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

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Abstract

Dysphagia (feeding and swallowing disorder) is associated with serious health complications and psychosocial sequelae. This review summarises international research relating to the prevalence of dysphagia in people with intellectual disability. Studies published from 1990 to July 2016 were identified using Medline, Cinahl, PsycINFO, Web of Science, email requests, and cross-citations. Twenty studies were identified. Dysphagia in people with intellectual disability appears to be associated with more severe intellectual disability, comorbid cerebral palsy, and motor impairments. However, further research with representative samples of people with intellectual disability using adequate methods of assessment are required in order to provide more precise prevalence estimates and clarify factors that may be associated with dysphagia in this population.

Keywords: dysphagia; intellectual disability; prevalence

Dysphagia (feeding and swallowing disorder) is associated with many health complications. Aspiration pneumonia is considered by many to be the main complication of dysphagia and is of significant concern due to its link with subsequent morbidity and mortality (Heslop et al., 2014; Hollins, Attard, von Fraunhofer, McGuigan, & Sedgwick, 1998; Martino et al., 2005). Other health complications include choking and airway blockage (National Patient Safety Agency, 2007; Samuels & Chadwick, 2006) and compromised nutritional status and dehydration (Kennedy, McCombie, Dawes, McConnell, & Dunnigan, 1997; MacDonald, McConnell, Stephen, & Dunnigan, 1989). In addition, urinary tract infections, headaches, constipation, oesophagitis and reduced ability to combat infections have all been associated with dysphagia (Cichero, 2006; Eustace, Gui, & Iacucci, 2015; Robbins et al., 2008).

In addition to health complications, the psychosocial impacts of dysphagia include loss of opportunity for communication during meals, increased stigma when eating in community settings, and loss of dignity associated with being supported to eat and drink (Chadwick, Jolliffe, Goldbart, & Burton, 2006; Miller, Noble, Jones, & Burn, 2006). Further, reduced choice may occur alongside a managed eating and drinking regime, with an associated loss of enjoyment of meals and drinks due to dietary modification (e.g., thickening drinks, mashed food, and food exclusions) reducing individual quality of life and wellbeing.

The association between dysphagia, its complications, and mortality appears pronounced in people with intellectual disability (ID). Respiratory disease, particularly bronchopneumonia, is a leading cause of death in people with ID, in particular in those with profound intellectual and multiple disabilities (PIMD), accounting for significantly more deaths than in the local general population (Hollins et al., 1998; Patja, Mölsä, & Iivanainen, 2001). Preventable lung inflammation caused by solids or liquids, and foreign bodies has also been associated with mortality in people with ID (Glover & Ayub, 2010). The importance of

dysphagia in relation to the well-being of people with ID led the United Kingdom National Patient Safety Agency (NPSA) to identify swallowing difficulties as one of five priority areas in relation to safety risks for people with ID using healthcare services (National Patient Safety Agency, 2004) and there has been a call for research investigating mealtime safety incidents involving people with intellectual and developmental disabilities (Hemsley, Balandin, Sheppard, Georgiou, & Hill, 2015).

In addition, the issue of dysphagia in people with ID may be complicated by medical co-morbidities, psychiatric, communicative, cognitive and behavioural issues. For example, there is a link between the side-effects of neuroleptic medications and dysphagia (Dziewas et al., 2007) and people with ID are more likely than others to be prescribed these (e.g., anti-psychotic medication; Glover et al., 2015). Further, specific syndromes associated with ID can result in both anatomical and neurological precursors for dysphagia, including Down Syndrome (O'Neill & Richter, 2013; Smith, Teo, & Simpson, 2014), Rubinstein Taybi Syndrome (Shashi & Fryburg, 1995), and Rett Syndrome (Abraham, Taragin, & Djukic, 2015).

The issues outlined above indicate the importance of considering dysphagia specifically in relation to people who have ID. Internationally, there appears to be a lack of research investigating the prevalence of dysphagia in people with ID (Chadwick & Jolliffe, 2009; Leslie, Crawford, & Wilkinson, 2009). A systematic review of 189 studies on dysphagia found that two thirds of the literature focussed on adults over 50 years of age (Roden & Altman, 2013), with only two studies (Calis et al., 2008; Henderson et al., 2009) focussing on people with ID. Further, although there have been systematic reviews conducted in relation to dysphagia and people with cerebral palsy (e.g., Benfer, Weir, & Boyd, 2012; Hirata & Santos, 2012), it cannot be assumed that people with cerebral palsy have ID (Enkelaar,

Ketelaar, & Gorter, 2008). No reviews exist specifically relating to people with ID and dysphagia.

The prevalence of dysphagia varies depending on the diagnostic instrument used, concomitant medical disorders, and the population studied (Wilkins, Gillies, Thomas, & Wagner, 2007). For the general population, literature reviews have reported prevalence rates ranging from 2.3-16% (Kertscher, Speyer, Fong, Georgiou, & Smith, 2015) and 1.7-11.3% (Roden & Altman, 2013), with prevalence being higher in older people (Kertscher et al., 2015; Roden & Altman, 2013). Although ID is a risk factor for dysphagia, with increased likelihood of dysphagia occurring with increasing severity of cognitive impairment (Chadwick & Jolliffe, 2009; Parkes, Hill, Platt, & Donnelly, 2010), it has been noted that the prevalence of dysphagia in populations with ID remains unknown (Leslie et al., 2009). Accurate estimates of dysphagia 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 ID to prevent it being overlooked. We conducted a systematic review of studies that include information on the prevalence of dysphagia in groups of people with ID published from 1990 to July 2016. The purpose of this article is to present the findings from our review, and to address the question of what is known about the prevalence of dysphagia in groups of people with ID.

Method

Electronic literature database searches were conducted in Medline, Cinahl and PsycINFO (all on EBSCO) and Web of Science (SCI-EXPANDED, SSCI, and A&HCI) in June 2015 and subsequently updated in July 2016. Searches combined terms for dysphagia and ID with the Boolean operator 'and'. An example of database specific search terms (Medline) is given in Appendix 1. Searches included broad terms relating dysphagia and people with ID 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 June 2015 a request for information on research relevant to dysphagia and people with ID 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, with the request subsequently being published in the TAC Bulletin in October 2015 (www.teamaroundthechild.com).

