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Published in:
Journal of Intellectual Disability Research

DOI:
10.1111/jir.12296

Publication date:
2017

JIDR Journal of Intellectual Disability Research
Published on behalf of mencap and in association with IASSID

Journal of Intellectual Disability Research doi: 10.1111/jir.12296

VOLUME 61 PART 1 pp 30–49 JANUARY 2017

Physical health issues in adults with severe or profound intellectual and motor disabilities: a systematic review of cross-sectional studies

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Abstract

Background People with severe or profound intellectual and motor disabilities (SPIMD) encounter several risk factors associated with higher mortality rates. They are also likely to experience a cluster of health problems related to the severe brain damage/dysfunction. In order to earlier detect physical health problems in people with SPIMD, first of all, knowledge regarding the prevalence of physical health problems is necessary. The aim of this systematic review was to methodically review cross-sectional studies on the prevalence of various types of physical health problems in adults with SPIMD.

Method MedLine/PubMed, CINAHL, Embase, PsycINFO and Web of Science were searched for studies published between 2004 and 2015. The quality of the incorporated studies was assessed utilising an adjusted 'risk of bias tool' for cross-sectional studies. To estimate the prevalence of the health problems, the proportion and corresponding confidence interval were calculated. A random effect meta-analysis was performed when at least three studies on a specific health problem were available.

Results In total, 20 studies were included and analysed. In the meta-analysis, a homogeneous prevalence rate of 70% (CI 65–75%) was determined for epilepsy. Heterogeneous results were ascertained in the meta-analysis for pulmonary/respiratory problems, hearing problems, dysphagia, reflux disease and visual problems. For the health problems identified in two studies or in a single study, the degree of evidence was low. As expected, higher prevalence rates were found in the current review compared with people with ID for visual problems, epilepsy and spasticity.

Conclusion This review provides an overview of the current state of the art research on the prevalence of health problems in adults with SPIMD. There is a substantial need for comprehensive epidemiological data in order to find clusters of health problems specific

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for people with SPIMD. This would provide insight into the excess morbidity associated with SPIMD.

Keywords adults, physical health, review, severe or profound intellectual and motor disabilities, severe or profound intellectual disabilities

Background

People experiencing severe or profound intellectual and motor disabilities (SPIMD) are more likely to have a number of specific health needs; therefore, they represent a vulnerable group. This population has two key characteristics: (1) severe or profound intellectual disability (ID) and (2) profound motor disability manifesting in an inability to move independently. The majority of people with SPIMD additionally experience sensory impairments and physical health problems (Nakken & Vlaskamp 2007). Research in the Netherlands indicates a population size of a maximum of 9639 adults with SPIMD (Vugteveen *et al.* 2014).

People with SPIMD encounter several risk factors associated with higher mortality rates such as a lower IQ, non-ambulation, poor motor skills, inability to feed oneself, poor communication and self-help limitations. They are likely to gradually have different patterns of health problems than those with a milder ID (Sutherland *et al.* 2002; Hayden 1998) and suffer more often from multimorbidity (Hermans & Evenhuis 2014). Furthermore, they frequently experience a cluster of health problems related to the severe brain damage/dysfunction such as dysphagia, epilepsy, spasticity, reflux disease, hearing impairment and visual impairment. Moreover, they have an increased risk for polypharmacy and secondary health problems (Hermans & Evenhuis 2014; Van Schrojenstein Lantman-de Valk & Walsh 2008). For example, undetected constipation may cause severe problems such as ileus (Van Schrojenstein Lantman-de Valk & Walsh 2008), and anticonvulsant medication is associated with osteoporosis (Mergler *et al.* 2009).

Early detection of physical health problems is important in order to improve or maintain health and quality of life (Kerr *et al.* 2003; Robertson *et al.* 2014). If health problems remain undiagnosed and untreated, secondary health complications can occur (McCarthy & O'Hara 2011; Cooper *et al.* 2004; May & Kennedy 2010); therefore, it is important to identify health issues in time. However, little is known about the prevalence of physical health problems in the SPIMD population.

The aim of this systematic review, therefore, is to methodically review cross-sectional studies on the prevalence of various types of physical health problems in adults with SPIMD.

Methods

Information sources

The Cochrane Library was initially searched in order to confirm that a systematic review did not already exist on physical health issues in adults with SPIMD. Subsequently, a systematic literature search was conducted in the databases of MedLine/PubMed, CINAHL, Embase, PsycINFO and Web of Science. Terms were searched as text words (in title, abstract) and key words that have been indexed by the databases. In addition, the reference lists of the included articles were screened.

Search strategy

To ensure capturing all potentially relevant studies, a broad range of search terms was employed. The search terms of profound intellectual and motor disabilities (PIMD) were extended to SPIMD as there is no standardised instrument to assess cognitive functioning in a person who also has severe/profound motor and sensory disabilities (Nakken & Vlaskamp 2007).

Along with searching for studies on individuals with SPIMD, studies regarding individuals with severe or profound intellectual disabilities (SPID) were also examined because most of them have an increased frequency of additional impairments. Furthermore, syndrome-specific terms such as Rett Syndrome were utilised.

To provide a broad overview of all physical health problems, physical health related terms were used in preference to specific health problem terms. An example of the database-specific search for MedLine/PubMed is shown in the Appendix.

Selection criteria for studies

Criteria for inclusion in this review included cross-sectional studies, peer-reviewed articles written in

English, published within the last 10 years (between January 2004 and December 2015). Furthermore, participants in the studies were required to be adults with SPID or SPIMD. For a comprehensive overview, studies involving both children and adults and all physical health problems were included. As the focus of the study was on physical health problems, we excluded mental and behavioural disorders such as sleep issues and challenging behaviours. For studies involving adults with a varying degree of ID, a separate analysis of the data on physical health problems for people with SPID was necessary for inclusion. The selection process was performed by the first author. The title and abstract of all of the obtained articles were screened utilising the selection criteria. A second reviewer (A. W.) independently and randomly screened 10% of the titles and abstracts, which yielded an inter-rater agreement of 100%.

Data extraction

Two reviewers (E. A. v. T. and H. A. S.) independently analysed the study characteristics utilising a protocol for data extraction, which was developed specifically for the review and included the following items:

1. Study design
2. Population/participants related to the review area
   - a Inclusion/exclusion criteria, setting, country
   - b Number of participants related to the review area
   - c Patients characteristics (age, gender)
   - d Non-response/non-participants
3. Health problem related to the review area
   - a Definition of health problem
   - b Measurement of health problem
   - c Prevalence rates of health problems
4. Specific issues raised by the study that are relevant to the review area

Discrepancies, for example, in the definition of a health problem or in the total number of participants, were resolved with discussions between the two reviewers.

Assessment of study quality

Because of the absence of standard criteria for the assessment of quality of the cross-sectional studies, an existing assessment tool (Hoy *et al.* 2012) for determining risk of bias was modified whereby the criteria of the tool and the scores were adapted to the focus of this current review. For external validity (EV), the risk of bias was assessed for the representativeness of the study group with the target group of this review, i.e. people with SPIMD. For internal validity (IV), the method of measurement and definition of the psychical health problem was evaluated. In the event that the physical health problems were not the primary focus of the included study, the quality assessment for IV was limited to the elements of the study that were relevant to the physical health problems related to the review area. The tool contains four items for EV and five items for IV. For the total score, the number of 'YES' judgements (i.e. risk of bias) were compiled. A total score of 0 to 1 for EV was considered as a high risk of bias, scores of 2 to 3 as a moderate risk of bias and scores of 4 as a low risk of bias. High risk of bias for internal validity was indicated by total scores of 0 to 1, a moderate risk of bias by scores of 2 to 3 and a low risk of bias by scores of 4 to 5. The two reviewers reached 100% consensus on the total number of quality points for each study.

