The neglected puzzle of dementia in people with severe/profound intellectual disabilities: A systematic literature review of observable symptoms

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Abstract

Background: Dementia is increasingly prevalent in people with severe/profound intellectual disabilities. However, early detection and diagnosis of dementia is complex in this population. This study aimed to identify observable dementia symptoms in adults with severe/profound intellectual disabilities in available literature.

Method: A systematic literature search was conducted in PubMed, PsycINFO and Web of Science with an exhaustive search string using a combination of search terms for severe/profound intellectual disabilities and dementia/ageing.

Results: Eleven studies met inclusion criteria. Cognitive decline, behavioural and psychological alterations, decline in activities of daily living as well as neurological and physical changes were found.

Conclusions: Only a very limited number of studies reported symptoms ascribed to dementia in adults with severe/profound intellectual disabilities. Given the complexity of signalling and diagnosing dementia, dedicated studies are required to unravel the natural history of dementia in this population.

KEYWORDS
ageing, dementia, Down syndrome, intellectual disabilities, severe or profound intellectual (and multiple) disabilities
1 | INTRODUCTION

The world’s population, including people with intellectual disabilities, is ageing. Life expectancy of people with intellectual disabilities has increased even more than in the general population (Coppus, 2013; Evans et al., 2013). Overall, life expectancy of this population is comparable to that of the general population, except for shorter life expectancy for people with more severe intellectual and multiple disabilities and people with Down syndrome (approximately 60 years; Bittles et al., 2007; Coppus, 2013). Given the fact that age is the major risk factor for dementia (Alzheimer’s Association, 2021), dementia is increasingly prevalent among the population of people with disabilities. In particular, people with Down syndrome (trisomy 21) have a high genetic risk of developing Alzheimer’s disease dementia: up to 77% will have developed dementia by the age between 60 and 69 years (Ballard et al., 2016). Moreover, prevalence rates of dementia for people with intellectual disabilities not due to Down syndrome vary across studies (Kinsky-McHale & Silverman, 2013).

A pre-existing intellectual disability, (lifelong) characteristic behaviour and co-morbidities which may mimic dementia symptoms are complicating factors in diagnosing dementia in people with intellectual disabilities (Esbensen et al., 2017; McKenzie et al., 2018; Sheehan et al., 2015). In fact, the more severe the level of intellectual disability, the more difficult diagnosing dementia becomes (Evans et al., 2013). Hence, diagnosing dementia is especially challenging in people with severe/profound intellectual disabilities, that is, Intelligence Quotient score below 35 (Evans et al., 2013; McKenzie et al., 2018). Firstly, their low cognitive baseline functioning makes it difficult to establish a decline in cognitive functioning from a previous higher level (Evans et al., 2013). Secondly, to show measurable changes in cognitive functioning using direct neuropsychological tests is virtually impossible due to floor effects (Elliott-King et al., 2016). Thirdly, observing a decline in functioning is highly complex because individuals with severe/profound intellectual disabilities have often multiple health conditions, that is, multimorbidity (Hermans & Evenhuis, 2014; Kinnear et al., 2018; Van Timmeren et al., 2017).

Fourthly, they need high levels of support to perform activities of daily living, because specific skills were not attained (Sheehan et al., 2015). Consequently, skills which have never been developed cannot alter and therefore not serve as symptoms indicative of dementia. Lastly, diagnosing dementia in this population is even more complicated because communication is mostly non-verbal, thus without self-reported complaints (Smiley & Cooper, 2003). Hence, those with severe/profound intellectual disabilities are largely reliant on caregivers/relatives for signalling observable dementia symptoms (McKenzie et al., 2018).

Evidently, diagnosing dementia in people with severe/profound intellectual disabilities is a complex puzzle, which necessitates a proper understanding of its presentation in this population. Early detection and diagnosis of dementia allows caregivers/professionals and relatives to make informed choices about adaptation of caregiving, support and treatment (Dekker, Wissing et al., 2021). However, caregivers indicate to have limited knowledge on the presentation of dementia in people with intellectual disabilities (Herron et al., 2015; Whitehouse et al., 2000). Limited knowledge about symptoms may cause early signs not be recognised, resulting in a (too) late diagnosis or no diagnosis at all (Cleary & Doody, 2017). Moreover, if dementia was diagnosed there was a gap in intellectual disability caregivers’ knowledge about the course of dementia (Furniss et al., 2011; Iacono et al., 2014). They struggled to understand whether changes were dementia symptoms or related to the intellectual disability (Iacono et al., 2014). Overall, a better understanding about (early) dementia symptoms, especially in those with severe/profound intellectual disabilities is essential to provide appropriate support and care in order to maintain quality of life (Janicki, 2011; Dekker, Wissing et al., 2021).

