Seizure 29 (2015) 46–62

Contents lists available at ScienceDirect

Seizure

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

Review

Prevalence of epilepsy among people with intellectual disabilities: A systematic review

Janet Robertson [a,*], Chris Hatton [a], Eric Emerson [a,b], Susannah Baines [a]

[a] Centre for Disability Research, Division of Health Research, Lancaster University, LA1 4YT, United Kingdom
[b] Centre for Disability Research and Policy, University of Sydney, Australia

ARTICLE INFO

Article history:
Received 11 December 2014
Received in revised form 24 February 2015
Accepted 25 March 2015

Keywords:
Epilepsy
Prevalence
Intellectual disabilities
Down syndrome

ABSTRACT

Purpose: Epilepsy is more common in people with intellectual disabilities than in the general population. However, reported prevalence rates vary widely between studies. This systematic review aimed to provide a summary of prevalence studies and estimates of prevalence based on meta-analyses.
Method: Studies were identified via electronic searches using Medline, Cinahl and PsycINFO and cross-citations. Information extracted from studies was tabulated. Prevalence rate estimates were pooled using random effects meta-analyses and subgroup analyses were conducted.
Results: A total of 48 studies were included in the tabulation and 46 studies were included in meta-analyses. In general samples of people with intellectual disabilities, the pooled estimate from 38 studies was 22.2% (95% CI 19.6–25.1). Prevalence increased with increasing level of intellectual disability. For samples of people with Down syndrome, the pooled estimate from data in 13 studies was 12.4% (95% CI 9.1–16.7), decreasing to 10.3% (95% CI 8.4–12.6) following removal of two studies focusing on older people. Prevalence increased with age in people with Down syndrome and was particularly prevalent in those with Alzheimer's/dementia.
Conclusion: Epilepsy is highly prevalent in people with intellectual disabilities. Services must be equipped with the skills and information needed to manage this condition.

1. Introduction

Intellectual disability (often referred to as 'learning disabilities' in the United Kingdom) refers to a significant general impairment in intellectual functioning that is acquired during childhood, typically operationalised as scoring more than two standard deviations below the population mean on a test of general intelligence [1]. While estimates of the prevalence of intellectual disability vary widely, it has been estimated that approximately 2% of the adult population have intellectual disability [2,3].

In the general population, estimates of the prevalence of epilepsy are in region of 0.6% [4,5] to 1% [6,7]. In people with intellectual disabilities, estimates of the prevalence of epilepsy vary due to differences in the methods used and inherent population biases [8]. Reported rates range, for example, from 16.1% of 1595 people with intellectual disabilities identified in South Wales [9] to 30.7% in a random sample of 753 people with intellectual disabilities aged 40 or more from Ireland's National Intellectual Disability Database (NIDD) [10]. In a systematic review of the prevalence of chronic health conditions in children with intellectual disabilities, the most common condition was epilepsy [11] with prevalence rates in the 14 studies identified ranging from 5.5% to 35.0%, with an overall weighted mean prevalence rate of 22.0% (95% CI 20.8–23.2).

Despite variation in reported prevalence figures, it is clear that the prevalence of epilepsy in people with intellectual disabilities is much greater than in the general population. Further, for people with intellectual disabilities and epilepsy, co-morbidities may be common. Over half of a representative sample of children with intellectual disability and active epilepsy were reported to have a psychiatric diagnosis [12]. However, conflicting findings exist and there is no consensus as to whether people with both intellectual disability and epilepsy are at increased risk of psychiatric morbidity compared to their peers with either epilepsy or intellectual disability alone [13].

The prevalence of epilepsy also increases with increasing severity of intellectual disabilities. In the Oeseburg et al. [11] review, the lower rate of 5.5% was for children with borderline to moderate intellectual disability [14], whilst the rate of 35.0% was

* Corresponding author. Tel.: +44 1524 592895; fax: +44 1524 592658.
E-mail address: j.m.robertson@lancaster.ac.uk (J. Robertson).

http://dx.doi.org/10.1016/j.seizure.2015.03.016
1059-1311/

for children with mild to profound intellectual disability [15]. Such wide differences highlight the need to examine prevalence rates taking into account factors such as the degree of intellectual disability of the sample. Samples based on, for example, those in contact with intellectual disability services are likely to miss out some people with less severe intellectual disabilities. A further issue is that the ascertainment of epilepsy is not consistent across studies, both in terms of the definition of epilepsy used, and how the information is collected.

The aim of this review is to summarise existing research on the prevalence of epilepsy in people with intellectual disabilities, including studies relating specifically to people with Down syndrome which is the most common genetic cause of intellectual disabilities [16]. The review also aims to provide pooled prevalence estimates for studies taking into account factors such as age and level of intellectual disability. Whilst existing reviews have considered the prevalence of epilepsy in people with intellectual disabilities, these reviews do not cover more recent studies on prevalence that now provide more data, particularly in relation to adults with intellectual disabilities. As highlighted in one earlier review, adults have previously been underrepresented in research on the epidemiology of epilepsy in people with intellectual disabilities, with the vast majority of published data pertaining to children [8]. As this review aims to estimate epilepsy prevalence in the general population of people with intellectual disabilities or Down syndrome, it does not include studies relating to less common specific genetic conditions associated with intellectual disabilities, although it is evident that work on such conditions has been published [17].

2. Method

Electronic literature database searches were conducted in Medline, Cinahl and PsycINFO on EBSCO. In addition, the reference lists of articles meeting the inclusion criteria were searched. The reference lists of key book chapters were also searched [18–20]. Searches were completed on 19 June 2014. Searches included terms relating to both prevalence and mortality to create a pool of articles on prevalence or mortality, with articles on mortality being retained for a separate review. Searches combined terms for epilepsy, intellectual disabilities, and prevalence/mortality with the Boolean operator 'and'. Full details of the search terms are given in Appendix A.

2.1. Inclusion criteria

- Peer reviewed
- English Language full text
- Published from 1990
- Primary research
- Present exact figures on the prevalence of epilepsy
- Samples where 50% or more have intellectual disabilities or mixed samples where results are disaggregated for people with intellectual disabilities
- Studies using representative samples of people with intellectual disabilities or samples representative of specific sub-groups of people with intellectual disabilities (e.g. specific level of intellectual disability, specific age band)

2.2. Exclusion criteria

- Case studies
- Case series
- Reviews
- Studies based on neonates (new born infants up to 28 days after birth)
- Studies on conditions where intellectual disabilities cannot be assumed (e.g. cerebral palsy) where results not disaggregated for people with intellectual disabilities
- Studies on specific syndromes associated with intellectual disabilities with the exception of Down syndrome
- Studies where ascertainment of epilepsy could be confounded with febrile seizures
- Studies employing samples unrepresentative of specific sub-groups of people with intellectual disability e.g. only those attending for inpatient specialist medical care
- Studies not presenting exact figures

Initially, titles and abstracts were used to exclude those studies which were obviously not within the scope of reviews on prevalence or mortality. 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 the first and second author and discussed until consensus was reached on whether or not they met the inclusion criteria. Those relevant to other future planned reviews (e.g. mortality) were filed for future reference.

Where multiple articles used the same sample or samples were likely to have considerable overlap, only the most recent study was included. One exception was a study based on adults with intellectual disabilities registered with the Leicestershire Intellectual Disability Register for the period 1993–2010 which reported a prevalence of 19.1% in a sample of 5391 [21]. As this study focuses on sudden and unexpected death in epilepsy (SUDEP), it does not outline the methodology for obtaining this estimate. As such, it was decided to include an earlier study based on the same register which focused on epilepsy prevalence [22]. A further study including only people with Down syndrome which was partly based on the Leicestershire Intellectual Disability Register was also included [23].

Information from the included studies was extracted by the first author and this information was tabulated (see Table 1).

2.3. Quality assessment

A gold standard to evaluate the quality of observational research does not exist [24]. A method for evaluating aspects of quality considered important in relation to obtaining valid estimates of the prevalence of epilepsy was developed. The selected quality indicators were:

1. Definition of epilepsy:
   - Score 2: Definition given (e.g. ILAE)
   - Score 1: Partial definition given – some information (e.g. database codes used, epilepsy diagnosis) but incomplete
   - Score 0: Not stated (no criteria for epilepsy given)
2. Ascertainment of epilepsy – this refers to the identification of those in the sample with epilepsy and not any subsequent follow up of those identified as having possible epilepsy. The following scores were allocated:
   - Score 1: Questionnaire self-completion by informant
   - Score 2: Interview with informant
   - Score 3: Extracted from records or databases
   - Score 4: Clinical examination

   If multiple methods were used, the highest level was entered as the score.
3. Prevalence figures presented for subgroup(s). A score of 1 was allocated for each of the following subgroups for which prevalence figures were reported.

Table 1
Summary of included studies giving prevalence rates for epilepsy in people with intellectual disabilities. Figures under male, and levels of ID columns relate to characteristics of the study sample. Sorted by author name. Studies only looking at Down syndrome listed separately at end of table.