The protocol for quality assessment included the following items:

External validity
1. Target population representative for the SPIMD group
2. Sampling frame representative for target population
3. Selection of the sample
4. Likelihood of non-response

Internal validity
5. Data collection
6. Definition health problem
7. Measurement health problem
8. Mode of data collection used for all subjects
9. Appropriate numerator(s) and denominator(s)

Data analyses

For this review, the prevalence rate of the physical health problems were classified according to the

categories of the tenth revision of the International Statistical Classification of Diseases (ICD-10). Prevalence data indicate the number of people with the health problem at a specified time. Therefore, proportion and relative frequency were notated and, if necessary, calculated. A 95% confidence interval (CI) was determined for each health problem using the Confidence Interval Calculator for Proportions (McCallum Layton n.d.). A narrow 95% CI indicates results that are more accurate based on the sample size.

A random effect meta-analysis (Viechtbauer 2010) on the proportions of the individual studies was performed to assess the prevalence of the health problem if three or more studies concerning the same health problem while using the same focus and design were identified. Forest plots were used to visualise meta-analysis results, and the degree of heterogeneity was described by the I^2 and Q statistic. The presence of heterogeneity was tested utilising the Q statistic.

Results

Process of study selection

The database search yielded 9319 results. After eliminating the duplicates, 7034 results remained. A total of 57 records were identified as being potentially relevant for this review.

After a full text review, 16 articles satisfied the eligibility criteria and were subsequently included. In addition, after screening the reference lists of the included articles and using the same selection criteria, another four studies were added. A total of 20 studies met the inclusion criteria. Figure 1 outlines the study selection and lists the explanations for excluding the other studies.

Description of the studies

The number of participants ranged from 34 to 562. In seven studies, both adults and children (age 0–82 years) were included (Van der Heide *et al.* 2009; Petry *et al.* 2009; Fellinger *et al.* 2009; Van den Broek *et al.*

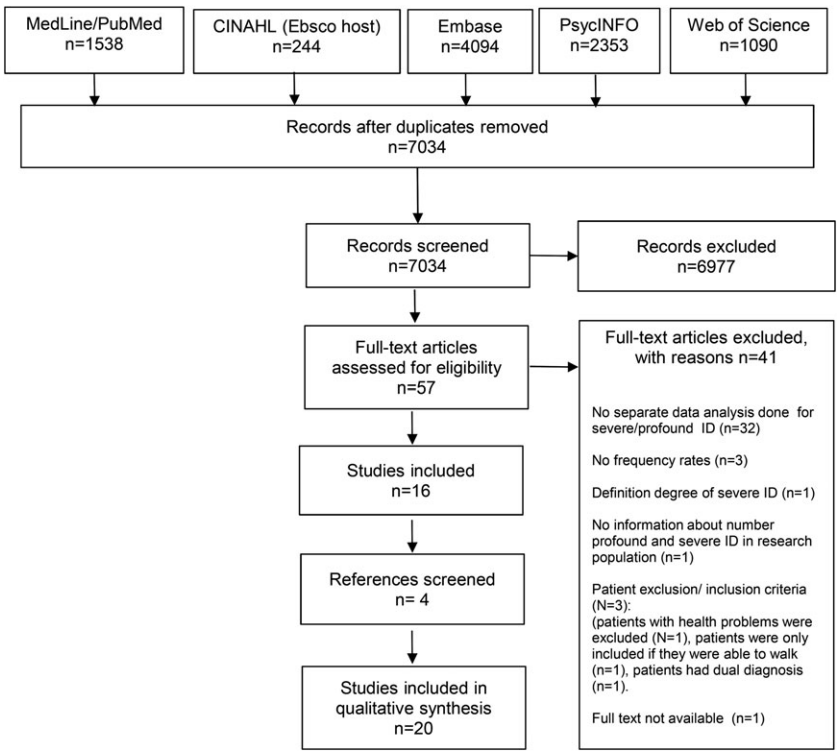

Figure 1 Flowchart of study selection process.

2; Poppes *et al.* 2010; Van den Akker *et al.* 2006; McGuire *et al.* 2010). Eight studies included a study population that was representative for SPIMD (Gittins & Rose 2008; Nagae *et al.* 2013; Ohwada *et al.* 2006; Ohwada & Nakayama 2008; Van den Broek *et al.* 2006; Poppes *et al.* 2010; Van der Heide *et al.* 2009; Petry *et al.* 2009). All of the studies included both male and female. One study analysed data for people with SPID and SPIMD (De Waal *et al.* 2009). Table 1 contains the characteristics and risk of bias for each study. The studies are listed in order from low to high risk of bias for IV, i.e. the method of measurement and definition of the psychical health problem.

Quality assessment

The quality rating of the studies ranged between moderate and high risk of bias for EV and between low and high risk of bias for IV.

External validity

1 Target population
Twelve studies included a broad range of ID levels in the study population. These studies were assessed as a less accurate representation of the SPIMD group.

2 Sampling frame
The sampling frame, in most of the studies, was a list from only one organisation/institute. In certain studies, the sampling frame consisted of several facilities; however, these studies investigated individuals aged ≥50 (De Winter *et al.* 2011; Bastiaanse *et al.* 2012) or did not include people who were living in settings that are more community-based (Poppes *et al.* 2010) or provided no information in regard to the sampling frame (Hove 2004). None of the sampling frames in the included studies was a genuine or accurate representation of the SPIMD population.

3 Selection sample
In most of the studies, the target population was residing in an institution wherein every person with SPIMD was sampled. There was an elevated risk of bias if not all of those with SPIMD were included (Van den Akker *et al.* 2006), if participation was open (Lin *et al.* 2012), if the selection was a component of the research (Hove 2004; Gittins & Rose 2008) or if no information was provided (McGuire *et al.* 2010; Petry *et al.* 2009).

4 Likelihood of non-response
In the SPIMD target population, non-response includes non-permission and missing data because of non-cooperation, physical impairments or limited understanding. A high risk was assessed if no information was provided (Poppes *et al.* 2010; Petry *et al.* 2009); if participation was voluntary (Lin *et al.* 2012); if the response rate was <75% and no analysis was performed to compare responders and non-responders (McGuire *et al.* 2010; Van der Heide *et al.* 2009; Ohwada *et al.* 2006; Lohiya *et al.* 2004; Hove 2004; Ohwada & Nakayama 2008); or if the analysis indicated a significant difference in relevant demographic characteristics between responders and non-responders (Bastiaanse *et al.* 2012; De Waal *et al.* 2009). In the remaining 11 studies, a low risk of bias was assessed because the entire sample was included or the response rate was >75%.

Internal validity

5 Data collection method
The methods of data collection in the studies can be divided into clinical examination ($n = 9$), data from an existing health check-up ($n = 4$), case file notes ($n = 3$) and questionnaire by proxy ($n = 4$). A clinical examination and data from an existing health check-up were considered as a low risk of bias. The remaining two forms of data collection (i.e. case file notes and questionnaire by proxy) were assessed as a high risk of bias.