Improving the diagnostic procedure in this population starts with understanding the natural history of dementia. Hence, this study reviews literature to identify observable dementia symptoms in adults with severe/profound intellectual disabilities. Given the diagnostic complexity, it is expected that dementia is often underdiagnosed. Therefore, we also reviewed ageing literature describing changes in cognitive functioning and/or behavioural and psychological alterations without explicitly referring to dementia.

2 | METHODS

This systematic literature review largely followed PRISMA criteria (Moher et al., 2009). All criteria were followed with the exception of a risk of bias assessment, since our core aim was to identify observable dementia symptoms in those with severe/profound intellectual disabilities in the scarce literature.

2.1 | Search strategy

In December 2020, a systematic literature search without any time period restrictions was performed in PubMed, PsycINFO and Web of Science. The search strategy involved three key search term clusters. The first cluster included search terms for severe/profound intellectual disabilities, using a broad range of synonyms for intellectual disabilities as well as older (sometimes abandoned) terminology to ensure that relevant studies using past terminology were obtained as well. Given that no specific indexed terms exist for severe/profound intellectual disabilities in the databases, all search terms were searched in fivefold (preceded by the adjectives Complex, Multiple, Profound, Serious or Severe) to discard articles not focusing on severe/profound intellectual disabilities. The second cluster included search terms for dementia, for example, Alzheimer, major/minor cognitive impairment as well as terms related to ageing, such as decline, changes, progressive, deterioration. Subsequently, the third cluster ensured that only results for an aged population were obtained, removing large numbers of irrelevant studies in children, adolescents, young adults and animals. These three clusters were combined using the Boolean operator ‘AND’, whereas ‘OR’.

In all three clusters,
truncation (*) accounted for different forms of words. Terms were searched in title and abstract (Table A1).

2.2 Study selection

To be included, studies had to describe (potential) dementia symptoms in people with severe/profound intellectual disabilities aged 30 years and over. If studies focused on a broader spectrum of intellectual disability levels, (potential) dementia symptoms had to be separately reported for people with severe/profound intellectual disabilities. Exclusion criteria: studies in the general population (without intellectual disabilities), people with mild or moderate intellectual disabilities, age under 30 years, studies focusing on persons with severe/profound intellectual disabilities caused by rare (genetic) disorders (i.e., fewer than 5 in 10,000 people; Nguengang Wakap et al., 2020), non-original research articles (e.g., reviews), animal studies and non-English articles.

All records obtained in the three databases were deduplicated using RefWorks bibliographic management software (ProQuest). Titles and abstracts were subsequently screened for eligibility (A.M.U.). A randomly selected 15% of the deduplicated records were screened by a second author (M.B.G.W.). Afterwards, two authors (A.M.U. and M.B.G.W.) independently determined eligibility of the selected articles by checking full-texts. Disagreements were resolved by consensus discussions, if necessary consulting a third author (A.D.D.). Lastly, reference lists of included studies were checked for additional articles. Figure 1 schematically depicts the selection procedure.

2.3 Data extraction and synthesis

Two authors (A.M.U. and M.B.G.W.) independently extracted relevant data from the selected full-text studies, namely: study population(s), intellectual disability, assessment of dementia/age-related changes that are potential dementia symptoms, main symptomatic results (Table 1). Discrepancies were resolved with discussions between the two authors. Additionally, two authors separately determined the limitations of the primary studies. To diagnose dementia, different sets of criteria are currently being used worldwide (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2018). Despite (minor) differences, these sets of criteria all include, in one way or another, a decline in cognitive functioning which interferes with the ability to perform activities of daily living, accompanied by behavioural and psychological symptoms of dementia (BPSD), (Dekker et al., 2015; Finkel, 2000). Therefore, in ageing studies only reported cognitive changes and behavioural and psychological alterations were considered to be potential dementia symptoms. In the present study, (potential) dementia symptoms were categorised according to the three diagnostic criteria domains. Additional findings reported in dementia studies were grouped in the category neurological and other physical changes (Nieuwenhuis-Mark, 2009; Strydom et al., 2010; Table 2).

