



# IMPROVING THE LIVES OF PEOPLE WITH A LEARNING DISABILITY WHO LIVE WITH EPILEPSY



## FINAL REPORT December 2013

East of England Managed Clinical Network Mental Health and  
Learning Disability

Learning Disabilities and Autism Workstream

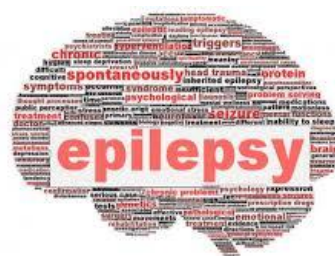


# An Easy Read Summary

Since March 2013, a small team from *Inclusion East* have worked on the East of England's Managed Clinical Network epilepsy project.



We wanted to find out what life is like for people who have learning disabilities and also have epilepsy.



We spoke to lots of people from this group who live throughout the region.



This wasn't always easy as sometimes people had to postpone meetings because they had had a seizure.



We also spoke to family carers and lots of different healthcare professionals.



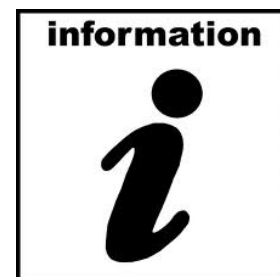
We heard that people are experiencing some of the same problems, including problems with medication and problems with having some of their seizures properly understood.



We think writing a long report that tells the stories of the people we met and the problems they are facing would be a good idea.



The team from *Inclusion East* also went to training events and looked at information that is available from epilepsy organisations.



We think people with learning disabilities who have epilepsy should be involved in producing both of these things to make things better for this group.



We have met and now know people who would be willing and able to do this beyond the end of 2013.



# CONTENTS

INTRODUCTION	1
AIMS OF THE PROJECT	1
WORK DONE	2
ACHIEVEMENTS	3
NEXT STEPS	4
APPENDICES	
1. INTERNATIONAL CLASSIFICATION OF SEIZURES	5
2. DIAGRAM OF SEIZURE TYPES	6
3. NICE GUIDELINES ON DIAGNOSIS & MANAGEMENT OF EPILEPSIES WITH REFERENCE TO PEOPLE WITH LEARNING DISABILITIES	7
4. SUDDEN UNEXPLAINED DEATH IN EPILEPSY	9
5. FURTHER READING	16
6. ACKNOWLEDGEMENTS	17



## INTRODUCTION

It is estimated that half of all people with learning disabilities have epilepsy. In those described as having a severe or profound learning disability, the number is thought to be as high as 80%. The Confidential Inquiry into Premature Deaths of People with Learning Disabilities (CIPOLD) identified epilepsy as the most common long-term health condition among those within this group.

A number of leading epilepsy organisations have acknowledged:-

- that, historically, epilepsy and the consequences of the condition were not taken as seriously in individuals with a learning disability as they were for those in the general population
- that *now*, it is only fair that this group of individuals should be able to access the same quality of epilepsy services as anyone else.

But what is the reality for the east of England's population of adults with learning disabilities who live with epilepsy on a daily basis?



## AIMS OF THE PROJECT

The primary aim of this project has been to respond to the above question in a co-productive manner:-

- by talking to relevant, interested professionals
- by talking to family carers
- and, most importantly, by talking to people with learning disabilities who, themselves, live with epilepsy.

In view of the prevalence of epilepsy among those with learning disabilities, the project also set out to:-

- find out to what extent information and resources available from leading epilepsy organisations was tailored specifically to this group of people
- and find out how relevant current training is to the specific needs of this group of people.



## WORK DONE

Since March 2013, a co-director and two associate members of *Inclusion East* – one of whom has learning disabilities and, himself, lives with epilepsy- have worked together on the East of England's Managed Clinical Network epilepsy project. We have identified, made contact and met with a range of others who have learning disabilities and epilepsy, living throughout the region. People we have met with:-

- live in Cambridgeshire, Suffolk, Norfolk and Hertfordshire
- are of both genders
- are of differing ages
- are from a variety of backgrounds and currently live in a variety of situations
- and access epilepsy services through both general neurological routes and through specialist learning disability services.

Being able to engage meaningfully with the people we met with was not always straightforward; time was needed to build relationships in which people felt comfortable sharing their personal, and sometimes painful, experiences, and meetings often had to be re-scheduled due to occurrence of seizures.