Authors, year and quality score[b]	Country of study	Key sample features	Sample source	Age range (mean (SD); median)	Male %	Borderline ID %	Mild ID %	Moderate ID %	Severe ID %	Profound ID %	Unspecified ID %
Airaksinen et al. (2000) [42] 10 (2/4/4)	Finland (Kuopio Province)	Children with ID born 1969–1972 in one province followed until age 22. CP 11%. LS ns.	School achievement tests and social services register	Prevalence at age 22 yrs given	55[a]	–	49[a]	51[a]	←[c]	←	–
Arvio and Sillanpää (2003) [43] 4 (0/3/1)	Finland	People with SPID. DS 14.3%, AE 19.3%, FXS 3.9%. LS ns	Register of District Centre for ID (all in catchment)	1–72 (ns; ns)	ns	–	–	–	52.5	47.5	–
Benassi et al. (1990) [52] 3 (0/3/0)	Italy	Children with 'severe' ID (IQ ≤ 50). DS 22.2%. LS ns.	System recording all school age children with ID	3–13 (ns; ns)	63.3	–	–	100	←	←	–
Christianson et al. (2002) [56] 3 (0/2/1)	South Africa	Children with ID in rural households, up to IQ 80. DS 2.1%, CP 8.4%	Rural villages	2–9 (ns; ns)	61.3[a]	→	81.9[a]	18.1[a]	←	←	–
David et al. (2014) [53] 4 (0/3/1)	France (Isère)	Children with mild ID born 1997 living in one county in 2008. LS ns	Maisons Départementales des Personnes Handicapées (MDPH) and Dept of Education	9–13[a] (ns; ns)	ns	–	100	–	–	–	–
Dekker and Koot (2003) [14] 2 (0/2/0)	Netherlands	Children with borderline to moderate ID living in family home. DS 5.3%	Schools for ID	7–20 (12.9 (3.0); ns)	61.8	100	←	←	–	–	–
Fernell (1998) [38] 3 (0/3/0)	Sweden	Children with 'severe' ID (IQ < 50–55). CP 23.4%, DS 20.3%. LS ns	Register of Board for Provision of Services to the Mentally Retarded BPSMR): all in one municipality	3–16 (ns; ns)	62.5	–	–	100	←	←	–
Forsgren et al. (1990) [39] 8 (2/2/4)	Sweden	All adults and children with ID in one County on a prevalence day. DS 13.7%, Fragile X 2.3%, RS 0.3%. LS any	Register of BPSMR, neurology and paediatric departments	All (ns)	ns	ns	ns	ns	ns	ns	100
Gittins and Rose (2008) [32] 5 (1/3/1)	England, West Midlands	Adults with PMLD in one health district. LS family home, residential care, family placement	Special needs register of LD service, CLDTs	18–51+ (37 (ns); ns)	ns	–	–	–	–	100	–
Goulden et al. (1991) [49] 9 (2/3/4)	Scotland, Aberdeen	Children with ID born 1951–1955 followed to age 22. CP 14.9%, DS 5.1%. LS ns	Receiving special services for ID prior to leaving school	Prevalence figure given is for age 22	ns	–	78.6[a]	21.4[a]	←	←	–
Hand and Reid (1996) [60] 1 (0/1/0)	New Zealand (NZ)	All NZ older adults with ID born before 1940, CP 4%, DS 13%. LS any	Multiple agencies and local networking	51–88 (ns; ns)	50.0	4.0	34.5	38.3	15.4	5.1	2.7
Haveman et al. (2011) [55] 4 (1/2/1)	14 European countries (1 of which upper middle income)	Adults with ID living in Europe. LS any	Mainly service provider registers	19–90 (41 (ns); ns)	50.6	–	22.7	28.2	20.7	11.8	16.6
Hove and Havik (2010) [47] 1 (0/1/0)	Norway	Adults with ID living in community. DS 16.4%, CP 9.1%. LS includes psychiatric wards if part of community care programme	Social services	18–97 (41.8 (14.5); ns)	53.1	–	21.6	41.0	18.0	13.0	6.4

Jelliffe-Pawlowski et al. (2003) [25] 5 (1/3/1)	US	Children with ID from a larger cohort born with or without birth defects. CP 46.8%. LS ns	ID those receiving services from California Department of Developmental Services	7–9 (ns; ns)	ns	–	52.7	47.3	←	←	–
Koskentausta et al. (2002) [15] 4 (0/3/1)	Finland	All children with ID born 1982 to 1988 in one district. LS mostly parental home	Patient register of Rehabilitation Centre, hospitals, special schools	6–13 (9.7 (ns); ns)	59.4	–	56.1	19.4	11.6	12.9	–
Lakhan (2013) [61] 7 (0/4/3)	India	Children with ID living in village households in one of poorest districts. DS 7.3%, CP 31.3%	Door to door survey in 63 villages	3–18 (ns; ns)	52.7	1.9	30.2	38.2	24.0	5.7	–
Lewis et al. (2000) [58] (2000) 3 (1/2/0)	Australia	Young people with ID, LS any	Services in five districts of New South Wales	8–22 (ns; ns)	52.0	→	29.8[a]	40.8[a]	24.2[a]	5.1[a]	–
Lin et al. (2003) [62] 1 (0/1/0)	Taiwan	People with ID registered with day-care institutions, 92.6% age <26. LS ns	Community-based day-care institutions	1–26+ (13.7 (ns); ns)	61.2	–	4.9	17.4	40.9	24.9	–
Matthews et al. (2008) [46] 3 (1/2/0)	Wales	Adults with ID registered with GP. LS independent 10%, family home 46%, staffed home 44%	40 general practices	17–86 (41 (ns); ns)	44	ns	ns	ns	ns	ns	100
McBrien and Macken (2009) [35] 3 (0/3/0)	Ireland	Children with moderate, to profound ID. Any chromosonal or genetic cause 48.5%, DS 24.7%. LS ns	Centre providing educational and health services for all individuals with moderate, severe and profound ID in one area	5–19 (ns; 12)	66.0	–	–	64.9	35.1	←	–
McCarron et al. (2014) [10] 6 (1/2/3)	Ireland	Older adults with ID, 3.1% with DS and dementia. LS all	National database (all ID eligible to receive services)	40–65+ (54.8 (9.6); ns)	45	–	24	46	24	5	–
McDermott et al. (2005) [26] 6 (2/3/1)	US	Adults with ID receiving primary health care. CP 24.9%, DS 8.9%. LS ns	Large urban or small rural primary care practice	20–60+ age[d] mean 36.5 (13.9); ns)	52.0[a]	–	35.9	22.9	41.2	←	–
McGrother et al. (2006) [22] 6 (1/2/3)	England, Leicestershire	Adults with ID known to services. LS any	Leicestershire LD Register	20–70+ (ns; ns)	56.6	ns	ns	ns	ns	ns	100
Memisevic and Sinanovic (2009) [51] 7 (1/3/3)	Bosnia and Herzegovina, Sarajevo	Children with moderate or mild ID. Organic brain injury 21.0%, DS 20.4%, other genetic syndromes 13.2%. LS ns	Two special schools	7–15 (ns; ns)	62.9[a]	–	50.9	49.1	–	–	–
Molteno et al. (2001) [57] 1 (0/1/0)	South Africa, Cape Town	Children with ID at special schools. CP 33.8%. LS ns	Two special schools and a training centre	6–18 (ns; ns)	55.5	–	35.8	38.6	13.8	10.7	1.1
Morgan et al. (2003) [9] 7 (1/3/3)	Wales	People with ID mainly age 16+ in contact with health or social services. LS any	Social services register, inpatient and outpatient databases, MH hospital dataset	15–85+ (ns; ns)	ns	ns	ns	ns	ns	ns	100
Murphy et al. (1995) [27] 4 (0/3/1)	US	Children with ID born 1975 to 1977 living in study area at age 10. CP 12.3%. LS ns	Schools, hospitals, other health and social services	10 year olds	59.2	–	69.9	30.1	←	←	–
Nordin and Gillberg (1996) [40] 6 (2/4/0)	Sweden	All children with ID born 1974 to 1988 in one region. CP 8.9%, DS 8.9%. LS ns	Habilitation and educational services	3–18 (ns; ns)	63.4[a]	–	56.4	43.6	←	←	–

Table 1 (*Continued*)

Authors, year and quality score[b]	Country of study	Key sample features	Sample source	Age range (mean (SD); median)	Male %	Borderline ID %	Mild ID %	Moderate ID %	Severe ID %	Profound ID %	Unspecified ID %
Pawar and Akuffo (2008) [33] 7 (1/3/3)	England, London Borough of Waltham Forest	Adults with ID in contact with services. LS residential homes, supported living, private homes	Adults in contact with one CLDT (active cases)	17–65+ (ns; ns)	53.7	ns	ns	ns	ns	ns	100
Schieve et al. (2009) [29] 4 (1/2/1)	US	Children in households with DS or with ID without DS. DS 19.5%	National survey of households	3–17 (ns; ns)	59.6[a]	ns	ns	ns	ns	ns	100
Schieve et al. (2012) [28] 3 (1/2/0)	US	Children in households with ID without autism	National survey of households	3–17 (ns; ns)	58.1	ns	ns	ns	ns	ns	100
Steffenburg et al. (1995) [41] 6 (2/3/1)	Sweden	Children with ID in one city born 1975–1986. CP 15.3%. LS ns	Education, inpatient, outpatient, child habilitation clinic and child neuropsychiatric clinic registers	6–13 (ns; ns)	ns	–	63.0	37.0	←	←	–
Strømme and Hagberg (2000) [48] 7 (2/4/1)	Norway	Children with mild or 'severe' ID (IQ < 50 assumed mod/sev/pro). Genetic cause 35%, CP 14%. LS ns	Multiple sources (education and medical) used to identify all in one County	8–13 (ns; ns)	57.8	–	55.6	44.4	←	←	–
Temtamy et al. (1994) [63] 4 (0/4/0)	Egypt	Children with ID in households. DS 2.6%, MCA 24.1%, primary CNS defect 12.9%	Households in three localities in Egypt	2–18 (ns; ns)	68.1	30.8[a]	47.9[a]	21.4[a]	←	←	–
Tenenbaum et al. (2012) [64] 1 (0/1/0)	Israel	All with ID living in residential centres. DS 8.1%, Fragile X 1.0%, Rett syndrome 0.2%	Residential care centres	0–60+ (ns; ns)	56.3	–	13.3	41.1	31.6	13.4	0.6
van Schrojenstein Lantman-de Valk et al. (2000) [45] 4 (1/3/0)	Netherlands	Any general practice patients with ID. LS ns	Registration Network Family Practices (RNH) of Maastricht University	ns. 20% aged over 50, includes children	62	ns	ns	ns	ns	ns	100
van Schrojenstein Lantman-de Valk et al. (1997) [44] 4 (0/1/3)	Netherlands	People with ID in institutions or group homes. CP 11.8%, dementia 3.8%, DS 22.3%	Institutions and group homes	0–70+ (ns; ns)	ns	ns	ns	ns	ns	ns	100
Wellesley et al. (1992) [59] 7 (2/3/2)	Australia	Children in Western Australia with ID born 1967–1976. CP 19.8%. LS ns	Multiple services and schools	6–16 (ns; ns)	59.6[a]	–	38.5[a]	31.0[a]	12.6[a]	7.1[a]	10.7[a]
Wong (2011) [65] 5 (1/1/3)	Hong Kong	Adults with ID in residential care. DS 13.2%, CP 16.7%	Residential care services	18–79 (44 (ns); ns)	53.3	–	4.9	41.8	51.9	←	–
Yousef (1995) [66] 6 (0/3/3)	Jordan	Children at special schools for ID. LS ns	Special education centres in one City	ns; school age	73.8	–	27.2	44.2	28.6	←	–
Down syndrome studies											
Collacott (1993) [23] 7 (2/3/2)	England, Leicester-shire	Adults with DS, dementia 5.1%. LS any	Leicestershire LD Register, health service records, day centres, residential services	<30–60+ (ns; ns)	ns	ns	ns	ns	ns	ns	100
Johannsen et al. (1996) [54] 7 (2/4/1)	Denmark	DS in age groups 14–16, 23–29 and 50–60. LS ns	All in one County identified via Danish register and city councils	Age groups 14–16, 23–29, 50–60	62.5	ns	ns	ns	ns	ns	100

