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Service responses to people with intellectual disabilities and epilepsy: a systematic review

Abstract

Background. Epilepsy is highly prevalent in people with intellectual disabilities and is associated with increased mortality and high healthcare usage. This systematic review summarises research on service responses to people with intellectual disabilities and epilepsy.

Method. Studies published from 1990 were identified via electronic searches using Medline, Cinahl, PsycINFO and Web of Science, email requests to researcher networks, and cross-citations.

Information extracted from studies was reviewed narratively in relation to identified themes.

Results. 35 studies met the inclusion criteria. Overall study quality was low, with no RCTs or similarly robust intervention study designs. Access to specialists was inconsistent. The importance of proxies and the need for education regarding epilepsy for staff, carers and people with intellectual disabilities was highlighted.

Conclusion. There are no methodologically robust studies on service related interventions for people with intellectual disabilities and epilepsy. Further research on improving service delivery is required to substantiate findings reported here.

Keywords: epilepsy; intellectual disabilities; service responses

Introduction

Epilepsy is one of the most common serious brain disorders, affecting over 50 million people worldwide (WHO, 2005), approximately 0.5% to 1.0% of the general population (Forsgren *et al.*, 2005, Linehan *et al.*, 2010, Joint Epilepsy Council, 2011). Despite variations in reported prevalence figures, the prevalence of epilepsy in people with intellectual disabilities is clearly much greater than in the general population. A systematic review and meta-analysis of 38 studies of general samples of people with intellectual disabilities found a pooled prevalence of 22.2% (95% CI 19.6, 25.1) (Robertson *et al.*, 2015b).

Epilepsy is associated with increased mortality in people with intellectual disabilities, particularly in those experiencing recent seizures (Robertson *et al.*, 2015a). For people with intellectual disabilities, epilepsy or convulsions has been identified as an important and to some extent preventable cause of death (Glover and Ayub, 2010). Based on Standardised Mortality Odds Ratios (SMORs), adjusting for ages at death, people where death involved epilepsy or unspecified convulsions were 9.7 times more likely than others to have an intellectual disability-related condition (95% confidence interval 9.1, 10.4).

There is little information available on the costs of epilepsy specifically for people with intellectual disabilities. One study examined the health and social care costs of supporting 91 adults with active epilepsy and intellectual disabilities living in the community in England (Pennington *et al.*, 2012). Costs of health and social services of supporting people with epilepsy and intellectual disabilities were found to be high, but epilepsy related health care costs were a small fraction of overall costs, with most being primarily due to the cost of providing appropriate accommodation and living support and appropriate activities for people with intellectual disabilities. The study suggests that the costs of epilepsy management are higher for people with intellectual disabilities than for the rest

of the population. Epilepsy has also been found to be associated with additional costs in an institutional setting in the United States (Burke *et al.*, 1999).

It is clear that people with intellectual disabilities and epilepsy have high health care usage. In the Dutch National Survey of General Practice, the most frequently presented health problem in people with intellectual disabilities in primary care settings was epilepsy (Straetmans *et al.*, 2007). Patients with intellectual disability and co-existing epilepsy in Wales also used secondary care services (inpatients, outpatients, and accident and emergency (A&E)) more frequently than those with intellectual disability only (Morgan *et al.*, 2003). In England, it has been reported that 'convulsions and epilepsy' accounted for more than 40% of all emergency admissions for ambulatory care sensitive conditions (ACSCs) for people with intellectual disabilities, accounting for 6,000 admissions and 28,000 bed days per year (Glover and Evison, 2013). Similarly, a Canadian study found that for people with intellectual disabilities, 27% of admissions for ACSCs were for epilepsy, a hospitalisation rate 54 times higher than for people without intellectual disabilities (Balogh *et al.*, 2010).

Given the high prevalence of epilepsy in people with intellectual disabilities, the associated increased mortality, and high health care usage, the issue of how services should respond to epilepsy in people with intellectual disabilities is important. The management of epilepsy in people with intellectual disabilities presents unique challenges, such as the possibility of misdiagnosis due to the misinterpretation of behavioural, physiological, syndrome related, medication related or psychological events by parents, paid carers and health professionals (Chapman *et al.*, 2011).

Clinical guidelines relating to the management of epilepsy in people with intellectual disabilities are available. The National Institute for Health and Clinical Excellence include a section on children, young people and adults with intellectual disabilities in their clinical guidelines for the diagnosis and management of the epilepsies in adults and children in primary and secondary care (National Institute for Health and Care Excellence, 2012), with the Guideline Development Group stating that

“this patient group has traditionally received sub-optimal care, and less access to specialist epilepsy services” (National Clinical Guideline Centre, 2012, p552). Consensus guidelines into the management of epilepsy in adults with an intellectual disability have been produced using a modified Delphi method by the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD) (Kerr *et al.*, 2009). International consensus clinical practice statements have also been developed regarding the treatment of neuropsychiatric conditions associated with epilepsy (Kerr *et al.*, 2011). Finally, earlier guidelines produced by IASSIDD include recommendations relating to the care context and has a specific section on standards for services (Kerr *et al.*, 2001).

For people with epilepsy generally, research on service approaches is evident. For example, a Cochrane review on care delivery and self-management strategies for adults with epilepsy (Bradley Peter and Lindsay, 2008) identified 13 trials and 16 reports, including five trials of specialist epilepsy nurses. There was some evidence of benefit for interventions based on specialist epilepsy nurses and self-management education. Since this review was conducted, further research has been conducted such as a non-randomised trial of a nurse led self-management intervention to reduce emergency visits by people with epilepsy (Noble *et al.*, 2014) and further research regarding self-management education is ongoing (Magill *et al.*, 2015). However, an earlier literature review found no research in the area of service delivery for people with intellectual disabilities and epilepsy (Bowley and Kerr, 2000).

This review aims to identify and summarise research on service responses to the general population of people with intellectual disabilities and epilepsy and as such it excludes studies on specific syndromes associated with intellectual disabilities. The effectiveness of either pharmacological or non-pharmacological treatments for epilepsy in people with intellectual disabilities are not covered for two reasons. First, recent Cochrane reviews exist (Beavis *et al.*, 2007a, Beavis *et al.*, 2007b; both

assessed as up to date in 2011), as well as a protocol for a future Cochrane review of pharmacological and non-pharmacological interventions for epilepsy in people with Down syndrome (Bashir *et al.*, 2013). Second, NICE (2012) guidelines state that: “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” (National Institute for Health and Care Excellence, 2012, p51). However, the review does include the use of rescue medication due to the impact service factors, such as training of staff or carers in its use, may have on outcomes. The review aims to cover research on a disparate range of topics within the broad area of service responses to epilepsy in people with intellectual disabilities published from 1990 onwards.

Method

Electronic literature database searches were conducted in Medline, Cinahl and PsycINFO (all on EBSCO) and Web of Science (SCI-EXPANDED, SSCI and A&HCI). Searches were also conducted in the following clinical trials registers: Cochrane Central Register of Controlled Trials (CENTRAL); ClinicalTrials.gov (U.S. National Institutes of Health); ISRCTN registry; and the UK Clinical Trials Gateway. The reference lists of articles meeting the inclusion criteria were searched and articles from authors’ personal collections included. A request for information on research relevant to the review was sent in January 2015 to the membership of the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD) Health Special Interest Research Group and the Intellectual Disability UK Research mailing list.

Searches combined word and index terms for epilepsy, intellectual disabilities, and service responses with the Boolean operator ‘and’. During the process of producing earlier reviews regarding people with intellectual disabilities who have epilepsy (Robertson *et al.*, 2015b, Robertson *et al.*, 2015a), research articles relevant to any aspect of service responses were identified and word search terms were developed based on these. Where available, each database index was also explored to identify

index terms relevant to service responses and these were combined with word search terms using the Boolean operator 'or'. Searches were completed in January 2015. Full details of the search terms are given in Appendix One.

Inclusion Criteria

Articles were required to meet all following criteria:

- Peer reviewed
- English language full text
- Published from 1990
- Primary research, service audit or evaluation
- Samples of people with intellectual disabilities or samples where 50% or more have intellectual disabilities or mixed samples where results are disaggregated for people with intellectual disabilities

In addition, articles had to meet one of the following criteria:

- Studies of interventions aiming to improve knowledge or practice in relation to service responses to people with intellectual disabilities and epilepsy; or
- Studies of current knowledge or practice in relation to service responses to people with intellectual disabilities and epilepsy; or
- Studies on opinions of professionals, carers or family with regard to services for people with intellectual disabilities and epilepsy; or
- Studies considering service related factors associated with outcomes for people with intellectual disabilities and epilepsy.

Exclusion Criteria

- Not peer reviewed or where peer review status was unclear
- Letters, commentaries, editorials, meeting or conference abstracts
- Case studies or case series

- Only includes information relating to specific syndromes eg. Dravet syndrome
- Narrative reviews
- Conditions where intellectual disabilities cannot be assumed (e.g. cerebral palsy, autistic spectrum disorder (ASD)) where results not disaggregated for people with intellectual disabilities
- Pharmacological or non-pharmacological treatment of epilepsy, except studies relating to the use of rescue medications by family or paid caregivers.

Initially, titles and abstracts were used to exclude articles obviously not within the scope of the review (1st and 2nd author). Those retained for further screening were those for which relevance could not be assessed without accessing full text, or those that were chosen as potentially within scope. These studies were screened by two authors (1st and 4th author) and discussed until consensus was reached on whether or not they met the inclusion criteria.

It was evident from the outset that there were no randomised controlled trials (RCTs) or studies with robust research designs and as such formal quality assessment of the literature identified was not undertaken. All relevant studies were included in the review regardless of methodological quality, although studies were categorised by research design in order to illustrate the overall number of studies identified in relation to established hierarchies of evidence (GRADE Working Group, 2004).

Data Extraction & Synthesis

Data were extracted from the full text of included articles. Textual descriptions were produced for each study by the 1st author. This included: bibliographic details; the country within which the study took place; details of the focus of the study; sample size and characteristics; study design and data sources; measures employed; main results; and issues raised in the discussion. This information was then tabulated.

The tabulated descriptions were reviewed by the 1st author to identify themes emerging from the literature for inclusion in the review results. An iterative approach was taken to determining themes with topics being listed as they arose and theme headings being developed which best categorised the topics. The 4th author also reviewed the tabulated descriptions of the studies in order to assess the appropriateness of the identified themes and no disagreement occurred. Studies providing evidence in relation to each theme were then identified from the textual descriptions and the information reviewed narratively. It was generally not possible to compare results between studies directly due to variation in the methods used. As such, no meta-analysis was conducted.

Results

The process of identifying studies for inclusion is summarised in Figure One. Electronic database searches identified 1,861 references, with 1,481 remaining after removal of 380 duplicates. Following the first screening, 1,333 references were excluded and 148 remained for further screening. After examination of full text articles and the addition of articles cited within these and from authors' personal collections, 35 articles met the criteria for inclusion. Searches of clinical trials registers identified 135 records of which 2 were relevant ongoing trials where results were not yet available (Durand *et al.*, 2014, Ring *et al.*, 2014) and these trials are considered further in the discussion section below.

Figure One

Country

Most studies identified were from the United Kingdom, including 17 from England (Ahmad *et al.*, 2007, Beber *et al.*, 1999, Branford and Collacott, 1994, Branford *et al.*, 1998, Cole *et al.*, 2009, Deepak *et al.*, 2012, Esan and Markar, 2010, Frost *et al.*, 2003, Kiani *et al.*, 2014, Markar and

Mahadeshwar, 1998, Mobbs *et al.*, 2002, Mukhopadhyay *et al.*, 2010, Redley *et al.*, 2013, Reuber *et al.*, 2008, Ring *et al.*, 2009, Tiffin and Perini, 2001, Whitten and Griffiths, 2007), three from Wales (Chubb *et al.*, 1995, Matthews *et al.*, 2008, Morgan *et al.*, 2003), and one from Scotland (Clark *et al.*, 2001). There were four studies from the United States (Baribeault, 1996, Buelow *et al.*, 2006, Hom *et al.*, 2015, Litzinger *et al.*, 1993) and in one survey most respondents were from the United States (US) (Camfield *et al.*, 2011). There were also two studies from Australia (Beran and McAulley, 1992, Kyrkou *et al.*, 2006), two from the Netherlands (Vallenga *et al.*, 2006, Vallenga *et al.*, 2008) and one each from Belgium (Peeters, 2000), Canada (Fridhandler *et al.*, 2012), and Ireland (McCarron *et al.*, 2014). One further study included both the UK and Ireland (Thompson *et al.*, 2013). Finally, an international online survey obtained responses from 14 countries although most responses were from the UK (Kerr *et al.*, 2014). There were no studies identified from low or middle income countries (LAMI).

