Observe
Once again

Dementia in people with severe/profound intellectual (and multiple) disabilities

Maureen B.G. Wissing

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The work presented in this dissertation was part of the research project *'Practice-based questions about dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D)'*, a collaborative effort of the University of Groningen, University Medical Center Groningen (UMCG) and Hanze University of Applied Sciences, with four Dutch care organizations spread across the country: Alliade, 's Heeren Loo, Ipse de Bruggen and Royal Dutch Visio. These care organizations are representative for the Dutch intellectual disability care sector given the high number of people with SPI(M)D for whom they provide diagnostic work-up, treatments and deliver care. The project was funded by the ZonMw Dementia Research and Innovation Programme 'Memorabel' (no. 733050863).

Dit onderzoek is onderdeel van het project *'Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen ((Z)EV(M)B)'*, een samenwerking tussen Rijksuniversiteit Groningen, Universitair Medisch Centrum Groningen, Hanzehogeschool Groningen, Alliade, 's Heeren Loo, Ipse de Bruggen en Koninklijke Visio. De laatste vier zijn zorgorganisaties verspreid over heel Nederland die diagnostiek, begeleiding en behandeling bieden aan mensen met verstandelijke beperkingen. Dit project wordt gesubsidieerd door ZonMW, dementie onderzoeks- en innovatieprogramma 'Memorabel' (projectnummer 733050863). Meer informatie: www.vb-dementie.nl.

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People
with severe/profound intellectual
(and multiple) disabilities

Aging

Dementia

The artist who crafted the crayon drawing and I
kept the above terms in mind as we underwent the endeavor of
creating the design for this dissertation.

As a reader, it is up to you to observe once again
to interpret the underlying meaning.

Mensen
met (zeer) ernstige verstandelijke
(en meervoudige) beperkingen

Veroudering

Dementie

met deze termen zijn
de kunstenaar die de krijttekening heeft gemaakt
en ik aan de slag gegaan met de vormgeving van dit proefschrift.

Het is aan u als lezer om nogmaals te observeren
om de onderliggende betekenis te ontdekken.

Contents

Chapter 1

Introduction to people with severe/profound intellectual (and multiple) disabilities and dementia

General introduction

People with SP(I(M)D

People with severe/profound intellectual (and multiple) disabilities (SPI(M)D) form a heterogeneous group because of major differences in intellectual functioning, sensory abilities, communication, motor functioning and abilities to perform activities of daily living (ADL) (Nakken & Vlaskamp, 2007; van der Putten et al., 2017). Each individual has a unique set of abilities. Some individuals, for example, use spoken words to communicate, whereas others communicate non-verbal via vocal sounds and facial expressions. Concerning motor functioning, some individuals can walk independently, while others make use of a wheelchair. Moreover, regarding performing ADL, some can put on a shirt, while others engage in dressing by helping to put their arm through a sleeve. Over time people with SPI(M)D can expand their abilities within the domains mentioned above (Belva & Matson, 2013; Houwen et al., 2014), but there can also be a decline from an individual's previous functioning.

Aging

A decline in functioning in people with SPI(M)D might be related to aging or age-related conditions, especially considering the trend of increased life expectancy of people with intellectual disabilities (ID) (Coppus, 2017; E. Evans et al., 2013; Torr & Davis, 2007). Improvements in medical care, supportive care and living circumstances in the past century have contributed to the substantial increase in life expectancy of people with ID, including those with SPI(M)D (Elliott-King et al., 2016; Rousseau et al., 2019; Whitehouse et al., 2000; Zigman et al., 2008). In 1990, Eyman et al. reported that the expected age of death for people with SPI(M)D was before the age of 20. Almost 30 years later, Rousseau et al. (2019) described that an increased number of people with SPI(M)D, particularly those with less severe health problems, survive for more than 50 years. Recent prevalence data is lacking. Estimations, for example, in The Netherlands, indicate that about 65% of the approximately 10.000 adults with SPIMD were older than 40 years (Vugteveen et al., 2014). Given that people with SPI(M)D are becoming older, age-related conditions will become more prevalent.

Dementia

Dementia is one of the conditions for which increased age is the main risk factor (Alzheimer's Association, 2022). Dementia is an umbrella term for

a group of symptoms associated with a progressive decline in cognitive functioning from an individual's previous higher level of functioning, which interferes with daily functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). Although cognitive changes, e.g., memory loss, are main indicators for dementia, behavioral and psychological changes, motor changes and medical comorbidities may also be present (American Psychiatric Association, 2013; Finkel, 2000; Kurrle et al., 2012; McKhann et al., 2011; Ries, 2018; World Health Organization, 2022). The manifestation of dementia depends partly on the underlying cause. Dementia has several causes, of which Alzheimer's disease (AD) is the most common (Alzheimer's Association, 2022).

Particularly, people with Down syndrome (DS) have an increased risk of developing dementia due to AD. This relates to the triplication of the human chromosome 21, which causes DS, i.e., trisomy 21 (Lott & Head, 2019; Snyder et al., 2020). One of the genes located on chromosome 21 encodes for the production of amyloid precursor protein (APP), which is the precursor of amyloid-β (Aβ). Accumulated amyloid-β is the main component of amyloid plaques, one of the characteristic pathological features of AD (Alzheimer's Association, 2022). Having an extra copy of chromosome 21 results in overproduction and accumulation of amyloid-β (Ballard et al., 2016; Coppus et al., 2012; Snyder et al., 2020; Wiseman et al., 2015). By the age of 40, nearly all individuals with DS have developed a significant amount of amyloid plaques (Lott & Head, 2019; Mann & Esiri, 1989; Wisniewski et al., 1985). This does not instantly result in clinical symptoms of dementia due to AD. About 9% of the individuals with DS have developed clinical AD in their 40s (Coppus et al., 2006), whereas this is approximately 75% by age 65 (Wiseman et al., 2015). Compared to the general population, where AD is present in about 5% of those aged between 65-74 (Alzheimer's Association, 2022), people with DS have a considerably higher risk of developing dementia due to AD. This also applies to those with DS having SPI(M)D, which is the case in approximately 20-30% of all individuals with DS (Coppus, 2017; Coppus et al., 2006).

Relatively little research has addressed the prevalence of dementia in people with ID not attributable to DS. Results of the few studies reporting on this appear to be variable (Elliott-King et al., 2016; Krinsky-McHale & Silverman, 2013). Some studies reported prevalence rates of 18,3% and 13,9% in people aged 65 and over (Strydom et al., 2007; Takenoshita et al.,

2020), which are higher than in the general population. Others reported a lower prevalence than in the general population. For example, Zigman et al. (2004) reported a prevalence of 4,2% in those aged 65 and over. Remarkably, in these studies, only people with mild, moderate, or severe ID were included, and thus not people with profound ID. Therefore, one could argue that those with profound ID do not develop dementia. Alternatively, it could be the case that dementia is not recognized in this population, and thus remains undiagnosed.

Complexity of recognizing and diagnosing dementia in people with SPI(M)D

Knowledge about symptoms and course of dementia in people with SPI(M)D is required to (early) recognize and diagnose dementia. However, scientific literature about dementia in people with SPI(M)D is scarce. This is related to the fact that in the past many people with SPI(M)D did not reach the age at which they could have developed dementia. Because of increased life expectancy, dementia will become increasingly prevalent among people with SPI(M)D, particularly those with DS. Consequently, dementia is a growing challenge in contemporary intellectual disability care.

Previous studies reported that caregivers of people with ID had difficulty recognizing dementia symptoms (Furniss et al., 2011; Herron & Priest, 2013; Whitehouse et al., 2000). They lacked knowledge about the symptoms and course of dementia. Similarly, care professionals working in Dutch intellectual disability care organizations and family members repeatedly indicated that they had limited knowledge about dementia in people with SPI(M)D. They wondered which dementia symptoms can be observed in people with SPI(M)D and how a dementia diagnosis can be established in this population. They also pointed out that they had observed a decline in functioning in elderly with SPI(M)D. These individuals were suspected of having dementia, but a diagnosis could not be established in many cases.

Dementia is likely to be underdiagnosed in people with SPI(M)D, which can be attributed to the lack of knowledge of dementia as well as the complexity of recognizing and diagnosing dementia in this population. Several factors make it difficult for informants (e.g., family members, caregivers/ direct support professionals) to observe symptoms and for clinicians (e.g., physicians, psychologists) and allied health care professionals

who support the diagnostic process (e.g., speech-language therapists, physiotherapists, occupational therapists, dieticians) to establish a dementia diagnosis in people with SPI(M)D. Figure 1.1 displays complicating factors subdivided into three levels: 1) characteristics of people with SPI(M)D, 2) recognition of symptoms, and 3) diagnostic process. For each level, the complicating factors are explained in more detail below by means of literature and practice-based experiences.

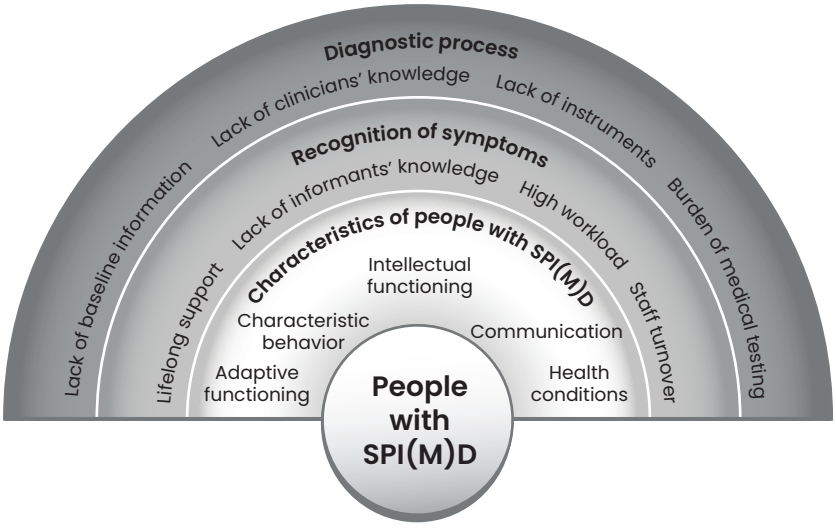


Figure 1.1 Complicating factors of recognizing and diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D).

Characteristics of people with SPI(M)D

People with SPI(M)D have significant limitations in both intellectual functioning and adaptive functioning/behavior, which originated during the developmental period, i.e., before the age of 22 (American Psychiatric Association, 2013; Schalock et al., 2021; World Health Organization, 2022). Moreover, individuals often have multiple other disabilities, such as motor, hearing, and/or visual impairments. In the past, the classification of the severity of the ID was based on IQ scores. People with an IQ score between 20-35 were classified as having a severe ID, and people with an IQ score of less than 20 as having a profound ID (American Psychiatric Association, 2010). Nowadays, the classification of the severity level is primarily based on the individual's level of adaptive functioning/behavior and the intensity of support (American Psychiatric Association, 2013; Schalock et al., 2021). People with severe ID need extensive support for performing daily

activities, whereas people with profound ID (almost) totally dependent on others for everyday tasks (American Psychiatric Association, 2013; Nakken & Vlaskamp, 2007; Schalock et al., 2021). The aforementioned and some other characteristics of people with SPI(M)D make it complex to recognize and diagnose dementia. These complicating factors are displayed in the first level of Figure 1.1.

Firstly, the pre-existing severe/profound limitations in intellectual functioning is a complicating factor. Dementia namely manifests in areas of cognitive functioning that are already less developed due to the ID (Elliott-King et al., 2016). Hence, a decline in cognitive functions may be less noticeable and thus be underrecognized in those with SPI(M)D. If cognitive changes are recognized, they may be incorrectly attributed to the underlying ID rather than to the development of dementia (Whitehouse et al., 2000).

> ***Physician T.:*** *''Cognitive symptoms are particularly difficult to observe if someone already has severe/profound limitations in cognitive functioning.''* *

A second complicating factor is that people with SPI(M)D have significant disabilities in adaptive functioning, such as the ability to ADL (Schalock et al., 2021). Consequently, skills needed to (independently) perform ADL may never have been developed. Skills that a person with SPI(M)D has never developed cannot decline, and therefore cannot be indicative of dementia (Llewellyn, 2011; Sheehan, Sinai, et al., 2015).

> ***Psychologist A.:*** *''What makes it complex [to recognize and diagnose dementia in people with SPI(M)D] is that there are a limited number of acquired skills which could decline.''*

Thirdly, individuals with SPI(M)D have (life-long) patterns of characteristic behavior related to the ID (Thompson et al., 2022). This makes it challenging to differentiate alterations in behavior due to dementia from fluctuations in characteristic behavior (Dekker, Strydom, et al., 2015; Jamieson-Craig et al., 2010; Zigman et al., 2008).

*Illustrative quotes from care professionals and family members who mentioned complicating factors for recognizing and diagnosing dementia in people with SPI(M)D in focus groups or interviews (Chapters 2 & 4) were selected.

A fourth complicating factor is that many people with SPI(M)D have multiple concurrent health problems (Hermans & Evenhuis, 2014; van Timmeren et al., 2017). This increases the possibility that they have conditions such as depression, delirium, hypothyroidism, sleep apnea, vision/hearing impairments, or vitamin B12 deficiency, which could result in symptoms that mimic dementia (Moriconi et al., 2015; Scott & Barrett, 2007).

> ***Psychologist R.:*** *''It is difficult that besides the possibility that someone [with SPI(M)D] has dementia, (...) also age-related vision and hearing problems can develop and cause [dementia-like] changes.''*

A fifth complexity concerns the communication of people with SPI(M)D, which is primarily non-verbal (Nakken & Vlaskamp, 2007). Given that they use little or no verbal communication, there are hardly any self-reported symptoms (Smiley & Cooper, 2003). Consequently, people with SPI(M)D primarily rely on observations of caregivers and family members, i.e., informants, for recognizing dementia symptoms (McKenzie et al., 2018).

> ***Psychologist C.:*** *''People with SPI(M)D do not tell me which symptoms they are experiencing. Consequently, they depend on [the observations of] those involved.''*

Recognition of symptoms

The second level in Figure 1.1. focuses on the ability of informants to recognize (potential) dementia symptoms.

A first complicating factor is that caregivers and family members may lack knowledge about the symptomatic presentation of dementia (Furniss et al., 2011; Herron & Priest, 2013; Whitehouse et al., 2000). Consequently, symptoms may not be recognized, or wrongly attributed to ID.

> ***Occupational therapist D.:*** *''It is important that [caregivers and family members] are aware of potential [dementia] symptoms, and have sufficient knowledge about dementia.''*

Secondly, recognizing symptoms is complicated because people with SPI(M)D already need lifelong extensive/pervasive support (Nakken & Vlaskamp, 2007; Schalock et al., 2021). Caregivers and family members

might automatically provide more support when needed, without being aware of subtle changes in, for example, ADL.

> ***Physician J.:*** *''People [with SPI(M)D] receive 24-hour care. Therefore, it might be possible that subtle changes are not directly noticed.''*

A third potential complicating factor concerns the high workload in intellectual disability care. An increasing problem is that care organizations are understaffed because of staff turnover and unfulfilled vacancies (Ministerie van Volksgezondheid Welzijn en Sport, 2022; Vereniging Gehandicaptenzorg Nederland, 2022)). In The Netherlands, for example, 45% of the 181.000 intellectual disability employees had to deal with prolonged understaffing (Centraal Bureau voor Statistiek, 2020). Additionally, among caregivers in intellectual disability care, a majority (50,8%) feel that the workload is too high. (Centraal Bureau voor Statistiek, 2022). Because of the high workload, and thus lack of time to extensively support each individual with SPI(M)D, symptoms might go unnoticed.

Fourthly, staff turnover further complicates the recognition of symptoms (Janicki et al., 1995; Whitehouse et al., 2000). To recognize dementia symptoms, knowledge about how a person has functioned throughout adult life (without decline) is needed (Janicki et al., 1995). Caregivers and family members who have known the person for a long time can compare the person's current level of functioning with his/her characteristic functioning in the past. However, when caregivers decide to quit their job, knowledge about the characteristic functioning of this person may (partly) disappear because the small details in functioning of people with SPI(M)D are difficult to transfer to others or describe in clinical records.

> ***Family member E.:*** *''I think that it is important to know the person [with SPI(M)D] for a long time to be able to observe [dementia-related] changes.''*

Diagnostic process

Today, a dementia diagnosis is still established based on the clinical expression of symptoms, whereby other potential causes should be ruled out (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). Clinical assessment (by experienced clinicians) to establish whether a person has dementia involves neuropsychological

evaluation, informant interviews, physical examination, blood tests, and/or brain imaging (Scott & Barrett, 2007). For people with SPI(M)D various complicating factors exist for diagnosing dementia. These factors are displayed in the third level of Figure 1.1.

A first complicating factor relates to the availability of information about the individual's premorbid level of functioning. To meet dementia diagnostic criteria, there needs to be a decline in cognitive functioning from one's baseline functioning interfering with daily functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). When information about baseline functioning is missing – either due to staff turnover or insufficient reporting in clinical records – it is complex for clinicians to diagnose dementia (Llewellyn, 2011; McKenzie et al., 2018; Zigman et al., 2008).

> ***Psychologist X.:*** *''What is reported in clinical records is often difficult to interpret. There are also continuous staff turnovers, whereby information about a person's functioning will go missing.''*

A second complexity concerns the clinicians' limited level of knowledge about dementia in people with SPI(M)D. This may cause them to mistakenly attribute dementia-related changes to the ID, other health conditions, or aging (Cleary & Doody, 2017).

Thirdly, the potential burden of medical testing for people with SPI(M)D is a complicating factor. Assessment of dementia should ideally include blood tests to rule out other health conditions that may mimic dementia, e.g., hypothyroidism and vitamin B12 deficiency (Scott & Barrett, 2007). However, taking a blood sample could be challenging when an individual with SPI(M)D demonstrates resistance behavior. Moreover, brain imaging and cerebrospinal fluid examination can be used to aid the diagnosis (Ahmed et al., 2014; Scott & Barrett, 2007). In The Netherlands, clinicians are generally hesitant to let people with SPI(M)D undergo such medical tests. For people with SPI(M)D, it may be very difficult to sit or lay still, which is required for both brain imaging and lumbar puncture. Clinicians may consider to perform such tests under anesthesia. However, there are potential risks associated with anesthesia (Choi & Doh, 2021).

> ***Psychologists G.:*** *''Brain imaging places a significant burden on a person [with SPI(M)D]. Using anesthesia [during the procedure] also involved risks. Therefore, it is not standard in the diagnostic process.''*

A last complicating factor relates to the absence of validated instruments for this population (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). Direct neuropsychological tests cannot be used for people with SPI(M)D. These tests namely require proper understanding of test instructions and well-developed verbal communication, which are very limited in people with SPI(M)D (Nieuwenhuis-Mark, 2009; Oliver & Kalsy, 2005). Therefore, floor effects occur when conducting neuropsychological tests (Elliott-King et al., 2016; Esbensen et al., 2017; McKenzie et al., 2018). Alternatively, informant-based dementia screening instruments could be used to aid the diagnosis of dementia. However, available instruments for people with ID are as a whole unsuitable for people with SPI(M)D (Elliott-King et al., 2016; Evenhuis, 1990; Hon et al., 1999; Margallo-Lana et al., 2007).

> ***Psychologist A.:*** *''I would like to see a more suitable dementia screening instrument [for people with SPI(M)D] focusing on subtle changes (…). [In existing dementia screening instruments] there are namely questions like 'Is someone able to independently dress up?'. Well the answer is always no [for people with SPI(M)D].''*

There are thus a considerable number of complicating factors for recognizing and diagnosing dementia in people with SPI(M)D. Given these complications, one could wonder whether it is possible and even necessary to observe and assess a decline in functioning due to dementia in people with SPI(M)D. Clearly, the complicating factors regarding the characteristics of people with SPI(M)D will not change. However, improving care professionals' and family members' abilities to recognize symptoms can lead to more accurate observation of symptoms and earlier diagnosis of dementia. To that end, engagement of care professionals and family members is vital. Dutch intellectual disability caregivers and family members expressed that to be able to recognize subtle changes in functioning due to dementia they need to have more knowledge about dementia in people with SPI(M)D. Moreover, clinicians indicated that there is a lack of suitable dementia screening instruments for people with

SPI(M)D, and therefore an instrument to aid the dementia diagnosis in this population should be developed.

Aim and outline of this dissertation

To provide answers to practice-based questions and fulfill needs as described above, this dissertation aims to gain insight into dementia in people with SPI(M)D by focusing on:

1. Examining the relevance of a dementia diagnosis in people with SPI(M)D
2. Identifying training/information needs regarding dementia in people with SPI(M)D
3. Identifying observable dementia symptoms in people with SPI(M)D
4. Developing an instrument to aid the dementia diagnosis in people with SPI(M)D

To achieve this, a range of studies were carried out in collaboration with Dutch care organizations that provide diagnostic work-up, and treatments and deliver care for a large number of people with SPI(M)D. The chapters that report on these studies and their relation to one another are depicted in Figure 1.2.

Examine the relevance of a dementia diagnosis

So far, it remained unclear whether it is important to know if individuals with SPI(M)D have dementia. A diagnosis might enable the environment to (timely) respond to the individual's changing wishes and needs. On the other hand, one may argue that the level of support will not change once dementia is diagnosed because people with SPI(M)D already receive lifelong extensive/pervasive support. In **Chapter 2,** care professionals and family members – having experience with people with SPI(M)D showing decline/dementia – were in multidisciplinary focus groups asked about the relevance of dementia diagnosis.

Identify training/information needs

There is a lack of understanding about training/information needs in the context of dementia in people with SPI(M)D. Therefore, in **Chapter 2**, we also asked care professionals and family members about their training/information needs regarding dementia in people with SPI(M)D.

Identify observable dementia symptoms

Dementia symptoms in people with SPI(M)D are not yet fully understood. Hence, an inventory of practice-based observations of dementia symptoms in people with SPI(M)D was obtained using quantitative and qualitative research methods. In **Chapter 2**, focus group participants were also asked which dementia symptoms they recognized in people with SPI(M)D in daily practice. In addition, **Chapter 3** presents a systematic literature review aiming to identify and summarize symptoms reported in already existing literature. Together, this led to a first inventory of observable dementia symptoms in people with SPI(M)D. Next, in **Chapter 4**, practice-based observable symptoms were further identified and deepened through a survey among care professionals and family members and semi-structured interviews with care professionals having vast experience with dementia in people with SPI(M)D. Moreover, **Chapter 5** focuses on characterizing the natural history of dementia in people with SPI(M)D by determining the prevalence and time of onset of symptoms identified in previous steps through a retrospective analysis of clinical records of people with SPI(M)D.

Develop an instrument to aid the dementia diagnosis

Until now, no standardized dementia screening instruments dedicated to people with SPI(M)D exist. Therefore, we aimed to develop a novel instrument to aid the diagnosis of dementia in this population. In addition to identifying observable symptoms through literature, focus groups, survey, interviews, and clinical records review (mixed methods, **Chapters 2-5**), we also studied applicable items for people with SPI(M)D in already existing dementia screening instruments available for people with ID were identified (**Chapter 6**). Next, the results of identified observable dementia symptoms and applicable items were summarized and assimilated to develop items for the novel instrument to aid the dementia diagnosis. **Chapter 7** describes the development of the diagnostic aid for dementia in people with SPI(M)D and the results of the first examination of its validity, reliability, and discriminative ability. Moreover, practice-based experiences with using the diagnostic aid are presented in this chapter.

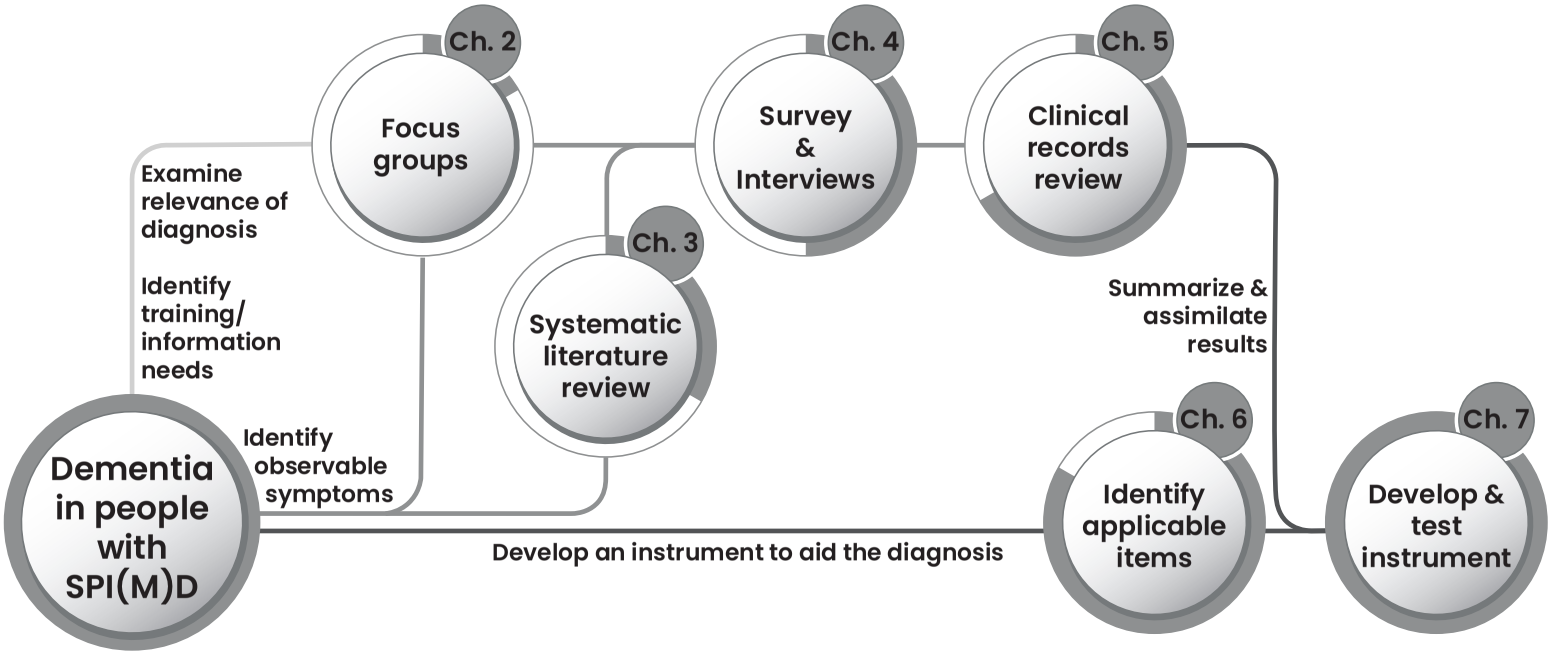


Figure 1.2 Schematic outline of the relation between the chapters of this dissertation.

Chapter 2

Focus group research into relevance, symptoms and training needs

Dekker, A. D., Wissing, M. B. G., Ulgiati, A. M., Bijl, B., van Gool, G., Groen, M. R., Grootendorst, E. S., van der Wal, I. A., Hobbelen, J. S. M., De Deyn, P. P. & Waninge, A. (2021). Dementia in people with severe or profound intellectual (and multiple) disabilities: Focus group research into relevance, symptoms and training needs. *Journal of Applied Research in Intellectual Disabilities*, *34*(6), 1602-1617.

