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Prevalence of constipation in people with intellectual disability: a systematic review

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Abstract

Background. Constipation can lead to serious health issues and death. This systematic review summarises international research pertaining to the prevalence of constipation in people with intellectual disability.

Method. Studies published from 1990 to January 2016 were identified using Medline, Cinahl, PsycINFO, Web of Science, email requests, and cross-citations. Studies were reviewed narratively.

Results. 31 studies were identified. Constipation rates of 50% or more were reported in 14 studies; 21 studies reported rates over 33%. Based on the most representative study, over 25% of people with intellectual disability received a repeat prescription for laxatives in one year, compared to 0.1% of people without intellectual disability. Constipation was more common in those with cerebral palsy and profound intellectual disability, and associated with immobility but not age. *Conclusion.* Constipation is a significant issue for people with intellectual disability across the life course and should be actively considered as a diagnosis in this population.

Introduction

Constipation is a syndrome defined by bowel symptoms of difficult or infrequent passage of stool, hardness of stool, or a feeling of incomplete evacuation (Bharucha, Pemberton, & Locke, 2013). Constipation can be classified as primary constipation (also referred to as chronic idiopathic constipation (CIC) or functional constipation) and secondary constipation (attributed to comorbid medical conditions or medications) (Sbahi & Cash, 2015).

The concept of constipation is complicated by disagreement among patients and doctors about its nature (Longstreth et al., 2006). Whilst physicians often regard constipation to be synonymous with infrequent bowel movements (usually less than three times a week), patients may have a broader set of symptoms as while bowel movement infrequency can be distressing to patients, it is the quality of, or difficulty associated with, defecation that is the primary determinant of patient-described constipation (Sbahi & Cash, 2015). As such, the Rome III diagnostic criteria for functional constipation (Longstreth et al., 2006) incorporates symptoms such as 'sensation of incomplete evacuation for at least 25% of defecations', and 'straining during at least 25% of defecations'.

The complications of chronic constipation can be serious and life-threatening, including: faecal incontinence (where overflow incontinence may confuse the diagnosis of chronic constipation); haemorrhoids; anal fissure; pelvic organ prolapse; faecal impaction and bowel obstruction necessitating surgery; and bowel perforation and stercoral peritonitis where extremely impacted faeces can compress the colonic wall, causing an ischemic ulcer and subsequent perforation, culminating in stercoral peritonitis and sometimes death (Leung, Riutta, Kotecha, & Rosser, 2011). Further, the impairment in health related quality of life (HRQoL) observed in adults with constipation is comparable with that seen in conditions that

might be regarded as more 'serious', such as osteoarthritis, rheumatoid arthritis, chronic allergies and diabetes (Belsey, Greenfield, Candy, & Geraint, 2010). In children, the level of impairment seen is greater than with gastro-oesophageal reflux and inflammatory bowel disease (Belsey et al., 2010). Early detection and management are crucial (De Hert et al., 2011).

There are a number of reasons underlying a need to focus on constipation in people with intellectual disability. The usual trigger for doctors to consider constipation is the specific mention of constipation by the patient or the communicated history of abdominal symptoms, but patients with intellectual disability may be unable to communicate these (Coleman & Spurling, 2010). In people with intellectual disability, pain from constipation may present as distress, sleep disturbance or behavioural changes (Coleman & Spurling, 2010), and may be associated with behavioural problems such as aggression and self-injury (Bosch, Van Dyke, Smith, & Poulton, 1997; Carr & Smith, 1995; Christensen et al., 2009). As a result, constipation may be missed.

Constipation can have serious consequences for people with intellectual disability if it is not identified and managed appropriately. Constipation constitutes an ambulatory care sensitive condition (ACSC) for people with intellectual disability (R. S. Balogh, Ouellette-Kuntz, Brownell, & Colantonio, 2011). A Canadian study on hospitalisation rates for ACSCs found that the hospitalisation rate for constipation for people with intellectual disability was 7.9 times higher (95% CI 4.4, 14.2) than for people without an intellectual disability (R. Balogh, Brownell, Ouellette-Kuntz, & Colantonio, 2010). In England, constipation was found to be one of the common causes of emergency hospital admissions for ACSCs (G. Glover & Evison, 2013). Constipation can also lead to death. The Safeguarding Adults Board in

Suffolk, England, commissioned two Serious Case Reviews in early 2014 into the deaths of two people with intellectual disability (Flynn & Eley, 2015b, 2015c), both of whom died from complications arising from constipation (Flynn & Eley, 2015a).

Several factors put people with intellectual disability at increased risk of constipation. Many medicines are constipating (National Institute for Health and Care Excellence, 2015) and people with intellectual disability are more likely to be prescribed some of these. For example, constipation is a common side effect of different antipsychotics (De Hert et al., 2011) and people with intellectual disability are more likely than others to be prescribed anti-psychotic medications (G Glover et al., 2015). Progression from constipation to ileus, intestinal obstruction, bowel ischaemia, megacolon and death is not uncommon, particularly in patients (not necessarily with intellectual disability) prescribed clozapine (Every-Palmer, Newton Howes, & Clarke Mike, 2014). People with intellectual disability are also more likely to have poor diet (Humphries, Traci, & Seekins, 2009), physical mobility limitations (Cleaver, Hunter, & Ouellette-Kuntz, 2009), and low levels of physical activity (E Emerson, 2005; Robertson et al., 2000), all factors associated with constipation (Mugie, Benninga, & Di Lorenzo, 2011). People with Down syndrome are more likely to have hypothyroidism (Goday-Arno et al., 2009) which is associated with constipation (National Institute for Health and Care Excellence, 2015). In addition, Down syndrome is associated with Hirschsprung's disease (Friedmacher & Puri, 2013) which is the most common congenital gut motility disorder, characterized by the absence of the enteric ganglion cells (aganglionosis) along the distal gut, which causes functional intestinal obstruction (Best et al., 2014).

In the general population, estimates of the prevalence of constipation vary. A systematic review and meta-analysis of the global prevalence of chronic idiopathic constipation (CIC) in

adults in the community found a pooled prevalence of 14% (95% CI 12%, 17%) (Suares & Ford, 2011). A further systematic review of 68 worldwide studies found prevalence ranged from 2.5% to 79% in adults (the latter figure relating to those based in geriatric long term care) and from 0.7% to 29.6% in children (Mugie et al., 2011). The mean value of constipation rates ascertained in Europe has been reported as 17.1 % (Peppas, Alexiou, Mourtzoukou, & Falagas, 2008). Variation in reported prevalence is partly due to differences in the definition for constipation used, variable age groups, and the methods used to collect the information. Female gender, increasing age, lower socioeconomic status and lower educational level are associated with higher constipation prevalence rates (Mugie et al., 2011).