Table One Here: Summary of Studies

Study Designs

No RCTs or other intervention studies with robust designs were identified. There were no matched comparison groups and no study used randomization. There were: 12 articles based on cross-sectional quantitative survey methods (Cole *et al.*, 2009, McCarron *et al.*, 2014, Branford *et al.*, 1998, Beran and McAulley, 1992, Frost *et al.*, 2003, Mobbs *et al.*, 2002, Reuber *et al.*, 2008, Branford and Collacott, 1994, Camfield *et al.*, 2011, Kyrkou *et al.*, 2006, Mukhopadhyay *et al.*, 2010, Deepak *et al.*, 2012); six articles based on qualitative methods (Vallenga *et al.*, 2006, Vallenga *et al.*, 2008, Buelow *et al.*, 2006, Redley *et al.*, 2013, Thompson *et al.*, 2013, Kerr *et al.*, 2014); and five articles based on audits (Chubb *et al.*, 1995, Esan and Markar, 2010, Markar and Mahadeshwar, 1998, Tiffin and Perini, 2001, Whitten and Griffiths, 2007). One article was based on description of clinical experience (Baribeault, 1996). One cross-sectional prevalence study was identified which contained information related to service and antiepileptic drug (AED) receipt (Matthews *et al.*, 2008). There

were three articles based on retrospective review of varied data sources (Fridhandler *et al.*, 2012, Hom *et al.*, 2015, Kiani *et al.*, 2014) and one combined retrospective review of data sources with interviews (Ring *et al.*, 2009). One used record linkage to obtain standardized activity ratios for health service use (Morgan *et al.*, 2003).

Whilst a small number of studies looked at outcomes prior to and following a service intervention, some did not employ a comparison group (Litzinger *et al.*, 1993, Ahmad *et al.*, 2007). One study on the use of a decision tree for responding to seizures compared data from a prospective study of its use of with retrospective data prior to its use (Peeters, 2000). One study on the use of an educational package for people with mild intellectual disabilities used a deferred treatment group as a comparison group (Clark *et al.*, 2001). One study on the establishment of an intellectual disabilities psychiatric service compared baseline and outcome measurements for those who did compared to those who did not receive psychiatric care (Beber *et al.*, 1999).

Themes identified in relation to service responses to epilepsy in people with intellectual disabilities

In the following sections, we present a narrative summary of information identified in the 35 studies in relation to specific themes: service provision/configuration; the impact of service setting; epilepsy reviews; epilepsy care plans; investigations; seizure diaries; medication adherence; management by proxy; risk assessment; managing prolonged or serial seizures (rescue medication); educating people with intellectual disabilities about their epilepsy; evaluations of initiatives in services; prescribing practices; and views of families, carers or professionals regarding services. Further details on the studies can be found in Table One.

Service Provision/Configuration

Within the UK, the management of epilepsy for people with intellectual disabilities has been provided by various combinations of primary care, specialist epilepsy and neurology services as well

as intellectual disability mental health services and social care agencies (Ring *et al.*, 2009). This section summarises information regarding patterns of service provision.

In one English county, 37% of those with intellectual disabilities and epilepsy received epilepsy care from a hospital-based neurology service (Ring *et al.*, 2009). The patients' GP also actively contributed to their epilepsy care, in terms of initiating or changing treatments, in 63% of cases and for 6% of participants the GP was the only clinician supporting epilepsy treatment. It was not clear what determined which treatment pathway individual patients followed. Obvious factors, such as markers of epilepsy severity or associated co-morbidities, did not appear to play a role (Ring *et al.*, 2009). However, the picture may be different in other areas. For example, in a study on the establishment of an intellectual disabilities psychiatric service in one English county after a five year period without one, it was noted that at baseline most received GP care only for their epilepsy (Beber *et al.*, 1999).

Further evidence points to a lack of comprehensive involvement by specialist services for those with intellectual disabilities and epilepsy. In one English county, the specialist intellectual disability service had no involvement for 29% of those in the community receiving AEDs (Branford and Collacott, 1994). In an English city, 60.6% of patients with ongoing seizures, 57.9% with major seizures and 68.7% of individuals taken to hospital with prolonged seizures had no access to specialist advice (Reuber *et al.*, 2008). A prevalence study in Wales found that for those with intellectual disabilities and epilepsy, slightly over one in four participants had not seen a specialist (Matthews *et al.*, 2008).

Matthews *et al.* (2008) suggest that lack of specialist input may reflect a failure of continuity of care from paediatric to adult services. In a survey of predominantly pediatric neurologists from the US, lack of an adult neurologist willing to care for adults with epilepsy and an intellectual disability was noted by 35% to be a barrier to transitioning a patient to adult care (Camfield *et al.*, 2011).

Further studies point to a lack of dedicated or specialist services. A survey of 215 health professionals in England regarding epilepsy care found that less than one third had a dedicated clinic for people with intellectual disabilities (Frost *et al.*, 2003). A survey regarding how community learning disability nurses (CNLDs) work within National Health Service (NHS) trusts in England found that 25 (23%) had CNLDs specializing in epilepsy (Mobbs *et al.*, 2002). In a study of professionals and carers from the UK and Ireland, respondents stated that specialist expertise was available regionally only if consultants had a specific interest in the intellectual disability field (Thompson *et al.*, 2013). This geographical distribution of expertise is termed a 'postcode lottery' within the UK. The role of specialist nurses was favourably regarded, with a call that more of these posts were required to meet need. One study found that 34% of people with intellectual disabilities and epilepsy had seen an epilepsy nurse (Reuber *et al.*, 2008).

The impact of service setting

A small number of studies present information indicating that elements of service provision may have an impact on some outcomes for people with intellectual disabilities and epilepsy. In the US, living situation was found to be associated with adherence to AEDs, with non-adherence being 6% in group homes, 20% in semi-independent living, and 32% in family homes (Hom *et al.*, 2015). In Ireland, those in residential care were more likely to have a seizure diary (93%) than those living independently (44%) or in the community (78%) (McCarron *et al.*, 2014). A study in an English county found an association between polypharmacy and the degree of involvement of specialist intellectual disability services (Branford and Collacott, 1994). Further, those in the NHS were more likely to receive carbamazepine and those outside NHS more likely to receive phenytoin, phenobarbitone and primidone. In Wales, it was found that institutionalized individuals with intellectual disabilities and epilepsy had less inpatient, outpatient, and A&E admissions compared to those in the community (Morgan *et al.*, 2003). It is suggested that this may be because many of

their health needs are dealt with within the institution or there is a greater threshold before an event is deemed as requiring an acute admission.

Epilepsy Reviews

In an English city, 70.7% of those with intellectual disabilities and epilepsy were reported to have had an epilepsy treatment review within the last 12 months (Reuber *et al.*, 2008). Overall, 38.7% were reported to be under review in secondary care (neurologist, psychiatrist or epilepsy nurse), 52.9% stated that reviews were carried out in primary care alone and 8.9% said that epilepsy treatment had not been reviewed at all (Reuber *et al.*, 2008). In a sample of 229 people aged 40 or more with intellectual disability and epilepsy from Ireland, 80.8% had had an epilepsy review within the past 12 months, 5.1% within the past 2 years, 11.6% over 2 years ago, and 2.5% had never had their epilepsy reviewed (McCarron *et al.*, 2014). In this study, 51% reported that a GP reviewed their epilepsy, 40% a psychiatrist, 42% a neurologist and 34.7% more than one group/professional.

Epilepsy Care Plans

There is very little research regarding the use of epilepsy care plans. A study in England examined the case notes of 20 people with intellectual disabilities who had died from SUDEP of whom 65% had an Epilepsy Care Plan (Kiani *et al.*, 2014). In a survey on the use of rescue medication 84 (60%) participants had an epilepsy care plan (Mukhopadhyay *et al.*, 2010).

Investigations

A national audit into SUDEP highlighted poor access to investigation for those with intellectual disabilities (Hanna *et al.*, 2002). In a Welsh study, 42 (93.3%) of people with intellectual disabilities and epilepsy had some investigations undertaken, with all 42 having had an electroencephalogram (EEG) (Matthews *et al.*, 2008). Other investigation rates were low, particularly the use of neuro-imaging, despite the recommendations of clinical guidelines that such investigations should be

considered (Kerr *et al.*, 2001). Of 225 people with intellectual disabilities and epilepsy in an English city, 46.2% were reported to have had an EEG, and 41.3% a brain scan (Reuber *et al.*, 2008). Of 75 in-patients in a treatment and continuing care service, 9% had no record of an EEG trace having been performed (Tiffin and Perini, 2001). In some cases this appeared to be due to the inability of the patient to cooperate with the investigation but previous EEG records may have been lost for those admitted from outside the region. Only around 24% of patients with partial seizures received an MRI or CT scan, with failures due to poor cooperation being reported.

Seizure diaries

Medical intervention relies heavily on the accuracy of seizure frequency reporting (Clark *et al.*, 2001). A small number of studies give information relating to seizure diaries. Of people aged 40 or more with intellectual disability and epilepsy from Ireland, most respondents (83.1%) kept a record of their seizures, with those living in residential care (92.7%) more likely to have such a record than those living independently (43.5%) or in the community (77.8%) (McCarron *et al.*, 2014). A study in England of 20 people with intellectual disabilities who had died from SUDEP found seizure frequency documented in all case files except one, but sometimes only the approximate numbers of seizures (of all types) were documented, partly due to lack of access to the seizure diary during review (Kiani *et al.*, 2014). In an evaluation of an educational programme for people with mild intellectual disabilities and epilepsy, outcomes included increased understanding of the importance of seizure diaries, and increased knowledge regarding what, and when, to write in a seizure diary (Clark *et al.*, 2001).

Medication adherence

Non-adherence to AEDs in the general population has been associated with increased morbidity, SUDEP, increased health care costs, and increased number of emergency hospital visits, hospitalisations and injuries (cf. Hom *et al.*, 2015). In a US study, the non-adherence rate for AEDs

was 6% for group homes, 20% for semi-independent settings, and 32% for family homes (Hom *et al.*, 2015). The authors suggest that people with intellectual disabilities and their family members should be educated about the importance of medication adherence as they may not fully understand the consequences of non-adherence. As noted in relation to an evaluation of an educational programme for people with mild intellectual disabilities, addressing lack of knowledge may help to ensure medication compliance (Clark *et al.*, 2001).

Management by proxy

In relation to the pharmacological management of epilepsy for people with intellectual disabilities, it has been noted that they may be disadvantaged by a lack of self-advocacy (National Clinical Guideline Centre, 2012). A small number of articles contain information relevant to the issue of management by proxy. One study considered the role of parent-proxies in treatment decisions based on interviewing mothers of adults with intellectual disabilities and epilepsy (Redley *et al.*, 2013). It is suggested that they were willing to live with what they considered to be an acceptable level of seizure activity and to reject possible changes in their son or daughter's treatment. Two questions are raised. First, do mothers refuse suggested changes during consultations, and second do clinicians avoid going against their wishes? In one survey, professionals commented on: high turnover of paid caregivers who accompanied patients to consultations; variation in accounts of individuals' seizures reported by different paid caregivers: an absence of accurate records; and the fact that some paid caregivers did not know the person they supported to the degree required in a consultation (Thompson *et al.*, 2013). Similarly, in an audit related to the setting up of a multi-disciplinary specialist epilepsy clinic, it was noted that nonpaid carers gave a better account of the patient's history, seizure descriptions and seizure frequency whilst in some cases paid carers were unfamiliar with the patient and their epilepsy (Whitten and Griffiths, 2007).

Risk Assessment

NICE guidelines state that people with intellectual 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; and the suitability of independent living, where the rights of the child, young person or adult are balanced with the role of the carer (National Institute for Health and Care Excellence, 2012).

Little research regarding risk assessment was identified. Following the setting up of a multi disciplinary specialist epilepsy clinic, all patients had risks discussed and 68% had risks identified, with appropriate referrals being made (Whitten and Griffiths, 2007).

Two linked studies at a specialized epilepsy residential centre in the Netherlands examined decision-making in relation to risk for people with intellectual disability and epilepsy (Vallenga *et al.*, 2006, Vallenga *et al.*, 2008). Protective measures were often taken in situations of immediate threat, allowing no time to consider their negative effects, with consideration of these effects coming later (Vallenga *et al.*, 2006). Systematic recording and analysis of accidents was lacking. The authors suggest that practice could be improved by a methodical approach to risk management, including observation and recording of specific individual risks, accidents and the circumstances in which they occur, including pre-existing protective measures. In a subsequent study aiming to improve decision making regarding risk, assessment of client risk became more systematic and 'steps to an individual framework for decision-making' were developed as a tool for a systematic approach (Vallenga *et al.*, 2007).