Study selection

Inclusion Criteria. Studies were required to meet all of the following criteria: peer reviewed; published between 1990 and 2016; English language full text; primary research; samples of people with ID, or samples (e.g., of people with cerebral palsy) where at least 50% of the sample are explicitly noted to have ID, or mixed samples where results are disaggregated for people with ID; findings that include exact figures on the number of people with dysphagia, or indicators of dysphagia such as particular items on the screening tool of eating problems (STEP).

Exclusion Criteria. Studies published before 1990 were excluded as they predate major changes in service provision for people with ID (Emerson, 2004). Additionally, the following exclusion criteria were applied: not peer reviewed or where peer review status unclear; reviews, letters, commentaries, editorials, meeting or conference abstracts; only included information relating to specific syndromes associated with ID (e.g. Rett syndrome), with the exception of Down syndrome which is the most common genetic cause of ID (Sherman, Allen, Bean, & Freeman, 2007)); conditions where ID cannot be assumed (e.g., cerebral palsy, autistic spectrum disorder) unless the proportion of the sample with ID is explicitly reported to be above 50% or results are disaggregated for people with ID; study relates solely to infants (less than one year of age).

Initially, titles and abstracts were used (by the first author) to exclude studies which were obviously not within scope. Those retained for further screening were those for which relevance could not be assessed without accessing full text, or those that were chosen as being 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 specific inclusion criteria with regards to dysphagia prevalence.

Data were extracted from the full text of included articles by the first author and entered into an Excel database. This included: bibliographic details; the country within which the study took place; details of the focus of the study; sample size and characteristics; study design and data sources; measures employed; main results; and issues raised in the discussion.

Quality Assessment

A method for evaluating aspects of quality considered important to obtaining unbiased estimates of the prevalence of dysphagia in people with ID was developed based on a subset of items from the checklist provided by Munn, Moola, Lisy, Riitano, and Tufanaru (2015) (e.g., Was the sample frame appropriate to address the target population?; Were valid methods used for the identification of the condition?). These items were expanded as indicated below. In addition, an item was developed to indicate the extent to which each study considered prevalence in relation to subgroups of the overall sample.

The following scores were allocated for the item 'Was the sample frame appropriate to address the target population?': Score 4 = study includes a population based sample of people with ID; Score 3 = study includes a sample of people with ID with limited generalisability (e.g., only includes certain types of accommodation, or a specific age band); Score 2 = study includes a representative sample of people with selected characteristics (e.g., limited to a specific severity of ID, or to people with Down syndrome); Score 1 = study includes a sample

of people with selected characteristics (as per score 2) but with limited generalisability (e.g., sample chosen from only one type of setting); Score 0 = study has a poorly defined sample.

The following scores were allocated for the item 'Were valid methods used for the identification of the condition?': Score 5 = dysphagia identified via instrumental assessment e.g., videofluoroscopic swallowing study (VFSS) specifically carried out for the study; Score 4 = dysphagia identified via clinical examination specifically carried out for the study including use of dysphagia screening tools by professionals; Score 3 = dysphagia identified based on information extracted from medical records or other health related documentation (which may or may not include results of methods specified for score 4 and score 5); Score 2 = dysphagia identified via interview with an informant (including STEP); Score 1 = questionnaire self-completion by an informant; Score 0 = method of identification unclear or not stated. If multiple methods were used, the highest level was entered as the score. Finally, for the item 'Prevalence figures presented for subgroup(s)', scores could range from 0 to 6 based on the extent to which additional prevalence figures were available. One point was allocated for each of the following criteria: severity of dysphagia; age; gender; level of ID; specific conditions (e.g., cerebral palsy); and other.

Quality assessment scores are presented in the third column of Table 1. All relevant studies were included in the review regardless of methodological quality. Ninety-five percent (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. It was not possible to compare results between studies directly due to variation in the methods used, and therefore a meta-analysis was not conducted.

Results

The process of identifying studies for inclusion is summarised in Figure 1. Electronic database searches identified 799 references, with 561 remaining after deletion of 238 duplicates. Following screening based on titles and abstracts, 441 references were excluded and a pool of 120 remained for further screening. After examination of full text and the addition of studies cited within these and from other sources, 20 studies met the criteria for inclusion with regards to dysphagia prevalence. These are summarised in Table 1 which gives the definition of dysphagia as used in the study, with some studies presenting figures for multiple items associated with dysphagia rather than a single figure for dysphagia overall. Only one of the 20 studies had a sample that was not entirely comprised of people with ID, with 89% of the sample explicitly noted to have ID (Waterman, Koltai, Downey, & Cacace, 1992).

Figure 1 Here

Geographical spread and Quality Scores

Table 1 shows that all studies were undertaken in high income countries. In terms of geographical spread, 8 studies were from the United States, 4 from the Netherlands, 3 from England, one from Belgium and the Netherlands jointly, and one each from Ireland, Israel, Japan and Spain. With regards to quality scores, 2 studies attained the maximum score of 4 for sample frame (Chadwick and Jolliffe, 2009, Ball et al., 2012), with 10 of the 20 studies being based on samples with selected characteristics (e.g., Down syndrome, PIMD) with limits to generalisability (e.g., only including those living in particular types of settings). The most common method of identifying dysphagia was via information from records (12 studies), with one study using instrumental assessment, and four conducting clinical evaluation. Only 8 studies provided results for subgroups.

Table 1 Here

Study Design

Studies were almost entirely cross-sectional and based on retrospective review of medical records (or other routinely collected information), questions completed either by self-report or interview, or clinical examination. One study conducted clinical assessment as part of prospective evaluation of esophageal motor dysfunction (Zárate, Mearin, Hidalgo, & Malagelada, 2001) and one study collected data as part of longitudinal study on recurrent lower respiratory tract infections (Calis et al., 2008).