6 Definition of health problem
Seventeen studies investigated one specific issue. In two cases, no acceptable definition of the health problem was indicated. One study on anaemia employed reference values of the institution instead of the standard value for anaemia proposed by the WHO (Ohwada *et al.* 2006), and the second study provided no instructions on how to observe pain in people with ID (McGuire *et al.* 2010). Furthermore, four studies investigated a range of physical health problems wherein no definitions of the specific physical health

Table 1 Characteristics and risk of bias of included studies ($n = 20$)

Author (year of publication)	Number of participants relevant for review (percentage of research population), research population, age (years), gender	Setting + country	Health problem	Methods of diagnosis health problem	Risk of bias: internal validity (score)	Risk of bias: external validity (score)
Nagae *et al.* (2013)	82, SMID, age mean 39.4 (SD 12.6; median 39), men 50%	A residential hospital, Japan	Vitamin K deficiency: based on vitamin K-dependent hepatic markers (indicating bleeding tendency) and vitamin K-dependent bone markers (indicating osteoporosis)	Blood samples for vitamin K status in the liver and bone	Low (5)	Moderate (3)
Van den Broek *et al.* (2006)	74, SPMD, age mean 33 (SD 12.6; range 4–74), men 58%	A care facility, The Netherlands	Visual functioning: low vision; blind	Visual screening by an orthoptist	Low (5)	Moderate (3)
Ohwada and Nakayama (2008)	39, SMID, men ($n = 21$), age mean 38.5 (SD 10.6), women ($n = 18$), age mean 35.2 (SD 9.9)	A public facility, Japan	Poor nutritional status indicated by serum albumin	Retrospectively analysed the existing health check-up data (blood profile tests) for each resident	Low (5)	Moderate (2)
Fodstad (2010)	131, SPID (78.9%), study population: 166 mild–profound ID and antipsychotic medication (Axis I diagnoses), age mean 51 (SD 14; range 17–82), Men 54%	A developmental centre, USA	Tardive dyskinesia	Tardive dyskinesia DSM-IV-TR criteria, Dyskinesia Identification System-Condensed User Scale (DISCUS)	Low (5)	Moderate (2)
Matson (2008)	123, SPID (76.4%), study population: 161 mild–profound ID and long-term psychotropic medication use (Axis I diagnoses), age mean 49.84 (SD 14.31; range 18–90), men 53%	Two developmental centres, USA	Tardive dyskinesia, tardive akathisia	DSM-IV-TR criteria, the outcome of a DISCUS or AIMS administration	Low (5)	Moderate (2)

Table 1. *(Continued)*

Author (year of publication)	Number of participants relevant for review (percentage of research population), research population, age (years), gender	Setting + country	Health problem	Methods of diagnosis health problem	Risk of bias: internal validity (score)	Risk of bias: external validity (score)
De Waal *et al.* (2009)	254, SPID (73.4%), study population: 346 moderate–profound ID, age mean 54.4 (SD 12.2; range 18–82), men 57% 56, PMD (16%)	A large care provider, The Netherlands	Post-void residual urine volume (PVR)/retention: after voiding, residual volumes above 150 mL	Ultrasound scanning	Low (5)	High (1)
De Winter *et al.* (2011)	210, SPID (51%), study population: 470 mild–profound ID, age mean 61.0 (range 50–90), men 27%	Three care providing agencies, The Netherlands	The metabolic syndrome: NCEP-ATP III criteria	Physical examinations and blood samples	Low (4)	Moderate (2) Moderate (2)
Fellinger *et al.* (2009)	195, SPID (87.1%), study population: 253 moderate–profound ID, age mean 30.7 (SD 8.5; range 3–55), men 62%	An institute for people with ID, Austria	Deaf-blind: hearing impairment and visual impairments	Clinical examination by orthoptists and an audiologist	Low (4)	Moderate (2)
Ohwada *et al.* (2006)	39, SMID, men ($n = 21$), age mean 38.5 (SD 10.6), women ($n = 18$), age mean 35.2 (SD 9.9)	A public facility, Japan	Anaemia: haemoglobin value in blood. Reference values of the institution. Co-morbid conditions with anaemia	Analysing existing health check-up data (blood profile tests) Review of the medical charts	Low (4) High (1)	Moderate (2) Moderate (2)
Bastiaanse *et al.* (2012)	194, SPID (21.9%), study population: 884 borderline–profound ID, age > 50, men 50.9%	Three care provider services, the Netherlands	Sarcopenia: muscle mass, muscle strength and muscle performance	Calf circumference, grip strength and walking speed	Low (4)	High (1)
Lohiya *et al.* (2004)	562, SID on whom they could measure bone mineral density (BMD), age mean 45 (range 30–82), men 61%	A long-term care facility for people with severe ID, USA	Low bone mass (measurement BMD)	Measure BMD by peripheral dual-energy X-ray absorptiometry (DXA) on the residents' middle fingers by Accudexa	Low (4)	High (1)

Table 1. *(Continued)*

Author (year of publication)	Number of participants relevant for review (percentage of research population), research population, age (years), gender	Setting + country	Health problem	Methods of diagnosis health problem	Risk of bias: internal validity (score)	Risk of bias: external validity (score)
Hsu *et al.* (2012)	87, SPID (53%), study population: 164 borderline–profound ID, age ≥ 20, mean 33 (SD 9.0), men 61%	A private disability welfare institution, Taiwan	Metabolic syndrome (MS)	Annual health examination chart of 2009	Moderate (3)	Moderate (2)
Lin *et al.* (2012)	267, SPID (32.1%), study population: 883 borderline–profound ID who freely participated in the annual health examination, age > 30, mean 61.16 (SD 16.59; range 31–98) men 59%	Health screening by a local government county, Taiwan	Hypertension: elevated blood pressure	Annual health examination chart of 2010	Moderate (3)	High (0)
Van den Akker *et al.* (2006)	149, SPID (36.5%), study population: 436 mild–profound ID (for level ID data for 28 persons were missing), age range 0–70+, men 52%	A residential service, The Netherlands	Cardiac diseases	Electronic file with information about cardiac check-up	Moderate (2)	High (1)
Hove (2004)	?, SID (15.5%), study population: 228 mild–severe ID, age > 18 years, men 51%	Local services from twenty communities, Norway	Underweight BMI < 18.5 Overweight: BMI 25–29.9 Obesity: BMI ≥ 30	Questionnaire sent to health workers	Moderate (2)	High (0)
McGuire *et al.* (2010)	34 carers of SPID (21.7%), study population: 157 mild–profound ID, age mean 36.9 (SD 11.7; range 16–70), men 54%	Service users of an organisation, Ireland	Chronic pain: pain experienced most days for a minimum of 6 months	Questionnaires sent to the primary carers	Moderate (2)	High (0)

Table 1. *(Continued)*

Author (year of publication)	Number of participants relevant for review (percentage of research population), research population, age (years), gender	Setting + country	Health problem	Methods of diagnosis health problem	Risk of bias: internal validity (score)	Risk of bias: external validity (score)
Poppes *et al.* (2010)	181, PIMD, age mean 35 (SD 19; range 3–62), men 56%	Seven facilities, The Netherlands	Sensory problems, health problems	Questionnaire completed by the direct support professional	High (1)	Moderate (2)
Van der Heide *et al.* (2009)	254, PIMD, age median 49 (range 6–82), men 46%	Eight residential facilities, The Netherlands	Motor disabilities; sensory problems; health problems	Information collected from the medical notes and if necessary additional information from the physician or nurse	High (1)	Moderate (2)
Gittins & Rose (2008)	61, PMLD, age > 18, mean 37, men ?	A local health district, UK	Visual impairment, hearing impairment, epilepsy, dysphagia, respiratory problems	Information from case files notes	High (1)	Moderate (2)
Petry *et al.* (2009)	49, PMD, age mean 23.7 (SD 12.2; range 5–57), men 53%	A day centre and a residential care facility, The Netherlands and Belgium	Motor limitations, sensory limitations, medical condition, feeding problems	A questionnaire on characteristics of person with PMD completed by 'indirect support staff' (e.g. behavioural scientist, therapist)	High (1)	High (1)

problems were signified (Poppes *et al.* 2010; Petry *et al.* 2009; Van der Heide *et al.* 2009; Gittins & Rose 2008).

