individual's care, and ensuring that there are regular scheduled reviews, particularly where there are ongoing seizures or hospital admissions. For these children, prolonged seizures may not be avoidable. However, it is important to ensure that the families are aware of the risks and have appropriate emergency care plans in place to respond effectively.

Four of the children who died as a result of status epilepticus had infrequent seizures and risk of prolonged seizures would not have been anticipated. Families should be made aware that, although rare, prolonged seizures can occur spontaneously in otherwise well-controlled epilepsy, and can be fatal. It is essential that all children with epilepsies and their families have information on the management of prolonged seizures and know how to respond appropriately. The potential risks need to be clearly discussed with the families following diagnosis and at regular intervals thereafter.

In none of the cases of SUDEP did the case assessors identify potentially modifiable factors in their care. As highlighted in section 6.1.3 above, the majority of these children had associated complex co-morbidities and also had difficult-to-control epilepsy. Nevertheless, there were two who had apparently well-controlled epilepsy with infrequent seizures. It is important therefore that all parents of children with epilepsies are alerted to the possibility of SUDEP, although the risks in otherwise uncomplicated epilepsies are low. Where the child has associated complex co-morbidities or difficult to control epilepsy, the risks are heightened and the parents should be carefully and sensitively advised of this. In addition, consideration should be given to monitoring the child appropriately, including overnight monitoring.

## 6.4 Children with life-limiting conditions: management prior to the death

*Having a specific diagnosis was really helpful in informing the management plan and discussions with the family about the child. They were clear they did not want long term ventilation and were involved in all discussions and decisions about this care and levels of intervention. There was discussion in advance with the coroner about the care plan, so all went smoothly at the time of death. (Case assessor comment)*

Eighteen of the 33 (55%) children whose cases were subject to more detailed case notes review were judged to have a recognised life-limiting condition. Seventeen (17/18) of these had evidence in the case records that the prognosis had been discussed with the family, and 12 (12/18) had an end of life plan agreed with the family and documented in the records. This suggests an overall good approach to anticipatory planning in keeping with the ACT guidelines.<sup>37</sup>

Every family should receive the disclosure of their child's prognosis in a face-to-face discussion in privacy and should be treated with respect, honesty and sensitivity. Information should be provided both for the child and family in language that they can understand. (ACT standard 1)

Every child and family should be helped to decide on an end of life plan and should be provided with care and support to achieve this as closely as possible. (ACT standard 5)

## 6.5 Management following the death

On reviewing the case notes, the case assessors deemed that 14 (42%) deaths were unexpected, 15 (46%) expected, and in the remaining four cases there was insufficient information to reach a conclusion. Overall, there was evidence from the case notes or clinical questionnaire that 17 cases (52%) had been referred to the coroner or procurator fiscal, including 10 of the 14 unexpected deaths. For the remainder, a referral may have been made, but this had not been recorded in the case notes. National guidelines in England stipulate that when a child dies unexpectedly, there should be a coordinated multi-agency 'rapid response'. Six out of 11 (58%) unexpected deaths in England had evidence in the case records that a 'rapid response' had been undertaken.

Those professionals involved (before and/or after the death) with a child who dies unexpectedly should come together to respond to the child's death.

The joint responsibilities of these professionals include:

- responding quickly to the unexpected death of a child
- making immediate enquiries into and evaluating the reasons for and circumstances of the death, in agreement with the coroner
- providing support to the bereaved family, and where appropriate referring on to specialist bereavement services

(Department for Education)

### 6.5.1 Investigation of the death

For 19 (58%) children there was evidence in the case records that an appropriately trained clinician had taken a clinical history, examined the child and arranged for appropriate investigations either before or after the child's death. In 11 (33%) cases, a report of the clinical findings was provided to the coroner or procurator fiscal either verbally or in writing (65% of those that had been referred). In five cases (15%) there was evidence that an autopsy had been carried out, and in a further eight (24%) this was not felt to be appropriate. There was rarely any record of the outcome of the autopsy in the clinical notes. In only four cases (12%) was there any evidence that there had been a case discussion or child death review to assess the clinical information around the child's death. In many occurrences this may have been due to the case being reported to the CHR-UK team shortly after the child's death but before such reviews had taken place. However, the lack of recording in the child's notes made it difficult for a judgement to be made.

### 6.5.2 Support for the family

*Immediate support offered by hospital staff, discussed possibility of post-mortem examination, provided written information, and started a memory box which nursing staff took to the home in the period after the death. Appointment offered for a consultant review six weeks after death, GP, local CDC team and the child's consultant were informed. The tertiary hospital and children's social care were also informed. (Case assessor comment)*

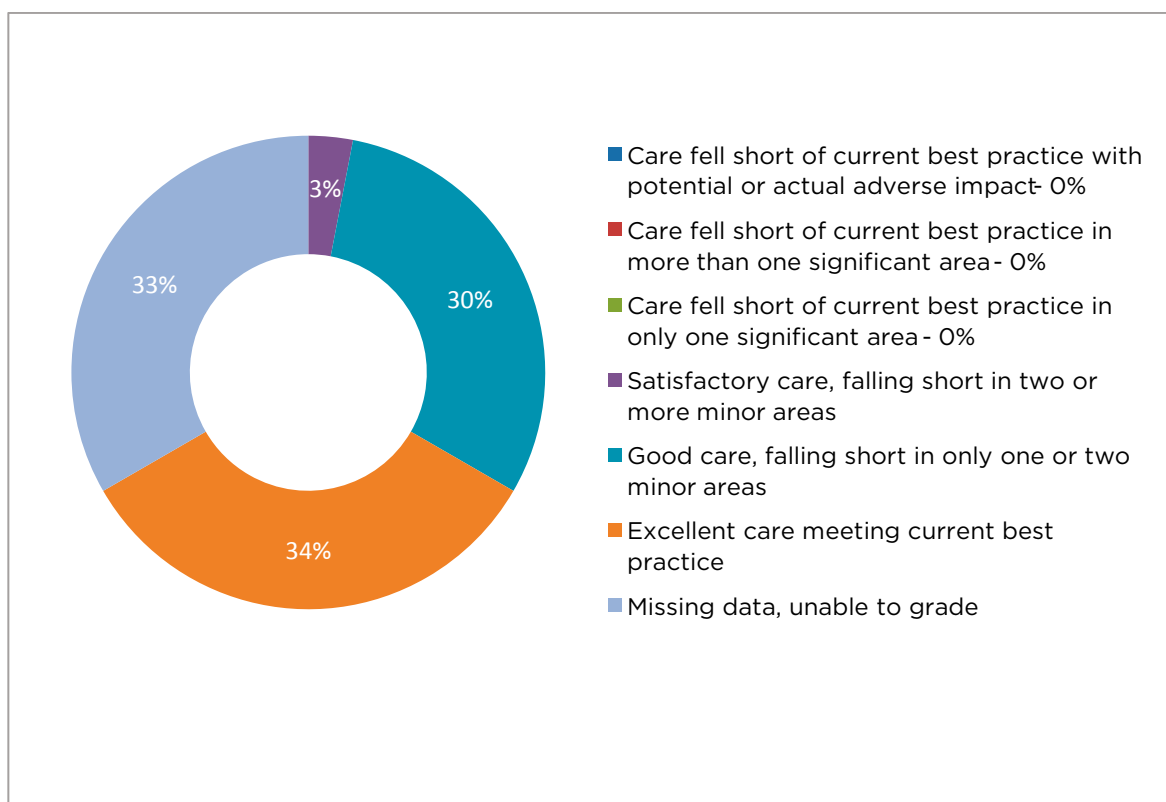
In 19 cases (58%) there was evidence in the notes that the family had been offered support from healthcare professionals. The case assessors frequently commented on the quality of support offered, and there were examples of letters sent to the family extending condolences and offering to meet with them. There was one case where a bereavement plan had been documented as part of a multidisciplinary case review. In a few cases, either a paediatrician or other members of the palliative care team had visited the family at home following the child's death. In other cases, there was no documentation of what support had been provided to the family.

## 6.6 Quality of care around the child's death

For many children the quality of record keeping was extremely poor following the child's death. Case assessors commented that the case records often came to an abrupt halt with documentation of signs of life being extinct and the acronym 'RIP'. Rarely was there any documentation of what happened subsequently, including any steps taken around certifying the death. This also included referring to the coroner or procurator fiscal, arrangements for an autopsy where appropriate, support for the family, or any review of the child's death.

In 22 (67%) cases the assessors judged the overall quality of care in relation to initial management of the child to be excellent and meeting current best practice, or falling short of current best practice in only minor areas (100% of those for whom they were able to make a judgement) (Figure 6.1). In 11 cases (33%) the case assessors were unable to rate the quality of care for the family in this phase.

**Figure 6.1: Quality of care around the child's death as judged by the case assessors**



An assessment of the quality of care, based on the criterion-based review showed that in no cases did the care fall short of the defined standards in all areas; there were seven (21%) cases where care received met the standards in all areas; and 19 (58%) where care fell short of defined standards in at least some areas. Table 6.8 shows the percentage of cases for which there was evidence that the key standards in relation to quality of care were met.

**Table 6.5: Standards around child's death**

	<b>Standard</b>	<b>Number (%) with evidence that this standard had been met</b>
1	If the death was unexpected, was there evidence that an appropriate multi-agency 'rapid response' was initiated?	6/11 (55%)
2	If the death was unexpected and the cause of death was not clear at the time, was the death referred to the coroner/procurator fiscal?	10/14 (71%)
3	If the child had a recognised life-limiting condition, had the prognosis been discussed appropriate with the family?	17/18 (94%)
4	If the child had a recognised life-limiting condition, had an end of life plan been agreed with the family?	12/18 (67%)
5	Were the parents offered support from relevant healthcare professionals, and counselling or other support?	19/33 (56%)
6	Either before or after the death, did an appropriately trained clinician take a clinical history, examine the child and arrange for appropriate investigations?	19/33 (56%)
7	If appropriate, was a report of the clinical findings provided to the coroner/procurator fiscal?	11/17 (65%)

## 6.7 Summary

The children who died in this review represent a mixed group of children with a range of co-morbidities. This is in keeping with other studies which have shown that deaths in children with epilepsies are rare in the absence of associated severe neurological disabilities.<sup>22,49,50</sup> Over half of children died of causes other than their epilepsy, typically related to a complex disability, and often with recognised life-limiting conditions. For these children and their families, the importance of advanced end of life planning has been emphasised. For many of these children, there was evidence in the review of good practice around working with the children and their families and involving them in appropriate decision making. Areas for improvement in the care of these children are highlighted through this review. It is important that all professionals acknowledge the needs of this group of children and take appropriate steps to provide for their care in partnership with their parents.

A small number of children died as a consequence of status epilepticus. The majority of these had associated developmental impairments in addition to their epilepsy. Some children had difficulty to control epilepsy with frequent ongoing seizures. For others the prolonged seizure that led to their death was unexpected. It is essential that all families, carers and healthcare professionals with children that have epilepsies are aware of the risks of prolonged seizures and are empowered to act promptly and appropriately according to national guidelines and in line with an agreed emergency care plan. Issues were identified in this review in relation to the emergency department response to prolonged seizures. All emergency providers should be able to respond in an appropriate, timely and coordinated way to a child presenting with prolonged seizures, in accordance with existing national and local guidance.

There is a small but appreciable risk of sudden unexpected death (SUDEP) in children and young people with epilepsies. This risk is greater in children with associated co-morbidities, and in those with difficult to control epilepsy<sup>51,52</sup>. All parents and carers need to be aware of SUDEP, and where particular risk factors are identified, should be advised on appropriate steps to minimise the risks.

Overall, nearly a quarter of the deaths reviewed were felt to have contributory factors identified which, by means of locally or nationally achievable interventions, can be modified to reduce the risk of future child deaths. These factors have been discussed in detail in this chapter and highlight areas for improvements in care. In particular they relate to the coordination of care in children with multiple complex needs, anticipatory planning and emergency care plans, and partnership working with families, to ensure they are empowered to recognise and respond appropriately to acute events in their child's health.

Although a number of issues were identified which could lead to improvements in care, the overall impression from this review was that good quality paediatric care is provided to these children and their families. Modifiable factors were identified in 24% of cases reviewed, in contrast to 59% of children's cases in the previous sentinel audit which were considered to be avoidable.<sup>22</sup> In the 2002 review, it was notable that a number of children had neither seen a paediatric consultant nor a paediatric neurologist. These were not issues that were encountered in the current review.

Once a child dies, there is a tendency for the case to be closed, with limited information subsequently recorded in the notes. While this may simply be an issue of recording, it raises the possibility that children's deaths may not be appropriately examined in order to establish an accurate cause of death, or to learn lessons. It may mean that families are not getting the support required following a child's death. Every death in a child with epilepsy, where it is unexpected (including all deaths related to an epileptic seizure, all SUDEP, and any deaths that may be related to epilepsy treatment) must be reported to the coroner or procurator fiscal, and should have a comprehensive multi-agency investigation, including a case review for the professionals involved to review all aspects of the child's care. The classification system developed for this review could assist clinical teams, coroners and procurators fiscal in assigning an appropriate cause of death for a child with epilepsy. It is important that information relating to the investigation of a child's death, and support provided to the family around the time of death, is recorded in the clinical records and where appropriate notified to the coroner or procurator fiscal.



In 2004 Baroness Helena Kennedy reported on sudden unexpected death in infancy and stated that 'it is every family's right to have their baby's death properly investigated'.<sup>53</sup> The same principle applies to the families of children and young people with epilepsies. These children and their families deserve appropriate care, respect and support both during life and at the time of their death. Their deaths could then lead to learning in order to prevent future children's deaths.

### 6.7.1 Key findings and recommendations

**Key finding 12:** Children with epilepsies who die do so from a variety of causes with over half of the children in this review dying of causes other than their epilepsy. This can lead to multiple professionals being involved in the child's care, allowing it to become fragmented. This further highlights the need for a single clinician taking charge of the coordination of the child's care.