Prevalence of dysphagia or indicators of dysphagia

The figures presented in Table 1 (also presented visually as a forest plot in Figure 2) range from 1% of adults with ID specifically noted not to have cerebral palsy (Henderson et al., 2009) to 99% of children with severe generalized cerebral palsy and ID (Calis et al., 2008). Variation in the characteristics of study samples, and methods used for the definition and ascertainment of dysphagia, prohibited the application of meta-analysis procedures.

Figure 2 here

Overall, only two studies employed samples that could be considered representative of the general population of adults with ID (Ball et al., 2012; Chadwick & Jolliffe, 2009). The first of these gives a crude estimate of 11.5% based on detailed investigation of a sample of those identified via speech and language therapists, social care providers and members of the community ID team as requiring mealtime support (Ball et al., 2012). The second gives an estimate of 8.1% based on referrals to a speech and language therapy service where dysphagia was subsequently confirmed via clinical assessment involving videofluoroscopy in 52% of cases (Chadwick & Jolliffe, 2009). The oral stage of swallow was affected for 94.1% of those with dysphagia, the pharyngeal stage for 51.5%, the oesophageal stage for 25.7%, and more than one stage for 58.4%. In both studies, estimates were reliant on dysphagia having been recognised initially by caregivers or professionals, and subsequently confirmed via further investigations.

Higher estimates have been reported in non-population based studies where estimates are not reliant on initial recognition of dysphagia by caregivers or professionals. In one study, 484 out of 929 adults with ID (52.1%) aged 50 or more were reported to have dysphagia based on mealtime observation by trained speech therapists using the *Dysphagia Disorders Survey* (Hermans & Evenhuis, 2014). In a second study, 290 of 416 adults and children with ID (69.7%) were reported to have mild to profound dysphagia based on clinical dysphagia evaluation (Sheppard, Hochman, & Baer, 2014).

There is some indication that there may be under-recognition of dysphagia by staff and family carers. In a study of children with severe generalised cerebral palsy and ID, parents' opinions on the presence of dysphagia did not correlate with the actual presence of clinical features of dysphagia, with parents tending to underestimate the severity of dysphagia (Calis et al., 2008). In one study using both oral motor evaluations during feeding and radiological studies, it was noted that although patients had generally presented to a Feeding Disorders Clinic with a prior diagnosis of rumination, or had unexplained regurgitation, the study identified many abnormalities of deglutition and gastroesophageal function that had not been diagnosed or treated previously (Rogers, Stratton, Victor, Kennedy, & Andres, 1992).

Factors Associated with the Presence of Dysphagia

There are a number of factors associated with the presence of dysphagia that contribute to the variation in prevalence rates reported in Table 1. A critical factor is the severity of ID. Namely, dysphagia appears to be more common in those with more severe ID. Problems on the eating skills category of the STEP (which includes independence) increased with severity of ID. In one study, 11 out of 25 children (44%) with mild ID were noted to have a problem on the eating skills category of the STEP, with the figure increasing to 24 out of 32 (75%) of those with moderate ID, and 33 out of 34 (97.1%) of those with severe or profound ID (Gal, Hardal-Nasser, & Engel-Yeger, 2011). In this study it was also reported that dysphagia

(defined as oral-motor disorders such as abnormalities in sucking, chewing, and lingual movement, and swallowing discoordination) was observed in half the children who had severe ID (but, the exact figure was not presented). In a U.S. study using the STEP, 14 out of 23 items (60.9%) were significantly more frequent in those with profound ID compared with those with mild to severe ID including: does not demonstrate ability to chew (13.2% of those with profound ID vs. 3.4% of those with mild to severe ID); chokes on food (3.5% vs. 2.3%); does not demonstrate ability to swallow (3.8% vs. 1.1%); swallows without chewing sufficiently (5.7% vs. 3.4%); special positioning for feeding (26.1% vs. 3.4%); and requires special equipment for feeding (43.4% vs. 11.4%) (Matson, Fodstad, & Boisjoli, 2008). Although based on small numbers of children with cerebral palsy, one study found increased dysphagia presence with increasing level of ID: none of 6 children without ID (0%) had dysphagia, compared to 2 of 16 children with mild or moderate ID (12.5%), 6 of 15 children with severe ID (40%), and 7 out of 16 children with profound ID (44%) (Waterman et al., 1992).

In studies on people with profound ID and multiple disabilities, swallowing difficulties have been reported in up to 49.0% of participants (Petry, Maes, & Vlaskamp, 2009), with 47.5% reported to have dysphagia or a feeding tube due to dysphagia (van Timmeren, van der Putten, van Schrojenstein Lantman-de Valk, van der Schans, & Waninge, 2016) and 29.5% reported to have dysphagia in a further study (Gittins & Rose, 2008). Although one study reports a lower rate of 20% for dysphagia in non-ambulatory people with profound ID, the definition of dysphagia is unclear, with it also being noted that 23 (44%) were at risk for aspiration pneumonia due to significant oral motor dysfunction (Kozma & Mason, 2003).

Cerebral palsy is also clearly associated with dysphagia. Calis et al. (2008) found that of a sample of 166 children with generalized cerebral palsy and ID, only 2 (1%) did not have dysphagia, with 8% having mild dysphagia, 76% having moderate to severe dysphagia and

15% having profound dysphagia. In one study of those with ID aged 40 or more living in small group homes, dysphagia was reported in 12 of 1196 (1%) of those who did not have cerebral palsy, compared to 11 of 177 (6%) of those who did have cerebral palsy (Henderson et al., 2009). Even when cerebral palsy was adjusted for Severity of Functional Impairment Index (SFII) and ID level, adults with cerebral palsy were still eight times more likely to have dysphagia than those without. Calis et al. (2008) also reported an association between motor impairment (as measured by the *Gross Motor Function Classification System*; GMFCS) and dysphagia severity in children with generalized cerebral palsy and ID, with 24 of 139 children (17.3%) at GMFCS level V having profound dysphagia compared to none of 27 children (0%) at GMFCS level IV (Calis *et al.*, 2008).