We also met with:-

- family carers
- community nurse practitioners (Peterborough City Council)
- epilepsy nurse specialists (National Hospital for Neurology and Neurosurgery)
- registrars and consultants (Hertfordshire Partnership NHS Foundation Trust)
- and academics (Universities of Cambridge and Hertfordshire).

The team from *Inclusion East* would like to take this opportunity to offer a heartfelt thank you to all of those who gave freely of their, often limited, time.

In addition to our engagement work, we also attended a number of training events and conferences – both locally and further afield – and researched a vast array of information and resources that are available from leading epilepsy organisations. Furthermore, we commissioned a final-year medical student to summarise features of both the National Institute of Clinical Excellence (NICE) and information available on sudden unexplained death in epilepsy (SUDEP) that are most pertinent to people with learning disabilities (see Appendices 3 & 4).



## ACHIEVEMENTS

From those we have engaged with, we have been able to successfully identify a number of issues that appear to be currently and commonly faced by those with learning disabilities who live with epilepsy and the families who support them:-

- uncertainty over whether there is a difference in good outcomes depending on whether epilepsy services are accessed via a general neurological route or a specialised learning disability route
- a breakdown/disjointedness in the epilepsy and learning disabilities pathways at the point of transition (childhood to adulthood)
- diagnostic overshadowing; misunderstanding between what is seizure activity and what is behaviour associated with the learning disability, particularly in relation to seizures of the partial type (descriptions of different seizure types as outlined by the International Classification of Seizures are offered in Appendices 1 and 2)
- lack of consideration given to the effects on quality of life when anti-epileptic medications are being prescribed/reviewed
- little evidence of thorough understanding and use of the Mental Capacity Act and best interest decisions particularly in relation to people with complex needs who very often have equally complex medication regimes
- not enough 'out of hours' support at times when problems arise during changes to medication regimes
- not enough information about SUDEP given to those who are at high risk of this and those who support them
- evidence that there is a lack of investment by providers of social care in the strong health facilitation and leadership that is needed by those with learning disabilities and complex epilepsy who are using their services.

In addition to establishing the above (the need to elaborate on these in a more detailed document is suggested under Next Steps) our engagement work has enabled us to bring together a wide group of individuals from different parts of the region who are now connected to one another and form a knowledgeable and informed network able to provide advice and support to one another and to others with learning disabilities who have epilepsy.

Within this group is a smaller number of skilled individuals who, with support, are keen to work with others in the co-production and co-delivery of training programmes, as well as on the co-production of information and resource packages



in relation to adults with learning disabilities who have epilepsy. Both of these have been identified by the team from *Inclusion East* as existing gaps in what is currently offered/available both at regional and national level. Our workshop at the Managed Clinical Network event on 9<sup>th</sup> December was a response to this; a quiz-based workshop, co-produced and co-delivered by people with learning disabilities, to broaden attendees understanding of epilepsy for those who have learning disabilities.



## NEXT STEPS

The team from *Inclusion East* believe the following could all contribute to improving outcomes for people with learning disabilities living with epilepsy, both regionally and nationally:-

- production of a document that will provide a more in-depth account of the stories we heard during the project, raising awareness of issues faced in an emotionally engaging and personalised manner
- development of a small team of individuals, comprising people with learning disabilities and epilepsy as well as family carers, to work together, and alongside professionals, to co-produce information packages and training materials aimed at better addressing the specific needs of this group
- further development of the team in order to enable, in addition to the above, participation in the co-delivery of training to those involved in the support of people with learning disabilities who have epilepsy
- opportunity for the team to liaise with professionals who are involved in providing information and advice to individuals and families of individuals who are at risk of SUDEP
- inclusion of epilepsy in Quality of Life audits, utilizing the skills and experience of those within our team and network.

**For further information please contact Inclusion East:**

Vicki Raphael: [vicki\\_raphael@hotmail.com](mailto:vicki_raphael@hotmail.com)

Matt Clark: [matthew.clark70@ntlworld.com](mailto:matthew.clark70@ntlworld.com)





## APPENDIX 1: INTERNATIONAL CLASSIFICATION OF SEIZURES

The International Classification of Seizures (published by the International League Against Epilepsy) is the most commonly used way of categorising epileptic seizures.