3 RESULTS

The literature search yielded a total of 668 hits in three databases. Deduplication resulted in 625 unique records (Figure 1). Based on title
<table>
<thead>
<tr>
<th>References</th>
<th>Study population(s)</th>
<th>ID classification</th>
<th>Assessment of dementia/age-related changes that are potential dementia symptoms</th>
<th>Main symptomatic results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reid and Aungle (1974)</td>
<td>155 ID, 63 ♂, 49–85 yrs - 1 severe ID (DS) + dementia, ♂, 52 yrs - 1 severe ID + dementia, ♂, 62 yrs</td>
<td>GMD</td>
<td>Clinical (re)assessment: - Case files - Informant interviews - Physical examination</td>
<td>activity, speech, self-care skills, personality change, disturbed sleep, incontinence, late-onset epilepsy (Case 1)</td>
</tr>
<tr>
<td>Day (1985)</td>
<td>357 ID, ≥ 40 yrs - 2 severe ID + late-onset dementia</td>
<td>ICD-9</td>
<td>- Case files</td>
<td>social skills, personal habits, behavioral disturbances, memory impairments: forgetfulness, confusion</td>
</tr>
<tr>
<td>Evenhuis (1990)</td>
<td>17 DS, 7 ♂, 45–63 yrs - 5 severe ID (DS) + AD, 2 ♂, 45–60 yrs</td>
<td>Severe ID: IQ 25–5</td>
<td>Non-standardised clinical assessment: - Observations - Informant interviews - Physical examination</td>
<td>self-help skills, gait, apathy/withdrawal, epileptic seizures, chair/bedridden (n = 5) urinary incontinence, daytime sleepiness, myoclonus (n = 4) disturbed night sleep, muscle hypertonia (n = 3) apraxia, irritability/aggression (n = 2) speech (n = 1)</td>
</tr>
<tr>
<td>Duggan et al. (1996)</td>
<td>12 ID + dementia, 3 ♂, 47–77 yrs - 1 severe ID (DS), ♂, 59 yrs</td>
<td>ICD-9</td>
<td>- Psychiatric and medical files - Informant interviews (BHI) - Physical examination</td>
<td>cognitive functioning, memory, everyday functioning, emotional/behavioural changes</td>
</tr>
<tr>
<td>Burt et al. (1998)</td>
<td>70 DS, 22–60 yrs - 2 severe ID (DS) + dementia - 3 profound ID (DS) + dementia</td>
<td>Severe: IQ 21–36</td>
<td>Profound: no basal on LIPS - Direct neuropsychological evaluation (DF &amp; SR, GPT, DTVMI, PD, PPVT-R, LIPS) - Informant interviews (VABS, DQMRP, RSMB, DSI, MAS)</td>
<td></td>
</tr>
<tr>
<td>Määttä et al. (2006)</td>
<td>129 DS, 76 ♂, 0–67 yrs - 1 moderate/severe ID (DS) + AD, ♂, 51 yrs</td>
<td>ICD-10</td>
<td>- Case files</td>
<td>self-care skills, forgetfulness, irritability, withdrawal, occasional aggressive outburst, late-onset epilepsy</td>
</tr>
<tr>
<td>Margallo-Lana et al. (2007)</td>
<td>92 DS, 63 ♂, 20–76 yrs - 6 profound ID (DS) + dementia</td>
<td>Unspecified</td>
<td>- Informant interviews - Medical files</td>
<td>Non-cognitive symptoms: everyday skills, mobility, interest in surroundings, uncharacteristic inappropriate behaviour, daytime sleepiness, wandering, getting lost, incontinence</td>
</tr>
<tr>
<td>Sauna-Aho et al. (2018)</td>
<td>128 DS, WS, FxS, 85 ♂, 36–85 yrs - 2 profound ID + dementia, 50 yrs (DS), 61 yrs (FxS)</td>
<td>Medical files</td>
<td>- Medical files - BPSLD - Brain imaging</td>
<td>Weight change, loss of energy, sleep disorder</td>
</tr>
<tr>
<td>References</td>
<td>Study population(s)</td>
<td>ID classification</td>
<td>Assessment of dementia/age-related changes that are potential dementia symptoms</td>
<td>Main symptomatic results</td>
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</tbody>
</table>
| Studies of potential dementia symptoms | Haveman et al. (1994) | 1580 ID, 0–60+ yrs  
- 209 severe ID (DS)  
- 477 severe ID (non-DS) | Unspecified | GQ:  
- Psychological functioning  
- Challenging behaviour |
|                             |                     |                   | Non-DS severe ID:  
= psychological problems, ↓ challenging behaviour  
DS severe ID:  
↑ psychological problems, = challenging behaviour |
| Cherry et al. (1997)        | 168 SPID  
- 84 young, 46 ♂, 20–29 yrs  
- 84 elderly, 45 ♂, 60–79 yrs | AAMD | DASH | Compared to younger SPID, older SPID showed:  
↓ durations anxiety, inappropriate sexual behaviour, impulse control problems  
↑ severity anxiety, stereotypies/tics, impulse control problems  
Result in relation to ID level:  
↓ duration self-injury behaviour in elderly with profound ID |
| Rousseau et al. (2019)      | 474 profound ID and severe motor deficiency  
- 219 young, 1.2 ♂/♀, 18–34 yrs  
- 151 middle-aged, 1 ♂/♀, 35–49 yrs  
- 104 elderly, 1.4 ♂/♀, 50–68 yrs | Medical files | Medical files | ↓ withdrawal, ↑ intermittent screaming, ↑ intermittent crying, ↑ agitation, ↑ self-aggressivity, ↓ language, ↓ posture-motor ability, ↓ coordination, ↓ sociability, = aggressivity, = stereotypies, = mercycsm, = sleep disorders |

Note: ↓, decrease; ↑, increase; =, remained stable; ♂, male; ♀, female.