Dutch version of the articles presented in chapters 2 & 3 |
Nederlandse versie van de artikelen gepresenteerd in hoofdstukken 2 & 3

Dekker, A. D., Wissing, M. B. G., Ulgiati, A. M., Bijl, B., van Gool, G., Groen, M. R., Grootendorst, E. S., van der Wal, I. A., Hobbelen, J. S. M., De Deyn, P. P. & Waninge, A. (2021). Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen: onderzoek. *NTZ*: *Nederlands Tijdschrift voor de Zorg aan mensen met een verstandelijke beperking*, *47*(4), 139–159.

Abstract

Introduction: Differentiating dementia from baseline level of functioning is difficult among people with severe/profound intellectual (and multiple) disabilities (SPI(M)D). Moreover, studies on observable dementia symptoms are scarce. This study examined 1) the relevance of dementia diagnosis, 2) observable symptoms, and 3) training/information needs.

Methods: Four explorative focus groups were held with care professionals and family members who have experience with people with SPI(M)D (≥40 years) and decline/dementia.

Results: Thematic analysis showed that participants wanted to know about a dementia diagnosis for a better understanding and to be able to make informed choices (question 1). Using a categorization matrix, cognitive and behavioral changes were shown to be most prominent (question 2). Participants indicated that they needed enhanced training, more knowledge development and translation, and supportive organizational choices/policies (question 3).

Conclusion: Timely identifying/diagnosing dementia allows for a timely response to changing needs. This requires a better understanding of symptoms.

Introduction

In the last decades, life expectancy of people with intellectual disabilities (ID) has increased even faster than in the general population (Bittles & Glasson, 2004; Coppus, 2013; E. Evans et al., 2013). Because advanced age is the greatest risk factor for dementia (Alzheimer's Association, 2020), dementia is a growing challenge in intellectual disability care. People with DS are at a particularly high genetic risk to develop Alzheimer's disease: approximately 75% would develop dementia by age 65 (Wiseman et al., 2015).

Diagnosing dementia in people with ID is complicated due to the level of intellectual disability, (life-long) patterns of characteristic/typical behavior related to the intellectual disability and the presence of comorbidities, which may be associated with dementia-like symptoms (Dekker, Strydom, et al., 2015; Jamieson-Craig et al., 2010; Sabbagh & Edgin, 2016; Zigman et al., 2008). Moreover, it may be hard to differentiate between aging and dementia. The diagnosis of dementia requires the presence of cognitive and behavioral decline from a previous higher level of functioning, and this decline must interfere with daily functioning (American Psychiatric Association, 2013; Fletcher et al., 2016; Sabbagh & Edgin, 2016; Zigman et al., 2008). However, the more severe and complex the present disabilities, the more difficult the assessment of decreasing skills due to dementia. This is particularly difficult in people with SPI(M)D (E. Evans et al., 2013; McKenzie et al., 2018).

For this population, there are hardly any validated direct neuropsychological tests and informant-based dementia questionnaires available for (early) identification and diagnosis of dementia (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). A diagnosis of dementia in this specific population is currently based on multidisciplinary clinical assessment (by experienced clinicians) involving observations, informant interviews, and/or screening case notes (Day, 1985; Duggan et al., 1996; Evenhuis, 1990; Määttä et al., 2006; Margallo-Lana et al., 2007; Reid & Aungle, 1974; Sauna-Aho et al., 2018). Moreover, studies on dementia symptoms in people with SPI(M)D are scarce because scientific research has primarily focused on dementia in people with mild/moderate ID (Wissing, Ulgiati, et al., 2022).

People with SPI(M)D have an estimated IQ of less than 35. Besides, they often experience serious health problems and sensory impairments that may adversely affect their functioning (Nakken & Vlaskamp, 2007; van Timmeren et al., 2016). In addition, they often experience profound neuromotor dysfunctions (Nakken & Vlaskamp, 2007). In these people, it is difficult to differentiate deterioration due to dementia from the severe or profound pre-existing limitations in functioning. Firstly, it is difficult to assess cognitive decline due to the developmental age below 36 months. Although memory changes are indicative of dementia in people with mild ID, decline in daily functioning is more visible in people with more severe ID (Jamieson-Craig et al., 2010). However, people with SPI(M)D often need lifelong support. They have never developed specific skills and have to be supported by care professionals for certain tasks. As a result, such skills cannot be considered as symptoms indicative of dementia (Llewellyn, 2011; Sheehan, Sinai, et al., 2015). Secondly, communication is mostly non-verbal and, therefore, there are no self-reported symptoms (Nakken & Vlaskamp, 2007; Smiley & Cooper, 2003). Thirdly, currently used dementia questionnaires are not suitable for SPI(M)D, and direct neuropsychological assessments are almost impossible due to floor effects (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). Fourthly, it is difficult to assess dementia-related decline due to the frequent presence of multiple concurrent health problems (van Timmeren et al., 2017).

Another obstacle for early identification and monitoring of deterioration in people with SPI(M)D is the dependence on observations of informants, such as caregivers/direct support professionals and family members (McKenzie et al., 2018), who often lack necessary background knowledge (Cleary & Doody, 2017b; Iacono et al., 2014) partly because information about symptoms and course of dementia in this population has been scarce until now (Wissing, Ulgiati, et al., 2022). On the other hand, family members and care professionals are often able to give concrete examples of minor signs of decline that they have observed. Until now, this knowledge has been individual-based and linked to one or a few people with SPI(M)D. Therefore, there is a great need for knowledge and education about dementia in this population in daily practice.

An explorative study paves the way for further research on dementia in people with SPI(M)D. This study focused on three practice-based questions:

- Why is it important to know if an individual with SPI(M)D has dementia? (question 1)
- Which dementia symptoms in people with SPI(M)D are recognized in daily practice? (question 2)
- What are training/information needs regarding dementia in people with SPI(M)D? (question 3)

Methods

Study consortium

This focus group study was part of the research project '*Practice-based questions about dementia in people with severe or profound intellectual (and multiple) disabilities,*' a collaborative effort of University of Groningen, University Medical Center Groningen (UMCG) and Hanze University of Applied Sciences, with four Dutch care organizations (Alliade, 's Heeren Loo, Ipse de Bruggen and Royal Dutch Visio) throughout The Netherlands, representative for the Dutch situation due to the high number of people with SPI(M)D for whom they provide care and treatments.

Study design

This explorative study was based on a qualitative research method using focus groups. Focus groups are group interviews that are not aimed at immediate problem-solving but at identifying practice-based experiences, attitudes and needs regarding a particular problem. Interaction between participants is key (van Royen & Peremans, 2007). We held four explorative focus group sessions with 11–13 participants each. To conduct and report this focus group study, we largely followed the method described by Breen (2006), the Consolidated Criteria for Reporting Qualitative Research (COREQ) (Tong et al., 2007) and van Royen & Peremans (2007).

Participants

Participants were selected based on the criterion that they would have something to say about dementia in people with severe/profound ID, that is, purposive sampling (Rabiee, 2004). Participants were purposefully selected using a two-stage procedure. First of all, care professionals and family members with experience with people (≥40 years) with severe/

profound ID (established according to clinical records and clinical judgment) and showing decline/dementia (with/without DS; with/without another (e.g., visual or motor) disability) were identified through contact persons at the four care organizations, the research advisory board in which family members and care professionals participated and through the project team members' network (snowball sampling method). In this process, the professions of potential participants were considered to ensure that the focus groups were multidisciplinary (like in daily practice). Therefore, the number of physicians/nurse specialists, allied health care professionals (occupational therapists, physiotherapists and speech therapists), psychologists (behavioral therapists who studied psychology or special needs education), psychological assistants, caregivers and family members were noted. If necessary, additional people were identified. A total of 53 potential participants were identified. This purposive sample subsequently received an invitation to participate by e-mail. Four people could not attend, resulting in 49 participants for this focus group study. These 49 participants were divided into the four focus groups in a multidisciplinary manner, that is, based on care professionals versus family members and based on different professions.

Ethics and consent

The Medical Ethical Committee of the UMCG decided that the Dutch Medical Research Human Subjects Act did not apply to this study (METc 2019/198). The study was registered in the UMCG Research Register (no. 201900193) and conducted in accordance with the UMCG Research Code and the EU General Data Protection Regulation. Each participant gave written consent for audiotaping of the focus group and analysis of this combined with questionnaire data.

Data collection

General participants' characteristics

Participants filled in a questionnaire stating their age, sex, highest level of education and relationship to people with SPI(M)D. Care professionals also stated how many years they have worked with people with SPI(M)D in general. Moreover, they answered on how frequently they work with people with SPI(M)D as well as with those with SPI(M)D and decline/dementia, respectively.

Focus group procedure

Four simultaneous focus group sessions were held, each lasting approximately 2 hours with a 15-min break. Each focus group was led by a moderator with considerable professional experience in intellectual disability care. For reasons of uniformity, the moderators received the same instructions and followed a procedural protocol drawn up in advance (Breen, 2006). Prior to the session, they welcomed participants, checked if participants signed informed consent forms and introduced the topic, the procedure, the rules of play (i.e., focus groups are not aimed at immediate problem-solving but at exploring and identifying experiences, attitudes and needs), the confidentiality and the multidisciplinary group composition. Furthermore, the three research questions were asked in the aforementioned order. The focus group interviews were semi-structured with three open research questions to guide the discussion. Moderators monitored time and ensured that all participants were able to speak.

Recording and transcription

The sessions were recorded with Tascam DR-40V2 digital audio recorders with an external omnidirectional microphone. Audio tapes were transcribed in Dutch (clean transcription) by the University Translation and Correction Service of the University of Groningen Language Centre. Fillers, hesitations and slips of the tongue were left out.

Data analysis

Three authors independently analyzed the transcripts using a qualitative method of content analysis called inductive content analysis (Elo & Kyngäs, 2008) also known as thematic analysis (Braun & Clarke, 2006) for question 1 and question 3. Following Braun & Clarke (2006), this analysis consists of five steps. In step 1 ('familiarizing with the data'), the three researchers independently read the full transcripts. In step 2 ('generating initial codes'), the transcripts were openly coded also independent of each other. In step 3 ('searching for themes'), the three researchers independently interpreted and divided into categories, which were then divided into overarching (sub) themes. This was an iterative process of reading, categorizing, rereading, refining and so forth. In step 4 ('reviewing themes'), the researchers met, discussed, compared and refined the division into categories and (sub) themes until they had reached consensus. In step 5 ('defining and naming themes'), phrasing of categories and (sub)themes was tailored to the research question. It is a recursive process, moving back and forth between

the steps (Braun & Clarke, 2006). To enhance trustworthiness and clarify participants' opinions and experiences, the thematic description (Section 3) was illustrated with authentic citations (Elo & Kyngäs, 2008). To improve readability, these quotes were linguistically corrected and, where possible, shortened (for instance, by leaving out unnecessary colloquial words) without the original meaning being affected.

For question 2, a qualitative method of content analysis combining aspects of deductive and inductive content analysis was used. Since dementia symptoms in people with SPI(M)D have hardly been studied in literature (Wissing, Ulgiati, et al., 2022), this study undertook an explorative approach to collect symptoms based on experiences in daily practice. To structure the broad range of symptoms, a categorization matrix (Elo & Kyngäs, 2008) was designed based on the most important clusters of dementia symptoms. The matrix rows were deductively designed in line with dementia diagnostic criteria (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022) showing the following themes: cognitive changes, behavioral changes (categories defined in accordance with the BPSD-DS evaluation scale (Dekker, Sacco, et al., 2018; Dekker, Ulgiati, et al., 2021a)), motor changes, and medical comorbidities (Strydom et al., 2010). To improve further interpretation, we categorized symptoms based on the daily contexts in which they are often observed in practice (columns). These daily contexts were inductively analyzed and defined based on the participants' descriptions of symptoms. In other words, symptoms in people with SPI(M)D mentioned by participants were coded and categorized in a matrix, which was partially deductively designed (rows consist of clusters of symptoms based on criteria/existing literature) and partially inductively designed (columns consist of daily contexts in which symptoms were seen according to participants).

Finally, the three researchers read the transcripts once more to compare these to the categories and (sub)themes that were ultimately defined per research question. This iterative process of reading, categorizing, rereading and refining also involved refining the naming of categories and (sub) themes. Since interaction between participants is key in focus groups (van Royen & Peremans, 2007) and participants thus respond to each other, this study did not intend to perform additional analyses with subgroups of participants. An integrated analysis was aimed for instead. The original Dutch manuscript with selected quotes was translated into English by the

University Translation and Correction Service of the University of Groningen Language Centre.

Results

To learn more about practice-based experiences, insights and needs regarding dementia in people with SPI(M)D, four focus group sessions were held with 13, 11, 12 and 13 participants, respectively. Each focus group had a multidisciplinary composition including different professions as well as family members. Based on the first analysis of these sessions, we concluded that answers were consistent with each other and saturation had been reached, that is, additional focus group sessions were not likely to provide new information. Table 2.1 shows the participants' characteristics.

Table 2.1 Participants' characteristics.

Characteristics	Total participant N=49	Family members n=8	Care professionals n=41
Age (years, mean ± SD (min. – max.))	49 ± 15 (25 – 76)	71 ± 5 (63 – 76)	45 ± 12 (25 – 63)
Gender (% female)	90	63	95
Care organization: Ipse de Bruggen, 's Heeren Loo, Alliade, Visio, other (%)	27, 33, 16, 14, 10	38, 13, 13, 13, 25	24, 37, 17, 15, 7
Level of education: MBO, HBO, WO (%)	31, 43, 27	38, 50, 13	29, 41, 29
Role: physician/nurse specialist, caregiver, psychologist, allied health care professional, psychologic assistant (%)		N/A	5, 34, 22, 34, 5
Experience working with SPI(M)D (years, mean ± SD (min. – max.))		N/A	15 ± 11 (0.3 – 43)
Working with SPI(M)D: D, W, M (%)		N/A	61, 37, 2
Working with SPI(M)D + decline/dementia: D, W, M (%)		N/A	37, 44, 20
Family relationship: parent, sibling, legal representative (%)		25, 50, 25	N/A
Characteristics of relative with SPI(M)D:			
- Age (years, mean ± SD (min. – max.))		56 ± 13 (40 – 72)	N/A
- Presence of DS (%)		75	N/A
- Presence of multiple disabilities (%)		63	N/A
- Decline/dementia: yes, no, don't know (%)		75, 13, 13	N/A
- Living situation: at home, care organization, combination (%)		13, 63, 25	N/A

Percentages (rounded off to the nearest whole number without decimals) are calculated based on the total number of participants per group (column). The group of psychologists is composed of behavioral therapists who studied psychology or special needs education (in Dutch: orthopedagogiek). Occupational therapists, physiotherapists and speech therapists were categorized as allied health care professionals. Abbreviations: D, daily; DS, Down syndrome; hbo, higher vocational education; M, monthly; mbo, intermediate vocational education; N/A, not applicable; SPI(M)D, severe/profound intellectual (and multiple) disabilities; W, weekly; wo, higher education.

In the focus groups, participants responded to the three questions. Answers are presented below as descriptions of categories and (sub)themes (question 1/ question 3) or by using a categorization matrix (question 2).

Question 1: Why is it important to know if an individual with SPI(M)D has dementia?

Thematic analysis revealed two themes (Figure 2.1): understanding and the ability to make informed choices.

Theme 1.1: Understanding
Participants stated that they want to know whether an individual has dementia in order to be able to explain problematic behavior.

> ***Psychologist G.:*** *"I am dealing with a man with a severe intellectual disability who is also becoming demented. He tends to display behavior that is very difficult to understand. Because we know that he is also becoming demented (…) we are better able to comprehend this behavior, and there is much more sympathy for it."*

Theme 1.2: Making informed choices
When we categorized the codes and divided categories into (sub) themes, it soon became clear that the majority of reasons were more or less related to the ability to make choices. Firstly, it was reported that choices concerned supporting care, for example, adjusting the aims of support, the way support is provided and the way of contact and interaction. Being aware that an individual also has dementia enables participants, for instance, to choose between an activating, development-oriented way of being supportive and a less development-oriented approach aimed at monitoring the dementia process and putting emphasis on comfort and maintaining skills. A similar consideration was reported regarding the choice between a behavioral way of being supportive, in which a client is talked to about and persuaded to change their conduct ('correcting' the person), and a more monitoring way of being supportive based on the fact that behavioral changes are caused by dementia ('following' the person).

> ***Psychologist X.:*** *"[Dementia] means that another approach must be used, in which we do not persuade [clients] to change their behavior but try to distract them and offer something else."*

> ***Caregiver C.:*** *"When do you continue to stimulate and when are you taking over? To decide on this, you have to observe someone all day long: what is the client able to do? (…) it is easier to accept that tasks must be taken over from a client who is becoming demented. In that case, you no longer persist in stimulating and assuming that the client is able to do the tasks."*

In addition to choices about support, choices about (medical) treatment were also consistently reported, for example, choices about adjusting a treatment plan and medication use.

> ***Caregiver E.:*** *"We have a client of whom we are not sure whether she suffers from dementia or depression (…) If she suffers from depression, you may give antidepressants which may revive her. However, if she suffers from dementia, you will need to adjust your actions."*

The population with SPI(M)D is diverse and includes not only people with severe ID who are (somewhat) able to express themselves verbally and move independently, but also people with profound intellectual and multiple disabilities who are not able to talk and are fully dependent on a wheelchair. Some participants raised the question as to whether the label 'dementia' would actually change the treatment plan for people with the most severe disabilities. The higher the level of functioning, the more likely it seems that the support and treatment can be adjusted. However, the majority of participants stated that they also wanted to know whether individuals with the most severe disabilities have dementia. In addition to obtaining clarity, it was also mentioned that it is not only about the label but also the preceding thorough diagnostic process. Diagnosing dementia requires a proper (differential) diagnostic procedure. This may also prevent misdiagnosis, which may result in clients receiving the wrong treatment.

The third subtheme concerned management of expectations/perspective. Participants stated that the diagnosis of dementia allows for anticipating the future, for example, anticipating the course of the disease, (timely)

entering a conversation with family members to prepare them for what may come and making choices about palliative care and the end of life.

> ***Occupational therapist D.:*** *"The earlier you can discover it, the better. (...) If you know the prognosis, that mental as well as physical [decline] will occur, you can adjust your actions."*

> ***Psychologist E.:*** *"Someone with Alzheimer's disease is, of course, more likely to die sooner. It is uncertain what is going to happen, but I think it may give the family something to hold on to. It is not a pleasant prospect, but it gives you a realistic view of what can happen and the opportunity to inform people about that."*

In view of the prospects, discussions were held about doubts as to whether or not it is useful and necessary to further examine clients, considering the burden and added value of this, for instance.

> ***Father M.:*** *"What can you achieve with all these examinations, how burdensome are they? (...) And how will they explain to our daughter what these tests entail?"*

Organizational choices were the final identified subtheme. Participants put forward that recognizing dementia may contribute to expanding the number/hours of staff involved and intensifying multidisciplinary collaboration.

> ***Psychologist X.:*** *"You notice that for people with dementia more intensive collaboration is required. It is important that transfer of information takes place more often. Being more in touch with each other, what do we see, what do we hear, which behavior do we observe, in order to be able to adjust our approach."*

With regard to organizational choices, the dilemma of changing activities during the daytime and/or housing was repeatedly mentioned. Although some participants stated that moving to a suitable house as early as possible enables clients to get used to their new surroundings, others wondered whether you should still introduce changes when dealing with an individual with dementia.

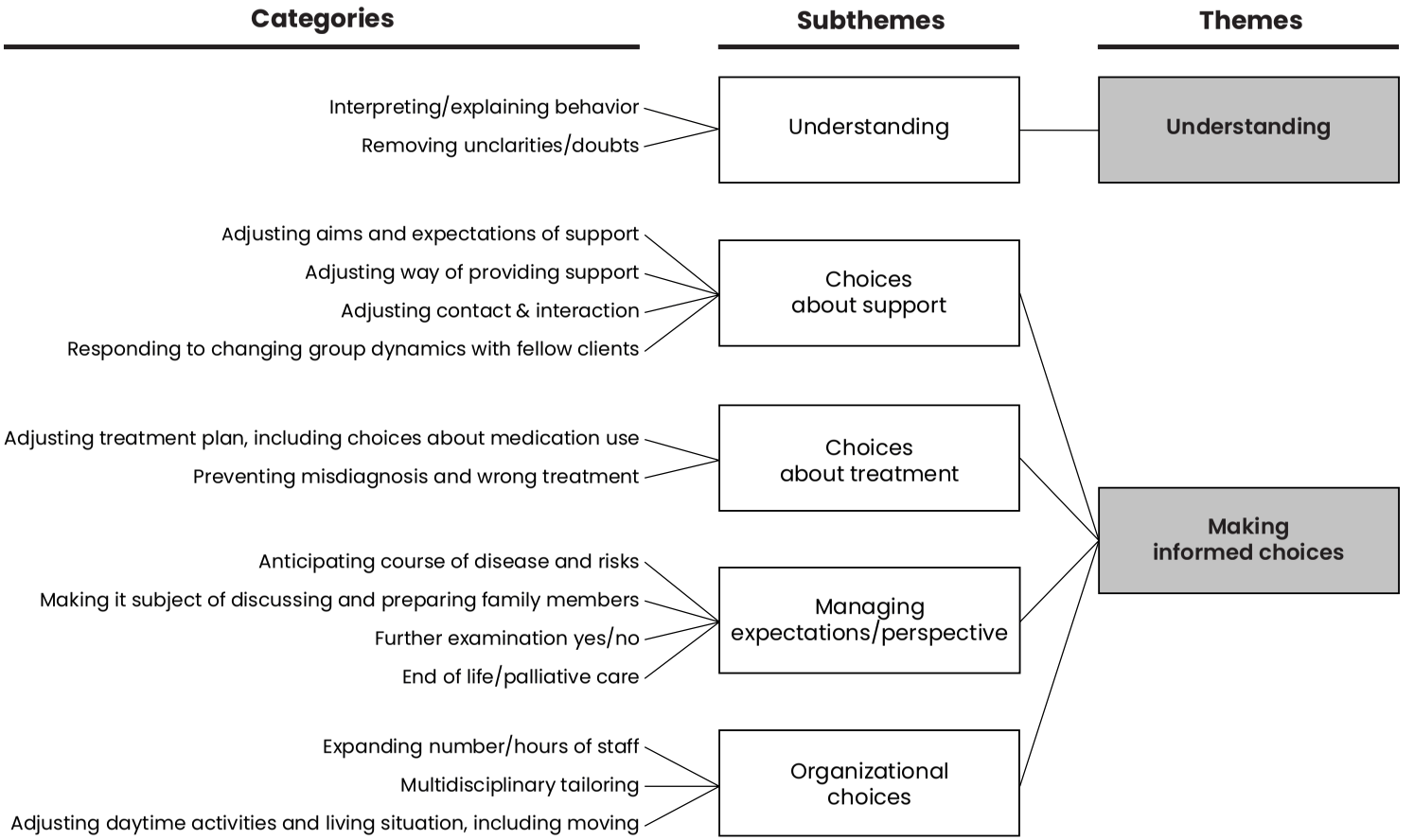


Figure 2.1 Thematic analysis of the answers to question 1: Why is it important to know if an individual with (SPI(M)D) has dementia? Figure based on the example in the methodological paper of Elo & Kyngäs (2008).

Question 2: Which dementia symptoms in people with SPI(M)D are recognized in daily practice?

The reported dementia symptoms were coded and subsequently categorized using a matrix (Table 2.2). Thematic analysis revealed that symptoms were generally observed in the context of nursing (for instance, bathing/showering, toilet use, getting dressed/undressed and external care), eating/drinking, mobility/transfers, communication and leisure activities. In addition, a category called 'context-independent' was created for symptoms of which the context was not sufficiently described or that did not seem to be not specifically related to particular contexts. Cognitive changes were often reported in relation to specific contexts such as nursing, eating/drinking and mobility/transfers. With regard to behavioral changes, symptoms of anxiety were clearly emphasized.

In this study, we identified symptoms based on practical experiences instead of neuropsychological assessment. As a result, a number of symptoms could not be uniformly classified because the description was not specific enough or the specific cause could not be ascertained within a focus group session. An example of this is the repeatedly mentioned loss of object permanence, that is, clients who always (have to) carry a certain object with them, for example, a little doll or a cuddly toy, suddenly lose interest in it. This may be due to amnesia (forgetting the object), agnosia (no longer recognizing the object), a decrease in compulsory behavior (the urge to always bring along the object has now subsided) or apathetic behavior (having lost interest). A few symptoms are, therefore, repeatedly described and italicized in Table 2.2.

Table 2.2 Categorization matrix structuring dementia symptoms provided by focus group participants.

		Contexts					Context-independent
		Nursing	Eating/drinking	Mobility/transfers	Communication	Leisure activities	
Cognitive changes	Amnesia	No longer understanding what is happening. Cannot remember that you went to the bathroom, where the toilet is, that you have to urinate in the toilet, that you have to go to the toilet, what a garment is for. ↓ trained to use the toilet at fixed moments	Inability to make choices. Cannot remember what a spoon/cup is for, what to do with food in your mouth, that you have to eat/drink, that you have to bring food with you when you go to day care, where food has been placed.	No longer understanding what is happening. Cannot remember that you were going somewhere, where to go on your bike, where you have put your rollator.	Cannot remember what has been said, what a caregiver is going to do, what has happened. Disguising behavior: making jokes to cover up what cannot be remembered.	Cannot remember what to do with a jigsaw piece, where a new sports venue is, which caregiver you need for what.	Cannot remember: what is happening around you, where you are in the room. ↓ *object permanence* ↑ *letting go of daily structure* ↑ putting objects in another place
	Aphasia			No longer able to tell that they do not know anymore.	Sound production changes (tone/ note), ↓ talking, ↑ mangling words, ↑ time needed to process what has been said.		
	Apraxia	No longer able to dry yourself off, put your arm into a sleeve, close your coat, get undressed without help.	No longer able to eat/drink properly, use cutlery, pick up/ use/put down a cup, continue to drink after stopping for a while, cut bread/ stick a fork in the bread, finish plate, leaves food in the mouth (↓ chewing/ swallowing)	No longer able to get into the car, use an aid, assume the right sitting position, stand up and take a step, transfer from chamber pot chair to regular chair, move forward. Freezing: not able to move anymore.		No longer able to do a jigsaw puzzle, exercise.	No longer able to perform actions/make movements, such as picking up an object from the floor. ↑ stagnation of actions

Table 2.2 Continued.

		Contexts					Context-independent
		Nursing	Eating/drinking	Mobility/transfers	Communication	Leisure activities	
Cognitive changes	Agnosia		↓ recognition of laid dinner table, spoon, cup, food		↓ *body awareness*, ↓ recognition of rooms, aids	↓ recognition of jigsaw	↓ recognition of caregiver, object, ↓ *object permanence*
	Cognitive other	↓ anticipating steps in getting dressed	↓ awareness of the proper order				↓ overview, ↓ awareness of proper order, ↓ quickness of response, ↓ confusion, ↑ disorientation in time/ space, colors are perceived differently, alternately bright and not bright.
Behavioral changes*	Anxious behavior	↑ fear of going to bed, fear before/ when taking a shower, fear of being touched during nursing (including reliving past sexual trauma during genital care). *Screaming during care.* Holding one's water.		↑ fear of transfers (hoist, rising, into wheelchair, reluctant to get out of bed, *Screaming during transfers.* Hesitant to walk/stand (sliding across the floor) or go through the door (fear of crossing thresholds).		↑ monitoring where someone is, ↑ *screaming*, ↑ being frightened, ↑ following caregiver, ↑ reassurance seeking	↑ fear of rain, ↑ fear of fellow clients, ↑ *screaming*, ↑ panic, ↑ despair, ↑ *lamenting*, ↑ *crying*, ↑ proximity seeking, ↑ tension, ↑ seeming unhappy, hesitant to let go, no longer feeling safe.
	Sleeping problems			↑ prowling about at night, ↑ wandering at night			Disturbance of day-night rhythm, ↑ waking up at night, ↑ sleeping/napping during the day
	Irritable behavior		↑ getting angry, ↑ throwing away food	↑ running away, ↑ complaining	↑ getting angry, ↑ yelling, ↑ grumbling, ↑ *screaming*, ↑ *groaning*		↑ getting angry, ↑ displeasure, ↑ touchy
	Obstinate behavior			No longer accepting aids, more/less running away.		↑ *groaning*	rigidity

Table 2.2 Continued.