No reviews have considered the prevalence of constipation in people with intellectual disability. Accurate estimates of constipation prevalence are important, allowing services and policy makers to be better informed when planning associated health and social care resources and raising awareness of this health issue in people with intellectual disability to prevent it being overlooked. The objective of this paper is to present the first systematic review of international research pertaining to the prevalence of constipation in people with intellectual disability. Studies relating to Hirschsprung's disease and Down syndrome are not included as a meta-analysis of 61 studies relating to incidence, outcomes and mortality already exists (Friedmacher & Puri, 2013).

Method

The review was conducted in accordance with PRISMA guidelines (Moher, Liberati, Tetzlaff, & Altman, 2009). Electronic literature database searches were conducted in Medline, Cinahl and PsycINFO (all on EBSCO) and Web of Science (SCI-EXPANDED,

SSCI and A&HCI) in January 2016. Searches combined terms for constipation and intellectual disability with the Boolean operator 'and'. An example of database specific search terms (Medline) is given in Appendix One. Searches included broad terms relating to constipation and people with intellectual disability to create a pool of studies, with studies on topics other than prevalence being retained for separate review. Specific criteria relating to prevalence studies were applied to this pool of studies as below. The reference lists of studies meeting the inclusion criteria were searched. In addition, in December 2015 a request for information on research relevant to the review was sent to members 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.

Inclusion Criteria

Articles were required to meet all the following criteria:

- Peer reviewed
- English language full text
- Published from 1990 to early 2016
- Quantitative research, evaluation or audit
- Samples where 50% or more have intellectual disability or mixed samples where results are disaggregated for people with intellectual disability
- Includes data regarding the prevalence of constipation, including laxative use as an indicator of constipation
- Presents exact figures on the outcome of interest

Exclusion Criteria

- Not peer reviewed or peer review status unclear

- Any study employing any research design with a sample size of 10 or less
- Reviews, letters, commentaries, editorials, meeting or conference abstracts
- Studies based on neonates (new born infants up to 28 days after birth)
- Studies on conditions where intellectual disability cannot be assumed (e.g. cerebral palsy) where results not disaggregated for people with intellectual disability
- Studies on specific syndromes associated with intellectual disability with the exception of Down syndrome which is the most common genetic cause of intellectual disability (Sherman, Allen, Bean, & Freeman, 2007). Less common specific syndromes such as Rett syndrome were excluded although it is evident that research on such syndromes exists (e.g. Baikie et al., 2014; Schwartzman, Vítolo, Schwartzman, & Morais, 2008)
- Studies not presenting exact figures (e.g. reporting that one third had constipation)
- Studies relating to encopresis (soiling)
- Studies relating to Hirschsprung's disease and Down syndrome, as a meta-analysis of 61 studies relating to incidence, outcomes and mortality already exists (Friedmacher & Puri, 2013).

Initially, titles and abstracts were used to exclude studies which were obviously not within scope (1st 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 the first and second author and discussed until consensus was reached on whether or not they met the inclusion criteria with regards to prevalence. All relevant studies were included in the review regardless of methodological quality. Study data was extracted from full text articles and entered into an excel database by the first author with regard to: authors, year, country, main focus of study, study design,

sample source, key sample features, sample size, sample age range (mean, SD and median), percentage of sample male, percentage of sample with specific levels of intellectual disability, definition of constipation used, method for ascertaining constipation, numerator and denominator for estimate of percentage with constipation, and other key results. 95% confidence intervals for prevalence rates were calculated using the Wilson Score Method (Eayres, 2008) in Microsoft Excel using the spreadsheet available at http://www.apho.org.uk/resource/view.aspx?RID=48617. All extracted data was subsequently checked for accuracy and completeness by a second reviewer. Whilst a third reviewer was available to resolve any disagreements, no instances of disagreement arose.

Risk of Bias

Risk of bias was assessed using an adaptation of a 9 item critical appraisal tool developed specifically for use in systematic reviews of prevalence data (Z. Munn, Moola, Lisy, Riitano, & Tufanaru, 2015). As this review includes studies relating to sub-populations of people with intellectual disability (such as people with profound intellectual disability, or people with Down syndrome), options for item 1 *'Was the sample frame appropriate to address the target population?'* were expanded from Yes (1)/No (0) to: population based sample of people with intellectual disability (score 4, allow some deviation e.g. can score 4 if sample frame only includes those in contact with services, or with GPs); sample of people with intellectual disability covering a range of levels of intellectual disability and etiologies but with limited generalisability (e.g. certain types of accommodation, restricted ages) (score 3); representative sample of people with selected characteristics (e.g. profound intellectual disability, Down syndrome) (score 2); sample of people with selected characteristics with limited generalisability (e.g. sample chosen from only one type of setting) (score 1); poorly defined sample (score 0).

Options for item 7 '*Was the condition measured in a standard, reliable way for all participants?'* were expanded from Yes (1)/No (0) to provide more detail in relation to identifying constipation: clinical examination or prospective recording of bowel movements (score 4); extracted from records or databases (score 3); interview with informant (score 2); questionnaire self-completion by informant (score 1); unclear or not stated (score 0).

For item 3 '*Was the sample size adequate?'*, using the formula proposed in relation to the checklist (Zachary Munn, Moola, Riitano, & Lisy, 2014), and adopting 50% as the estimated rate, a sample size of 385 was judged as adequate (Naing, Winn, & Rusli, 2006).

No studies were excluded based on risk of bias scores. In Table 1 individual scores are given for items 1, 6 ('*Were valid methods used for the identification of the condition?*' scored as Yes (1)/No (0)) and 7 as these were considered to be key scores in relation to obtaining unbiased estimates of the prevalence of constipation in people with intellectual disability. Total score is also presented (possible range 0-15).