Abbreviations: AAMD, American Association on Mental Deficiency; AD, Alzheimer’s disease; DASH, Diagnostic Assessment for the Severely Handicapped; BHI, Behaviour History Inventory; BPSLD, British Present Psychiatric State-Learning Disabilities assessment; DF & SR, Digits Forward and Sentence Recall subscales; DS, Down syndrome; DSI, Depression Status Inventory; DTVMI, Developmental Test of Visual-Motor Integration; DQMRP, Dementia Questionnaire for Mentally Retarded Persons; FxS, Fragile X syndrome; GMD, Glossary of Mental Disorder; GPT, Grooved Pegboard Test; GQ, Gerontological Questionnaire subscales; ICD, International Classification of Diseases and Related Health Problems; ID, intellectual disabilities; IQ, Intelligence Quotient; LIPS, Leiter International Performance Scale; MAS, Mood Assessment Scale for MR; PD, Picture Discription test; PPVT-R, Peabody Picture Vocabulary Test-Revised; RSMB, Reiss Screen for Maladaptive Behaviour; SPID, severe/profound intellectual disabilities; VABS, Vineland Adaptive Behaviour Scale, WS, Williams syndrome; yrs, years.
and abstract 141 records were considered potentially relevant for this review. Inter-rater agreement of screening a randomly selected 15% of the deduplicated records was 96.7%. The 141 articles were read full-text, 9 studies satisfied all criteria and were subsequently included. Inter-rater agreement of this full-text screening process was 96.2%. Two additional articles were identified by screening reference lists of included articles (Figure 1). In total, 11 studies met inclusion criteria. Table 1 provides a detailed overview of characteristics and main results.

### 3.1 Symptoms in dementia studies

The first study to report dementia symptoms in adults with severe/profound intellectual disabilities was published by Reid and Aungle (1974). Among 155 persons with intellectual disabilities, two individuals with a severe intellectual disability were diagnosed with dementia based on clinical (re)assessment (Table 1). For a 52-year-old man with Down syndrome, reported dementia symptoms consisted of a personality change, disturbed sleep, diminution of activity, reduction of speech and a deterioration in self-care skills. Incontinence and late-onset epilepsy were also reported. The second case concerned an ongoing dementia process in a 65-year-old woman with a severe intellectual disability of unknown aetiology. Dementia began in her middle 50s and signs were slowly progressive loss of self-care skills, reduction of speech, a personality change and asymmetrical spasticity in the limbs.

Subsequently, Day (1985) reported the prevalence of dementia in 357 people with intellectual disabilities. A diagnosis of late-onset dementia was recorded in case files of nine individuals, including two persons with a severe intellectual disability. Both individuals had memory impairments presented as forgetfulness and confusion as well as loss of social skills and deterioration in personal habits. Furthermore, they exhibited behavioural disturbances, which were not further specified for these two persons.

Evenhuis (1990) conducted a prospective longitudinal study in seventeen individuals with Down syndrome of whom five had a severe intellectual disability. Dementia was suspected in these five individuals based on progressive decline in activities of daily living. However, it was not possible to formally diagnose dementia because memory and orientation could not be evaluated. Decreased self-help skills were observed in these five individuals as well. In one person, a reduction of speech was seen. In the other four persons speech had hardly developed, and was consequently not indicative of dementia.

### Table 2 Overview of (potential) dementia symptoms in people with severe/profound intellectual disabilities

<table>
<thead>
<tr>
<th>Categories</th>
<th>Dementia symptoms</th>
<th>Potential dementia symptoms</th>
</tr>
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<tbody>
<tr>
<td>Cognitive changes</td>
<td>↓ speech1,2,3, ↓ social skills2, ↓ cognitive functioning5, ↓ memory2,3, forgetfulness2,6, confusion2, aimless walking4, wandering5, getting lost7, ↓ personal habits2, apraxia3, inappropriate food placing4, wrong utensils use3</td>
<td>↓ language11, ↓ sociability11, ↓ posture-motor11, ↓ coordination11</td>
</tr>
<tr>
<td>Behavioural and psychological changes</td>
<td>aggression3,6, withdrawal3,6, apathy3, ↓ interest in surroundings1, irritability3,6, daytime sleepiness3,7, disturbed sleep1, sleep disorder1, ↓ food intake6, ↓ drinking4, pica4, uncharacteristic inappropriate behaviour7, hypermetamorphosis4, personality change1, emotional/behavioural changes9</td>
<td>↑ self-aggressivity11, ↑ duration self-injury behaviour (PID)10, ↑ withdrawal11, ↑ agitation11, ↑ intermittent screaming11, ↑ intermittent crying11, ↑ durations anxiety10, ↑ durations inappropriate sexual behaviour10, ↑ durations impulse control problems10, ↑ severity anxiety10, ↑ severity stereotypies/tics10, ↑ severity impulse control problems10, ↑ psychological problems (DS)9</td>
</tr>
<tr>
<td>Changes in activities of daily living</td>
<td>↓ self-care skills1,3,6, ↓ everyday functioning/skills1,3,7, ↓ activity1</td>
<td></td>
</tr>
<tr>
<td>Neurological and other physical changes</td>
<td>incontinence1,3,7, (late-onset) epilepsy1,3,6, weight change/loss4,5, ↓ energy4, ↓ amount of walking4, ↓ mobility1, ↓ gait1, chair/bedridden3, asymmetrical spastic signs in limbs1, muscle hypertonia1, myoclonus5</td>
<td></td>
</tr>
</tbody>
</table>