		Contexts					Context-independent
		Nursing	Eating/drinking	Mobility/transfers	Communication	Leisure activities	
Behavioral changes*	Restless & Stereo-typical beahvior	↑ repeatedly getting undressed			↑ repeating questions		↓ *object permanence*, ↑ restlessness, ↑ speeding up daily program, increase/ decrease in compulsive acts
	Aggressive behavior						↑ biting, beating, kicking ↑ self-mutilation
	Apathetic behavior		↓ focus on eating, drinking	↓ interest in using aids	↑ distancing yourself from the group, ↑ withdrawn into yourself	↓ interest in favorite toy/ object, pleasant things, sports	↓ initiative, ↓ interest, ↓ *object permanence*, ↑ masklike face, ↑ *letting go of daily structure*
	Depressive behavior						↓ enjoying, ↑ emotional, ↑ mood changes, ↑ *crying*, ↑ *lamenting*, ↑ gloominess
	Psychotic behavior						Visual hallucinations
	Dis-inhibited behavior	↓ decorum: pulling up trousers in the hall, getting undressed in front of others	↓ decorum: eating with hands		↑ cuddling, ↑ kissing		Recurrence of problematic behavior
	Eating/ drinking behavior		↓ appetite, ↓ drinking, ↓ taste sensation, ↑ pica, changes in preferences: ↑ sweet, ↓ hot				

Table 2.2 Continued.

		Contexts					Context-independent
		Nursing	Eating/drinking	Mobility/transfers	Communication	Leisure activities	
Motor changes	Motor skills	↓ swinging, ↑ stiffness	↓ oral skills, ↓ holding spoon	↓ standing up, ↓ turning, ↓ walking, gait changes, ↑ wheelchair use			↓ muscle strength, ↓ motor skills, ↑ cramps
	Balance			↓ balance, falling			
	Body awareness	↓ *body awareness*		↓ *body awareness,* ↑ slanting			
	Swallo-wing		↓ swallowing function				
Medical comorbidities		↑ incontinence	↓ weight				↑ incontinence, ↑ pain, ↑ epilepsy, body temperature dysregulation

Symbols: ↓, decreased; ↑, increased; *, the categorization of dementia-related behavioral changes (or: behavioral and psychological symptoms of dementia) is based on the BPSD-DS scale (Dekker, Sacco, et al., 2018).

Question 3: What are training/information needs regarding dementia in people with SPI(M)D?

Thematic analysis revealed three overarching themes: 1) enhancement of training, 2) knowledge development and translation, and 2) organizational choices/policies (Figure 2.2). Participants defined information needs not only in terms of education and knowledge but also in terms of information about the client that they want to have. In addition, participants tended to describe their training needs particularly in terms of problems currently encountered. This was taken into account when phrasing the categories and (sub)themes, so that the question was properly answered.

Theme 3.1: Enhancement of education

Participants stated that the focus of their (preliminary) training had not or hardly been on dementia-related knowledge. Except for medicine, this goes for intermediate vocational education (mbo) received by most caregivers as well as higher vocational education (hbo) and higher education (wo).

> ***Caregiver Ch.:*** *"The intermediate vocational education that I received did not focus at all on the aging client with dementia, let alone dementia and severe intellectual disabilities. It was mainly focused on young people with mild intellectual disabilities."*

> ***Psychologist M.:*** *"Trainees receiving higher vocational education or higher education are completely unaware. When they start their apprenticeship, they think: I am going to administer a standard questionnaire, that is my diagnostic tool. Well, it does not take long to open their eyes."*

It was mentioned that caregivers, who have generally received intermediate vocational education, are expected to be among the first to (early) identify decline. Participants stated that caregivers have usually received agogic training, whereas dementia requires a more medical/ nursing background.

> ***Caregiver C.:*** *"I have colleagues who have only received intermediate vocational education in social care. Although that is very nice, they completely lack experience with dementia (…). It is also preferable to have some individual healthcare or nursing (…) background."*

> ***Physician B:*** *"Medical school focuses a lot on dementia. However, we depend on [information provided by] caregivers. Therefore, they must be fully aware of what they are expected to observe."*

Participants discussed whether specific knowledge about dementia in people with ID, and SPI(M)D in particular, can be (partly) included in (preliminary) training, or whether a specific module about dementia in people with SPI(M)D is more suitable. In addition, participants expressed their needs in terms of training content, including a link with existing methods in intellectual disability care and greater emphasis on observation techniques, such as the repeatedly reported need, when dealing with this population, to pay attention to subtle changes of a client's—often limited—specific functions.

> ***Psychologist R.:*** *"It is necessary to meticulously observe what (…) could previously be accomplished, but not anymore. (…) When dealing with profound intellectual disabilities in particular, these features are key (…) it is advisable to focus on these very critical features. (…) In my opinion, this is a crucial part that is, unfortunately, really lacking."*

With regard to learning formats, participants mentioned that training should be practice-based (i.e., concrete, easily manageable) and that it should be possible to learn from colleagues' experiences and by doing experience-based exercises. It was considered desirable to combine various learning formats and to present information in an inviting, for example, visual, manner. It was also stated that e-learnings do not provide the perfect solution, and it is advisable to combine new learning formats with in-person meetings.

Theme 3.2: Knowledge development and translation

Participants stated that information about dementia in people with SPI(M)D is lacking. More research is needed into, for instance, the development of (standardized) diagnostic tools suitable for this population, such as dementia questionnaires and the application of video observations to monitor decline.

> ***Caregiver H.:*** *"I think that the lack of training programs or additional courses is due to the fact that there is not enough information available about dementia in intellectual disability care. That is the heart of the problem."*

> ***Psychologist E.:*** *"I would like to have some tools, e.g., a questionnaire, (...) instead of the process only taking place in my head (...) I would like to have a standardized tool to help me examine and monitor clients."*

In addition to developing new knowledge, participants stated that they would like to see available knowledge being made more accessible by joining forces more and promoting collaboration between care organizations to prevent them from reinventing the wheel independently from each other. It was also considered desirable that information about dementia in the general population and in people with mild/moderate ID should be translated to SPI(M)D, if possible.

> ***Legal representative F.:*** *"[In regular elderly care] the development of materials regarding dementia is much more advanced. It is not realized that the same approach can be used in people with intellectual disabilities."*

> ***Speech therapist M.:*** *"I think that many organizations set up [training programs] themselves. However, it may be advisable to combine all these into one new program."*

Theme 3.3: Organizational choices/policies

Increasing the level of knowledge also depends on choices made by care organizations. Participants stated that time and money should be made available to take courses, and that such courses are often optional and without (many) obligations. It was also mentioned that specialized staff can be of added value to an organization.

> ***Caregiver H.:*** *"I regret the fact that going to specialize is not really rewarded. I think that providing a reward might be a way to stimulate staff a little bit more."*

With regard to information needs, it was stated that it is important to systematically monitor clients using standardized methods to improve the transferability of client information. Improvement of multidisciplinary collaboration (also with family members) was emphasized, as well as the importance of reducing staff changes to prevent the loss of knowledge and experience.

> ***Speech therapist A.:*** *"It is always important to document all information gathered about a client. (…) It may be very difficult to make a comparison with the client's previous situation, let's say two years ago, if there are now other caregivers working. (…) Has the situation really deteriorated or got worse?"*
>
> ***Sister M.:*** *"Sometimes, I only talked to the caregiver during the annual evaluation meeting. Then I used to think: where is the doctor? Where is the psychologist? Where is the team leader?"*
>
> ***Psychologist G.:*** *"It is very important to adopt a multidisciplinary approach. That it is not only the caregiver's responsibility (…) There are so many perspectives that may help to make a good diagnosis."*

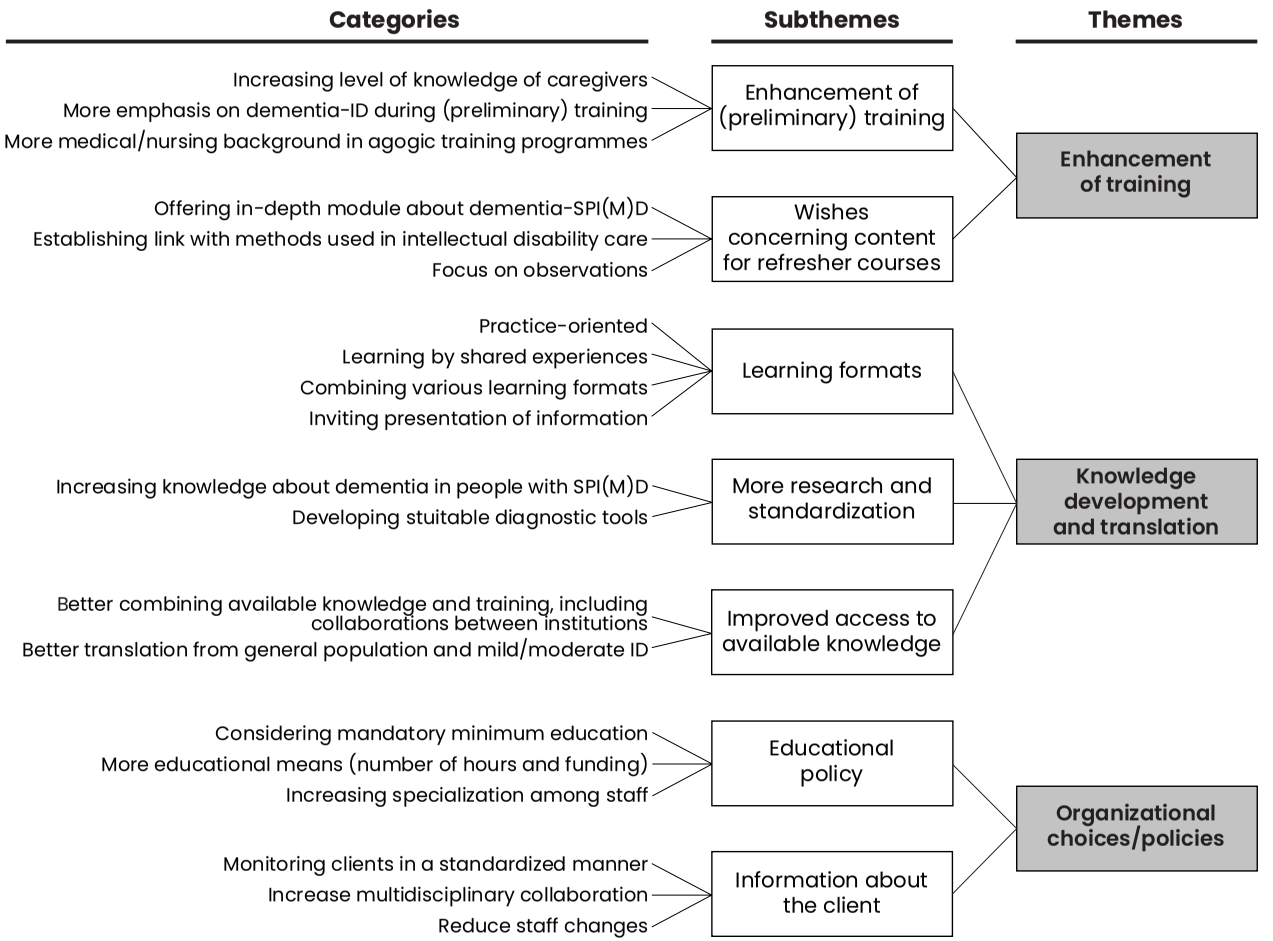


Figure 2.2 Thematic analysis of the answers to question 3: What are training/information needs regarding dementia in people with SPI(M)D? Figure based on the example in the methodological paper of Elo & Kyngäs (2008). Abbreviations: ID, intellectual disabilities; SPI(M)D, severe/profound intellectual (and multiple) disabilities.

Discussion

In this explorative focus group study on dementia in people with SPI(M)D, we examined the 1) relevance of the diagnosis, 2) symptoms, and 3) training/information needs. Thematic analysis revealed that participants want to know whether a person has dementia for a better understanding and to be able to make informed choices. The reported dementia symptoms were categorized using a matrix, in which cognitive changes and behavioral changes were the most prominent. With regard to education, participants expressed their need for enhancement of education, more knowledge development and translation and supportive organizational choices/policies.

The results concerning relevance (question 1) are consistent with Australian research that showed that caregivers often struggle to understand whether behavioral changes are deliberate and people with ID can be called to account for them or whether these are dementia-related and, therefore, beyond the person's control (Iacono et al., 2014). Knowing whether someone has dementia enables earlier intervention and more appropriate support and resources (Chapman et al., 2018). A few participants wondered whether the label 'dementia' would actually change care/treatment for people with the most severe disabilities since they already receive care all their life. Importantly, although dementia cannot (yet) be prevented or cured, non-pharmacological, psychosocial interventions and (behavior-modifying) medication may be used to improve the person's well-being and quality of life (Bessey & Walaszek, 2019; Keller et al., 2016; MacDonald & Summers, 2020). Without proper diagnosis, treatment might be withheld or the wrong treatment may be provided. Participants disagreed about the use of and need for further testing, for instance in a clinical setting. Consistent with previous research (Chapman et al., 2018), concerns were expressed about the impact on the person.

Not only were choices regarding supportive care and treatment emphasized, but also organizational choices. Whether or not it is advisable to move house was subject of discussion, which was also addressed in scientific literature (Chaput, 2003; Heller et al., 2018; Llewellyn, 2011). To enable an individual with dementia to continue to live at a familiar location, adjustments must be made to the house, supportive care and interactions (Chapman et al., 2018; Janicki et al., 2005; Watchman, 2003). According to

focus group participants, a timely diagnosis helps to achieve this.

Although the relevance of the diagnosis was frequently emphasized (question 1), answers to question 3 also showed that the knowledge level needed to identify dementia must be vastly improved. The (timely) identification of signs is almost always the task of caregivers. However, participants stated that there is much room for improvement of caregivers' level of knowledge and expertise, as also found in a review article (Cleary & Doody, 2017b). Limited knowledge and the early signs of dementia not being identified may result in delayed dementia diagnosis and subsequent decision-making delays (Cleary & Doody, 2017b), for example, regarding supportive care and treatment. Specific training programs can improve knowledge, understanding, trust and quality of care (Chapman et al., 2018; Cleary & Doody, 2017b). The focus group sessions revealed that this is desirable not only for caregivers but also for professionals from other disciplines that play an important role in the diagnostic process.

The lack of scientific knowledge and dedicated diagnostic tools for dementia in people with SPI(M)D (Elliott-King et al., 2016; Esbensen et al., 2017; Wissing, Ulgiati, et al., 2022) on which to base training programs, makes it more difficult to improve care professionals' knowledge and expertise. As a result, care professionals involved with SPI(M)D have to rely on their practice-based observations, experiences and anecdotal knowledge. Participants emphasized the necessity to develop new knowledge and suitable diagnostic tools. It was frequently stated that knowledge should be made more accessible and closer collaboration is needed between regular elderly care and intellectual disability care, a perspective promoted in literature as well (Heller et al., 2018; Iacono et al., 2014). Participants also emphasized that knowledge development and training cannot take place without making organizational choices and adopting policies focused on dementia in people with ID, such attributing more time to caregivers for individuals with dementia (Janicki et al., 2005; Mccarron et al., 2005).

To facilitate the development of new knowledge and diagnostic tools, participants shared their practice-based symptoms of dementia in people with SPI(M)D. In their daily work, they observe decline across different domains. During these observations, subtle signs are key. The categorization matrix revealed that cognitive decline and a variety of behavioral changes were observed in particular, which may or may not be

associated with specific everyday life situations. Remarkably, participants mentioned a considerable number of cognitive changes, whereas a British study concluded that deterioration in everyday functional skills is more indicative of dementia in people with more severe ID (Jamieson-Craig et al., 2010). This apparent discrepancy can be explained by the categorization matrix. By dividing symptoms into deductively ascertained themes (cognitive, behavioral and motor changes; rows) and by linking these to specific contexts inductively obtained from the transcripts (columns), it appears that underlying cognitive changes can be observed in contexts of activities of daily living. Cognitive decline in people with SPI(M)D appears to be mainly apparent during activities of daily living, such as nursing, eating/drinking and mobility/transfers.

Strengths

To the best of our knowledge, this study is the first to specifically address dementia in people with SPI(M)D. This focus group research is strong because of its solid study design, large number of participants (N=49), multidisciplinary composition and representativeness of intellectual disability care in The Netherlands. Although care professionals in this study were primarily female, this resembles the overrepresentation of women in health care. Indeed, 80% of employees in the Dutch intellectual disability care sector is female (Vereniging Gehandicaptenzorg Nederland, 2019). Within the care organizations, participants were purposefully selected based on their practice-based experience with decline/dementia in people with SPI(M)D. The structured overview of symptoms is an important first step towards an evidence-based approach to the diagnosis of dementia in this vulnerable, severely disabled population. This study also provides important information about the relevance of a dementia diagnosis and the training needs of staff and family members. Although training needs were asked in the context of dementia, the (sub) themes emerging from the provided answers (Figure 2.2) may appear to be applicable to other diseases as well, suggesting that these needs are of essence in good care for people with SPI(M)D in general. For most (sub)themes, the underlying categories specify the needs in the context of dementia.

Limitations

Considering the multidisciplinary composition, a first limitation was the fact that participation of a physician or nurse specialist could not be achieved in each focus group. Whereas one unspecialized physician and one nurse

specialist participated, involvement of specialized intellectual disability physicians would have been desirable.

Secondly, although the focus on people with SPI(M)D was continuously emphasized, care professionals may have referred to some signs of dementia in people with mild/moderate ID (question 2) because they often provide care to people with different levels of functioning. It is also important to mention the considerable heterogenicity of the SPI(M)D population. A number of symptoms, particularly a decline in speech and ability to walk in people with severe ID, were not widely recognized by care professionals who work with clients who are non-verbal, profoundly disabled and totally dependent on wheelchairs. This underlines the importance of identifying changes within a person by assessing how his/her functioning develops over time. Therefore, a timely baseline measurement of the level of functioning, that is, prior to the occurrence of decline, is essential (Keller et al., 2016).

Thirdly, based on symptoms and contexts mentioned in the transcripts, the categorization matrix — a simplification of the real situation — was created. However, classification of symptoms was not always straightforward because a detailed description or contextualization was missing, symptoms appeared to fit into more than one category or a specific cause could not be ascertained within the focus group session. Nevertheless, this is indicative of the struggle faced by participants in daily practice.

A fourth limitation concerned the sudden ending of the audio recording of one of the four focus group sessions approximately 15 min before the session actually ended. However, the fact that some data (question 3) were lost had no impact on saturation.

Finally, in literature there is an ongoing discussion about the necessity to report inter-rater reliability in qualitative research. Braun & Clarke (2013) argue that reliability is not an appropriate criterion for judging qualitative work, because there is no single true meaning inherent in the data. Instead, to enhance trustworthiness of the analysis, the analysis process and the results should be described in sufficient detail and it is advised to provide authentic citations (Braun & Clarke, 2006, 2013; Elo & Kyngäs, 2008) like in this study.

Future implications

Scientific literature on dementia in people with SPI(M)D has been scarce until now (Wissing, Ulgiati, et al., 2022), although dementia-related decline on top of severe/profound ID is very complex. Here, the results clearly emphasize the relevance of research on dementia in this population, show obvious practice-based needs for more knowledge and suitable diagnostic tools and provide direction for further research. More in-depth studies of symptoms, for example, clinical records analysis and interviews with experienced care professionals to obtain a detailed overview of their practice-based experiences are needed. Finally, it is important to focus more on dementia in people with SPI(M)D in training programs. Development of training products related to this topic must be tailored to the wishes and needs in daily practice.

Conclusion

This focus group study examined the 1) relevance of the diagnosis, 2) symptoms and 3) training/information needs. It is important to identify dementia (early on) in people with SPI(M)D to be able to make informed choices. To be able to diagnose dementia, a better understanding of dementia symptoms in this population is necessary. This focus group study paves the way for further study of symptoms. In training programs, dementia in people with SPI(M)D should be incorporated and the provided information should be tailored to practice-based wishes. People with SPI(M)D are not or hardly able to express the occurrence of deterioration and strongly depend on care professionals. Therefore, improving the knowledge level of these professionals helps to (better) timely identify dementia. As a result, the client's changing wishes and needs can be better responded to by making informed choices.

Chapter 3

A systematic literature review of observable symptoms

Wissing, M. B. G., Ulgiati, A. M., Hobbelen, J. S. M., De Deyn, P. P., Waninge, A., & Dekker, A. D. (2022). The neglected puzzle of dementia in people with severe/profound intellectual disabilities: A systematic literature review of observable symptoms. *Journal of Applied Research in Intellectual Disabilities, 35*(1), 24–45.

Dutch version of the articles presented in chapters 2 & 3 |
Nederlandse versie van de artikelen gepresenteerd in hoofdstukken 2 & 3

Dekker, A. D., Wissing, M. B. G., Ulgiati, A. M., Bijl, B., van Gool, G., Groen, M. R., Grootendorst, E. S., van der Wal, I. A., Hobbelen, J. S. M., De Deyn, P. P. & Waninge, A. (2021). Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen: onderzoek. *NTZ*: *Nederlands Tijdschrift voor de Zorg aan mensen met een verstandelijke beperking*, *47*(4), 139–159.

Abstract

Introduction: Dementia is increasingly prevalent in people with severe/profound (and multiple) intellectual disabilities (SPI(M)D). However, early recognizing and diagnosing dementia in this population is complex. This study aimed to identify observable dementia symptoms in adults with SPI(M)D in available literature.

Methods: 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 SPI(M)D and dementia/aging.

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

Conclusion: Only a very limited number of studies reported symptoms ascribed to dementia in adults with SPI(M)D. Given the complexity of recognizing and diagnosing dementia, dedicated studies are required to unravel the natural history of dementia in this population.

Introduction

The world's population, including people with intellectual disabilities (ID), is aging. Life expectancy of people with ID has increased even more than in the general population (Coppus, 2013; E. 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 (DS) (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 DS (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 ID not due to DS vary across studies (Krinsky-McHale & Silverman, 2013).

A pre-existing intellectual disability, (lifelong) characteristic behavior and comorbidities which may mimic dementia symptoms are complicating factors in diagnosing dementia in people with ID (Esbensen et al., 2017; McKenzie et al., 2018; Sheehan, Sinai, et al., 2015). In fact, the more severe the level of intellectual disability, the more difficult diagnosing dementia becomes (E. Evans et al., 2013). Hence, diagnosing dementia is especially challenging in people with SPI(M)D, that is, IQ score below 35 (E. 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 (E. 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 SPI(M)D 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 ADL, because specific skills were not attained (Sheehan, Sinai, et al., 2015). Consequently, skills that 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 SPI(M)D are largely reliant on caregivers/relatives

for recognizing observable dementia symptoms (McKenzie et al., 2018).

Evidently, diagnosing dementia in people with SPI(M)D is a complex puzzle, which necessitates a proper understanding of its presentation in this population. Early recognizing and diagnosing dementia allows care professionals and relatives to make informed choices about adaptation of caregiving, support and treatment (Dekker, Wissing, et al., 2021a). However, caregivers indicate to have limited knowledge of the presentation of dementia in people with ID (Herron et al., 2015; Whitehouse et al., 2000). Limited knowledge about symptoms may cause early signs not to be recognized, resulting in a (too) late diagnosis or no diagnosis at all (Cleary & Doody, 2017b). 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 of (early) dementia symptoms, especially in those with SPI(M)D is essential to provide appropriate support and care in order to maintain quality of life (Dekker, Wissing, et al., 2021a; Janicki, 2011).

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 SPI(M)D. Given the diagnostic complexity, it is expected that dementia is often underdiagnosed. Therefore, we also reviewed aging literature describing changes in cognitive functioning and/or behavioral and psychological alterations without explicitly referring to dementia.

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 SPI(M)D in the scarce literature.

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 SPI(M)D, using a broad range of synonyms for ID 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 SPI(M)D 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 SPI(M)D. The second cluster included search terms for dementia, for example, Alzheimer, and major/minor cognitive impairment as well as terms related to aging, such as decline, changes, progressive, and 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.

Study selection

To be included, studies had to describe (potential) dementia symptoms in people with SPI(M)D 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 SPI(M)D. Exclusion criteria: studies in the general population (without ID), people with mild or moderate ID, age under 30 years, studies focusing on persons with SPI(M)D 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.). Afterward, 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 3.1 schematically depicts the selection procedure.

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, and main symptomatic results (Table 3.1). 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, 2022). 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 ADL, accompanied by behavioral and psychological symptoms of dementia (BPSD), (Dekker, Strydom, et al., 2015; Finkel, 2000). Therefore, in aging studies only reported cognitive changes and behavioral and psychological alterations were considered to be potential dementia symptoms. In the present study, (potential) dementia symptoms were categorized 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 3.2).

Results

The literature search yielded a total of 668 hits in three databases. Deduplication resulted in 625 unique records (Figure 3.1). Based on title 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 3.1). In total, 11 studies met inclusion criteria. Table 3.1 provides a detailed overview of characteristics and main results.

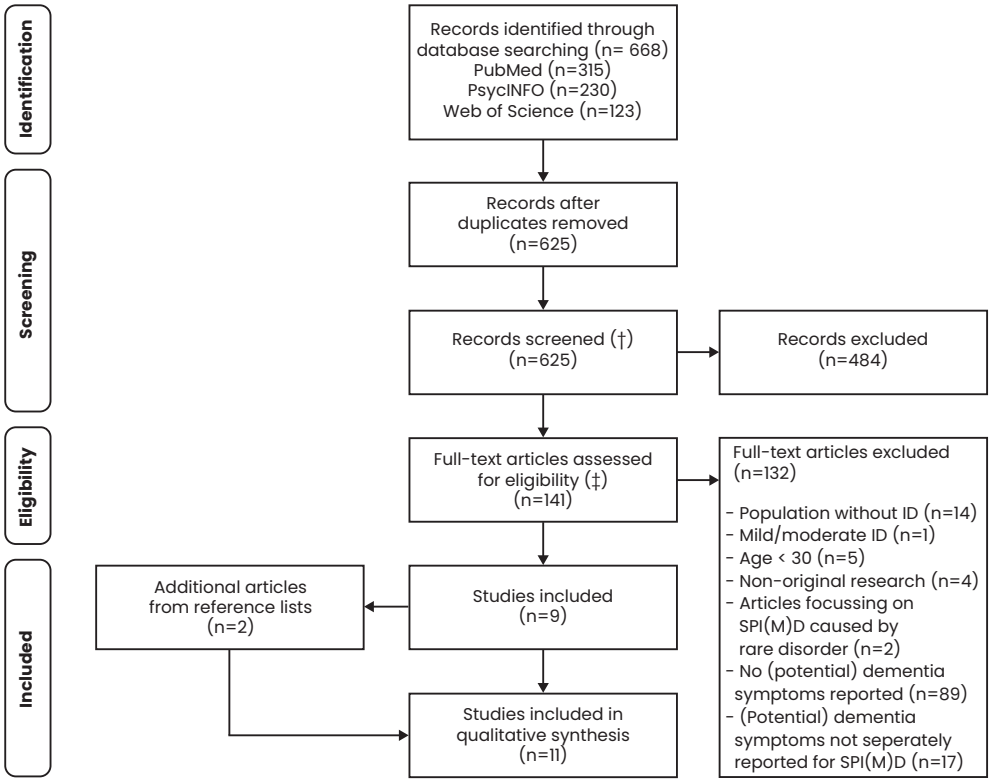


Figure 3.1 Flowchart of the study selection process. †, inter-rater agreement in 15% of deduplicated records; ‡, inter-rater of full-text screening. Abbreviations: ID, intellectual disabilities; SPI(M)D, severe/profound intellectual disabilities.