**Key finding 13:** Many of the children's deaths were anticipated deaths in the context of known life-limiting illnesses. A careful review of all the clinical findings and the circumstances of death are important for accuracy in classifying and registering the death. This should include discussion with the coroner or procurator fiscal and a joint agency 'rapid response' when a child with epilepsy dies unexpectedly, including those cases that appear to meet criteria for SUDEP and those as a result of status epilepticus. This supports the approach being taken by English Child Death Overview Panels, and the all-Wales Child Death Review Programme to reviewing all children's deaths and of a rapid response for unexpected deaths.

#### **Recommendation 8**

Child Death Overview Panels in England and the All-Wales Child Death Review Programme should ensure that the case of each child with epilepsy who dies is subject to a child death review, including, where appropriate, a multi-agency rapid response to investigate the death and provide support to the family. NHS Scotland, HSC Northern Ireland, Public Health Jersey, Public Health Guernsey and Department of Health Isle of Man should consider how such reviews could be built into any plans for development of child death review in their devolved nations.

**Key finding 14:** For many of the children with known life-limiting conditions there was evidence in this review of good supportive and anticipatory planning for the children and their families, in keeping with guidelines from the Association for Children's Palliative Care/Together for Short Lives (ACT). This should be a standard adhered to for all such children.

**Key finding 15:** Overall, in eight of the deaths reviewed the case assessors identified contributory factors which, by means of locally or nationally achievable interventions, could be modified to reduce the risk of future child deaths. This emphasised the value of a thorough approach to reviewing each child's death in order to identify lessons at a local and wider level as highlighted in as highlighted in **key finding 13** and **Recommendation 8** (Chapter 6).

**Key finding 16:** When a child with epilepsy or other neuro-developmental impairments dies, he or she does not cease to be a part of their family. It is important that clinicians recognise this and ensure that the family receive appropriate support, advice and information.

**Recommendation 9**

The consultant responsible for the care of any child with epilepsy who dies should ensure that all subsequent actions after death, including registration of the death, referrals to the coroner or procurator fiscal, and follow up of the family together with child death review are documented in the child's notes and shared with other members of the clinical team.

**Key finding 17:** The review findings showed that there were potentially modifiable factors leading to children's deaths in relation to the communication with parents. This highlights the need for clear information and advice to parents and carers, in a manner they can understand, on the signs indicating when a child is unwell. Furthermore the clinician responsible for the care of the child should ensure there are clear and careful discussions around the risks of seizures and SUDEP, as set out in the recommendations in the NICE guidelines. This would help empower parents and carers to recognise and respond promptly in such situations.

**Key finding 18:** This review identified some concerns with recognition of and response to status epilepticus in hospitals, as highlighted in **key finding 10** (Chapter 5). Ensuring clear channels of communication between different hospital teams are established will help to ensure that acute episodes are managed effectively.

## 7. The process of clinical outcome reviews

The RCPCH was commissioned to carry out the CHR-UK themed review of mortality and morbidity in children and young people with epilepsies. This chapter reflects on the process of the review, presenting relevant data enabling some consideration of the validity of the results, and address the fourth aim of the review:

### **What can be learnt through different approaches to reviewing cases of death or serious morbidity in children and young people with epilepsies?**

This review has built upon the work undertaken by the CMACE (formally known as CEMACH), *Why Children Die study* which identified the feasibility of using confidential enquiry methodology to reduce child deaths and make significant contributions to child health.<sup>8</sup> The report highlighted that carrying out a child health review provided the opportunity to reflect on the specialist nature of child healthcare and its provision. This led to establishing the first UK-wide themed review of children and young people's healthcare, which has uniquely incorporated both mortality and serious morbidity. The concept of serious morbidity is complex and the CHR-UK team recognised the full scope of this could not be covered within this programme. In order to fully assess this concept, long-term neurodevelopmental, health and behavioural measures would need to be considered and would have required a longitudinal study design and a wider case group to address. In consultation with the TEG, the population of children and young people receiving intensive or high dependency care for prolonged seizures was selected as one measure of serious morbidity. This was because it was achievable and clear to define.

CHR-UK examined the entire care pathway for each child who met the inclusion criteria, from the initial diagnosis of the child's epilepsy until the sentinel incident, including primary care and emergency care. This is a divergence from standard confidential enquiry programmes which tend to look at a specific incident in a child's healthcare. Reviewing the entire care pathway of each child provided an overview of the child's care, and put the sentinel incident into context.

### 7.1 Case notification process

The overall response rate for notifications in the review was 39%, rising from 32% in the first two months of the review to 42% in the final two months (Table 7.1). The response rate for each month was calculated from the number of clinicians who notified a case, the number of clinicians reporting they had not seen a case and the number of clinicians that opted out of the review, divided by the total number of emails successfully sent out that month. This was lower than that obtained from similar data capture systems such as the British Paediatric Surveillance Unit (BPSU), but in keeping with response rates for many other studies. The BPSU began its orange card surveillance over 20 years ago with an initial response rate of 73%, increasing to 91% in 2011 (54,55).

**Table 7.1: Notifications over the 10 month data collection period**

	England & Wales		England, Wales & Scotland		England, Wales, Scotland, Northern Ireland, Isle of Man & Jersey						Total
	June	July	August	Sept	Oct	Nov	Dec	Jan	Feb	March	
Total number of emails sent	2624	2591	2779	2583	2720	2603	2548	2478	2423	2343	<b>26,692</b>
Number of responders that had no case to report	811	749	892	948	1021	943	935	961	903	893	<b>9056</b>
Number of responders that reported a case(s)	31	36	37	40	36	34	49	25	47	74	<b>409</b>
Number of responders that opted out of review	25	10	159	102	106	56	48	28	32	30	<b>596</b>
<b>Response rate</b>	<b>33%</b>	<b>31%</b>	<b>39%</b>	<b>42%</b>	<b>43%</b>	<b>40%</b>	<b>41%</b>	<b>41%</b>	<b>41%</b>	<b>43%</b>	<b>39%</b>
<b>Reported cases</b>	<b>46</b>	<b>42</b>	<b>49</b>	<b>48</b>	<b>43</b>	<b>37</b>	<b>59</b>	<b>36</b>	<b>55</b>	<b>111</b>	<b>526</b>

During the entire data collection period, 2183<sup>c</sup> clinicians responded to the notification emails. Twenty-six of these responded only when they had seen a case. Just over half the notifications received during the period (226; 55%) were from general paediatric consultants. The remaining notifications were comprised of community paediatricians (38; 9%), paediatric neurologists (31; 8%), paediatric intensivists (24; 6%), and other subspecialties (36; 9%).

Surveillance systems such as BPSU have been running for long periods of time and are firmly embedded into clinicians' work practices. Keeping the CHR-UK case notification system running continuously would enable a system which clinicians can engage with on an ongoing basis. In addition, by expanding the case notification system to include other professionals, beside RCPCH clinicians (such as epilepsy specialist nurses, child death overview panels, and other specialist groups) would have allowed engagement with other professionals who could have notified cases, therefore increasing the overall notification rate. However, the time constraints during this review meant that a robust notification system could not be established within the review time frame.

<sup>c</sup> Approx. 3500 clinicians emailed each month. The exact figure changed on a month basis as new clinicians were added to the distribution list and some clinicians were subtracted depending on their willingness for their data to be held on the RCPCH database.

## 7.2 Challenges encountered collecting data in the devolved nations

CHR-UK encountered some delays in establishing the data collection processes in Scotland, Northern Ireland, the Channel Islands and the Isle of Man. In addition to approvals from the National Information Governance Board (NIGB), permissions were required from the Scottish Caldicott Guardians (SCG), and the Patient Advisory Group (PAG) in Northern Ireland. Assistance from the relevant government offices in each devolved nation was extremely helpful in achieving this. Permission was granted from the Scottish Caldicott Guardians and data collection for the programme extended to Scotland from August 2012.

In Northern Ireland the PAG provided advice on the legal position on using patient-identifiable information. Northern Ireland legislation on data protection and secondary use of patient-identifiable information meant that consent must be gained for all living patients. Therefore CHR-UK, in collaboration with the Northern Ireland Maternal and Child Health (NIMCH) office, who managed the data collection process, had to establish a protocol specifically for use in Northern Ireland. On the advice of PAG, a consent form was created and issued for all children with a diagnosis of epilepsy who received intensive or high dependency care. The lead consultant for the child's care, who was known to the family, was charged with asking parents or carers to opt in or out of the review. In most cases the child had been discharged from hospital and parents or carers were contacted at home and asked if their child's details could be included in the review. Gaining consent was a lengthy process and therefore the CHR-UK team were unable to obtain any completed questionnaires from Northern Ireland for children meeting the intensive or high dependency care criteria during the time frame of the review.

## 7.3 Clinical questionnaire completion

In total CHR-UK received 421 valid case notifications, and 173 completed questionnaires with a completion rate of 41%. To understand the reasoning behind the lower than expected questionnaire return rate, a short survey (Appendix 6) consisting of seven questions was sent out to clinicians who had notified CHR-UK of a case meeting the project criteria, but had not submitted a questionnaire. Out of the 214 clinicians who were asked to complete the survey 84 (39%) responded, and 41 (49%) reported difficulties completing the questionnaire.

The majority of clinicians reported that they had technical issues (9; 22%) or that the questionnaire was too time consuming to complete (9; 21%). Twenty-two per cent (9; 22%) were unable to recall the case. This was exacerbated by a two month gap between starting the notification system and having the on-line questionnaire available for use. Twelve per cent (5) did not complete a questionnaire due to it already being completed for the case, and 7% (3) realised that the case did not meet the inclusion criteria. From free text responses, clinicians additionally noted they found it difficult to retrieve patient notes, particularly as there were huge volumes of notes for patients with epilepsy and complex co-morbidities. This, in turn, proved an added pressure on their already demanding workloads.

## 7.4 How representative was the sample?

The 173 completed questionnaires over a 10 month period included 46 deaths (of which 32% were primarily due to epilepsy, 59% due to other causes, and 9% unclassifiable), 67 episodes of intensive care and 70 episodes of high dependency care.

During 2008-10 the average annual number of registered deaths in the UK for children and young people (1-17 years inclusive) with epilepsy recorded as the underlying cause of death was 52 (equivalent to 43 deaths in a 10 month period). In addition, there were 107 registered deaths with any mention of epilepsy (equivalent to 89 deaths in a 10 month period). For this review, notification of all deaths in children with epilepsies were requested, suggesting some under-ascertainment of cases of children who died. However, it is important to recognise that the figures may not be directly comparable. The ONS data<sup>(19)</sup> record death registrations, and are dependent on the certifying clinician/coroner/procurator fiscal listing epilepsy either as the underlying cause, or as another mention on the death certificate. While it is likely that most deaths directly attributable to epilepsy would be classified as such, there are some potential gaps. For example, there is no ICD-10 code for SUDEP; some deaths might be attributed to epilepsy when, in fact, this was not the cause (e.g. a symptomatic seizure); and other deaths may be coded according to the final terminal event (e.g. a respiratory infection), even if the precipitating cause was epilepsy. Those deaths in children with epilepsies, where the epilepsy was not the primary cause of death may or may not include epilepsy on the death certificate, thus these are likely to be an underestimate in official statistics.

The PICANet annual report 2012, indicated that 1,101 children and young people, aged 0-15 were admitted to paediatric intensive care units between January 2009 and December 2011 due to status epilepticus.<sup>56</sup> A previous analysis of PICU admissions indicated that around 24% of intensive care admissions for status epilepticus were due to underlying epilepsy.<sup>57</sup> Extrapolating from these data, it is expected that there would be approximately 73 intensive care admissions of 0-15 year olds within a 10 month period<sup>D</sup>. A significant proportion of these would have been infants aged less than a year (the PICANet report documented 26% of admissions with neurological conditions were aged less than a year). Therefore CHR-UK's figure of 67 episodes of intensive care admissions for prolonged seizures would seem to be consistent with that finding. It was not possible to ascertain the number of 16 and 17 year olds who are admitted to intensive care with prolonged seizures from epilepsy. It is likely that this review under-ascertained these cases, as not all would come under the care of paediatricians or be admitted to paediatric, as opposed to adult, intensive care units.

It is clear, therefore, that the data obtained for this review do not provide a completely accurate picture of the incidence of deaths or intensive care admissions for children with epilepsies in the UK. In particular, there would appear to be some under-ascertainment of deaths in children with epilepsies, although the figures for intensive care admissions suggest that ascertainment of these cases was more complete. The findings therefore should be interpreted with some caution, and cannot be said to be representative of the care of all children with epilepsies who die or who receive intensive or high dependency care for prolonged seizures. Nevertheless, as a themed review, the findings can give us an insight into the quality of care for children and young people with epilepsies, and can highlight issues from which all clinicians caring for such children can learn.