One Japanese study of adults with ID and severe motor disabilities with Lennox-Gastaut syndrome suggests an association between seizure outcome and dysphagia (Ogawa *et al.*, 2001). For those with unfavourable seizure outcome (with a favourable outcome defined as a reduction of total seizure frequency to below half of the initial frequency) 14 of 21 individuals (66.7%) had progression of dysphagia compared to 1 of 17 individuals (5.9%) with a favourable outcome (Ogawa *et al.*, 2001). The authors suggest that dysphagia was caused by the persistent epileptic activity itself.

The professional literature provides very scant findings regarding the association of age with dysphagia. For psychiatric inpatients with ID, dysphagia was reported more often in patients in age groups 46-60 and 60 or more than in those aged 16-25 or 26-45 (Exact Permutation Test, $p < 0.0001$; % figures for age groups not given) (Charlot et a*l.*, 2011).

Discussion

This review found extremely wide variation in the figures reported for the prevalence of dysphagia in people with ID. This can be attributed to differences in the characteristics of the samples included, the definition of dysphagia used, and the method of dysphagia

ascertainment. This variation makes comparison between studies difficult. From the international evidence reviewed, two estimates from population based samples of people with ID from areas of England are available and these indicate that around 8.1% to 11.5% of adults known to formal ID services have dysphagia (Chadwick & Jolliffe, 2009; Ball et al., 2012). However, both estimates are reliant on dysphagia having been initially recognised by caregivers or professionals prior to further investigation. Moreover, there is some indication that there may be under-recognition of dysphagia (Calis et al., 2008; Rogers et al., 1992), and therefore the actual prevalence may be higher. Indeed, for less representative samples rates of over 50% have been reported based on mealtime observation by trained speech therapists (Hermans & Evenhuis, 2014) and clinical dysphagia evaluation (Sheppard, Hochman, & Baer, 2014).

The primary aim of many of the included studies was not focused on identifying the prevalence dysphagia in people with ID, with some studies collecting information on a wide range of health issues in people with ID (e.g., McBrien & Macken, 2009). As such, procedures for identifying dysphagia, understandably, did not generally attain high quality scores although the quality of studies in relation to their primary aim may have been high. Chadwick and Jolliffe (2009) acknowledge that the prevalence estimate they give is likely to be an underestimate as it is based on those referred to speech and language therapists with a confirmed diagnosis of dysphagia and thus may only include those with more severe dysphagia. Accurate prevalence estimates are important to those planning services and screening studies of representative samples of people with ID using adequate methods of assessment (e.g., videofluoroscopy) alongside clinical assessment are required (Chadwick & Jolliffe, 2009).

Studies suggest that increasing severity of ID (Gal et al., 2011; Matson et al., 2008; Parkes et al., 2010; Waterman et al., 1992) and cerebral palsy (Calis et al., 2008; Henderson et al.,

2009; Perez et al., 2015) are associated with dysphagia. Motor impairment has also been associated with dysphagia (Calis et al., 2008) and this may contribute to the high prevalence of dysphagia found in people with profound and multiple ID (van der Heide, van der Putten, van den Berg, Taxis, & Vlaskamp, 2009; van Timmeren et al., 2016). There is little evidence regarding an association between age and dysphagia in people with ID, but given that dysphagia has been found to increase with age in the general population (Kertscher et al., 2015; Roden & Altman, 2013), it is reasonable to assume that this will be the case for people with ID (even taking into account possible differential mortality with those with severe dysphagia dying younger) and the association is likely to be compounded by co-morbidities occurring in people with ID such as dementia in people with Down syndrome. Larger scale studies employing multivariate analytic methods would help clarify the factors associated with dysphagia in people with ID.

Limitations

There are a number of limitations to this review. First, although studies were identified from a range of countries, the review is restricted to English language publications and this may have contributed to the lack of any identified studies from low and middle income countries. Second, all data was extracted by one reviewer and extraction of data by two reviewers independently would have reduced the possibility of errors. Third, the quality scores employed were developed specifically for this review and there is no information on the reliability or validity (for example in relation to the accuracy of methods for identifying dysphagia) of the scores. Fourth, studies on relatively rare specific syndromes associated with ID were excluded and future review should consider the unique issues that may arise in these syndromes in relation to dysphagia (c.f., Shashi & Fryburg, 1995; Abraham, Taragin, & Djukic, 2015). Finally, no studies have included the 'hidden majority' of adults with ID who are not known to formal ID services (Emerson, 2011).

Conclusion

Dysphagia is common in people with ID and may be under-recognised. There is a need to discern more accurately the prevalence of dysphagia in people with ID, especially those in low and middle income countries. Improved recognition and management of dysphagia may reduce the occurrence of associated health conditions and reduce hospital admissions and premature death. Those providing services and supports to people with ID need access to resources that provide comprehensive information concerning dysphagia. A recent report by Marriot and Turner (2016) would be an excellent starting point for those seeking additional information. They not only provide essential information, but also offer numerous examples of best practices in relation to reasonable adjustments, as well as other practical ideas to improve dysphagia care for people with ID (Marriott & Turner, 2016).

References

Abraham, S. S., Taragin, B., & Djukic, A. (2015). Co-occurrence of Dystonic and Dyskinetic Tongue Movements with Oral Apraxia in Post-regression Dysphagia in Classical Rett Syndrome Years of Life 1 Through 5. *Dysphagia, 30*, 128-138. doi:10.1007/s00455-014-9587-9

Ball, S. L., Panter, S. G., Redley, M., Proctor, C. A., Byrne, K., Clare, I. C. H. (2012). The extent and nature of need for mealtime support among adults with intellectual disabilities. *Journal of Intellectual Disability Research, 56*, 382-401. doi:10.1111/j.1365-2788.2011.01488.x

Benfer, K. A., Weir, K. A., & Boyd, R. N. (2012). Clinimetrics of measures of oropharyngeal dysphagia for preschool children with cerebral palsy and neurodevelopmental disabilities: a systematic review. *Developmental Medicine and Child Neurology, 54*, 784-795. doi:10.1111/j.1469-8749.2012.04302.x