Symptoms in dementia studies

The first study to report dementia symptoms in adults with SPI(M)D was published by Reid and Aungle (1974). Among 155 persons with ID, two individuals with a severe intellectual disability were diagnosed with dementia based on clinical (re)assessment (Table 3.1). For a 52-year-old man with DS, reported dementia symptoms were 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 etiology. 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.

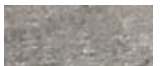
Table 3.1 Characteristics and main symptomatic results of included studies.

	Reference	Study population(s)	ID classification	Assessment of dementia/age-related changes that are potential dementia symptoms	Main symptomatic results
Studies of dementia symptoms	Reid and Aungle (1974)	155 ID, 63 ♂, 49-85 yrs - 1 severe ID (DS) + dementia, ♂, 52 yrs - 1 severe ID + dementia, ♀, 62 yrs	GMD	Clinical (re)assessment: - Clinical records - Informant interviews - Physical examination	↓ activity, ↓ speech, ↓ self-care skills, personality change, disturbed sleep, incontinence, late-onset epilepsy (case 1) ↓ self-care skills, ↓ speech, personality change, asymmetrical spastic signs in limbs (case 2)
	Day (1985)	357 ID, ≥ 40 yrs - 2 severe ID + late-onset dementia	ICD-9	- Clinical records	↓ social skills, ↓ personal habits, behavioral disturbances, memory impairments: forgetfulness, confusion
	Evenhuis (1990)	17 DS, 7 ♂, 45-63 yrs - 5 severe ID (DS) + AD, 2 ♂, 45-60 yrs	Severe ID: IQ 25-35	Non-standardized clinical assessment: - Observations - Informant interviews - Physical examination	↓ 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)
	Duggan et al. (1996)	12 ID + dementia, 3 ♂, 47-77 yrs - 1 severe ID (DS), ♀, 59 yrs	ICD-9	- Psychiatric and clinical records - Informant interviews (BHI) - Physical examination	↓ amount of walking, ↓ food intake, ↓ drinking, weight loss, inappropriate food placing, wrong utensils use, pica, aimless walking hypermetamorphosis
	Burt et al. (1998)	70 DS, 22-60 yrs - 2 severe ID (DS) + dementia - 3 profound ID (DS) + dementia	Severe: IQ 21-36 Profound: no basal on LIPS	- Direct neuropsychological evaluation (DF & SR, GPT, DTVMI, PD, PPVT-R, LIPS) - Informant interviews (VABS, DQMRP, RSMB, DSI, MAS)	↓ cognitive functioning, ↓ memory, ↓ everyday functioning, emotional/behavioral changes
	Määttä et al. (2006)	129 DS, 76 ♂, 0-67 yrs - 1 moderate/severe ID (DS) + AD, ♂, 51 yrs	ICD-10	- Clinical records	↓ self-care skills, ↑ forgetfulness, ↑ irritability, withdrawal, occasional aggressive outburst, late-onset epilepsy
	Margallo-Lana et al. (2007)	92 DS, 63 ♂, 20-76 yrs - 6 profound ID (DS) + dementia	Unspecified	- Informant interviews - Clinical records	Non-cognitive symptoms: ↓ everyday skills, ↓ mobility, ↓ interest in surroundings, uncharacteristic inappropriate behavior, daytime sleepiness, wandering, getting lost, incontinence

Table 3.1 Continued.

	Reference	Study population(s)	ID classification	Assessment of dementia/age-related changes that are potential dementia symptoms	Main symptomatic results
	Sauna-Aho et al. (2018)	128 DS, WS, FxS, 85 ♂, 36-85 yrs - 2 profound ID + dementia, 50 yrs (DS), 61 yrs (FxS) - 1 profound ID + vascular dementia, 53 yrs (WS)	Clinical records	- Clinical records - BPSLD - Brain imaging	Weight change, loss of energy, sleep disorder
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 behavior	Non-DS severe ID: = psychological problems, ↓ challenging behavior DS severe ID: ↑ psychological problems, = challenging behavior
	Cherry et al. (1997)	168 SPI(M)D - 84 young, 46 ♂, 20-29 yrs - 84 old, 45 ♂, 60-79 yrs	AAMD	DASH	Compared to younger SPI(M)D, older SPI(M)D showed: ↑ durations anxiety, inappropriate sexual behavior, impulse control problems ↑ severity anxiety, stereotypies/tics, impulse control problems Result in relation to ID level: ↑ duration self-injury behavior 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 seniors, 1.4 ♂/♀, 50-68 yrs	Clinical records	Clinical records	↑ withdrawal, ↑ intermittent screaming, ↑ intermittent crying, ↑ agitation, ↑ self-aggressivity, ↓ language, ↓ posture-motor ability, ↓ coordination, ↓ sociability, = aggressivity, = stereotypies, = mericysm, = sleep disorders

Symbols: ↓, decreased; ↑, increased; =, remained stable, ♂, male; ♀, female. Abbreviations: AAMD, American Association on Mental Deficiency; AD, Alzheimer's disease dementia; BHI, Behaviour History Inventory; BPSLD, British Present Psychiatric State-Learning Disabilities assessment; DASH, Diagnostic Assessment for the Severely Handicapped; 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; GMP, Glossary of Mental Disorder; GPT, Grooved Pegboard Test; GQ, Gerontological Questionnaire subscales; ICD, International Classification of Diseases and Related Health Problems; ID, intellectual disability; IQ, Intelligence Quotient; LIPS, Leiter International Performance Scale; MAS, Mood Assessment Scale for mental retardation; PD, Picture Description test; PPVT-R, Peabody Picture Vocabulary Test-Revised; RSMB, Reiss Screen for Maladaptive Behavior; SPI(M)D, severe/profound intellectual disabilities; VABS, Vineland Adaptive Behavior Scale, WS, Williams syndrome; yrs, years.



Subsequently, Day, (1985) reported the prevalence of dementia in 357 people with ID. A diagnosis of late-onset dementia was reported in clinical records 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 behavioral disturbances, which were not further specified for these two persons.

Evenhuis (1990) conducted a prospective longitudinal study in seventeen individuals with DS of whom five had a severe intellectual disability. Dementia was suspected in these five individuals based on progressive decline in ADL. 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 3.1). 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 behavioral 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 behavioral changes in a population with ID 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 Behavioral History Inventory, this 59-year-old woman with DS was described as showing changes in walking behavior, 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 compulsion to touch furniture (Table 3.1).

Similar to Evenhuis (1990), also Burt et al. (1998)used a prospective longitudinal approach for studying dementia in 70 people with DS, of whom at entry 16 had a severe intellectual disability and had 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 3.1). 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 behavioral changes.

Furthermore, Määttä et al. (2006) focused on mental health and adaptive behavior of 129 people with DS (Table 3.1). One case of an adult with a moderate–severe intellectual disability and Alzheimer's disease was described. Observations during the past five years revealed increasing forgetfulness, irritability, withdrawal, occasional aggressive outbursts, and declining self-care skills. In line with Reid and Aungle (1974), late-onset epilepsy was found, here present for two and a half years.

Margallo-Lana et al. (2007) studied the extent of cognitive changes and dementia in people with DS, emphasizing that clinically diagnosing dementia in those with more severe ID could be problematic. For six persons with profound ID, the diagnosis of dementia was based on non-cognitive dementia characteristics such as behavioral symptoms like loss of interest in surroundings, daytime sleepiness, and uncharacteristic inappropriate behavior. Further non-cognitive signs were a decline in everyday skills, wandering and getting lost, as well as decreasing mobility and incontinence (Table 3.1).

The last study reporting dementia symptoms in people with SPI(M)D was published by Sauna-Aho et al. (2018). Using the British Present and Psychiatric State learning Disabilities assessment dementia was screened in 128 individuals consisting of subjects with DS (n=62), Williams syndrome (n=22), and Fragile X syndrome (n=44). A total of 50 individuals had a severe intellectual disability, and 3 had a profound intellectual disability.

However, specific dementia symptoms were only separately reported for the three individuals with profound ID (one DS, one Williams, one Fragile X syndrome), namely: weight change, loss of energy and sleep disorder (Table 3.1).

Potential dementia symptoms in aging studies

It is expected that dementia is often underdiagnosed in people with SPI(M)D due to the complexity of diagnosing dementia in this population. Therefore, changes in cognitive functioning and/or behavioral 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 SPI(M)D was published by Haveman et al. (1994). They evaluated challenging behavior and psychological problems in 1580 persons with ID according to age, level of intellectual disability and presence of DS. Specifically, for people with a severe intellectual disability without DS, they found less challenging behavior with advanced age, whereas psychological problems were evenly distributed. In people with DS, elderly with severe ID had more psychological problems (Table 3.1). The authors concluded that these psychological problems could be explained as symptoms of dementia, given that 39 of the 85 persons with DS (mild and severe ID) 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 SPI(M)D (Table 3.1). 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 behavior, 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 ID were compared. Older persons with a profound intellectual disability had longer durations of self-injury behavior 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 aging 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 behavioral 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.

Table 3.2 Overview of (potential) dementia symptoms in people with SPI(M)D.

Categories	Dementia symptoms	Potential dementia symptoms
Cognitive changes	↓ speech[1,3], ↓ social skills[2], ↓ cognitive functioning[5], ↓ memory[2,5], forgetfulness[2,6], confusion[2], aimless walking[4], wandering[7], getting lost[7], ↓ personal habits[2], apraxia[3], inappropriate food placing[4], wrong utensils use[4]	↓ language[11], ↓ sociability[11], ↓ posture-motor ability[11], ↓ coordination[11]
Behavioral and psychological changes	aggression[3,6], withdrawal[3,6], apathy[3], ↓ interest in surroundings[7], irritability[3,6], daytime sleepiness[3,7], disturbed sleep[1], sleep disorder[8], ↓ food intake[4], ↓ drinking[4], pica[4], uncharacteristic inappropriate behavior[7], hypermetamorphosis[4], personality change[1], emotional/behavioral changes[2,5]	↑ self-aggressivity[11], ↑ duration self-injury behavior (PID)[10], ↑ withdrawal[11], ↑ agitation[11], ↑ intermittent screaming[11], ↑ intermittent crying[11], ↑ durations anxiety[10], ↑ durations inappropriate sexual behavior[10], ↑ durations impulse control problems[10], ↑ severity anxiety[10], ↑ severity stereotypies/tics[10], ↑ severity impulse control problems[10], ↑ psychological problems (DS)[9]
Changes in ADL	↓ self-care skills[1,3,6], ↓ everyday functioning/skills[5,7], ↓ activity[1]	
Neurological and other physical changes	incontinence[1,3,7], (late-onset) epilepsy[1,3,6], weight change/loss[4,8], ↓ energy[8], ↓ amount of walking[4], ↓ mobility[7], ↓ gait[3], chair/bedridden[3], asymmetrical spastic signs in limbs[1], muscle hypertonia[3], myoclonus[3]	

Symbols: ↓, decreased; ↑, increased. Abbreviations: ADL, activities of daily living; DS, Down syndrome; PID, profound intellectual disability. 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).

Synthesis of results

In summary, eight studies reported dementia symptoms for in total 25 adults with SPI(M)D and dementia. Of these 25 individuals, 20 had DS, 1 had Williams syndrome, 1 had Fragile X syndrome, and for 3 individuals the etiology was unspecified. Additionally, three studies focusing on age-related changes in people with SPI(M)D found a decline in cognitive functioning and an increase in emergence of BPSD, which could potentially relate to dementia-related symptoms given the complexity of diagnosing dementia in this population. Table 3.2 provides an overview of reported

cognitive changes, BPSD, changes in the ability to perform ADL, and neurological and other physical changes.

Limitations of primary literature

The very limited number of studies that studied – in part – dementia in people with SPI(M)D 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 summarized in general, grouped according to the data extraction categories presented in Table 3.1.

Study population(s)

The first limitation concerned the small number of people with SPI(M)D for whom observable dementia symptoms were reported (ranging from n=1-6 in each dementia study). Secondly, the etiology 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 DS in the aging study of Haveman et al. (1994) and 474 persons in the aging 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 DS and the person with Fragile X syndrome had Alzheimer's disease dementia or vascular dementia Sauna-Aho et al. (2018).

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.

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 SPI(M)D varied across studies. Two

studies obtained diagnoses from clinical records 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 clinical records 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.

Discussion

To the best of our knowledge, this review is the first to systematically identify observable dementia symptoms in people with SPI(M)D 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 behavioral and psychological alteration in the context of aging were also included. Summarizing 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, particularly withdrawal and aggressiveness. Furthermore, specifically in those with dementia, skills necessary to perform ADL 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 ID (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). Two studies reported that specifically in those with profound ID, 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 ADL, making it complex to determine whether dementia-related cognitive impairments interfere with the ability to perform ADL. 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 aging studies all reported on BPSD. Four studies found symptoms of apathetic behavior 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 DS, in which apathy was found one of the most commonly observed BPSD symptoms (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a). Furthermore, in this study a substantial proportion of people with DS and Alzheimer's disease displayed an increased frequency of aggressive behavior (Dekker, Sacco et al., 2018; Dekker, Ulgiati, et al., 2021a). Similarly, two dementia studies included in this review reported aggression in three individuals with a severe intellectual disability, DS 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 aging studies can be signs of dementia in persons with SPI(M)D.

Furthermore, in those with a clinically and/or postmortem confirmed diagnosis of dementia, reported BPSD were irritability, alterations in eating/drinking behavior, and sleep problems. Irritability was observed particularly in individuals with a severe intellectual disability and DS (Evenhuis, 1990; Määttä et al., 2006), which is in line with results of other studies focusing on dementia symptoms in persons with DS (Lai & Williams, 1989; Moss & Patel, 1995). Moreover, Duggan et al., (1996) found alterations in eating and drinking behavior also specifically in a person with a severe intellectual disability and DS. This suggests that eating and drinking behavior is affected by dementia (Dekker, Sacco, et al., 2018; Dekker, Ulgiati, et al., 2021a). Additionally, individuals with SPI(M)D 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 ID (Van de Wouw et al., 2012). Nevertheless, they are important to consider given that they may aggravate cognitive

decline and BPSD (Dekker, Strydom, et al., 2015).

Besides emerging BPSD, the ability to perform ADL declined. Similar to Lai and Williams, (1989), three dementia studies reported losses of self-care skills in individuals with a severe intellectual disability and DS (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.

Strengths

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

Limitations

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

Given the complexity of diagnosing dementia in people with SPI(M)D, it is questionable whether early dementia symptoms were observed or whether observed symptoms were attributed to the intellectual disability, aging, 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 SPI(M)D (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 SPI(M)D. Given the functionalities of the databases PubMed, PsychINFO, and Web of Science, studies assessing a broad level of ID 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 ID (in the broadest sense) or persons with DS published in the last five years were potentially missed. No additional broad intellectual disability and DS studies reporting dementia symptoms in those with SPI(M)D were identified. This emphasizes the lack of focus on dementia in this population.

Future implications

This systematic review provides a first overview of observable dementia symptoms in people with SPI(M)D. 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 analyzing existing clinical records 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) recognizing and diagnosing dementia in this population. This enables care professionals and family members to adequately adapt daily care (Dekker, Wissing, et al., 2021a; Janicki, 2011). 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 (Dekker, Wissing, et al., 2021a; Janicki, 2011). Altogether, (early) diagnosis of dementia may contribute to the well-being of individuals with SPI(M)D.

Conclusion

Dementia in people with SPI(M)D 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 summarized observable symptoms in available literature. Despite the few small-sized studies, a range of dementia symptoms were identified, subdivided into cognitive decline (e.g., memory loss, forgetfulness, deterioration in speech, losses of social skills), decline in ADL (e.g., self-cares skills, everyday functioning/skills), BPSD (e.g., apathy, aggression, irritability, altered eating/drinking behavior) 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 SPI(M)D. 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 SPI(M)D, further establishing the natural history. This would enable (early) recognizing and diagnosing dementia which contributes to maintaining quality of life in people with SPI(M)D and dementia.

Chapter 4

Practice-based observations of symptoms identified through a survey and interviews

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Abstract

Introduction: Observable dementia symptoms are hardly studied in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D). Insight into symptomatology is needed for timely recognizing and diagnosing dementia. This study aimed to identify practice-based observations of dementia symptoms in this population.

Methods: Care professionals and family members were invited to complete a survey about symptoms. Quantitatively analyzed survey data were further deepened through semi-structured interviews with care professionals having vast experience in recognizing/diagnosing dementia in this population. Symptoms were categorized using a symptom matrix.

Results: Survey respondents and interviewees frequently observed a decline in activities of daily living (ADL) functioning and behavioral and psychological changes, like increased irritability, anxiety, apathy and decreased eating/drinking behavior. Cognitive symptoms were particularly recognized in persons with verbal communication and/or walking skills. To lesser extent motor changes and medical comorbidities were reported.

Conclusion: Increased insight into dementia symptoms contributes to developing a dedicated screening instrument for dementia in people with SPI(M)D.

Introduction

Life expectancy of people with intellectual disabilities (ID) has increased substantially over the last decades (Bittles & Glasson, 2004; Coppus, 2013; E. Evans et al., 2013). Since aging greatly increases the risk of dementia (Alzheimer's Association, 2021), dementia is becoming increasingly prevalent among people with ID. Moreover, people with Down syndrome (DS) have a particularly high genetic risk to develop dementia due to Alzheimer's disease (AD); up to 75% will have developed dementia by age 65 (Wiseman et al., 2015). Consequently, dementia is becoming a greater challenge in ID care. Particularly, the pre-existing ID and (life-long) patterns of characteristic/typical behavior make it complex to recognize and diagnose dementia in people with ID (Dekker, Strydom, et al., 2015; Jamieson-Craig et al., 2010; Sabbagh & Edgin, 2016; Zigman et al., 2008). Furthermore, there are a variety of comorbidities that may result in dementia-like symptoms (Moriconi et al., 2015; Scott & Barrett, 2007). For example, there are similarities in symptoms between depression and dementia, and therefore depression could mistakenly be diagnosed as dementia, or vice versa (Dekker, Strydom, et al., 2015; Dierckx et al., 2008; Prasher, 2009).

Recognizing and diagnosing dementia is particularly challenging in people with SPI(M)D, with an estimated IQ of less than 35 points (E. Evans et al., 2013; McKenzie et al., 2018). A diagnosis of dementia requires a decline in cognitive functioning interfering with performing ADL (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). Due to their low level of cognitive baseline functioning it is difficult to determine a decline in cognitive functioning resulting from the development of dementia (Ball et al., 2004; E. Evans et al., 2013). Moreover, they often need lifelong support to perform ADL, because they may have never developed specific skills. Never acquired skills cannot decline, and therefore cannot be indicative of dementia (Llewellyn, 2011; Sheehan, Sinai, et al., 2015). Furthermore, the frequent presence of multiple concurrent health problems in people with SPI(M)D make it highly complex to assess a dementia-related decline in functioning (van Timmeren et al., 2017). Lastly, there are hardly any self-reported symptoms, because of limited verbal communication skills in people with SPI(M)D (Smiley & Cooper, 2003).

Another obstacle to (early) recognizing and diagnosing dementia is the absence of validated and feasible direct neuropsychological tests and informant-based dementia screening instruments dedicated to people with SPI(M)D (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). Therefore, it is hard to establish a general diagnosis of dementia, let alone that a diagnosis of a subtype of dementia (e.g., AD, dementia with Lewy bodies, vascular dementia, or frontotemporal dementia) can be established (Burt et al., 1998; Day, 1985; Duggan et al., 1996; Margallo-Lana et al., 2007; Reid & Aungle, 1974). Currently, a diagnosis of dementia is based on multidisciplinary clinical assessment (by experienced clinicians) comprising observations, interviews with informants, such as family members and direct support professionals/caregivers, and/or screening case notes (Day, 1985; Duggan et al., 1996; Evenhuis, 1990; Määttä et al., 2006; Margallo-Lana et al., 2007; Reid & Aungle, 1974; Sauna-Aho et al., 2018). However, information about the presentation of symptoms and course of dementia in this population is scarce (Wissing, Ulgiati, et al., 2022). Therefore, dementia symptoms may not be recognized or may mistakenly be attributed to the ID, resulting in a (too) late diagnosis or no diagnosis at all (Cleary & Doody, 2017b). Nevertheless, it is essential to diagnose dementia in people with SPI(M)D to be able to timely respond to the person's changing wishes and needs by making informed choices (Dekker, Wissing, et al., 2021a; Janicki, 2011).

Early recognizing and diagnosing dementia in people with SPI(M)D requires a proper understanding of the presentation of dementia symptoms in this population. Recently, we obtained a first inventory of observable symptoms from the scarce literature (Wissing, Ulgiati, et al., 2022) and focus groups (Dekker, Wissing, et al., 2021a). This study aimed to further identify and deepen observable dementia symptoms in people with SPI(M)D through a survey and semi-structured interviews.

Methods

Study consortium

This study is part of the research project '*Practice-based questions about dementia in people with SPI(M)D*' (Dekker, Wissing, et al., 2021a; Wissing, Ulgiati, et al., 2022), a collaborative effort of University of Groningen, University Medical Center Groningen (UMCG) and Hanze University of Applied Sciences, with four Dutch care organizations (Alliade, 's Heeren

Loo, Ipse de Bruggen, and Royal Dutch Visio). These care organizations are representative of the Dutch ID care sector given the high number of people with SPI(M)D for whom they provide diagnostic work-up, treatments, and deliver care.

Study design

A mixed methods design was adopted comprising a survey and semi-structured interviews. Firstly, a survey was developed to identify practice-based observations of dementia symptoms in people with SPI(M)D. Secondly, interviews with care professionals were conducted to collect richer and more in-depth perspectives on symptoms covered in the survey. The Good Reporting of A Mixed Methods Study (GRAMMS; O'Cathain et al., 2008) and Consolidated Criteria for Reporting Qualitative Research (COREQ; Tong et al., 2007) were used as guidance for reporting this study.

Ethics and consent

The Medical Ethical Committee of the UMCG decided that the Dutch Medical Research Human Subjects Act did not apply to this study (METc 2019/198). The study was registered in the UMCG Research Register (no. 201900193) and conducted in accordance with the UMCG Research Code and the EU General Data Protection Regulation. Survey respondents provided consent for analyzing their responses by answering a consent question before the start of the survey. Interviewees provided written informed consent for audiotaping and analyzing the interview.

Survey

Respondents

Care professionals and family members of people with SPI(M)D and questionable dementia – i.e., presenting decline, but not (yet) meeting dementia criteria – or diagnosed dementia (established according to clinical judgment and (medical) records) were invited to participate in an online Dutch survey. The project team, consisting of representatives from consortium partners, identified eligible care professionals and family members within the four participating care organizations, partly through snowball sampling. Eligible respondents were purposefully selected based on the criterion that they had relevant experience/had a relative with SPI(M)D and questionable dementia or diagnosed dementia, and thus were able to provide information about observable dementia symptoms,

i.e., purposive sampling (Palinkas et al., 2015). Consequently, respondents were excluded if they only had experience with/their relative had mild/moderate ID or when they had no experience with questionable dementia or diagnosed dementia in those with SPI(M)D. The project team emailed the survey link to eligible respondents. A reminder was sent two weeks after initial invitation. Moreover, the survey link was disseminated via websites and newsletters of the research project and consortium partners. Five family members received a paper version of the survey due to limited computer accessibility/skills. Responses on paper were digitalized after completion.

Data collection

To construct the survey, we followed the steps described by Passmore et al. (2002). The first part consisted of two closed-ended questions to check whether respondents met inclusion criteria, i.e., having relevant experience with/having a relative with SPI(M)D and questionable dementia or diagnosed dementia. The survey ended if respondents did not meet the inclusion criteria. The second part gathered demographic data about age, sex, highest level of education, and relationship to people with SPI(M)D. The third part included items evaluating the observation of dementia symptoms, subdivided into four symptom domains based on diagnostic dementia criteria (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022) and literature (Dekker, Ulgiati et al., 2021a; Ries, 2018; Strydom et al., 2010). The fourth part contained an open-ended question in which respondents were asked whether they had observed changes not addressed in the survey.

Within part three, the first domain focused on cognitive functioning. In the general population, AD is the most common cause of dementia, accounting for 60–80% of all diagnoses (Alzheimer's Association, 2021). Moreover, people with DS have an extremely high genetic risk of developing dementia due to AD (Lott & Head, 2019; Wiseman et al., 2015). Therefore, items within this domain consisted of cognitive functions affected by AD: memory, planning, problem solving, orientation in time, orientation in place, understanding visual images/spatial relationships, language skills, losing objects, and judgment (Alzheimer's Association, 2021). These items were complemented by cognitive changes addressed in the focus group study of Dekker, Wissing, et al., (2021a): person recognition, object recognition, preference for (favorite) objects, responsiveness, and awareness of proper

order. Moreover, one item within this domain focused on ADL functioning (Alzheimer's Association, 2021). Given that dementia is characterized by a decline of cognitive and ADL functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022), response options were defined as: decrease (increase for losing objects, given that people with dementia may lose objects more frequently), unaltered, never shown or unknown.

The second domain contained behavioral and psychological items according to the sections described in the Behavioral and Psychological Symptoms of Dementia in Down Syndrome scale version II (BPSD-DS II) (Dekker, Ulgiati et al., 2021a). In the BPSD-DS II, restless and stereotypic behavior constitutes one section. To ensure that the survey addressed one aspect at a time, restless and stereotypic behavior were addressed as two separate items. Although most individuals with dementia display an increased frequency of behavioral changes, decreased frequency are also observed (Dekker, Ulgiati et al., 2021a). Therefore, response options were defined as: increase, decrease, unaltered, never shown, or unknown.

The third domain comprised motor items: walking, balance, fall frequency, movement speed (Ries, 2018), stiffness, muscle strength, cramps, wheelchair use, and choking (Dekker, Wissing, et al., 2021a). The fourth domain focused on medical comorbidities: epilepsy, weight, and incontinence (Strydom et al., 2010). Depending on the item, response options were defined as: increase, decrease, unaltered, never shown, or unknown. Moreover, each domain was followed by a comment field.

Subsequently, two survey versions were created: one for family members (answering about an individual case) and one for care professionals (answering about multiple cases). Given that the observation of changes can vary for different persons, two additional response options, 1) decrease for some persons, increase for others, and 2) unaltered for some persons, never shown for others, were added in the version for care professionals. The online survey versions were constructed in REDCap (Harris et al., 2009), hosted within the secured network of the UMCG.

During survey construction, project team members reviewed structure and content, leading to optimizing structure, removing redundant questions, and rephrasing items. Subsequently, draft versions were pilot tested with

two family members and six care professionals. Based on this pilot, the expected time needed to fill out the survey was between 10 and 15 minutes. Moreover, analysis of the pilot survey findings resulted in expanding survey introduction, refining texts (incl. shortening response options), reordering questions, and changing visual presentation. The pilot survey respondents did not fill out the final survey. Final survey versions were launched in August 2020. Data collection lasted three months.