Managing prolonged or serial seizures (rescue medication)

Rescue medication for managing prolonged or serial seizures can reduce the need for hospital admissions. Clinical guidelines suggest that any individual with intellectual disabilities and epilepsy should have an assessment of the management of prolonged or cluster seizures and where rescue

medication is prescribed a clear care plan should be completed (Kerr *et al.*, 2001). NICE guidelines recommend the administration of buccal midazolam as first-line treatment in children, young people and adults with prolonged or repeated seizures in the community, with administration of rectal diazepam if preferred or if buccal midazolam is not available (National Institute for Health and Care Excellence, 2012). Guidelines also state that buccal midazolam or rectal diazepam should only be prescribed for those who have had a previous episode of prolonged or serial convulsive seizures.

A number of studies consider the issue of rescue medication. In one English city, 15.6% of people with intellectual disabilities and epilepsy had to attend A&E at least once over the preceding year because seizures would not stop, and 5.3% attended more often (Reuber *et al.*, 2008). Of the 27.6% of respondents who had a supply of rescue medication at home, only 42.6% had received training from a nurse or a doctor on how to give this medication. Only 40% of carers of people with intellectual disabilities and epilepsy who had been to A&E over the last year with a prolonged seizure had access to emergency medication.

Two audits demonstrated increased recording regarding the use of rescue medication following the setting of standards (Whitten and Griffiths, 2007, Esan and Markar, 2010). In the latter study, it was noted that some patients had no one to administer it or there were training issues with staff involved. In relation to staff training, a survey of staffed homes in an English town in the non-health sector (private, voluntary or local authority) found that of 11 homes with a client with epilepsy, less than half had staff trained to administer emergency AEDs (Deepak *et al.*, 2012). Prescriptions written by the GP frequently did not include emergency AEDs. It is noted that whilst the NHS trust has policy guidelines on training staff in emergency AED use, this did not seem to have been implemented at the community level.

Buccal midazolam has been reported to be 'vastly superior' to rectal diazepam, being less likely to sedate patients, protecting their dignity, and being attributed with reducing the need to engage with emergency services (Thompson *et al.*, 2013). There was a general call for training in administration by family, paid caregivers, and GPs. In a community population of people with intellectual disabilities and epilepsy, the use of buccal midazolam was much more prevalent than the use of rectal diazepam (Mukhopadhyay *et al.*, 2010). Buccal midazolam was rated as better across a range of parameters: convenience of administration; invasiveness of procedure; gender issues; social acceptability; consent issues; and onset of action. An Australian study on the use of intranasal midazolam found that it was effective at controlling seizures, with no instances of respiratory arrest, and only one report of apparent shallow breathing (Kyrkou *et al.*, 2006). However, the authors stress that adequate training is required in its use.

One study evaluated individual epilepsy guidelines (IEG's) which gave directions for administering emergency medication for people with intellectual disabilities and refractory epilepsy living in the community (Cole *et al.*, 2009). These were produced in response to concerns that in community settings many carers had difficulty recognising when rectal diazepam should be administered. Carers found the IEG's easy to understand and were more informed about administering rectal diazepam as a result of the guidelines. IEG's could be viewed as a means of minimising patient/carers anxieties and stress during emergency events and reduce the need for hospital admissions with possible cost savings for the NHS. Guidelines were also going to be developed for the use of buccal midazolam.

Educating people with intellectual disabilities about their epilepsy

People with intellectual disabilities and epilepsy may benefit from education regarding the management of their condition. There has been little research on this issue. A Scottish study has evaluated a video assisted educational package 'Epilepsy and You' for people with mild intellectual

disabilities and epilepsy (Clark *et al.*, 2001). Epilepsy and You' involved participants participating in three, weekly sessions each lasting one hour. Participants increased greatly in their knowledge about what an EEG is and the importance of seizure diaries, with increased knowledge about what, and when, to write in a seizure diary. The authors suggest that addressing lack of knowledge may help in relation to ensuring medication compliance and prevent the generation of false and distressing beliefs regarding epilepsy. Two other articles concerning educating people with intellectual disabilities and epilepsy, which did not meet the inclusion criteria for the review, are discussed in the discussion section (Codling, 2010, Durand *et al.*, 2014).

Evaluations of initiatives in service provision

In an English county, an intellectual disabilities psychiatric service was established after a five year period without one (Beber *et al.*, 1999). At baseline, most received GP care only for their epilepsy. Intellectual disabilities psychiatric care was found to be associated with reductions in seizure frequency, drug side-effects and polypharmacy. Those receiving intellectual disabilities psychiatric care were more likely to have: type of seizure determined; medical reviews; appropriate blood test monitoring; active interventions to improve seizure control; and to have unnecessary drugs withdrawn. Some of those not under intellectual disabilities psychiatric care tended just to receive repeat prescriptions for their epilepsy.

In one area of England, an audit of adherence to standards based on NICE guidelines was conducted in a specialist epilepsy clinic for people with intellectual disabilities (Esan and Markar, 2010). At initial audit, 11 of 21 standards were met and the following recommendations were made: all patient records kept in one file; user friendly leaflet developed giving information on medication changes, care plan, contacts with clinic and next appointment date; NICE information booklet given to patients and carers after appointments; checklist developed detailing main areas that needed

improvement to serve as reminder to psychiatrist at clinic; standardised risk assessment form developed to complete and share with carers or other agencies; leaflet on SUDEP developed. The subsequent audit found an improvement in meeting standards which was of benefit to both service users and clinicians.

In another area of England NICE guidelines were implemented through the introduction of a specialist multi-disciplinary epilepsy clinic (Whitten and Griffiths, 2007). There were improvements to seizure assessments, matching of medication to seizure type, patient care and epilepsy management. All patients prescribed three or more AEDs received a medication reduction plan and all patients received an individualised risk assessment. In Wales, audit was used to look at the impact of standard setting in a clinic for people with intellectual disabilities and epilepsy (Chubb *et al.*, 1995). There was improvement in several areas of out-patient management but the recording of side effects and comments on quality of life were poor both before and after audit.

A study in Belgium looked at the introduction of a decision tree, for non-medically trained staff in two establishments for adults with intellectual disabilities, which treats seizures as medical emergencies (Peeters, 2000). Treatment of seizures as medical emergencies reduced frequency, duration, and severity (in comparison with results from a retrospective review). No patient was hospitalized. The authors note that use of the decision tree could also be taught to caregivers and parents.

An early study in the United States looked at outcomes for 15 people with severe to profound intellectual disabilities and complex epilepsy who were moved from an institution to specially built community based group homes despite the expression of concerns that the needs of this medically fragile group could not be met in the community (Litzinger *et al.*, 1993). It was found that simplification of anticonvulsants, early intervention for seizures, and improved staff education resulted in fewer seizures, decreases in emergency room visits for status seizures and drug-related

side effects, and increased levels of functioning as measured by activities of daily living. The success of the community placement was attributed by authors to the availability and proper usage of the new generation of AEDs.

A study in an area of England piloted three care pathways (Ahmad *et al.*, 2007), of which one was for epilepsy (cf. Ahmad *et al.*, 2002). The percentage attained for training offered to clients and carers, and results sent to the GP, increased after the introduction of the epilepsy care pathway.

Prescribing Practices

Polypharmacy was common reflecting difficult to treat epilepsy in this population. Of 183 people with intellectual disabilities and epilepsy in one geographic area of England, 36% were being treated with two AEDs and 23% were prescribed three or more AEDs (Ring *et al.*, 2009). In Wales, of 57 participants 40% were prescribed two AEDs, 9% three AEDs and 4% four AEDs (Matthews *et al.*, 2008). It was noted that whilst polypharmacy was common, this and dosage were related to higher seizure frequency, which may reflect an appropriate response to the morbidity. One study suggests that some people may benefit from a reduction in the number of AEDs prescribed. In an audit of AED use in adult in-patients with intellectual disabilities in a treatment and continuing care service, 13 (17%) were identified as potentially benefiting from a reduction in the number of AEDs they were prescribed (Tiffin and Perini, 2001).

In an early study of people with intellectual disabilities who were receiving AEDs in one English county, no difference in polypharmacy was found between those in NHS and community facilities, but there was an association between polypharmacy and the degree of involvement of specialist intellectual disability services (Branford and Collacott, 1994). There were differences in the choice of AEDs with those outside the NHS more likely to receive phenytoin, phenobarbitone & primidone which have been shown to have particularly poor effects on cognitive performance. A follow-up

study found continued reduction of the prescribing of phenobarbitone and increased prescribing of sodium valproate and carbamazepine (Branford *et al.*, 1998).

A theoretical concern is that patients with intellectual disabilities might not be offered treatment with newer AEDs as often as other patients, potentially due to a lack of advocacy in favour of using newer AEDs by patients with intellectual disabilities or their caregivers (Fridhandler *et al.*, 2012). A recent Canadian study looked at AED prescribing practices for patients with and without intellectual disabilities in a tertiary epilepsy center (Fridhandler *et al.*, 2012). There was no evidence for a discrepancy in access to new AEDs in patients with intellectual disabilities. Similarly, the authors of one study suggest that over the years the pharmacological management of epilepsy in people with intellectual disabilities has been modified according to emerging changes in clinical practice recommendations (Ring *et al.*, 2009). However, a survey in an English city found that only one in 10 patients under primary care review was taking one of the “newer” AEDs (Reuber *et al.*, 2008). Finally, in an Irish study, a number of people who reported a history of seizure activity were not taking medication despite experiencing seizures, with this group tending to be those with mild or moderate intellectual disabilities who lived in the community or independently (McCarron *et al.*, 2014).

Views of families, carers or professionals regarding services

A qualitative study explored the views of carers and professionals from the UK and Ireland into the management of people with intellectual disabilities and epilepsy (Thompson *et al.*, 2013). In relation to ‘medical care and services’, some respondents argued that the medical profession had lower expectations of whether seizure freedom was possible for this population. Respondents, largely family members, commented on the ‘trade-off’ between the goal of achieving seizure control on the one hand and the impact of side effects from medication on the other hand. Some family members reported their distress at the price paid in side effects in order to gain seizure control.

A small number of professionals stated that caregivers should be better informed about the 'blind target' to eliminate seizures given the negative impact treatment may have on the individual's overall quality of life.

A US study identified sources of stress for parents of children with both epilepsy and intellectual disability (Buelow *et al.*, 2006). Most seemed fairly happy with their children's healthcare providers, but did discuss concerns about medications and a lack of information regarding their child's illness. Whilst some complained about medications and side effects, they did not have suggestions as to actions healthcare providers could take regarding seizure treatment.

In an audit on communication between GPs and psychiatrists following an initial outpatient assessment of patients with intellectual disabilities, 30 GPs responded to a survey and one of the comments made on how to improve the service was that psychiatrists should take over the management of epilepsy (Markar and Mahadeshwar, 1998).

An international online survey obtained responses regarding service delivery for people with intellectual disability and epilepsy from professionals and carers, mainly from the UK (Kerr *et al.*, 2014). There were calls for: greater resources including multidisciplinary team approaches; specialist nursing; provision of local services; consistency in quality of service provision; and greater recognition of family. There were concerns regarding lack of expertise in primary care; brief and ineffective consultations. Calls were made for greater use of epilepsy nurse specialists in training.

Discussion

Despite the broad inclusion criteria which allowed for the identification of research on disparate topics within the broad area of service responses to epilepsy in people with intellectual disabilities,

no RCTs or studies employing similarly robust designs were identified. The articles identified consisted mainly of small scale surveys, audits, and qualitative studies. Nonetheless, the findings of this review elucidate key issues emerging from research in this area.

Descriptions of current service provision suggest that access to specialists is inconsistent, with geographical distribution of expertise being termed a 'postcode lottery' within the UK (Thompson *et al.*, 2013). In addition, a report based on a survey in England found that 49% (40/79) of acute and foundation trusts did not offer patients access to intellectual disability epilepsy specialist nurses (Epilepsy Action, 2013). A report based on a survey undertaken under the auspices for the Intellectual Disability Task Force of the International League against Epilepsy (ILAE) and the International Bureau for Epilepsy (IBE) found that an overarching theme across many responses was an endorsement for specialist care, ranging from epilepsy nurse specialists through to multidisciplinary teams (Kerr *et al.*, 2013).

Despite calls for specialist care, there was very little evidence in relation to the relative efficacy of different models of service provision although there was some indication that service settings and service provision may make a difference to outcomes for people with intellectual disabilities and epilepsy. Further evidence is crucially needed in this area. In this respect, there is an ongoing cluster randomised controlled trial which is aiming to establish whether nurses with expertise in epilepsy and intellectual disabilities can improve clinical and quality of life outcomes in the management of epilepsy in adults with intellectual disabilities compared to treatment as usual (Ring *et al.*, 2014).

In some areas, the lack of research is surprising. There is very little research regarding the use of epilepsy care plans, seizure diaries, or risk assessments. There is some research with regards to managing prolonged or serial seizures which generally supports the usefulness of rescue medication

(in particular buccal midazolam). However, one issue was the lack of suitably trained staff or carers to administer such medications (Deepak *et al.*, 2012, Esan and Markar, 2010). Only one evaluation on the impact of a care pathway for people with intellectual disabilities and epilepsy was identified (Ahmad *et al.* 2007), although a care pathway has also been developed in an English county (Kiani *et al.* 2014).