Results

The process of identifying studies for inclusion is summarised in Figure 1. Searches identified 1,757 articles, with 1,169 remaining follow deletion of 588 duplicates. 1,070 articles were excluded based on the title/abstract, leaving a pool of 99 articles for further screening. After examination of full text and the addition of articles cited within these and from other sources, 32 articles met the criteria for inclusion with regards to prevalence, two of which were based on data from the same study (Haveman et al., 2011; Martínez-Leal et al., 2011) giving a total of 31 studies. Studies are summarised in Table 1.

Figure 1 Here

Geographical spread

Most studies were from high income countries. Studies were identified from the following countries: United States (8), Netherlands (7), Australia (3), United Kingdom (3), Belgium (2), Belgium and the Netherlands jointly (1), Italy (2), Brazil (1), India (1), Ireland (1), Israel (1), and one study was based on a sample from 14 European countries.

Table 1 Here

Risk of Bias

Scores indicate quality in relation to obtaining estimates of the prevalence of constipation in the population of people with intellectual disability. As this was not the primary aim of most of the studies, scores are often low even though the quality of the study in relation to its primary aim may have been very high.

Item 1. Sample frame. Only one study employed a sample frame (general practices in the Netherlands) broadly representative of people with intellectual disability (Straetmans, van Schrojenstein Lantman-de Valk, Schellevis, & Dinant, 2007). A further six studies employed a sample broadly representative of people with intellectual disability from a particular sub-population: Down syndrome (Alexander et al., 2015; Yin, Boyd, Pacheco, Schonfeld, & Bove, 2012), children with Down syndrome (Leonard, Bower, Petterson, & Leonard, 1999; Thomas et al., 2011), children with severe generalized cerebral palsy (Veugelers et al., 2010), and adults with ASD (Jones et al., 2015).

Items 2, 5 & 9. Many studies were based on small convenience samples where the entire sampling frame was included. As such, these items regarding sampling method, response rate and coverage bias tended to be redundant in this context.

Item 3. Seven studies employed a sample size of 385 or above; 14 studies had a sample size of less than 100.

Item 4. 11 studies did not describe study subjects in sufficient detail, with level of intellectual disability not being specified.

Item 6. In over half (16) of the studies there is a high risk of bias due to the definition of constipation employed (e.g. only including laxative use, not specifying what is meant by constipation).

Item 7. Only 6 studies ascertained constipation via clinical examination or prospective recording of bowel movements.

Item 8. Only one study reported the 95% confidence interval for the percentage reported to have constipation (Veugelers et al., 2010).

Overall, all studies were at some risk of bias in relation to one or more aspects of selected key items 1, 6 and 7. The results regarding prevalence rates below highlight studies with a lower risk of bias.

Prevalence Rates

The rates presented for constipation in Table 1 range from 96.4% of people with profound intellectual disability living in an institution (Kozma & Mason, 2003) to 4.5% of people on a Down syndrome register with constipation of sufficient severity to warrant investigation for Hirschsprung's disease (HD) (Yin et al., 2012). The wide variation in the figures can be

attributed to differences in the characteristics of the samples included, the definition of constipation used, and the method of ascertainment which has been found to be related to variation in prevalence estimates (Peppas et al., 2008; Suares & Ford, 2011). Due to this variation, meta-analysis was inappropriate and prevalence rates are reported narratively below and illustrated visually in Figure 2.

Figure 2 here

General population of people with intellectual disability. In relation to the general population of people with intellectual disability, based on the most representative sample available, 5.2% were diagnosed with constipation during GP contact in one year, 14.3% were prescribed laxatives, and 25.7% received a repeat prescription for laxatives (257 per 1000, compared to 1 per 1000 for controls) (Straetmans et al., 2007). Other studies including a range of levels of intellectual disability and etiologies with a relatively low risk of bias report figures of: 69.3% of 215 people with intellectual disability living in institutions found via prospective recording of bowel movements to have less than 3 bowel movements a week or to need laxatives over 3 times a week (Böhmer, Taminiau, Klinkenberg-Knol, & Meuwissen, 2001); 57.1% of 70 adults aged 60 or more being identified with constipation over a 10 year period (Evenhuis, 1997); and 36.2% of 58 people with intellectual disability aged 16 or more without scoliosis or swallowing disorder found to have less than 3 defecations a week based on a prospective diary (Vande Velde, Van Biervliet, Van Goethem, De Looze, & Van Winckel, 2010).

Down syndrome. In relation to people with Down syndrome, estimates range from (as noted above) 4.5% of people on a Down syndrome register with constipation of sufficient severity to warrant investigation for HD (Yin et al., 2012) to 50.0% of 84 adults and children with Down syndrome attending one clinic being identified as having functional constipation

(Rome III criteria) based on interviews with the person/parent using a validated questionnaire (de Carvalho Mrad et al., 2014).

For people with Down syndrome, the most representative sample, consisting of nearly 3,000 adults and children with Down syndrome in the UK, found that the one year prevalence of laxative prescription was 18.8% (Alexander et al., 2015). This compared to 3.4% of 8,910 matched controls, giving a yearly prevalence rate ratio of 5.5 (95% CI 4.8, 6.4). Further studies with a relatively low risk of bias report figures of: 4.5% of people on a Down syndrome register with constipation of sufficient severity to warrant investigation for HD (Yin et al., 2012); and based on a retrospective chart review of 105 children and adolescents with Down syndrome attending the inaugural year of a newly established Down syndrome clinic, 16 had a pre-existing diagnosis of constipation, and a new constipation diagnosis as a result of the clinical visit was made in 20 (total with constipation 34.3%) (Skotko, Davidson, & Weintraub, 2013).

People with profound intellectual disability. Four studies present figures for constipation in people with profound intellectual disability. The study with the lowest risk of bias and largest sample was a retrospective analysis of the pharmacy and medical records of 254 people with profound intellectual and multiple disabilities which identified 59.8% as having constipation as a registered health problem and 65.0% having been prescribed laxatives in the previous year (van der Heide, van der Putten, van den Berg, Taxis, & Vlaskamp, 2009). Whilst the other three studies are based on small samples, rates are consistently extremely high: 73.1% of 26 women with profound intellectual disability reported to be receiving medication to stimulate bowel movements (Giesbers et al., 2012); 96.4% of 55 non-ambulatory, institutionalized adults with profound intellectual disability identified as having constipation via review of individual habilitation plans (Kozma & Mason, 2003); and 44% of

49 adults and children with profound multiple disability having constipation reported as a medical condition (Petry, Maes, & Vlaskamp, 2009).