McCarron et al. (2005) [36] 4 (0/3/1)	Ireland	Adults with DS aged 35+ in out-of-home placements, AD 50.8%	Care settings (out-of-home placements)	>35-ns (AD 55.4 (7.0); ns. Non-AD 50.8 (5.8); ns)	33.9	–	–	69.4	30.6	–	–
McVicker et al. (1994) [50] 7 (2/3/2)	Northern Ireland	Adults with DS living in community (82%) or hospital	Adults training centres, social services register, MH hospital	19–50+ (community 33.5 (ns); ns, hospital 54.5 (ns); ns)	ns	ns	ns	ns	ns	ns	100
Prasher (1995) [34] 4 (2/2/0)	England, West Midlands	Adults with DS. LS hospital, community or family home	Cohort with DS in West Midlands	16–72 (44.2 (12.5); ns)	50.7	–	18.9	66.7	13.4	–	1.0
Pueschel et al. (1991) [30] 4 (1/3/0)	US, Rhode Island	Children and adults with DS. LS family home, other types ns	Child development centre (enables near complete ascertainment of DS)	0.5–45 (ns; ns)	52.1	ns	ns	ns	ns	ns	100
Roizen et al. (2014) [31] 2 (1/1/0)	US, New York State	Children with DS in New York State. LS ns	Families registered in the New York Congenital Malformations Registry (NYCMR)	3–14 (7.5 (3.1); ns)	51.7	ns	ns	ns	ns	ns	100
Tyrrell et al. (2001) [37] 7 (2/4/1)	Ireland	Adults with DS over age 35. Dementia 13.3%. LS institutional, residential, community	Learning disability services	35–70+ (ns; ns)	ns	–	ns	ns	ns	ns	100

Authors, year and quality score[b]	Method epilepsy ascertainment	Epilepsy definition	Epilepsy prevalence % in main subgroup conditions	Epilepsy cases *n*	Sample size *N*	Epilepsy prevalence %
Airaksinen et al. (2000) [42] 10 (2/4/4)	Parent questionnaire and interview, medical records, examination, EEG	ILAE, epilepsy	CP 62.5%	32	151	21.2
Arvio and Sillanpää (2003) [43] 4 (0/3/1)	ns assume medical records	Epilepsy, ns	DS, 30% AE 83% FXS 5.5%	239	461	51.8
Benassi et al. (1990) [52] 3 (0/3/0)	Medical records and discussion with school health service	Epilepsy, ns	ns	27	90	30.0
Christianson et al. (2002) [56] 3 (0/2/1)	Phase 1 TQQ screening, phase 2 paediatric evaluation	Epilepsy, ns	ns	37	238	15.5
David et al. (2014) [53] 4 (0/3/1)	Carer telephone interview and medical records	Epilepsy, ns	ns	5	181	2.8
Dekker and Koot (2003) [14] 2 (0/2/0)	Parent interview	Epilepsy, ns	ns	26[a]	474	5.5
Fernell (1998) [38] 3 (0/3/0)	Medical records, author personal knowledge	Epilepsy, ns	ns	17	64	26.6
Forsgren et al. (1990) [39] 8 (2/2/4)	Asked staff in institutions and letters to parents or carers, medical records examined if reported epilepsy	Active ≥ 1 SZ last 5 yrs and/or on AED	FXS 23.5% DS 5.9% RS 80% (4/5)	299	1479	20.2
Gittins and Rose (2008) [32] 5 (1/3/1)	Case notes	Epilepsy in case notes	ns	39	61	63.9
Goulden et al. (1991) [49] 9 (2/3/4)	Parent interview and/or records (medical, education, social work)	ILAE, epilepsy	CP 37.5%, postnatal injury 73.3%, Genetic or malformation e.g. DS 12.5%	33	215	15.3
Hand and Reid (1996) [60] 1 (0/1/0)	Questionnaire completed by carer, staff or GPs	Epilepsy, ns	ns	177	1063	16.7

Table 1 (*Continued*)

Authors, year and quality score[b]	Method epilepsy ascertainment	Epilepsy definition	Epilepsy prevalence % in main subgroup conditions	Epilepsy cases *n*	Sample size *N*	Epilepsy prevalence %
Haveman et al. (2011) [55] 4 (1/2/1)	Carer interview	Diagnosis epilepsy	ns	351[a]	1253	28
Hove and Havik (2010) [47] 1 (0/1/0)	Informant questionnaire (personnel)	Epilepsy, ns	ns	134[a]	593	22.6
Jelliffe-Pawlowski et al. (2003) [25] 5 (1/3/1)	Service records	Diagnosis epilepsy	ns	160	603	26.5[a]
Koskentausta et al. (2002) [15] 4 (0/3/1)	Case records	Epilepsy, ns	Psychiatrically non-disturbed 35%, disturbed 37%	55	155	35.5
Lakhan (2013) [61] 7 (0/4/3)	Examination. EEG if symptoms of epilepsy	Epilepsy, ns	CP 46.3%, DS 10.5%	62	262	23.7
Lewis et al. (2000) [58] (2000) 3 (1/2/0)	Carer interview	Seizures or epilepsy, lifetime	ns.	115	392	29.3
Lin et al. (2003) [62] 1 (0/1/0)	Parent or carer questionnaire	Epilepsy, ns	ns	262	1116	23.5[a]
Matthews et al. (2008) [46] 3 (1/2/0)	Carer interview. If epilepsy, visit by epilepsy nurse and information assessed by 2 doctors (and neuropsychiatrist if needed)	Diagnosis epilepsy	ns	58	318	18.2
McBrien and Macken (2009) [35] 3 (0/3/0)	Medical records (moderate), routine medical review (severe or profound)	Epilepsy, ns	ns	35	97	36.1
McCarron et al. (2014) [10] 6 (1/2/3)	Questionnaire and interview (carer)	Diagnosis epilepsy	DS and dementia 52.2%, DS controlling for dementia 13.4%	229	747	30.7
McDermott et al. (2005) [26] 6 (2/3/1)	Medical records	≥1 AED	DS 13.6%, CP 40%	186[a]	663	28.1
McGrother et al. (2006) [22] 6 (1/2/3)	Carer interviews	Suffers epilepsy (ns), seizures ≥occasionally, or on AED	ns	620	2393	25.9
Memisevic and Sinanovic (2009) [51] 7 (1/3/3)	Medical records	Diagnosis epilepsy	DS 0%, other genetic cause 31.8%, brain injury 48.6%	34	167	20.4
Molteno et al. (2001) [57] 1 (0/1/0)	Teacher questionnaire	Epilepsy, ns	ns	84	355	23.7
Morgan et al. (2003) [9] 7 (1/3/3)	Codes in multiple databases	Epilepsy code in inpatient, MH hospital or mortality datasets, or on epilepsy clinic database	ns	257	1595	16.1
Murphy et al. (1995) [27] 4 (0/3/1)	Records	Epilepsy, ns	ns	157	1074	14.6[a]
Nordin and Gillberg (1996) [40] 6 (2/4/0)	Clinical interview and medical examination	≥1 SZ or AED in last yr or SZs important part of medical history	ns	22	101	21.8

Pawar and Akuffo (2008) [33] 7 (1/3/3)	Case records	Diagnosis epilepsy	ns	53	177	29.9
Schieve et al. (2009) [29] 4 (1/2/1)	Family carer interview	SZ past 12 mths	DS 1.4%, non-DS 16.3% (weighted estimates)	98	750	13.1[a]
Schieve et al. (2012) [28] 3 (1/2/0)	Carer interview	SZ past 12 mths	ns	ns 36[a]	238	15.1 (weighted)
Steffenburg et al. (1995) [41] 6 (2/3/1)	Medical files	ILAE Active ≥2 unprovoked SZ and ≥1 SZ in last 5 yrs	CP 72.4%[a]	98	378	25.9
Strømme and Hagberg (2000) [48] 7 (2/4/1)	Parent interview and examination	ILAE, epilepsy	ns	35	178	19.7
Temtamy et al. (1994) [63] 4 (0/4/0)	Clinical examination	Epilepsy, ns	ns	5	116	4.3
Tenenbaum et al. (2012) [64] 1 (0/1/0)	Residential centre report, assumed records	Epilepsy, ns	ns	2313	7067	32.7
van Schrojenstein Lantman-de Valk et al. (2000) [45] 4 (1/3/0)	Electronic GP medical records	Epilepsy code N88 ICPC	ns	35	318	11.0
van Schrojenstein Lantman-de Valk et al. (1997) [44] 4 (0/1/3)	GP questionnaire	Epilepsy, ns	DS 10.7%, non-DS 17.4%. OR for epilepsy if dementia 8.8 (95% CI 4.8–16.2)	167[a]	1020	16.4
Wellesley et al. (1992) [59] 7 (2/3/2)	Records. Clarification if needed via examination or contacting doctor (main source records)	≥2 major or minor convulsions in absence of fever	ns	208	1590	13.1
Wong (2011) [65] 5 (1/1/3)	Nursing staff questionnaire	Diagnosis epilepsy	DS 13.2%, non-DS 37.0%	276	811	34.0
Yousef (1995) [66] 6 (0/3/3)	School records and teachers to clarify if necessary	Epilepsy, ns	ns	75	379	19.8
Down syndrome studies						
Collacott (1993) [23] 7 (2/3/2)	Carer interviews and medical records	≥3 SZ in 2 yrs, lifetime	Dementia 27.8%	35	351	10.0
Johannsen et al. (1996) [54] 7 (2/4/1)	Parent/carer interview and examination	ILAE, epilepsy	ns	12	72	16.7
McCarron et al. (2005) [36] 4 (0/3/1)	Medical records	Epilepsy, ns	AD 55.5%, non-AD 11.4%, end-stage AD 84.0%, mid-stage AD 39.4%	42	124	33.9
McVicker et al. (1994) [50] 7 (2/3/2)	Medical records	≥1 SZ in prior 2 yrs and/or on AED	ns	18	191	9.4
Prasher (1995) [34] 4 (2/2/0)	Carer interview	≥3 SZ in a 2 yr period and/or on AED (excludes partial complex SZ)	Dementia in 34.4% of those with epilepsy, total *n* with dementia not identified	32	201	15.9
Pueschel et al. (1991) [30] 4 (1/3/0)	Medical records and parent questionnaire	SZ disorder, exclude single provoked or unprovoked SZ	ns	33	405	8.1

Table 1 (*Continued*)

Authors, year and quality score[b]	Method epilepsy ascertainment	Epilepsy definition	Epilepsy prevalence % in main subgroup conditions	Epilepsy cases *n*	Sample size *N*	Epilepsy prevalence %
Roizen et al. (2014) [31] 2 (1/1/0)	Parental questionnaire	Diagnosis seizures	ns	30	440	6.8
Tyrrell et al. (2001) [37] 7 (2/4/1)	Medical notes, assessment (carer present)	ILAE, epilepsy	Dementia 65.8%, non-dementia 13.5%	58	283	20.5[a]

Abbreviations: ns, not stated; SZ, seizures; ILAE, International League Against Epilepsy; AED, anti-epileptic drug; DS, Down syndrome; ID, intellectual disabilities; LD, learning disabilities; CLDT, community learning disability team; MH, 'mental handicap'; SPID, Severe or profound intellectual disabilities; LS, living situation; CP, cerebral palsy; BPSMR, Board for Provision of Services to the Mentally Retarded; MCA, multiple congenital anomalies; AE, acquired encephalopathy; FXS, Fragile X syndrome; CNS, central nervous system; TQQ, the 'Ten Questions' Questionnaire; OR, odds ratio; RS, Rett syndrome.