One evaluation of an educational package for people with mild intellectual disabilities and epilepsy met the inclusion criteria (Clark *et al.*, 2001). However, it is evident that other educational programmes have been developed, but research evaluating their use is currently limited. For example, an English version of a German multi-media programme (PEPE: a psycho-educative programme about epilepsy for people with intellectual disabilities) designed to give people with intellectual disabilities and epilepsy information and support has been developed in association with the National Society for Epilepsy (Kushinga, 2007). However, there have been relatively few initiatives that attempt to measure the effectiveness of this resource (Codling, 2010). One study (which has not been included in the main review as peer review status was not determined), which used the PEPE programme alongside other methods such as group discussions and role-play, found that the groups had an impact on the ability of people with intellectual disabilities to manage their epilepsy, including an increased awareness amongst people with intellectual disabilities about risk (Codling, 2010). Group sessions highlighted anxiety caused by interpretation of information given by health staff. For instance one member was informed that people can die from epilepsy and so they became frightened to go to sleep, leading to increased seizures. One feasibility study was identified which is being used to inform the design and methodology of a study to determine the impact of an intervention to improve the management of epilepsy in people with intellectual disabilities based on a 'Books Beyond Words' booklet that uses images to help people with intellectual disabilities manage their epilepsy (Durand *et al.*, 2014). Finally, evaluations of training initiatives for staff have not been identified although such initiatives exist (Sterrick and Foley, 1999, Pointu and Cole, 2005).

In addition, a number of relevant measures have been developed, but there is little research evaluating their use. For example, the epilepsy risk awareness checklist (ERAC) provides a measure of risk, and initial research suggests that it is a useful tool in the care of people with intellectual disability and epilepsy (Cole *et al.*, 2010). The Epilepsy and Learning Disabilities Quality of Life scale (ELDQOL) has been reported to be a promising instrument for assessing quality of life in children or young adults with epilepsy and intellectual disabilities (Buck *et al.*, 2007). The Glasgow Epilepsy Outcome Scale (GEOS; GEOS-35 and GEOS-90 versions) has been developed to measure concerns of family carers, staff carers and clinicians for people with intellectual disabilities and epilepsy (Espie *et al.*, 2003, Espie *et al.*, 2001). A client version of the GEOS (GEOS-C) has also been developed for use with people with mild to moderate intellectual disabilities to complement existing GEOS measures (Watkins *et al.*, 2006). Development of the GEOS-C revealed some qualitatively different concerns to those of family carers, staff carers and clinicians, such as wetting self during seizure, feeling embarrassed if having a fit when out, and people calling me names when I have a fit. The existing scales may have useful applications in both research and practice.

Further research is also being carried out on the identification of situations associated with increased or decreased seizure likelihood in people with intellectual disabilities due to the potential for informing nonpharmacological approaches to seizure management that may provide a useful adjunct to AED treatment (Illingworth *et al.*, 2014).

Limitations

There are a number of limitations to this review. A main weakness is the potential loss of information from two main sources due to the focus on studies relating to the general population of people with intellectual disabilities. Firstly, the review has not considered how studies regarding services approaches to epilepsy in general may be applicable to people with intellectual disabilities,

and indeed some of these studies may include information relating to intellectual disabilities. Secondly, the review does not consider research relating to specific syndromes associated with intellectual disabilities such as Lennox Gastaut or Dravet syndrome. These sources of information remain to be reviewed. In addition, whilst studies were identified from a range of countries, the review is restricted to English language publications. No information was identified regarding services responses to people with intellectual disabilities and epilepsy in low and middle income (LAMI) countries. Indeed, the majority of studies were conducted in the UK. Further, whilst no publications prior to 1990 were included, descriptions of service provision or initiatives from older articles based on historically prevalent models of service provision may not be as relevant as more current studies. Finally, all data was extracted by one reviewer and extraction of data by two reviewers independently would have reduced the possibility of errors.

Conclusion

Research into service responses to epilepsy in people with intellectual disabilities is at an embryonic stage. Whilst initiatives to improve service responses to people with intellectual disabilities and epilepsy exist, evaluations of such initiatives are lacking. Further research is needed to substantiate some of the findings reported here and the results of ongoing research will be a welcome addition to the evidence base. It is important that services are equipped with the information and skills needed to manage epilepsy in this population. A recent report provides information on reasonable adjustments that can be made to improve epilepsy care for people with intellectual disabilities (Marriott *et al.*, 2014). The ideas, information and examples of good practice in relation to reasonable adjustments provided within this report should help services improve provision for this highly prevalent condition and potentially reduce the excess deaths associated with epilepsy in people with intellectual disabilities.

Appendix One: Electronic Search Strategy

CINAHL (searched 20.1.15)

Limits: English, Human, Peer reviewed, published from 1990.

Results 176

(TI (learning N1 (disab* or difficult* or handicap*)) OR TI (mental* N1 (retard* or disab* or deficien* or handicap*)) OR TI (intellectual* N1 (disab* or impair* or handicap*)) OR TI development* N1 disab* OR TI (multipl* N1 (handicap* or disab*)) OR TI "Down* syndrome" OR (MH "Developmental Disabilities") OR (MH "Intellectual Disability+") OR (MH "mentally disabled persons")) OR (AB (learning N1 (disab* or difficult* or handicap*)) OR AB (mental* N1 (retard* or disab* or deficien* or handicap*)) OR AB (intellectual* N1 (disab* or impair* or handicap*)) OR AB development* N1 disab* OR AB (multipl* N1 (handicap* or disab*)) OR AB "Down* syndrome")

AND

(MH "Epilepsy+/CO/DI/DH/ED/NU/PC/PR/PF/RH/RF/TH") OR (TI epilep* OR TI seizure* OR AB epilep* OR AB seizure*)

AND

AB training OR AB (care n1 (plan* or manag* or pathway)) OR AB (risk n1 (manag* or assess*)) OR AB (medic* n1 (emergency or rescue)) OR AB (diar* n1 (seizure or epilep*)) OR AB (medic* n1 (compliance or adherence)) OR AB guidelines OR AB (review n1 (epilep* or annual)) OR AB (manag* n1 (seizure or epilep*)) OR TI training OR TI (care n1 (plan* or manag* or pathway)) OR TI (risk n1 (manag* or assess*)) OR TI (medic* n1 (emergency or rescue)) OR TI (diar* n1 (seizure or epilep*)) OR TI (medic* n1 (compliance or adherence)) OR TI guidelines OR TI (review n1 (epilep*

or annual)) OR TI (manag* n1 (seizure or epilep*)) OR (MH "Health Services Administration+") OR (MH "Health Services+")

Medline (searched 20.1.15)

Limits: English, Human, published from 1990.

Results 1164

As Cinahl (above) with amended subheadings as follows:

(MH "Epilepsy+/CO/DI/DH/NU/PC/PX/RH/TH")

PsycINFO (searched 21.1.15)

Limits: peer reviewed, human, English language, published from 1990, exclude dissertations.

Results 184

((((((DE "Social Services" OR DE "Community Services" OR DE "Outreach Programs" OR DE "Protective Services"))) OR (DE "Health Care Services" OR DE "Continuum of Care" OR DE "Long Term Care" OR DE "Mental Health Services" OR DE "Primary Health Care"))) OR (DE "Health Care Administration" OR DE "Hospital Administration"))) OR (DE "Caregivers")) OR (DE "Managed Care" OR DE "Health Maintenance Organizations" OR DE "Mental Health Programs" OR DE "Deinstitutionalization" OR DE "Home Visiting Programs" OR DE "Quality of Care" OR DE "Quality of Services" OR DE "Quality of Care") OR (AB training OR AB (care n1 (plan* or manag* or pathway)) OR AB (risk n1 (manag* or assess*)) OR AB (medic* n1 (emergency or rescue)) OR AB (diar* n1 (seizure or epilep*)) OR AB (medic* n1 (compliance or adherence)) OR AB guidelines OR AB (review n1 (epilep* or annual)) OR AB (manag* n1 (seizure or epilep*)) OR TI training OR TI (care n1 (plan* or manag* or pathway)) OR TI (risk n1 (manag* or assess*)) OR TI (medic* n1 (emergency or rescue)) OR TI (diar* n1 (seizure or epilep*)) OR TI (medic* n1 (compliance or adherence)) OR TI guidelines OR TI (review n1 (epilep* or annual)) OR TI (manag* n1 (seizure or epilep*))

AND

DE "Intellectual Development Disorder" OR DE "mental retardation" OR DE "developmental disabilities" OR (TI (learning N1 (disab* or difficult* or handicap*)) OR TI (mental* N1 (retard* or disab* or deficien* or handicap* or disorder*)) OR TI (intellectual* N1 (disab* or impair* or handicap*)) OR TI development* N1 disab* OR TI (multipl* N1 (handicap* or disab*)) OR TI "Down* syndrome") OR AB (mental* N1 (retard* or disab* or deficien* or handicap* or disorder*)) OR AB (intellectual* N1 (disab* or impair* or handicap*)) OR AB development* N1 disab* OR AB (multipl* N1 (handicap* or disab*)) OR AB "Down* syndrome"

AND

DE "Epilepsy" OR DE "Epileptic Seizures" OR (DE "Seizures" OR DE "Audiogenic Seizures" OR DE "Epileptic Seizures" OR DE "Grand Mal Seizures" OR DE "Petit Mal Seizures" OR DE "Status Epilepticus") OR (TI epilep* OR TI seizure* OR AB epilep* OR AB seizure*)

Web of Science (searched 21.1.15)

Limits: English language; published from 1990; articles.

Results 337.

learning NEAR/1 (disab* or difficult* or handicap*) OR mental* NEAR/1 (retard* or disab* or deficien* or handicap*) OR intellectual* NEAR/1 (disab* or impair* or handicap*) OR development* NEAR/1 disab* OR multipl* NEAR/1 (handicap* or disab*) OR "Down* syndrome"

(In TOPIC which includes title, abstract, keywords)

AND

Epilep* or seizure*

AND

training OR care NEAR/1 (plan* or manag* or pathway) OR risk NEAR/1 (manag* or assess*) OR medic* NEAR/1 (emergency or rescue) OR diar* NEAR/1 (seizure or epilep*) OR medic* NEAR/1

(compliance or adherence) OR guidelines OR review NEAR/1 (epilep* or annual) OR manag* NEAR/1
(seizure or epilep*) OR service*

Clinical Trials Registers (searched 28.9.15)

ClinicalTrials.gov (U.S. National Institutes of Health): Intellectual and epilepsy (15 results, 0 selected); "Mental retardation" and epilepsy (5 results, 0 selected)

Cochrane Central Register of Controlled Trials (CENTRAL): Intellectual and epilepsy (56 results, 0 selected); "Mental retardation" and epilepsy (26 results, 0 selected)

ISRCTN registry (<http://www.controlled-trials.com/>): Intellectual (text) and epilepsy (condition) (2 results, both selected, both ongoing trials); Mental retardation (text) and epilepsy (condition) (5 results, 0 selected)

UK Clinical Trials Gateway: Intellectual and epilepsy (11 results, 2 selected, both already identified in ISRCTN search); Mental retardation and epilepsy (15 results, 0 selected)