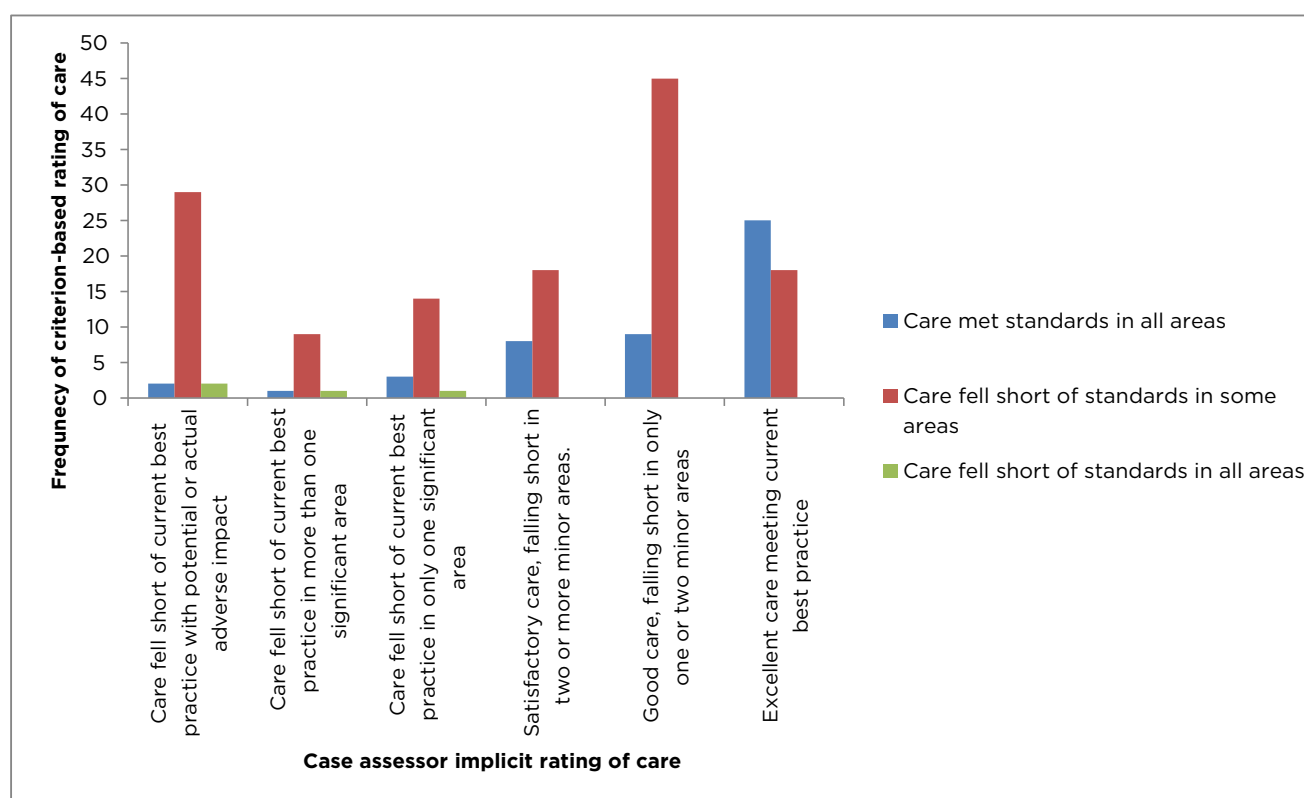
<sup>D</sup> Calculated as  $1101 * 10/36 * 24/100$

## 7.5 Comparison of the explicit and implicit methods of review

From this review, both criterion-based and structured implicit assessments were used. Historically, criterion-based assessments have been criticised for their reduced flexibility in identifying unexpected factors that influence standards of care. Similarly holistic assessments have been criticised for being reviewer-dependent, relying on the reviewer's personal knowledge and perspectives to judge the quality of care.<sup>32</sup> In view of this, a mixed approach was used in this review.

As the two methods of review measured different aspects of care, it was not possible to carry out any statistical correlation. However, for 213 ratings of the quality of care in different phases of the care pathway there were both implicit and criterion-based assessments available for comparison. In Figure 7.1, those cases which, on the criterion-based assessments, were found to be falling short of standards in all areas were all rated by case assessors as falling short of current best practice in one or more significant areas, with or without the potential for, or actual, adverse impact on the patient. Those which, on the criterion-based assessments, were found to be meeting standards in all areas, tended to be rated by the case assessors as demonstrating excellent or good quality care. However, a small number were rated as falling short of current best practice in one or more significant areas.

**Figure 7.1: Comparison of criterion-based and structured implicit evaluations of the quality of care.**



The combination of the criterion-based and implicit reviews proved valuable in enabling the CHR-UK research team and the TEG to gain a comprehensive picture of the quality of care provided for these children and their families. While the criterion-based review provided an objective assessment of the quality of care when compared to recognised standards, this depended on the information being available, and did not give any insights as to why particular standards might not have been

met, nor of the relative clinical importance of individual standards. For example, a child who was prescribed the incorrect anti-epileptic drug would be given the same grading on the criterion-based review as one who had been prescribed the correct drug but by a non-specialist clinician. Both fall short of current best standards, but one has a more direct potential for harm to the child. In contrast, the implicit review ratings were more subjective and dependent on the expertise and views of the case assessors; however, they did provide a rich source of narrative information enabling a deeper understanding of why particular standards might not have been met.

The tools themselves had some limitations, and the case assessors sometimes found it difficult to reach conclusions. For example, distinguishing between care that fell short of current best practice in one or more significant areas with potential for adverse impacts on the patient, and care that fell short of current best practice in one or more significant areas without potential for adverse impacts on the patient.

Overall, the CHR-UK team concluded combining the two approaches to review was valuable, but that some of the tools and scales used would require modifying to make them easier to use and more relevant to the topic being studied.



## 7.6 Case assessment process; hospital and RCPCH based assessments

Case assessments were carried out both in hospitals and at the RCPCH. A major criticism of confidential enquiry programmes<sup>31,58</sup> is the poor quality and readability of photocopied notes received by programmes and the effect this has on the assessor's ability to make judgements on the quality of care, thus impacting on the overall quality of the review. NCEPOD<sup>59</sup> have recognised the limitations associated with standard confidential enquiry panel reviews where decisions are based on information they are presented with and where they do not have access to other clinical notes or health professionals. CHR-UK opted to carry out some reviews in hospitals, to determine whether the quality of information available was improved and whether the case assessors were better able to carry out the review.

Eleven hospital-based case reviews were carried out. These were found to be significantly less time consuming administratively as the CHR-UK team organised case note retrieval with clinical audit departments to ensure case notes were available on the day, rather than being sent to the RCPCH for anonymisation. Once at the hospital, if the case assessors found that some notes were missing, they had the opportunity to request these and liaise directly with staff. Some hospitals were unwilling to send notes to RCPCH for assessments as the case notes were voluminous, therefore these case assessments could not have been completed without hospital-based assessments.

The hospital-based case reviews have proved particularly valuable in being able to access all relevant hospital information directly, including some information which is only stored electronically, or in different sets of case files within the same hospital. They also reduced the time spent by the CHR-UK team in preparation for case assessment days with regard to case anonymisation and copying of notes. The hospital reviews were subsequently backed up with a review of community-based records at RCPCH, which did incur additional costs with travel to the College. CHR-UK found that including records from a range of healthcare settings provided extra information which is often not available in the hospital records. This provided the opportunity to gain further insights into the child's care across the entire care pathway.

Case assessors completed a short survey reflecting on the case assessment process (Appendix 7). Of the 24 that were asked to complete the questionnaire, nine responded. All the case assessors found the case assessment tool easy to use and felt it allowed them to reflect on the cases they were reviewing. Two assessors commented that the assessment tool was too lengthy with some of the questions being too repetitive and needing further clarification. All case assessors who carried out hospital-based reviews (6/6) found these easier compared to RCPCH-based reviews, due to direct access to documents on-site. It was easier to navigate the original notes in comparison to photocopied notes and the assessors were able to gain a more comprehensive understanding of the case. The assessors reflected that the assessment would have been made easier if they had access to the other healthcare settings notes at the hospital. This was one limitation of the hospital-based assessments. The case assessors had to attend the hospital to complete the primary assessment, and then attend the RCPCH to complete the assessment on the other relevant notes (e.g. GP notes, community care notes). Future studies should aim to ensure all relevant case notes for a child are transferred to the hospital prior to the review so a thorough case assessment can take place in one sitting.

## 7.7 Inter-rater reliability

For quality assurance purposes, 11 cases were assessed by more than one pair of case assessors. In order to ascertain the degree of agreement between pairs of case assessors reviewing the same case, the intra-class correlation coefficient was calculated for each case assessor's overall rating of the phase of care (Table 7.2). For each phase of care the intra-class coefficient was found to be highly significant showing agreement between case assessor pairs reviewing the same case. Overall, from investigating completed case assessments, separate pairs of case assessors reviewing the same case were found to have similar results.

**Table 7.2: Intra-class correlations**

Phase of care*	Intra-class correlation coefficient	F value	Significance ( $p < 0.005$ )
Initial diagnosis (N=52)	0.978	89.508	<0.001
Ongoing management (N=36)	0.943	33.760	<0.001
Pre-hospital care (N=50)	0.989	179.542	<0.001
Emergency department care (N=31)	0.740	6.831	<0.001
Intensive care and high dependency care (N=23)	0.985	129.364	<0.001
Management after death (N=25)	0.867	15.170	<0.001
Overall care received by the child (N=41)	0.909	21.749	<0.001

\*All single assessment (assessments carried out by one case assessor) and missing values were removed from the analysis

## 7.8 Clinician interviews and comparator group

The review aimed to learn and incorporate aspects into the methodology which were lacking from previous confidential enquiries,<sup>31</sup> and initially proposed to include clinician-led interviews as well as a comparator group. The inclusion and practicalities of these two aspects were discussed in detail with the TEG. The TEG concluded that, the inclusion of a comparator group (case-control) was unachievable within the time frame and resources of the review. Any valid comparator group would need to include large numbers in order to make useful comparisons; it would also need to differ significantly from the sample group in order to evaluate different aspects of care.

The TEG considered the incorporation of clinician-led interviews into the review would be problematic and not feasible for this review. Hospitals already have internal reviews and it was felt clinicians may be unwilling to volunteer to undergo further reviews. Time and personnel constraints would limit the extent to which the team could carry out a valid and worthwhile review incorporating clinician-led assessments.

## 7.9 Summary

CHR-UK has built upon the work by CMACE<sup>8</sup> to develop a review of serious morbidity and mortality of children with epilepsies, and where possible answer criticisms and failings of previous confidential enquiry programmes. This has provided valuable lessons which future programmes of work can learn from:

- Establishing a robust continuously operational case notification system would enable the system to be embedded in clinicians' work practice. Incorporating nurses, specialist groups and child death overview panels would help to ensure all relevant cases are notified
- Keeping the questionnaire brief and focusing on key points, while ensuring this is rolled out at the same time as the case notification process, would help to increase participation levels
- The use of both a criterion-based and structured implicit assessment in the case review process has been shown to successfully draw out different aspects of care, helping to gain a comprehensive picture of the care of both the child and the family
- The case assessment tool has been demonstrated to be intuitive, easy to use while allowing the case assessors to reflect on the care of the child received. Future programmes can learn from this and look to reduce the length of the tool and refine scales to make them easier to use and more relevant to the topic being investigated
- Building on the success of the hospital-based reviews, future work should aim to ensure the transfer of all healthcare settings case records to the hospital for a more comprehensive case review
- Future programmes should further investigate and consider the inclusion of a comparator group and clinician-based interviews

## 8. Summary

This themed case review of mortality and serious morbidity in children and young people with epilepsies has examined the quality of care provided for children and their families at all stages of the care pathway, including primary and emergency care. Clinical questionnaire data were obtained for 173 incidents (deaths and intensive or high dependency care admissions) in 162 children from across the UK. Although steps were taken to maximise case ascertainment, comparison with national figures suggests there may have been some under-ascertainment, particularly in relation to the deaths. Nevertheless, the review was able to identify examples of both high quality care and less adequate care, and highlight a number of key learning points from these.

The children included in this review may not necessarily be representative of all children and young people living with epilepsies, the majority of whom are likely to live healthy independent lives with their epilepsy well controlled. It is not clear whether this represents a bias for children notified in this review. All of the children in this review were included either because they had died or because they had experienced a prolonged seizure requiring intensive or high dependency care. Thus they represent the more complex end of the spectrum of children with epilepsies. The children in this review tended to present with epilepsy at a very young age. There was a high proportion from ethnic minorities and from deprived socio-economic areas. One particularly notable finding was the extremely high proportion of children who had associated developmental impairments or other co-morbidities. Such children may represent a particular subgroup of children with epilepsies who have an increased risk of prolonged seizures or mortality. However, further epidemiological studies would be needed to confirm this hypothesis.

A detailed case notes review was undertaken on 69 incidents in 61 children. Case assessments were undertaken on 33 children who died, along with 17 children receiving intensive care and 19 receiving high dependency care for prolonged seizures, of whom eight subsequently died.

The review identified reasonable standards of care around the initial diagnosis and management of epilepsy in these children. There was evidence that clinicians with expertise in epilepsy were involved in most cases from an early stage. In addition, there was evidence that the approach to diagnosis had taken account of seizure type, aetiology of the epilepsy and any associated co-morbidities. In most cases, there was evidence that appropriate anti-epileptic drugs were being prescribed according to standard guidelines, together with appropriate follow up of children and of the involvement of relevant specialists in their care.

There was a lack of evidence in this review around the provision of information to parents and carers and across the wider clinical team. This was reflected in the absence of individual treatment plans in a number of cases and poor documentation of discussions with parents and carers. The findings of the review emphasise the importance of clinical teams working in partnership with children and families to improve their overall care.

The complex nature of these children's epilepsies and co-morbidities gave rise to the potential for fragmentation of care. This was particularly prominent in those children requiring hospital admissions for prolonged seizures. This could result in a focus on individual acute episodes in isolation. This

emphasises the importance of overall coordination of care for these children and the relevance of considering the implications of specific episodes within the context of the overall care. These findings support the NICE and SIGN guidelines around having a named clinician with overall responsibility for the child's care. They also support the value of epilepsy nurse specialists in coordination of care, but emphasise that, where they are involved, there must be good communication and documentation of their input.

Two further systemic changes were identified which could help to improve the coordination of care for these children, together with communication with families and between different members of the clinical team. The first systematic change is that the use of a parent or carer-held 'epilepsy passport' could facilitate good documentation of management, including emergency care plans and ensuring that all those looking after the child are kept informed and up to date. Similar health passports have been used in the care of people with diabetes,<sup>60</sup> and this could be developed as a standard of care.