Cerebral palsy. One study with a relatively low risk of bias and including use of a two week diary found that in a representative sample of 152 children with severe generalized cerebral palsy, 57.2% had constipation (Veugelers et al., 2010). A further study found that 74.1% of 58 children with cerebral palsy referred to a paediatric neurology outpatient clinic had three or fewer bowel movements a week (Del Giudice et al., 1999).

Other groups. Other reported constipation figures were: 59.1% of 93 adults with intellectual disability and dysphagia (Chadwick & Jolliffe, 2009); 59.6% of 198 people with intellectual disability admitted to an inpatient psychiatric unit (Charlot et al., 2011); 55.7% of 122 paediatric outpatients with intellectual disability in India (Jauhari, Bhargava, Bhave, Kumar, & Kumar, 2012); 61.7% of 47 children with severe brain damage and intellectual disability (Staiano & Del Giudice, 1994); lifetime prevalence of 38.0% in 50 ambulatory children with developmental disability (60% of whom had intellectual disability) and no genetic disorder (Valicenti-McDermott et al., 2006); and for adults with autism (73% of whom had intellectual disability) 35.9% were reported to have ever had constipation (Jones et al., 2015).

Presentation issues

In one study it is noted that whilst hard or infrequent stools were brought to the attention of caregivers by those with mild intellectual disability, this was not the case for those with moderate or severe intellectual disability (Evenhuis, 1997). One study notes that reported symptoms included abdominal cramps, bloating, flatus, and concomitant behavioural problems, with constipation being newly diagnosed in 19 (33.9%) of 56 attending a clinic where parents had an initial concern of behavioural issues (Skotko et al., 2013). Finally, of

80 patients with a diagnosis of mental disorder due to a medical disorder which provoked admission to an acute care specialised inpatient psychiatric unit, of 72 people where the medical cause was listed the most commonly reported was constipation, which affected 15 patients (Charlot et al., 2011).

Factors associated with constipation

A small number of studies present univariate associations with constipation. One study found non-ambulancy, cerebral palsy, the use of anticonvulsant therapy, the use of benzodiazepines, the use of H_2-receptor antagonists or proton pump inhibitors, and an IQ < 35 (severe to profound intellectual disability) to be significantly more frequent in patients with constipation compared to those without (Böhmer et al., 2001). A further study found constipation to be more common in those who were non-ambulatory (73.2%) and those with spastic quadriplegia (76.3%) (Chadwick & Jolliffe, 2009). One study found that constipation was strongly associated with lower urinary tract symptoms, presenting in 95.7% (22/23) of individuals with lower urinary tract symptoms and 32.8% (20/61) without (OR 45.1; 95% CI 5.6, 301) (de Carvalho Mrad et al., 2014). One study found constipation to be related to mobility impairment, with a rate of 87.5% in those who were non-ambulant compared to 50% who were ambulant, and a rate of 62.5% of those who routinely used psychotropic or anti-epileptic drugs (Evenhuis, 1997). In paediatric outpatients in India, constipation was identified in 49/68 (72.1%) of those with cerebral palsy, and 19/54 (35.2%) without (OR 4.8 (95% CI 2.2, 10.2)) (Jauhari et al., 2012). Age was not been found to be associated with constipation in some studies (Böhmer et al., 2001; Haveman et al., 2011; Morad, Nelson, Merrick, Davidson, & Carmeli, 2007).

Only two studies conducted a multivariate analysis of risk factors for constipation. One study found that immobility (OR 2.4; 95% CI 1.3, 4.5), no exercise (OR 1.5; 95% CI 1.1, 2.1) and

any neurological disease (OR 1.9; 95% CI 1.4, 2.7) were significantly associated with constipation (Morad et al., 2007). Gender and age were not significant. The second study found that female gender (OR 2.0; 95% CI 1.1, 3.4), non-ambulancy (OR 3.4; 95% CI 1.6, 7.4), pureed or tube feeding (OR 2.7; 95% CI 1.4, 5.2) and use of medication (non-constipating OR 3.9 (95% CI 1.8, 8.3); constipating OR 4.0 (95% CI 1.7, 9.2)) were associated with laxative use (Van Winckel, Vander Stichele, De Bacquer, & Bogaert, 1999).

Characteristics of constipation

A small number of studies note the absence of faecal soiling in people with intellectual disability and constipation. Faecal soiling was found in 14.9% of institutionalised people with intellectual disability with constipation (Böhmer et al., 2001), no children with cerebral palsy (Del Giudice et al., 1999), and brain damaged children were noted to have constipation without faecal soiling (Staiano & Del Giudice, 1994).

A number of studies have assessed colonic transit times. Evaluation of colonic segmental transit times for 25 children with cerebral palsy and constipation showed a delay at the level of the more proximal segments of the colon in 13 of 25 (52%) patients, at the level of the left colon-rectum in 9 (36%) and at the level of the rectum only in 3 (12%), suggesting that constipation is mainly due to prolonged transit at the level of the more proximal segments of the colon (Del Giudice et al., 1999). One study found that whilst colonic transit times indicated that most children with typical mental development and functional faecal retention had delayed transit only in the rectum (80%) (with soiling typically present), the majority with brain damage (56%) had delayed transit in both the left colon and rectum, without soiling and at rectum only in 25% (Staiano & Del Giudice, 1994). One study found that colonic transit time was prolonged in patients with intellectual disability compared to healthy controls, irrespective of the presence of constipation (Vande Velde et al., 2010). In patients

with intellectual disability with a low defecation frequency, transit was slow in all colon segments.

Discussion

Estimated prevalence rates of constipation in people with intellectual disability vary depending on the sample and methods used, with variation in methods for the definition and ascertainment of constipation making comparison between studies difficult. However, it is clear that prevalence is high, with rates of 50% or more being reported in 14 of the 31 studies, and 21 studies reporting rates of over a third. The study with the most representative sample of the population of people with intellectual disability, suggests that over 1 in 4 people with intellectual disability received a repeat prescription for laxatives in one year, compared to 1 in 1,000 people without intellectual disability (Straetmans et al., 2007). In the most representative sample of people with Down syndrome, one year prevalence of laxative prescription was 18.8% compared to 3.4% of matched controls (Alexander et al., 2015). However, laxative prescription may underestimate actual constipation prevalence as it is clear that constipation can be missed (Coleman & Spurling, 2010) and not all of those with constipation will be prescribed laxatives. The high prevalence rates reported support the suggestion that doctors should actively consider and exclude the diagnosis of constipation in people with intellectual disability (Coleman & Spurling, 2010) and awareness of constipation should be raised among caregivers.