References

- Ahmad F., Bissaker S., De Luc K., Pitts J., Brady S., Dunn L. & Ashok R. (2002) Partnership for Developing Quality care pathway initiative for people with learning disabilities. Part iic: epilepsy. *Journal of Integrated Care Pathways*, 6, 90-93.
- Ahmad F., Roy A., Brady S., Belgeonne S., Dunn L. & Pitts J. (2007) Care pathway initiative for people with intellectual disabilities: impact evaluation. *Journal of Nursing Management*, 15, 700-702.
- Balogh R., Brownell M., Ouellette-Kuntz H. & Colantonio A. (2010) Hospitalisation rates for ambulatory care sensitive conditions for persons with and without an intellectual disability-a population perspective. *Journal of Intellectual Disability Research*, 54, 820-832.
- Baribeault J. J. (1996) Clinical advocacy for persons with epilepsy and mental retardation living in community-based programs. *Journal of Neuroscience Nursing*, 28, 359-372.
- Bashir C. I., Kerr M. & Isaac A. (2013) The effect of pharmacological and non-pharmacological interventions for epilepsy in people with Down syndrome: Protocol. Cochrane Library 2013 Issue 8.
- Beavis J., Kerr M., Marson Anthony G. & Dojcinov I. (2007a) Non-pharmacological interventions for epilepsy in people with intellectual disabilities (Assessed as up to date 2011). *Cochrane Database of Systematic Reviews*, Issue 4. Art. No.: CD005502. DOI: 10.1002/14651858.CD005502.pub2.
- Beavis J., Kerr M., Marson Anthony G. & Dojcinov I. (2007b) Pharmacological interventions for epilepsy in people with intellectual disabilities (Assessed as up to date 2011). *Cochrane Database of Systematic Reviews*, Issue 3. Art. No.: CD005399. DOI: 10.1002/14651858.CD005399.pub2.
- Beber E., Bailey N. M. & Cooper S.-A. (1999) Health gain for epilepsy associated with learning disabilities psychiatric care. *Irish Journal of Psychological Medicine*, 16, 46-50.
- Beran R. G. & McAulley R. (1992) Nurses' knowledge attitudes, preparation and skills for serving people with epilepsy and developmental disability. *Australia & New Zealand Journal of Developmental Disabilities*, 18, 9-16.
- Bowley C. & Kerr M. (2000) Epilepsy and intellectual disability. *Journal of Intellectual Disability Research*, 44, 529-543.
- Bradley Peter M. & Lindsay B. (2008) Care delivery and self-management strategies for adults with epilepsy. *Cochrane Database of Systematic Reviews*, Issue 1. Art. No.: CD006244. DOI: 10.1002/14651858.CD006244.pub2.
- Branford D., Bhaumik S., Duncan F. & Collacott R. A. (1998) A follow-up study of adults with learning disabilities and epilepsy. *Seizure - European Journal of Epilepsy*, 7, 469-472.
- Branford D. & Collacott R. A. (1994) Comparison of community and institutional prescription of antiepileptic drugs for individuals with learning disabilities. *Journal of Intellectual Disability Research*, 38, 561-566.
- Buck D., Smith M., Appleton R., Baker G. A. & Jacoby A. (2007) The development and validation of the Epilepsy and Learning Disabilities Quality of Life (ELDQOL) scale. *Epilepsy and Behavior*, 10, 38-43.
- Buelow J. M., McNelis A., Shore C. P. & Austin J. K. (2006) Stressors of parents of children with epilepsy and intellectual disability. *Journal of Neuroscience Nursing*, 38, 147-154.
- Burke T. A., McKee J. R., Pathak D. S., Donahue R. M., Parasuraman T. V. & Batenhorst A. S. (1999) Costs of epilepsy in an intermediate care facility for persons with mental retardation. *American Journal Of Mental Retardation*, 104, 148-157.
- Camfield P. R., Gibson P. A. & Douglass L. M. (2011) Strategies for transitioning to adult care for youth with Lennox-Gastaut syndrome and related disorders. *Epilepsia*, 52 Suppl 5, 21-27.

- Chapman M., Iddon P., Atkinson K., Brodie C., Mitchell D., Parvin G. & Willis S. (2011) The misdiagnosis of epilepsy in people with intellectual disabilities: a systematic review. *Seizure - European Journal of Epilepsy*, 20, 101-106.
- Chubb H., Kerr M. & Joyce J. (1995) Out-patient care for people with learning disability and epilepsy: evaluating the audit process. *Psychiatric Bulletin*, 19, 686-688.
- Clark A. J., Espie C. A. & Paul A. (2001) Adults with learning disabilities and epilepsy: knowledge about epilepsy before and after an educational package. *Seizure - European Journal of Epilepsy*, 10, 492-499.
- Codling M. (2010) The ME (My epilepsy) Project: an active learning approach for people with learning disabilities and epilepsy. *Epilepsy Care*, 2, 3-7.
- Cole C., Pointu A., Mahadeshwar S. & Dudley A. (2009) Community survey of carer's: individual epilepsy guidelines (IEG) for rescue medication. *Seizure - European Journal of Epilepsy*, 18, 220-224.
- Cole C., Pointu A., Wellsted D. M. & Angus-Leppan H. (2010) A pilot study of the epilepsy risk awareness checklist (ERAC) in people with epilepsy and learning disabilities. *Seizure - European Journal of Epilepsy*, 19, 592-596.
- Deepak S., Obe E. & Attavar R. (2012) Administration of emergency antiepileptics in staffed care homes for people with intellectual disabilities in High Wycombe: Survey of staff training status. *Advances in Mental Health and Intellectual Disabilities*, 6, 41-46.
- Durand M. A., Gates B., Parkes G., Zia A., Friedli K., Barton G., Ring H., Oostendorp L. & Wellsted D. (2014) Wordless intervention for epilepsy in learning disabilities (WIELD): study protocol for a randomized controlled feasibility trial. *Trials*, 15:455.
- Epilepsy Action (2013) A Critical Time for Epilepsy in England. Epilepsy Action. Available online at: <https://www.epilepsy.org.uk/sites/epilepsy/files/campaigns/ACT/Epilepsy%20Action%20-%20A%20Critical%20Time%20%282013%29.pdf>. (accessed 10th February 2015).
- Esan F. & Markar N. (2010) Audit on adherence to NICE guidelines in a specialist epilepsy clinic for people with learning disabilities. *British Journal of Developmental Disabilities*, 56, 57-66.
- Espie C. A., Watkins J., Duncan R., Espie A., Sterrick M., Brodie M. J., McGarvey C. & Curtice L. (2001) Development and validation of the Glasgow Epilepsy Outcome Scale (GEOS): a new instrument for measuring concerns about epilepsy in people with mental retardation. *Epilepsia*, 42, 1043-1051.
- Espie C. A., Watkins J., Duncan R., Sterrick M., McDonach E., Espie E. & McGarvey C. (2003) Perspectives on epilepsy in people with intellectual disabilities: comparison of family carer, staff carer and clinician score profiles on the Glasgow Epilepsy Outcome Scale (GEOS). *Seizure - European Journal of Epilepsy*, 12, 195-202.
- Forsgren L., Beghi E., Öun A. & Sillanpää M. (2005) The epidemiology of epilepsy in Europe - a systematic review. *European Journal of Neurology*, 12, 245-253.
- Fridhandler J. D., Coelho F. M., Tai P., Jette N. & Andrade D. M. (2012) A comparison of antiepileptic drug therapy in patients with severe intellectual disability and patients with normal intellect. *Epilepsy & Behavior: E&B*, 25, 196-199.
- Frost S., Crawford P., Mera S. & Chappell B. (2003) Implementing good practice in epilepsy care. *Seizure - European Journal of Epilepsy*, 12, 77-84.
- Glover G. & Ayub M. (2010) How People with Learning Disabilities Die. Available online at <http://www.improvinghealthandlives.org.uk/gsf.php5?f=8586&fv=9033> (accessed 25 November 2014).
- Glover G. & Evison F. (2013) Hospital Admissions That Should Not Happen: Admissions for Ambulatory Care Sensitive Conditions for People with Learning Disabilities in England. Available online at www.ihal.org.uk/gsf.php5?f=16714 (accessed 21st April 2015).
- Hanna N., Black M., Sander J., Smithson W., Appleton R., Brown S. & Fish D. (2002) The National Sentinel Clinical Audit of Epilepsy-Related Death: Epilepsy – death in the shadows. The Stationery Office. Available at:

- <https://www.sudep.org/files/sudepaction/nationalsentinelreport1.pdf> (accessed 22 July 2014).
- Hom C. L., Touchette P., Nguyen V., Fernandez G., Tournay A., Plon L., Himber P. & Lott I. T. (2015) The relationship between living arrangement and adherence to antiepileptic medications among individuals with developmental disabilities. *Journal of Intellectual Disability Research*, 59, 48-54.
- Illingworth J. L., Watson P. & Ring H. (2014) Why do seizures occur when they do? Situations perceived to be associated with increased or decreased seizure likelihood in people with epilepsy and intellectual disability. *Epilepsy & Behavior*, 39, 78-84.
- Joint Epilepsy Council (2011) Epilepsy prevalence, incidence and other statistics. Available at: <http://www.jointepilepsycouncil.org.uk/downloads/2011/Joint%20Epilepsy%20Council%20Prevalence%20and%20Incidence%20September%202011.pdf> (retrieved 22 July 2014).
- Kerr M., Linehan C., Thompson R., Glynn M., Mula M., Gil-Nagel A. & Zuberi S. (2013) Listening for a change: the medical and social needs of people with epilepsy and intellectual disability. International League Against Epilepsy, Available at: <http://www.ilae.org/visitors/documents/ListeningForChange-2013.pdf> (accessed 28th January 2015).
- Kerr M., Linehan C., Thompson R., Mula M., Gila-Nagal A., Zuberi S. M. & Glynn M. (2014) A White Paper on the medical and social needs of people with epilepsy and intellectual disability: The Task Force on Intellectual Disabilities and Epilepsy of the International League Against Epilepsy. *Epilepsia*, 55, 1902-1906.
- Kerr M., Scheepers M., Arvio M., Beavis J., Brandt C., Brown S., Huber B., Iivanainen M., Louise A. C., Martin P., Marson A. G., Prasher V., Singh B. K., Veendrick M. & Wallace R. A. (2009) Consensus guidelines into the management of epilepsy in adults with an intellectual disability. *Journal of Intellectual Disability Research*, 53, 687-694.
- Kerr M., Scheepers M., Besag F., Bowley C., Brown S., Espie C., Foley J., Paul A., Webb J. O., Ahmed Z., Baker G., Betts T., Bjorkman M., Cornaggia C., Iivanainen M., Wallace R., Kaski M., Koot H., Marson A., Mohammed N., Prasher V. & Visser F. (2001) Clinical guidelines for the management of epilepsy in adults with an intellectual disability. *Seizure - European Journal of Epilepsy*, 10, 401-409.
- Kerr M. P., Mensah S., Besag F., de Toffol B., Ettinger A., Kanemoto K., Kanner A., Kemp S., Krishnamoorthy E., LaFrance W. C., Jr., Mula M., Schmitz B., van Elst L. T., Trollor J. & Wilson S. J. (2011) International consensus clinical practice statements for the treatment of neuropsychiatric conditions associated with epilepsy. *Epilepsia*, 52, 2133-2138.
- Kiani R., Tyrer F., Jesu A., Bhaumik S., Gangavati S., Walker G., Kazmi S. & Barrett M. (2014) Mortality from sudden unexpected death in epilepsy (SUDEP) in a cohort of adults with intellectual disability. *Journal of Intellectual Disability Research*, 58, 508-520.
- Kushinga K. (2007) The PEPE project: trialling a German epilepsy education programme for English use. *Learning Disability Practice*, 10, 10-13.
- Kyrkou M., Harbord M., Kyrkou N., Kay D. & Coulthard K. (2006) Community use of intranasal midazolam for managing prolonged seizures. *Journal of Intellectual & Developmental Disability*, 31, 131-138.
- Linehan C., Kerr M. P., Walsh P. N., Brady G., Kelleher C., Delanty N., Dawson F. & Glynn M. (2010) Examining the prevalence of epilepsy and delivery of epilepsy care in Ireland. *Epilepsia*, 51, 845-852.
- Litzinger M. J., Duvall B. & Little P. (1993) Movement of individuals with complex epilepsy from an institution into the community: Seizure control and functional outcomes. *American Journal on Mental Retardation*, 98, 52-57.
- Magill N., Ridsdale L., Goldstein L. H., McCrone P., Morgan M., Noble A. J., Baker G., Richardson M., Taylor S. & Landau S. (2015) Self-management education for adults with poorly controlled

