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Review

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

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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

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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.