7 Measurement of health problem
No information regarding the reliability and validity of the measurement of the health problems that are specific for the target population of SPIMD was considered as a high risk of bias. This was the case in most of the studies ($n = 13$). Only objective measurements such as blood samples, ultrasound scanning of the bladder or clear criteria assessed by specialists were assessed as low risk of bias ($n = 7$).

8 Mode of data collection
In studies that investigated a range of physical health problems, there was no information on whether the same method was employed when collecting information on the subjects concerning specific physical health problems (Poppes *et al.* 2010; Petry *et al.* 2009; Van der Heide *et al.* 2009; Gittins & Rose 2008). Furthermore, in four studies, information on the mode of data collection was not evidenced (Hove 2004; Van den Akker *et al.* 2006; Lin *et al.* 2012; Hsu *et al.* 2012). In the remaining studies, the same mode of data collection or standardised methods of data collection were utilised.

9 Numerator(s) and denominator(s)
In all of the studies, appropriate numbers and prevalence percentages were provided.

Physical health problems

Table 2 lists, per ICD-10 chapter, the prevalence rate of the physical health problems ascertained in the studies.

The results are listed according to whether health problems are addressed in at least three studies, two studies or one study. CIs and risk of bias for IV and EV are provided for each health problem.

Health problems in three or more studies

A random effect meta-analysis was performed for epilepsy, pulmonary/respiratory problems, hearing problems, dysphagia, reflux disease and visual problems. All of the studies included people with SPIMD. Three studies included both adults and children (Poppes *et al.* 2010; Petry *et al.* 2009; Van der Heide *et al.* 2009).

Epilepsy, pulmonary/respiratory problems, hearing problems and dysphagia. The four studies included in the meta-analysis investigated a range of health problems (IV high, EV moderate to high; Poppes *et al.* 2010, Petry *et al.* 2009, Van der Heide *et al.* 2009, Gittins and Rose, 2008).

For *epilepsy,* the prevalence rate ranged from 64% to 80%. The results of the meta-analysis indicated a prevalence rate of 70% (CI 65–75%) with homogeneity among the observed prevalence across studies ($I^2 = 27\%$; $Q = 4.94$, $df = 3$, $P = 0.18$; Fig. 2).

The prevalence of *pulmonary/respiratory problems* ranged from 8% to 27%. The meta-analysis indicated a prevalence rate of 21% (CI 12–30%) with heterogeneity among the observed prevalence across studies ($I^2 = 84\%$; $Q = 17.90$, $df = 3$, $P = 0$; Fig. 3).

The prevalence rate of *hearing problems* varied between 8% and 32.9%. The meta-analysis indicated a prevalence rate of 21% (CI 6–36%) with heterogeneity among the observed prevalence across studies ($I^2 = 95\%$; $Q = 56.08$, $df = 3$, $P < 0.0001$; Fig. 4).

Three studies investigated d*ysphagia*, and the prevalence rates vary from 15% to 50%. The meta-analysis indicated a prevalence rate of 30% (CI 11–50%) with heterogeneity among the observed prevalence across studies ($I^2 = 92\%$; $Q = 24.87$, $df = 2$, $P < 0.0001$; Fig. 5).

Reflux disease. Reflux disease was investigated in three studies. One study included adults (Ohwada *et al.* 2006), and two studies included both adults and children (Petry *et al.* 2009; Van der Heide *et al.* 2009). Prevalence rates ranged between 2.6% and 24%. The meta-analysis indicated a prevalence rate of 16% (CI 2–29%). with heterogeneity among the observed prevalence across studies ($I^2 = 93\%$; $Q = 36.49$, $df = 2$, $P < 0.0001$; Fig. 6).

Visual problems. Prevalence rates of visual problems ranged from 32.7% to 92% in five studies. Four studies used comparable designs and based their findings on a questionnaire by proxy or medical

Table 2 Prevalence rate of the physical health problems per ICD-10 chapter

ICD-10 chapter	Prevalence rate (frequency/total) and physical health problem	Author
Blood and blood-forming organs and the immune mechanism (D50-D89)	41% (16/39) SMID and **anaemia**	Ohwada *et al.* 2006
	52.4% (43/82) SMID and high **vit. K-dependent hepatic marker**	Nagae *et al.* 2013
	30.5% (25/82) SMID and high **vit. K-dependent bone markers**	
Endocrine, nutritional and metabolic (E00-E90)	22.9% (48/210) SPID and **metabolic syndrome**	De Winter *et al.* 2011
	51.9% (109/210) SPID and **abdominal obesity**	
	23.3% (49/210) SPID and **low HDL cholesterol**	
	24.8% (52/210) SPID and **hypertriglyceridemia**	
	12.4% (26/210) SPID and **insulin resistance**	
	8% (7/87) SPID and **metabolic syndrome**	Hsu *et al.* 2012
	25.6% (10/39) SMID and **thyroid dysfunction**	Ohwada *et al.* 2006
	0% (0/39) SMID and **parathyroid dysfunction**	
	5% (2/39) SMID and **low serum albumin level**	Ohwada and Nakayama, 2008
Nervous system (G00-G99)	63% (161/254) PIMD and **spasticity**	Van der Heide *et al.* 2009*
	75% (37/49) PMD and **spasticity**	Petry *et al.* 2009*
	45% (22/49) PMD and **limitations in movement of upper limbs**	Petry *et al.* 2009*
	59% (29/49) PMD and **limitations in movement of lower limbs**	
	45% (59/131) SPID and **tardive dyskinesia**	Fodstad, 2010**
	30.1% (37/123) SPID and **tardive dyskinesia**	Matson, 2008**
	17.9% (22/123) SPID and **tardive dyskinesia and akathisia**	
	63.9% (39/61) PMLD and **epilepsy**	Gittins & Rose 2008
	71% (180/254) PIMD and **epilepsy**	Van der Heide *et al.* 2009*
	79% (39/49) PMD and **epilepsy**	Petry *et al.* 2009*
	66.3% (120/181) PIMD and **epilepsy**	Poppes *et al.* 2010*
Eye and adnexa (H00-H59)	62% (158/254) PIMD and **visual problems**	Van der Heide *et al.* 2009*
	32.7% (20/61) PMLD and **visual impairment**	Gittins & Rose 2008
	92% (68/74) SPMD and **visual impairment**	Van den Broek *et al.* 2006*
	(impaired visual acuity) 65% (48/74) range severe-blindness	
	54% (26/49) PMD and **visual impairments**	Petry *et al.* 2009*
	72.9% (132/181) PIMD and **visual problems**:	Poppes *et al.* 2010*
	10% (18/181) PIMD and **blind**	
	63% (114/181) PIMD and **weak-sighted**	
Ear and mastoid process (H60-H95)	29% (73/254) PIMD and **auditory problems**	Van der Heide *et al.* 2009*
	8.1% (5/61) PMLD and **hearing impairment**	Gittins & Rose 2008
	8% (4/49) PMD and **auditory impairments**	Petry *et al.* 2009*
	39.2% (71/181) PIMD and **auditory problems**	Poppes *et al.* 2010*
	5% (9/181) PIMD and **deaf**	
	22.7% (41/181) PIMD and **hard of hearing**	
	11.6% (21/181) PIMD and **hypersensitive**	
	24.6% (48/195 SPID and **deaf-blind**	Fellinger *et al.* 2009*
	2.6% (5/195 profound/severe hearing and visual impairment	
	8.2% (16/195) moderate hearing and profound/severe visual impairment	
	5.1% (10/195) profound/severe hearing and moderate visual impairment	
	8.7% (17/195) moderate hearing and visual impairment	
Circulatory system (I00-I99)	60% (126/210) SPID and **hypertension**	De Winter *et al.* 2011
	20.2% (54/267) SPID and **hypertension**	Lin *et al.* 2012
	9.4% (14/149) SPID had **cardiac diseases**	Van den Akker *et al.* 2006*
	15% (39/254) PIMD and **cardiovascular problems**	Van der Heide *et al.* 2009*
	6% (16/254) PIMD and **congenital heart disease**	
	9% (24/254) PIMD and **other cardiovascular problems**	
Respiratory system (J00-J99)	2.6% (1/39) SMID and **tonsillitis**	Ohwada *et al.* 2006
	0% (0/39) SMID and **sinusitis**	
	8.1% (5/61) PMLD and **respiratory problems**	Gittins & Rose 2008