- epilepsy (SMILE (UK)): statistical, economic and qualitative analysis plan for a randomised controlled trial. *Trials*, 16, 269-269.
- Markar T. N. & Mahadeshwar S. (1998) Audit on communication between general practitioners and psychiatrists following an initial outpatient assessment of patients with learning disabilities. *British Journal of Developmental Disabilities*, 44, 38-41.
- Marriott A., Turner S., Hatton C., Glover G. & Robertson J. (2014) Making reasonable adjustments to epilepsy services for people with learning disabilities. Available on-line at <http://www.improvinghealthandlives.org.uk/gsf.php5?f=313318&fv=20779> (accessed 13 November 2014).
- Matthews T., Weston N., Baxter H., Felce D. & Kerr M. (2008) A general practice-based prevalence study of epilepsy among adults with intellectual disabilities and of its association with psychiatric disorder, behaviour disturbance and carer stress. *Journal of Intellectual Disability Research*, 52, 163-173.
- McCarron M., O'Dwyer M., Burke E., McGlinchey E. & McCallion P. (2014) Epidemiology of epilepsy in older adults with an intellectual disability in Ireland: associations and service implications. *American Journal On Intellectual And Developmental Disabilities*, 119, 253-260.
- Mobbs C., Hadley S., Wittering R. & Bailey N. M. (2002) An exploration of the role of the community nurse, learning disability, in England. *British Journal of Learning Disabilities*, 30, 13-18.
- Morgan C. L., Baxter H. & Kerr M. P. (2003) Prevalence of epilepsy and associated health service utilization and mortality among patients with intellectual disability. *American Journal Of Mental Retardation*, 108, 293-300.
- Mukhopadhyay A., Harsha G., da Costa E., Anderton S. & Karki C. (2010) A survey of carers' views on the use of rescue medication in people with intellectual disability having prolonged seizures: Are the winds of change blowing? *British Journal of Developmental Disabilities*, 56, 11-18.
- National Clinical Guideline Centre (2012) The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care. Pharmacological update of clinical guideline 20. National Clinical Guideline Centre, London.
- National Institute for Health and Care Excellence (2012) The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care. NICE Clinical guideline 137. National Institute for Health and Care Excellence, London.
- Noble A., McCrone P., Seed P., Goldstein L. H. & Ridsdale L. (2014) Clinical- and cost-effectiveness of a nurse led self-management intervention to reduce emergency visits by people with epilepsy. *Plos One*, 9, e90789.
- Peeters E. (2000) Treatment of epileptic seizures as medical emergencies: a prospective analysis of a decision tree for nonmedically trained staff. *Seizure - European Journal of Epilepsy*, 9, 473-479.
- Pennington M., Prince E., Bateman N., Gray J., Croudace T. J., Redley M., Wood N. & Ring H. (2012) Factors influencing the costs of epilepsy in adults with an intellectual disability. *Seizure - European Journal of Epilepsy*, 21, 205-210.
- Pointu A. & Cole C. (2005) An education programme for social care staff: Improving the health of people who have a learning disability and epilepsy. *British Journal of Learning Disabilities*, 33, 39-43.
- Redley M., Prince E., Bateman N., Pennington M., Wood N., Croudace T. & Ring H. (2013) The involvement of parents in healthcare decisions where adult children are at risk of lacking decision-making capacity: a qualitative study of treatment decisions in epilepsy. *Journal of Intellectual Disability Research*, 57, 531-538.
- Reuber M., Gore J., Wolstenhome J., Jonas P., Frankson C., Murray C. & Styring L. (2008) Examining a community model of epilepsy care for people with learning disabilities. *Seizure - European Journal of Epilepsy*, 17, 84-91.
- Ring H., Donaldson C., Jones L., Mander A., Murphy C., Pennington M., Rawnsley M., Rowe S., Wason J. & Redley M. (2014) Clinical trial protocol: Improving outcomes in adults with epilepsy and

- intellectual disability: A cluster randomised controlled trial of nurse-led epilepsy management (EpAID). Available online at: http://www.nets.nihr.ac.uk/data/assets/pdf_file/0014/81131/PRO-10-104-16.pdf (accessed 12 May 2015).
- Ring H., Zia A., Bateman N., Williams E., Lindeman S. & Himlok K. (2009) How is epilepsy treated in people with a learning disability? A retrospective observational study of 183 individuals. *Seizure - European Journal of Epilepsy*, 18, 264-268.
- Robertson J., Hatton C., Emerson E. & Baines S. (2015a) Mortality in people with intellectual disabilities and epilepsy: A systematic review. *Seizure: European Journal of Epilepsy*, 29, 123-133.
- Robertson J., Hatton C., Emerson E. & Baines S. (2015b) Prevalence of epilepsy among people with intellectual disabilities: A systematic review. *Seizure: European Journal of Epilepsy*, 29, 46-62.
- Sterrick M. & Foley J. (1999) Educating lay carers of people with learning disability in epilepsy awareness and in the use of rectal diazepam: a suggested teaching protocol for use by healthcare personnel. *Health Bulletin*, 57, 198-204.
- Straetmans J., Lantman-de Valk H., Schellevis F. & Dinant G. (2007) Health problems of people with intellectual disabilities: the impact for general practice. *British Journal of General Practice*, 57, 64-66.
- Thompson R., Linehan C., Glynn M. & Kerr M. P. (2013) A qualitative study of carers' and professionals' views on the management of people with intellectual disability and epilepsy: a neglected population. *Epilepsy & Behavior*, 28, 379-385.
- Tiffin P. A. & Perini A. F. (2001) The use of antiepileptic drugs in learning disabled people with epilepsy: an audit of adult in-patients in a treatment and continuing care service. *Seizure - European Journal of Epilepsy*, 10, 500-504.
- Vallenga D., Grypdonck M. H. F., Tan F. I. Y., Lendemeijer B. H. G. M. & Boon P. A. J. M. (2006) Decision-making about risk in people with epilepsy and intellectual disability. *Journal of Advanced Nursing*, 54, 602-611.
- Vallenga D., Grypdonck M. H. F., Tan F. I. Y., Lendemeijer B. H. G. M. & Boon P. A. J. M. (2007) Improving decision-making in caring for people with epilepsy and intellectual disability: an action research project. *Journal of Advanced Nursing*, 61, 261-272.
- Vallenga D., Grypdonck M. H. F., Tan F. I. Y., Lendemeijer B. H. G. M. & Boon P. A. J. M. (2008) Improving decision-making in caring for people with epilepsy and intellectual disability: an action research project. *Journal of Advanced Nursing*, 61, 261-272.
- Watkins J., Espie C. A., Curtice L., Mantala K., Corp A. & Foley J. (2006) Development of a measure to assess the impact of epilepsy on people with an intellectual disability: the Glasgow Epilepsy Outcome Scale - Client version (GEOS-C). *Journal of Intellectual Disability Research*, 50, 161-171.
- Whitten E. & Griffiths A. (2007) Implementing epilepsy guidelines within a learning disability service. *Seizure - European Journal of Epilepsy*, 16, 471-478.
- WHO (2005) Atlas: Epilepsy Care in the World 2005. World Health Organization, International Bureau for Epilepsy & International League Against Epilepsy, Geneva.

Table One: Summary of studies regarding service responses to people with intellectual disabilities & epilepsy¹

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Ahmad et al 2007	England	Impact evaluation of epilepsy care pathway initiative for people with intellectual disabilities	Auditor assessed pilot site service on parameters prior to & 3 months after introduction of care pathways (3 pathways in initiative, 1 of which epilepsy)	6 pilot sites in West Midlands	6 pilot sites	Each site displayed regional variations in models of service delivery & local recruitment difficulties	Percentage for training offered to clients & carers, & results sent to the general physician pre- & post epilepsy care pathway AED use	% pre-/post approximated from bar chart: client training 16% v 35%; carer training 50% v 88%; results to GP 16% v 54%
Baribeault 1996	US	Clinical advocacy	Descriptive information based on practice	Adults living in residential group homes with epilepsy for whom the author had acted as clinical advocate/healthcare coordinator	21	Adults with intellectual disabilities & epilepsy living in residential group homes over period 1988-1993		After the clinical advocate collaborated with the prescribing neurologist to complete a review of seizure history & AED use, 12 (58%) had AEDs reduced or eliminated. The 21 participants saw neurologists scattered throughout the community from group practice to teaching hospitals. The author suggests that many neurologists in the community may tend towards a 'status quo' or maintenance type of prescribing so if no worse & no AED effects, AEDs remain unchanged. A clinically skilled advocate participating in the consultation led to more precise objectives being formulated.
Beber et al 1999	England	Establishment of a learning disabilities psychiatric service	Learning disabilities psychiatric service established. Comparisons made between baseline & outcome measurements for those who did (for min 1 year, max 2 years), compared to those who did not receive psychiatric care	Those living in NHS accommodation or referred to the learning disabilities psychiatric service; or using NHS respite care & not accessing learning disabilities psychiatric care (comparison group)	42 psychiatric care; 12 comparison group	Adults with learning disabilities & epilepsy aged 17-66 years, 37 men & 31 women. Only 7 referred to psychiatric service specifically for epilepsy	Seizure frequency, AED use, blood tests, side effects	Learning disabilities psychiatric care was found to effect reduced seizure frequency, with a reduced frequency of drug side-effects & reduced frequency of polypharmacy. The type of the person's seizures was more likely to be determined, medical reviews & appropriate blood test monitoring conducted & active interventions made to improve seizure control & to withdraw unnecessary drugs, if the person was receiving learning disabilities psychiatric care. Almost all of these results were highly statistically significant.
Beran et al 1992	Australia	Knowledge & attitudes of nurses working with people with epilepsy & developmental disability	Postal survey	Random sample of 100 institutionally based nurses out of 385 employed by a regional developmental disability service.	85 out of 100 (85% response rate)	Modal age between 35 & 45 (30 nurses). 40% less than age 35	Includes % correctly ascribing seizure type to unambiguous descriptions of a variety of seizures	75% unable to correctly diagnose a complex partial seizure. > 60% confused terms such as absences, atonic seizures & myoclonic seizures. None correctly identified a pseudoseizure. They had no problem diagnosing a generalized tonic clonic seizure or status epilepticus. In open ended comments, the most frequent response was an identified need for in-service lectures to include discussion of AEDs, clarification of terminology, discussion of aetiology & management of epilepsy & the issuing of frequent information updates. They also wanted greater input to teach them what they should observe during seizures.

¹ Terms for intellectual disabilities are given as used in the studies e.g. 'learning disabilities' is often used in the UK

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Branford et al 1994	England	AED prescribing practices	Cross-sectional survey based on NHS facility records or day centres, homes, hostels & interviewing carers. Comparison of NHS & community facilities	Leicestershire Learning Disability Register (LIDR)	381	Adults with seizure disorder for which AED received	Polypharmacy, drugs used, residential placement & specialist involvement	240 (63.0%) on monotherapy, 125 (32.8%) on two AEDs & 16 (4.2%) on 3 AEDs. The most frequently used were carbamazepine (80.1% of NHS, 54.8% of community, chi square 27.06, $p < .001$) & sodium valproate (31.6% NHS, 29.5% community, ns). Phenytoin (5.3% NHS, 21.4% community, chi square 20.15, $p < .001$), phenobarbitone (11.1% NHS, 23.3% community, chi square 9.58, $p < .001$), & primidone (which is metabolized to phenobarbitone) (0.0% NHS, 5.7% community, chi square 10.11, $p < .001$) were used significantly more frequently by those living in non-NHS settings. The specialist learning disability service was directly involved with the supervision of seizure control for all in NHS facilities & 49% in community facilities, with a lesser degree of involvement for a further 22%. There was no involvement for less than 30% of those in the community. Degree of specialist service involvement was associated with reduced polypharmacy (chi square 9.69, $p < .05$). There was an association between increased age & the prescribing of phenytoin & phenobarbitone with the association being similar for both NHS & community facilities.
Branford et al 1998	England	AED prescribing practices	Survey, questionnaire completion by key carers, comparison to findings of previous study involving same participants who were initially living in institutional settings	Leicestershire Learning Disability Register (LIDR)	138	80 (58%) male & 58 (42%) female. Mean age 47 years (SD 12.59) & 63% profound learning disabilities. 74 (54%) in NHS accommodation, 64 (46%) in community accommodation	Seizure frequency, polypharmacy & drugs used	A greater proportion had active epilepsy in 1997 (71%) than in 1985 (58%). The deterioration in seizure control occurred despite an increase in polypharmacy, the introduction of new AEDs & in some cases increases in dosage. There was continued reduction of the prescribing of phenobarbitone & increased prescribing of sodium valproate & carbamazepine. This change would be in line with the aims of the original review programme & constitutes good medical practice.
Buelow et al 2006	US	Sources of parental stress for parents of children with intellectual disabilities & epilepsy	Semi-structured face to face interviews & qualitative analysis	Pediatric neurology & epilepsy clinics, a pediatric neurologist in private practice, & school nurses	20	18 mothers, 1 father, 1 dyad. Children age 9-19 (mean 12.2, SD 2.5), all definite diagnosis epilepsy	Qualitative analysis of open-ended responses	Five categories of sources of stress were identified, one of which was communication with healthcare providers. There were concerns about medications & a lack of information regarding their child's illness. Several had experienced problems obtaining a diagnosis. Some felt doctors ignored what they felt to be significant side effects of medication. Parents commonly expressed the need for more information about their child's condition in relation to future course, situations about which they should be concerned, & a need for information on medications particularly side effects.