Data analysis

Responses were exported to SPSS Statistics version 27 (IBM Corp), surveys that were not completed were excluded. Standard descriptive statistics and stacked bar graphs were used to present results. From left to right in the stacked bar graphs, changes per domain were depicted from most frequently to least frequently reported. Additionally, responses to open-ended fields/question were analyzed by coding symptoms as described in the interview data analysis section.

Interviews

Participants

The project team purposefully selected 28 eligible care professionals (not necessarily persons who had filled out the survey) having vast experience in recognizing/diagnosing dementia in people with SPI(M)D. They were particularly knowledgeable about and experienced in dementia in people with SPI(M)D and could thus provide a richer and more in-depth perspective. Eligible care professionals were consecutively invited until data saturation was reached, which was defined as the moment no new dementia symptoms were mentioned. Furthermore, to ensure that the interviewees reflected the multidisciplinary composition of professionals in daily practice, we selected interviewees based on their profession. Until data saturation was reached, 19 eligible participants had received an invitation by e-mail. Two persons did not respond, and three persons were unable to attend because of scheduling issues. Two eligible participants suggested including their direct colleagues (same profession, same care organization), who could provide a wealth of information as well (snowball sampling). Consequently, two interviews were held, in which two participants were simultaneously interviewed. Also, these two interviews were considered in the process of determining data saturation.

Data collection

Semi-structured interviews were conducted via Microsoft Teams by one author (A.S.F.) and lasted 45 to 75 minutes. An interview protocol was developed in advance, in consultation with project team members, and based on the guidelines by Boyce and Neale (2006). The interviewer followed instructions (protocol) which entailed a series of steps. Each interview started with welcoming interviewee(s), introducing the topic, checking if interviewee(s) had signed informed consent forms, asking permission for audiotaping, and explaining procedure and confidentiality. Furthermore, interviewees were asked to provide demographic information: age, sex, highest level of education, and working experience with SPI(M)D and questionable dementia or diagnosed dementia. Subsequently, the interviewer addressed symptom domains covered in the survey by asking open-ended questions. The first question was "On the basis of which symptoms do you conclude that someone with SPI(M)D has dementia?" Next, the interviewer asked follow-up questions (protocol) or could ask additional questions about relevant brought up symptoms. Each interview ended with summarizing discussed themes, asking if they would like to share anything else, and thanking the interviewee(s). After the first interview, the protocol was refined, i.e., rephrasing questions.

Interviews were recorded with a Philips audio recorder (DVT6510). To evaluate whether saturation was achieved, another researcher (M.B.G.W.) – not present during the interview – listened after each interview to the recording and summarized dementia symptoms. Data saturation was discussed with the project team members. Audiotapes were transcribed in Dutch (clean transcription) by the University Translation and Correction Service of the University of Groningen Language Center. Fillers, hesitations, and slips of the tongue were left out.

Data analysis

Transcripts of all 14 interviews were analyzed using a qualitative method of content analysis combining aspects of deductive and inductive content analysis (Elo & Kyngäs, 2008). Firstly, one researcher (M.B.G.W.) read all transcripts to familiarize herself with the data. Secondly, this researcher openly coded symptoms within the transcripts in ATLAS.ti version 8 (Scientific Software Development GmbH). A second researcher (A.S.F.) coded selected symptom text fragments of six interviews (in total 395 symptoms) by using the codes generated by the other researcher (M.B.G.W.). The number of

concordant coded symptoms was 296. Intercoder percent agreement, i.e., number of concordant coded symptoms/total number of coded symptoms x 100 (Gisev et al., 2013), was 74.9%.

To structure the broad range of symptoms, a categorization matrix (Elo & Kyngäs, 2008), here called a symptom matrix, was designed, similar to Dekker, Wissing, et al. (2021a). The symptom matrix rows were deductively designed in line with the symptom domains and items addressed in the survey. To further improve interpretation, the symptom matrix columns were thereafter inductively designed. Project team members discussed and refined categorization and (sub)thematization until reaching consensus. To improve trustworthiness, illustrative quotes were selected to support results (Elo & Kyngäs, 2008). One researcher (M.B.G.W.) translated selected Dutch quotes to English, which were where possible shortened (e.g., by leaving out unnecessary colloquial words). Project team members checked whether translations were accurate and if intentional meanings were maintained.

Results

Survey

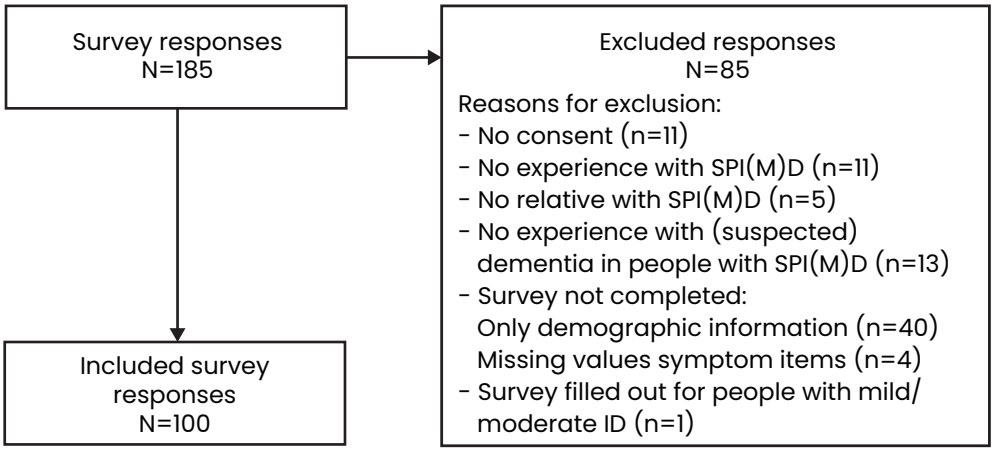

Figure 4.1 Schematic overview of included and excluded survey respondents. Abbreviations: ID, intellectual disabilities; SPI(M)D, severe/profound intellectual disabilities.

In total, 185 respondents started filling out the survey, of whom 85 were excluded for various reasons (Figure 4.1), primarily surveys that were not completed. Of the total 85 excluded responses, 61% were from care professionals and the remaining 33% were from family members. Data from 100 respondents, i.e., 87 care professionals and 13 family members (Table 4.1) were eligible for analysis.

Table 4.1 Characteristics of respondents.

Characteristics	All respondents N=100	Care professionals n=87	Family members n=13
Age (years [median (IQR), min-max])	46 (22), 21-83	42 (19), 21-68	63 (20), 44-83
Sex (% female)	91	94	69
Level of education: primary school, high school, mbo, hbo, wo (%)	2, 2, 37, 39, 20	0, 0, 39, 38, 23	15, 15, 23, 46, 0
Care organization: Ipse de Bruggen, 's Heeren Loo, Talant, Visio, other (%)		38, 21, 24, 9, 8	N/A
Role: physician, nurse specialist, caregiver, psychologist, psychologic assistant, occupational therapist, speech therapist, physiotherapist, dietician (%)		6, 6, 51, 16, 2, 5, 8, 6, 1	N/A
Experience working with SPI(M)D (years [median (IQR), min-max])		15 (13), 0-44	N/A
Working with SPI(M)D: D, W, M, other (%)		39, 43, 13, 6	N/A
Experience working with SPI(M)D + questionable dementia or dementia (years [median (IQR), min-max])		10 (12), 0-33	N/A
Working with SPI(M)D + questionable dementia or diagnosed dementia: D, W, M, other (%)		35, 38, 15, 13	N/A
Family relationship: parent, sibling, no family member but legal representative (%)		N/A	8, 77, 15
Years knowing relative ([median (IQR), min-max])		N/A	57 (13), 1-67
Frequency of visits (% W, M, Q)		N/A	31, 62, 8
Characteristics of relative with SPI(M)D + questionable dementia or diagnosed dementia			
- Age (years [median (IQR), min-max])		N/A	60 (9), 47-73
- Level of intellectual disability: severe, profound, not determined but probably severe/profound (%)		N/A	62, 31, 8
- Presence of Down syndrome (%)		N/A	54
- Presence of multiple disabilities (%)		N/A	62
- Living situation: care organization, at home (%)		N/A	92, 8

Percentages (rounded off to the nearest whole number without decimals) are calculated based on the total number of respondents per group (column). The group of psychologists is composed of behavioral therapists who studied psychology or special needs education (in Dutch: orthopedagogiek). Abbreviations: D, daily; hbo, higher vocational education; IQR, Interquartile range; M, monthly; mbo, intermediate vocational education; N/A, not applicable; Q, quarterly; SPI(M)D, severe/profound intellectual (and multiple) disabilities; W, weekly; wo, higher education.

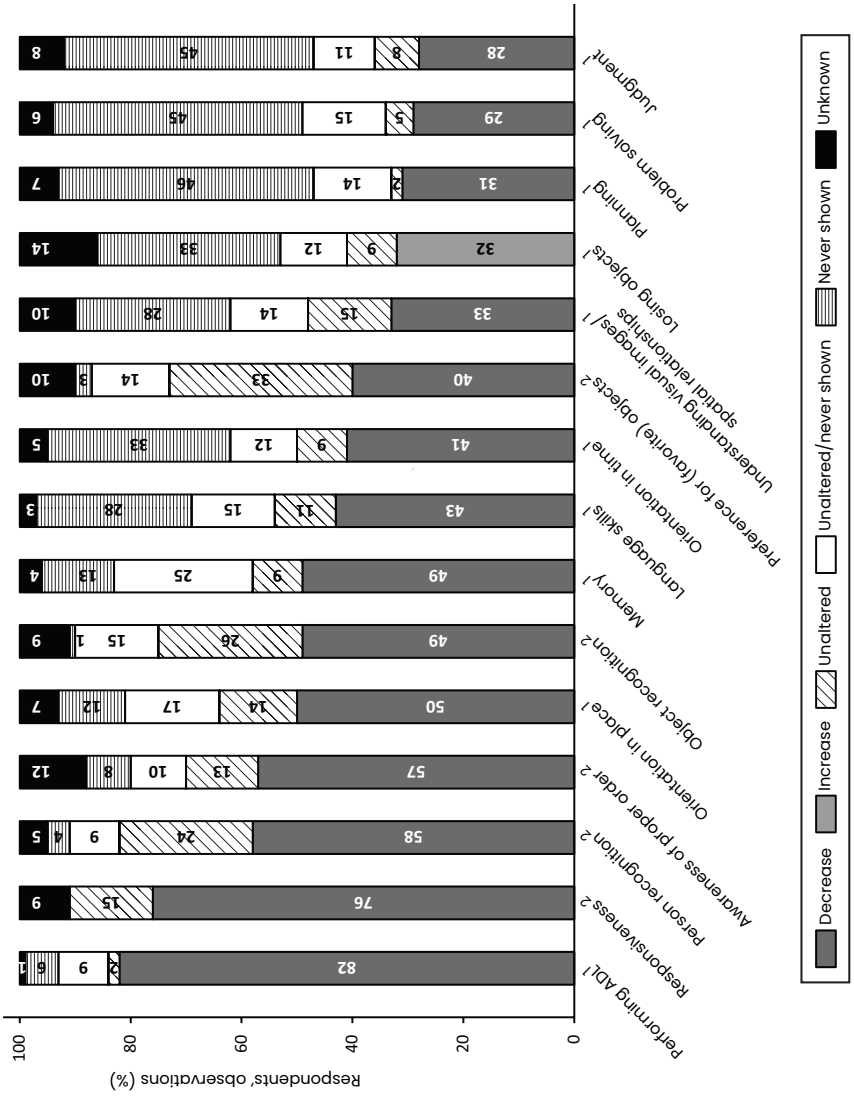

Figure 4.2 Respondents' observations of cognitive and activities of daily living (ADL) changes in people with SPI(M)D since the onset of questionable dementia or diagnosed dementia. Per item, the proportion (%) of decrease, unaltered, unaltered/never shown (i.e., unaltered for some persons, never shown for others), never shown and unknown are presented within each bar. From left to right, items are ordered from highest to lowest percentage of respondents observing a decrease (increase for losing objects) since questionable dementia or diagnosed dementia. References: 1, (Alzheimer's Association, 2021); 2, (Dekker, Wissing, et al., 2021a).

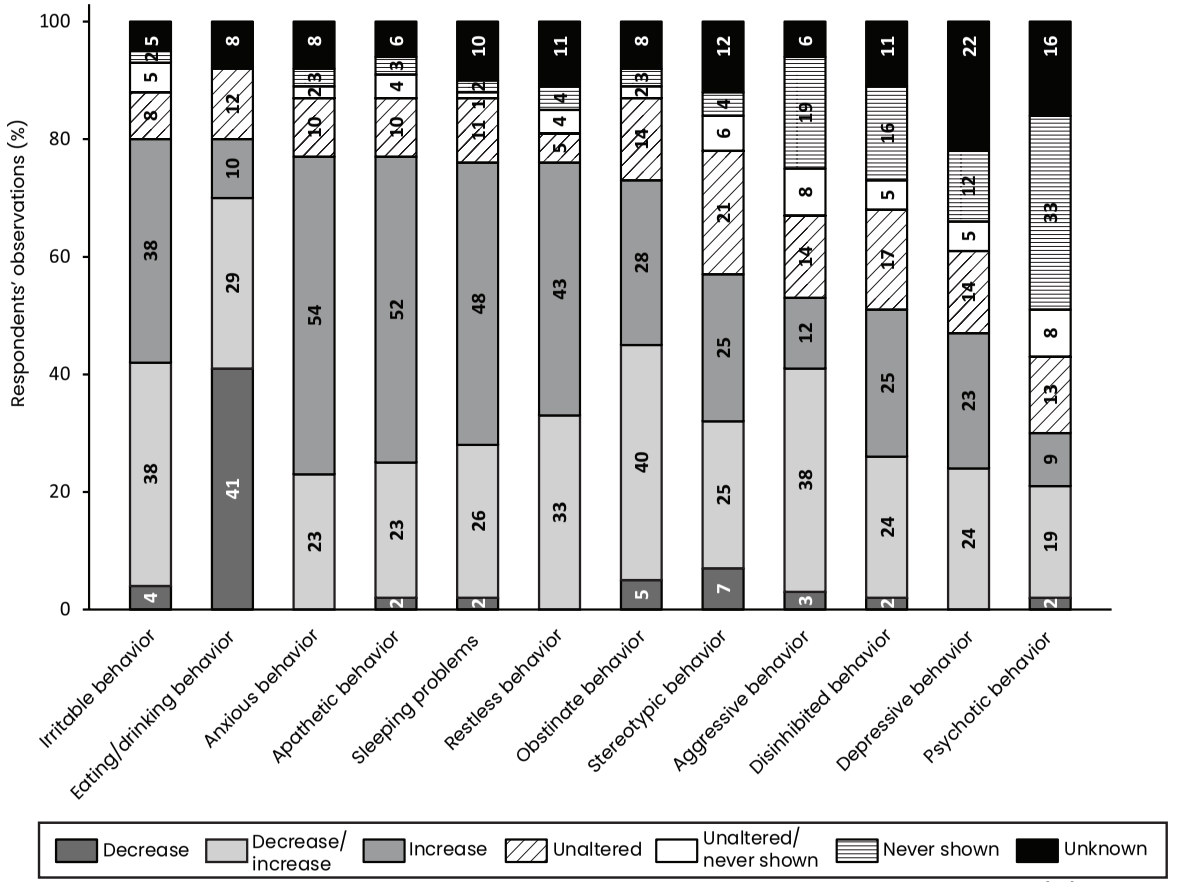

Figure 4.3 Respondents' observations of behavioral and psychological changes in people with SPI(M)D since the onset of questionable dementia or diagnosed dementia. Per item, the proportion (%) of decrease, decrease/increase (i.e., decrease for some persons, increase for others), increase, unaltered, unaltered/never shown (i.e., unaltered for some persons, never shown for others), never shown and unknown are presented within each bar. From left to right, changes are depicted from most frequently reported (either a decrease, an increase or a combination of both) to least frequently reported. Behavioral and psychological categories are provided in accordance with the sections of the BPSD-DS II (Dekker, Ulgiati et al., 2021a).

Cognitive and ADL changes

Figure 4.2 shows the respondents' observations of cognitive and ADL changes in people with SPI(M)D since the onset of questionable dementia or diagnosed dementia. The survey revealed that the most frequently observed dementia symptom in this population was a decline in ADL functioning (82%). Among cognitive items, most respondents (76%) indicated a decrease in responsiveness since questionable dementia or diagnosed dementia. Less frequent changes in planning (31%), problem solving (29%), and judgment (28%) were observed. For these items, respondents often reported that individuals had never shown these cognitive functions. Additionally, in the open text field respondents indicated that since the onset of questionable dementia or diagnosed dementia they had observed changes in sensory sensitivities (n=3) and a decreased ability to concentrate (n=1).

Behavioral and psychological changes

Figure 4.3 provides an overview of the percentage of respondents reporting behavioral and psychological changes. Evidently, changes in irritable behavior (80%) and eating/drinking behavior (80%) were most frequently reported. For all items within this domain, an increase as well as a decrease in behavior relative to the pre-existing life-long characteristic behavior was observed since the onset of questionable dementia or diagnosed dementia. For instance, concerning irritable behavior, 38% of the respondents observed an increase, 4% a decrease, and another 38% observed a decrease for some persons and an increase for others. Moreover, individuals ate and drank less/slower according to 41%, more/faster according to 29%, and variable according to 10%. Furthermore, respondents frequently highlighted changes in anxious behavior (77%), apathetic behavior (77%), sleeping problems (76%), restless behavior (76%), and obstinate behavior (73%). Changes in depressive (47%) and psychotic behavior (30%) were less commonly observed. Often respondents had never observed this behavior (12% and 33%, respectively), or they did not know whether behavior had changed (22% and 16%, respectively). Additionally, respondents reported changes in compulsive behavior, which was not addressed in the survey. One had observed an increase in compulsive behavior, whereas two others had observed a decrease relative compared to the pre-existing life-long characteristic behavior.

Motor changes

Figure 4.4 visualizes the responses about motor changes. The majority of respondents (80%) noticed that, since the onset of questionable dementia or diagnosed dementia, walking skills had declined. Moreover, the wheelchair was more frequently used (72%), choking was more common (70%), and movements were slower (57%) since the onset of questionable dementia or diagnosed dementia. Changes in muscle strength and cramps were less common, often it either remained stable (19% and 35%, respectively) or unknown (31% and 30%, respectively). Additionally, respondents described motor symptoms not addressed in the survey: decreased body awareness (n=3), decreased motor skills (n=2), increased tremor (n=1), and sitting/laying more in fetal position (n=1).

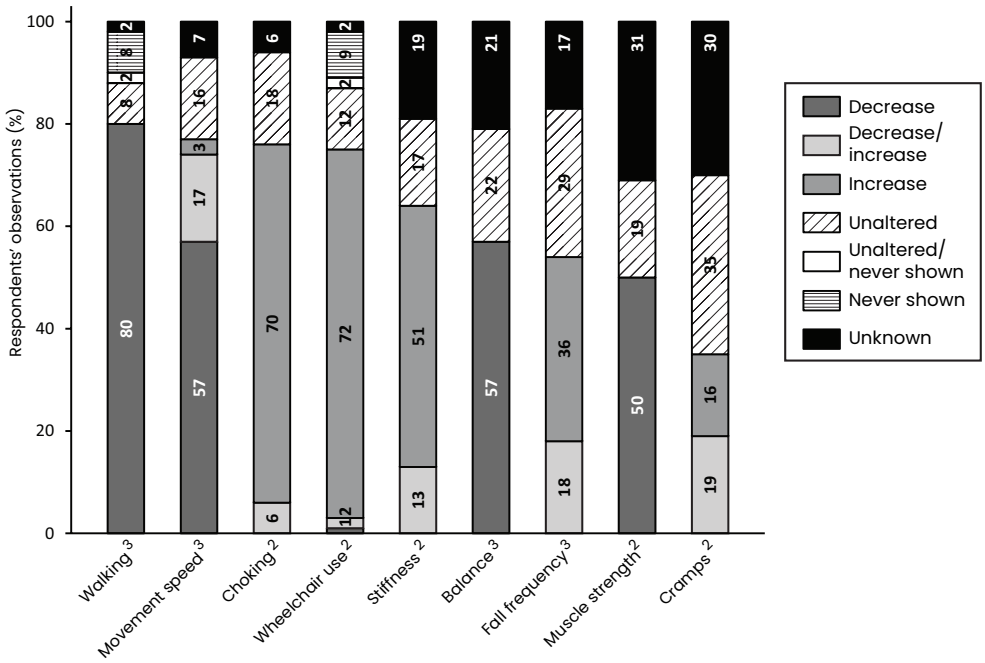

Figure 4.4 Respondents' observations of motor changes in people with SPI(M)D since the onset of questionable dementia or diagnosed dementia. Per item, the proportions (%) of decrease, decrease/increase (i.e., decrease for some persons, increase for others), increase, unaltered, unaltered/never shown (i.e., unaltered for some persons, never shown for others), never shown and unknown are presented within each bar. From left to right, motor changes are presented from most frequently reported (either a decrease, an increase or a combination of both) to least frequently reported. References: 2, (Dekker, Wissing, et al., 2021a); 3, (Ries, 2018).

Medical comorbidities

Figure 4.5 visualizes respondents' observations of medical comorbidities in people with SPI(M)D and questionable dementia or diagnosed dementia. Respondents observed weight changes (57%), mainly weight loss (27%), increased incontinence (48%), and increased frequency and severity of epileptic seizures (34%). In the open text fields, three medical comorbidities not included in the survey were addressed: decreased taste sensation (n=2), increased pain (n=1), and becoming bedridden (n=1).

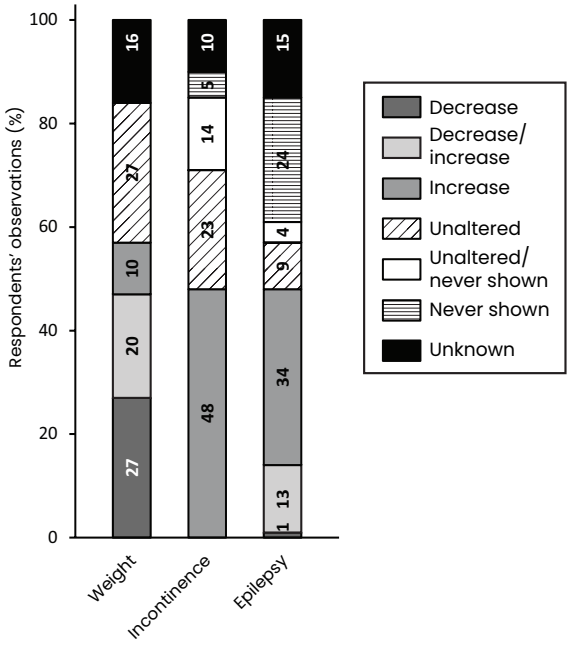

Figure 4.5 Respondents' observations of changes in medical comorbidities in people with SPI(M)D since the onset of questionable dementia or diagnosed dementia. Per item, the proportion (%) of decrease, decrease/ increase (i.e., decrease for some persons, increase for others), increase, unaltered, unaltered/ never shown (i.e., unaltered for some persons, never shown for others), never shown and unknown are shown within each bar. From left to right, motor changes are presented from most frequently reported (either a decrease, an increase or a combination of both) to least frequently reported. Reference: (Strydom et al., 2010).

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Interviews

Based on the first analysis, we concluded that observable dementia symptoms mentioned during interviews 12, 13, and 14 were consistent with earlier interviews, and thus saturation had been reached. Hence, a total of 14 interviews were conducted with 16 care professionals (Table 4.2).

Table 4.2 Interviewees' characteristics.

Characteristics	Care professionals N = 16
Age (years [median (IQR), min-max])	50 (21), 29-64
Sex (% female)	88
Level of education: mbo, hbo, wo (%)	13, 38, 50
Care organization: Ipse de Bruggen, 's Heeren Loo, Talant, Visio, other (%)	25, 19, 19, 31, 6
Role: physician, nurse specialist, caregiver, psychologist, occupational therapist, speech therapist, physiotherapist (%)	19, 6, 19, 25, 13, 13, 6
Experience working with SPI(M)D (years [median (IQR), min-max])	22 (16), 5-33
Experience working with SPI(M)D + questionable dementia or diagnosed dementia (number of people [median (IQR), min-max])	127 (186), 2-500

Percentages (rounded off to the nearest whole number without decimals) are calculated based on the total number of interviewees. The group of psychologists is composed of behavioral therapists who studied psychology or special needs education (in Dutch: orthopedagogiek). Abbreviations: hbo, higher vocational education; mbo, intermediate vocational education; SPI(M)D, severe/profound intellectual (and multiple) disabilities; wo, higher education.

Reported symptoms were coded and subsequently categorized using a symptom matrix (Table 4.3). Inductive content analysis revealed that symptoms were generally observed in relation to having verbal communication or walking skills, which is in accordance with a survey respondent stating in the open text field that the observation of dementia symptoms in persons with SPI(M)D depended on whether at baseline individuals were verbal or could walk. In addition, a category called "category-independent" was created for symptoms not affected by the presence or absence of verbal communication or walking skills.

> ***Psychologist C.:*** *"Many persons within the SPI(M)D group are dependent on a wheelchair, they cannot walk and/or cannot talk. Then it is far more difficult to examine (. . .)."*

Table 4.3 shows that interviewees observed cognitive changes, particularly when individuals had verbal communication or walking skills at baseline,

whereas behavioral and psychological changes were mostly observed irrespective of such skills. Motor changes were particularly observed when persons were able to walk at baseline. Furthermore, changes in ADL functioning and medical comorbidities were observed in people with and people without walking skills at baseline. Results within each domain are described below in more detail, supported by quotes to clarify interviewees' observations of symptoms.

Cognitive changes

Interviewees stated that observing cognitive symptoms in people with SPI(M)D is very complex. Nevertheless, cognitive symptoms like deterioration in language skills, memory loss, and disorientation in time were (mainly) recognized when individuals had verbal communication skills at baseline.

> ***Physician T.:*** *"Language is something that is very obvious. People are going to use fewer words and eventually stop talking. If someone has the ability to speak, then loss of speech could certainly be a signal."*

Disorientation in place, losing objects, and trouble understanding visual images/spatial relationships were particularly observed in people with walking skills.

> ***Speech therapist M.:*** *"Walking in the wrong direction or suddenly going to the toilet, but walking into the laundry room instead, that are signals when someone is able to walk. (. . .) That is not observable when someone is dependent on a wheelchair."*

Cognitive symptoms like reduced responsiveness, declined person recognition, and increased sensory sensitivities were observed regardless of having verbal communication or walking skills. This also applied to reduced sound recognition, which was a symptom not addressed in the survey.

ADL changes

Particularly, a decline in eating/drinking skills was observed in persons with SPI(M)D and questionable dementia or diagnosed dementia. A decline in dressing, toilet use, and stair climbing were only noticed in individuals

more capable of performing ADL. Moreover, interviewees emphasized that when someone is less able to perform ADL independently, this complicates the observation of alterations in ADL.

> ***Speech therapist M.:*** *"Symptoms like not understanding how to perform a task, how to brush your teeth (. . .), how to dress up, those are not indicative for dementia in people with SPI(M)D, because in general we do that for them."*

Behavioral and psychological changes

All interviewees highlighted that since the onset of questionable dementia or diagnosed dementia they had observed behavioral and psychological changes. Particularly, an increase in anxious behavior as compared to the pre-existing life-long characteristic behavior was noticed.