[a] Calculated from available figures not reported directly.

[b] Presented as Total score (epilepsy definition score/ascertainment of epilepsy score/subgroup analysis score).

[c] ←, included in previous figure; →, included in next figure.

[d] entry age includes those without ID in main study

- Age
- Gender
- Level of intellectual disability
- Other – prevalence for other subgroup(s) given (e.g. those with dementia)

A score was awarded if the information was presented in a bar chart, or in an alternative format such as relative risk. Scores could range from 0 to 4. Studies were not excluded based on quality scores and scores are presented in the first column of Table 1.

2.4. Meta-analysis

For each study, the sample size and number of cases of epilepsy in the sample were entered as effect size data in Comprehensive Meta-Analysis Version 2.2 software (www.Meta-Analysis.com). Prevalence estimates were pooled using random effects meta-analysis. For the main random effects pooled estimates, heterogeneity between studies was summarised using I^2 and Q statistics. Subgroup analyses were conducted using between study moderator variables and within study subgroups. To compare across subgroups, the Q-test was used. Statistical significance was set at p value $< .05$.

3. Results

The process of identifying studies for inclusion is summarised in Appendix B. Electronic database searches identified a total of 1332 references, with 1099 remaining after removal of duplicates. Following the first examination of studies, 144 remained in a pool of articles relating to prevalence or mortality. After examination of full text articles from this pool and the addition of articles cited within these, 48 articles met the criteria for inclusion in relation to the prevalence of epilepsy and these are summarised in Table 1. Studies only including people with Down syndrome are presented separately at the end of Table 1.

3.1. Geographical spread

The majority of studies (42) were from high income countries, with just six studies from Low and Middle Income countries. The studies included a wide range of countries, with the greatest number for one country being seven studies from the United States [25–31].

A large number were from European countries: five were from England [22,23,32–34]; four from Ireland [10,35–37]; four from Sweden [38–41]; three from Finland [15,42,43]; three from the Netherlands [14,44,45]; two from Wales [9,46]; two from Norway [47,48]; one study each from Scotland [49], Northern Ireland [50], Bosnia and Herzegovina [51], Italy [52], France [53], and Denmark [54]; and one study included 14 European countries [55].

In addition, there were two studies from South Africa [56,57] and two from Australia [58,59]. Finally, one study each was included from the following countries: New Zealand [60]; India [61]; Taiwan [62]; Egypt [63]; Israel [64]; Hong Kong [65]; and Jordan [66].

3.2. Study design

Studies were almost entirely cross-sectional and based on retrospective review of records, questions completed either by self-report or interview, or clinical examination. There were three prospective cohort studies [42,44,49] although in the latter authors only present prevalence rates for the last data collection round. In

one retrospective study people could be included in more than one age-band estimate as there was an average of 12 years of follow-up for those with disabilities [26].

3.3. Meta-analysis

Two studies were excluded from meta-analyses as they focused on seizures in the last 12 months rather than epilepsy per se [28,29]. For prospective or retrospective cohort studies where people could be included in prevalence estimates at more than one time point, only the most recent data collection point was included. Analyses looked at subgroups using between study moderator variables, and also within study subgroups.

3.3.1. General samples versus samples of people with Down syndrome

An a priori decision was taken to compare studies based solely on samples of people with Down syndrome to general samples of people with intellectual disabilities. This was done in view of evidence suggesting that the prevalence of epilepsy is lower in people with Down syndrome than in general samples of people with intellectual disabilities (although these general sample figures are likely to include a number of people with Down syndrome). Whether or not studies included only people with Down syndrome was used as a between study moderator variable (see Table 2). The pooled estimate for 38 studies of general samples of people with intellectual disabilities was 22.2% (95% CI 19.6–25.1). There was significant heterogeneity between the studies (I^2 = 96.4%, Q = 1025.2, df = 37, $p < .001$). The pooled estimate for studies including only people with Down syndrome was 13.6% (95% CI 9.9–18.4). There was significant heterogeneity between studies (I^2 = 91.7%, Q = 84.3, df = 7, $p < .001$). Fig. 1 presents a forest plot of the 38 studies based on general samples and the 8 studies based on samples of people with Down syndrome only.

3.3.2. Level of intellectual disability

For studies using general samples of people with intellectual disabilities, level of intellectual disability was used as a between study moderator variable (see Table 2). This classified studies as: 'All' (study representative of all levels of intellectual disability); 'Less' (study representative of those with less severe intellectual disabilities e.g. excludes those with severe/profound intellectual disability); 'More' (study representative of those with more severe intellectual disabilities e.g. excludes those with mild intellectual disability). The pooled estimate for studies including all levels of intellectual disability was 22.2% (95% CI 19.6–25.0), whereas the estimate for studies classed as 'less severe' was 7.3% (95% CI 4.5–11.6) and the estimate for 'more severe' 41.6% (95% CI 32.1–51.8). In view of the effect of level of intellectual disability on pooled prevalence estimates, subsequent analyses only included those 29 studies which included all levels of intellectual disability.

3.3.3. Age group

Broad age group was used as a between study moderator variable for the 29 studies which included all levels of intellectual disability and which were not restricted to people with Down syndrome. Age group was classed as adult, child, or mixed (adult and child). This was based on the main age group of the study sample, so for example a study would be classed as 'adult' if it included mainly adults and a small number of 16 year olds, and a

Table 2
Random effects meta-analysis pooled estimates of prevalence of epilepsy.

Subgroup	Number of studies	Prevalence %[a]	95% CI lower	95% CI upper	
Down syndrome					Q 8.7, df 1, p .003
Mixed sample	38	22.2	19.6	25.1	
Down syndrome only	8	13.6	9.9	18.4	
Level of ID[b]					Q 43.4, df 2, $p < .001$
All	29	22.2	19.6	25.0	
Less severe	4	7.3	4.5	11.6	
More severe	5	41.6	32.1	51.8	
Age[b,c]					Q 0.8, df 2, p .661
Adult	12	23.5	19.5	28.0	
Child	12	21.7	17.9	26.1	
Mixed	5	20.2	15.0	26.7	
High/LAMI[b,c]					Q 0.2, df 1, p .626
High	25	22.4	19.7	25.4	
LAMI	4	20.5	14.5	28.2	
Subgroup (including within study subgroups)					
Level of ID					Q 56.0, df 1, $p < .001$
Mild	13	9.8	7.6	12.4	
Moderate/severe/profound	14	30.4	25.5	35.7	
Level of ID (where moderate, severe, profound available separately)					Q 16.6, df 2, $p < .001$
Moderate	5	16.7	10.8	25.0	
Severe	3	27.0	16.1	41.5	
Profound	4	50.9	36.1	65.5	
Gender (any study where male/female figures given separately)					Q 0.4, df 1, p .524
Male	9	24.8	19.6	30.8	
Female	9	22.2	17.3	28.1	
Age groups[b,c]					Q 2.2, df 2, p .339
0–18	11	21.6	17.9	25.9	
19–49	8	26.0	21.2	31.5	
50+	7	21.5	17.0	26.9	

[a] Estimates based on meta-analysis using random effects model.
[b] Excludes DS only studies.
[c] Excludes less/more severe ID studies.

Group by DS	Study name	Event rate	Lower limit	Upper limit	Z-Value	p-Value
No	Airaksinen, Matilainen, Mononen et al (2000)	0.212	0.154	0.284	-6.596	0.000
No	Arvio & Sillanpää (2003)	0.518	0.473	0.564	0.792	0.429
No	Benassi, Guarino, Cammarata et al (1990)	0.300	0.214	0.402	-3.684	0.000
No	Christianson, Zwane, Manga et al (2002)	0.155	0.115	0.207	-9.460	0.000
No	David, Dieterich, de Villemeur et al (2014)	0.028	0.012	0.065	-7.852	0.000
No	Dekker & Koot (2003)	0.055	0.038	0.079	-14.112	0.000
No	Fernell (1998)	0.266	0.172	0.386	-3.593	0.000
No	Forsgren, Edvinsson, Blomquist et al (1990)	0.202	0.182	0.223	-21.204	0.000
No	Gittins & Rose (2008)	0.639	0.512	0.749	2.147	0.032
No	Goulden, Shinnar, Koller et al (1991)	0.153	0.111	0.208	-9.025	0.000
No	Hand (1996)	0.167	0.145	0.190	-19.562	0.000
No	Haveman, Perry, Salvador-Carulla et al (2011)	0.280	0.256	0.306	-15.003	0.000
No	Hove & Havik (2010)	0.226	0.194	0.261	-12.539	0.000
No	Jelliffe-Pawlowski, Shaw, Nelson et al (2003)	0.265	0.232	0.302	-11.041	0.000
No	Koskentausta, Iivanainen & Almqvist (2002)	0.355	0.284	0.433	-3.561	0.000
No	Lakhan (2013)	0.237	0.189	0.292	-8.057	0.000
No	Lewis, Tonge, Mowat et al (2000)	0.293	0.250	0.340	-7.925	0.000
No	Lin, Wu & Lee (2003)	0.235	0.211	0.261	-16.731	0.000
No	Matthews, Weston, Baxter et al (2008)	0.182	0.144	0.229	-10.331	0.000
No	McBrien & Macken (2009)	0.361	0.272	0.461	-2.704	0.007
No	McCarron, O'Dwyer, Burke et al (2014)	0.307	0.275	0.341	-10.286	0.000
No	McDermott, Moran, Platt et al (2005)	0.281	0.248	0.316	-10.894	0.000
No	McGrother, Bhaumik, Thorp et al (2006)	0.259	0.242	0.277	-22.520	0.000
No	Memisevic & Sinanovic (2009)	0.204	0.149	0.271	-7.098	0.000
No	Molteno, Molteno, Finchilescu et al (2001)	0.237	0.195	0.284	-9.379	0.000
No	Morgan, Baxter & Kerr (2003)	0.161	0.144	0.180	-24.225	0.000
No	Murphy, Yeargin-Allsopp, Decouflé et al (1995)	0.146	0.126	0.169	-20.434	0.000
No	Nordin & Gillberg (1996)	0.218	0.148	0.309	-5.303	0.000
No	Pawar & Okuffo (2008)	0.299	0.237	0.371	-5.179	0.000
No	Steffenburg, Hagberg, Viggedal et al (1995)	0.259	0.218	0.306	-8.945	0.000
No	Strømme & Hagberg (2000)	0.197	0.145	0.262	-7.463	0.000
No	Temtamy, Kandil, Demerdash et al (1994)	0.043	0.018	0.099	-6.781	0.000
No	Tenenbaum, Fuchs, Raskas et al (2012)	0.327	0.316	0.338	-28.418	0.000
No	van Schrojenstein Lantman-de Valk et al (2000)	0.110	0.080	0.149	-11.665	0.000
No	van Schrojenstein Lantman-de Valk et al (1997)	0.164	0.142	0.188	-19.272	0.000
No	Wellesley, Hockey, Montgomery et al (1992)	0.131	0.115	0.148	-25.463	0.000
No	Wong (2011)	0.340	0.309	0.374	-8.931	0.000
No	Yousef (1995)	0.198	0.161	0.241	-10.855	0.000
No		0.222	0.196	0.251	-15.273	0.000
Yes	Collacott (1993)	0.100	0.072	0.136	-12.352	0.000
Yes	Johannsen, Christensen, Goldstein et al (1996)	0.167	0.097	0.271	-5.089	0.000
Yes	McCarron, Gill, McCallion et al (2005)	0.339	0.261	0.426	-3.526	0.000
Yes	McVicker, Shanks & McClelland (1994)	0.094	0.060	0.145	-9.137	0.000
Yes	Prasher (1995)	0.159	0.115	0.216	-8.632	0.000
Yes	Pueschel, Louis & McKnight (1991)	0.081	0.059	0.112	-13.337	0.000
Yes	Roizen, Magyar, Kuschner et al (2014)	0.068	0.048	0.096	-13.826	0.000
Yes	Tyrrell, Cosgrave, McCarron et al (2001)	0.205	0.162	0.256	-9.206	0.000
Yes		0.136	0.099	0.184	-10.031	0.000
Overall		0.178	0.108	0.280	-5.143	0.000