Table 2. *(Continued)*

ICD-10 chapter	Prevalence rate (frequency/total) and physical health problem	Author
	26% (13/49) PMD and **problems with the bronchial tubes**	Petry *et al.* 2009*
	27.1% (49/181) PIMD and **pulmonary/ respiratory problems**	Poppes *et al.* 2010*
	23% (58/254) PIMD and **pulmonary problems**	Van der Heide *et al.* 2009*
Digestive system (K00-K93)	5.1% (2/39) SMID and **liver disease**	Ohwada *et al.* 2006
	26% (47/181) PIMD and **dental problems**	Poppes *et al.* 2010*
	44% (21 of 22/49) PMD and **constipation**	Petry *et al.* 2009*
	2.6% (1/39) SMID and **gastroesophageal reflux disease**	Ohwada *et al.* 2006
	5.1% (2/39) SMID and **gastrointestinal disease**	
	75.7% (137/181) PIMD and **bowel and abdominal problems**	Poppes *et al.* 2010*
	72% (183/254) PIMD and **gastrointestinal problems**	Van der Heide *et al.* 2009*
	24% (62/254) PIMD and **reflux**	
	60% (152/254) PIMD and **constipation**	
	15% (37/254) PIMD and **hiatus hernia**	
	14% (35/254) PIMD and **others gastrointestinal problems**	
	29.5% (18/61) PMLD and **dysphagia** (3/18 were using PEG feeds)	Gittins & Rose 2008
	76% (37/49) PMD and **feeding problems**	Petry *et al.* 2009*
	29% (14/49) PMD and **tube fed**	
	61% (30/49) PMD and **chewing difficulties**	
	50% (24/49) PMD and **swallowing difficulties**	
	21% (10/49) PMD and **reflux disease**	
	34% (87/254) PIMD and **feeding/drinking problems**	Van der Heide *et al.* 2009*
	13% (32/254) PIMD and **feeding tube**	
	15% (37/254) PIMD and **problems with swallowing**	
	9% (22/254) PIMD and **other feeding/ drinking problem**	
	5% (13/254) PIMD and **dental problems**	
Musculoskeletal system and connective tissue (M00-M99)	69% (34/49) PMD and **deformities**	Petry *et al.* 2009*
	20% (52/254) PIMD and **deformities/contractures**	Van der Heide *et al.* 2009*
	30% (77/254) PIMD and **scolioses**	
	19% (47/254) PIMD and **hip problems**	
	45.2% (254/562) SPID and **low bone mineral density** (BMD)	Lohiya *et al.* 2004
	17.4% (98/562) SPID and **osteoporotic**	
	27.8% (156/562) SPID and **osteopenic**	
	24.2% (47/194) SPID and **sarcopenia**	Bastiaanse *et al.* 2012
Genitourinary system (N00-N99)	0% (0/39) SMID and **renal disease**	Ohwada *et al.* 2006
	10% (5/49) PMD and **problems with the urinary tract system**	Petry *et al.* 2009*
	17% (43/254) PIMD and **urinary tract problems**	Van der Heide *et al.* 2009*
	9.4% (24/254) SPID and **post-void residual urine**	De Waal *et al.* 2009
	14% (8/56) PMD and **post-void residual urine**	
Symptoms, signs and abnormal clinical and laboratory findings, not elsewhere classified (R00-R99)	33.7% (61/181) PIMD and **tactile problems**	Poppes *et al.* 2010*
	23.2% (42/181) PIMD and **hypersensitive**	
	10% (18/181) PIMD and **undersensitive**	
	14.9% SID and **underweight**	Hove, 2004
	21.3% SID and **overweight**	
	10.6% SID and **obesity**	
	12.1% (4/33) SPID and mild level of **pain**	McGuire *et al.* 2010*
	26% (47/181) PIMD and **pain**	Poppes *et al.* 2010*
	17% (44/254) PIMD and **skin problems**	Van der Heide *et al.* 2009*

*Studies included adults and children

**Population with ID, diagnosed with an Axis I disorder and use of atypical antipsychotic medication

SMID, severe motor and intellectual disabilities; SPMD, severe and profound intellectual and motor disabilities; SPID, severe or profound intellectual disability; PMD, profound multiple disabilities; SID, severe intellectual disability; PIMD, profound intellectual disability and a profound or severe motor disability; PIMD, profound intellectual and multiple disabilities; PMLD, profound and multiple learning disabilities; PSMI, persons with severe motor and intellectual disabilities.

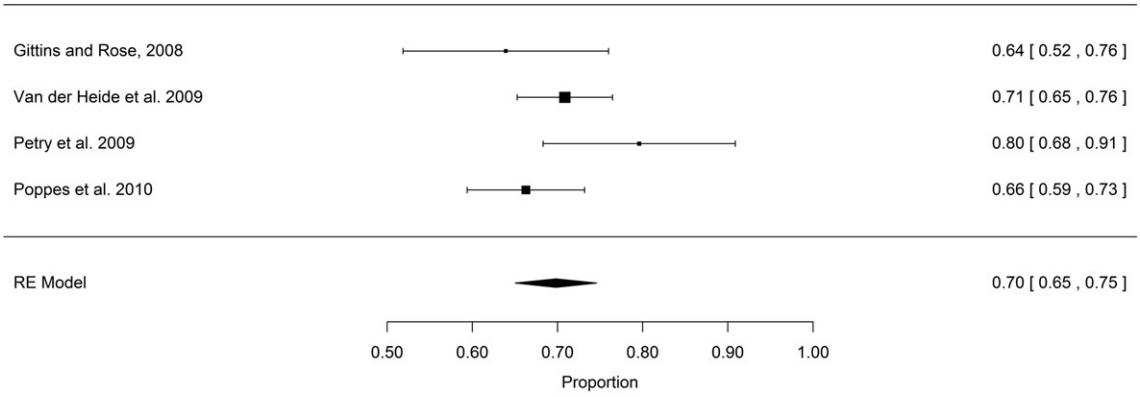

Figure 2 Forest plot: epilepsy, $N = 545$.

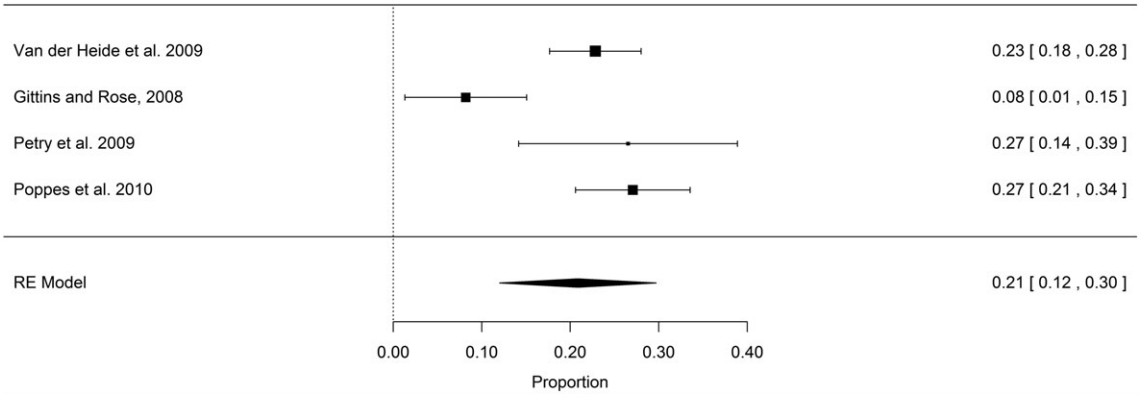

Figure 3 Forest plot: pulmonary/respiratory problems, $N = 545$.