Reported constipation rates are higher for those with profound intellectual disability and those with cerebral palsy. Non-ambulancy has also consistently been reported to be associated with constipation (Böhmer et al., 2001; Chadwick & Jolliffe, 2009; Evenhuis, 1997; Morad et al., 2007; Van Winckel et al., 1999). Age has not consistently been found to be associated with constipation (Böhmer et al., 2001; Haveman et al., 2011; Morad et al.,

2007) in contrast to the general population (Mugie et al., 2011; Suares & Ford, 2011) suggesting that for people with intellectual disability, constipation is a significant issue across the life course. However, there is limited information available regarding factors associated with constipation in this population. Additional large scale studies employing multivariate analytic methods would help clarify the factors associated with constipation in people with intellectual disability. Future research should also further evaluate the pathogenic mechanisms of constipation in people with intellectual disability as chronic constipation should be managed according to its etiology (Sbahi & Cash, 2015).

Böhmer et al (2001) note that until research provides clear answers as to how to manage constipation in people with intellectual disabilities, the basic principles for treatment are the same for individuals with intellectual disabilities as for the general population. As a basic principle, it has been suggested that bowel management should begin with the "eight keys to bowel success" prior to developing an individualised bowel programme: physical exercise, high fibre intake, high fluid intake, consistent habit time, an upright position on toilet or commode, privacy, medication management, and patient and family education (Weeks, Hubbartt, & Michaels, 2000).

Appropriate toilet training appears necessary for normal defecation and improper training has been implicated as a cause of constipation in children (Palit, Lunniss, & Scott, 2012). Toilet sitting in a correct position at the toilet facilitates defecation by opening the anorectal angle at its maximal width (Altomare et al., 2001; Sakakibara et al., 2010). Establishing a consistent habit time that coincides with when people are most likely to evacuate their bowels may reduce the possibility of people suppressing the urge to defecate. Voluntary suppression of defecation has been found to lead to a decrease in stool frequency, stool volume, and increases in total transit times, and rectosigmoid and right hemicolon transit times, suggesting

that constipation can be ‘‘learned’’ (Klauser, Voderholzer, Heinrich, Schindlbeck, & Muller-Lissner, 1990).

It has been suggested that children with intellectual disabilities should begin toileting training when they are ready physiologically, rather than waiting until they are “ready” based on a standard toilet readiness checklist (Rogers, 2002). However, some people with intellectual disabilities may not receive appropriate toilet training. Whilst incontinence should not be attributed to intellectual disability without ruling out other causes (Nair, Sagayaraj, Rajan, & Kumar, 2015), it tends to be viewed as an integral part of having a severe intellectual disability (Stenson & Danaher, 2005). As such, children with intellectual disabilities may not undergo a comprehensive bladder and bowel assessment but may instead be given a simple “pad assessment” and be issued with nappies in the mistaken belief that they are not ready to be toilet trained (Rogers, 2002; Rogers & Patricolo, 2014). Future research could consider the relationship between appropriate toilet training and constipation in people with intellectual disabilities. Research could also consider the extent to which the “eight keys to bowel success” are adhered to by those supporting people with intellectual disabilities. Ultimately, research needs to address the management of constipation in people with intellectual disabilities. This will be the focus of a future review where we will consider issues such as medication (e.g., Migeon-Duballet et al., 2006), abdominal massage (e.g., Connor, Hunt, Lindley, & Adams, 2014) and the knowledge of those supporting people with intellectual disabilities with regards to constipation (e.g., Marsh & Sweeney, 2008).

Limitations

There are a number of limitations to this review. First, ascertaining the prevalence of constipation was not the primary aim of the majority of the included studies and as such the risk of bias inherent in the definitions and procedures employed for identifying constipation was high. Second, whilst studies were identified from a range of countries, the review is

restricted to English language publications and this may have contributed to the fact that only one study was identified from low and middle income (LAMI) countries. Third, all data was extracted by one reviewer and checked for accuracy and completeness by a second reviewer. Whilst this is an accepted minimum (Centre for Reviews and Dissemination, 2009), extraction of data by two reviewers independently would have reduced the possibility of errors. Fourth, studies relating to specific syndromes were excluded (e.g. Baikie et al., 2014; Kuhlmann, Joensson, Froekjaer, Krogh, & Farholt, 2014; Schwartzman et al., 2008) and should be the focus of future review. Finally, no studies have included the 'hidden majority' of adults with intellectual disability who are not known to intellectual disability services (E. Emerson, 2011).

Conclusion

Constipation is common in people with intellectual disability across the life course and may be under-recognised. There is a need to discern more accurately the prevalence of, and risk factors for, constipation in people with intellectual disability. Improved recognition and management of constipation may reduce the occurrence of associated health conditions, reduce hospital admissions, and improve quality of life. A recent report provides information on reasonable adjustments that can be made for managing constipation in people with intellectual disability (Marriott & Emly, 2016). The ideas, information and examples of good practice in relation to managing constipation provided within this report should help services improve provision for this highly prevalent condition and potentially reduce ill health and deaths associated with constipation in people with intellectual disability.

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Appendix 1: Example of database specific search strategy (Medline)

Medline searches run on 26.1.16. Total 794 citations identified.

Limiters: Jan 1990 – 2016; English language.