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Camfield et al 2011	Mainly US	Transition to adult care	Survey of symposium attendees	254 attendees at symposium on Lennox Gastaut syndrome & related epilepsies	133 (response rate 52%)	Majority were pediatric neurologists, 91.7% from US	% responses to survey items	Factors that make it challenging to transition a patient with epilepsy & intellectual disability to adult care: attachment of the family & patient to the respondent (62%); concern that an adult neurologist will take less time with the patient & his/her family than the respondent has in the past (42%); respondent's attachment to the patient/family (41%), lack of adult neurologist willing to care for adults with epilepsy & intellectual disability (35%); concern that an adult neurologist would not be familiar with the patient's neurologic or medical condition (33%); other (18%); & too much time needed to coordinate care & complete the transition (17%). For 'other' the most common specified response was "financial/insurance issues" (e.g., with neurologists not wanting to take Medicaid patients).
Chubb et al 1995	Wales	Audit of out-patient care	Classical audit cycle: recognition of areas of concern, setting of standards, measuring of a baseline, & evaluation of effect of standard setting on this baseline	Patients attending Developmental Epilepsy Clinic (established in 1991) run by the learning disability services at the University Hospital of Wales, Cardiff.	24	Mean age 31.5 years (range 6-66), 58% females	% of times standard reached	Significant improvements were seen in recording of monthly seizure rate by seizure type; legibility of signature; regular letter to GP; writing seizure frequency in letter to GP. Some standards showed no significant improvement because the pre-audit standard was high. Others, in particular the recording of side effects (22.3% before, 33.3% after) & comments on quality of life (52.8% before, 58.3% after) were poor both before & after audit.
Clark et al 2001	Scotland	Evaluation of a video-assisted educational package for adults with mild learning disabilities & epilepsy	Deferred entry to treatment design: 8 in treatment group & 10 in deferred treatment group	Adult training centres (n =2), a residential village for adults with a learning disability (n =3), a residential hostel for adults with a learning disability & epilepsy (n =8) & an adult training centre for individuals with a learning disability & epilepsy (n =5).	18	Mild intellectual disabilities. One or more seizures in past 12 months, prescribed AEDs, some verbal communication.	Before/after scores on Epilepsy & You checklist, & EKQ-LD (epilepsy knowledge questionnaire)	Knowledge increased on the majority of items on the checklist. Subjects increased greatly in their knowledge about what an EEG is & about the importance of seizure diaries; both issues fundamental to the management of epilepsy. Subjects also demonstrated increases in their knowledge about what, & when, to write in a seizure diary & demonstrated increased understanding of why seizures happen. There was no increase in knowledge for the deferred treatment group. All subjects completed the Evaluation Questionnaire & stated that they enjoyed 'Epilepsy & You'. Increased knowledge was maintained at 4 week follow-up.
Cole et al 2009	England	Use of individual epilepsy guidelines (IEGs) by carers; management of emergency seizure events in the community (rescue medication)	Service evaluation of IEGs through a postal survey of carers	Carers (support workers/family carers) of 44 people who had received an IEG from 2000-2006 approached	42 (response rate 96%)	Carers of patients with learning disability & complex refractory epilepsy from an outpatient's learning disability clinic list	% responses to survey items	34 (80%) of the carers utilised the IEGs every time an individual had an epileptic seizure. 39 (93%) carers found the IEGs easy to understand & 37 (88%) carers were more informed about carrying out the procedure of administering rectal diazepam as a result of IEGs. 40 (95%) carers were certain when to administer rectal diazepam & 36 (86%) claimed that the IEGs made it easier for clarification. Only 23 individuals were prescribed oral clobazam medication, alongside the prescription of rectal diazepam. The responses indicated that 20 (93%) carers were certain when to administer oral clobazam. Only 3 (7%) of the responses indicated they were uncertain. 37 (88%) carers agreed that they had become familiar with their clients epilepsy due to the IEGs.

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Deepak et al 2012	England	Care home staff training for use of rescue medication	Survey, semi-structured telephone questionnaire completed with care home managers	Staffed homes in High Wycombe in the non-health sector (private, voluntary or local authority)	21 care homes (of total 22)	Care home managers in 21 non-health sector care homes	% responses to survey items	11 of 21 care homes had a client with epilepsy. 5 of these 11 (45%) had staff trained to administer emergency AEDs. Hence, 55% of the 11 care homes with a client with epilepsy did not have staff trained in emergency AED administration. Of the 10 care homes with no clients with epilepsy, none had staff trained to administer AEDs but 7 had arrangements for in house training if a client with epilepsy were placed there. One home had a policy of only accepting clients with well controlled epilepsy. Others reasons for non-training included non-awareness of need for training, & not having patients with prescribed emergency AEDs.
Esan et al 2010	England	Audit on adherence to NICE 2004 guidelines in a specialist epilepsy clinic for people with learning disabilities	Audit 2005, recommendations made, & audit repeated 2007. Based on review of letters to GPs not handwritten notes	Multidisciplinary epilepsy clinic for people with learning disabilities running > 10 years. Held at outpatient dept of general hospital.	All 45 regular patients	All regular attendees. Male 47%, mild intellectual disability 38%, moderate 13%, severe 20%, ns 29%	% where standards met	At initial audit, 11 of 21 standards were met. At re-audit, some improvement in assessment of AED effectiveness, AED tolerability, requesting EEG, CT/MRI, measuring of serum calcium & vitamin D levels, tests of bone metabolism, providing written & visual information about epilepsy, discussing quality of life, safety issues, provision of diazepam protocol, & provision of information about SUDEP. The specialist epilepsy service showed improvement in meeting standards & this was of benefit to both service users & clinicians. With regards to rectal diazepam, some patients had no one to administer it or there were training issues with staff involved.
Fridhandler et al 2012	Canada	AED prescribing practices	Retrospective review of physician notes, letters, & pharmacy reports. Comparison of those with & without intellectual disabilities	Patients seen between 2009 & 2011 at Epilepsy Clinic at the Toronto Western Hospital	50 with & 50 without intellectual disabilities	50 patients with severe intellectual disabilities, mean age 33.1 (SD 1.92), 44% male, 84% drug resistant (ILAE criteria). Control group mean age 45.4 (SD 2.05), 34% male, 74% drug resistant.	AED receipt (current & past)	Those with intellectual disabilities currently & historically prescribed a greater total number of AEDs (currently 2.7 v 1.8). More were taking at least one old AED (72% v 58%) but there was no difference in the number historically prescribed an old AED (76% v 88%). There was no difference in the number currently (70% v 64%) or previously (66% v 60%) taking a new AED. More of those with intellectual disabilities were currently (54% v 24%) & previously (58% v 28%) prescribed a benzodiazepine. No evidence for a discrepancy in access to new AEDs in patients with intellectual disabilities.
Frost et al 2003	England	Implementation of good practice guidelines	Cross-sectional postal survey	750 randomly selected health professionals: specialist epilepsy nurses (200), adult consultants (neurologist & learning disability consultants) (300), paediatric consultants/neurologists (150) & GPs (100).	241 (32%) questionnaires returned	81 specialist epilepsy nurses, 88 adult consultants, 57 paediatric consultants, 15 GPs	% responses to survey items	Most questions relate to epilepsy care generally. A dedicated clinic for people with learning difficulties was noted to have been implemented by 31.6% of 215 who answered the question, not implemented by 62.8% & attempted by 5.6%

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Hom et al 2015	US	Adherence to AEDs & living situation	Retrospective analysis of a closed Medicaid pharmacy billing system over a 30 month period (2000 to 2002).	Client data of regional centre coordinating services for people with intellectual disabilities (linked by social security number to billing system). All included had filled prescriptions for AEDs spanning at least 6 months.	793	107 children (13.5%), 479 males (60.4%), 563 Caucasian (71%). Living situation group homes (69.9%), family homes (23.5%), semi-independent settings (6.7%). Most adults lived in group homes (74.3%), most children lived in family homes (58.8%)	Calculated the medication possession ratio (number of days each participant was in possession of an AED), & defined non-adherence as 25% or more of the exposure days without the possession of an AED.	The non-adherence rate for AEDs was 5.96% for group homes, 20.00% for semi-independent settings, & 31.72% for family homes. Controlling for age & gender using logistic regression, participants living in semi-independent settings had a 4.14-fold increase in the odds of prescription non-adherence (95% CI: 1.93, 8.84; P < 0.001), & participants living in family homes had a 6.05-fold increase in the odds of prescription non-adherence compared with those living in group homes (95% CI: 3.54, 10.36; P < 0.001).
Kerr et al 2014	Total 14 but most from UK	Views on supports for people with an intellectual disability who have epilepsy	International online survey of professionals, paid caregivers, & family members, qualitative analysis	15 organizations, including national epilepsy associations, support groups, & professional networks, located in 7 countries promoted the survey either via their website &/or by distributing documentation to their members	133	Mostly from UK. 67 professionals, 48 family members, 18 paid caregivers	Open ended responses to survey questions	Within the area of service delivery, the key themes identified from responses were: calls for greater resources including multidisciplinary team approaches; specialist nursing; provision of local services; consistency in quality of service provision; & greater recognition of family. Concerns regarding lack of expertise in primary care; brief & ineffective consultations. Calls for greater use of epilepsy nurse specialists in training.
Kiani et al 2014	England	SUDEP	Retrospective study of adults who died from SUDEP between 1993 & 2010	Leicestershire Learning Disability Register (LIDR)	26 definite or probable SUDEP, case notes investigated for 20	Adults on LIDR who died from definite or probable SUDEP between 1993 & 2010	Information recorded in case notes	13 (65%) had an Epilepsy Care plan & similar number had a prescription/written protocol for the use of a rescue medication. No recorded evidence found of SUDEP being discussed with patients or their families/carers. Although seizure frequency was documented in all case files except one, this was not reported in detail; sometimes only the approximate numbers of seizures (of all types) were documented, partly owing to lack of access to the seizure diary during the review. Generally record keeping following the adult's death was poor. Only 5 medical case files (25%) recorded the news of the patient's death & any communication between the clinical team & family after the death of the patient including discussion on referral for bereavement counselling.

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Kyrkou et al 2006	Australia	Community use of intranasal midazolam (INM)	Survey of parents, carers & education staff	Villa managers in a large institution for people with intellectual disabilities identified residents who had been administered INM. Also parents of children (not necessarily with intellectual disabilities) who had been ordered INM to manage prolonged seizures; education & support staff trained to administer INM to those children.	80 people with intellectual disabilities; 51 children (not necessarily with intellectual disabilities) & 47 parents of these children responded to survey	80 with intellectual disabilities aged between 23 & 74 years who received INM	% responses to survey items	Seizures were controlled in 125 of these 131 people (95.4%), increasing to 127 (96.9%) when a higher dose based on weight was administered. There was only one minor adverse event. Whilst not necessarily parents of children with intellectual disabilities, parents expressed a preference for INM over rectal diazepam because of the shorter time it took to take effect & wear off, & the ability to administer it in public if necessary.
Litzinger et al 1993	US	Community placement for those with severe to profound intellectual disabilities & complex epilepsy	Outcomes of community placement assessed. Involved visits from neurologist to oversee medication changes, education of home staff & emergency room physicians, & early intervention for seizures	15 men & women relocated from institution to community home	15	7 women & 8 men with severe to profound intellectual disabilities & complex epilepsy, age 22-40 years, average 27 years	Score on Scales of Independent Behavior, times hospitalized, emergency room visits, seizure control	Simplification of anticonvulsants, early intervention for seizures, & improved staff education resulted in fewer seizures, decreases in emergency room visits for status seizures & drug-related side effects, & increased levels of functioning as measured by activities of daily living. None of the individuals returned to the institution, & only 1 was relocated to a more structured living situation after several hospitalizations for pneumonia.
Markar et al 1998	England	Communication between GPs & psychiatrists after initial outpatient assessment & satisfaction with service provided by Community Team for Learning Disability (CTLD)	Audit including survey of GPs regarding satisfaction with service provided by CTLD	57 GPs in one area (all using services of CTLD)	30 usable responses	GPs using service provided by CTLD in one area	% responses to survey items	83.3% expressed overall satisfaction with the service. The remaining 16.6% made comments regarding the improvement of the service which include the comment that psychiatrists should take over the management of epilepsy in these patients

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Matthews et al 2008	Wales	Level of investigation & AED receipt	Cross-sectional information collected by interview with carer	40 general practices	58 with epilepsy from total of 318	Adult, mean age 39.6 (SD 13.3)	% having seen specialist, receiving investigations, or AEDs	40 out of 55 participants (72.7%) had been referred to a specialist, two (5%) to a neurologist, 13 (32.5%) to an epilepsy clinic, 23 (57.5%) to a learning disability psychiatrist & two (5%) to another specialist. Medication details were available for 57 participants: three (5.3%) were prescribed no AED, & aside from prescriptions for pro re nata (PRN) medication, 24 (42.1%) were prescribed a single AED, 23 (40.4%) were prescribed two AEDs, 5 (8.8%) three & 2 (3.5%) four. The most common AEDs were carbamazepine (n = 29, prescribed to 53.7% of those receiving medication), sodium valproate (n = 26, 48.1%) & lamotrigine (n = 15, 27.8%). Phenytoin, primidone & one or other of the benzodiazepines were each prescribed to five participants (9.3%). Other AEDs were prescribed to three or fewer participants.
McCarron et al 2014	Ireland	Epilepsy management & support for older adults with intellectual disabilities	Data on epilepsy were drawn from the first wave of data collected as part of the Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA).	Randomly selected from Ireland's National Intellectual Disability Database (NIDD)	229 with epilepsy in total sample of 753	Age 40 or more, eligible for receiving services	% responses to survey items	Epilepsy review: within past 12 months 80.8%; past 2 years 5.1%; over 2 years ago 11.6%; never 2.5%. Review by: GP 51%; psychiatrist 40%; neurologist 42%; more than one group/professional 34.7%. The majority of respondents (83.1%) kept a record of their seizures, with those living in residential care (92.7%) more likely to have such a record than those living independently (43.5%) or in the community (77.8%). 89.5% (n=205) were taking AEDs. Of the 24 not taking AEDs, nine (37.5%) had not had a seizure in the past 2 years, three (12.5%) had had a seizure more than once a month, & three (12.5%) had had a seizure less than once a month but within the past 2 years, 9 (37.5%) did not answer the question on seizure frequency.
Mobbs et al 2002	England	Role of learning disability community nurse (CNLD)	Cross-sectional postal survey	194 Managers of CNLDs working in 170 NHS Trusts	136 (81%) of 170 NHS Trusts represented	Managers of CNLDs working in NHS	% responses to survey items	25 (23%) had CNLDs specializing in epilepsy. CNLDs were offering training in services outside the NHS trust, such as social services & the independent/voluntary sectors in relation to epilepsy in 126 trusts (95%)
Morgan et al 2003	Wales	Health service use	District-wide registers for epilepsy & intellectual disabilities created using record-linkage techniques	Social services register, inpatient & outpatient databases, 'mental handicap' hospital dataset	257 with epilepsy out of 1595	Mainly age 16+ in contact with health or social services	Standardized activity ratios (SAR)	Patients with intellectual disability & epilepsy from the institutional group were less likely to be admitted as inpatients compared with those in the community group (SAR 0.63 (95% CI 0.54, 0.73)). This was also true for admissions primarily for seizure (SAR 0.31 (95% CI 0.21, 0.40)). Patients with intellectual disability & epilepsy from the institutional group were less likely to be seen as outpatients compared with those in the community group (SAR 0.19 (95% CI 0.15, 0.22)). For A&E admissions for those institutionalized individuals with co-existing epilepsy & intellectual disability, the standardized admission ratio was 0.34 (95% CI 0.23, 0.44) relative to the community based population.