### PARTIAL/FOCAL SEIZURES

The disturbance in brain activity begins or involves an isolated area of the brain. The nature of the seizure is usually determined by the function of the part of the brain involved.

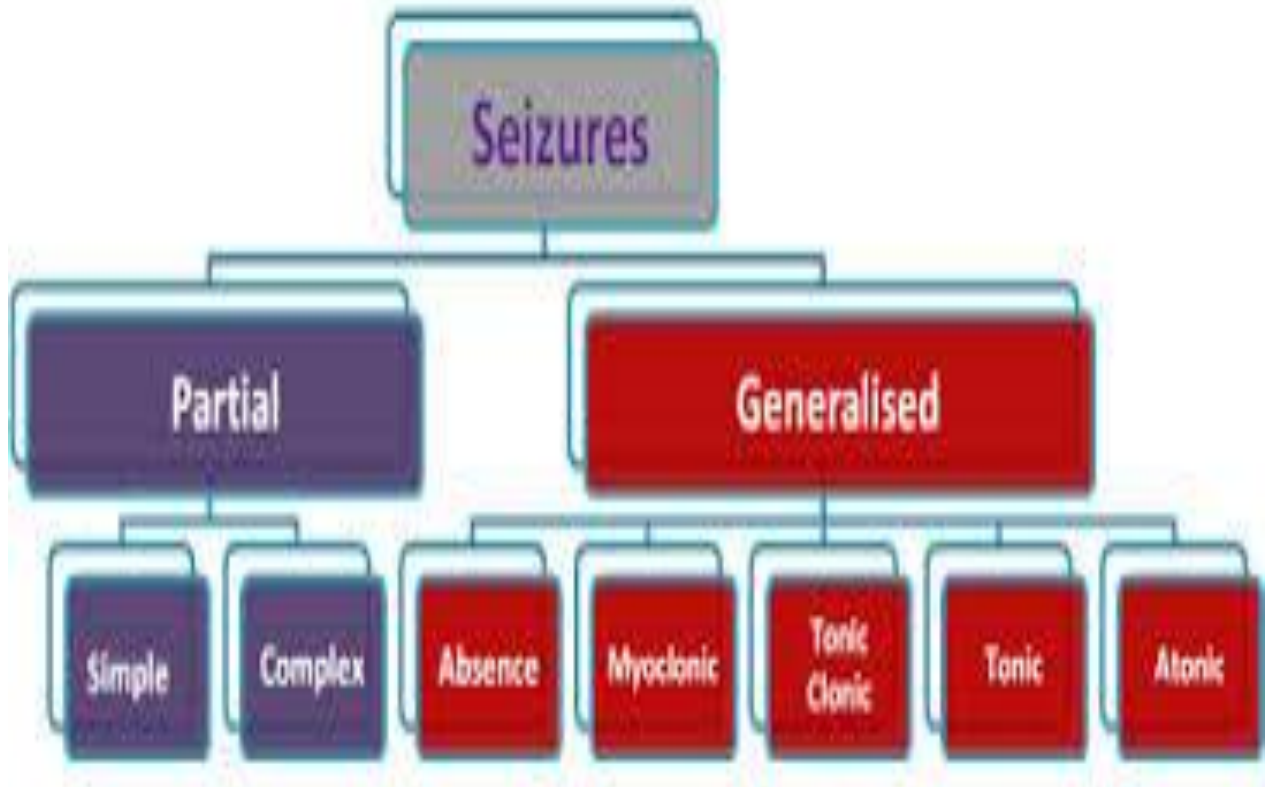
- **Simple Partial:** Seizure activity with no loss of consciousness.
- **Complex Partial:** Seizure activity with consciousness impaired.
- **Secondary Generalisation:** Begins in one small part and spreads to involve the whole brain. Consciousness is always impaired.

### GENERALISED SEIZURES

The whole brain is affected and consciousness is always impaired. There may be a period of convulsion after the seizure and there will be no memory of the event.

- **Tonic:** Whole body stiffens as muscles tighten.
- **Tonic-Clonic:** Body stiffens and muscles then convulse.
- **Atonic:** Body loses all muscle tone.
- **Absence:** Blanking out for a period of time, sometimes brief.
- **Myoclonic:** Arms, head and sometimes whole body jerk briefly.