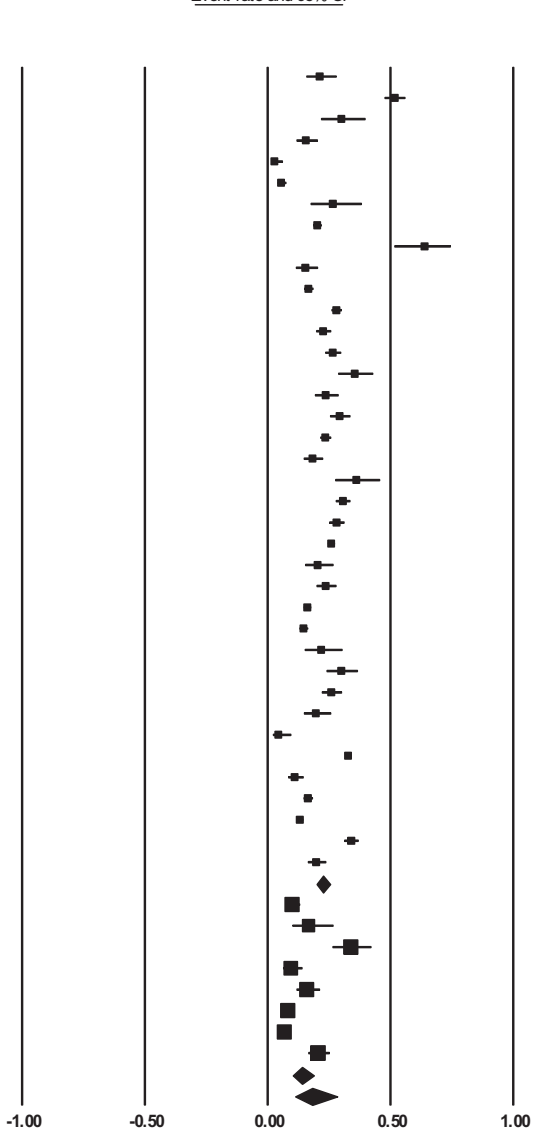


Fig. 1. Forest plot of prevalence for mixed samples versus Down syndrome only.

study would be classed as 'child' if it included mainly children and a small number of 20 year olds. Estimates for these broad age groups did not differ significantly (see Table 2).

3.3.4. Country economy

Country economy (High or Low and Middle Income (LAMI)) was also used as a between study moderator variable for the 29 studies which included all levels of intellectual disability and which were not restricted to people with Down syndrome. Countries in which studies were undertaken were classed as 'high income' or 'low and middle income' based on the World Bank list of economies [67]. This classifies countries according to 2013 gross national income (GNI) per capita: low income, $1045 or less; lower middle income, $1046–4125; upper middle income, $4126–12,745; and high income, $12,746 or more. Taiwan (not included in country classification) was classed as High Income. One study included 14 European countries of which one was upper middle income and this study overall was classed as 'high income' [55]. There was no significant difference in the pooled estimates (see Table 2).

3.4. Within study subgroup analyses

Further meta-analyses were then conducted which included information on prevalence from within study subgroups, for example where studies presented prevalence rates separately by level of intellectual disability, gender or age bands. Studies which only included a relevant subgroup (e.g. a sample including only people with mild intellectual disability) were also included in these analyses.

3.4.1. Level of intellectual disability

For level of intellectual disability, firstly prevalence rates were included for those with mild intellectual disability and the combined prevalence for those with moderate, severe or profound intellectual disability. Combining moderate, severe and profound intellectual disability was done to maximise the number of studies that could be included as few studies presented results for each of these three levels of intellectual disability separately. The pooled estimate for moderate/severe/profound intellectual disability from 14 studies was 30.4% (95% CI 25.5–35.7) compared to

9.8% (95% CI 7.6–12.4) from 13 studies for those with mild intellectual disability (see Fig. 2).

Pooled estimates were also calculated for the studies which did provide separate estimates for any of the moderate, severe or profound categories. For moderate intellectual disability, the pooled estimate was 16.7% (95% CI 10.8–25.0), compared to 27.0% (95% CI 16.1–41.5) for severe intellectual disability and 50.9% (95% CI 36.1–65.5) for profound intellectual disability.

3.4.2. Gender

Where male and female prevalence figures were given separately, pooled estimates were male 24.8% (95% CI 19.6–30.8) and female 22.2% (17.3–28.1). One study in the male/female subgroup analysis only included those with mild or moderate intellectual disability but was nonetheless included in the analysis [51].

3.4.3. Age group

Studies presenting results separately for age bands were considered using age band as a subgroup within study. Studies presenting results for only one age band were also included in this analysis. The broad age bands used were 0–18, 19–49, and 50+. However, a 5 year leeway was given for these age bands at both the upper and lower limit so, for example, a figure for those aged 19–54 or 17–54 would be included in the 19–49 category. Age bands from McDermott et al. [26] were not included as due to participants having an average of 12 years of follow-up time a person could be in more than one age band and they were thus not independent subgroups. Figures for a specific age (e.g. age 22) were included in the appropriate age band. Overall, there was not a significant difference by age band although the prevalence for age band 19–49 (26.0% (95% CI 21.2–31.5)) was slightly higher than that for the 0–18 age band (21.6% (95% CI 17.9–25.9) and the 50+ age group (21.5% (95% CI 17.0–26.9).

3.5. Down syndrome

Eight studies focussed exclusively on people with Down syndrome [23,30,31,34,36,37,50,54]. A further eight studies included some results disaggregated for people with Down syndrome in the overall sample. Results from meta-analyses in relation to people with Down syndrome are given in Table 3. In these analyses, prevalence rates from studies looking only at people with Down syndrome were combined with prevalence rates given in other studies which presented results for people with Down syndrome as a within study subgroup (excluding studies which did not include all levels of intellectual disability). No rates disaggregated by gender were identified.

Firstly, pooled prevalence for people with Down syndrome was estimated by combining the prevalence rates from studies looking only at people with Down syndrome with prevalence rates for people with Down syndrome presented as a within study subgroup (excluding studies which did not include all levels of intellectual disability). The pooled estimate was 12.4% (95% CI 9.1–16.7). There was significant heterogeneity between studies, $I^2 = 87.4\%$, $Q = 95.3$, df = 12, $p < .001$.

Pooled prevalence was also estimated for age bands. This showed a significant effect of age band, with the pooled estimate rising from 6.9% (95% CI 3.8–12.0) at age 0–18, to 9.0% (95% CI 5.9–13.5) at age 19–49, and 26.0% (95% CI 16.1–39.2) at age 50+.

In view of the increased rate of epilepsy in older people with Down syndrome, overall prevalence was then estimated excluding two studies which looked at samples of people with Down syndrome aged 35+ only [36,37]. Based on data from 11 studies, the pooled estimate was 10.3% (95% CI 8.4–12.6), $I^2 = 57.0\%$, $Q = 23.2$, df = 10, $p < .01$. However, it should be noted that these studies did not include all age bands, with some including only adults and other including only children.

Finally, a small number of studies presented prevalence rates separately for those with and without Alzheimer's disease/dementia. The pooled prevalence for those with Alzheimer's/dementia was 53.3% (95% CI 41.9–64.4) compared to 12.8% (95% CI 7.7–20.4) for those specifically noted not to have Alzheimer's/dementia. It is not possible to give the mean age for those with and without Alzheimer's disease/dementia overall. However, the mean age for both groups is available in two studies: 54.7 (SD 7.5) for those with Alzheimer's disease/dementia compared to 45.6 (SD 7.3) for those without [37]; and 55.4 (SD 7.0) for those with and 50.8 (SD 5.8) for those without [36] (see Fig. 3).

3.6. Co-morbidity

A number of studies presented data on co-morbidities in people with intellectual disabilities and epilepsy.