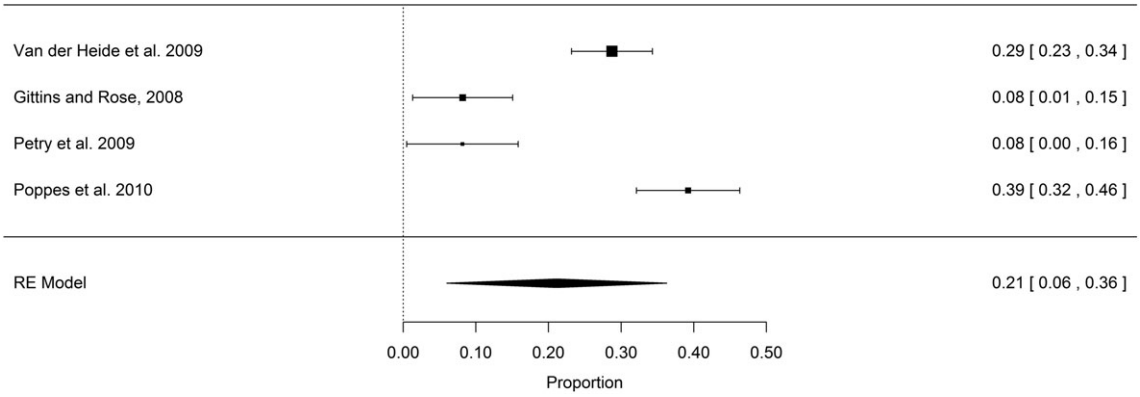

Figure 4 Forest plot: hearing problems, $N = 545$.

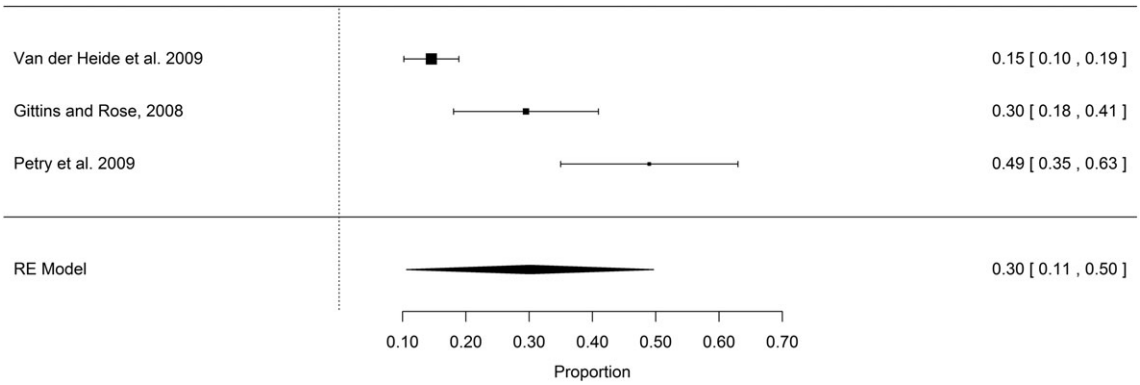

Figure 5 Forest plot: dysphagia, $N = 364$.

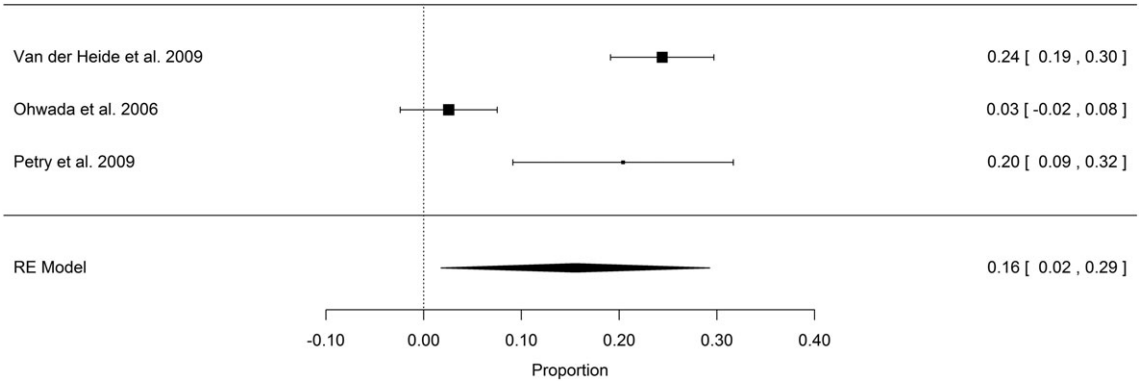

Figure 6 Forest plot: reflux disease, $N = 342$.

records. One study included adults (Gittins & Rose 2008), and three studies included both adults and children (Petry *et al.* 2009; Van der Heide *et al.* 2009; Poppes *et al.* 2010). The meta-analysis indicated a prevalence rate of 56% (CI 39–73%) with heterogeneity among the observed prevalence across studies ($I^2 = 93\%$; $Q = 36.14$, $df = 3$, $P < 0.0001$; Fig. 7).

The fifth study employed a different focus and design and, therefore, was not included in the meta-analysis. Van den Broek *et al.* (2006) ascertained a prevalence rate of 92% (CI 85.8–98.2%; IV low, EV moderate) based on visual screening by an orthoptist.

Health problems identified in two studies

Thirteen health problems were identified in two studies. The prevalence (CI, IV and EV) are described per health problem/per study and listed according to risk of bias for IV.

Tardive dyskinesia. A prevalence rates of 30.1% (CI 22–38.2%; IV low, EV moderate; Matson *et al.* 2008) and 45% (CI 36.5–53.5%; IV low, EV moderate; Fodstad *et al.* 2010) were reported for tardive dyskinesia in individuals with SPID who used antipsychotic medication.

Metabolic syndrome. Prevalence rates determined for metabolic syndrome were 8% (CI 2.3–13.7%; IV moderate, EV moderate; Hsu *et al.* 2012) and 22.9% (CI 17.2–28.6%; IV low, EV moderate; De Winter *et al.* 2011).

Hypertension. The prevalence rates of hypertension were 20.2% (CI 15.4–25%; IV moderate, EV high;

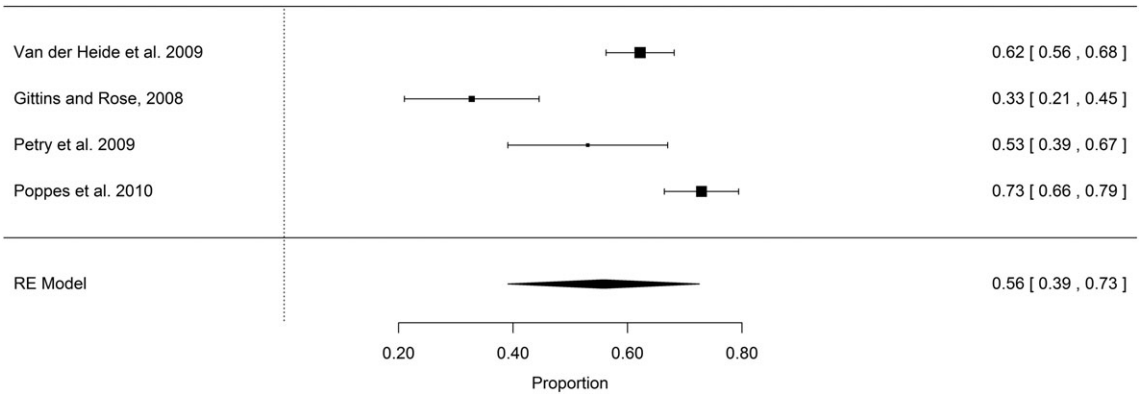

Figure 7 Forest plot: visual problems, $N = 545$.