Search terms:

(((MH "Constipation") OR (MH "laxatives") OR (MH "fecal impaction") OR (TI constipat* OR TI fecal OR TI faecal OR TI laxative* OR TI defecat* OR TI defaecat* OR TI bowel OR TI colon* OR AB constipat* OR AB fecal OR AB faecal OR AB laxative* OR AB defecat* OR AB defaecat* OR AB bowel OR AB colon*))) AND (((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") 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 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")))

Figure 1

Flowchart of Study Identification

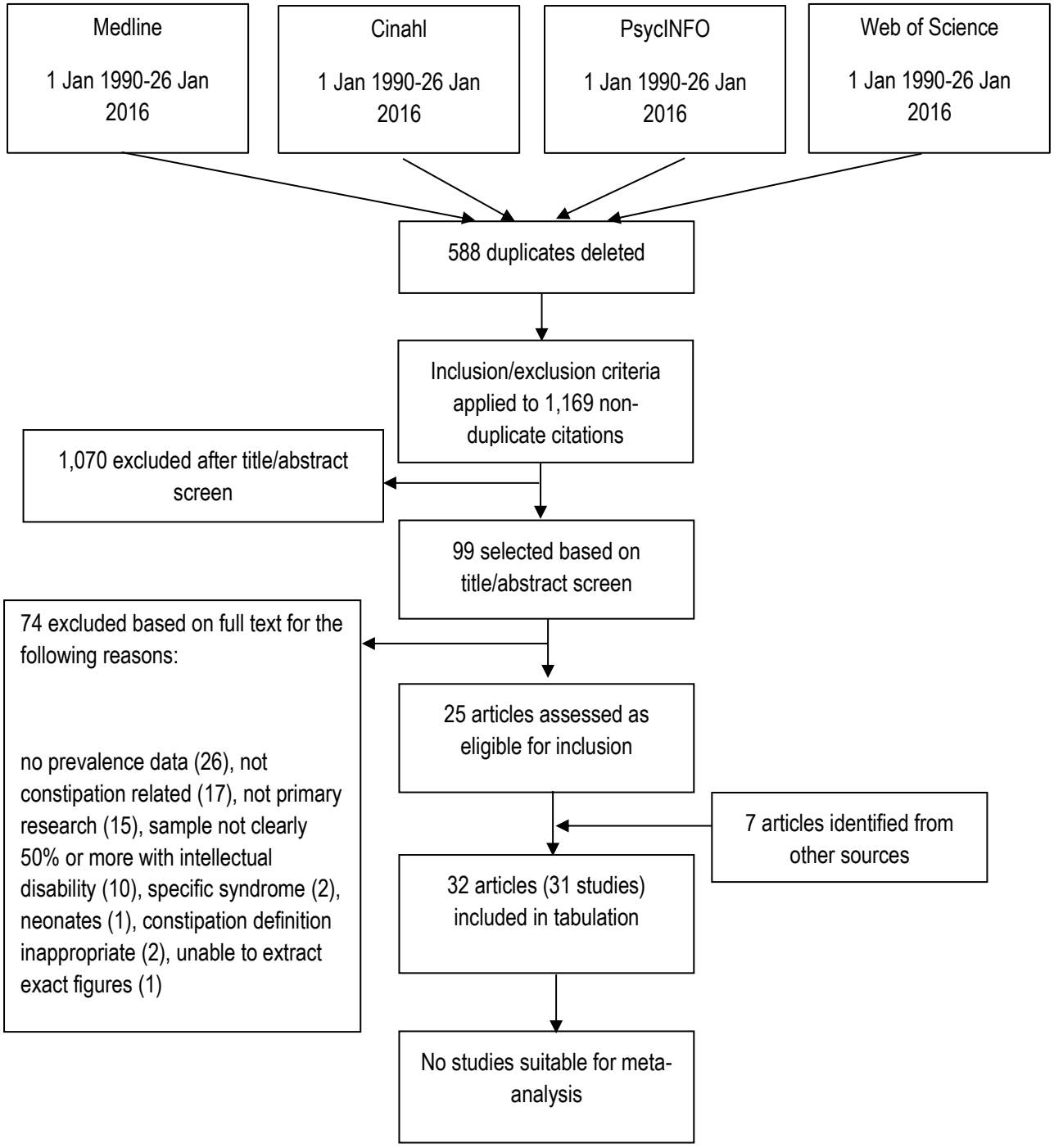


Table 1: Summary of studies giving constipation rates for people with intellectual disability

1st Author & Year (1/6/7/total)*	Country	Key sample features	Age range (mean (SD); Mdn)	% male	Border-line % (IQ > 70)	Mild % (50 or 55 - 70)	Moderate % (IQ 35-50/54)	Severe % (IQ 20-34)	Profound % IQ <20	Not specified	Definition of constipation (or other definition)	Cases	N	%
Alexander 2015 (2/0/3/9)	UK	Adults & children with DS on CPRD	ns 30.4% < 18; 49.5% < 30	53.8	ns	ns	ns	ns	ns	100	Prescribed laxatives in 2013	551	2926	18.8
Böhmer 2001 (3/1/4/12)	Netherlands	Institutionalized people with ID, IQ < 50	6-80 (constipated 31.8 (15.8); ns; not constipated 34.1 (18.6); ns)	60.0	0	0	17.7	82.3	←	0	Bowel movement < 3 week or need laxatives > 3 week	149	215	69.3
Chadwick 2009 (1/0/3/8)	England	Adults with ID & dysphagia	18-74 (40.7 (13.96); ns[b]	43.6[b]	→	7.1	24.2	38.4	30.3	0	Abnormally delayed or infrequent passage of dry hardened faeces & taking medication for this	55	93	59.1
Chaidez 2014 (3/0/1/4)	US	Children ID or DD without autism. Score for MSEL *or* VABS of < 70 & < 77 on both	24-60 months (ns (ns); ns)	63.5	0	ns	ns	ns	ns	100	Constipation (ns) frequently or always in past 3 months	21.7[a]	137	15.8
Charlot 2011 (1/1/3/9)	US	People with ID admitted to a specialised inpatient psychiatric unit	ns (39 (10.06); ns)	56.0	0	46.0	40.0	13.0	0	0	Medical diagnosis at time of discharge	118	198	59.6
de Carvalho Mrad 2014 (1/1/2/7)	Brazil	People with DS[d]	4-30 (ns (5); 16)	33.3	ns	ns	ns	ns	ns	100	Rome III criteria for functional constipation	42	84	50.0
Del Giudice 1999 (1/1/4/10)	Italy	Children with CP referred to paediatric neurology outpatient clinic	6mths-12yrs (5.2 (4.9); ns)	43.1	0	→	10.0	90.0	←	0	Reduced bowel frequency with 3 or less bowel movements per week	43	58	74.1
Evenhuis 1997 (3/1/4/12)	Netherlands	Adults aged 60+	60-92 (70.1 (ns); ns)	37.1	0	38.6	51.4	10.0	0	0	10 year prevalence of chronic constipation (ns)	40	70	57.1