Group by Subgroup within study	Study name	Subgroup within study	Statistics for each study				
			Event rate	Lower limit	Upper limit	Z-Value	p-Value
Mild	Airaksinen, Matilainen, Mononen et al (2000)	Mild	0.068	0.028	0.152	-5.667	0.000
Mild	Christianson, Zwane, Manga et al (2002)	Mild	0.133	0.092	0.189	-8.885	0.000
Mild	David, Dieterich, de Villemeur et al (2014)	Mild	0.028	0.012	0.065	-7.852	0.000
Mild	Forsgren, Edvinsson, Blomquist et al (1990)	Mild	0.112	0.084	0.148	-12.665	0.000
Mild	Goulden, Shinnar, Koller et al (1991)	Mild	0.101	0.063	0.156	-8.566	0.000
Mild	Jelliffe-Pawlowski, Shaw, Nelson et al (2003)	Mild	0.135	0.102	0.177	-11.315	0.000
Mild	Lakhan (2013)	Mild	0.038	0.012	0.111	-5.491	0.000
Mild	Memisevic & Sinanovic (2009)	Mild	0.165	0.100	0.259	-5.552	0.000
Mild	Murphy, Yeargin-Allsopp, Decouflé et al (1995)	Mild	0.071	0.054	0.091	-18.093	0.000
Mild	Steffenburg, Hagberg, Viggedal et al (1995)	Mild	0.147	0.107	0.198	-9.605	0.000
Mild	Strømme & Hagberg (2000)	Mild	0.091	0.048	0.166	-6.586	0.000
Mild	Wellesley, Hockey, Montgomery et al (1992)	Mild	0.113	0.091	0.141	-16.196	0.000
Mild	Yousef (1995)	Mild	0.068	0.033	0.136	-6.688	0.000
Mild			0.098	0.076	0.124	-15.943	0.000
MSP	Airaksinen, Matilainen, Mononen et al (2000)	MSP	0.351	0.253	0.463	-2.580	0.010
MSP	Benassi, Guarino, Cammarata et al (1990)	MSP	0.300	0.214	0.402	-3.684	0.000
MSP	Christianson, Zwane, Manga et al (2002)	MSP	0.256	0.148	0.405	-3.055	0.002
MSP	Fernell (1998)	MSP	0.266	0.172	0.386	-3.593	0.000
MSP	Forsgren, Edvinsson, Blomquist et al (1990)	MSP	0.233	0.209	0.259	-16.728	0.000
MSP	Goulden, Shinnar, Koller et al (1991)	MSP	0.348	0.225	0.495	-2.031	0.042
MSP	Jelliffe-Pawlowski, Shaw, Nelson et al (2003)	MSP	0.411	0.355	0.469	-3.005	0.003
MSP	Lakhan (2013)	MSP	0.331	0.266	0.404	-4.406	0.000
MSP	McBrien & Macken (2009)	MSP	0.361	0.272	0.461	-2.704	0.007
MSP	Murphy, Yeargin-Allsopp, Decouflé et al (1995)	MSP	0.322	0.273	0.375	-6.253	0.000
MSP	Steffenburg, Hagberg, Viggedal et al (1995)	MSP	0.450	0.370	0.533	-1.181	0.238
MSP	Strømme & Hagberg (2000)	MSP	0.329	0.235	0.440	-2.974	0.003
MSP	Wellesley, Hockey, Montgomery et al (1992)	MSP	0.151	0.128	0.178	-17.620	0.000
MSP	Yousef (1995)	MSP	0.247	0.200	0.302	-7.964	0.000
MSP			0.304	0.255	0.357	-6.706	0.000
Overall			0.179	0.053	0.461	-2.186	0.029

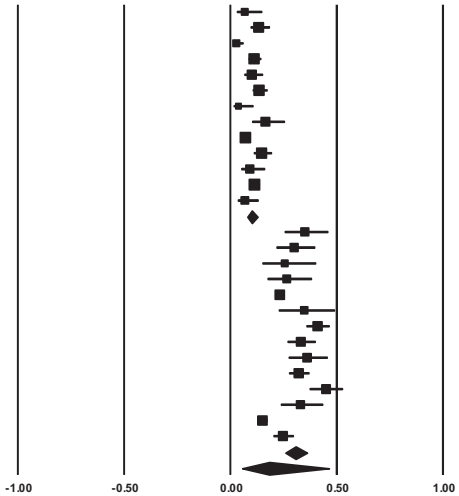


Fig. 2. Forest plot for prevalence mild versus moderate/severe/profound intellectual disability.

Table 3
Meta-analysis estimates for people with Down syndrome.

Subgroup	Number of studies	Prevalence %[a]	95% CI lower	95% CI upper	
Overall prevalence including subgroups in non-DS only studies[b]	13	12.4	9.1	16.7	
Overall prevalence including subgroups in non-DS only studies[b] excluding two studies on older people	11	10.3	8.4	12.6	
Age[b]					*Q* 15.0, df 2, *p* .001
0–18	2	6.9	3.8	12.0	
19–49	3	9.0	5.9	13.5	
50+	3	26.0	16.1	39.2	
Has Alzheimer's/dementia					*Q* 30.9, df 1, $p < .001$
Yes	4	53.3	41.9	64.4	
No	2	12.8	7.7	20.4	

[a] Estimates based on meta-analysis using random effects model.
[b] Excludes less/more severe ID studies.

3.6.1. Psychiatric and behavioural problems

One study reported that epilepsy was associated with higher levels of psychopathology [57] and one study (controlling for age, gender and level of understanding) found associations with epilepsy and some psychological and behaviour problems [22]. However, other studies found that people with intellectual disability and epilepsy were not more likely to have co-morbid psychiatric and/or behavioural problems than those with intellectual disabilities without epilepsy. Reported findings include: being significantly less likely to have behavioural disturbances (17.6% vs 27.9%) [43]; no significant difference in the prevalence of psychiatric disorders [15]; no significant differences in behavioural and emotional disturbance when controlling for level of intellectual disability [58]; no significant differences in psychopathology between matched epilepsy and non-epilepsy groups [46]; no association between epilepsy and mental health concerns, with 46.7% of those with epilepsy reporting mental health problems compared with 48.1% of those without epilepsy [10]; no association between epilepsy and the prevalence of challenging behaviour or psychiatric conditions [33]; and no significant difference in maladaptive behaviour scores for those with Down syndrome and epilepsy [34].

3.6.2. Physical impairments

People with intellectual disabilities and epilepsy were found to have more associated impairments (2.7) than those without epilepsy (1.2) and were more likely to have: speech handicap (73.6% versus 50.0%), motor handicap (54.4% versus 14.4%), and blindness (14.2% versus 1.4%) [43]. Other reported co-morbidities in those with epilepsy were: cerebral palsy (33.4%) and visual impairment (12.4%) [39]; cerebral palsy (36.4%) [49]; and cerebral palsy (43%) and visual impairment (24.5%) [41]. After adjusting for age, gender and level of understanding, those with epilepsy were more likely to have: a range of physical disabilities (adjusted OR 1.8, 95% CI 1.5–2.2); problems with wetting (OR 2.7, 95% CI 2.1–3.4), soiling (OR 2.2, 95% CI 1.6–3.1) and walking (OR 2.5, 95% CI 2.0–3.2) [22]. Those with intellectual disability were also found to be more likely to have joint disease (29.3% versus 16.8% for those with intellectual disability without epilepsy, adjusted OR 2.1, 95% CI 1.5–3.1), gastrointestinal disease (34.5% versus 23.4%, adjusted OR 1.8, 95% CI 1.3–2.5), and stroke (5.2% versus 1.9%, adjusted OR 3.3, 95% CI 1.4–9.0) [10].

4. Discussion

Despite the variation in reported prevalence rates between studies, it is clear that the prevalence of epilepsy is high in people with intellectual disabilities worldwide. The results suggest that in general samples of people with intellectual disabilities, approximately one in five people will have epilepsy, with the pooled estimate from 38 studies being 22.2% (95% CI 19.6–25.1). For samples of people with Down syndrome excluding two studies focusing on older people, the rate is lower with approximately one in ten people having epilepsy, with the pooled estimate from data in 11 studies being 10.3% (95% CI 8.4–12.6). In studies where this information was available, those with intellectual disabilities and epilepsy had more physical impairments than those without epilepsy. However, whilst psychiatric or behavioural co-morbidity was common, rates were not necessarily higher than in those with intellectual disabilities without epilepsy. A review specifically addressing co-morbidity in people with intellectual disabilities

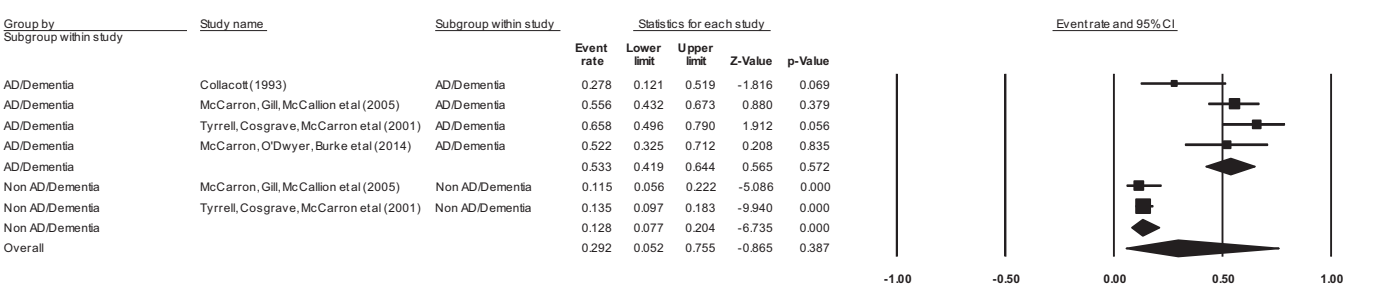


Fig. 3. Forest plot for prevalence by Alzheimer's/dementia for people with Down syndrome.

and epilepsy extending beyond studies that present figures on prevalence (e.g. [68]) would be a useful addition to the literature.

The prevalence of epilepsy is related to level of intellectual disability. In 29 studies which included all levels of intellectual disability, the pooled estimate was 22.2% (95% CI 19.6–25.0), whilst for four studies with samples skewed towards less severe intellectual disability the pooled estimate was 7.3% (95% CI 4.5–11.6) and for five studies skewed towards more severe intellectual disability the pooled estimate was 41.6% (95% CI 32.1–51.8). Similarly, data from 13 studies gives a pooled estimate for those with mild intellectual disability of 9.8% (95% CI 7.6–12.5) compared to 30.4% (95% CI 25.5–35.7) for those with moderate, severe or profound intellectual disability. Few studies give figures separately for those with moderate, severe or profound intellectual disability but it is clear that prevalence increases with level of intellectual disability. The pooled estimate for moderate intellectual disability from five studies was 16.7% (95% CI 10.8–25.0), for severe intellectual disability from three studies 27.0% (95% CI 16.1–41.5) and for profound intellectual disability from four studies 50.9% (95% CI 36.1–65.5).

Age was not found to be a significant factor for general samples of people with intellectual disabilities, although the rate for those aged 19–49 was slightly higher at 26.0% (95% CI 21.2–31.5) than for 0–18 year olds (21.6%, 95% CI 17.9–25.9) and 50+ year olds (21.5%, 95% CI 17.0–26.9). However, for people with Down syndrome there was a clear increase in prevalence with age. Data from two studies for those aged 0–18 gave a pooled estimate of 6.9% (95% CI 3.8–12.0), compared to 9.0% (95% CI 5.9–13.5) for three studies giving data for 19–49 year olds and 26.0% (95% CI 16.1–39.2) for three studies giving data for those aged 50 or more. An increase with age was also found for people with Down syndrome in a study by van Schrojenstein Lantman-de Valk et al. [44], with the rates being 4.9% at age 0–19, rising to 36.4% for those age 60 or more. However, it was not possible to include these figures in the meta-analysis as sample sizes for individual age bands were not identified. Similarly, an increasing prevalence of epilepsy with age was found for a small sample of people with Down syndrome [26] but these figures could not be included in the meta-analysis due to participants being included in more than one age band estimate depending on the number of years the person was followed up for.

Overall, it is clear that for people with Down syndrome, epilepsy prevalence increases with age. This increase is likely to be mainly accounted for by the increasing presence of Alzheimer's disease/dementia in people with Down syndrome as they age. The pooled estimate for those with Alzheimer's disease/dementia from four studies was 53.3% (95% CI 41.9–64.4) compared to 12.8% (95% CI 7.7–20.4) for two studies explicitly giving data for those without Alzheimer's/dementia. Further, in one study, epilepsy was found here to be significantly more common in persons at end-stage (84.0%) versus persons at mid-stage Alzheimer's disease (39.4%) [36].