The second systematic change is that the use of peer review by clinical teams could allow opportunities for reviewing the care provided to children, monitoring and improving standards of care. Such systems have proved to be helpful in the child protection field and are now considered an important component of good clinical care.<sup>61</sup>

## 8.1 Management of prolonged seizures

The children who required intensive or high dependency care for prolonged seizures were often children with difficult to manage epilepsies. This was not, however, universal. Serious prolonged seizures can present unexpectedly in children with otherwise well-controlled epilepsies. It is important that all those caring for children with epilepsies are aware of the risks of prolonged seizures, while knowing how to recognise and respond promptly to such seizures. The review identified some concerns around the care received prior to arrival at hospital. This was considered by the case assessors to often reflect a lack of forward planning and absent or unclear emergency care plans.

One specific finding in a number of cases was evidence of delayed or inadequate emergency treatment for seizures by ambulance crews. This led to a specific recommendation to ambulance trusts around the use of buccal midazolam as a first line drug for the management of prolonged seizures.

There was good evidence in this review of high standards of care in emergency departments and intensive or high dependency care. Those cases where there was evidence that standards had not been met emphasised the importance of clinical staff being thoroughly trained and supported in following standard guidelines, together with documenting carefully when decisions are made to deviate from those guidelines.

## 8.2 Management around and after death

As found with those children presenting with prolonged seizures, the children who died had a high rate of co-morbidities. Many of the children died as a consequence of their co-morbidities, but a substantial proportion died following status epilepticus or possible SUDEP. While the majority of those dying as a direct consequence of their epilepsy had significant developmental impairments, or difficult to control epilepsies, this was not universal. Both events can occur in children with seemingly well-controlled epilepsy and all parents and carers need to be aware of the risks of SUDEP and of the risks associated with prolonged seizures. This is particularly important where additional risk factors are identified in children.

There was some evidence of good advance care planning for children with known life limiting conditions, but this was not universal and could be improved in line with national standards. The review highlighted the lack of documentation of events and care following a child's death. Such documentation should form part of the clinical record. It is important in relation to support for the parents, understanding of the causes and circumstances of the child's death, and for learning lessons for future prevention.

Almost a quarter of all deaths in this review were felt to have contributory factors which could be modified to reduce the risks of further deaths. These particularly related to coordination of care, anticipatory planning and emergency care plans, together with empowering parents and carers to recognise and respond appropriately to acute events.

## 8.3 Conclusion

Epilepsies are an important childhood condition that impact on both acute hospital-based care and ongoing care in the community. This is highlighted in the introduction. This review found an overall positive picture of good clinical care provided by clinical teams working in partnership with families. However, such care is not universal, and lessons can be learnt and improvements can be made. This report aims for all clinical teams caring for children and young people with epilepsies to learn from the findings of this review. The specific recommendations made can lead to more systemic improvements in the care provided to these children and their families. Many of the lessons learnt are not specific to epilepsies, and can be applied to a wide range of acute and chronic childhood conditions. Therefore, individual practitioners should all be striving to provide the best possible care for children, reflecting on their own clinical practice and be committed to learning lessons when things do not go to plan. Those responsible for commissioning and managing services need to consider how their services can be improved in ways that support clinicians in providing care and will, ultimately, lead to better outcomes for children and families.

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## Glossary and abbreviations

### General Terms

<b>British National Formulary for Children (BNFC)</b>	Provides authoritative and practical information on the selection and clinical use of medicines.
<b>British Paediatric Surveillance Unit (BPSU)</b>	A unit run by the Royal College of Paediatrics and Child Health which enables doctors and researchers to find out how many children in the UK and Republic of Ireland are affected by particular diseases or conditions each year.
<b>CMACE (formally known as CEMACH)</b>	Centre for Maternal and Child Enquiries
<b>Child/children</b>	For the purposes of the review, “child” or “children” encompasses both children and young people.
<b>Child Death Overview Panel (CDOP)</b>	CDOPs are in place in every local authority area in England. When a child dies the CDOP will collect and analyse information about each death to identify any wider public health or safety concerns or any matters of concern affecting the safety and welfare of children in the area of authority. The CDOP has the responsibility of reviewing the deaths of all children, with priority given to those deaths that are unexpected and unexplained.
<b>Child Death Review Programme</b>	A programme in Wales, similar to that of the CDOPs, which aims to identify and describe patterns and causes of child death and recommend actions to reduce risk of avoidable factors contributing to child deaths.

<b>Clinical Outcome Review Programme (CORP)</b>	<p>The clinical outcome review programmes which now encompass Confidential Enquiries are designed to help assess the quality of healthcare, and stimulate improvement in safety and effectiveness by systematically enabling clinicians, managers and policy makers to learn from adverse events and other relevant data. The programme aims to complement and contribute to the work of other agencies such as NICE; CQC, the Royal Colleges and academic research studies with the aim of supporting changes that can help improve the quality and safety of healthcare delivery. This programme of work is run by the Health Quality Improvement Partnership.</p>
<b>HQIP (Healthcare Quality improvement partnership)</b>	<p>HQIP is contracted by the NHS England to deliver outcome focused quality improvement programmes structured around collection of clinical data, including clinical audits, registers and confidential enquiries.</p>
<b>IAG (Independent Advisory Group)</b>	<p>The Independent Advisory Group were appointed by HQIP in conjunction with the Medical Directors for the four nations following nomination via the Academy of Royal Colleges, UK Health Departments and Patient Voices. Their role is to provide overall governance to the programmes including final selection of programme topics.</p>
<b>ILAE (International League Against Epilepsy)</b>	<p>The ILAE is a global, professional and non-profit international organisation and a non-governmental organisation with an official relationship with the WHO (World Health Organisation). The ILAE's objectives are: to advance and disseminate knowledge about epilepsy (having developed guidelines for the classification of epilepsy and the design of investigative trials); to promote research, education and training; and to improve overall patient care.</p>
<b>Incident</b>	<p>For the purposes of the review the "incident" refers to the sentinel incident; either the child's death or the child's intensive or high dependency care admission following a prolonged seizure.</p>

<b>NICE</b>	National Institute for Health and Clinical Excellence (NICE) sets standards for quality healthcare and produces guidance on medicines, treatment and procedures.
<b>SIGN</b>	Scottish Intercollegiate Guidelines Network (SIGN) develops an evidence based clinical practice guidelines for the National Health Service (NHS) in Scotland.

**Clinical Terms**

<b>Absence seizure</b>	A seizure characterised by behavioural arrest associated with generalised spike and slow wave activity on the EEG. May be typical or atypical.
<b>Acute symptomatic seizures</b>	Seizures occurring at the time of a diagnosis of an acute disorder (e.g. meningitis, encephalitis, electrolyte disturbance) and seizures occurring within a week of traumatic head injury.
<b>Anti-epileptic drug (AED)</b>	Medication taken to prevent the recurrence of epileptic seizures.
<b>Atonic seizure</b>	A generalised seizure characterised by sudden onset of loss of muscle tone often resulting in a fall or 'drop'.
<b>Benign epilepsy with centrotemporal spikes (BECTS)</b>	An epilepsy syndrome that usually occurs between 4 and 11 years of age, with the epilepsy entering a spontaneous remission by the age of 14/15. The syndrome is characterised by focal motor seizures, typically involving the mouth and face that usually occur during sleep, in an otherwise normal individual and a characteristic EEG pattern.
<b>Childhood-onset absence epilepsy (CAE)</b>	An epilepsy syndrome with an onset age of 4-9 years, characterised by frequent (daily) absence seizures associated with 3Hz spike and slow wave activity on EEG.

<b>Convulsive status epilepticus</b>	When a convulsive (tonic-clonic or clonic) seizure continues for a prolonged period (longer than 5 minutes), or when convulsive seizures occur one after the other with no recovery between. Convulsive status epilepticus is an emergency and requires immediate medical attention.
<b>Dravet syndrome</b>	Previously known as severe myoclonic epilepsy of infancy. An epilepsy syndrome with onset in epilepsy, characterised by initial prolonged, typically lateralised, febrile seizures, subsequent development of multiple seizure types including myoclonic, absence, focal and generalised tonic-clonic seizures, with developmental plateau or regression. This epilepsy syndrome has a poor prognosis.
<b>Electrocardiogram (ECG)</b>	A test that records the heart's electrical activity
<b>Electroencephalography (EEG)</b>	An investigation that involves recording of the electrical activity of the brain. Electrodes are attached to the standardised points on the individual's head with collodion or with a rubber cap. Recordings are usually taken across two points for the role of EEG in diagnosis of epilepsy and epilepsy syndrome.
<b>Epilepsy</b>	A chronic neurological condition characterised by two or more epileptic seizures (ILAE). A pragmatic definition for epilepsy in this review is "two or more epileptic seizures more than 24 hours apart that are not acute symptomatic seizures or febrile seizures" to aid consistent application of these criteria. 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
<b>Epileptic seizure</b>	A transient occurrence of clinically-manifest signs and/or symptoms, resulting from a primary change to the electrical activity (abnormally excessive or synchronous) in the brain.

<b>Epilepsy syndrome</b>	A complex of clinical features, including seizure type(s), age at onset of the seizure(s), developmental status and EEG findings that collectively define a distinctive, recognizable type of epilepsy.
<b>Febrile seizure</b>	A seizure occurring in childhood between 6 months and five years of age that is associated with a febrile illness not caused by an infection of the central nervous system, without previous neonatal seizures or a previous unprovoked seizure, and not meeting the criteria for other acute symptomatic seizures.
<b>Focal seizure</b>	A seizure that originates within networks limited to one hemisphere, discretely localised or more widely distributed. Replaces the terms 'partial' seizure and 'localisation-related' seizure.
<b>Generalised seizure</b>	A seizure that originates in, and rapidly engages, bilaterally distributed neuronal networks. Such bilateral networks can include cortical and subcortical structures but do not necessarily include the entire cortex.
<b>Generalised tonic-clonic seizure</b>	A seizure of sudden onset involving generalised stiffening and subsequent rhythmic jerking of the limbs, the result of rapid widespread engagement of bilateral cortical and subcortical networks in the brain.
<b>High dependency care</b>	For the purposes of this review, any child or young person who has a prolonged seizure and requires 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.
<b>Idiopathic</b>	Used to describe a condition with no known cause.



<b>Infantile spasm</b>	A specific seizure type presenting in the first year of life, most commonly between 4 and 9 months. Spasms are brief axial movements lasting 0.2-2 seconds, most commonly flexor in nature, involving flexion of the trunk with extension of the upper and lower limbs. They are occasionally referred to as 'salaam seizures'.
<b>Intensive care</b>	For the purposes of this review, any child or young person who has a prolonged seizures and requires admittance to an intensive care ward, or is receiving this level of care.
<b>Intravenous</b>	Administration of medication into a vein.
<b>Juvenile absence epilepsy</b>	An epilepsy syndrome with an age of onset of 9-13 years characterised by absence seizures, associated with 3-4 Hz spike wave on EEG. Generalised tonic-clonic seizures may also occur.
<b>Juvenile myoclonic epilepsy (JME)</b>	An epilepsy syndrome with an age of onset of 5-20+years (peak, 10-16), characterised by myoclonic seizures which most commonly occur soon after waking. Generalised tonic-clonic and absence seizures may occur in between 60 and 40% of people retrospectively. EEG demonstrates 3-6Hz generalised polyspike and wave activity, with photosensitivity in >40%.
<b>Ketogenic Diet</b>	A specific diet that is high in fats but low in carbohydrates and protein. It is used to treat a number of paediatric epilepsy syndromes
<b>Landau-Kleffner syndrome</b>	A very rare epilepsy syndrome with an age of onset of 3-6 years characterised by loss of language (after a period of normal language development) associated with an epilepsy of centrotemporal origin, more specifically bitemporal spikes on EEG with enhancement in sleep or continuous spike wave activity in slow sleep (CSWS).

<b>Late-onset childhood occipital epilepsy (Gastaut type)</b>	Epilepsy with an age of onset in mid-childhood to adolescence with frequent brief seizures characterised by initial visual hallucinations, ictal blindness, vomiting and post-ictal headache. EEG typically shows interictal occipital spikes attenuated by eye opening.
<b>Lennox-Gastaut syndrome</b>	A rare epilepsy syndrome with an age of onset of 2-10 years (peak 4-7) characterised by multiple seizure types (including atonic, tonic, tonic-clonic, atypical absence and focal seizures), cognitive impairment and specific EEG features of diffuse slow spike and wave (<2Hz) as well as paroxysmal fast activity (10 Hz or more) during tonic seizures which frequently occur in sleep.
<b>Magnetic resonance imaging (MRI)</b>	A medical imaging technique used to visualise detailed internal structures. The imaging modality of choice in patients with epilepsy.
<b>Monotherapy</b>	Use of a single drug in treatment.
<b>Myoclonic seizure</b>	Sudden brief (<100ms) and almost shock-like involuntary single or multiple jerks due to abnormal excessive or synchronous neuronal activity and associated with polyspikes on EEG.
<b>Ohtahara syndrome</b>	A rare epilepsy syndrome usually presenting in babies in their first 10 days of life, and characterised by tonic spasms and focal seizures and a markedly abnormal EEG, usually a burst-suppression pattern.
<b>Panayiotopoulos syndrome</b>	Epilepsy syndrome presenting in early childhood (mean 3-7 yrs) with infrequent seizures which may be prolonged lasting over 30 minutes. Characterised by autonomic features including vomiting, pallor, and sweating followed by tonic eye deviation, impairment of consciousness with possible evolution into secondary generalisation. Prognosis is excellent and treatment with an anti-epileptic medication may not be necessary.