> ***Psychologist C.:*** *"Observable behavior related to dementia is anxiety and nervousness, for instance, an anxious facial expression, (. . .) or hesitant to walk. Screaming, that can of course also be a sign of anxiety."*

Moreover, since the onset of questionable dementia or diagnosed dementia interviewees had frequently observed an increase in apathetic behavior, sleeping problems, irritability, obstinate behavior, restlessness/stereotypic behavior, and a decrease in eating/drinking behavior. Interviewees emphasized that often a combination of such changes was observed in specific situations.

> ***Speech therapist M.:*** *"Signs which are often observed during eating and drinking are restlessness, crying, or falling asleep at the table. (. . .) A person could also be less alert."*

Additionally, it was emphasized that it is difficult to observe psychotic and depressive behavior in people with SPI(M)D.

> ***Psychologist C.:*** *"Unhappiness can be observed when the entire appearance of a person changes, for instance, hollow eyes or keeping your head down. It remains very complex (. . .). It could be dementia, but it could also be a depression."*

Table 4.3 Symptom matrix structuring dementia symptoms in people with SPI(M)D observed by interviewees.

			Verbal communication skills	Walking skills	Category-independent
Cognitive changes[1,2,3]	Memory	P	↓ remembering names, ↑ saying incorrect things, ↑ repeatedly asking something	↓ remembering: how to walk/dress up/set table, that you went to toilet, were to hang coat, that you were asked to sit on chair.	↓ understanding: what is (about to) happen, expectations, activities, changes, jokes, communication (verbal, augmentative and alternative).
		A			↓ remembering: new information, that you were going to eat/drink, how to use cutlery, how to chew/swallow/ brush teeth, what to do with food in mouth, that dining chairs belong to table.
	Orientation in time	P	Saying "good morning" in afternoon	Put on pajamas in morning, regular clothes in evening	
		A			
	Orientation in place	P		↓ remembering: where you/rooms are, direction/destination, new routes. ↑ getting lost, ↑ suddenly standing still in rooms	
		A		↑ stretching arms when passing doorways, ↑ agitation when wheelchair is turned	
	Understanding visual images/ spatial relationships	P		↑ bumping into things,difficulty with floor transitions	
		A		↑ bumping into things with wheelchair	
	Language skills	P	↓ talking, ↓ speech intelligibility, ↓ number of words used, ↓ expression with words, ↓ language comprehension, stop talking mid-sentence, speaking confusedly		
		A	↓ frequency of producing sounds		
	Losing objects	P		↓ remembering where you put toys	
		A			
	Person recognition	P			↓ recognizing caregivers/family members
		A			

Table 4.3 Continued.

			Verbal communication skills	Walking skills	Category-independent
Cognitive changes[1,2,3]	Object recognition	P		↓ recognizing walking lines on floor	↓ recognizing food/cutlery/table/ chair/doll
		A			
	Sound recognition	P			↓ recognizing sounds/songs
		A			
	Preference for (favorite) objects	P		Not going to preferred seat	Liking things that were previously disliked, no longer touching cup/toys that someone used to hold
		A			
	Responsiveness	P			No response while previously full of expectations, ↓ quickness of response, ↓ contact, ↓ alertness, ↑ alertness, ↓ keeping up with pace, ↓ reacting on the environment, ↑ staring ahead, ↑ not making eye contact, ↑ not responding to songs/activities
		A			
	Awareness of proper order	P		↓ understanding sequence of showering before dressing up, ↑ dependency on structure, put underwear over pants, start activities at wrong moment	↓ remembering: daily routines, consecutive steps. Shift daily routines, skip steps, ↓ understanding daily activity icons
		A			
	Sensory sensitivities	P			Sensory overload, negative reactions to stimuli, cannot bear sounds/songs/light, ↓ toleration of other residents, perceiving being touched as unpleasant, clothes feel uncomfortable, ↑ seeking proprioceptive input
		A			
	Concentration	P			↓ concentration
		A			
ADL[1,2,3] changes	Performing ADL	P		↓ ability to: (un)dress, put on socks/ coat, go to toilet, stair climbing.	↓ ability to: eat/drink, take food from spoon, open mouth, use cutlery/cup, washing vegetables, fold laundry, pick up puzzle pieces.
		A		↓ ability to propel wheelchair	

Table 4.3 Continued.

			Verbal communication skills	Walking skills	Category-independent
Behavioral and psychological changes[4]	Anxious behavior	P		↑ hesitant to walk, sliding across floor	↑ screaming, ↑ crying, ↑ whining, ↑ nervous, ↑ tension, ↑ panic, ↑ stress, ↑ freezing, inability to relax, anxious facial expression, fear in someone's eyes, eyes wide open, forehead sweating, ↑ feeling unsafe, ↑ fear of being alone/hoist/going outside/hard noises/at night/ when eating, ↑ frightened when being touched/ball is thrown, clinging to table, avoiding things, ↑ contact seeking, ↑ seeking security, hesitant to let caregiver go
		A		↑ anxiety for hoist	
	Sleeping problems	P		↑ wandering at night, crawl out of bed	day-night rhythm disturbance, ↑ insomnia, ↑ prowling about/ restlessness/waking up/screaming at night, ↑ difficulty getting up, ↑ sleeping during day, ↑ being tired during day, falling asleep earlier
		A			
	Irritable behavior	P			↑ touchy, ↑ irritability, ↑ frustration, ↑ anger, ↑ yelling, ↑ screaming during ADL, ↑ sounds of discomfort
		A			
	Obstinate behavior	P		↓ willingness to walk	↑ resistance against eating/dressing up/showering/ activities. ↑ being uncooperative, ↑ being self-willed, no longer accepting aids, ↑ turning head away
		A			
	Restless/ stereotypic behavior	P	↑ repeating words/questions, excessive talking	↑ walking, ↑ wandering	↑ restlessness, ↑ compulsive acts, ↑ stereotypical acts, ↑ picking behavior, ↑ fecal smearing, ↑ rituals preventing to sleep
		A			
	Aggressive behavior	P		↑ slam doors	↑ verbal/physical aggression against self and/or others, ↑ biting/beating, ↑ throwing objects
		A			
	Apathetic behavior	P		↓ motivation to walk, not getting of sofa	↑ withdrawn, ↑ being passive, ↑ laziness, ↓ initiative, ↓ motivation, ↓ jovial, ↓ enjoying food/music, ↓ interest in activities they used to like, ↓ focus on eating, ↓ (eye) contact, ↓ noticing things, ↑ staring ahead, ↑ letting go daily structure
		A		↑ sitting still in wheelchair	

Table 4.3 Continued.

			Verbal communication skills	Walking skills	Category-independent
Behavioral and psychological changes[4]	Depressive behavior	P			↓ emotion regulation, ↓ smiling, ↑ crying, ↑ discourages, ↑ mood changes, very unhappy, hollow eyes, head down
		A			
	Psychotic behavior	P	↑ mentioning things that are not there		↑ suddenly looking at something/ noticing things
		A			
	Eating/drinking behavior	P			↓ appetite/eating/drinking, ↓ preference favorite food, eating slowly
		A			
Motor changes[5]	Motor skills	P		↓ walking distance/speed, gait changes, ↓ lower limb coordination, ↓ standing up, ↑ bottom shuffle, ↑ wheelchair use	↓ movement speed, ↑ stiffness, ↑ clumsiness, ↓ muscle strength, ↑ muscle tension, ↑ cramps, ↑ overstretching muscles, ↑ fetal sleep position, ↑ sitting cross-legged
		A			
	Balance	P		↑ gait clumsiness/unsteadiness, ↑ insecure walking, ↑ tripping, ↑ falling, ↓ body awareness	
		A		↓ maintaining body posture	
	Chewing/ swallowing	P			↓ chewing, ↓ swallowing, ↑ unsafe swallowing, ↑ swallowing without chewing, ↑ choking
		A			
Medical comorbidities[6]		P		↑ incontinence, ↑ weight, ↓ bowel movements, ↑ bedridden	↑ epilepsy, ↓ weight
		A			

Dementia symptoms reported by interviewees were categorized based on symptom domains and items addressed in the survey (rows) and verbal/walking skills at baseline, i.e., highest level of functioning before dementia-related decline occurred (columns). Symbols: ↓ , decrease compared to baseline level of functioning/behavior; ↑, increase compared to baseline level of functioning/behavior. Abbreviations: ADL, activities of daily living; A, capacity/skill already absent at baseline; P, capacity/skill present at baseline. References: 1, (American Psychiatric Association, 2013); 2, (McKhann et al., 2011); 3, (World Health Organization, 2022); 4, (Dekker, Ulgiati et al., 2021a); 5, (Ries, 2018); 6, (Strydom et al., 2010).

Motor changes
A deterioration of walking skills accompanied by increased balance problems and wheelchair use were frequently observed by interviewees.

> ***Physiotherapist P.:*** *"Persons with walking skills at baseline, lose at a certain moment their ability to walk and eventually become dependent on a wheelchair. However, then it is often already obvious that those individuals have dementia."*

Furthermore, interviewees stated that chewing and swallowing became progressively more difficult with the onset of questionable dementia or diagnosed dementia.

> ***Physician J.:*** *"People with SPI(M)D often already have swallowing problems, but it becomes progressively worse (. . .). I think that is a sign."*

Medical comorbidities
Interviewees stated that they had observed medical comorbidities like the onset of epilepsy, becoming incontinent and weight changes with the onset of questionable dementia or diagnosed dementia.

> ***Nurse specialist S.:*** *"Particularly in people with Down syndrome, epilepsy is something that can be associated with dementia."*

Discussion

Using a survey and semi-structured interviews, an inventory of practice-based observations of dementia symptoms in people with SPI(M)D was obtained. Survey data indicated that the most frequently observed symptom concerned a decline in ADL functioning, followed by behavioral and psychological symptoms of dementia, in particular changes in irritable, eating/drinking, anxious, and apathetic behavior. To a lesser extent, cognitive symptoms, motor changes, and medical comorbidities were observed. Subsequently, interviews provided a richer and more in-depth perspective on symptoms covered in the survey. Cognitive symptoms were generally observed when persons had verbal communication or walking skills at baseline, whereas behavioral and psychological changes were mostly noticed regardless of having such baseline skills. Moreover,

motor changes were particularly observed when persons were at baseline able to walk. Lastly, changes in ADL functioning and medical comorbidities were observed in people with and people without walking skills at baseline.

To timely recognize and diagnose dementia in SPI(M)D, insights into the symptomology are needed. This also contributes to better understanding and making informed choices (Dekker, Wissing, et al., 2021a). Recently, we conducted a systematic literature review to identify observable symptoms in the scarce literature about dementia in this population (Wissing, Ulgiati, et al., 2022). Given the very limited number of studies, we conducted an explorative focus group study to obtain practice-based experiences (Dekker, Wissing, et al., 2021a). This study was the next step to further identify practice-based observations of dementia symptoms in this population. Hereafter, we contextualize the survey and interview results with the outcomes of the systematic literature review and focus groups (Table 4.4; Dekker, Wissing, et al., 2021a; Wissing, Ulgiati, et al., 2022).

Cognitive changes

An ID is characterized by deficits in cognitive functioning, e.g., deficits in reasoning, problem solving, planning, and judgment (American Psychiatric Association, 2013). If cognitive skills have not or hardly been developed at baseline, such skills cannot decline, and therefore cannot be indicative of dementia (Llewellyn, 2011; Sheehan, Sinai, et al., 2015). Consequently, one could hypothesize that cognitive decline would be less observable in people with SPI(M)D. However, results from the survey, interviews as well as previous findings in focus groups (Dekker, Wissing, et al., 2021a) and literature (Wissing, Ulgiati, et al., 2022) jointly show that, despite the low baseline level of functioning, cognitive alterations like memory loss, disorientation in place and deterioration in language skills are observable in this population (Table 4.4). It should be noted that interviewees emphasized that such changes are (more easily) observed when individuals have verbal communication or walking skills at baseline. As expected, higher cognitive functions such as planning, problem solving, and judgment were not mentioned in interviews, focus groups (Dekker, Wissing, et al., 2021a) and literature (Wissing, Ulgiati, et al., 2022) and hardly addressed in the survey (Table 4.4).

Table 4.4 Comparison of dementia symptoms in people with SPI(M)D obtained with different research methods.

	Symptoms	Survey	Interviews	Focus groups[1]	Literature review[2]
Cognitive changes	↓ Memory	✓	✓	✓	✓
	↓ Orientation in place	✓	✓	✓	✓
	↓ Language skills	✓	✓	✓	✓
	↓ Responsiveness	✓	✓	✓	
	↓ Person recognition	✓	✓	✓	
	↓ Awareness of proper order	✓	✓	✓	
	↓ Object recognition	✓	✓	✓	
	↓ Orientation in time	✓	✓	✓	
	↓ Preference for (favorite) objects	✓	✓	✓	
	↓ Understanding visual images/spatial relationships	✓	✓		
	↓ Concentration	✓	✓		
	↑ Losing objects	✓	✓		
	↓ Sensory sensitivities	✓	✓		
	↓ Planning	✓			
	↓ Problem solving	✓			
	↓ Judgment	✓			
	↓ Sound recognition		✓		
ADL	↓ Performing ADL	✓	✓	✓*	✓
Behavioral and psychological changes	↑ Irritable behavior	✓	✓	✓	✓
	↓ Eating/drinking behavior	✓	✓	✓	✓
	↑ Apathetic behavior	✓	✓	✓	✓
	↑ Sleeping problems	✓	✓	✓	✓
	↑ Restless/stereotypic behavior	✓	✓	✓	✓
	↑ Aggressive behavior	✓	✓	✓	✓
	↑ Anxious behavior	✓	✓	✓	
	↑ Obstinate behavior	✓	✓	✓	
	↑ Disinhibited behavior	✓		✓	✓
	↑ Depressive behavior	✓	✓	✓	
	↑ Psychotic behavior	✓	✓	✓	
Motor changes	↓ Walking	✓	✓	✓	✓
	↑ Wheelchair use	✓	✓	✓	
	↓ Balance	✓	✓	✓	
	↑ Fall frequency	✓	✓	✓	
	↑ Swallowing problems	✓	✓	✓	
	↑ Stiffness	✓	✓	✓	
	↑ Cramps	✓	✓	✓	
	↓ Body awareness	✓	✓	✓	
	↓ Muscle strength	✓	✓	✓	
	↓ Motor skills	✓	✓	✓	
	↓ Movement speed	✓	✓		

Table 4.4 Continued.

	Symptoms	Survey	Interviews	Focus groups[1]	Literature review[2]
	↑ Fetal sitting/laying position	✔	✔		
	↑ Tremor	✔			
Medical comorbidities	↓ Weight	✔	✔	✔	✔
	↑ Incontinence	✔	✔	✔	✔
	↑ Epilepsy	✔	✔	✔	✔
	↑ Bedridden	✔	✔		✔
	↑ Pain	✔		✔	
	↓ Taste sensation	✔		✔	
	↓ Bowel movements		✔		

This table provides a comparison of dementia symptoms reported in the survey, interviews with previously published findings using two other research methods, namely focus groups (Dekker, Wissing, et al., 2021a) and systematic literature review (Wissing, Ulgiati, et al., 2022). Symptoms are categorized in five symptom domains, which is in line with dementia criteria (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022) and literature (Dekker, Ulgiati et al., 2021a; Ries, 2018; Strydom et al., 2010). ✔ indicates that a symptom was reported in a research method. For behavioral and psychological changes, motor changes and medical comorbidities only the most prominently reported symptoms are presented. Symbols: ↓, decrease compared to baseline level of functioning/behavior; ↑, increase compared to baseline level of functioning/behavior; Baseline level of functioning is the highest level of functioning before dementia-related decline occurred, *, Symptoms reported in focus groups were categorized based on the daily contexts in which they were often observed in practice. Therefore, a decline in activities of daily living (ADL) functioning was addressed in various contexts and symptoms.

ADL changes

In addition to cognitive decline, another prominent sign of dementia concerns a decline in ADL functioning (Alzheimer's Association, 2021; American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). In people with SPI(M)D and questionable dementia or diagnosed dementia this was prominently observed by survey respondents and interviewees. These findings underline previous findings in focus groups (Dekker, Wissing, et al., 2021a) and literature (Wissing, Ulgiati, et al., 2022); Table 4.4). However, the way in which a decline in ADL manifests depends on someone's baseline functioning, as already addressed by Benejam (2009). Interviews showed that in most persons a decline in eating/drinking skills was observed, whereas a decline in dressing, toilet use, and stair climbing was only observable in individuals more capable of performing ADL.

Behavioral and psychological changes

Behavioral and psychological symptoms of dementia can be observed in all types of dementia and are most observable for caregivers (Engelborghs et al., 2005; Finkel, 2000). Indeed, results from the survey, interviews as well as previous findings of focus groups (Dekker, Wissing, et al., 2021a) and literature

(Wissing, Ulgiati, et al., 2022) jointly showed that behavioral changes such as increased irritable, restless/stereotypic, aggressive, apathetic behavior and decreased eating/drinking behavior can be observed in persons with SPI(M)D and questionable dementia or diagnosed dementia (Table 4.4). In line with recent findings in two large studies on dementia in people with DS with mild, moderate, and severe ID (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a), prominent behavioral and psychological symptoms of dementia were changes in irritable, eating/drinking, anxious, apathetic, restless/ stereotypic behavior and sleeping problems, whereas psychotic behavior was less frequently observed. Communication of individuals with SPI(M)D is less verbal, making it complex to accurately elucidate the inner experience of delusions and hallucinations (Cooper & Smiley, 2007). In contrast to Dekker et al. (2018, 2021a), changes in depressive behavior were less frequently reported in individuals with SPI(M)D and questionable dementia or diagnosed dementia. Recognizing and differentiating between depression and depressive symptoms related to dementia is particularly difficult in this population because persons with limited verbal communication skills cannot report their mood and do not have the cognitive level for specific symptoms that classically characterize depression, such as doom-mongering or being tired of life (Dekker, Sacco, et al., 2018; Dekker, Strydom, et al., 2015; K. M. Evans et al., 1999).

Motor changes

In the general population, motor changes such as gait changes and diminished postural control (balance and falls) are observed in individuals with dementia (Ries, 2018). Survey and interview results demonstrated that in people with SPI(M)D and questionable dementia or diagnosed dementia, such motor changes were also observed in those with walking skills at baseline. Moreover, both research methods showed that since the onset of questionable dementia or diagnosed dementia swallowing problems increased in this population. These motor changes were also found in the focus group study (Dekker, Wissing, et al., 2021a; Table 4.4).

Medical comorbidities

Incontinence, onset of epilepsy, and weight changes are recognized as medical comorbidities with dementia in the general population (Kurrle et al., 2012) and people with DS (Strydom et al., 2010). Similarly, these comorbidities were reported in survey and interviews and are also consistent with the findings of the focus groups (Dekker, Wissing, et al.,

2021a) and literature (Wissing, Ulgiati, et al., 2022). With respect to epilepsy, interviewees stressed that the onset of epilepsy was particularly observed in those with DS, which is in line with results of other studies focusing on late onset myoclonic epilepsy in DS (Aller-Alvarez et al., 2017; Altuna et al., 2021; Menéndez, 2005).

Strengths

One of the strengths of this study is the mixed methods design comprising a quantitative survey and qualitative interviews to identify practice-based observations of dementia symptoms in people with SPI(M)D. To the best of our knowledge, this study is the first to examine whether dementia symptoms described in literature are observed in daily practice in this population. Furthermore, a richer and more in-depth perspective on symptoms covered in the survey was obtained by conducting interviews with care professionals. A strength of the interviews is the purposive sampling of care professionals having vast experience in recognizing/diagnosing dementia in this population. Furthermore, the heterogeneity of this population was considered through inductive content analysis of transcripts, enabling us to refine results (categorization of symptoms) in relation verbal communication and walking skills at baseline.

Limitations

A first potential limitation concerns the fact that a rather small number of family members completed the survey. This might be related to the fact that most elderly with SPI(M)D have spent much of their lives in a care organization (Johnson & Traustadóttir, 2005), which could have an impact on the (more distant) family involvement. Secondly, providing care is key priority when working with people with SPI(M)D, and therefore care professionals might not always had time to complete the survey. Thirdly, care professionals provide care to people with different levels of functioning, and therefore could have referred to some signs of dementia in people with mild/moderate ID. In the survey (introductory texts) and interviews, the focus on SPI(M)D was clearly emphasized. Fourthly, given the complexity of diagnosing dementia in this population a diagnosis is often not formally established. Therefore, family members and care professionals could have referred to symptoms caused by other conditions that mimic dementia. This is the result of limited knowledge about dementia symptoms in people with SPI(M)D. It underlines the relevance of research on the symptomatology of dementia in this population.

Future implications

Diagnosing dementia in people with SPI(M)D is an often long and complex process. It is already quite difficult to establish a general diagnosis of dementia, let alone that a diagnosis of the subtype of dementia (e.g., AD, dementia with Lewy bodies, vascular dementia, or frontotemporal dementia) is established (Burt et al., 1998; Day, 1985; Duggan et al., 1996; Margallo-Lana et al., 2007; Reid & Aungle, 1974). Diagnosing dementia requires a proper diagnostic procedure. Dementia-like symptoms could be caused by – often treatable – conditions, also referred to as pseudo-dementias (Zigman, 2013; Zigman et al., 2008). Therefore, potential other causes, such as depression, delirium, vision or hearing problems, hypothyroidism, medication use, sleep apnea, or vitamin B12 deficiency – should be ruled out as much as possible before establishing a diagnosis of dementia (Moriconi et al., 2015; Scott & Barrett, 2007). Moreover, tests could be used to monitor the progression of reduction in functioning over time. However, currently there are hardly any validated direct neuropsychological tests and informant-based dementia questionnaires available to (early) diagnose dementia in people with SPI(M)D (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). An inventory of observable dementia symptoms through a survey and interviews, together with findings from literature (Wissing, Ulgiati, et al., 2022) and focus groups (Dekker, Wissing, et al., 2021a) provide a first essential step for developing a dedicated dementia screening instrument for people with SPI(M)D. The development process may be aided by identifying relevant items within existing dementia screening instruments primarily applicable to people with mild and moderate ID.

Conclusion

This study provided an overview of observable dementia symptoms in people with SPI(M)D. Particularly, a decline in ADL functioning and behavioral and psychological symptoms like increased irritable, anxious, apathetic behavior and decreased eating/drinking behavior were recognized. To a lesser extent, cognitive symptoms like memory loss, disorientation in place, and deterioration in language skills were observed, particularly in those with verbal communication or walking skills at baseline. Furthermore, motor changes and medical comorbidities were reported. The inventory of symptoms in this study, together with findings from literature (Wissing, Ulgiati, et al., 2022) and focus groups (Dekker, Wissing, et al., 2021a), pave the way for developing a dedicated screening instrument for dementia in people with SPI(M)D.

Chapter 5

Characterizing the natural history of dementia through retrospective analysis of clinical records

Wissing, M. B. G., Hobbelen, J. S. M., De Deyn, P. P., Waninge, A., & Dekker, A. D. (2023). Dementia in people with severe/profound intellectual (and multiple) disabilities, and its natural history. *Journal of Mental Health Research in Intellectual Disabilities*.

Abstract

Introduction: Although the prevalence of dementia increases among people with severe/profound intellectual (and multiple) disabilities (SPI(M)D), dementia in people with SPI(M)D is not yet fully understood. Therefore, this study aimed to characterize the natural history of dementia in people with SPI(M)D, in particular, the prevalence and time of onset of dementia symptoms.

Methods: An explorative retrospective review of clinical records was conducted for people with SPI(M)D without dementia (n = 103), with questionable dementia (n = 19), and with diagnosed dementia (n = 19). Presence and time of onset of symptoms were extracted and compared between groups.

Results: People with questionable dementia or diagnosed dementia had compared to people without dementia more symptoms regarding the cognitive, activities of daily living, behavioral/psychological, and motor domains. The most prevalent early symptoms were memory loss, declined walking skills, increased anxious, apathetic, and irritable behavior. Predictors for dementia were the number of cognitive, behavioral/psychological, and motor symptoms.

Conclusion: These results contribute to enhance our understanding of dementia in people with SPI(M)D, which is essential for earlier recognizing and diagnosing dementia.

Introduction

With aging, people are prone to develop age-related conditions such as dementia (Alzheimer's Association, 2022). Dementia is the overarching term for a group of symptoms associated with a progressive decline in cognitive functioning from an individual's previous level of functioning, which is severe enough to interfere with daily functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2018). Dementia has several causes, Alzheimer's disease (AD) being the most common one (Alzheimer's Association, 2022). Recognizing and diagnosing dementia and its underlying etiology requires proper understanding of its natural history.

A vast number of studies have described which (early) dementia symptoms are generally observed among people with dementia in the general population (among others: Brodaty et al., 2015; Engelborghs et al., 2005; Giebel et al., 2021; Gilmore-Bykovskyi et al., 2020; Hendriks et al., 2022; Jost & Grossberg, 1996; Priefer & Robbins, 1997; Ramakers et al., 2007) as well as among people with intellectual disabilities (ID) and dementia, particularly those with Down syndrome (DS) (among others: Arvio & Bjelogrlic-Laakso, 2021; Benejam et al., 2020; Blok et al., 2017; Cosgrave et al., 2000; Dekker et al., 2015, 2018, 2021a; Fonseca et al., 2020; Huxley et al., 2005; Moss & Patel, 1995; Nelson et al., 2001; Oliver et al., 1998; Temple & Konstantareas, 2005). DS is associated with an extremely high genetic risk of developing dementia due to AD (Ballard et al., 2016; Lott & Dierssen, 2010) and is actually considered according the International Working Group (IWG) 2 criteria as presymptomatic stage of AD (Dubois et al., 2014). While dementia research in people with ID is a growing field, only very few studies have focused on dementia in people SPI(M)D (Wissing, Ulgiati, et al., 2022).

Dementia in people with SPI(M)D may differ from that in the general population and even from people with mild(er) ID. After all, already at baseline, people with SPI(M)D have severe/profound limitations in intellectual and adaptive functioning, i.e., conceptual, social, and practical skills (Schalock et al., 2021). They also often experience serious physical health problems, sensory impairments, and motor disabilities (Nakken & Vlaskamp, 2007; van Timmeren et al., 2017). Because of pre-existing severe/profound disabilities, people with SPI(M)D have not attained

specific skills and often need lifelong support. As a result, never developed skills cannot alter and, therefore, cannot be considered as symptoms that may be indicative of dementia (Llewellyn, 2011; Sheehan, Sinai, et al., 2015). Moreover, dementia symptoms may be less noticeable in those with SPI(M)D: people with SPI(M)D have difficulty to self-report symptoms because their communication is limited and mainly non-verbal (Cooper & Smiley, 2007; Nakken & Vlaskamp, 2007). For the observation of symptoms, they thus depend on informants, such as family members and direct support professionals/caregivers (McKenzie et al., 2018).