Calis, E. A. C., Veugelers, R., Sheppard, J. J., Tibboel, D., Evenhuis, H. M., & Penning, C. (2008). Dysphagia in children with severe generalized cerebral palsy and intellectual disability. *Developmental Medicine & Child Neurology, 50*, 625-630. doi:10.1111/j.1469-8749.2008.03047.x

Chadwick, D. D., & Jolliffe, J. (2009). A descriptive investigation of dysphagia in adults with intellectual disabilities. *Journal of Intellectual Disability Research, 53*, 29-43. doi:10.1111/j.1365-2788.2008.01115.x

Chadwick, D. D., Jolliffe, J., Goldbart, J., & Burton, M. H. (2006). Barriers to Caregiver Compliance with Eating and Drinking Recommendations for Adults with Intellectual Disabilities and Dysphagia. *Journal of Applied Research in Intellectual Disabilities, 19*, 153-162. doi:10.1111/j.1468-3148.2005.00250.x

Charlot, L., Abend, S., Ravin, P., Mastis, K., Hunt, A., & Deutsch, C. (2011). Non-psychiatric health problems among psychiatric inpatients with intellectual disabilities. *Journal of Intellectual Disability Research, 55,* 199-209. doi:10.1111/j.1365-2788.2010.01294.x

Cichero, J. (2006). Conditions commonly associated with dysphagia. In J. Cichero & B. Murdoch (Eds.), *Dysphagia: Foundations, Theory and Practice*. Chichester: Wiley & Sons.

Dziewas, R., Warnecke, T., Schnabel, M., Ritter, M., Nabavi, D., Schilling, M. (2007). Neuroleptic-Induced Dysphagia: Case Report and Literature Review. *Dysphagia, 22,* 63-67. doi:10.1007/s00455-006-9032-9

Eayres, D. (2008). Technical Briefing 3: Commonly used public health statistics and their confidence intervals. Retrieved from www.apho.org.uk/resource/view.aspx?RID=48457 (last accessed 1 September 2015).

Emerson, E. (2004). Deinstitutionalisation in England. *Journal of Intellectual and Developmental Disability, 29,* 79-84. doi:10.1080/13668250410001662838

Emerson, E. (2011). Health status and health risks of the "hidden majority" of adults with intellectual disability. *Intellectual and Developmental Disabilities, 49,* 155-165. doi:10.1352/1934-9556-49.3.155

Enkelaar L., Ketelaar M. & Gorter J. W. (2008) Association between motor and mental functioning in toddlers with cerebral palsy. *Developmental Neurorehabilitation, 11,* 276-282. doi:10.1080/17518420802581164

Eustace, G., Gui, X. Y., & Iacucci, M. (2015). Recent advances in the recognition and management of eosinophilic esophagitis. *Canadian Journal of Gastroenterology and Hepatology, 29,* 237-240.

Gal, E., Hardal-Nasser, R., & Engel-Yeger, B. (2011). The relationship between the severity of eating problems and intellectual developmental deficit level. *Research in Developmental Disabilities, 32*, 1464-1469. doi:10.1016/j.ridd.2010.12.003

Gittins, D., & Rose, N. (2008). An audit of adults with profound and multiple learning disabilities within a West Midlands Community Health Trust: Implications for service development. *British Journal of Learning Disabilities, 36*, 38-47. doi:10.1111/j.1468-3156.2007.00480.x

Glover, G., & Ayub, M. (2010). How People with Learning Disabilities Die. Improving Health & Lives: Learning Disabilities Observatory. Available online at http://www.improvinghealthandlives.org.uk/gsf.php5?f=8586&fv=9033 (accessed 25 November 2014).

Glover, G., Williams, R., Branford, D., Avery, R., Chauhan, U., & Hoghton, M. (2015). Prescribing of psychotropic drugs to people with learning disabilities and/or autism by general practitioners in England. Improving Health & Lives: Learning Disabilities Observatory. Available online at http://www.improvinghealthandlives.org.uk/gsf.php5?f=313881&fv=21599 (accessed 24 September 2015).

Guan, X.-L., Wang, H., Huang, H.-S., & Meng, L. (2015). Prevalence of dysphagia in multiple sclerosis: a systematic review and meta-analysis. *Neurological Sciences, 36*, 671-681. doi:10.1007/s10072-015-2067-7

Hemsley, B., Balandin, S., Sheppard, J. J., Georgiou, A., & Hill, S. (2015). A call for dysphagia-related safety incident research in people with developmental disabilities. *Journal of Intellectual and Developmental Disability, 40*, 99-103. doi:10.3109/13668250.2014.994172

Henderson, C. M., Rosasco, M., Robinson, L. M., Meccarello, J., Janicki, M. P., Turk, M. A. (2009). Functional impairment severity is associated with health status among older persons with intellectual disability and cerebral palsy. *Journal of Intellectual Disability Research, 53*, 887-897. doi:10.1111/j.1365-2788.2009.01199.x

Hermans, H., & Evenhuis, H. M. (2014). Multimorbidity in older adults with intellectual disabilities. *Research in Developmental Disabilities, 35*, 776-783. doi:10.1016/j.ridd.2014.01.022

Heslop, P., Blair, P. S., Fleming, P., Hoghton, M., Marriott, A., & Russ, L. (2014). The Confidential Inquiry into premature deaths of people with intellectual disabilities in the UK: a population-based study. *Lancet, 383*, 889-895. doi: 10.1016/S0140-6736(13)62026-7

Hirata, G. C., & Santos, R. S. (2012). Rehabilitation of oropharyngeal dysphagia in children with cerebral palsy: A systematic review of the speech therapy approach. *International Archives of Otorhinolaryngology, 16*, 396-399. doi:10.7162/s1809-97772012000300016

Hollins, S., Attard, M. T., von Fraunhofer, N., McGuigan, S., & Sedgwick, P. (1998). Mortality in people with learning disability: risks, causes, and death certification findings in London. *Developmental Medicine and Child Neurology, 40*, 50-56. doi:10.1111/j.1469-8749.1998.tb15356.x