Lin *et al.* 2012) and 60% (CI 53.4–66.6%; IV low, EV moderate; De Winter *et al.* 2011).

Cardiovascular problems. A prevalence of 9.4% was found in adults and children with SPID (CI 4.7–14.1%: IV moderate, EV high; Van den Akker *et al.* 2006) and 15% with cardiovascular problems in adults and children with SPIMD (CI 11–19.8%; IV high, EV moderate; Van der Heide *et al.* 2009).

Pain. The prevalence rates were 12.1% mild pain in adults and children with SPID (CI 1–23.2%; IV moderate, EV high; McGuire *et al.* 2010) and 26% pain in adults and children with SPIMD (CI 19.6–32.4%; IV high, EV moderate; Poppes *et al.* 2010).

Spasticity, constipation, feeding/drinking problems, feeding tube, deformities and urinary tract problems. Two studies investigated spasticity, constipation, feeding/drinking problems, feeding tube, deformities and urinary tract problems in adults and children (IV high EV moderate; Van der Heide *et al.* 2009, IV high, EV high; Petry *et al.* 2009).

Prevalence rates were *spasticity* 63% (CI 57.1–68.9%; Van der Heide *et al.* 2009) and 75% (CI 62.9–87.1%; Petry *et al.* 2009); *constipation* 44% (CI 30.1–57.9%; Petry *et al.* 2009) and 60% (CI 54–66%; Van der Heide *et al.* 2009); *feeding/drinking problems* 34% (CI 28.2–39.8%; Van der Heide *et al.* 2009) and 76% (CI 64–88%; Petry *et al.* 2009); *deformities* (including scoliosis and hip problems) 69% (CI 63.3–74.7%; Van der Heide *et al.* 2009) and 69% (CI 56.1–82%; Petry *et al.* 2009); *urinary tract problems* 10% (CI 1.6–18.4%; Petry *et al.* 2009) and 17% (CI 12.4–21.6%; Van der Heide *et al.* 2009); *feeding tube* 13% (CI 8.9–17.1%; Van der Heide *et al.* 2009) and 29% (CI 16.3–41.7%; Petry *et al.* 2009). There was no information regarding the type of feeding tube.

Dental problems. Prevalence rates for dental problems in adults and children varied from 5% (CI 2.3–7.7%; IV high, EV moderate; Van der Heide *et al.* 2009) to 26% (CI 19.6–32.4%; IV high, EV moderate; Poppes *et al.* 2010).

Gastrointestinal problems. Prevalence rates of gastrointestinal problems were 5.1% (CI −1.8–12%; IV high, EV moderate; Ohwada *et al.* 2006) and 72% in adults and children (CI 66.5–77.5%; IV high, EV moderate; Van der Heide *et al* 2009).

Health problems identified in one study

Sixteen health problems were identified in a single study. The prevalence of the health problems (CI, IV and EV) are described per study and listed according to risk of bias for IV.

Vitamin K deficiency. The vitamin K-dependent liver marker exceeded the upper normal range in 52% (CI 41.6–63.2%), and the vitamin K-dependent bone marker was above the upper reference in 30% (CI 20.5–40.5%; IV low, EV moderate; Nagae *et al.* 2013).

Low serum albumin. One study reported a prevalence rate of 5% of low serum albumin level (CI −1.8–11.8%; IV low, EV moderate; Ohwada & Nakayama 2008).

Post-void residual urine. The prevalence rates of post-void residual urine were 9.4% in people with SPID (CI 5.8–13%; IV low, EV high) and 14% for those with SPIMD (CI 4.9–23.1%; IV low, EV moderate; De Waal *et al*. 2009).

Deaf-blind. A prevalence rate of 24.6% of deaf/blindness in people was reported in adults and children (CI 18.6–30.6%; IV low, EV moderate; Fellinger *et al*. 2009).

Anaemia, thyroid dysfunction, tonsillitis and liver disease. A prevalence rate of 41% was found for *anaemia* (CI 25.6–56.4%, IV low, EV moderate).

Furthermore, Ohwada *et al.* (2006) reported thyroid dysfunction 25.6% (CI 11.9–39.3%), tonsillitis 2.6% (CI −2.4–7.4%) and liver disease 5.1% (CI −1.8–11.8%) (IV high, EV moderate).

Sarcopenia. A prevalence of 24.2% of sarcopenia was reported (CI 18.2–30.2%; IV low, EV high; Bastiaanse *et al*. 2012).

Osteoporosis. A prevalence of 45.2% of low bone mineral density was found (CI 41.1–49.3%; IV low, EV high; Lohiya *et al*. 2004).

Underweight, overweight and obesity. The prevalence rates on weight were 14.9% underweight, 21.3% overweight and 10.6% obesity (IV moderate, EV high; Hove 2004). The number of persons with severe ID was unclear; therefore, no confidence interval could be calculated.

Bowel/abdominal problems and tactile problems. For bowel and abdominal problems, a prevalence rate was determined of 75.7% (CI 69.5–82%), and for tactile problems, 33.7% (CI 26.8–40.6%) was reported in adults and children (IV high, EV moderate; Poppes *et al*, 2010).

Hiatus hernia and skin problems. A prevalence rate of 15% hiatus hernia (CI 10.3–18.9%) and 17% for skin problems (CI 12.4–21.6%) was reported in adults and children (IV high, EV moderate; Van der Heide *et al*. 2009).

Chewing difficulties. A prevalence rate of 61% chewing difficulties was reported in adults and children (CI 47.3–74.7%; IV high, EV high; Petry *et al*. 2009).

Discussion

The aim of this study was to systematically review cross-sectional studies on the prevalence of various types of physical health problems in adults with SPIMD. Our systematic review identified 35 physical health problems in 20 different studies.

For six health problems, a meta-analysis was conducted. A homogeneous prevalence rate was ascertained for epilepsy 70% (CI 65–75%). For the remaining five health problems, heterogeneous results were found, with a significant degree of inconsistency between the results of the studies which were based on case file notes or questionnaire by proxy and, therefore, these prevalence estimates are likely to be an underestimation of the true prevalence (Haveman 2004).

The results of the meta-analysis of visual problems indicated a prevalence rate of 56% (CI 39–73%). However, Van den Broek *et al.* (2006) determined a prevalence rate of 92% (CI 85.8–98.2%) in people with SPIMD based on visual screening by an orthoptist, which confirms that problems with sensory functions are easily overlooked, and health screenings can identify previously unrecognised health problems (Felce *et al.* 2009; Felce *et al.* 2008; Robertson *et al.* 2014; Kerr *et al.* 2003).

For the health problems identified in two studies or in a single study, the degree of evidence is minimal. There were three important variabilities between the studies: (1) diversity in the definition of a health problem, for example, metabolic syndrome (De Winter *et al.* 2011; Hsu *et al.* 2012), hypertension (De Winter *et al.* 2011; Lin *et al.* 2012) and feeding problems (Petry *et al.* 2009; Van der Heide *et al.* 2009); (2) lack of definition of a health problem, for example, feeding/drinking problems and urinary tract problems (Petry *et al.* 2009; Van der Heide *et al.* 2009); (3) differences in the study population, e.g. differences in mean age (De Winter *et al.* 2011; Hsu *et al.* 2012) and differences in sample selection (De Winter *et al.* 2011; Lin *et al.* 2012).