## APPENDIX 2: DIAGRAM OF SEIZURE TYPES



## **APPENDIX 3: NICE GUIDELINES ON DIAGNOSIS AND MANAGEMENT OF EPILEPSIES WITH REFERENCE TO PEOPLE WITH LEARNING DISABILITIES/INTELLECTUAL IMPAIRMENTS.**

What do the guidelines say about epilepsy?

Keywords: “Disability”, “learning”, “impairment”.

Below are the clauses mentioning the above keywords within the 2012 NICE guideline 137 “The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care”.

Section: **Coping with epilepsy**

1.3.9 The possibility of having seizures should be discussed, and information on epilepsy should be provided before seizures occur, for children, young people and adults at high risk of developing seizures (such as after severe brain injury), with a learning disability, or who have a strong family history of epilepsy. [2004]

Section : **1.16 Children, young people and adults with learning disabilities (see also sections 1.15 and 1.17)**

**1.16.1 Diagnosis (see also section 1.5)**

1.16.1.1 It can be difficult to diagnose epilepsy in children, young people and adults with learning disabilities, and so care should be taken to obtain a full clinical history.

Confusion may arise between stereotypic or other behaviours and seizure activity. [2004]

1.16.1.2 It is important to have an eye witness account supplemented by corroborative evidence (for example, a video account), where possible. [2004]

1.16.1.3 Clear, unbiased reporting is essential. Witnesses may need education to describe their observations accurately. [2004]

**1.16.2 Investigations (see also section 1.6)**

1.16.2.1 Those with learning disabilities may require particular care and attention to tolerate investigations. [2004]

1.16.2.2 Facilities should be available for imaging under anaesthesia, if necessary. [2004]

1.16.2.3 In the child or young person presenting with epilepsy and learning disability, investigations directed at determining an underlying cause should be undertaken. [2004]

### **1.16.3 Management (see also section 1.8)**

1.16.3.1 Enable children, young people and adults who have learning disabilities, and their family and/or carers where appropriate, to take an active part in developing a personalised care plan for treating their epilepsy while taking into account any co-morbidities. [new 2012]

1.16.3.2 Ensure adequate time for consultation to achieve effective management of epilepsy in children, young people and adults with learning disabilities. [new 2012]

1.16.3.3 In making a care plan for a child, young person or adult with learning disabilities and epilepsy, particular attention should be paid to the possibility of adverse cognitive and behavioural effects of AED therapy. [2004]

1.16.3.4 The recommendations on choice of treatment and the importance of regular monitoring of effectiveness and tolerability are the same for those with learning disabilities as for the general population. [2004]

1.16.3.5 Do not discriminate against children, young people and adults with learning disabilities, and offer the same services, investigations and therapies as for the general population. [new 2012]

1.16.3.6 Every therapeutic option should be explored in children, young people and adults with epilepsy in the presence or absence of learning disabilities. [2004]

1.16.3.7 Healthcare professionals should be aware of the higher risks of mortality for children, young people and adults with learning disabilities and epilepsy and discuss these with them, their families and/or carers. [2004]

1.16.3.8 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. [2004]

## **APPENDIX 4: SUDDEN UNEXPECTED DEATH IN EPILEPSY**

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### **Author**

Steven C Schachter, MD

### **Section Editor**

Timothy A Pedley, MD

### **Deputy Editor**

April F Eichler, MD, MPH

All topics are updated as new evidence becomes available and our peer review process is complete.

**Literature review current through:** Jul 2013. | **This topic last updated:** Jul 23, 2012.

**INTRODUCTION** — Patients with epilepsy have a small risk of sudden unexpected death, a condition referred to as sudden unexpected death in epilepsy (SUDEP) [1,2]. SUDEP is defined specifically as the sudden, unexpected, witnessed or unwitnessed, nontraumatic and nondrowning death in patients with epilepsy with or without evidence for a seizure, and excluding documented status epilepticus, in which post mortem examination does not reveal a structural or toxicologic cause for death [3].

The cause of SUDEP is uncertain. Observations in individual cases have suggested possible cardiogenic, pulmonary, and primary neurologic etiologies. It may be that SUDEP is a heterogeneous condition.

This topic discusses risk factors, causes, and prevention strategies for SUDEP. The management and other complications of seizures and epilepsy are discussed separately. (See "Overview of the management of epilepsy in adults" and "Evaluation and management of drug-resistant epilepsy".)