Kennedy, M., McCombie, L., Dawes, P., McConnell, K. N., & Dunnigan, M. G. (1997). Nutritional support for patients with intellectual disability and nutrition/dysphagia disorders in community care. *Journal of Intellectual Disability Research, 41*, 430-436. doi:10.1111/j.1365-2788.1997.tb00731.x

Kertscher, B., Speyer, R., Fong, E., Georgiou, A. M., & Smith, M. (2015). Prevalence of Oropharyngeal Dysphagia in the Netherlands: A Telephone Survey. *Dysphagia, 30,* 114-120. doi:10.1007/s00455-014-9584-z

Kozma, C., & Mason, S. (2003). Survey of nursing and medical profile prior to deinstitutionalization of a population with profound mental retardation. *Clinical Nursing Research, 12,* 8-22. doi:10.1177/1054773803238737

Leslie, P., Crawford, H., & Wilkinson, H. (2009). People with a learning disability and dysphagia: a cinderella population? *Dysphagia, 24*, 103-104. doi:10.1007/s00455-008-9153-4

MacDonald, N. J., McConnell, K. N., Stephen, M. R., & Dunnigan, M. G. (1989). Hypernatraemic dehydration in patients in a large hospital for the mentally handicapped. *British Medical Journal, 299*, 1426-1429.

Marriott, A., & Turner, S. (2016). Making reasonable adjustments to dysphagia services for people with learning disabilities. Improving Health & Lives: Learning Disabilities Observatory. Available online at http://www.improvinghealthandlives.org.uk/gsf.php5?f=314186&fv=22099 (accessed 22 August 2016).

Martino, R., Foley, N., Bhogal, S., Diamant, N., Speechley, M., & Teasell, R. (2005). Dysphagia After Stroke: Incidence, Diagnosis, and Pulmonary Complications. *Stroke, 36*, 2756-2763. doi:10.1161/01.STR.0000190056.76543.eb

Matson, J. L., Fodstad, J. C., & Boisjoli, J. A. (2008). Cutoff scores, norms and patterns of feeding problems for the Screening Tool of fEeding Problems (STEP) for adults with intellectual disabilities. *Research in Developmental Disabilities, 29*, 363-372. doi: 10.1016/j.ridd.2007.06.001

McBrien, J., & Macken, S. (2009). Meeting the health care needs of school-age children with intellectual disability. *Irish Medical Journal, 102*, 252-255.

Miller, N., Noble, E., Jones, D., & Burn, D. (2006). Hard to swallow: dysphagia in Parkinson's disease. *Age and Ageing, 35*, 614-618. doi:10.1093/ageing/afl105

Munn, Z., Moola, S., Lisy, K., Riitano, D., & Tufanaru, C. (2015). Methodological guidance for systematic reviews of observational epidemiological studies reporting prevalence and cumulative incidence data. *International Journal of Evidence Based Healthcare, 13*(3), 147-153. doi:10.1097/XEB.0000000000000054

National Patient Safety Agency. (2004). Understanding the patient safety issues for people with learning disabilities. Retrieved from http://www.npsa.nhs.uk/EasySiteWeb/GatewayLink.aspx?alId=839 (accessed 14 May 2015).

National Patient Safety Agency. (2007). Problems swallowing? Ensuring safer practice for adults with learning disabilities who have dysphagia: resources for healthcare staff. Retrieved from http://www.nrls.npsa.nhs.uk/EasySiteWeb/getresource.axd?AssetID=60131&type=full&servicetype=Attachment (accessed 14 May 2015).

O'Neill, A. C., & Richter, G. T. (2013). Pharyngeal Dysphagia in children with Down syndrome. *Otolaryngology Head & Neck Surgery, 149*, 146-150. doi:10.1177/0194599813483445

Ogawa, K., Kanemoto, K., Ishii, Y., Koyama, M., Shirasaka, Y., Kawasaki, J. (2001). Long-term follow-up study of Lennox-Gastaut syndrome in patients with severe motor and intellectual disabilities: with special reference to the problem of dysphagia. *Seizure, 10*, 197-202. doi:10.1053/seiz.2000.0483

Parkes, J., Hill, N., Platt, M. J., & Donnelly, C. (2010). Oromotor dysfunction and communication impairments in children with cerebral palsy: a register study. *Developmental Medicine and Child Neurology, 52*, 1113-1119. doi:10.1111/j.1469-8749.2010.03765.x

Patja, K., Mölsä, P., & Iivanainen, M. (2001). Cause-specific mortality of people with intellectual disability in a population-based, 35 year follow-up study. *Journal of Intellectual Disability Research, 45*, 30-40. doi:10.1111/j.1365-2788.2001.00290.x

Perez, C. M., Ball, S. L., Wagner, A. P., Clare, I. C. H., Holland, A. J., & Redley, M. (2015). The incidence of healthcare use, ill health and mortality in adults with intellectual disabilities and mealtime support needs. *Journal of Intellectual Disability Research, 59*, 638-652. doi:10.1111/jir.12167

Petry, K., Maes, B., & Vlaskamp, C. (2009). Measuring the Quality of Life of People with Profound Multiple Disabilities Using the QOL-PMD: First Results. *Research in Developmental Disabilities, 30*, 1394-1405. doi:10.1016/j.ridd.2009.06.007

Robbins, J., Gensler, G., Hind, J., Logemann, J. A., Lindblad, A. S., Brandt, D. (2008). Comparison of two interventions for liquid aspiration on pneumonia incidence. *Annals of Internal Medicine, 148*, 509-518. doi:10.7326/0003-4819-148-7-200804010-00007

Roden, D. F., & Altman, K. W. (2013). Causes of dysphagia among different age groups: a systematic review of the literature. *Otolaryngologic Clinics of North America, 46*, 965-987. doi:10.1016/j.otc.2013.08.008

Rogers, B., Stratton, P., Victor, J., Kennedy, B., & Andres, M. (1992). Chronic regurgitation among persons with mental retardation: A need for combined medical and interdisciplinary strategies. *American Journal on Mental Retardation, 96*, 522-527.