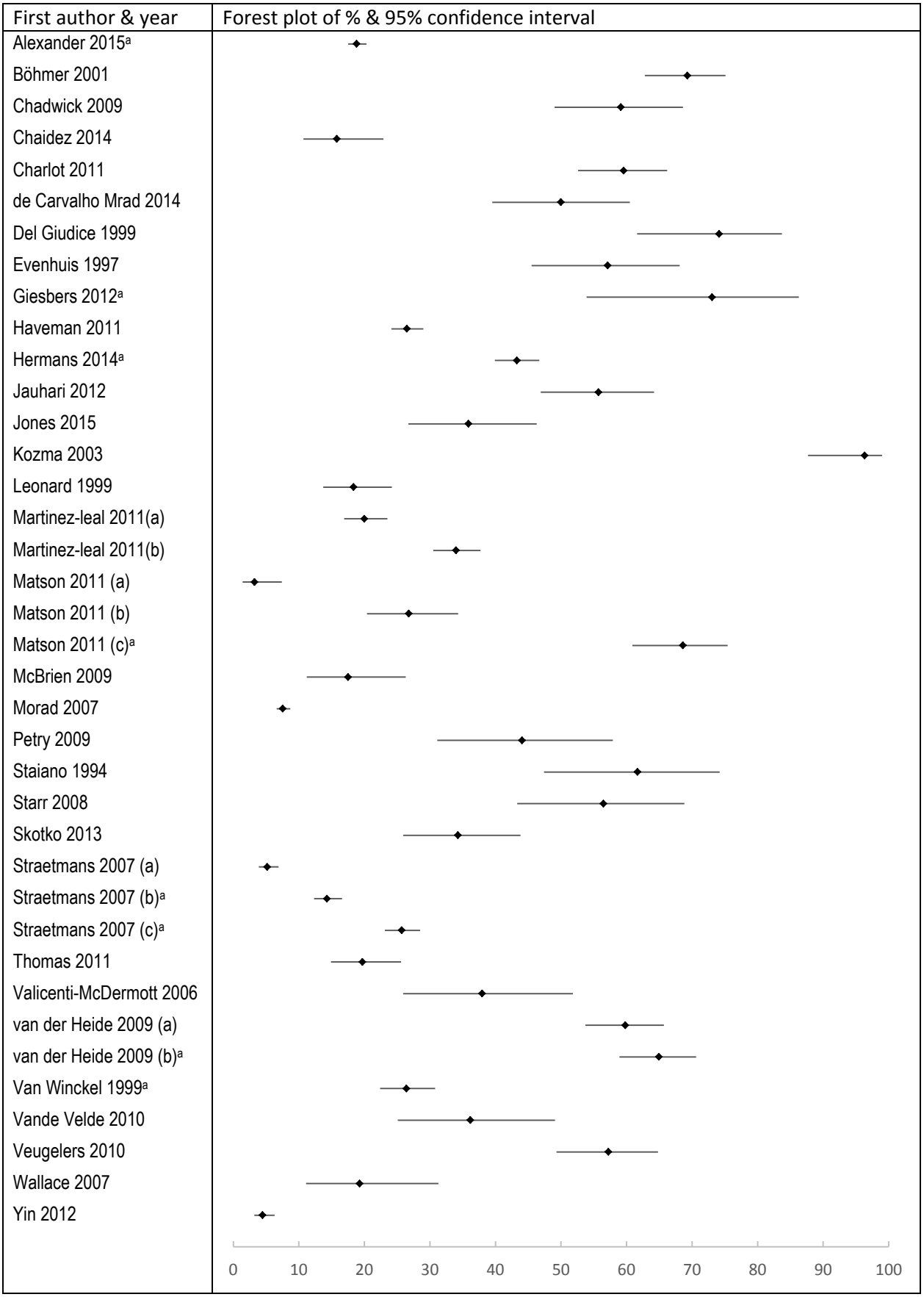
1st Author & Year (1/6/7/total)*	Country	Key sample features	Age range (mean (SD); Mdn)	% male	Border-line % (IQ > 70)	Mild % (50 or 55 - 70)	Moderate % (IQ 35-50/54)	Severe % (IQ 20-34)	Profound % IQ <20	Not specified	Definition of constipation (or other definition)	Cases	N	%
Giesbers 2012 (1/0/1/5)	Netherlands	Control group of females with profound ID	5.3–47.3 (21.6 (11.5); ns)	0%	0	0	0	0	100	0	Receiving medication to stimulate bowel movements	19	26	73.1
Haveman 2011 (3/0/2/9)	14 European countries	Adults with ID in Europe, 20.3% with DS	19-90 (41 (ns); ns)	50.6	0	22.7	28.2	20.7	11.8	16.6	Constipation (ns) in the last 12 months	332[a]	1253	26.5
Martinez-leal 2011(a)	As above	As above, unstaffed homes	ns (38.1 (13.7); ns)	53.3	0	27.9	28.8	13.9	8.2	21.2	As above	111	555[a]	20.0
Martinez-leal 2011 (b)	As above	As above, staffed homes	ns (44.3 (15.0); ns)	48.1	0	16.5	25.3	24.8	14.1	19.3	As above	223	656[a]	34.0
Hermans 2014 (3/0/3/10)	Netherlands	Aged 50+ known to ID services	50-ns (60.9 (8.1); ns)	51.3	→	24.3	48.2	16.2	8.7	2.6	Use of laxative medication	349	806	43.3
Jauhari 2012 (1/1/1/6)	India	Paediatric outpatients assessed as having ID using validated tests	6-159 months (ns (ns); 30)	68.8	0	31.1	23.8	45.1	←	0	Scybalous, pebble-like, hard stools in > 25% defecations & defecation < 3 per week during a 2-week period; or large stools palpable on abdominal examination; or laxative use or manual disimpaction of faeces	68	122	55.7
Jones 2015 (2/0/2/8)	US	Adults with autism, 73% with ID	23.5–50.5 (ns (ns); 36.1)	75.0	27.0[e]	24.0	38.0	←	←	11.0	Ever had constipation (ns)	32	89.1[a]	35.9
Kozma 2003 (1/0/3/8)	US	Non-ambulatory, institutionalized adults with profound ID	28-63 (ns (ns); ns)	47.3	0	0	0	0	100	0	Constipation ns	53	55	96.4
Leonard 1999 (see Thomas 2011) (2/0/1/6)	Australia	School age children with DS	5-17 (ns (ns); ns)	57.3	ns	ns	ns	ns	ns	100	Ever had bowel problem: constipation (ns)	38	207	18.4