<b>Polytherapy</b>	Two or more medications used in combination therapy.
<b>Post-ictal state</b>	A period of confusion, tiredness or memory loss after a seizure. This can last for minutes, hours or, in rare cases, days.
<b>Prolonged seizure</b>	Any tonic-clonic seizure lasting longer than five minutes, or serial, repeated seizures continuing over a period of more than thirty minutes.
<b>Secondary generalised seizures</b>	Focal seizures that spread to become generalised.
<b>Spasms</b>	A sudden, involuntary contraction of a muscle or group of muscles. These can be epileptic or non-epileptic.
<b>Sudden unexpected death in epilepsy (SUDEP)</b>	Sudden, unexplained, witnessed or unwitnessed, nontraumatic and nondrowning death in individuals with epilepsy, with or without evidence for a seizure, and excluding documented status epilepticus, in which post-mortem examination does not reveal a toxicological or anatomic cause for death. <i>Provided by Nashef L. Sudden unexpected death in epilepsy: Terminology and definitions. Epilepsia 1997;38:S20-S22.</i>
<b>Symptomatic</b>	Exhibiting symptoms of a particular disease but having a different cause
<b>Tonic seizure</b>	An epileptic seizure characterised by abrupt generalised muscle stiffening which often results in a fall or drop. The seizure usually lasts less than a minute and recovery is rapid. The EEG usually shows fast spike activity (at least 10-12 spikes per second) during the seizure.

<b>Tonic-clonic seizure</b>	An epileptic seizure characterised by initial generalised muscle stiffening (tonic phase), followed by rhythmical jerking of the limbs (clonic phase), usually lasting a few minutes. The person may bite their tongue and may be incontinent. They may feel confused, complain of a severe headache and be sleepy afterwards, and take a while to recover fully.
<b>West syndrome</b>	A rare epilepsy syndrome occurring from birth to 13 months of age (peak age, 4-9 months), characterised by epileptic spasms and an EEG pattern called hypsarrhythmia. There are many causes of West syndrome. The long-term prognosis depends predominantly on the underlying cause.

**Miscellaneous acronyms**

<b>SPR</b>	Specialist Registrar
<b>OPD</b>	Outpatient Paediatric Department

## Appendix 1: Topic selection and TEG

This topic was chosen by the HQIP Independent Advisory Group (IAG) from a shortlist compiled by the RCPCH following a process of stakeholder consultation and a structured review of published literature on child mortality. A transparent approach to topic nomination and selection was developed by the RCPCH aligning with the requirements set out in the HQIP Protocol for topic selection for the Clinical Outcomes Review Programmes. A total of 62 topics were suggested during the stakeholder consultation, from which a shortlist of 18 potential topics was drawn up. This shortlist was reached by removing duplicates, those that did not involve a healthcare setting and those not fitting the criteria. The remaining 18 were assessed using a criteria based proformas provided by HQIP and ranked by the CHR-UK team. Clinical leads then used this proforma to reach the final four topics. These topics were identified as high priority topics which met the criteria for review and for which a review using this framework would be achievable. The IAG then selected epilepsy as the topic, to include serious morbidity as well as mortality while emphasising that the entire patient pathway is reviewed.

The RCPCH convened the TEG to support the CHR-UK team in developing the data collection tools and defining the standards that formed the basis of the review. The TEG included a broad range of relevant clinical and methodological expertise and several members had experience in quality improvement and methods of case review.

The TEG Expertise was made up as follows:

- Hospital and community based paediatricians at career grade and higher specialty trainee levels
- Tertiary paediatric neurologists with specific expertise in childhood epilepsy
- Paediatricians with particular expertise in childhood epilepsy, neurodisability, emergency care, and intensive care
- Paediatric epilepsy specialist nurses
- A General Practitioner with expertise in case review and clinical improvement
- Two parents of children and young people with epilepsy
- A representative from each devolved nation

The CHR-UK team and TEG recognised that many of the deaths in children and young people with epilepsy would be from causes other than their epilepsy, and that there was potential for misdiagnosis and inappropriate classification of deaths in epilepsy. In order to explore these issues, ensure that no relevant cases were excluded, and to look at broader aspects of care for these children, the scope of the review was expanded to include all deaths in children and young people with epilepsy, rather than just seizure-related deaths.

The concept of serious morbidity is complex, and it was recognised that the full scope of this could not be covered within the timeframe of this programme. For epilepsy, children and young people receiving intensive or high dependency care for prolonged seizures was used as a measure of serious morbidity, which was considered achievable and clear to define. This enabled the consideration of the extent to which any change in the child's condition following an intensive or high dependency care admission could be identified and the ability of this review to inform other studies on appropriate measures of longer term morbidity.

## Appendix 2: Clinical questionnaire

### Themed case review of epilepsy care

#### Instructions

**Please complete all questions to the best of your ability. We recognise that users may not be able to answer all questions, and ask that you complete the sections and questions on which you hold information. Please circle or tick your answer as appropriate. If you do not have information for any particular item, please respond 'Not known' as this indicates to the Child Health Reviews-UK team that you have considered the question but are not able to answer.**

Date of completion of questionnaire:

Project identifier number:

#### Section A: Inclusion criteria

1. Did the child/young person receive intensive care or high dependency care following a prolonged seizure?
2. Did the child/young person die (of any cause)?
3. Was the child/young person aged between their 1st and 18th birthday at the time of the incident?
4. Prior to the incident had the child/young person had two or more epileptic seizures more than 24 hours apart that were not acute symptomatic seizures or febrile seizures?
5. Please select the country that you are in: (Northern Ireland, England, Scotland, Wales, Channel Islands, Isle of Man)

#### Section B: Case details

6. Child young person's name
7. Gender
8. Date of birth
9. Ethnic group
10. NHS number
11. CHI number
12. Postcode of usual residence
13. Hospital presented to
14. Hospital where the child/young person received intensive care or high dependency care (if different from above)
15. Date of incident
16. Date of admission to intensive care or high dependency care
17. Date of death (if applicable)
18. Date of discharge from intensive care or high dependency care (if applicable):
19. Date of discharge from hospital (if applicable)

20. Please list other health professionals involved in the child/young person's care: E.g. GP (practice and location), Intensive care consultant (role and hospital/trust)
21. Has this incident been reviewed, or is it planned to be reviewed through any other inquiry/audit process? E.g. Serious Untoward Incident Investigation

### Section C: Background factors

22. Were there any known neonatal conditions (e.g. low birth weight, prematurity, neonatal encephalopathy)?
23. Was there any known developmental impairment or disability at the time of incident?
24. Apart from the epilepsy, were there any known medical conditions at the time of incident?
25. Was the child/young person on any medication at the time of incident?
26. Who was directly looking after the child/young person immediately prior to the incident? E.g. parents
27. Were there any concerns raised about family engagement with services?
28. Were there any concerns raised with adherence to treatment?
29. Factors in relation to the child, their environment or the care received: Please provide a description of any relevant factors within the domestic and social environment, or factors intrinsic to the child, that may have contributed to the incident or its management

### Section D: Pre-incident care

30. What was the date of the child/young person's first epileptic seizure?
31. What was the date the child/young person's epilepsy was diagnosed?
32. Which clinician was responsible for confirming the diagnosis of epilepsy? E.g. consultant paediatric neurologist
33. What seizure types did the child/young person experience? (Please refer to the seizure table and write the codes for any seizures the child/young person experienced)
34. Did the child/young person have an identified epilepsy syndrome? (Please refer to the epilepsy syndrome table and write the codes for any syndrome the child/young person experienced)
35. Was there an identified cause of the child/young person's epilepsy? E.g. cerebral malformation
36. Which health professionals were involved in the ongoing care of the child? E.g. consultant paediatric neurologist
37. Which health professional was primarily responsible for the management of the child/young person's epilepsy? E.g. consultant paediatrician with expertise in epilepsy
38. Prior to this incident, had the child/young person had any other intensive care or high dependency care admissions with prolonged seizures?
39. Prior to this incident, had the child/young person had any other intensive care or high dependency care admissions with prolonged seizures? (If yes, please provide the date of the most recent admission)
40. What was the date of the last accident and emergency attendance?
41. In the 12 months prior to this incident, how many times had the child/young person been admitted to hospital with prolonged seizures?
42. What was the date of the last hospital admission?
43. In the 6 months prior to this incident, how frequently was the child/young person having tonic-clonic seizures? E.g. daily

44. In the 6 months prior to this incident, how frequently was the child/young person having tonic-clonic seizures? E.g. at least weekly
45. In the 6 months prior to this incident had there been any changes to medication? (If yes, please specify)
46. In the 6 months prior to this incident had there been any other changes to management? (If yes, please specify)
47. In the 12 months prior to the incident, how many scheduled reviews were there of the child/young person's epilepsy? E.g. not known
48. Prior to the incident, what was the most recent contact between the child/young person or their family and any health professional regarding the epilepsy? (Include any contact, including unscheduled attendances or telephone consultations) E.g. scheduled outpatient clinic review
49. What was the date of that contact?
50. In the 6 months prior to this incident were there any appointments that were missed or cancelled without good cause? (Do not include missed/cancelled appointments for which an appropriate reason was given, for example, the child/young person was an inpatient at the time)
51. Did the child/young person have any documented care plans? (tick all that apply) E.g. emergency care plan for management of prolonged seizures, **not** including rescue medication
52. Were there any identified unmet needs/gaps in services (please specify)
53. Were there any problems around the initial diagnosis or ongoing management which may have affected the outcome or which you feel are relevant for CHR-UK team to consider

#### Section E: Pre-hospital care

54. Where was the child/young person at the time of the incident? E.g. acute hospital(if ticked please select an answer in the column to the right, and go to question 64), Emergency Department
55. If the child/young person had an emergency care plan, was this followed?
56. Was any emergency treatment given prior to the arrival of an ambulance crew?(tick all that apply) E.g. buccal Midazolam
57. What management was provided by the ambulance crew? (tick all that apply) E.g. ventilatory support (intubation)
58. What was the child/young person's condition on arrival at the emergency department? E.g. buccal Midazolam
59. What management did the emergency department initiate? (tick all that apply) E.g. thiopentone
60. Was a departmental protocol for the management of status epilepticus/prolonged seizures followed? E.g. Yes - departmental protocol followed
61. What was the outcome of the emergency department management? E.g. discharged home
62. Were there any problems around the pre-hospital or emergency department management which may have affected on the outcome or which you feel are relevant for the CHR-UK team to consider?

#### Section E: First line hospital management

63. What was the child/young person's condition on arrival? E.g. fully alert, no longer seizing
64. What emergency management was undertaken? (tick all that apply)E.g. buccal midazolam



65. Was a departmental protocol for the management of status epilepticus/prolonged seizures followed? E.g. yes - departmental protocol followed
66. What was the outcome of the first line management? E.g. discharged home
67. Were there any problems around the first line hospital management which may have affected the outcome or which you feel are relevant for the CHR-UK team to consider?

#### **Section F: Intensive care and high dependency management**

68. What was the primary reason for this child/young person receiving high dependency or intensive care? E.g. prolonged seizure
69. What was the child/young person's condition on admission to intensive care or high dependency care? E.g. fully alert, no longer seizing
70. What seizure management did the child/young person receive in intensive care or high dependency care? E.g. thiopentone
71. What was the length of ventilation the child/young person received in intensive care or high dependency care? (Hours, Not Ventilated, Not known)
72. What was the outcome for the child/young person on discharge from intensive care or high dependency care? E.g. fully recovered to pre-admission state
73. Were there any problems around the first line hospital management which may have affected the outcome or which you feel are relevant for the CHR-UK team to consider?

#### **Section G Management following the death**

74. Has a medical certificate of the cause of death been issued?
75. Registered cause of death (if known) (Ia, Ib, Ic, II or not done)
76. What is your understanding of the cause of death?
77. Was this death referred to the coroner/procurator fiscal?
78. Was a post-mortem examination carried out?
79. Where was the child/young person when the death was confirmed E.g. acute Hospital
80. Was an interagency rapid response initiated to respond to the child/young person's death?
81. Was an interagency rapid response initiated to respond to the child/young person's death?

#### **Section H: Further issues**

Are there any issues not already covered in any of the previous sections which you feel are important for the CHR-UK team to consider in relation to this child's admission or death? Please include any action or learning you consider should be taken as a result of the child's admission or death, issues that you feel require a more in-depth evaluation, or problems that you feel should be considered as part of this review of clinical outcomes.