**DEFINITION** — Sudden unexpected death in epilepsy (SUDEP) is defined specifically as the sudden, unexpected, witnessed or unwitnessed, nontraumatic and nondrowning death in patients with epilepsy with or without evidence for a seizure, and excluding documented status epilepticus, in which post mortem examination does not reveal a structural or toxicologic cause for death [3,4].

**INCIDENCE** — SUDEP causes between 2 and 18 percent of all deaths in patients with epilepsy [5-9]. This proportion may be somewhat higher in children, as high as 30 percent in one cohort study [10]. The overall mean incidence is estimated at about 2 cases per 1000 patient years [6,8,10,11]. The incidence of SUDEP increases with severity of epilepsy, and may be as high as 0.5 to 1 percent a year in those with severe refractory epilepsy [1,5].

**RISK FACTORS** — Risk factors for SUDEP include early age of epilepsy onset, frequent generalized tonic-clonic seizures, and intractable epilepsy [1,2,5,10,12-17]. Case-control and cohort studies of SUDEP have identified certain clinical and demographic features as potential risk factors, although these are not all consistently found in all studies [2,5,8,11,13,17-20]:

- Seizure frequency (>1/month)
- Medication noncompliance, subtherapeutic AED level
- Age 20 to 45 years
- Generalized tonic-clonic seizures
- Polytherapy
- Duration of epilepsy (>10 years)
- Alcoholism
- Male gender
- Nocturnal seizures

Most reported cases of SUDEP are in young adults, but children younger than one year can be affected [6,8,21-23]. SUDEP is relatively unusual in individuals greater than 45 years. However, the latter observation may be due to under-reporting of sudden death as SUDEP in an older individual, particularly if there are medical comorbidities.

**ETIOLOGIES** — While SUDEP is only rarely witnessed, most observations suggest that SUDEP occurs in the context of a seizure [2,3,23,24]. While a few cases of witnessed SUDEP in the absence of clinical seizure activity have been described, subclinical seizure activity remains a possibility in these individuals [16,22,25]. Many SUDEP deaths occur while the patient is asleep in bed [3,8,16,21,23].

Possible causes of SUDEP include cardiac arrhythmia, central apnea, neurogenic pulmonary edema, and laryngospasm [2,8,12,24,26-31].

**Cardiogenic** — Indirect evidence suggests that cardiogenic mechanisms may be involved in SUDEP.

- Some patients with epilepsy have ictal bradycardia and even asystole [32-35]. Atrioventricular conduction block is a less frequently described ictal phenomenon [36]. The frequency of this phenomenon is uncertain [37]. One case series monitored electrocardiograms using an implantable loop recorder for an average of 18 months

in 19 patients with refractory epilepsy [38]. Four patients had bradycardia or periods of asystole, prompting subsequent pacemaker placement; three of these episodes occurred during a clinical seizure. In another study, ictal asystole was observed on long-term EEG monitoring in 10 of 6825 patients (0.15 percent) [33]. This phenomenon was not associated with cardiovascular risk factors or abnormal baseline ECG. Another series that reviewed monitoring data on 1277 seizures in 69 individuals identified five patients with ictal bradycardia occurring in 18 percent of their seizures; two of these individuals had asystolic episodes [35]. This phenomenon is similarly unusual in children. In one case series of 49 children who had seizures on a monitoring study, ictal bradycardia occurred in 3.7 percent of seizures and only during partial complex seizures of extratemporal onset [39]. Monitoring studies have identified clinical features that suggest an ictal arrhythmia. Diffuse atonia has been observed to accompany episodes of asystole lasting longer than eight seconds [33,34]. Another report documented sudden falls due to seizure-induced asystole in three patients with partial epilepsy [40]. These observations suggest that when seizures include a delayed loss of tone, clinicians should consider the possibility of ictal asystole and need for cardiac monitoring. Pacemaker implantation appears to reduce falls and secondary morbidity in patients with seizure-induced asystole and bradycardia when seizures are not controlled by other therapies [41,42].