Author	Country	Focus	Design	Sample source	Sample Size	Sample features	Outcome measures	Results
Mukhopadhyay et al 2010	England	Carers views on the use of rescue medication	Postal survey (classed as service evaluation)	People with intellectual disabilities & epilepsy identified from lists of two specialist intellectual disability services in Essex. Questionnaires to care home managers, care staff or relatives.	58 out of 93 care home managers responded. 102 care staff responded. 15 out of 25 relatives responded.	Care home managers, care staff or relatives of 140 people with intellectual disabilities & epilepsy	% responses to survey items	Of 140 people with epilepsy, 84 (60%) had a care plan for epilepsy. 23 carers had used rescue medication in the last year, 61% used buccal midazolam (BM), 26% rectal diazepam (RD) & 13% both. 17 episodes of RD use were recorded in the past year, compared to 46 episodes of BM use. Carers perceived BM to be more socially acceptable compared to RD. It was rated as better across a range of parameters: convenience of administration, invasiveness of procedure, gender issues, social acceptability, consent issues, & onset of action. When those who had used both BM & RD were asked which they preferred, of 27 carers who responded 26 preferred BM.
Peeters 2000	Belgium	Decision tree for non-medically trained staff which treats seizures as medical emergencies	Prospective analysis of decision tree over 16 months compared to 6 months retrospective review. Non-medically trained staff instructed on proper identification of epileptic seizures; verbal & written instructions on antiepileptic treatment provided. Initiation of AED treatment 5 or 7 minutes after seizure onset indicated in the tree.	Two establishments, both non-hospital settings	20	At least 18 years of age, moderate to profound intellectual disabilities & had been receiving maintenance therapy with oral AEDs.	Dose, time of treatment, seizure type, duration, severity, & frequency were recorded	Treatment of seizures as medical emergencies reduced frequency, duration, & severity (in comparison with results from the retrospective review). In patients who had cluster seizures with loss of consciousness, treatment with rectal diazepam or lorazepam resulted in good recovery of consciousness between intervals. In seizures involving a loss of consciousness, none lasted longer than 10 minutes, whereas in the 10 months prior to implementation of the decision tree, these types of seizures lasted for 20 minutes or more. For seizures without a loss of consciousness, none lasted longer than 37 minutes. No patient was hospitalized. Seizures lasted up to 18 hours in the retrospective review.
Redley et al 2013	England	Parental involvement in clinical decision making	Semi-structured interviews & qualitative analysis	Convenience sample of mothers of adults with intellectual disabilities reflecting familiarity with specialist community based learning disability services (n=8) & mainstream hospital neurology services (n=9), & where both services were involved but neither appeared to have overall clinical responsibility (n=4)	21	Mothers of adult children with intellectual disabilities & epilepsy (accompanied by father at 3 interviews)	Process of analytic induction to present a model of the role of parent-proxies in the management of epilepsy in adult children with intellectual disabilities	Mothers were willing to live with what they considered to be an acceptable level of seizure activity, considered as a state of equipoise, & to reject possible changes in their son or daughter's treatment. Clinicians may avoid going against mother's wishes. There was no evidence to indicate that clinicians were following the Convention on the Rights of Disabled Persons (CRPD) or Mental Capacity Act (MCA) & involving these adult patients in decisions about their own treatment.

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Reuber et al 2008	England	Health service use & AED receipt	Cross-sectional postal survey	Sent to carers of all 442 individuals from the Sheffield Case Register who also had a recorded diagnosis of epilepsy	236/442 returns (53.4%). Further 11 returns excluded as said they did not have epilepsy	Mean age 43.5 (SD 16.1, range 18–86 years), 53.3% male, 44.6% lived in residential care or supported living, 43.2% with carers, 6.9% on their own, 1.2% married or living with a partner, unknown 4.2%.	% accessing health services	22.7% had been free of seizures for over 1 year. 15.6% attended A&E at least once or in preceding year because seizures would not stop, 5.3% attended more often. 27.6% had a supply of RD or BM or both at home for prolonged seizures (27.4% RD, 4.5% BM), of whom 42.6% stated that they had received training from a nurse or a doctor on how to give the medication. 40% of those who had been to A&E over the last year with a prolonged seizure had access to emergency medication. One in 10 patients under primary care review was taking one of the “newer” AEDs. 53.3% of epilepsy diagnoses made by a seizure expert (neurologist or paediatrician). 46.2% had had an EEG, & 41.3% a brain scan. 68.9% had been assessed by an expert in secondary care (neurologist, psychiatrist or epilepsy nurse) at some point. 60.4% had seen a neurologist, 34.2% an epilepsy nurse, & 20.9% a psychiatrist. 70.7% had an epilepsy treatment review within the last 12 months, the rest less frequently. 38.7% were under review in secondary care (neurologist, psychiatrist or epilepsy nurse), 52.9% were under review in primary care alone, & 8.9% epilepsy treatment had not been reviewed at all. Patients with more severe epilepsy more likely to be under specialist care but 60.6% of patients with ongoing seizures, 57.9% with major seizures & 68.7% of individuals taken to hospital with prolonged seizures had no access to specialist advice.
Ring et al 2009	England	Management strategies & clinical outcomes	Retrospective observational study; data from clinical notes (preceding 3 months) & interviews with carers, learning disability team members, any neurology-based clinicians involved in their management & GP	All those with intellectual disability & epilepsy in 5 community learning disability services that covered one county (excluding those without capacity to consent); 71% of those identified took part	183	Mean age 40 years, range 16–72 years; 55% male. 20% mild intellectual disabilities, 16% moderate, 57% severe, & 7% profound. 43% diagnosed with epilepsy for at least 17 years	Current seizure types & frequency, nature of clinical support & management received for epilepsy, current AEDs	37% received epilepsy care from a hospital-based neurology service. The patients' GP also actively contributed in terms of initiating or changing treatments in 63% of cases & for 6% of participants the GP was the only clinician supporting epilepsy treatment. Around a third were receiving epilepsy management from hospital-based neurology services but it was not clear what factors determined which treatment pathway patients followed. 1% were not taking any AEDs, 40% were receiving monotherapy, 36% were being treated with two AEDs & 23% were prescribed three or more AEDs. There was a significant difference between the mean number of AEDs prescribed to those whose epilepsy was only managed by learning disability services (1.7 AEDs) & those who also received neurology service input (2.14 AEDs) ($t = 3.257, P = 0.001$). Clobazam was prescribed relatively more frequently to those receiving neurology-based AED management (chi square 6.627, $df = 1, p = 0.018$), whilst sodium valproate was prescribed relatively less often (chi square 5.027, $df = 1, p = 0.025$). Other AEDs were prescribed at approximately similar rates in learning disability & neurology services.

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Thompson et al 2013	UK & Ireland	Carers' & professionals' views on management (medical care & services)	Online survey with open-ended comments analysed qualitatively	International online survey promoted by various epilepsy associations, support groups, & networks in 7 countries. Only responses from UK & Ireland used due to low numbers from other countries	113	Family 38 (34%); paid carer 16 (14%); professional 59 (52%).	Qualitative analysis of open-ended responses	Professionals & carers differ in their expectations of treatment & with regard to maximizing communication during consultations. Findings suggest that the potential of consultations to provide information for carers, as well as to allay concerns, may not be realized. Rescue medications were viewed favorably; however, respondents expressed less satisfaction with routine medications. These findings may reflect a failure by professionals to appropriately transfer knowledge of these treatments.
Tiffin et al 2001	England	AED use	Audit based on case notes	23 wards which housed adult in-patients with learning disabilities	75	Mean age 38.6 years (SD 11.6). 26.7% female. 41.3% mild intellectual disabilities, 25.3% moderate, 25.3% severe, 8% profound	AED receipt; investigations performed	As a result of information obtained during the audit, one patient received a diagnosis of factitious epilepsy & his treatment was subsequently withdrawn. All patients with a history of status epilepticus were prescribed RD or, more rarely, INM. 44% were on monotherapy, 40% on 2 AEDs, 13.3% on 3 AEDs, & 2.7% on 4 AEDs. 13 (17%) were identified as potentially benefiting from a reduction in the number of AEDs they were prescribed. 15 other patients were currently having a trial AED reduction. Only around 24% of patients with partial seizures received an MRI or CT scan although this may be due to sound practical reasons. 11 (14.7%) had received CT scans of the head with 2 having had failed attempts due to poor co-operation. 6 had received MRIs and one had a failed attempt at an MRI scan due to poor cooperation. The average number of standard EEGs performed was 3 per patient (SD 2.9). 7 patients had no record of having had an EEG.
Vallenga et al 2008	Netherlands	Decision making concerning risks	Participative action research. Data gathered continuously over a 22-month period in 2004–2006 by interviews, observation, written reports of meetings & personal stories	Two residential units of an epilepsy centre. Only nursing team members (registered nurses & nursing assistant staff) fully participated in the action research groups	Nursing team members for 2 units each with 12 clients	Registered nurses & nursing assistants	Ongoing process of improvement	During the action research, caregiving became more client-centred, & the insight that clients should be involved in decision-making grew. Assessment of client risk became more systematic & 'steps to an individual framework for decision-making' were developed as a tool for a systematic approach.

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Vallenga et al 2006	Netherlands	Decision making concerning risks	Qualitative method of multiple embedded case study. Based on interviews with client's parent, brother or sister; the care-manager, who was the physician or psychologist responsible for management of the client's care; the nurse, who provided the care & the client when this was possible (possible for n=7).	Specialized epilepsy residential centre	15	Clients at specialised epilepsy residential centre, age 7-64	Qualitative analysis of interview comments	Measures were taken on an individual basis & included (poly)pharmacy, supervision, avoidance of higher risk activities, provision of protective clothing, or physical restraint of movement. General measures were taken in the furnishing of rooms (eg induction cooking equipment with no hot surfaces). Protective measures were often taken in situations of immediate threat, allowing no time to consider their negative effects. Continuous anxiety about the possibility of seizure & injury caused constant vigilance in parents & nurses. Systematic recording & analysis of accidents was lacking in all of the cases, making it hard for those involved to examine to what extent their fear was proportionate to the risk.
Whitten et al 2007	England	Introduction of a specialist multi-disciplinary epilepsy clinic in line with NICE guidelines	Audit prior to & following implementation of epilepsy clinic (previously seen at a learning disability mental health outpatient clinic); based on clinical notes	Patients with diagnosis of epilepsy within one consultant's catchment area of the learning disability service	23	All with diagnosis of epilepsy in catchment area	% meeting items in audit tool	83% compared to 6% of patients had accurate name & detailed seizure descriptions. 100% compared to 81% & 57% had recording of seizure frequency & severity. 76% of patients had changes made to their seizure diagnosis. 91% compared to 50% of consultations led to changes in treatment plans. 96% compared to 64% had medication prescribed relevant to their seizure type. All patients prescribed 3 or more AED's received a medication reduction plan. All patients received an individualised risk assessment compared to 4.5% beforehand & 68% had risks identified. The implementation of NICE guidelines in this study showed improvements to seizure assessments, matching of medication to seizure type, patient care & epilepsy management.