## Reference document for questionnaire

### Question 33: What seizures types did the child/young person experience?

Code	Seizure type	Code	Seizure type
S1	Absence	S14	Massive bilateral myoclonus
S2	Atonic	S15	Myoclonic absence seizures
S3	Clonic	S16	Myoclonic atonic seizures
S4	Eyelid myoclonic	S17	Myoclonic seizures
S5	Focal motor seizures	S18	Negative myoclonus
S6	Focal sensory seizures	S19	Occipital seizures
S7	Focal seizures	S20	Parietal seizures
S8	Frontal seizures	S21	Reflex seizures
S9	Gelastic seizures	S22	Secondarily generalized seizures
S10	(Generalised) tonic-clonic seizures	S23	Spasms
S11	Hemiclonic seizures	S24	Temporal seizures
S12	Infantile seizures	S25	Unclassified seizures
S13	Massive bilaterial myoclonic	S26	Not known

**Question 34: Did the child/young person have an identified epilepsy syndrome?**

Code	Syndrome	Code	Syndrome
ES1	No identified syndrome	ES26	Hemiconvulsion-hemiplegia syndrome
ES2	Not known	ES27	Hot water epilepsy
ES3	Autosomal-dominant nocturnal frontal lobe epilepsy (ADNFLE)	ES28	Idiopathic focal epilepsy of childhood
ES4	Autosomal dominant partial epilepsy with auditory features	ES29	Idiopathic photosensitive occipital lobe epilepsy
ES5	Bathing epilepsy	ES30	Juvenile absence epilepsy (JAE)
ES6	Benign familial neonatal seizures	ES31	Juvenile myoclonic epilepsy (JME)
ES7	Benign focal epilepsy of childhood	ES32	Landau-Kleffner syndrome
ES8	Benign infantile seizures	ES33	Late onset childhood occipital epilepsy (Gastaut type) (idiopathic childhood occipital epilepsy)- Lennox-Gastaut syndrome
ES9	Benign neonatal seizures	ES34	Migrating partial (focal) seizures of infancy
ES10	Benign non-familial neonatal seizures {fifth day fits	ES35	Myoclonic encephalopathy in nonprogressive disorders {myoclonic status in nonprogressive encephalopathies}
ES11	Childhood absence epilepsy(CAE)	ES36	(Benign) myoclonic epilepsy in infancy
ES12	(Benign) childhood epilepsy with centrotemporal spikes [BECTS] also known as benign rolandic epilepsy)	ES37	Occipital lobe epilepsy
ES13	Childhood epilepsy with occipital paroxysms	ES38	Ohtahara syndrome
ES14	Dravet syndrome (severe myoclonic epilepsy of infancy or SMEI)	ES39	Panayiotopoulos syndrome (early onset (benign) childhood occipital epilepsy)
ES15	Electroclinical syndrome	ES40	Parietal lobe epilepsy
ES16	Early myoclonic encephalopathy	ES41	Perioral myoclonia with absences
ES17	Epilepsy with generalized tonic-clonic seizures only (epilepsy with generalised tonic-clonic seizures on awakening)	ES42	Phantom absences

ES18	Epilepsy with myoclonic absences	ES43	Primary reading epilepsy
ES19	Epilepsy with myoclonic astatic seizures (Doose syndrome: myoclonic astatic epilepsy)	ES44	Progressive myoclonus (myoclonic) epilepsies (PME)
ES20	Eyelid myoclonia with absences	ES45	Rasmussen's syndrome(chronic progressive epilepsy partialis continua also known as Kozhevnikov syndrome)
ES21	Familial focal epilepsy with variable foci	ES46	Reflex epilepsies - Startle epilepsy
ES22	Familial temporal lobe epilepsy	ES47	Temporal lobe epilepsy
ES23	Frontal lobe epilepsy		Visual sensitive epilepsies
ES24	Generalized epilepsy with febrile seizures plus (FS+)	ES48	West syndrome - Unclassified
ES25	Gelastic seizures due to hypothalamic hamartoma	ES49	

## Appendix 3: CHR-UK Case assessment tool

Seizure-related deaths and serious morbidity, and sudden unexpected death in children and young people with epilepsy

### Instructions for case assessors

The questionnaire has been colour coded: the blue sections should be completed for all cases; yellow sections for deaths only; green for intensive/high dependency care only. The criterion based assessment requires the assessors to document specific case information and to answer, to the best of their ability based on the information in the case records, key criterion based questions. These key questions are highlighted in bold. Where it is not possible to answer these questions on the basis of the information in the notes, the 'unclear' response should be used.

For more guidance on how to complete the assessment tool please refer to the case assessor handbook.

*Please complete the below*

<b>Case number</b>	
<b>Case assessor A</b>	
<b>Case assessor B</b>	

### A1. Initial diagnosis

The case assessors should review any records relating to the initial presentation and management, including any hospital admissions with initial seizures, and subsequent outpatient follow up. Discharge summaries, clinic letters, and clinical records may need to be reviewed.

1. How old was the child/young person at the time of the first epileptic seizure?
2. How old was the child/young person when epilepsy was diagnosed?
3. Which clinician was responsible for confirming the diagnosis of epilepsy? E.g. consultant paediatric neurologist
- 3a. Was the diagnosis established by a paediatric neurologist or specialist paediatrician with training and expertise in epilepsy?
4. What seizure types did the child/young person experience? (please tick all that apply) E.g. absence seizures
- 4a. Was there evidence seizure type was considered in establishing the diagnosis?
5. Did the child/young person have an identified epilepsy syndrome? E.g. Dravet Syndrome
- 5a. Was there evidence that an epilepsy syndrome was considered in establishing the diagnosis?
6. Did the child/young person have an identified cause for their epilepsy? E.g. cerebral malformation
- 6a. Was there evidence that aetiology was considered in the diagnostic process?
7. Were there any known neonatal conditions (e.g. low birth weight, prematurity, neonatal encephalopathy)? If yes, please provide details.
8. Was there any known developmental impairment or disability at the time of diagnosis? If yes, please provide details.

9. Apart from the epilepsy, were there any other known medical conditions at the time of diagnosis? If yes, please provide details.
- 9a. Was there evidence that co-morbidities were considered in the diagnostic process?
10. Who initiated AED treatment? E.g. consultant paediatric neurologist?
- 10a. Was AED treatment initiated by a paediatric neurologist or specialist paediatrician with training and expertise in epilepsy?
11. What AED treatment was the child started on? (Name, dose, schedule)
- 11a. Were appropriate anti-epileptic drugs administered according to the seizure type, epilepsy syndrome, co-medication and co-morbidity?
12. Was there evidence that the child and family were given information about their diagnosis and prognosis within 6 months of the diagnosis?

### **A1. Initial diagnosis: structured implicit review**

Please comment on the care received by the patient during this phase.

From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase:

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

### **A2. Ongoing management**

This section will primarily be based on the most recent scheduled review. The case assessors will need to supplement this with data from case records (hospital and community) and possibly from patient administration systems for the local hospital.

1. Prior to the incident, what was the most recent contact between the child/family and any health professional regarding the epilepsy? (include any contact, including unscheduled attendances or telephone consultations) E.g. scheduled outpatient clinic review. What was the date of that contact?
2. Prior to the incident, what was the most recent scheduled review of the child/young person's epilepsy? E.g. consultant paediatric neurologist. What was the date of that contact?
3. Which health professionals were involved in the ongoing care of the child?
  - 3a. Was there evidence in the records that the child and family had access to a named individual to contact for advice and support?
  - 3b. Did the child and family have access to an epilepsy specialist nurse?
  - 3c. Was a named clinician responsible for the ongoing management of the child/young person's epilepsy?

4. Reviewing the records from diagnosis to the most recent scheduled review, were any of the following criteria present?
  - the child was aged under 2 years at diagnosis
  - there was a unilateral structural lesion
  - the epilepsy was not controlled with medication within 2 years of diagnosis
  - management was unsuccessful after two drugs
  - the child experienced unacceptable side effects from medication
  - there was clearly identified psychological and/or psychiatric co-morbidity
  - there was diagnostic doubt as to the nature of the seizures and/or seizure syndrome
  - there was identified behavioural or developmental regression
- 4a. If yes, had the child/young person been referred to a tertiary specialist?
5. Prior to this incident, had the child/young person had any other intensive care or high dependency care admissions with prolonged seizures? If yes, date of last intensive care/high dependency care admission
6. In the 12 months prior to this incident, how many times had the child/young person attended an accident and emergency department with prolonged seizures? What was the date of the last accident and emergency attendance?
7. In the 12 months prior to this incident, how many times had the child/young person been admitted to hospital with prolonged seizures? What was the date of the last hospital admission?
8. In the 6 months prior to this incident, how frequently was the child/young person having tonic-clonic seizures? E.g. daily.
9. In the 6 months prior to this incident, how frequently was the child/young person having any other type of seizures? E.g. daily.
10. In the 6 months prior to this incident were there any appointments that were missed or cancelled without good cause? (do not count missed/cancelled appointments for which an appropriate reason was given, e.g. child was an inpatient at the time).
11. Were there any identified difficulties in family engagement with services?
12. Were there any identified issues with adherence with treatment?
13. In the 12 months prior to this incident how often was the child/young person's epilepsy reviewed? (all scheduled reviews by any means; not including unscheduled reviews). E.g. once a month or more frequent.
- 13a. Was the child's epilepsy reviewed at least annually (for uncomplicated epilepsy) or more frequently (for epilepsy that was difficult to control, or where other factors such as co-morbidities, compliance indicated)?
14. In the 6 months prior to this incident had there been any changes to medication?
15. In the 6 months prior to this incident had there been any other changes to management?
16. At the last scheduled review what was the child's weight? If not recorded, what was the child's most recent recorded weight and when was that from? (weight in kg, date).
17. At the last scheduled review, what medication was the child being prescribed? (If more than 1 type of medication being prescribed then please separate name and dosages using '/e.g. clonazepam/ leviracetam/ topiramate) (Name, dose, schedule).
- 17a. Were appropriate antiepileptic drugs administered according to the seizure type, epilepsy syndrome, co-medication and co-morbidity?
- 17b. For each AED, was the child/young person on an appropriate dose for his/her age and weight?
18. In the last 6 months, had the child/young person had any anti-epileptic drugs withdrawn?

- 18a. If yes, was the treatment withdrawn slowly over at least 2-3 months with a clear plan, and under the guidance of a specialist?
19. Did the child/young person have any documented care plans? (tick all that apply) E.g. emergency care plan for management of prolonged seizures, including rescue medication.
- 19a. Was there evidence in the clinical records that the child/young person had an appropriate individual care plan/treatment pathway?
20. Was there evidence in the clinical records that the following had been discussed, either in the most recent scheduled review, or at a previous point?
  - Treatment plan
  - Management of seizures
  - Effectiveness
  - Side effects of medication
  - Concordance and adherence
  - Information about support
  - Academic progress
  - Risks and hazards including SUDEP
21. Had a risk assessment been undertaken?
22. Had the young person been involved in discussions about medication and lifestyle issues?

## **A2. Ongoing management: structured implicit review**

Please comment on the care received by the patient during this phase.

From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase:

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.



## B1. Pre-hospital care

This section will examine the sentinel incident for any incidents occurring in the community, and the management of that incident prior to arrival in hospital, including lay responses, and ambulance responses. The case assessors will need a copy of the ambulance call sheet and emergency department records. Case assessors should refer to appendix F of the NICE guidelines: Guidelines for treating convulsive status epilepticus in children (published in 2011).

If the incident occurred in hospital, rather than in the community, then the assessors should continue to section B3.

1. Where was the child/young person at the time of the incident? E.g. acute hospital, Foster home. If Acute hospital is ticked, go to section B3.
2. Who was present with the child at the time of the incident? E.g. parent
- 2a. Was an appropriate trained person available to administer first aid and emergency treatment?
3. Did the child/young person have an emergency care plan for management of prolonged seizures?
4. What emergency treatment was given prior to the arrival of an ambulance crew? (Tick all that apply) Doses, timings.
- 4a. If the child/young person had an emergency care plan, was this followed?
5. What time did the incident start or was the child discovered?
6. What time was the ambulance called?
7. What time did the ambulance arrive?
- 7a. Was an ambulance called at an appropriate time?
8. What management was given by the ambulance crew? (Tick all that apply)
- 8a. Did the first responder/ambulance crew take appropriate steps to assess the situation, secure the airway/breathing/circulation, and administer appropriate emergency treatment, taking account of any treatment already given?
- 8b. Was buccal midazolam or other benzodiazepines given in an appropriate dose?

### B1. Pre-hospital care: structured implicit review

Please comment on the care received by the patient during this phase.

From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase:

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

## **B2. Emergency department management**

This section will examine the management of the sentinel incident in the emergency department. The case assessors will need a copy of the emergency department records.

Case assessors should refer to appendix F of the NICE guidelines: Guidelines for treating convulsive status epilepticus in children (published in 2011)

1. What time did the ambulance arrive at the emergency department?
2. What was the child/young person's condition on arrival at the emergency department? E.g. fully alert, no ongoing seizing
3. What were the most senior grade and specialty of doctor/health professional on hand to manage the seizure? E.g. consultant, Anaesthetics
4. At what time did an anaesthetist/intensivist arrive?
5. What management did the emergency department initiate? (tick all that apply) E.g. buccal midazolam, thiopentone
- 5a. On arrival in the emergency department, were appropriate steps taken to assess and secure the airway, breathing and circulation?
- 5b. Was appropriate medication given, taking account of treatment already given before arrival?
- 5c. Was appropriate expertise (including an anaesthetist/intensivist) sought in a timely manner?
6. What was the outcome of the emergency department management? E.g. discharged home, Admitted to Intensive care

## **B2. Emergency department management: structured implicit review**

Please comment on the care received by the patient during this phase.

From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase.

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

### B3. First line hospital management

If the incident occurred in hospital, rather than in the community, then the assessors will complete these questions below instead of section B1 and B2 above. The assessor will need to review the hospital records. Case assessors should refer to appendix F of the NICE guidelines: Guidelines for treating convulsive status epilepticus in children (published in 2011)

1. Where was the child/young person at the time of the incident? E.g. emergency Department, General paediatric ward
2. What time did the incident start or was the child discovered?
3. What was the child/young person's condition on arrival of the first health staff? E.g. fully alert, no longer seizing
4. What were the most senior grade and specialty of doctor/health professional on hand to manage the seizure? E.g. consultant, Anaesthetics
5. At what time did an anaesthetist/intensivist arrive?
6. What management did the hospital staff initiate? (tick all that apply) E.g. buccal midazolam, rectal diazepam
  - 6a. Were appropriate steps taken to assess and secure the airway, breathing and circulation?
  - 6b. Was appropriate medication given?
  - 6c. Was appropriate expertise (including an anaesthetist/intensivist) sought in a timely manner?
7. What was the outcome of the first line hospital management? E.g. discharged home, admitted to Intensive care

### B3. First line hospital management: structured implicit review

Please comment on the care received by the patient during this phase.  
From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase.

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

### C. Intensive care and high dependency management

To be completed for all cases receiving intensive or high dependency care. The assessors will need to review the intensive care or other inpatient notes. Case assessors should refer to appendix F of the NICE guidelines: Guidelines for treating convulsive status epilepticus in children (published in 2011)

1. What was the child/young person's condition on admission to intensive or high dependency care? E.g. post-ictal with normal level of consciousness, Intubated and ventilated, seizures controlled.
2. What seizure management did the child/young person receive on the intensive care or high dependency care unit? (tick all that apply). E.g. thiopentone boluses, thiopentone infusion
- 2a. If the child had ongoing seizure activity, was appropriate treatment with intravenous midazolam or thiopental sodium given?
3. If the child was ventilated, for how long was this continued?
4. What other medication was given?
5. What neurophysiological monitoring was used? E.g. isolated EEG, continuous EEG/cerebral function monitor
- 5a. Was adequate monitoring of the child in place throughout the intensive care high dependency care stay?
6. Was appropriate tertiary expertise consulted?
7. What was the outcome for the child/young person on discharge from intensive care or high dependency care?