- While bradycardia and asystole are more commonly reported, partial seizures have been associated with tachyarrhythmias as well. In one case report, a 51 year old woman developed ictal ventricular tachycardia evolving into a ventricular fibrillation arrest requiring resuscitation [43].
- A number of case series of combined EEG-ECG telemetry have noted prolongation of the QT interval during seizures in 12 to 23 percent of patients [44-48]. This finding may be associated with potentially fatal ventricular arrhythmias. However, in a matched case-control study, a prolonged QT interval was not specifically associated with SUDEP [46]. A preictal shortened QT interval has also been observed and is of uncertain clinical significance [47]. Other ECG markers for cardiovascular arrhythmia and mortality (T-wave alternans, ventricular late potentials) have also been found to be more prevalent in patients with epilepsy compared with controls in small case series [49,50].

These studies suggest the possibility that cardiac arrhythmias may cause some cases of SUDEP [51]. However, direct evidence of mortality from seizure-induced cardiac arrhythmias is lacking [37]. It is not definitely known if the risk of SUDEP is increased for patients with ictal arrhythmias compared with patients who do not have ictal changes in heart rhythm. In one study of 21 patients with SUDEP, a prior EEG-ECG recording had revealed ictal cardiac repolarization and rhythm abnormalities in 56 percent of SUDEP patients compared to 39 percent of controls [52]. SUDEP patients also had higher ictal heart rate elevations, particularly in seizures that arose from sleep. However, this data does not indicate that routine ECG recording or



ambulatory ECG monitoring can identify which epilepsy patients are at increased risk for SUDEP. One investigation observed that the cardiovascular characteristics of ictal asystole were similar to tilt-table induced vasovagal asystole [53]. The authors speculated that ictal asystole might share a pathophysiologic mechanism with vasovagal syncope and perhaps be similarly benign.

The etiologic link between seizures and cardiac arrhythmias and possibly sudden cardiac death is not known [54,55]. Channelopathies (diseases caused by abnormal function of ion channels) underlie some causes of epilepsy as well as some cardiac arrhythmias. Some suggest that release of catecholamines during seizures may lead to cumulative cardiac injury (takotsubo cardiomyopathy) and possibly a susceptibility to arrhythmia or sudden hemodynamic compromise and cardiogenic shock [56]. The influence of antiepileptic drug treatment on SUDEP is not defined. Both carbamazepine and phenytoin have been implicated as possibly exacerbating some of the autonomic and cardiac effects of seizures, but an increased risk of either ictal arrhythmias or SUDEP with these or other AEDs has not been defined [37]. Lamotrigine has been identified in some reports as increasing the risk of SUDEP, possibly through a proarrhythmic effect [17,57].

Epilepsy has also been linked to cardiovascular disease. A case-control study in Stockholm found that a history of epilepsy was a risk factor for acute myocardial infarction (MI) (OR = 4.8) and was associated with a poor prognosis after MI [58]. Other studies have also linked epilepsy and cardiovascular mortality [59,60]. Proposed mechanisms underlying this association include an adverse effect of AEDs on cholesterol and lipid metabolism and obesity; increased smoking and decreased physical activity in persons with epilepsy [61]; as well as a possible adverse effect of chronic epilepsy upon the heart.

**Pulmonary** — Alternatively, ventilatory failure with ictal hypoxemia and hypercapnia from centrally-mediated apnea may underlie some cases of SUDEP; frank apnea is sometimes observed [25,62,63].

- In a series of 56 patients with intractable epilepsy, oxygen saturation dropped below 90 percent in 33 percent of both partial and secondarily generalized seizures, below 80 percent in 10 percent of seizures, and below 70 percent in 3.6 percent of seizures [63]. Seizure duration and temporal lobe localization were risk factors for desaturation. The timing of the apnea appeared to coincide with contralateral spread [64]. Most apnea episodes appeared to be of central origin; 9 percent appeared obstructive. Two patients have been described with seizures whose sole or major ictal manifestation was apnea [25]. In children, the prevalence of ictal hypoxia appears to be similar. In one case series of 225 seizures in 49 monitored children, 27 percent of seizures were associated with ictal hypoxia; in one third of these, oxygen saturation dropped below 60 percent [39].
- Another case series of 94 recorded seizures in 33 patients revealed elevations in

ictal/postictal endtidal CO<sub>2</sub> in eleven patients [65]. Peak endtidal CO<sub>2</sub> was above 50 mmHg in 35 seizures, above 60 mmHg in 15, and above 70 mmHg in 5 seizures. Because the degree of hypercapnia was not associated with apnea or seizure duration, the investigators speculated that it possibly resulted from ventilation-perfusion mismatch or transient pulmonary edema.