Note: ↓, decrease; ↑, increase; =, remained stable. References: 1, (Reid & Aungle, 1974); 2, (Day, 1985); 3, (Evenhuis, 1990); 4, (Duggan et al., 1996); 5, (Burt et al., 1998); 6, (Määttä et al., 2006); 7, (Margallo-Lana et al., 2007); 8, (Sauna-Aho et al., 2018); 9, (Haveman et al., 1994); 10, (Cherry et al., 1997); 11, (Rousseau et al., 2019). Abbreviations: DS, Down syndrome; non-DS, without Down syndrome; PID, profound intellectual disabilities.
(Table 1). Two persons had developed apraxia, whereas apraxia was not assessed in the other three persons. Additionally, apathy, for example, social withdrawal, was observed in all five participants. Furthermore, reported behavioural changes were daytime sleepiness (n = 4), disturbed sleep (n = 3) and irritability/aggression (n = 2). Resembling Reid and Aungle (1974) late-onset of epileptic seizures (n = 5), myoclonus (n = 4) and incontinence (n = 4) were also reported. Additionally, all five individuals developed muscle hypertonia and presented gait deterioration. Over the course of dementia they became chair ridden or bedridden. Postmortem neuropathological examination confirmed Alzheimer’s disease dementia in these five persons.

Duggan et al. (1996) described behavioural changes in a population with intellectual disabilities and dementia. Among the twelve individuals, one had a severe intellectual disability and was diagnosed with dementia. Based on an informant interview using the Past Behavioural History Inventory, this 59-year-old woman with Down syndrome was described as showing changes in walking behaviour, specifically aimless walking in the last eight months and a noticeable decrease in the amount of walking over the last eighteen months. Additional symptoms were weight loss, decreased food intake since eight months, inappropriate placing of food for eighteen months, wrong use of utensils, pica in the last six months and a decrease in the drinking amount over the last eight months. Lastly, she exhibited hypermetamorphosis which manifested as a compulsive touch to furniture (Table 1).

Similar to Evenhuis (1990), also Burt et al. (1998) used a prospective longitudinal approach for studying dementia in 70 people with Down syndrome, of whom at entry 16 had a severe intellectual disability and 5 a profound intellectual disability. Based on the International Classification of Diseases and Related Health Problems 10th revision, two persons with a severe intellectual disability and three with a profound intellectual disability had dementia (Table 1). A decline in memory was found in three persons and a decline in other cognitive functions was found in four individuals. In one person, direct memory and/or other cognitive tests were not possible, but decline was assumed to be present based on informant reports. All five persons declined in everyday functioning along with emotional and psychological problems can be explained as symptoms of dementia, particularly weight change, loss of energy and sleep disorder (Table 1).

3.2 Potential dementia symptoms in ageing studies

It is expected that dementia is often underdiagnosed in people with severe/profound intellectual disabilities due to the complexity of diagnosing dementia in this population. Therefore, changes in cognitive functioning and/or behavioural and psychological alterations were considered to be potential symptoms of dementia.

The first study reporting age-related changes, which can in fact be dementia symptoms in adults with severe/profound intellectual disabilities was published by Haveman et al. (1994). They evaluated challenging behaviour and psychological problems in 1580 persons with intellectual disabilities according to age, level of intellectual disability and presence of Down syndrome. Specifically for people with a severe intellectual disability without Down syndrome, they found less challenging behaviour with advanced age, whereas psychological problems were evenly distributed. In people with Down syndrome, elderly with severe intellectual disabilities had more psychological problems (Table 1). The authors concluded that these psychological problems can be explained as symptoms of dementia, given that 39 of the 85 persons with Down syndrome (mild and severe intellectual disabilities) aged 50 years or older had a diagnosis of dementia.