### C. Intensive care and high dependency management: structured implicit review

Please comment on the care received by the patient during this phase.  
From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase.

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

## D. Management after death

This section looks at the response to a child's death and support for the family. Although there are limited standards within NICE epilepsy guidelines, there are other standards that can be drawn from the child death review process.

1. Was the mode of death? E.g. unwitnessed, found dead, witnessed death, no response to resuscitation (dead on arrival)
2. If appropriate, was there documentation of discussion with the parents or carers about withdrawal of care?
3. If applicable, was organ/tissue donation discussed with the parents or carers?
4. Was the death unexpected?
5. Which professional were involved in responding to the child's death? E.g. paediatrician, emergency department physician
- 5a. If the death was unexpected, was there evidence that an appropriate multi-agency 'rapid response' was initiated?
- 5b. If the death was unexpected and the cause of death was not clear at the time, was the death referred to the coroner/procurator fiscal?
6. Did the child have a recognized life-limiting condition?
- 6a. If the child had a recognized life-limiting condition, had the prognosis been discussed appropriate with the family?
- 6b. If the child had a recognized life-limiting condition, had an end of life plan been agreed with the family?
7. Having reviewed the case records, what is your judgment of the cause of death? (1a,1b,1c) (see separate notes on classification of death for options) E.g. SUDEP, Severe aspiration or airway obstruction secondary to a seizure
8. Having reviewed the case records, can you identify any factors in any of the following domains which may have contributed to the death? E.g. factors intrinsic to the child, Factors in the family and environment#
9. Having reviewed the records, cause of death and any identified contributory factors, in your opinion was the death preventable according to the DfE definition? E.g. yes- modifiable factors identified, No modifiable factors identified or Insufficient information
10. If the death is considered preventable, please list the modifiable factors identified
11. Was a medical certificate of the cause of death issued?
12. If so, what was the cause of death as listed on the MCCD?
13. If the death was referred to the coroner/procurator fiscal, what was their verdict on the cause of death?
14. Did the MCCD or coroner's/procurator fiscal's verdict accurately reflect the cause of death?
15. What support was offered to the parents and family?
- 15a. Were the parents offered support from relevant healthcare professionals, and counselling or other support?
16. Either before or after the death, did an appropriately trained clinician take a clinical history, examine the child and arrange for appropriate investigations?
17. Was a report of the clinical findings provided to the coroner/procurator fiscal?
18. If appropriate, was an autopsy carried out by an appropriate pathologist?

19. Was a final case discussion convened within 4 months of the death to review all the clinical information?
20. If held, were the results of the final case discussion reported to the coroner/procurator fiscal and, where appropriate (England & Wales) the CDOP?
21. For deaths in England & Wales, was the death reviewed by the CDOP within 6 months?

#### **D. Management after death: structured implicit review**

Please comment on the care received by the patient during this phase.  
From the records, was there anything in particular worth noting?

Please rate the care received by the patient during this phase.

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

#### **E. Overall care**

Please comment on the overall care received by the patient and family.

Please identify any factors in relation to the child, the family (including parental care) or the environment that may have impacted on the care received or the outcome for the child and family.

From the records, was there anything in relation to the overall care pathway that has not previously been mentioned which may have impacted on the care received or the outcome for the child and family?

Please rate the care received by the patient during this phase.

1. Care fell short of current best practice in one or more significant areas resulting in the potential for, or actual, adverse impact on the patient.
2. Care fell short of current best practice in more than one significant area, but is not considered to have the potential for adverse impact on the patient.
3. Care fell short of current best practice in only one significant area, but is not considered to have the potential for adverse impact on the patient.
4. This was satisfactory care, falling short of current best practice in two or more minor areas.
5. This was good care, which fell short of current best practice in only one or two minor areas.
6. This was excellent care and met current best practice.

## **F. Quality of records**

What records were available to you for this review (please list)

Were there any gaps in your assessment because the relevant records were not available? (please describe)

We are interested in your view about the quality of the patient records in enabling good quality care to be provided. E.g. the patient record contains gaps in three or more significant areas, the patient record contains gaps in two significant areas.

## Appendix 4: Classification of death in children and young people with epilepsy

Having reviewed the case notes, the assessors should assign a cause(s) of death using the system below, incorporating an overall classification and ICD10 coding following the format used in death certification. The causes assigned should be based on the appropriate ICD 10 codes and specifically described. The full list of ICD codes can be found at: <http://apps.who.int/classifications/icd10/browse/2010/en> (accessed on 07 August 2013)

	Classification	ICD 10 coding	Notes
A	Death from a cause unrelated to the epilepsy	Ia: relevant code for the primary cause of death  Ib/Ic: any relevant conditions leading to Ia	Where the death is from a completely unrelated cause. E.g. a child with epilepsy who develops an untreatable cancer, or is involved in a road traffic accident with no evidence that a seizure contributed. In these situations it would not be appropriate to include epilepsy in II.
B	Death from a co-morbidity associated with the epilepsy	Ia: relevant code for the primary cause of death  Ib/Ic: any relevant conditions leading to Ia  II: Specific epilepsy syndrome (G40.0 - 40.8 / F80.3) or G40.9 Epilepsy, unspecified	Use this for any child with epilepsy and a significant co-morbidity who dies as a consequence of that co-morbidity and whose death is not directly related to the epilepsy. E.g. a child with epilepsy and cerebral palsy who dies following aspiration, with no evidence that the aspiration was related to a seizure.
C	Death as a consequence of treatment given for epilepsy	Ia relevant code for primary cause of death (R09.2 or other cause as applicable)  Ib Y46-Y48 or other drugs (Y40-59) as appropriate; misadventures to patients during surgical or medical care (Y60-69); medical devices associated with adverse incidents in diagnostic and therapeutic use (Y70-Y82) surgical and medical procedures as the cause of abnormal reaction of the patient (Y83-84)  Ic G40 Epilepsy, or where applicable, Specific epilepsy syndrome (G40.0 - 40.8 / F80.3)  II: Any co-morbidity or other disease condition which contributed to the death	Use this category where the death was clearly caused by the treatment given for the epilepsy, including treatment given for management of status epilepticus, where there was a clear temporal link between the treatment and the death. Include deaths resulting from reactions to or consequences of treatment given appropriately (e.g. overwhelming infection in a child on high dose steroids, recognised side effects of medication), and deaths where treatment was given inappropriately (e.g. excessive doses of benzodiazepines). Include deaths resulting from non-pharmacological treatments, e.g. neurosurgery, dietary treatments.



D	<p>Trauma associated with seizure</p> <p>1. Head injury</p> <p>2. Drowning</p> <p>3. Burns</p> <p>4. Foreign body inhalation or aspiration</p> <p>5. Other</p>	<p>Ia: appropriate code for the pathological cause of death, e.g. S00-09 Injuries to the head; T17 Foreign body in respiratory tract; T20-T32 burns and corrosions; T71 asphyxiation</p> <p>Ib: Appropriate code from W65-74 accidental drowning or submersion; X00-09 Exposure to smoke, fire and flames; or relevant other code from V01-X59 Accidents</p> <p>Ic: G40 Epilepsy, or where applicable, Specific epilepsy syndrome (G40.0 - 40.8 / F80.3)</p> <p>II: Any co-morbidity or other disease condition which contributed to the death</p>	<p>Use this category where it is clear that the child died of trauma that was related to a seizure.</p> <p>Trauma without any evidence of a seizure would be classified in category A</p> <p>It is important that if a seizure did actually cause the head injury which then caused the intracranial insult (haemorrhage, extra- or subdural haematoma) which caused the child's death, this must be captured. So 1a) would be the intracranial insult, 1b) would be the head injury and 1c) would be epilepsy (and obviously specified if possible).</p> <p>Include severe aspiration or airway obstruction secondary to a seizure - use this where there is clear clinical or pathological evidence of severe aspiration or airway obstruction sufficient to cause death. Do not use for minor degrees of aspiration or pulmonary oedema noted at autopsy.</p>
E	<p>Death secondary to status epilepticus</p>	<p>Ia: R09.2 Respiratory arrest/ cardiorespiratory failure or other relevant code for the primary cause of death</p> <p>Ib: G41.0 Grandmal status epilepticus, or other subclassification of G41 Status epilepticus as appropriate</p> <p>Ic: G40 Epilepsy, or where applicable, Specific epilepsy syndrome (G40.0 - 40.8 / F80.3)</p> <p>II: Y46-48 Drugs, medicaments and biological substances causing adverse effects in therapeutic use - Y46 Anti-epileptics; Y47 Sedatives, hypnotics and anti-anxiety drugs; Y48 Anaesthetics and therapeutic gases</p> <p>Any co-morbidity or other disease condition which contributed to the death</p>	<p>Only use this if there is documented evidence of convulsive status epilepticus (continuous, convulsive seizures lasting at least 30 minutes* or two or more seizures during which the patient does not return to baseline consciousness)</p> <p>If the death is a result of a cardiorespiratory arrest during status epilepticus, code as R09.2; if from some other complication of status epilepticus (e.g. secondary infection acquired on ITU)</p> <p>Where treatment has been given for management of the seizures that could potentially have a respiratory depressant effect, give details of that treatment in II.</p> <p>If there is clear evidence that the cardiorespiratory arrest was a direct consequence of treatment given, use category D (e.g. cardiorespiratory arrest immediately after giving benzodiazepines; benzodiazepines given in excessive doses prior to the cardiorespiratory arrest)</p>

F	<p>SUDEP</p> <p>4. Definite SUDEP</p> <p>1a. Definite SUDEP Plus</p> <p>5. Probable SUDEP</p> <p>2a. Probable SUDEP Plus</p> <p>6. Possible SUDEP</p>	<p>Ia: R96 Other sudden death, cause unknown</p> <p>Ib: G40 Epilepsy, or where applicable, Specific epilepsy syndrome (G40.0 - 40.8 / F80.3)</p> <p>II: Any co-morbidity or other disease condition which contributed to the death</p>	<p>Use this classification only if the criteria for SUDEP are met and the death does not fit into any of the other categories.</p> <p>In addition to the ICD coding, the assessors should document the degree of certainty and whether or not a seizure was witnessed:</p> <p>1. Definite SUDEP: Sudden, unexpected, witnessed or unwitnessed, nontraumatic and nondrowning death, occurring in benign circumstances, in an individual with epilepsy, with or without evidence for a seizure and excluding documented status epilepticus (seizure duration <math>\geq 30</math> min or seizures without recovery in between), in which postmortem examination does not reveal a cause of death</p> <p>1a. Definite SUDEP Plus: Satisfying the definition of Definite SUDEP, if a concomitant condition other than epilepsy is identified before or after death, if the death may have been due to the combined effect of both conditions, and if autopsy or direct observations/recordings of terminal event did not prove the concomitant condition to be the cause of death. In such cases, include the co-morbidity or other disease condition in II</p> <p>2. Probable SUDEP/Probable SUDEP Plus: Same as Definite SUDEP but without autopsy. The victim should have died unexpectedly while in a reasonable state of health, during normal activities, and in benign circumstances, without a known structural cause of death</p> <p>3. Possible SUDEP: A competing cause of death is present but not proven (e.g. found dead in water, but no pathological confirmation of drowning.)</p> <p>For each, confirm whether:</p> <ul style="list-style-type: none"> <li>With witnessed seizure</li> <li>Without witnessed seizure</li> </ul>
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**Death certification**

The Medical Certificate of the Cause of Death (MCCD) requires the medical practitioner certifying the death to specify the cause of death as follows, with the condition thought to be the ‘underlying cause of death’ appearing in the lowest completed line of part I.

Ia Disease or condition directly leading to death (this does not mean the mode of dying, such as heart failure, asphyxia etc. it means the disease, injury or complication which caused death)

Ib Other disease or conditions (if any) leading to Ia

Ic Other disease or conditions (if any) leading to Ib

II Other significant conditions CONTRIBUTING TO THE DEATH but not related to the disease or condition causing it.

NB. In Scotland, there is provision for four diseases/conditions in part I.

In addition to listing the various causes, the certifier also has to specify the approximate interval between onset and death for each category used.

The death certificate should reflect the sequence of events/conditions leading directly to the death. The initiating condition on the lowest line of part I is the **underlying cause of death**. This is defined as 'a) the disease or injury which initiated the train of morbid events leading directly to the death, or b) the circumstances of the accident or violence which produced the fatal injury.'