4.1. Limitations

There are a number of limitations to this review. Whilst studies were identified from a large range of countries, the review is restricted to English language publications. All data was extracted by one reviewer and extraction of data by two reviewers independently would have reduced the possibility of errors. In some instances it was necessary to calculate figures from reported data as they were not reported explicitly (e.g. obtaining the number of epilepsy cases from the overall sample number and reported prevalence rate or vice versa) and two minor discrepancies arose. Firstly, calculating figures from McVicker et al. [50] on prevalence by age band resulted in a total number of epilepsy cases of 19 compared to a reported number of 18. Secondly, calculating figures from Wong [65] on prevalence in a subgroup with Down syndrome resulted in a prevalence rate of 13.1% compared to a rate of 13.2% as reported in the article.

Ideally, the same definition of epilepsy should be used across studies to allow comparison of prevalence rates [4]. However, many of the studies identified did not present a definition of epilepsy, generally referring to either a diagnosis of epilepsy or the presence of epilepsy. The lack of detail given regarding the definition of epilepsy in many studies means that it is not possible to determine whether reported prevalence rates related to active epilepsy or lifetime epilepsy. The issue of defining epilepsy is not straightforward [69]. Where definitions were provided, these included standard definitions based on International League Against Epilepsy (ILAE) criteria and other definitions specifying variable criteria in relation to number of seizures, anti-epileptic drug (AED) use and time spans. In addition, the source of information used to ascertain epilepsy is variable between studies which may lead to varying levels of accuracy in obtained rates.

In addition to the variation in prevalence rates that is likely to be due to differences in the definition of epilepsy used and the source of data in studies, there is also likely to be an unknown number of cases where epilepsy has been misdiagnosed due to the misinterpretation of behavioural, physiological, syndrome related, medication related or psychological events by parents, paid carers and health professionals [70].

Finally, the review has focussed on prevalence in the general population of people with intellectual disabilities or Down syndrome and has not included studies on less common syndromes such as Fragile X syndrome. Future review work could consider prevalence in a greater range of specific syndromes associated with intellectual disabilities.

5. Conclusion

This review aims to provide an up to date summary of research on the prevalence of epilepsy in people with intellectual disabilities. The pooling of estimates from studies, and the examination of factors which account for some of the heterogeneity of reported prevalence rates between studies, allows for the provision of more robust figures on prevalence. With around one in five people with intellectual disabilities having epilepsy, 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 [71] 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.

Conflict of interest statement

There is no conflict of interest.

Acknowledgements

This work was supported by Public Health England. However, the findings and views reported in this paper are those of the authors and should not be attributed to Public Health England. We would like to thank Howard Ring for comments on the draft of this manuscript.

Appendix A. Electronic search strategy

MEDLINE AND CINAHL

Limits: 1990; English; Human

(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 (MH "Developmental Disabilities/EP/MO") OR (MH "Intellectual Disability+/EP/MO") 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 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

(MH "Epilepsy+/MO/EP") OR (TI epilep* OR TI seizure* OR TI convulsi* OR AB epilep* OR AB seizure* OR AB convulsi*)

AND

(TI incidence OR TI prevalence OR TI mortality OR TI death OR AB incidence OR AB prevalence OR AB mortality OR AB death) OR (MH "Incidence") OR (MH "Prevalence") OR (MH "Mortality+")

PSYCINFO

Limits: 1990, Peer review, English, Exclude dissertations

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 TI convulsi* OR AB epilep* OR AB seizure* OR AB convulsi*)

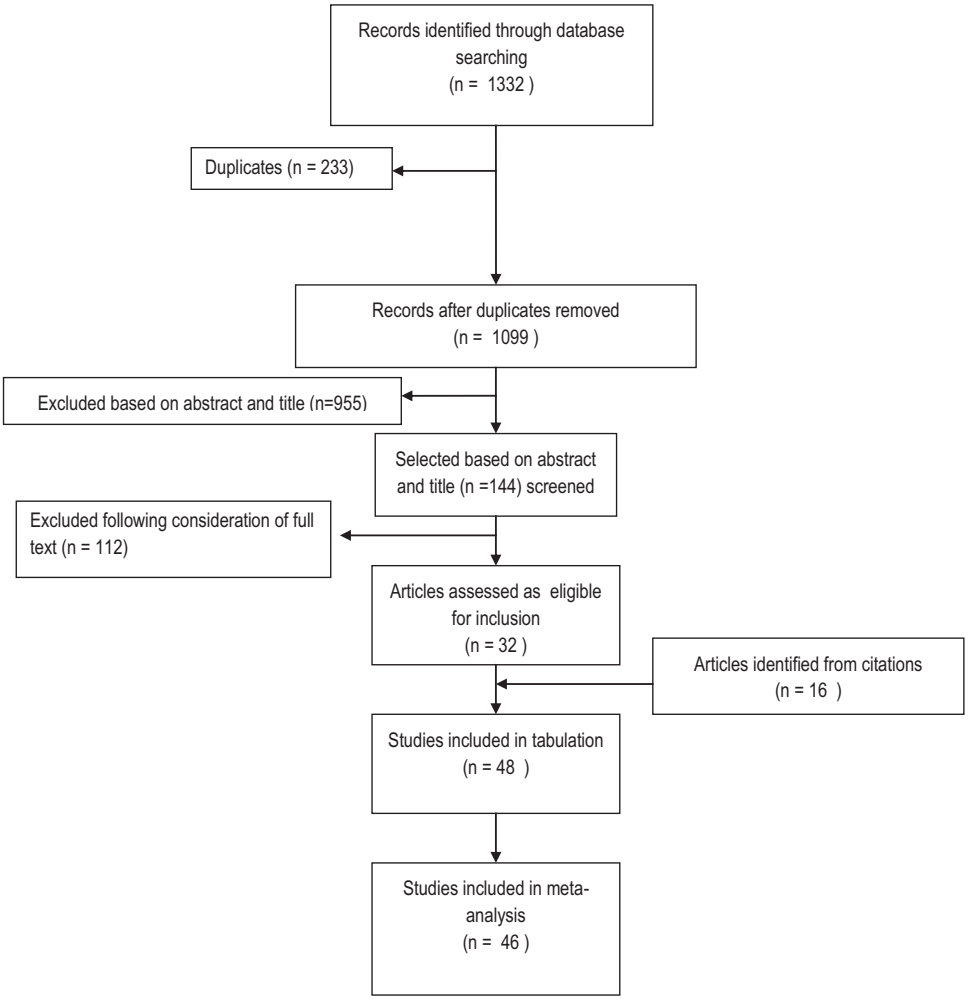
AND

(TI incidence OR TI prevalence OR TI mortality OR TI death OR AB incidence OR AB prevalence OR AB mortality OR AB death) OR DE "Epidemiology" OR DE "death and dying" OR DE "mortality rate"

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"