The first studies to thoroughly identify practice-based observations of dementia symptoms in people with SPI(M)D have indicated which symptoms – aside from the pre-existing disabilities – are often observed by care professionals and family members (Dekker, Wissing et al., 2021a; Wissing, Fokkens, et al., 2022). Although cognitive changes, e.g., memory loss, are main indicators for dementia in the general and mild ID population (Jamieson-Craig et al., 2010; World Health Organization, 2018), changes in activities of daily living (ADL) as well as behavioral and psychological changes were more prominent in people with SPI(M)D (Wissing, Dijkstra, et al., 2022). Such changes may indeed be indicative of dementia but could also be caused by – often treatable – conditions such as depression, delirium, vision problems, hearing problems, hypothyroidism, medication use, sleep apnea, or vitamin B12 deficiency (Moriconi et al., 2015; Scott & Barrett, 2007). Furthermore, such changes might relate to 'normal' aging (Alzheimer's Association, 2022). Correctly differentiating between, i.e., attributing changes to, dementia, comorbidities, or aging, is important to prevent over- and underdiagnosed dementia. Accurately diagnosing dementia requires thus a thorough process of ruling out other potential causes and proper understanding of differences between dementia and aging. However, very little research has examined observed differences between people with SPI(M)D with and without dementia (Wissing, Ulgiati, et al., 2022).

Increasing knowledge about dementia in people with SPI(M)D is necessary to improve recognition and diagnosis of dementia in early stages. Early identification of dementia allows to timely respond to a person's changing wishes and needs by making informed choices (Dekker, Wissing et al., 2021a; Janicki, 2011). Care can, for example, be tailored to the individual with SPI(M)D and dementia (Chapman et al., 2018; Dekker, Wissing et al., 2021a).

Moreover, diagnostic errors – missed, wrong or delayed diagnosis – as well as incorrect treatments can be avoided (Dekker, Wissing et al., 2021a; Garcia et al., 1981). Early diagnosis also facilitates anticipation of the progression of dementia, for example, making choices about palliative care and end of life (Dekker, Wissing et al., 2021a; Hughes et al., 2007; Roger, 2006).

To enhance understanding of dementia in people with SPI(M)D, this study aimed to characterize the natural history of dementia in people with SPI(M)D by determining the prevalence and time of onset of symptoms.

Methods

Study consortium

This study was part of a larger research project designed to identify dementia symptoms in people with SPI(M)D and develop a dedicated dementia screening instrument for people with SPI(M)D. The project '*Practice-based questions about dementia in people with severe/profound intellectual (and multiple) disabilities*' (Dekker, Wissing et al., 2021a; Wissing, Dijkstra, et al., 2022; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022) is a collaborative effort of University of Groningen, University Medical Center Groningen (UMCG), Hanze University of Applied Sciences and with four Dutch care organizations spread across the country: Alliade, 's Heeren Loo, Ipse de Bruggen, and Royal Dutch Visio. In addition, data of people with SPI(M)D obtained within a similar study of clinical records in three Dutch care organizations Cosis, Philadelphia, and De Trans were used.

Study design

This study is an explorative retrospective analysis of clinical records of people with SPI(M)D. Different care organizations use different electronic clinical record systems. Nevertheless, for each participant, the same data were obtained from different components of clinical records, namely demographic information, physical examinations, diagnostic information, laboratory results, information about medication use, multidisciplinary consultations, psychological assessments, case notes drawn up by involved physicians, ID psychologists, and allied health care professionals.

Ethics and consent

The Medical Ethical Committee of the UMCG concluded that the Dutch Medical Research Human Subjects Act did not apply to this study (METc

2019/198). The study was registered in the UMCG Research Register (no. 201900193) and conducted in compliance with the UMCG Research Code and the EU General Data Protection Regulation. Legal representatives of people with SPI(M)D provided written informed consent for obtaining data from clinical records and processing/analyzing coded data for this study.

Participants

Participants were purposefully recruited through the participating care organizations according the following inclusion criteria: severe, severe to profound or profound ID that originated before the age of 22, aged ≥40 years, with/without the presence of diagnosed syndromes (e.g., DS) or other disabilities (e.g., visual or motor impairments), with/without questionable dementia or diagnosed dementia. Participants were excluded from this study if no intellectual disability level was reported or when they had mild, mild to moderate, moderate, or moderate to severe ID. ID psychologists working within the care organizations were asked to identify eligible participants. Legal representatives of identified eligible participants received an information letter with informed consent forms. After providing informed consent, the intellectual disability level was checked before extracting data from clinical records.

Data collection

To extract data from clinical records, a data extraction form was developed in consultation with the project team, students (medicine, nursing, physiotherapy, and physician assistant), and care professionals working with people with SPI(M)D and experienced in keeping clinical records. The draft version of the data extraction form was pilot tested by extracting data from clinical records of 10 participants. The pilot allowed to improve the clarity and efficiency of the data extraction form. The ease of use was further optimized by constructing the data collection form in REDCap (Harris et al., 2009), hosted within the secured network of the UMCG.

The final version of the data extraction form consisted of two parts. The first part focused on participants' characteristics, i.e., age, sex, living situation, attending day care, deaths, intellectual disability level, etiology of ID, a formal diagnosis of autism spectrum disorder, intelligence quotient, social-emotional functioning, baseline presence of verbal communication and walking skills. Additionally, information was collected about the presence

of treated or untreated conditions – cerebrovascular accident, chronic pain, depression, delirium, epilepsy, hearing problems, hypothyroidism, sleep apnea, vision problems, vitamin B12 deficiency – which could cause dementia-like symptoms (Moriconi et al., 2015; Scott & Barrett, 2007). Furthermore, data were extracted about psychoactive medication use. Finally, data were collected about the presence of questionable dementia or diagnosed dementia, including information about the first year an individual was suspected of having dementia, the year of clinical diagnosis, and the etiology of dementia.

The second part focused on extracting data about the prevalence and time of onset of symptoms. According to diagnostic dementia criteria (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2018) and literature (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a; Ries, 2018; Strydom et al., 2010) symptoms were categorized into five domains: cognitive symptoms, ADL symptoms, behavioral and psychological symptoms, motor symptoms, and medical comorbidities. Each domain consisted of symptoms observed in people with SPI(M)D, which were obtained in one or more of the previous studies concerning dementia symptoms in SPI(M)D (Dekker, Wissing et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). The total number of symptoms was 44, subdivided into 14 cognitive symptoms, 6 ADL symptoms, 11 behavioral and psychological symptoms, 10 motor symptoms, and 3 medical comorbidities.

The first domain contained 14 cognitive functions: awareness of proper order, judgment, language skills, losing objects, memory, object recognition, orientation in place, orientation in time, person recognition, planning, preference for (favorite) objects, problem solving, responsiveness, and understanding visual images/spatial relationships. Within the ADL domain, the six items concerned dressing, eating/drinking skills, grooming, showering/bathing, toilet use, and stair climbing. In the behavioral and psychological domain, the 11 items comprised aggressive, anxious, apathetic, depressive, disinhibited, eating/drinking, irritable, obstinate, psychotic, restless/stereotypic behavior, and sleeping problems. Motor functions were balance, choking, cramps, fall frequency, movement speed, muscle strength, stiffness, transfers/mobility, walking skills, and wheelchair use. Finally, epilepsy, incontinence, and weight formed the last domain.

Similar to the data extraction method of Jost and Grossberg (1996), we identified in clinical records the presence and time of onset of the symptoms. For each item, text fragments describing such an item, including the year in which a text fragment was written, were extracted from clinical records. If text fragments indicated a change, the item was coded as 'presence of symptom.' Given that dementia is characterized by a decline in cognitive and ADL functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2018), items within the cognitive and ADL domain were coded as 'presence of symptom that decreased' (except for losing objects, which was coded as 'presence of symptom that increased'). For behavioral and psychological symptoms, mainly an increase but also a decrease in frequency/severity of behavior may be observed (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a). Therefore, behavioral and psychological items were coded as either 'presence of symptom that increased' or 'presence of symptom that decreased.' Depending on the item, motor changes and changes in medical comorbidities were coded as 'presence of symptom that decreased' or 'presence of symptom that increased.' If the text fragments for a specific item indicated multiple changes, e.g., a decrease in 2018 and an increase in 2020, the first reported change was coded and added to the data extraction form. When text fragments did not comprise any indication of a change, the item was coded as 'absence of symptom.' Lastly, if no text fragments for a particular item were identified, the code 'not reported' was assigned to that item.

From June 2021 until September 2022, one researcher (M.B.G.W.) collected raw data and completed the data extraction form by coding the raw data for all participants, including the 10 clinical records of the pilot. Doubts about whether a symptom was absent or present were resolved in consultation with the project team. Two months after initial data extraction, the researcher coded identified item text fragments (in total 455) once more for a subset of 14 randomly selected participants, i.e., 10% of the total sample. The number of concordant codes was 429. Intracoder percent agreement, i.e., number of concordant codes/total number of identified item text fragments × 100 (Gisev et al., 2013), was 94.3%.

Data analysis

Extracted data were exported from REDCap to SPSS Statistics version 28 (IBM, Corp). Based on data about the absence/presence of questionable dementia or diagnosed dementia, participants were categorized into

three groups: 1) SPI(M)D without dementia, 2) SPI(M)D with questionable dementia. i.e., the individual was suspected of having dementia but does not (yet) clearly meet the diagnostic criteria, and 3) SPI(M)D with clinically diagnosed dementia. For each group, participants' characteristics were presented using descriptive statistics: chi-squared tests were used to compare categorical data and ANOVA to compare normally distributed continuous data (age) between groups.

To determine the prevalence and time of onset of dementia symptoms, we followed the analyzing method as described in the study by Jost and Grossberg (1996). Firstly, the prevalence, i.e., the proportion of individuals with diagnosed dementia exhibiting a symptom, was calculated for all identified symptoms. Secondly, the time of onset of symptoms was calculated by subtracting the year of diagnosis from the year at which the first change was reported. This could only be calculated if items were coded as 'presence of symptom that decreased' or 'presence of symptom that increased,' the year of diagnosis and year at which the first change was reported was known. Thereafter, the mean time of onset – separately for increase and decrease of a symptom – was calculated for each symptom. A time-density plot, in which the mean time of onset of a symptom was plotted against the prevalence of a symptom, was used to present results. Time zero represented the time of diagnosis. A negative time value indicated that the mean time of onset of a symptom was before the diagnosis and for a positive time value the mean time of onset of a symptom was after the diagnosis. In the plot also the mean time dementia was first suspected (year of diagnosis minus the first-year dementia was suspected) was displayed.

Furthermore, the prevalence of each symptom was also calculated for the group without dementia and the group with questionable dementia. Chi-squared tests were applied to identify differences in the prevalence between the three groups. When nothing was reported about an item in the clinical records, data were considered to be missing and thus not included in the analysis. Moreover, the number of symptoms per domain was calculated for each participant. Differences between the groups were compared using Kruskal–Wallis tests. Bonferroni–Dunn's multiple comparisons post hoc tests were carried out when significant differences were found.

Finally, multinomial logistic regression, with the odds ratio (OR) being the main outcome measure, was used to analyze whether the number of symptoms per domain – cognitive, ADL, behavioral and psychological, motor, and medical comorbidities – could predict whether a person had questionable dementia or diagnosed dementia. The group which had no dementia was considered as reference category to which the other two groups were compared.

Additionally, the analysis was performed again with the questionable dementia group as a reference category to also compare the questionable and diagnosed dementia group. For the statistical tests – except post hoc tests – a p-value <0.05 was considered significant.

Results

Legal representative of 266 identified eligible participants received an information letter with an informed consent form. Legal representatives of 168 eligible participants provided written informed consent, 19 did not provide consent, and 79 did not respond. Out of the 168 participants, 27 individuals were excluded based on exclusion criteria: no severe/profound ID ($n=14$), intellectual disability level not reported ($n=4$), deceased between consent and data extraction causing clinical records not to be accessible anymore ($n=9$). The 141 included participants were grouped by the absence/presence of questionable dementia or diagnosed dementia: 103 had no dementia, 19 had questionable dementia, and 19 had diagnosed dementia.

Participants' characteristics

All participants lived in residential facilities of care organizations and attended day care. Further participants' characteristics are presented separately for each group in Table 5.1. What stands out in this table is that the presence of a syndrome significantly differed between groups ($p<0.001$). Among the 103 persons without dementia, 9.7% had DS, whereas 52.6% and 63.2% had DS in the group with questionable and diagnosed dementia, respectively. Additionally, the baseline presence of walking skills differed significantly between groups ($p=0.009$). All persons with questionable dementia and 94.7% with diagnosed dementia were at baseline able to walk while this was true for 80.6% in the group without dementia. No significant differences were found between groups concerning the

prevalence of conditions that could cause dementia-like symptoms. Often nothing was reported about such conditions as chronic pain, sleep apnea, vitamin B12 deficiency, cerebrovascular accident and delirium in clinical records of those with questionable and diagnosed dementia. Lastly, the use of any psychoactive medication use (yes/no) ($p = 0.040$) and the total number of psychoactive medication used ($p = 0.001$) differed significantly between groups.

Prevalence and time of onset of symptoms

The time-density plot (Figure 5.1) displays the mean time of onset and prevalence of dementia symptoms in those with diagnosed dementia. Among the 19 persons, 7 individuals with DS also had a diagnosis of AD, 3 for whom the cause of intellectual disability was unknown had a diagnosis of vascular dementia, and for the remaining 9 (5 with DS and 4 with an unknown cause of intellectual disability) no etiology of dementia was reported. The mean time between the first suspicions of dementia and the clinical diagnosis was 5.7 years (SD = 4.0, min.–max. = 0–16 years). Figure 5.1 shows that aggressive behavior – either an increase or decrease – was reported earlier than the mean time dementia was first suspected. Moreover, within the 5.7 years before the diagnosis, the most prevalent early reported symptoms (75–100%) were decreased memory, walking skills and increased anxious, apathetic, and irritable behavior. Decreased orientation in place was also prevalent in more than 75% of individuals with dementia, and was reported two years before the diagnosis. Furthermore, early symptoms with a prevalence between 50 and 75% were increased depressive, restless/stereotypic behavior, and decreased language skills. In the four to two years before the diagnosis, commonly reported symptoms (50–75%) were increased incontinence, fall frequency, obstinate behavior, and decreased balance, transfers/mobility, movement speed, and dressing. Two years before diagnosis, there were five symptoms with a prevalence between 50% and 75%, namely decreased eating/drinking skills, orientation in time, weight and increased sleeping problems, and wheelchair use. Finally, decreased toilet use, responsiveness, and eating/drinking behavior were also prevalent symptoms in more than 50%, but were, generally, reported after the diagnosis of dementia.

Table 5.1 Characteristics of the three study groups.

Participants' characteristics	No dementia n=103	Questionable dementia n=19	Diagnosed dementia n=19	p
Age (years, mean ± SD (min.–max.))	64.6 ± 1.1 (43.0–89.0)	61.0 ± 8.9 (47.0–81.0)	65.6 ± 10.1 (48.0–85.0)	0.122
Sex (% female)	49.5	36.8	36.8	0.408
Deaths among included participants (%)	12.6	15.8	15.8	0.890
Intellectual functioning: severe, severe/profound, profound (%)	56.3, 1.0, 42.7	68.4, 5.3, 26.3	52.6, 10.5, 36.8	0.195
Presence of syndrome: DS, other genetic syndrome, no/unknown (%)	9.7, 14.6, 75.7	52.6, 0.0, 47.4	63.2, 0.0, 36.8	<0.001*
Autism spectrum disorder: formal diagnosis, signs but no diagnosis (%)	14.6, 27.2	5.3, 21.1	0.0, 21.1	0.449
IQ-score available (%)	2.9	10.5	10.5	0.225
Social-emotional functioning: 0-6 months, 6-18 months, 18-36 months, 3-7 years, not reported (%)	19.4, 18.4, 5.8, 1.9, 54.5	10.5, 15.8, 5.3, 10.5, 57.9	15.8, 10.5, 15.8, 57.9	0.318
Verbal communication: able, no longer, never (%)	39.8, 2.9, 57.3	52.6, 0.0, 47.4	42.1, 5.3, 52.6	0.679
Walking skills: able, never (%)	80.6, 19.4	100.0, 0.0	94.7, 5.3	0.009*
Conditions which could cause dementia-like symptoms		**Not reported (%)**		**Not reported (%)**	
Vision problems: treated, untreated (%)	25.2, 62.1	36.8, 52.6	0.0	42.1, 57.9	0.0	0.162
Hearing problems: treated, untreated (%)	15.5, 38.8	21.1, 52.6	0.0	26.3, 52.6	0.0	0.257
Epilepsy: treated, untreated (%)	48.5, 10.7	15.8, 15.8	5.3	47.4, 5.3	0.0	0.051
Hypothyroidism: treated, untreated (%)	10.7, 0.0	15.8, 5.3	10.5	36.8, 0.0	10.5	0.022
Depression: treated, untreated (%)	3.9, 1.0	10.5, 0.0	36.8	5.3, 0.0	52.6	0.870
Chronic pain: treated, untreated (%)	4.9, 0.0	10.5, 5.3	73.7	10.5, 0.0	63.2	0.380
Sleep apnea: treated, untreated (%)	1.0, 1.9	0.0, 5.3	78.9	0.0, 0.0	94.7	0.831
Vitamin B12 deficiency: treated, untreated (%)	0.0, 0.0	0.0, 0.0	42.1	0.0, 0.0	36.8	-
CVA (%)	10.7	15.8	63.2	21.1	63.2	0.616
Delirium (%)	1.0	0.0	100.0	5.3	94.7	0.306

Table 5.1 Continued.

Participants' characteristics	No dementia n=103	Questionable dementia n=19	Diagnosed dementia n=19	p
Psychoactive medication use				
Any psychoactive medication use (%)	72.8	47.4	63.2	0.040*
- *Antiepileptics (N03A, %)*	51.5	26.4	42.1	0.090
- *Antipsychotics (N05A, %)*	30.1	21.1	26.3	0.673
- *Anxiolytics (N05B, %)*	6.8	10.5	10.6	0.789
- *Hypnotics and sedatives (N05C, %)*	3.9	0.0	5.3	0.458
- *Antidepressants (N06A, %)*	14.6	10.5	15.8	0.862
- *Antidementia (N06D, %)*	0.0	0.0	0.0	-
- *Opioids (N02A, %)*	0.0	0.0	0.0	-
Number of psychoactive medications (% n=0, 1, 2, 3, 4, not reported)	19.4, 29.1, 26.2, 14.6, 2.9, 7.8	47.4, 31.6, 5.3, 0.0, 10.5, 5.3	31.6, 36.8, 0.0, 15.8, 10.5, 5.3	0.001*

With respect to the prevalence of (un)treated conditions which could cause dementia-like symptoms, one would expect that these conditions were ruled out in the diagnostic process of dementia. Therefore, the proportion of individuals for whom nothing was reported about the prevalence of these (un)treated conditions are presented for both the group with questionable dementia and the group with diagnosed dementia. To compare differences between groups, ANOVA was used for normally distributed continuous data (age) and chi-squared tests were used for categorical data. Symbol: *, $p < 0.05$. Abbreviations: CVA, cerebrovascular accident; DS, Down syndrome; max., maximum; min., minimum; SD, standard deviation.

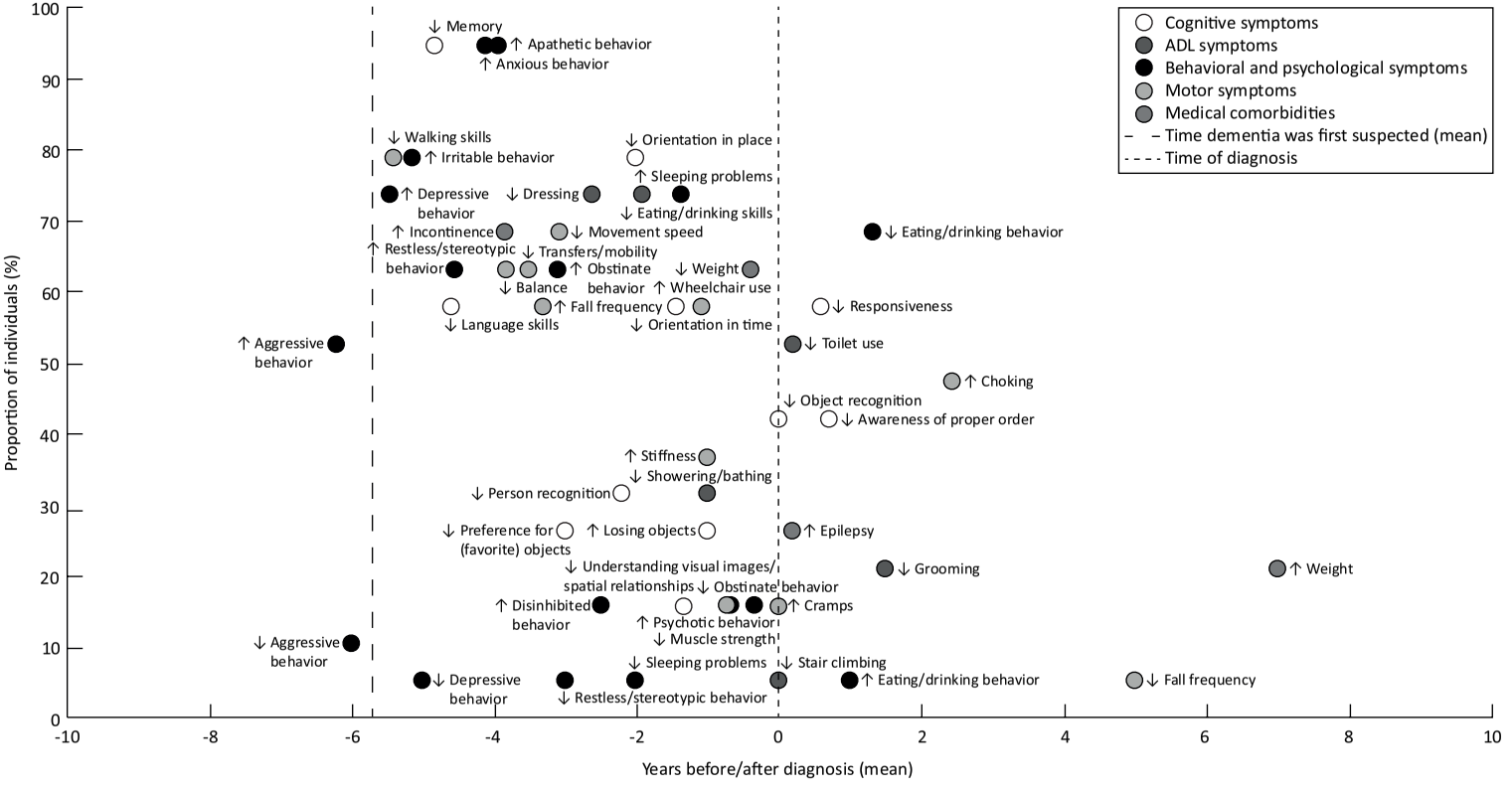


Figure 5.1 Time-density plot of dementia symptoms reported in clinical records of people with diagnosed dementia. The mean time of onset of a symptom is plotted against the prevalence of that symptom. Dashed lines represent the mean time dementia was first suspected -5.7years before diagnosis – and the time of diagnosis, respectively. Symbols: ↓, decrease; ↑, increase. Abbreviation: ADL, activities of daily living.

Differences in the prevalence of symptoms

Figures 5.2–5.6 visualizes the prevalence of cognitive symptoms, ADL symptoms, behavioral and psychological symptoms, motor symptoms, and medical comorbidities per group: no dementia, questionable dementia, and diagnosed dementia. What stands out in these figures is that for the majority of symptoms the proportion of individuals exhibiting the symptom was lowest in the group without dementia, intermediate for those with questionable dementia and highest in the group with diagnosed dementia. Significant differences in the prevalence of symptoms between groups were found within all domains, which are explained in more detail in the subsequent paragraphs.

Cognitive symptoms

Between groups, the following cognitive symptoms differed significantly: memory, orientation in place, orientation in time, language skills, responsiveness, object recognition, awareness of proper order, and person recognition (all p-values<0.001). For all eight symptoms, the proportion of individuals exhibiting the symptom was lowest in the group without dementia and highest in the group with diagnosed dementia. Preference for (favorite) objects differed significantly between groups as well (p = 0.011). Particularly, people with diagnosed dementia showed a decrease in preference for (favorite) objects. Regarding judgment, for which the groups also significantly differed (p = 0.020), only some persons with questionable dementia showed decreased judgment (Figure 5.2).

ADL symptoms

ADL symptoms that differed significantly between groups (all p-values<0.05) were eating/drinking skills, dressing, toilet use, showering/bathing, and grooming. For these symptoms, the prevalence of a reported decrease was lowest in the group without dementia and highest in the group with diagnosed dementia (Figure 5.3).

Proportion of individuals (%)
0
20
40
60
80
100
Decrease
Increase
Unaltered
Not reported
Memory
Orientation in place
Orientation in time
Language skills
Responsiveness
Object recognition
Awareness of proper order
SPI(M)D
SPI(M)D + QD
SPI(M)D + D
15.5
35.9
48.5
63.2
5.3
31.6
94.7
5.3
14.6
69.9
15.5
57.9
26.3
15.8
78.9
10.5
10.5
5.8
63.1
31.1
26.3
57.9
15.8
13.6
86.4
42.1
57.9
57.9
42.1
7.8
56.3
35.9
47.4
26.3
26.3
57.9
31.6
10.5
1.0
65.0
34.0
10.5
63.2
26.3
42.1
36.8
21.1
5.8
71.8
22.3
26.3
52.6
21.1
42.1
42.1
15.8
p<0.001*

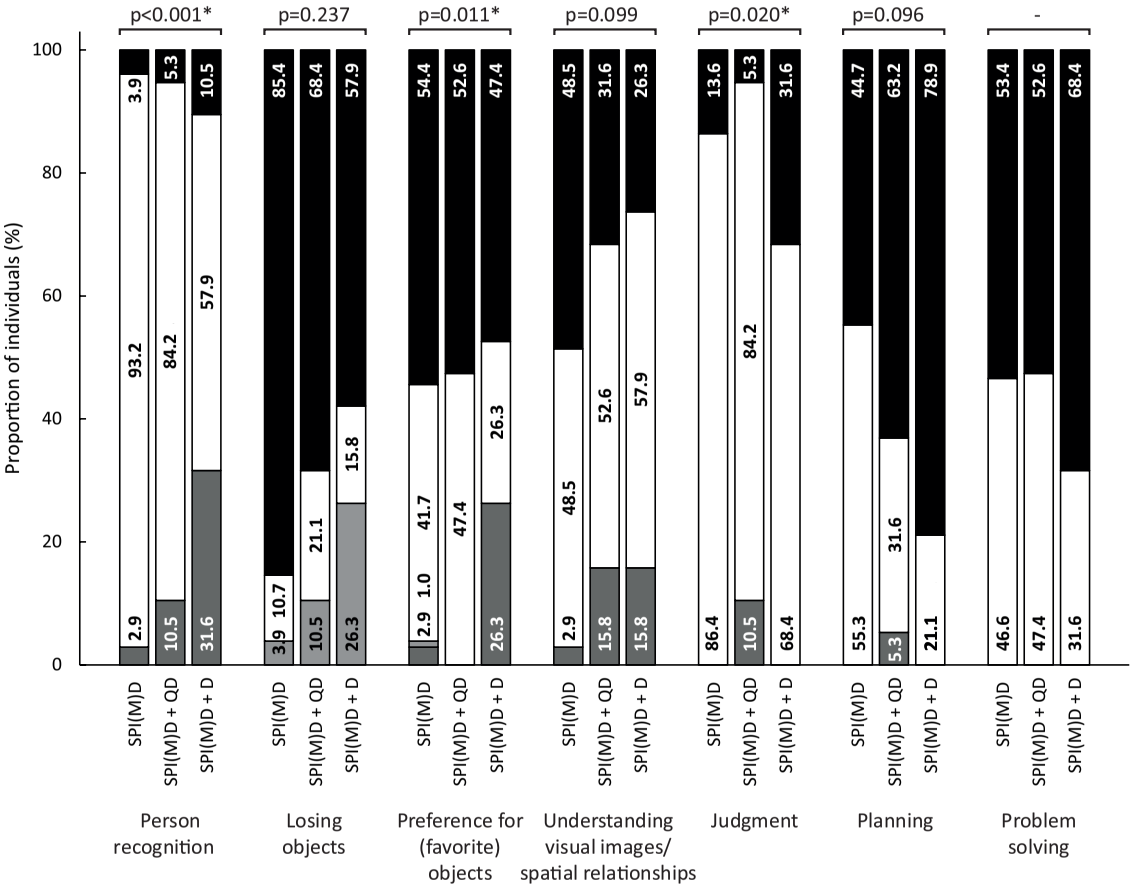


Figure 5.2 Prevalence of cognitive symptoms per group: no dementia (SPI(M)D), questionable dementia (SPI(M)D + QD) and diagnosed dementia (SPI(M)D + D). From left to right, symptoms are presented from most to least frequently reported for those with diagnosed dementia. Chi-squared tests were used to identify differences between groups. Symbol: *, $p < 0.05$. Abbreviation: SPI(M)D, severe/profound intellectual (and multiple) disabilities.