Where the death is deemed due to the epilepsy (SUDEP, status or consequences thereof) or its treatment, then epilepsy, or the specific epilepsy syndrome should form the bottom line of part I

Where the epilepsy is secondary to another health condition, this should be included with the epilepsy on the bottom line (e.g. 'G40.3 Generalized idiopathic epilepsy and epileptic syndromes secondary to S06.5 traumatic subdural haemorrhage in infancy')

### Sudden Unexpected Death in Epilepsy

SUDEP has been defined as the sudden, unexpected, witnessed or unwitnessed, non-traumatic, and nondrowning death in patients with epilepsy, with or without evidence for a seizure, with exclusion of documented status epilepticus, and when post-mortem examination does not reveal a structural or toxicological cause for death.(Nashef, 1997)

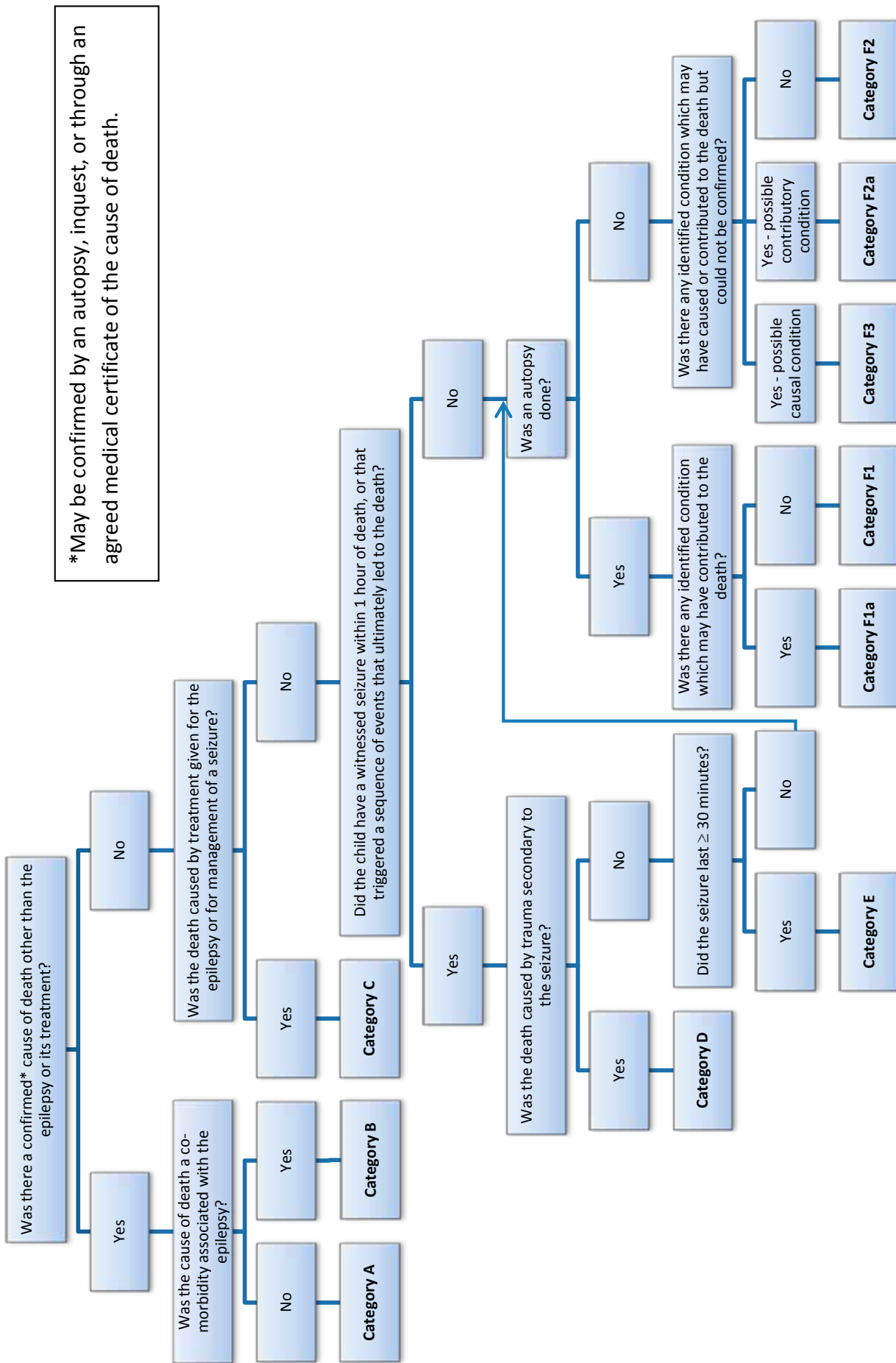
Accepted practice is to classify all such deaths when there has been an autopsy "definite SUDEP" and those in which there has been no autopsy as "probable SUDEP". This is a broad definition that encompasses heterogeneous cases. Depending on the purpose (eg, for studies on mechanisms of SUDEP), separation of SUDEP cases that occur in seizures and those (much rarer) cases that occur without a seizure might be worthwhile, because the pathophysiology in these groups is probably quite different. Furthermore, in clinical practice, there are many cases that, because information is scarce or because there are plausible explanations for death, are sometimes considered as "possible SUDEP".(Shorvon and Tomson, 2011)

An updated classification has been proposed by Nashef (Nashef et al., 2012). We will use this unified classification system.

### References

- ONS (2010) Guidance for doctors completing medical certificates of cause of death in England and Wales. [http://www.gro.gov.uk/images/medcert\\_July\\_2010.pdf](http://www.gro.gov.uk/images/medcert_July_2010.pdf)
- Scottish Government (2009) Guidance on completion of medical certificates of the cause of death. [http://www.sehd.scot.nhs.uk/cmo/CMO\(2009\)10.pdf](http://www.sehd.scot.nhs.uk/cmo/CMO(2009)10.pdf)

**Flow chart for classifying death in a child with epilepsy**



## Appendix 5: Child Health Reviews-UK: Service description questionnaire for secondary service

Audit unit:

Audit lead:

Epilepsy12 census date:

Please check this questionnaire for accuracy, making any amendments as necessary and sign in the box provided below.

Signature:	Date:
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- I understand that in signing this form I am confirming that the information provided is correct and refers to all paediatric services contained within the Epilepsy12 'audit unit' as defined within the 'audit unit' profile.*

<b>Question</b>	<b>Response (given in Epilepsy12 audit)</b>	<b>Response (given in Epilepsy12 audit)</b>	<b>Notes</b>
1. How many whole time equivalent (WTE) general paediatric consultants (community or hospital based) are there employed within the 'audit unit'?			Audit Unit - The audit unit is defined by your audit unit profile. Most audit units will include one or more secondary tier paediatric services grouped together using pragmatic boundaries agreed by the paediatric audit unit lead, the project team and the tertiary link. WTE = whole time equivalent. E.g. One full time post is 1 WTE; Someone working 3 days a week = 0.6 WTE; 2 people both working 3 days a week = 1.2 WTE.

<p>2. How many whole time equivalent (WTE) general paediatric consultants with 'expertise in epilepsy' are there employed within the 'audit unit'?</p>			<p>Paediatrician with expertise - Paediatric consultant (or associate specialist) defined by themselves, their employer and tertiary service/network as having: training and continuing education in epilepsies AND peer review of practice AND regular audit of diagnosis (e.g. participation in Epilepsy12).</p> <p>Paediatric neurologists should not be included in your response.</p>
<p>3. How many whole time equivalent (WTE) epilepsy specialist nurses (ESNs) are there employed within the 'audit unit'?</p>			<p>ESN - A children's nurse with a defined role and specific qualification and/or training in children's epilepsies</p>
<p>4. On average, how many consultant (or associate specialist) led secondary level 'epilepsy clinics' for children or young people take place within your audit unit per week?</p>			<p>A secondary level 'epilepsy clinic' is a clinic run just for children with seizures or epilepsy that takes referrals direct from GPs or emergency department (decimal answers are allowed). An "Epilepsy Clinic" is defined as a paediatric clinic where all children and young people attending have epilepsy or possible epileptic seizures.</p>
<p>5. Do any of the paediatric services within the 'audit unit' maintain a database or register of children with epilepsies?</p>	<p>Yes for all children/ Yes for some children/No</p>		
<p>6. Which of the following investigations can be obtained at a location within the 'audit unit'?</p>			
<ul style="list-style-type: none"> <li>• 12 lead ECG</li> </ul>	<p>Yes/No/Uncertain</p>		
<ul style="list-style-type: none"> <li>• 'awake' MRI</li> </ul>	<p>Yes/No/Uncertain</p>		

• MRI with sedation	Yes/No/Uncertain		
• MRI with general anaesthetic	Yes/No/Uncertain		
• Routine EEG	Yes/No/Uncertain		
• Sleep-deprived EEG	Yes/No/Uncertain		
• Melatonin induced EEG	Yes/No/Uncertain		
• Sedated EEG	Yes/No/Uncertain		
• 24-48h ambulatory EEG	Yes/No/Uncertain		
• Video telemetry	Yes/No/Uncertain		
• Portable EEG on paediatric ward within audit unit	Yes/No/Uncertain		
7. Does the 'audit unit' host paediatric neurology clinics? (e.g. a paediatric neurologist visits a site within the audit unit or is based within that 'audit unit')	Yes/No		
8. Which of the following 'transition services' are available within the 'audit unit'?			Handover Clinic - A clinic where a young person 'leaves the paediatric service and joins an adult service' and comprises both adult and paediatric health professionals
• A specific clinic for 'young people' or 'teenagers' with epilepsies	Yes/No/Uncertain		
• a 'Handover clinic'	Yes/No/Uncertain		
• Other defined handover or referral process	Yes/No/Uncertain		
• Local adult specialist epilepsy nurse	Yes/No/Uncertain		
• Youth worker	Yes/No/Uncertain		

<ul style="list-style-type: none"> <li>From what age do 'outpatient' adult services within your audit unit begin to accept referrals from General Practitioners (GPs) for young people with a seizure or seizures?</li> </ul>	<p>Number</p>		
<p>9. How many whole time equivalent (WTE) general paediatric consultants (community or hospital based) are there employed within the 'audit unit'?</p>			<p>Audit Unit - The audit unit is defined by your audit unit profile. Most audit units will include one or more secondary tier paediatric services grouped together using pragmatic boundaries agreed by the paediatric audit unit lead, the project team and the tertiary link. WTE = whole time equivalent. E.g. One full time post is 1 WTE; Someone working 3 days a week = 0.6 WTE; 2 people both working 3 days a week = 1.2 WTE.</p>
<p>10. How many whole time equivalent (WTE) general paediatric consultants with 'expertise in epilepsy' are there employed within the 'audit unit'?</p>			<p>Paediatrician with expertise -Paediatric consultant (or associate specialist) defined by themselves, their employer and tertiary service/network as having: training and continuing education in epilepsies AND peer review of practice AND regular audit of diagnosis (e.g. participation in Epilepsy12).</p> <p>Paediatric neurologists should not be included in your response.</p>



11. How many whole time equivalent (WTE) epilepsy specialist nurses (ESNs) are there employed within the 'audit unit'?			ESN - A children's nurse with a defined role and specific qualification and/or training in children's epilepsies
12. On average, how many consultant (or associate specialist) led secondary level 'epilepsy clinics' for children or young people take place within your audit unit per week?			A secondary level 'epilepsy clinic' is a clinic run just for children with seizures or epilepsy that takes referrals direct from GPs or emergency department (decimal answers are allowed). An "Epilepsy Clinic" is defined as a paediatric clinic where all the children and young people attending have epilepsy or possible epileptic seizures.
13. Which of the following investigations can be obtained at a location within the 'audit unit'?			
• 12 lead ECG	Yes/No/Uncertain		
• 'awake' MRI	Yes/No/Uncertain		
• MRI with sedation	Yes/No/Uncertain		
• MRI with general anaesthetic	Yes/No/Uncertain		
• Routine EEG	Yes/No/Uncertain		
• Sleep-deprived EEG	Yes/No/Uncertain		
• Melatonin induced EEG	Yes/No/Uncertain		
• Sedated EEG	Yes/No/Uncertain		
• 24-48h ambulatory EEG	Yes/No/Uncertain		
• Video telemetry	Yes/No/Uncertain		
• Portable EEG on paediatric ward within audit unit	Yes/No/Uncertain		
14. Does the 'audit unit' host paediatric neurology clinics? (e.g. a paediatric neurologist visits a site within the audit unit or is based within that 'audit unit')	Yes/No		

15. Which of the following 'transition services' are available within the 'audit unit'?		Handover Clinic - A clinic where a young person 'leaves the paediatric service and joins an adult service' and comprises both adult and paediatric health professionals	
• A specific clinic for 'young people' or 'teenagers' with epilepsies	Yes/No/Uncertain		
• a 'Handover clinic'	Yes/No/Uncertain		
• Other defined handover or referral process	Yes/No/Uncertain		
• Local adult specialist epilepsy nurse	Yes/No/Uncertain		
• Youth worker	Yes/No/Uncertain		
• From what age do 'outpatient' adult services within your audit unit begin to accept referrals from General Practitioners (GPs) for young people with a seizure or seizures?	Number		

## Appendix 6: Clinical questionnaire feedback

### CHR-UK Clinical Questionnaire - User Feedback

**Child Health Reviews-UK would like to understand any issues or problems you experienced when using the RCPCH web portal and completing the clinical questionnaire. Please complete the short survey below. This will take no longer than 2 minutes**

1. Please select the country you are in: E.g. England
2. What is your specialty/area of work? E.g. general paediatrician
3. Did you complete the questionnaire?
4. Did you receive a link to the questionnaire?
5. Did you register on the web portal?
6. If you answered no, can you tell us why you didn't register on the web portal? (Go to question 8 after answering this) E.g. The registration instructions were not clear
7. Why were you unable to complete the questionnaire? E.g. technical issues
8. Do you have any further comments about the clinical questionnaire or RCPCH web portal?

## Appendix 7: Feedback from case assessors

### CHR-UK Case Assessor Feedback

**Child Health Reviews-UK would like feedback relating to the case assessments that took place both at the College and at various hospital locations. Please complete the following short survey. This should take no longer than 5 minutes.**

1. Did you carry out any hospital based assessments?
2. Which case assessment did you find easier? (Please explain your reasoning)
3. If all patient notes (e.g. GP and community care notes) could be available at the hospital; would you prefer to carry out the case assessment at the hospital? (Please explain your reasoning)
4. Did you find the case assessment tool easy to use? (if no, explain your reasoning)
5. Did the case assessment tool allow you to capture all the relevant information? (if no, explain your reasoning)
6. Did the case assessment tool allow you to reflect on the learning from cases? (if no, explain your reasoning)
7. Does the CHR-UK analysis in the report capture what you felt was coming out of your case assessments? (if no, please explain your reasoning)
8. What aspects of the case assessments do you think worked well?
9. What aspects of the case assessments do you think didn't work so well?
10. If CHR-UK carried out the case assessment process again, what is the one thing you would change?

For one intensive care case, there was no information on whether or not the child had any impairment or disability