These observations don't clearly link apnea to SUDEP; however, in one case, a woman undergoing EEG monitoring developed persistent apnea in the setting of a convulsive seizure [29]. Her cardiac rhythm remained intact initially and she underwent successful cardiopulmonary resuscitation. Another case series found that seizures associated with oxygen desaturation were also more likely to have ictal QT prolongation (OR=4.3) [48].

Alternative causes of ictal respiratory compromise include laryngospasm, asphyxiation, aspiration, and neurogenic pulmonary edema [54]:

- The former was observed in a single case of a 40-year-old man who developed laryngospasm with stridor and cardiopulmonary arrest during a generalized tonic-clonic seizure and was successfully resuscitated [30].
- It has been noted in some case series, although not in others, that patients with SUDEP are often found in the prone position, suggesting that asphyxiation may have played a role in their death [3,16,21,66]. In a few cases of witnessed SUDEP, airway obstruction was evident [24].
- Case series with post mortem examination often find pulmonary edema in patients with SUDEP [12,16,21,23]. In one study, the prevalence of this finding was significantly higher in those with SUDEP compared to patients with epilepsy who died of an alternative cause (62 versus 27 percent) [16]. However, the degree of edema observed is typically felt to be insufficiently severe to cause death.

**Neurologic** — Sudden, persistent cerebral electrical silence after a seizure, suggesting a primary central nervous system mechanism, has been documented in a few cases of SUDEP [67-69]. This may be the cause of some cases of ictal central apnea.

In two cases of SUDEP captured on video-EEG monitoring, marked attenuation of EEG activity was followed by irregular respirations and then apnea, followed by ECG changes and asystole [70]. A retrospective review of video-EEG telemetry studies in 10 patients who subsequently died of SUDEP found that postictal generalized EEG suppression was seen in half of these patients compared with 38 percent of seizures in control patients [71]. This period of postictal EEG suppression was significantly longer in patients with SUDEP; in particular, a duration of >50 seconds was associated with a significantly elevated odds of SUDEP. Other series of monitored patients without SUDEP have suggested possible pathogenic roles for this phenomenon in SUDEP. One such study found that postictal EEG suppression was associated with longer and more severe periods of postictal oxygen desaturation

[72]; another correlated the duration of postictal EEG suppression with the presence and magnitude of sympathetic activation and parasympathetic suppression [73].

Some have suggested that brainstem serotonergic pathways may be involved in the pathogenesis of SUDEP, as they have been proposed to play a role in sudden infant death syndrome [74]. In an animal model of SUDEP in which audiogenic seizures precipitate respiratory arrest, this phenomenon was prevented by administration of a selective serotonin reuptake inhibitor, and induced by the serotonin receptor antagonist, cyproheptadine [75].

**PREVENTION AND COUNSELING** — There are no data based on clinical trials regarding the prevention of SUDEP. Existing strategies come from knowledge of risk factors identified in cohort and case-control studies as well as expert recommendations.

- Identifying patients at high risk, educating patients and their families about SUDEP, ascertaining seizure precipitants, and promoting compliance with treatment are strategies to reduce risk [2,15].
- Aggressive treatment of refractory epilepsy, including referral to a comprehensive epilepsy center and consideration of epilepsy surgery, appears to be appropriate. However, SUDEP was not an outcome measure in randomized trials evaluating the efficacy of these measures. In one nonrandomized study, mortality was compared in patients who had epilepsy surgery to those who had pre-surgical assessment but no surgery [76]. Non-operated patients were 2.4 times more likely to die as those who had surgery and were 4.5 times more likely to die a probably epilepsy-related death. In a longitudinal follow-up study of patients after temporal lobe resection, the risk for premature death decreased over time, although it remained somewhat higher than the standard population [77]. Efforts to reduce seizure frequency with additional trials of adjunctive AEDs also seems appropriate for patients who are not surgical candidates [2]. In a meta-analysis of placebo-controlled randomized trials in patients with refractory seizures, the risks of definite or probable SUDEP, all SUDEP, and all-cause death were lower in patients who received adjunctive AEDs in efficacious doses compared to patients in the placebo group (OR=0.17, 0.17 and 0.37, respectively) [78]. The management of drug-resistant epilepsy is discussed in detail separately. (See "Evaluation and management of drug-resistant epilepsy".)
- Supervision at night (defined as the supervising person sharing the same bedroom, or use of special precautions such as regular checks throughout the night, or use of a monitoring device) was associated with a decreased risk of SUDEP in a large case-control study [14]. The efficacy of this intervention has not been confirmed in a prospective, controlled trial.
- ECG and/or ECG monitoring should be considered for patients with intractable epilepsy [37,79]. However, this approach has not been evaluated prospectively to evaluate its impact on managing the risk of SUDEP.