Appendix B. Flowchart of study identification



References

[1] Einfeld S, Emerson E. Intellectual disability. In: Rutter M, Bishop D, Pine D, Scott S, Stevenson J, Taylor E, et al., editors. Rutter's child and adolescent psychiatry. Oxford: Blackwell; 2008.
[2] Maulik PK, Mascarenhas MN, Mathers CD, Dua T, Saxena S. Prevalence of intellectual disability: a meta-analysis of population-based studies. Res Dev Disabil 2011;32:419–36.
[3] Hatton C, Emerson E, Glover G, Robertson J, Baines S, Christie A. People with learning disabilities in England 2013. London: Public Health England; 2014.
[4] Forsgren L, Beghi E, Öun A, Sillanpää M. The epidemiology of epilepsy in Europe – a systematic review. Eur J Neurol 2005;12:245–53.
[5] Ngugi AK, Bottomley C, Kleinschmidt I, Sander JW, Newton CR. Estimation of the burden of active and life-time epilepsy: a meta-analytic approach. Epilepsia 2010;51:883–90.
[6] Linehan C, Kerr MP, Walsh PN, Brady G, Kelleher C, Delanty N, et al. Examining the prevalence of epilepsy and delivery of epilepsy care in Ireland. Epilepsia 2010;51:845–52.
[7] Joint Epilepsy Council. Epilepsy prevalence, incidence and other statistics; 2011, Available from: http://www.jointepilepsycouncil.org.uk/downloads/2011/Joint%20Epilepsy%20Council%20Prevalence%20and%20Incidence%20September%2011.pdf [retrieved 22.07.14].
[8] Lhatoo SD, Sander JW. The epidemiology of epilepsy and learning disability. Epilepsia 2001;42(Suppl. 1):6–9.
[9] Morgan CLI, Baxter H, Kerr MP. Prevalence of epilepsy and associated health service utilization and mortality among patients with intellectual disability. Am J Mental Retard 2003;108:293–300.
[10] McCarron M, O'Dwyer M, Burke E, McGlinchey E, McCallion P. Epidemiology of epilepsy in older adults with an intellectual disability in Ireland: associations and service implications. Am J Intellect Dev Disabil 2014;119:253–60.
[11] Oeseburg B, Dijkstra GJ, Groothoff JW, Reijneveld SA, Jansen DEMC. Prevalence of chronic health conditions in children with intellectual disability: a systematic literature review. Intellect Dev Disabil 2011;49:59–85.
[12] Steffenburg S, Gillberg C, Steffenburg U. Psychiatric disorders in children and adolescents with mental retardation and active epilepsy. Arch Neurol 1996;53:904–12.
[13] Beavis J, Kerr M, Marson Anthony G, Dojcinov I. Pharmacological interventions for epilepsy in people with intellectual disabilities. Cochrane Database Syst Rev 2007;(3). http://dx.doi.org/10.1002/14651858.CD005399.pub2 [art. no.: CD005399].
[14] Dekker MC, Koot HM. DSM-IV disorders in children with borderline to moderate intellectual disability. I: prevalence and impact. J Am Acad Child Adolesc Psychiatry 2003;42:915–22.
[15] Koskentausta T, Iivanainen M, Almqvist F. Psychiatric disorders in children with intellectual disability. Nord J Psychiatry 2002;56:126–31.
[16] Sherman SL, Allen EG, Bean LH, Freeman SB. Epidemiology of Down syndrome. Mental Retard Dev Disabil Res Rev 2007;13:221–7.
[17] Leung HTT, Ring H. Epilepsy in four genetically determined syndromes of intellectual disability. J Intellect Disabil Res 2013;57:3–20.
[18] Blake P, Kerr M. Epilepsy. In: Taggart L, Cousins W., editors. Health promotion for people with intellectual and developmental disabilities. Maidenhead: McGraw Hill (Open University Press); 2014. p. 77–87.
[19] Brown S. Epidemiology of epilepsy in persons with intellectual disabilities. In: Prasher V, Kerr M, editors. Epilepsy and intellectual disabilities. London: Springer; 2008. p. 29–42.
[20] Cardoza B, Kerr M. Diseases of the nervous system. I: epilepsy, hydrocephalus and nervous system malformations. In: O'Hara J, McCarthy J, Bouras N, editors. Intellectual disability and ill health: a review of the evidence. Cambridge: Cambridge University Press; 2010.
[21] Kiani R, Tyrer F, Jesu A, Bhaumik S, Gangavati S, Walker G, et al. Mortality from sudden unexpected death in epilepsy (SUDEP) in a cohort of adults with intellectual disability. J Intellect Disabil Res 2014;58:508–20.
[22] McGrother CW, Bhaumik S, Thorp CF, Hauck A, Branford D, Watson JM. Epilepsy in adults with intellectual disabilities: prevalence, associations and service implications. Seizure 2006;15:376–86.
[23] Collacott RA. Epilepsy, dementia and adaptive behaviour in Down's syndrome. J Intellect Disabil Res 1993;37:153–60.
[24] Shamliyan T, Kane RL, Dickinson S. A systematic review of tools used to assess the quality of observational studies that examine incidence or prevalence and risk factors for diseases. J Clin Epidemiol 2010;63:1061–70.
[25] Jelliffe-Pawlowski LL, Shaw GM, Nelson V, Harris JA. Risk of mental retardation among children born with birth defects. Arch Pediatr Adolesc Med 2003;157:545–50.
[26] McDermott S, Moran R, Platt T, Wood H, Isaac T, Dasari S. Prevalence of epilepsy in adults with mental retardation and related disabilities in primary care. Am J Ment Retard 2005;110:48–56.
[27] Murphy CC, Yeargin-Allsopp M, Decouflé P, Drews CD. The administrative prevalence of mental retardation in 10-year-old children in metropolitan Atlanta, 1985 through 1987. Am J Public Health 1995;85:319–23.
[28] Schieve LA, Gonzalez V, Boulet SL, Visser SN, Rice CE, Van Naarden Braun K, et al. Concurrent medical conditions and health care use and needs among children with learning and behavioral developmental disabilities. National Health Interview Survey, 2006–2010. Res Dev Disabil 2012;33:467–76.
[29] Schieve LA, Boulet SL, Boyle C, Rasmussen SA, Schendel D. Health of children 3 to 17 years of age with Down syndrome in the 1997–2005 national health interview survey. Pediatrics 2009;123:e253.
[30] Pueschel SM, Louis S, McKnight P. Seizure disorders in Down syndrome. Arch Neurol 1991;48:318–20.
[31] Roizen NJ, Magyar CI, Kuschner ES, Sulkes SB, Druschel C, van Wijngaarden E, et al. A community cross-sectional survey of medical problems in 440 children with Down syndrome in New York State. J Pediatr 2014;164:871–5.
[32] Gittins D, Rose N. An audit of adults with profound and multiple learning disabilities within a West Midlands Community Health Trust – implications for service development. Br J Learn Disabil 2008;36:38–47.
[33] Pawar DG, Akuffo EO. Comparative survey of comorbidities in people with learning disability with and without epilepsy. Psychiatr Bull 2008;32:224–6.
[34] Prasher VP. Epilepsy and associated effects on adaptive behaviour in adults with Down syndrome. Seizure 1995;4:53–6.
[35] McBrien J, Macken S. Meeting the health care needs of school-age children with intellectual disability. Ir Med J 2009;102:252–5.
[36] McCarron M, Gill M, McCallion P, Begley C. Health co-morbidities in ageing persons with Down syndrome and Alzheimer's dementia. J Intellect Disabil Res 2005;49:560–6.
[37] Tyrrell J, Cosgrave M, McCarron M, McPherson J, Calvert J, Kelly A, et al. Dementia in people with Down's syndrome. Int J Geriatr Psychiatry 2001;16:1168–74.
[38] Fernell E. Aetiological factors and prevalence of severe mental retardation in children in a Swedish municipality: the possible role of consanguinity. Dev Med Child Neurol 1998;40:608–11.
[39] Forsgren L, Edvinsson SO, Blomquist HK, Heijbel J, Sidenvall R. Epilepsy in a population of mentally retarded children and adults. Epilepsy Res 1990;6:234–48.
[40] Nordin V, Gillberg C. Autism spectrum disorders in children with physical or mental disability or both. I. Clinical and epidemiological aspects. Dev Med Child Neurol 1996;38:297–313.
[41] Steffenburg U, Hagberg G, Viggedal G, Kyllerman M. Active epilepsy in mentally retarded children. I. Prevalence and additional neuro-impairments. Acta Paediatr 1995;84:1147–52.
[42] Airaksinen EM, Matilainen R, Mononen T, Mustonen K, Partanen J, Jokela V, et al. A population-based study on epilepsy in mentally retarded children. Epilepsia 2000;41:1214–20.
[43] Arvio M, Sillanpää M. Prevalence, aetiology and comorbidity of severe and profound intellectual disability in Finland. J Intellect Disabil Res 2003;47:108–12.
[44] van Schrojenstein Lantman-de Valk HM, van den Akker M, Maaskant MA, Haveman MJ, Urlings HF, Kessels AG, et al. Prevalence and incidence of health problems in people with intellectual disability. J Intellect Disabil Res 1997;41(Pt 1):42–51.
[45] van Schrojenstein Lantman-De Valk HMJ, Metsemakers JFM, Haveman MJ, Crebolder HFJM. Health problems in people with intellectual disability in general practice: a comparative study. Fam Pract 2000;17:405–7.
[46] Matthews T, Weston N, Baxter H, Felce D, Kerr M. 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. J Intellect Disabil Res 2008;52:163–73.
[47] Hove O, Havik OE. Developmental level and other factors associated with symptoms of mental disorders and problem behaviour in adults with intellectual disabilities living in the community. Soc Psychiatry Psychiatr Epidemiol 2010;45:105–13.
[48] Strømme P, Hagberg G. Aetiology in severe and mild mental retardation: a population based study of Norwegian children. Dev Med Child Neurol 2000;42:76–86.
[49] Goulden KJ, Shinnar S, Koller H, Katz M, Richardson SA. Epilepsy in children with mental retardation: a cohort study. Epilepsia 1991;32:690–7.
[50] McVicker RW, Shanks OEP, McClelland RJ. Prevalence and associated features of epilepsy in adults with Down's syndrome. Br J Psychiatry 1994;164:528–32.
[51] Memisevic H, Sinanovic O. Epilepsy in children with intellectual disability in Bosnia and Herzegovina: effects of sex, level and etiology of intellectual disability. Res Dev Disabil 2009;30:1078–83.
[52] Benassi G, Guarino M, Cammarata S, Cristoni P, Fantini MP, Ancona A, et al. An epidemiological study on severe mental retardation among schoolchildren in Bologna, Italy. Dev Med Child Neurol 1990;32:895–901.
[53] David M, Dieterich K, de Villemeur AB, Jouk PS, Counillon J, Larroque B, et al. Prevalence and characteristics of children with mild intellectual disability in a French county. J Intellect Disabil Res 2014;58:591–602.
[54] Johannsen P, Christensen JEJ, Goldstein H, Nielsen VK, Mai J. Epilepsy in Down syndrome: prevalence in three age groups. Seizure 1996;5:121–5.
[55] Haveman M, Perry J, Salvador-Carulla L, Walsh PN, Kerr M, Van Schrojenstein Lantman-de Valk H, et al. Ageing and health status in adults with intellectual disabilities: results of the European POMONA II study. J Intellect Dev Disabil 2011;36:49–60.
[56] Christianson AL, Zwane ME, Manga P, Rosen E, Venter A, Downs D, et al. Children with intellectual disability in rural South Africa: prevalence and associated disability. J Intellect Disabil Res 2002;46:179–86.
[57] Molteno G, Molteno CD, Finchilescu G, Dawes ARL. Behavioural and emotional problems in children with intellectual disability attending special schools in Cape Town, South Africa. J Intellect Disabil Res 2001;45:515–20.
[58] Lewis JN, Tonge BJ, Mowat DR, Einfeld SL, Siddons HM, Rees VW. Epilepsy and associated psychopathology in young people with intellectual disability. J Paediatr Child Health 2000;36:172–5.

[59] Wellesley DG, Hockey KA, Montgomery PD, Stanley FJ. Prevalence of intellectual handicap in Western Australia: a community study. Med J Aust 1992;156:94.
[60] Hand JE, Reid PM. Older adults with lifelong intellectual handicap in New Zealand: prevalence, disabilities and implications for regional health authorities. N Z Med J 1996;109:118–21.
[61] Lakhan R. Intelligence quotient is associated with epilepsy in children with intellectual disability in India. J Neurosci Rural Pract 2013;4:408–12.
[62] Lin J-D, Wu JL, Lee PN. Healthcare needs of people with intellectual disability in institutions in Taiwan: outpatient care utilization and implications. J Intellect Disabil Res 2003;47:169–80.
[63] Temtamy SA, Kandil MR, Demerdash AM, Hassan WA, Meguid NA, Afifi HH. An epidemiological/genetic study of mental subnormality in Assiut Governorate, Egypt. Clin Genet 1994;46:347–51.
[64] Tenenbaum A, Fuchs BS, Raskas M, Carmeli E, Aspler S, Merrick J. National survey 2009 on medical services for persons with intellectual disability in residential care in Israel. Int J Disabil Hum Dev 2012;11:75–9.
[65] Wong CW. Adults with intellectual disabilities living in Hong Kong's residential care facilities: a descriptive analysis of health and disease patterns by sex, age, and presence of Down syndrome. J Policy Pract Intellect Disabil 2011;8:231–8.
[66] Yousef JMS. Epilepsy in a sample of children with intellectual disability in Jordan. Austr N Z J Dev Disabil 1995;20:63–6.
[67] World Bank. World Bank list of economies; 2014, July, Available from: siteresources.worldbank.org/DATASTATISTICS/Resources/CLASS.XLS [accessed 01.10.14].
[68] Arshad S, Winterhalder R, Underwood L, Kelesidi K, Chaplin E, Kravariti E, et al. Epilepsy and intellectual disability: does epilepsy increase the likelihood of comorbid psychopathology? Res Dev Disabil 2011;32:353–7.
[69] Fisher RS, Acevedo C, Arzimanoglou A, Bogacz A, Cross JH, Elger CE, et al. ILAE official report: a practical clinical definition of epilepsy. Epilepsia 2014;55:475–82.
[70] Chapman M, Iddon P, Atkinson K, Brodie C, Mitchell D, Parvin G, et al. The misdiagnosis of epilepsy in people with intellectual disabilities: a systematic review. Seizure 2011;20:101–6.
[71] Marriott A, Turner S, Hatton C, Glover G, Robertson J. Making reasonable adjustments to epilepsy services for people with learning disabilities; 2014, Available from: http://www.improvinghealthandlives.org.uk/gsf.php5?f=313318&fv=20779 [accessed 13.11.14].