1st Author & Year (1/6/7/total)*	Country	Key sample features	Age range (mean (SD); Mdn)	% male	Border-line % (IQ > 70)	Mild % (50 or 55 - 70)	Moderate % (IQ 35-50/54)	Severe % (IQ 20-34)	Profound % IQ <20	Not specified	Definition of constipation (or other definition)	Cases	N	%
Matson 2011 (a) (3/1/2/9)	US	Adults with ID from two large developmental centres	16-89 (49.81 (12.66); ns)	58.8	0	8.5	11.1	11.8	68.0	0.7	Regularly does not experience a bowel movement every 3 days	5	153	3.3
Matson 2011 (b)											Had constipation prior to the age of 3 mths	41	153	26.8
Matson 2011 (c)											Requires the use of fibre supplements or laxatives to defecate	105	153	68.6
McBrien 2009 (3/0/3/10)	Ireland	Children with moderate to profound ID	5-19 (ns; 12)	66.0	0	0	64.9	35.1	←	0	Being treated for constipation	17	97	17.5
Morad 2007 (3/0/3/11)	Israel	Adults with ID 40+ living in residential care centres	40-ns (49.8 (7.6); ns)	51.4	0	→	52.1	47.9	←	0	Treated for: constipation (ns) within past 24 months	173	2283	7.6
Petry 2009 (1/0/1/5)	Belgium & Netherlands	Children & adults with PMD	5-57 (23.7 (12.2); ns)	53.1	0	0	0	0	100	ns	Medical condition: constipation (ns)	21.6[a]	49	44.1
Staiano 1994 (1/1/4/9)	Italy	Children with severe brain damage, all IQ <35	ns (5.8 (4.2); ns)	53.2	0	0	0	100	←	0	Chronic constipation at least 6 months duration	29	47	61.7
Starr 2008 (3/0/2/8)	Scotland	Adults 40+ with a wide range of ID, living in the community	40-79 (ns (ns); 53)	42.1	0	0	0	0	0	100	Constipation (ns)	30.5[a]	54	56.5
Skotko 2013 (1/1/3/8)	US	Children & adolescents with DS	3.2–20.9 (9.5 (3.8); ns)	63.8	ns	ns	ns	ns	ns	100	Diagnosis of constipation (ns)	36	105	34.3
Straetmans 2007 (a) (4/1/3/12)	Netherlands	People with ID registered with GPs	ns	ns	ns	ns	ns	ns	ns	100	Diagnosis of constipation (ns) during 2001 GP contact	45	868	5.2
Straetmans 2007 (b)											Prescribed laxative during GP contact	143	1000[c]	14.3
Straetmans 2007 (c)											Repeat prescription for laxative	257	1000[c]	25.7

1st Author & Year (1/6/7/total)*	Country	Key sample features	Age range (mean (SD); Mdn)	% male	Border-line % (IQ > 70)	Mild % (50 or 55 - 70)	Moderate % (IQ 35-50/54)	Severe % (IQ 20-34)	Profound % IQ <20	Not specified	Definition of constipation (or other definition)	Cases	N	%
Thomas 2011 (see Leonard 1999) (2/0/1/5)	Australia	Children with DS	5.02 -17.98 (11.7 (ns); ns)	56.7	ns	ns	ns	ns	ns	100	Constipation ns	41	208	19.7
Valicenti-McDermott 2006 (1/1/2/6)	US	Children with developmental disabilities, ambulatory, no genetic disorder, 60% ID	1-18 (7.9 (4); ns)	74	ns	ns	ns	ns	ns	100	Chronic constipation: Rome II criteria. Lifetime prevalence	19	50	38.0
van der Heide 2009 (a) (1/1/3/9)	Netherlands	People with PIMD	6-82 (ns (ns); 49)	46.1	0	0	0	0	100	0	Constipation registered as a health problem	152	254	59.8
(b)											Prescribed laxatives in prior 12 mths	165	254	65.0
Van Winckel 1999 (3/0/3/11)	Flanders, Belgium	Patients with moderate to profound ID in institutions	2-72 (ns (ns); 29)	62.6	0	0	26	40	34	0	Point prevalence of laxative use as indicator of constipation	111	420	26.4
Vande Velde 2010 (3/1/4/12)	Belgium	People with ID 16+ without scoliosis or swallowing disorder	27-41 (35.5 (ns); ns)	50.0	0	0	10.3	41.4	48.3	0	< 3 defecations a week	21	58	36.2
Veugelers 2010 (2/1/4/12)	Netherlands	Children with severe generalized CP	2-18 (9.6 (4.6); ns)	53.3	0	0	→	48.3	51.7	0	As Jauhari 2012	87	152	57.2
Wallace 2007 (1/0/3/7)	Australia	Adults with DS	17-63 (37 (13); ns)	59.6	ns	ns	ns	ns	ns	100	Constipation (not HD) no further definition given	11	57	19.3
Yin 2012 (2/1/3/10)	US	People on DS register	ns for N, only for cases	ns	ns	ns	ns	ns	ns	100	Constipation of severity to warrant investigation for HD	32	713	4.5

*Risk of bias scores for items 1 (possible range 0-4), 6 (0-1), 7 (0-4), and total score (0-15) (higher score = lower risk of bias)
Abreviations: CPRD, Clinical Practice Research Datalink; DS, Down syndrome; MSEL, Mullen Scales of Early Learning; VABS, Vineland Adaptive Behavior Scales; CP, cerebral palsy; ID, intellectual disabilities; HD, Hirschsprung's disease; DD, developmental delay; PIMD, profound intellectual and multiple disabilities;
[a] Exact figure not given, approximated from sample size and percentage figures given
[b] Figures given include some cases that are not included in the final sample size N
[c] Figure reported as per 1,000, actual sample size 868
[d] Excludes those with current urogenital disorder, untreated hypothyroidism, current use of medications known to interfere in bladder or sphincter function, or still using 'diapers'
[e] Figure includes those with IQ >=70 who were classed as having normal intellectual ability in study
← included in previous figure; → included in next figure

Figure 2: *Forest plot of estimates relating to the prevalence of constipation in people with intellectual disability*



[a] Constipation definition relates wholly to laxative prescription or use