Informing patients about SUDEP is controversial, as there are no proven effective

preventive measures [11]. Most clinicians counsel high risk patients, particularly in the setting of risky, noncompliant behaviour or when discussing the role of epilepsy surgery [80,81]. However, a joint task force of the American Epilepsy Society and the Epilepsy foundation and concluded that information regarding the risk of SUDEP should be disclosed to ALL patients with a diagnosis of epilepsy as part of a comprehensive education program [54,82]. A survey of parents of children with epilepsy indicated that more than 90 percent thought that this information should be provided to them [83].

The potential benefits of counselling were outlined by a panel of experts and included [54]:

- Helps physicians and patients share in treatment goals
- Helps establish “truth-telling relationship”
- Avoids a false sense of security and complacency regarding epilepsy and its treatment
- Allows expression of patients’ anxiety about their diagnosis and encourages constructive discussion
- Allows people with epilepsy to organize their lives with reasonable expectations
- Allows people with epilepsy and caregivers to reduce potential risk factors for SUDEP, promoting compliance and minimizing behaviours that increase seizure-risk
- Reduces the fear of SUDEP in low-risk populations
- Lessens pain, grief, and blame in families in the event SUDEP does occur

**SUMMARY AND RECOMMENDATIONS** — Patients with epilepsy have a small but significant risk of sudden unexpected death (SUDEP).

- The most consistently observed risk factors for SUDEP are frequent convulsive seizures. (See 'Risk factors' above.)
- The etiology of SUDEP is uncertain; most cases appear to occur in the context of a seizure. Theories include a cardiac arrhythmia, central apnea, or a primary central nervous system event complicating the seizure ictus. It may be that causes are heterogeneous. (See 'Etiologies' above.)
- There are no strategies that have been shown to reduce the risk of SUDEP. Maximizing seizure control is recommended, including timely consideration of epilepsy surgery.
- We suggest that clinicians counsel patients and when appropriate, family members, about SUDEP (especially patients with risk factors) to promote medical compliance and when discussing aggressive treatment of refractory epilepsy (**Grade 2C**). (See 'Prevention and counselling' above.)

## APPENDIX 5: FURTHER READING

- Confidential Inquiry into Premature Deaths of People with Learning Disabilities (CIPOLD) available from:  
<http://www.bris.ac.uk/cipold/>
- Wasted Money, Wasted Lives available from:  
<http://jointepilepsycouncil.org.uk/downloads/2011/Wasted%20Money,%20Wasted%20Lives.pdf>
- Learning Disabilities and Epilepsy available from:  
<http://www.epilepsy.org.uk/info/learning-disabilities>
- Learning Disability and Epilepsy available from:  
<http://www.epilepsysociety.org.uk/learning-disability-and-epilepsy>
- Using NICE Guidance to Standardise Epilepsy Management available from:  
<http://www.nursingtimes.net/using-nice-guidance-to-standardise-epilepsy-management/1044290.article>

## APPENDIX 6: ACKNOWLEDGEMENTS

The team from *Inclusion East* who worked on the Managed Clinical Network's epilepsy project were Vicki Raphael, Matthew Clark and Christian Raphael. Thanks also to Jo Hough and other members of *Inclusion East* for their support during the project.

Thank you to all the people with learning disabilities who have epilepsy, and their family carers, with whom we met and spoke with. These individuals are not named, in order to protect their privacy.

Thank you to all the professionals with whom we met and spoke with, particularly Jane Swannell and Anthony Linklater.

Thank you to all those who attended and took part in our advisory group meetings; Dr Asif Zia, Dr Inderbir Sidhu, Dr Indermeet Sawhney, Dr Howard Ring and Professor Robert Gates.

Thank you to George Bainbridge who researched and wrote Appendices 3 and 4 of this report.

Image on front cover is taken from *The Portrait Anatomised* by Susan Aldworth and was chosen by someone with learning disabilities who has epilepsy who took part in our engagement work.