Next, Cherry et al. (1997) undertook a cross-sectional study focusing on symptoms associated with psychiatric disorders in younger (20–29 years) versus older adults (60–79 years) with severe/profound intellectual disabilities (Table 1). Using the Diagnostic Assessment for the Severely Handicapped, psychopathologic symptoms were assessed based on frequency, duration and severity. Older adults showed longer durations for anxiety, inappropriate sexual behaviour and impulse control problems, as well as increased severity for anxiety, stereotypies/tics and impulse control problems. Moreover, the results implicated that anxiety and impulse control problems were more problematic in older adults. Additionally, age-related changes in severe versus profound intellectual disabilities were compared. Older persons with a profound intellectual disability had longer...
durations of self-injury behaviour compared to those with a severe intellectual disability. The authors reported that the prevalence of diagnosis for psychiatric disorders, particularly classic forms of mental illness like anxiety was low.

Lastly, Rousseau et al. (2019) evaluated ageing in 474 people with a profound intellectual disability and severe motor deficiency. Compared to younger individuals (18–34 years), older adults (50–68 years) presented more frequently behavioural problems like withdrawal, intermittent screaming, intermittent crying, agitation and self-aggressivity. Similar proportions of aggressivity, stereotypies, mericysm and sleep problems were found in young, middle-aged (35–49 years) and older persons. Moreover, cognitive skills including language, posture-motor ability, coordination and sociability decreased with age.

3.3 | Synthesis of results

In summary, eight studies reported dementia symptoms for in total 27 adults with severe/profound intellectual disabilities and dementia. Of these 27 individuals, 22 had Down syndrome 1 had Williams syndrome, 1 had Fragile X syndrome and for 3 individuals the aetiology was unspecified. Additionally, three studies focusing on age-related changes in people with severe/profound intellectual disabilities found a decline in cognitive functioning and an increase of emergence of BPSD, which could potentially relate to dementia-related symptoms given the complexity of diagnosing dementia in this population. Table 2 provides an overview of reported cognitive changes, BPSD, changes in the ability to perform activities of daily living, neurological and other physical changes.

3.4 | Limitations of primary literature

The very limited number of studies that explicated studied dementia in people with severe/profound intellectual disabilities evidently showed that this population has been largely neglected in literature so far. Here, a first inventory of observable symptoms in this population is provided. Given that the retrieved articles had similar limitations, these limitations were not discussed per primary article but were summarised in general, grouped according to the data extraction categories presented in Table 1.

3.4.1 | Study population(s)

The first limitation concerned the small number of people with severe/profound intellectual disabilities for who observable dementia symptoms were reported (ranging from n = 1 to 6 in each dementia study). Secondly, the aetiology of the intellectual disability was not specified in two dementia studies for a total of three persons (Day, 1985; Reid & Aungle, 1974) as well as for the 477 persons without Down syndrome in the ageing study of Haveman et al. (1994) and 474 persons in the ageing study of Rousseau et al. (2019). Thirdly, the exact (sub)type of dementia was not reported in five dementia studies (Burt et al., 1998; Day, 1985; Duggan et al., 1996; Margallo-Lana et al., 2007; Reid & Aungle, 1974). In another study it was not clearly reported whether the person with Down syndrome and the person with Fragile X syndrome had Alzheimer’s disease dementia or vascular dementia Sauna-Aho et al. (2018).

3.4.2 | Intellectual disability classification

Criteria to determine the level of intellectual disability varied across studies, introducing a potential degree of variation. In fact, two studies did not specify how the level of intellectual disability was established (Haveman et al., 1994; Margallo-Lana et al., 2007). Surprisingly, Burt et al. (1998) determined the level at the entry of the study, introducing uncertainty about the premorbid level of intellectual disability, i.e., if those persons had always functioned in the severe/profound range at baseline.

3.4.3 | Assessment of dementia/age-related changes that are potential dementia symptoms

Similar to determining the level of intellectual disability, assessment procedures for dementia in those with severe/profound intellectual disabilities varied across studies. Two studies obtained diagnoses from case files without elaborating on the exact diagnostic procedure (Day, 1985; Määttä et al., 2006). Five studies reported that they had used instruments to identify (potential) dementia symptoms (Burt et al., 1998; Cherry et al., 1997; Duggan et al., 1996; Haveman et al., 1994; Sauna-Aho et al., 2018). In the remaining studies (potential) symptoms were retrieved from case files and/or information obtained from staff.

Taken together, studies retrieved in this systematic review displayed rather similar limitations with respect to small sample sizes and variation in assessment criteria/procedures.