Samuels, R., & Chadwick, D. D. (2006). Predictors of asphyxiation risk in adults with intellectual disabilities and dysphagia. *Journal of Intellectual Disability Research, 50*, 362-370. doi:10.1111/j.1365-2788.2005.00784.x

Shashi, V., & Fryburg, J. S. (1995). Vascular ring leading to tracheoesophageal compression in a patient with Rubinstein-Taybi syndrome. *Clinical Genetics, 48*, 324-327. doi:10.1111/j.1399-0004.1995.tb04119.x

Sheppard, J. J., Hochman, R., & Baer, C. (2014). The Dysphagia Disorder Survey: Validation of an assessment for swallowing and feeding function in developmental disability. *Research in Developmental Disabilities, 35*, 929-942. doi:10.1016/j.ridd.2014.02.017

Sherman, S. L., Allen, E. G., Bean, L. H., & Freeman, S. B. (2007). Epidemiology of Down syndrome. *Mental Retardation and Developmental Disabilities Research Reviews, 13*, 221-227. doi:10.1002/mrdd.20157

Smith, C. H., Teo, Y., & Simpson, S. (2014). An observational study of adults with Down syndrome eating independently. *Dysphagia, 29*, 52-60. doi:10.1007/s00455-013-9479-4

van der Heide, D. C., van der Putten, A. A. J., van den Berg, P. B., Taxis, K., & Vlaskamp, C. (2009). The Documentation of Health Problems in Relation to Prescribed Medication in People with Profound Intellectual and Multiple Disabilities. *Journal of Intellectual Disability Research, 53*, 161-168. doi:10.1111/j.1365-2788.2008.01141.x

van Timmeren, E. A., van der Putten, A. A. J., van Schrojenstein Lantman-de Valk, H. M. J., van der Schans, C. P., & Waninge, A. (2016). Prevalence of reported physical health problems in people with severe or profound intellectual and motor disabilities: a cross-sectional study of medical records and care plans. *Journal of Intellectual Disability Research, Online Early*. doi:10.1111/jir.12298

Waterman, E. T., Koltai, P. J., Downey, J. C., & Cacace, A. T. (1992). Swallowing disorders in a population of children with cerebral palsy. *International Journal of Pediatric Otorhinolaryngology, 24*, 63-71.

Wilkins, T., Gillies, R. A., Thomas, A. M., & Wagner, P. J. (2007). The Prevalence of Dysphagia in Primary Care Patients: A HamesNet Research Network Study. *The Journal of the American Board of Family Medicine, 20*, 144-150. doi:10.3122/jabfm.2007.02.060045

Zárate, N., Mearin, F., Hidalgo, A., & Malagelada, J. R. (2001). Prospective evaluation of esophageal motor dysfunction in Down's syndrome. *The American Journal of Gastroenterology*, *96*(6), 1718-1724. doi:10.1111/j.1572-0241.2001.03864.x

Appendix 1: Example of Database Specific Search (Medline)

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

Limits: English language, human, published from 1990.

This is the authors' accepted version of a paper due to appear in a forthcoming issue of *Intellectual and Developmental Disability*