As expected, higher prevalence rates were found in our review compared with people with ID for a cluster of health problems that are associated with brain damage/dysfunction. Reported prevalence rates in individuals with ID were visual problems 19.2% (Van Splunder *et al.* 2006) against 56% and 92% in the present review; epilepsy 22.2% (Robertson *et al.* 2015) against 70% in the present review; and spasticity 14.6% (Maaskant *et al.* 1994) against 63% and 75% in the present review. Unexpected lower prevalence rates were compared with people with ID ascertained for constipation 70% (Böhmer *et al.* 2001) against 44% and 60% in the present review; dysphagia 33% (Rogers *et al.* 1994) against 30% in the present review; reflux disease 48% (Böhmer *et al.* 1999) against 16% in the present review; and hearing problems 30% (Meuwese-Jongejeugd *et al.* 2006) against 21% in the present review. This is of concern because undiagnosed and untreated dysphagia and reflux disease can lead to recurrent respiratory tract infections, which is a possible leading cause of death for people with ID (Emerson & Baines 2010).

This systematic review is restricted to recent publications because increase in the quality of support combined with advances in medical care may possibly influence the detection rate of physical health problems. As a consequence, relevant studies published prior to 2004 were probably missed. A strength of this study is the thorough search strategy in six databases with the utilisation of a broad range of search terms. For a comprehensive overview, studies were included in which the physical health problems were not the primary focus but were collected as characteristics of individuals with SPIMD. Furthermore, studies regarding both children and adults were included. This may affect the type and prevalence of the reported health problems. For example, long-term use of medication may lead to different physical health problems during adulthood. In addition, studies that focused on people with SPIMD used slightly different terms to describe the target population. However, as the alternative was a less complete overview of the available literature, we considered this the most appropriate inclusion method. Another strength of this study is the detailed critical appraisal of the studies assessing a risk of bias for IV and EV. At the time that this systematic review was conducted, there was no standard method for critical appraisal of studies of prevalence data; therefore, an existing risk of bias checklist was modified. Recently, a new tool was developed specifically for studies reporting prevalence data (Munn *et al.* 2014).

It is believed that the current review is the first that provides an overview of the current state of the art research on the prevalence of health problems in adults with SPIMD. Only eight studies included a study population representative for SPIMD, and not all of the studies focused primarily on physical health issues. The included studies were diverse, and most studies focused on a single or a minimal number of somatic problems rather than on the entire range that were experienced. There is a strong need for comprehensive epidemiological data in order to determine clusters of health problems that are specific for people with SPIMD. This would provide insight into the excess morbidity associated with SPIMD.

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Accepted 8 April 2016

Appendix 1: Search string Database MedLine/ PubMed

((("Williams Syndrome"[Mesh Terms] OR "Prader-Willi Syndrome"[MeSH Terms] OR "Fragile X Syndrome"[MeSH Terms] OR "Cerebral Palsy"[MeSH Terms] OR "Rett Syndrome"[MeSH Terms] OR "De Lange Syndrome"[MeSH Terms] OR "Rubinstein-Taybi Syndrome"[MeSH Terms] OR "Angelman Syndrome"[MeSH Terms] OR "Smith-Lemli-Opitz Syndrome"[MeSH Terms] OR "Orofaciodigital Syndromes"[MeSH Terms] OR "Smith-Magenis Syndrome"[MeSH Terms] OR "Mental Retardation, X-Linked"[MeSH Terms] OR "intellectual disability"[MeSH Terms] OR "mentally disabled persons"[MeSH Terms] OR "developmental disabilities"[MeSH Terms] OR "Williams Syndrome"[Title/Abstract] OR "Prader-Willi Syndrome"[Title/Abstract] OR "Fragile X

Syndrome"[Title/Abstract] OR "Cerebral Palsy"[Title/Abstract] OR "Rett Syndrome"[Title/Abstract] OR "De Lange Syndrome"[Title/Abstract] OR "Kabuki syndrome" [Title/Abstract] OR "Rubinstein-Taybi Syndrome"[Title/Abstract] OR "Angelman Syndrome"[Title/Abstract] OR "Smith-Lemli-Opitz Syndrome"[Title/Abstract] OR "Orofaciodigital Syndromes"[Title Abstract] OR "Taybi Syndrome"[Title/Abstract] OR "Smith-Magenis Syndrome"[Title/Abstract] OR "Cohen syndrome"[Title/Abstract] OR "X-Linked Mental Retardation"[Title/Abstract] OR "X-Linked Mental Retardations"[Title/Abstract] OR "Profound multiple disabilities"[Title/Abstract] OR "Severe multiple disabilities"[Title/Abstract] OR "intellectual disability"[Title/Abstract] OR "intellectual disabilities"[Title/Abstract] OR "intellectually disabled"[Title/Abstract] OR "intellectual disorder"[Title/Abstract] OR "intellectual disorders"[Title/Abstract] OR "intellectual impairment"[Title/Abstract] OR "intellectual impairments"[Title/Abstract] OR "intellectually impaired"[Title/Abstract] OR "intellectual deficiency"[Title/Abstract] OR "intellectual deficiencies"[Title/Abstract] OR "intellectually deficient"[Title/Abstract] OR "intellectual retardation"[Title/Abstract] OR "intellectual retardations"[Title/Abstract] OR "intellectually retarded"[Title/Abstract] OR "intellectual handicap"[Title/Abstract] OR "intellectual handicaps"[Title/Abstract] OR "intellectually handicapped"[Title/Abstract] OR "mental deficit"[Title/Abstract] OR "mental deficits"[Title/Abstract] OR "mental deficiency"[Title/Abstract] OR "mental deficiencies"[Title/Abstract] OR "mentally deficient"[Title/Abstract] OR "mental retardation"[Title/Abstract] OR "mental retardations"[Title/Abstract] OR "mentally retarded"[Title/Abstract] OR "mental incapacity"[Title/Abstract] OR "mental incapacities"[Title/Abstract] OR "mentally incapacitated"[Title/Abstract] OR "mental handicap"[Title/Abstract] OR "mental handicaps"[Title/Abstract] OR "mentally handicapped"[Title/Abstract] OR "mental disability"[Title/Abstract] OR "mental disabilities"[Title/Abstract] OR "mentally disabled"[Title/Abstract] OR idiocy[Title/Abstract] OR "cognitive disability"[Title/Abstract] OR "cognitive disabilities"[Title/Abstract] OR "cognitively disabled"[Title/Abstract] OR "cognitive retardation"[Title/Abstract] OR "cognitive retardations"[Title/Abstract] OR "cognitively retarded"[Title/Abstract] OR "developmental disability"[Title/Abstract] OR "developmental disabilities"[Title/Abstract] OR "developmentally disabled"[Title/Abstract] OR "developmental disorder"[Title/Abstract] OR "developmental disorders"[Title/Abstract] OR "multiple disabilities"[Title/Abstract] OR "multiple handicaps"[Title/Abstract]) AND ("severe"[Title/Abstract] OR "profound"[Title/Abstract]) AND ("Comorbidity"[MeSH Terms] OR "risk factors"[MeSH Terms] OR comorbidity[Title/Abstract] OR comorbidities[Title/Abstract] OR comorbid[Title/Abstract] OR "ill health"[Title/Abstract] OR "health problem"[Title/Abstract] OR "health problems"[Title/Abstract] OR "health indicator"[Title/Abstract] OR "health indicators"[Title/Abstract] OR "health issue"[Title/Abstract] OR "health issues"[Title/Abstract] OR "secondary condition"[Title/Abstract] OR "secondary conditions"[Title/Abstract] OR somatic[Title/Abstract] OR "physical health"[Title/Abstract] OR "risk factor"[Title/Abstract] OR "risk factors"[Title/Abstract] OR "medical condition"[Title/Abstract]))