4 | DISCUSSION

To the best of our knowledge, this review is the first to systematically identify observable dementia symptoms in people with severe/profound intellectual disabilities and a clinically and/or postmortem confirmed diagnosis of dementia in the – very scarce – available literature. Using an extensive search strategy, only eight studies were identified focusing – in part – on dementia in this population. Additionally, given the complexity of diagnosing dementia, three studies describing a decline in cognitive functioning and/or behavioural and psychological alteration in the context of ageing were also included. Summarising symptoms of (potential) dementia, this review revealed a decline in cognitive functioning, involving a deterioration in speech and losses of social skills as well as BPSD, in particular withdrawal and
aggressiveness. Furthermore, specifically in those with dementia, skills necessary to perform activities of daily living declined. Lastly, neurological and physical changes like, incontinence, (late-onset) epilepsy and a deterioration in gait were reported.

In line with the diagnostic criteria of dementia, eight studies reported cognitive changes, merely for those with severe intellectual disabilities (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2018). Two studies reported that specifically in those with profound intellectual disabilities, it is virtually impossible to show measurable changes in cognitive function with neuropsychological tests (Evenhuis, 1990; Margallo-Lana et al., 2007), which is in accordance with the findings of Elliott-King et al. (2016). Cognitive functions may never have been acquired and therefore cannot decline (Holland et al., 2000). Furthermore, they have to be supported by care professionals for activities of daily living, making it complex to determine whether dementia-related cognitive impairments interfere with the ability to perform activities of daily living. Nevertheless, it might be possible to determine dementia in this population based on BPSD, given that BPSD are found in all types of dementia and are most observable for caregivers (Engelborghs et al., 2005; Finkel, 2000).

In fact, the eight dementia studies as well as the three ageing studies all reported on BPSD. Four studies found symptoms of apathetic behaviour including withdrawal and loss of interest in surroundings (Evenhuis, 1990; Määttä et al., 2006; Margallo-Lana et al., 2007; Rousseau et al., 2019). These results are consistent with recent findings in a large study on dementia in people with Down syndrome, in which apathy was found one of the most commonly observed BPSD symptoms (Dekker et al., 2021, 2018). Furthermore, in this study a substantial proportion of people with Down syndrome and Alzheimer's disease displayed an increased frequency of aggressive behaviour (Dekker et al., 2021, 2018). Similarly, two dementia studies included in this review reported aggression in three individuals with a severe intellectual disability, Down syndrome and dementia (Evenhuis, 1990; Määttä et al., 2006). Additionally, the prevalence of self-aggressivity increased with age in people with a profound intellectual disability without an official diagnosis of dementia (Rousseau et al., 2019). Taken together, apathy and aggression reported in ageing studies can be signs of dementia in persons with severe/profound intellectual disabilities.

Furthermore, in those with a clinically and/or postmortem confirmed diagnosis of dementia, reported BPSD were irritability, alterations in eating/drinking behaviour and sleep problems. Irritability was observed particularly in individuals with a severe intellectual disability and Down syndrome (Evenhuis, 1990; Määttä et al., 2006), which is in line with results of other studies focusing on dementia symptoms in persons with Down syndrome (Lai & Williams, 1989; Moss & Patel, 1995). Moreover, Duggan et al. (1996) found alterations in eating and drinking behaviour also specifically in a person with a severe intellectual disability and Down syndrome. This suggests that eating and drinking behaviour is affected by dementia (Dekker et al., 2021, 2018). Additionally, individuals with severe/profound intellectual disabilities and dementia presented sleep problems including disturbed sleep and daytime sleepiness (Evenhuis, 1990; Reid & Aungle, 1974; Sauna-Aho et al., 2018). Sleep problems are common in the population of people with intellectual disabilities (Van de Wouw et al., 2012). Nevertheless, they are important to consider given that they may aggravate cognitive decline and BPSD (Dekker et al., 2015).

Besides emerging BPSD, the ability to perform activities of daily living declined. Similar to Lai and Williams (1989), three dementia studies reported losses of self-care skills in individuals with a severe intellectual disability and Down syndrome (Evenhuis, 1990; Määttä et al., 2006; Reid & Aungle, 1974). Furthermore, dementia studies reported symptoms related to neurological and other physical changes like, incontinence, (late-onset) epilepsy, hypertonia and gait deterioration. In the study of Prasher (1995), these symptoms were associated with increasing severity of dementia.

4.1 | Strengths

This systematic review is a first step towards a proper understanding of the presentation of dementia in the population of people with severe/profound intellectual disabilities. A thorough search strategy using a broad range of searching terms, including older (sometimes abandoned) terminology, was performed to identify studies reporting dementia symptoms. Besides, we reviewed ageing literature describing changes in cognitive functioning and/or behavioural and psychological alterations which can in fact be symptoms of dementia given the complexity of diagnosing dementia in this population. Indeed, in one ageing studies the authors confirmed that observed psychological problems were symptoms of dementia.

4.2 | Limitations

Although this study provides the first steps towards a proper understanding of the natural history of dementia in people with severe/profound intellectual disabilities, various limitations were found across the primary literature retrieved here. The rather small number of people with severe/profound intellectual disabilities and dementia poses a threat to representativeness of these results for the entire population. Consequently, further analyses on aetiological subgroups were not possible (further complicated by evident lack of reporting of aetiologies). Establishing patterns of symptoms associated with different (sub)types of dementia could also not be determine because studies did not report the exact type of dementia.