Figure 1

Flowchart of Study Identification

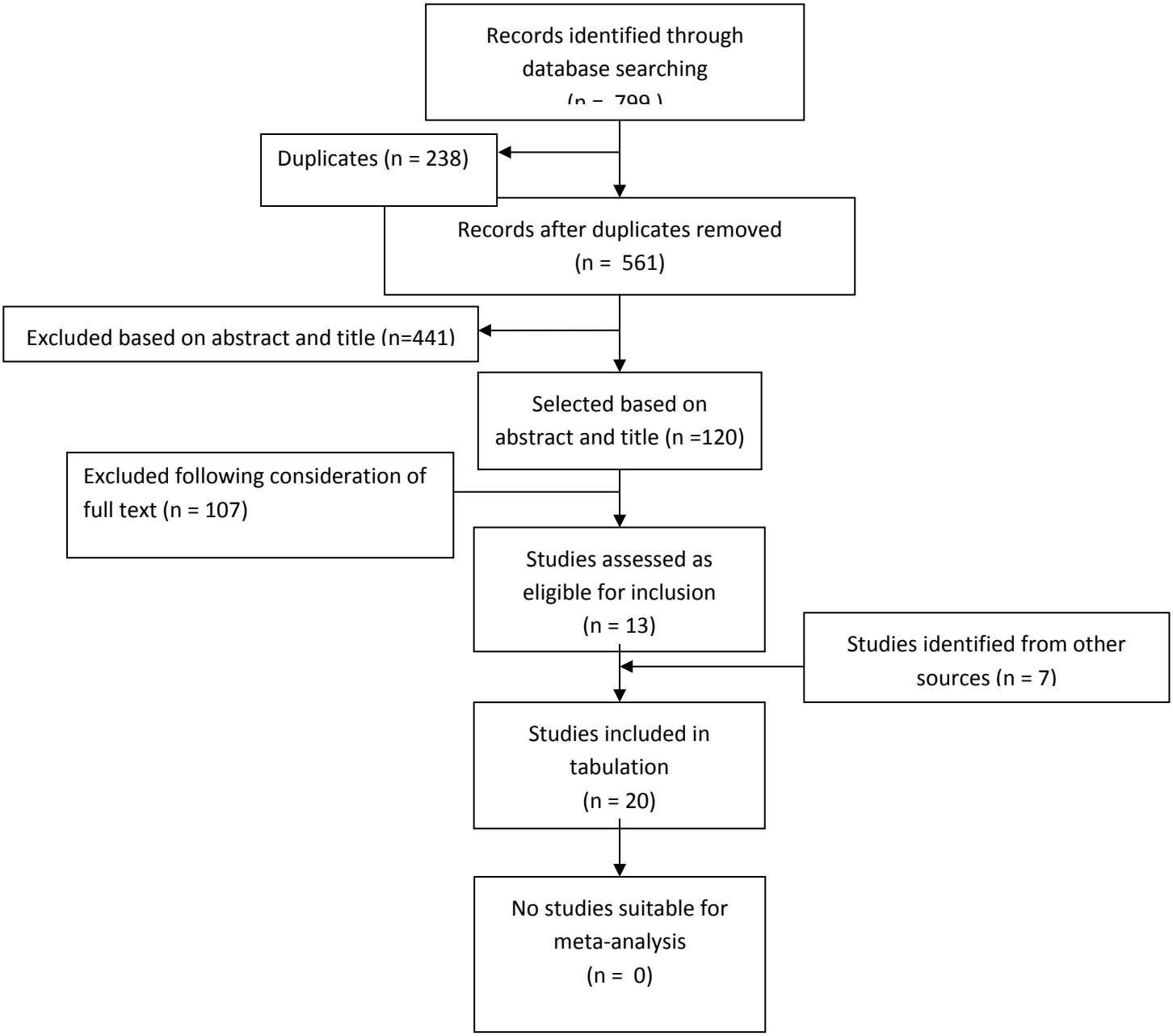

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Table 1. *Summary of Studies Including Figures Relating to the Prevalence of Dysphagia in People with Intellectual Disability*

First author & year	Country	Quality Score*	Sample characteristics (in addition to ID)	Dysphagia definition	Cases/N	%	95% CI
Ball 2012	England	4/3/0	Adults known to ID services	Chewing, oral control, swallowing	256/2230 (extrapolated)	11.5	10.2, 12.9
Calis 2008	Nether-lands	2/4/2	Children with severe cerebral palsy	DDS part 2	164/166	98.8	95.7, 99.7
Chadwick 2009	England	4/3/0	Adults referred to SLTs	Oral, pharyngeal or oesophageal dysphagia	99/1215	8.1	6.7, 9.8
Charlot 2011	US	1/3/0	Psychiatric inpatients	Dysphagia (ns)	20/198	10.1	6.6, 15.1
Gal 2011	Israel	3/2/1	Children in segregated education mild to profound ID	STEP feeding skills category	69[a]/91	75.8	66.1, 83.5
Gittins 2008	England	2/3/0	Adults with PMLD	Dysphagia (problems with eating & drinking)	18/61	29.5	19.6, 41.9
Henderson 2009	US	3/3/1	a. Age 40+ small group homes (not CP)	ICD9 code 787.2x dysphagia (swallowing disorder)	12[a]/1196	1.0	0.6, 1.7
			b. Age 40+ small group homes (CP)		11[a]/177	6.0	3.5, 10.8
Hermans 2014	Nether-lands	3/4/1	Age 50+ known to ID services	DDS mealtime observation	484/929	52.1	48.9, 55.3
Kozma 2003	US	1/3/0	Institutionalized adults profound ID	Dysphagia (ns) - abnormal esophageal motility mentioned	11/55	20.0	11.6, 32.4
Matson 2008	US	3/2/2	Adolescent/adult inpatients (81% with profound ID)	a. STEP item does not demonstrate ability to chew	52/459	11.3	8.7, 14.6
				b. STEP item chokes on food	15/459	3.3	2.0, 5.3
				c. STEP item does not demonstrate ability to swallow	15/459	3.3	2.0, 5.3
				d. STEP item special positioning for feeding	100/459	21.8	18.3, 25.8
				e. STEP item special equipment for feeding	171/459	37.3	33.0, 41.8
				f. STEP item swallows without chewing sufficiently	24/459	5.2	3.5, 7.7
McBrien 2009	Ireland	3/3/0	Children moderate to profound ID	Feeding difficulties – GI/neurological/ behavioural	8/97	8.2	4.2, 15.4
Ogawa 2001	Japan	1/3/1	Adults with Lennox-Gastaut syndrome & severe motor & ID	Swallowing difficulties &/or having 2+ episodes aspiration pneumonia	15/38	39.5	25.6, 55.3
O'Neill 2013	US	1/3/0	Children with DS at tertiary care hospital	Pharyngeal dysphagia	116/201	57.7	50.8, 64.3
Petry 2009	Belgium & Nether-lands	1/1/0	Children & adults with PMD	a. Swallowing difficulties	24[a]/49	49.0	35.6, 62.5
				b. Tube fed	14[a]/49	28.6	17.8, 42.4
				c. Chewing difficulties	30[a]/49	61.2	47.2, 73.6
Rogers 1992	US	1/5/0	Chronic regurgitation & profound ID	Oral or pharyngeal dysphagia	19/23	82.6	62.9, 93.0
Sheppard 2014	US	3/4/1	Adult & child residents at two centers	CDE; rated as none, mild, moderate, severe, profound	290/416	69.7	65.1, 73.9
van der Heide 2009	Nether-lands	1/3/0	Children & adults with PIMD	a. Problems with swallowing	37/254	14.6	10.8, 19.4
				b. Feeding tube	32/254	12.6	9.1, 17.2
van Timmeren 2016	Nether-lands	1/3/0	Adults with severe or profound ID and motor disabilities	Dysphagia & feeding tube due to dysphagia	47[a]/99	47.5	37.9, 57.2
Waterman	US	1/3/1	Children with CP	Swallowing disorder - oral	15/56	26.8	17.0, 39.6

1992				preparatory/phayrngeal, esophageal			
Zárate 2001	Spain	1/4/0	Adults & children with DS	a. Esophageal symptom dysphagia for liquids	9/58	15.5	8.4, 26.9
				b. Esophageal symptom dysphagia for solids	10/58	17.2	9.6, 28.9

*Sample frame (range 0-4)/identification method (range 0-5)/subgroups reported (range 0-6).

[a] numerator not given, approximated from percentage rate presented & sample size. *Abbreviations:* CDE clinical dysphagia evaluation; CP cerebral palsy; DDS Dysphagia Disorders Survey; DS Down syndrome; GI gastrointestinal; ns not specified; PIMD profound intellectual & multiple disabilities; PMD profound multiple disabilities.

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Figure 2

Forest plot of estimates relating to the prevalence of dysphagia in people with intellectual disability