Given the complexity of diagnosing dementia in people with severe/profound intellectual disabilities, it is questionable whether early dementia symptoms were observed or whether observed symptoms were attributed to the intellectual disability, ageing or another condition rather than dementia. For instance, cognitive changes and BPSD may also be caused by other causes than dementia, for example, sensory impairments and psychiatric disorders (Moriconi et al., 2015). In fact, studies reported high prevalence rates of visual or hearing deficits and psychiatric disorders including depression and
schizophrenia in elderly with severe/profound intellectual disabilities (Evenhuis et al., 2001; Haveman & Maaskant, 1989; Kirkpatrick-Sanchez et al., 1996; Van Splunder et al., 2006).

Furthermore, our search strategy was targeted to retrieve studies focusing on severe/profound intellectual disabilities. Given the functionalities of the databases PubMed, PsychINFO and Web of Science, studies assessing a broad level of intellectual disabilities without specifying the level in title or abstract were likely missed in the search strategy. To that end, we performed an additional search to assess how many studies focusing on dementia in people with intellectual disabilities (in the broadest sense) or persons with Down syndrome published in the last five years were potentially missed. No additional broad intellectual disability and Down syndrome studies reporting dementia symptoms in those with severe/profound intellectual disability were identified. This emphasises the lack of focus on dementia in this population.

4.3 | Future implications

This systematic review provides a first overview of observable dementia symptoms in people with severe/profound intellectual disabilities. There is an evident need for further study of the natural history of dementia in this population. Future studies should focus on identification of observable dementia symptoms. Evidently, literature only provided limited clues. Therefore, it is of utmost importance to make an inventory of practice-based observations, among others by analysing existing medical files and by collecting observed symptoms by care professionals with vast experience in intellectual disability care through surveys and interviews. Such information about symptomology of dementia is relevant to enable (early) detection and diagnosis of dementia in this population. This enables family and health care professionals to adequately adapt daily caregiving (Janicki, 2011; Dekker, Wissing et al., 2021). Furthermore, early diagnosis allows development of an individual treatment plan, including choices about medication use, to reduce specific symptoms or slow the rate of further decline (Janicki, 2011; Dekker, Wissing et al., 2021). Altogether, (early) diagnosis of dementia may contribute to the well-being of individuals with severe/profound intellectual disabilities.

5 | CONCLUSION

Dementia in people with severe/profound intellectual disabilities has received very little attention so far, as shown by the limited number of studies focusing on this complex combination. Here, we have identified and summarised observable symptoms in available literature. Despite the few small-sized studies, a range of dementia symptoms were identified, subdivided in cognitive decline (e.g., memory loss, forgetfulness, deterioration in speech, losses of social skills), decline in activities of daily living (e.g., self-cares skills, everyday functioning/skills), BPSSD (e.g., apathy, aggression, irritability, altered eating/drinking behaviour) as well as neurologic and other physical symptoms (e.g., incontinence, late-onset epilepsy, hypotonia, gait deterioration). Because of increasing life expectancy, dementia will become more prominent in people with severe/profound intellectual disabilities. This review is a first step in improving the diagnostic procedure in this population. Future studies are required to specifically address dementia in people with severe/profound intellectual disabilities, further establishing the natural history. This would enable (early) detection and diagnosis of dementia which contributes to maintaining quality of life in people with severe/profound intellectual disabilities and dementia.

ACKNOWLEDGEMENTS

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#2 Alzheimer* OR Change OR Changed OR Changes OR Changing OR Decline OR Dement* OR Deteriorat* OR Major cognitive dysfunction* OR Major cognitive impairment* OR Mild cognitive dysfunction* OR Mild cognitive impairment* OR Creutzfeldt Jakob OR Degeneration OR Neurodegenerat* OR K Kluever-Bucy OR Lewy Body OR Cerebrovascular OR Wilhelmsen Lynch OR Pick* ORBinswanger OR Progressive OR Leukoencephalopathy OR Aging OR Aging OR Neurodegeneration*

#3 Elder* OR Geriatric* OR Old age OR Older participant* OR Old population* OR Older individual* OR Older adult* OR Older patient* OR Older people OR Older person* OR Older subject* OR Oldest participant* OR Oldest population* OR Oldest individual* OR Oldest adult* OR Oldest patient* OR Oldest people OR Oldest person* OR Oldest subject* OR Senior* OR Aged individual* OR Aged population* OR Aged participant* OR Aged adult* OR Aged patient* OR Aged people OR Aged person* OR Aged subject*

#4 [#1 TI AND #2 TI AND #3 TI] OR [#1 AB AND #2 AB AND #3 AB]