Figure 5.3 Prevalence of activities of daily living (ADL) symptoms per group: no dementia (SPI(M)D), questionable dementia (SPI(M)D+QD) and diagnosed dementia (SPI(M)D+D). From left to right, symptoms are presented from most to least frequently reported for those with diagnosed dementia. Chi-squared tests were used to identify differences between groups. Symbol: *, $p < 0.05$. Abbreviation: SPI(M)D, severe/profound intellectual (and multiple) disabilities.

Behavioral and psychological symptoms

A substantial number of behavioral and psychological symptoms were found to differ significantly between groups, namely anxious, apathetic, obstinate, depressive, irritable, restless/stereotypic, and aggressive behavior (all p-values<0.05). For these seven symptoms, the proportion of individuals exhibiting a symptom – either increase or decrease – was, again, lowest in the group without dementia and increased toward the group with diagnosed dementia. Sleeping problems ($p = 0.027$) as well as eating/drinking behavior ($p = 0.002$) significantly differed between groups. For these two symptoms, the prevalence was highest in the group with questionable dementia (Figure 5.4).

Motor symptoms

Seven out of ten motor symptoms significantly differed between groups (all p-values<0.05): walking skills, movement speed, balance, fall frequency, wheelchair use, choking, and stiffness. For the symptoms movement speed, fall frequency, wheelchair use, and choking the prevalence was highest in the diagnosed dementia group, whereas for walking skills and stiffness the prevalence was highest in the group with questionable dementia. The prevalence for decreased balance was similar for the group with questionable and diagnosed dementia but lower in the group without dementia (Figure 5.5).

Medical comorbidities

Concerning medical comorbidities, only incontinence differed significantly between groups ($p = 0.002$). The proportion of individuals showing increased incontinence was lowest in the group without dementia and highest in the group with diagnosed dementia (Figure 5.6).

Decrease
Increase
Unaltered
Not reported
Proportion of individuals (%)
0
20
40
60
80
100
Anxious behavior
Apathetic behavior
Sleeping problems
Obstinate behavior
Depressive behavior
Irritable behavior
SPI(M)D
SPI(M)D + QD
SPI(M)D + D
p<0.001*
p=0.027*
8.7
17.5
59.2
14.6
68.4
31.6
94.7
5.3
6.8
29.1
53.4
10.7
68.4
26.3
5.3
94.7
10.7
51.5
36.9
1.0
84.2
15.8
5.3
73.7
21.1
6.8
3.9
68.9
20.4
52.6
42.1
5.3
15.8
63.2
21.1
14.6
22.3
49.5
13.6
68.4
26.3
5.3
5.3
73.7
15.8
5.3
1.0
23.3
50.5
25.2
73.7
10.5
15.8
78.9
10.5
10.5

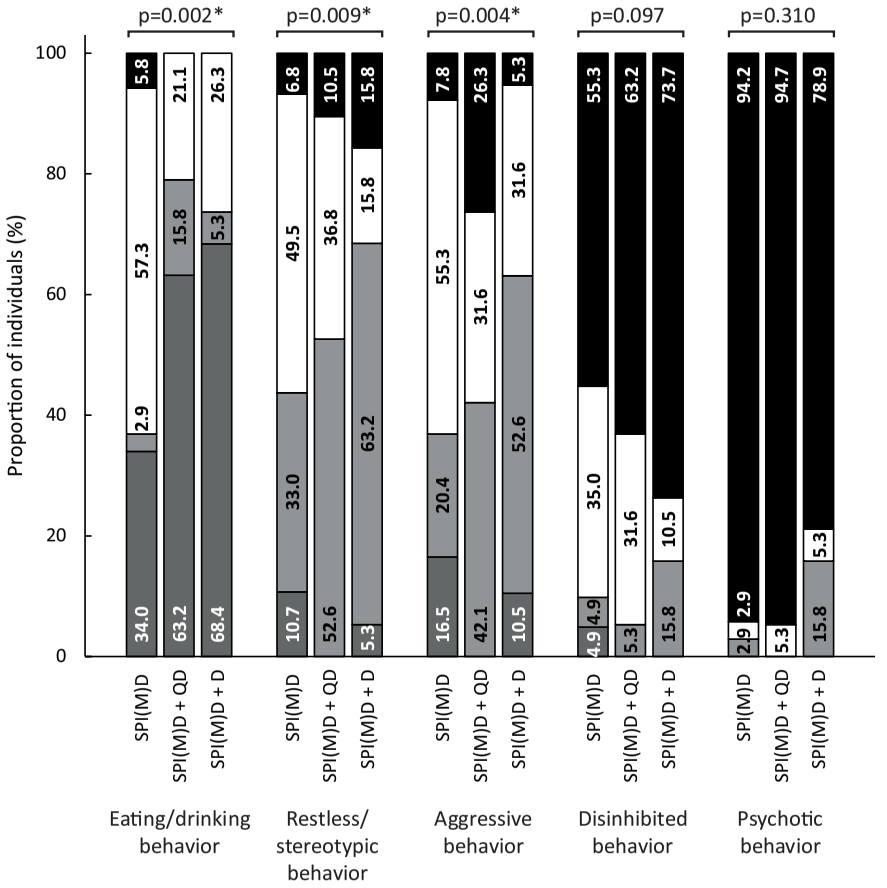


Figure 5.4 Prevalence of behavioral and psychological symptoms per group: no dementia (SPI(M)D), questionable dementia (SPI(M)D + QD) and diagnosed dementia (SPI(M)D + D). From left to right, symptoms – either decrease or increase – are presented from most to least frequently reported for those with diagnosed dementia. Chi-squared tests were used to identify differences between groups. Symbol: *, $p < 0.05$. Abbreviation: SPI(M)D, severe/profound intellectual (and multiple) disabilities.

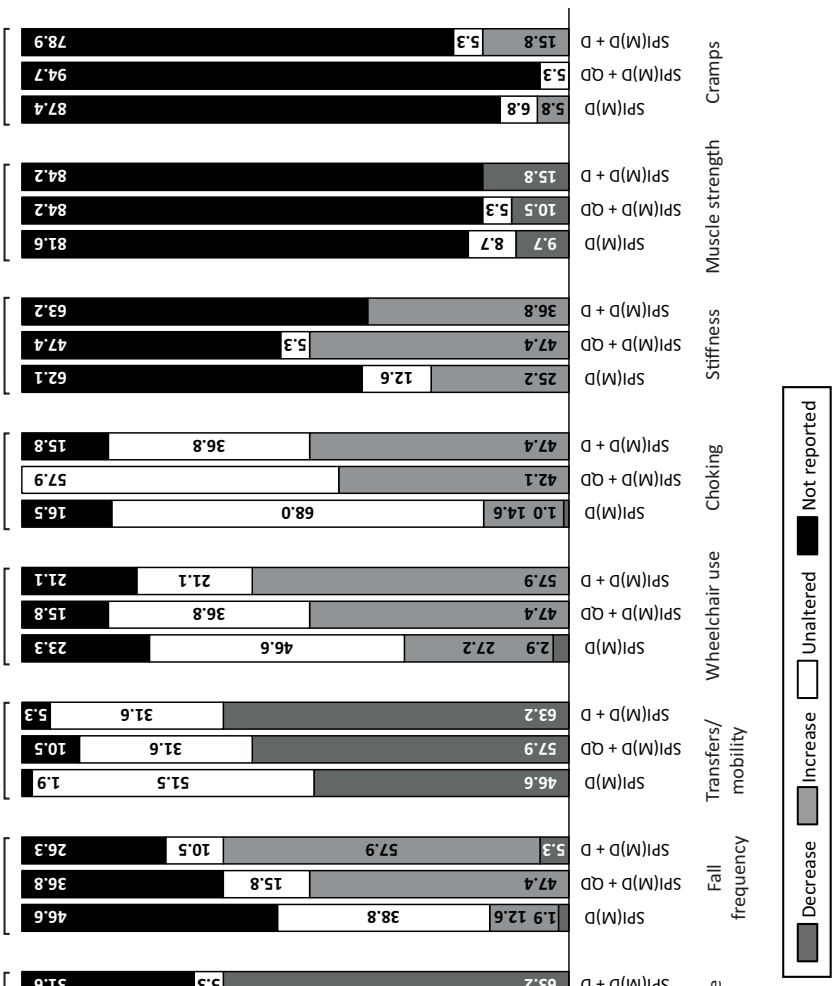


Figure 5.5 Prevalence of motor symptoms per group: no dementia (SPI(M)D), questionable dementia (SPI(M)D + QD) and diagnosed dementia (SPI(M)D + D). From left to right, symptoms – either decrease or increase – are presented from most to least frequently reported for those with diagnosed dementia. Chi-squared tests were used to identify differences between groups. Symbol: *, $p < 0.05$. Abbreviation: SPI(M)D, severe/profound intellectual (and multiple) disabilities.

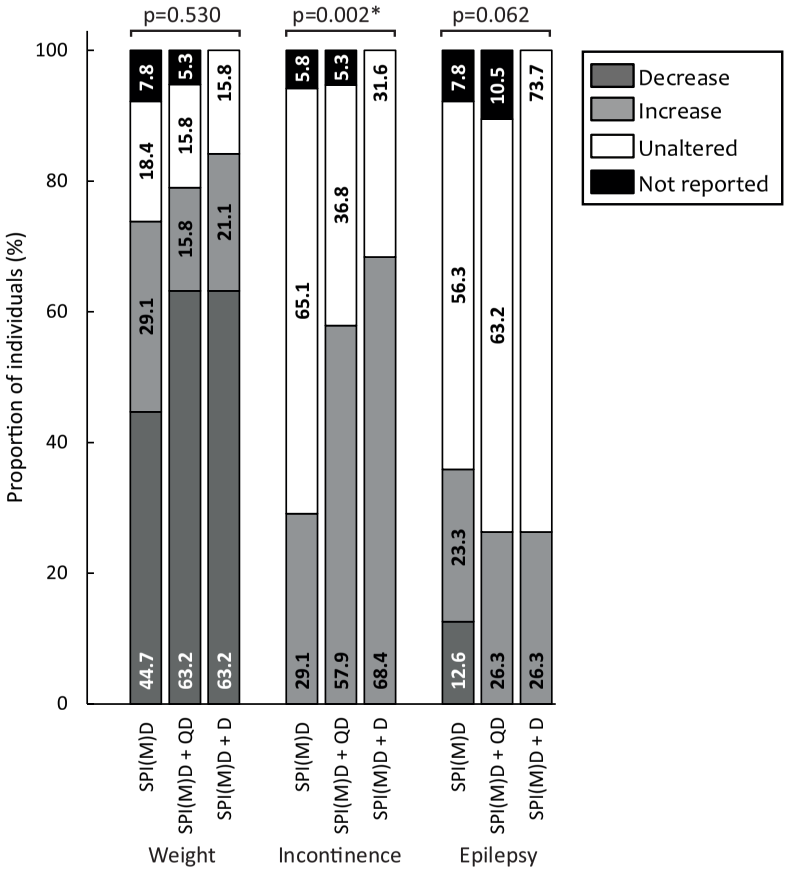


Figure 5.6 Prevalence of medical comorbidities per group: no dementia (SPI(M)D), questionable dementia (SPI(M)D + QD) and diagnosed dementia (SPI(M)D + D). From left to right, symptoms – either decrease or increase – are presented from most to least frequently reported for those with diagnosed dementia. Chi-squared tests were used to identify differences between groups. Symbol: *, $p < 0.05$. Abbreviation: SPI(M)D, severe/profound intellectual (and multiple) disabilities.

Differences in the number of symptoms per domain

The number of symptoms for each domain are displayed per group in Table 5.2. Except for medical comorbidities, the number of symptoms per domain was lowest in the group without dementia and highest for those with diagnosed dementia. Significant differences between groups were found for cognitive, ADL, behavioral and psychological, and motor domain (all *p*-values<0.001). Post hoc tests showed that significantly more symptoms were reported for people with questionable dementia and people with diagnosed dementia compared to those without dementia (all *p*-values<0.05), whereas no differences were found between the group with questionable dementia and the group with diagnosed dementia.

Table 5.2 Number of symptoms per domain.

Domain	No dementia n=103	Questionable dementia n=19	Diagnosed dementia n=19	p
Cognitive symptoms	0 (1), 0 – 7	3 (3), 0 – 8	6 (3), 2 – 8	<0.001*
ADL symptoms	1 (1), 0 – 5	2 (1), 0 – 5	2 (3), 0 – 6	<0.001*
Behavioral and psychological symptoms	3 (2), 0 – 9	6 (3), 2 – 9	8 (1), 4 – 10	<0.001*
Motor symptoms	2 (3), 0 – 7	4 (4), 0 – 9	6 (3), 0 – 9	<0.001*
Medical comorbidities	1 (1), 0 – 3	2 (1), 1 – 3	2 (2), 0 – 3	0.120

The number of symptoms for each domain (median (interquartile range), min.–max.) are presented separately for the groups. Kruskal-Wallis tests were used to compare differences between groups. Symbol: *, p < 0.05. Abbreviation: ADL, activities of daily living.

Predictors of dementia

The number of cognitive symptoms (OR = 2.12), behavioral and psychological symptoms (OR = 1.67), and motor symptoms (OR = 1.34) significantly predicted whether a person had questionable dementia versus no dementia (Table 5.3). The number of cognitive symptoms (OR = 3.92), behavioral and psychological symptoms (OR = 3.39), and motor symptoms (OR = 1.67) were also significant predictors for having diagnosed dementia versus no dementia. In both comparisons, the number of cognitive symptoms was associated with the highest risk for either questionable (OR = 2.12) or diagnosed dementia (OR = 3.92), followed by the number of behavioral and psychological symptoms (OR = 1.67 and 3.39, respectively). Lastly, persons with questionable dementia were compared to those with diagnosed dementia. The number of cognitive symptoms (OR = 1.85) as well as the number of behavioral and psychological symptoms (OR = 2.03) significantly predicted whether a person had diagnosed dementia, but not the number of motor symptoms (Table 5.3).

Table 5.3 Predictive values of the number of symptoms per domain for the development of dementia.

Domain	No dementia versus questionable dementia[I]		No dementia versus diagnosed dementia[I]		Questionable versus diagnosed dementia[II]	
	OR (95% CI)	p	OR (95% CI)	p	OR (95% CI)	p
Cognitive symptoms	2.12 (1.38–3.26)	<0.001*	3.92 (2.05–7.51)	<0.001*	1.85 (1.08–3.17)	0.025*
ADL symptoms	0.67 (0.36–1.29)	0.240	0.44 (0.18–1.08)	0.730	0.65 (0.31–1.37)	0.260
Behavioral and psychological symptoms	1.67 (1.20–2.32)	0.002*	3.39 (1.61–7.16)	0.001*	2.03 (1.01–4.11)	0.048*
Motor symptoms	1.34 (1.01–1.78)	0.042*	1.67 (1.06–2.64)	0.028*	1.25 (0.82–1.90)	0.310
Medical comorbidities	0.89 (0.37–2.11)	0.790	0.46 (0.12–1.75)	0.250	0.52 (0.16–1.70)	0.280

Multinomial logistic regression was used to identify predictors for the development of dementia. Symbols: I, the reference category was no dementia; II, the reference category was questionable dementia; *, $p < 0.05$. Abbreviations: ADL, activities of daily living; CI, confidence intervals; OR, odds ratio.

Discussion

This explorative study aimed to characterize the natural history of dementia in people with SPI(M)D by determining the prevalence and time of onset of symptoms. Regarding the prevalence of symptoms, the results showed that the majority of symptoms were more frequently reported when people had questionable dementia and most prevalent when dementia was diagnosed. People with questionable dementia or diagnosed dementia had in total more cognitive, ADL, behavioral and psychological, and motor symptoms than those without dementia. With respect to the time of onset of symptoms, the results showed that the most frequent early symptoms were memory loss, declined walking skills, increased anxious, apathetic, and irritable behavior. The earliest symptom was aggressive behavior, for which mainly an increase but also a decrease in frequency/severity was reported. Before the diagnosis also changes in ADL, i.e., decreased dressing and eating/drinking skills, and medical comorbidities, i.e., increased incontinence and weight loss, were reported. The number of cognitive symptoms, behavioral and psychological symptoms, and motor symptoms were predictive for questionable dementia and for diagnosed dementia.

Cognitive changes

One of the earliest sign of dementia, particularly of AD, is memory loss (Alzheimer's Association, 2022; Stern et al., 1993). This study confirms that memory loss also appeared in almost all persons with SPI(M)D and diagnosed dementia, on average already 4.8 years before the diagnosis. This result is likely to be related to the large number of people having DS, predisposed to develop AD with memory decline as a predominant symptom (Ballard et al., 2016; Lott & Dierssen, 2010). A decline in cognitive functions, such as memory can, however, also be part of 'normal' aging (Alzheimer's Association, 2022; Deary et al., 2009). Results showed that cognitive alterations were indeed observed in people without dementia as well, though cognitive symptoms were more common in those with questionable dementia or diagnosed dementia. In fact, results showed that cognitive symptoms were associated with a high risk on developing dementia, which is in line with findings in the general and the population with mild ID (Jamieson-Craig et al., 2010; Ramakers et al., 2007; World Health Organization, 2018). Overall, findings of this study thus indicate that alterations in cognitive functions – aside from pre-existing cognitive

limitations – are indicative for dementia in people with SPI(M)D (Dekker, Wissing et al., 2021a; Wissing, Fokkens, et al., 2022).

Behavioral and psychological changes

In all types of dementia, behavioral and psychological alterations are noticeable (Engelborghs et al., 2005; Finkel, 2000). The results of this study and previous studies demonstrated that behavioral and psychological changes are commonly observed in people with SPI(M)D and dementia (Dekker, Wissing et al., 2021a; Wissing, Fokkens, et al., 2022). In line with findings in the general population (Hendriks et al., 2022; Ramakers et al., 2007), this study also demonstrated that behavioral changes are predictive for dementia specifically in people with SPI(M)D. Before diagnosis, increased anxious, apathetic, irritable, depressive and restless/stereotypic, obstinate behavior, and sleeping problems were reported. Moreover, these symptoms were also prevalent in more than half of the group with questionable dementia. Consistent with findings of dementia in people with DS (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a), this indicates that these symptoms are likely early 'alarm signals' for dementia in people with SPI(M)D. The earliest dementia symptom was aggressive behavior, which is contrary to previous findings which showed that aggressive behavior was observed after AD diagnosis in the general population (Jost & Grossberg, 1996). A possible explanation for this might be that care professionals particularly report aggression earlier, because aggressive behavior may be disturbing and harmful for the individual as well as their fellow residents, caregivers, and family members (Emerson, 2001; Jones & Kroese, 2007; Sheehan, Hassiotis, et al., 2015).

Motor changes

Not only cognitive symptoms and behavioral and psychological symptoms but also motor symptoms were found to be predictive for dementia in people with SPI(M)D. This finding is in agreement with previous research on dementia in the general population, reporting that gait disturbances predict dementia (Ramakers et al., 2007). Interestingly, in our study decline in walking skills was an early frequently reported motor symptom, which could only be observed if individuals are able to walk at baseline (Wissing, Fokkens, et al., 2022). The presence of baseline walking skills was higher in people with questionable dementia or diagnosed dementia than in those without dementia. It is likely that dementia is underrecognized and -diagnosed in people with SPI(M)D who have profound motor disabilities because certain

symptoms like a decline in walking skills cannot be recognized within those persons (Dekker, Wissing et al., 2021a; Wissing, Dijkstra, et al., 2022; Wissing, Fokkens, et al., 2022).

ADL changes

The number of ADL symptoms differed between those with/without questionable dementia or diagnosed dementia, but was no predictor. This might be related to the pre-existing limitations in the ability to perform ADL (Wissing, Dijkstra, et al., 2022; Wissing, Fokkens, et al., 2022), which vary among people with SPI(M)D (Nakken & Vlaskamp, 2007). Changes in ADL are less noticeable when people only perform small tasks within an ADL task and are not at all noticeable when someone fully dependent on others for performing ADL. Despite pre-existing limitations, ADL symptoms, like decline in eating/drinking skills, dressing, and toilet use were often reported in people with SPI(M)D and questionable dementia or diagnosed dementia. Previous dementia research in the general population described that ADL deteriorate particularly in later stages of dementia (Giebel et al., 2021; Marshall et al., 2012). In line with these findings, ADL symptoms are in those with SPI(M)D and diagnosed dementia generally later reported than cognitive symptoms, behavioral and psychological symptoms, and motor symptoms. This potentially also has to do with the tendency of caregivers and family members to provide more support if needed without being aware that the subtle decline in ADL functioning can be due to dementia.

Medical comorbidities

Medical comorbidities were no predictor for dementia in people with SPI(M)D. Differences in medical comorbidities between people with/without questionable dementia or diagnosed dementia were only found for incontinence. This might be related to the fact that people with SPI(M)D already experience physical health problems like incontinence and epilepsy (Nakken & Vlaskamp, 2007; van Timmeren et al., 2017).

Strengths

To the best of our knowledge, this explorative study is the first to extensively describe the prevalence and time of onset of dementia symptoms in people with SPI(M)D. This knowledge is of great essence to improve early recognition and diagnosis of dementia in people with SPI(M)D. Furthermore, a strength of this study is that not only the cognitive domain – main indicators for dementia in the general population and mild ID population

(Jamieson-Craig et al., 2010; World Health Organization, 2018) – but also other domains, i.e., ADL, behavioral and psychological, motor domain and medical comorbidities were considered. To that end, we used previously found dementia symptoms in people with SPI(M)D (Dekker, Wissing et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). Lastly, a strength is the use of the time-density plot, which is a convenient method of demonstrating both the prevalence of symptoms as well as the onset of symptoms over time in people with dementia (Jost & Grossberg, 1996).

Limitations

Extracting data from clinical records allowed to identify if and when symptoms were reported. However, reliance on clinical records can also be considered as a limitation since reports could be incomplete: symptoms might have been observed but not reported or were underrecognized and thus not reported. Especially since knowledge about dementia in people with SPI(M)D is limited, it is conceivable that care professionals are often not educated sufficiently to recognize all symptoms, especially not in early stages. Moreover, documentation varied among care organizations. For example, certain organizations extensively reported on motor skills, whereas others reported on different prioritized areas. This may have resulted in underreporting symptoms in less prioritized areas. Altogether, these limitations may have caused an underestimation of the prevalence of symptoms. Another limitation of this study is that no second researcher independently extracted and coded data from (part of) the clinical records, and thus intercoder percent agreement could not be determined. Furthermore, a limitation was that the sizes of the subgroups were rather unequal. In each of the two groups of interest, i.e., questionable dementia and diagnosed dementia, 19 participants were included. The small subgroup sizes seem to be related to the complexity of recognizing and diagnosing dementia in people with SPI(M)D. Until recently, hardly any literature about dementia in people with SPI(M)D was available, and therefore it is very likely that dementia is underdiagnosed in people with SPI(M)D (Wissing, Ulgiati, et al., 2022). Despite the small subgroup sizes, the results of this study contribute to a better understanding of the natural history of dementia in people with SPI(M)D, which is essential to (earlier) recognize and diagnose dementia. Reducing underdiagnosis of dementia would allow future studies to more easily include larger and more equally divided study groups. Moreover, with a larger sample size also similarities and differences in the natural history of dementia in those with SPI(M)D with and without DS could be examined.

Future implications

Due to the complexity of recognizing and diagnosing dementia, dementia is diagnosed later in people with SPI(M)D (5.7 years after dementia was first suspected) compared to the general population (2.7 years after the onset of symptoms (Jost & Grossberg, 1995)). Earlier diagnosing dementia, and thus preventing delayed or underdiagnosed dementia, requires a diagnostic procedure dedicated to people with SPI(M)D. First of all, conditions causing dementia-like symptoms should be more thoroughly ruled out (differential diagnosis). This study showed that often nothing was reported about the presence of such treated and untreated conditions in people with questionable dementia or diagnosed dementia. Enhancing knowledge and understanding about dementia in people with SPI(M)D would also significantly benefit the process of diagnosing dementia. To facilitate the diagnostic process, dementia-related changes should be systematically identified and monitored by, for example, using a dementia screening instrument. However, today no such standardized instrument exists for people with SPI(M)D (Wissing, Dijkstra, et al., 2022). Therefore, there is a need to develop a tool to aid the diagnosis of dementia in people with SPI(M)D. Additionally, videotaping could potentially become a standard part of dementia diagnostics in people with SPI(M)D because it allows to examine the onset and progression of subtle dementia symptoms in more detail. Overall, steps toward an early diagnostic process and thus early diagnosis of dementia in people with SPI(M)D can only be undertaken when symptoms are recognized early by informants, such as family members and caregivers. Therefore, it is essential to develop training products about dementia in people with SPI(M)D to increase informants' knowledge.

Conclusion

This explorative study focused on the natural history of dementia in people with SPI(M)D. Presence and time of onset of symptoms were extracted from clinical records of purposefully selected participants with SPI(M)D with/without questionable dementia or diagnosed dementia. Differences in the prevalence of symptoms were found between those with/without questionable dementia or diagnosed dementia. Most symptoms were more common in people with questionable dementia and most prevalent in those with diagnosed dementia. People with questionable dementia or diagnosed dementia showed compared to those without dementia more cognitive, ADL, behavioral and psychological, and motor symptoms. Regarding the time of onset, memory loss, declined walking skills and increased anxious, apathetic, and irritable behavior were found to be early signs of dementia, present in almost all people with diagnosed dementia. The number of cognitive symptoms, behavioral and psychological symptoms, and motor symptoms were predictors for questionable dementia and diagnosed dementia. Together these results provide important insight into the natural history of dementia in people with SPI(M)D, which is essential to early recognize and diagnose dementia in people with SPI(M)D.

Chapter 6

Applicability of items in dementia screening instruments for people with intellectual disabilities

Wissing, M. B. G., Dijkstra, R., van der Wal, I. A., Grootendorst, E. S., Hobbelen, J. S. M., van der Putten, A. A. J, De Deyn, P. P., Waninge, A., & Dekker, A. D. (2022). Dementia in people with severe/profound intellectual (and multiple) disabilities: Applicability of items in dementia screening instrument for people with intellectual disabilities. *Journal of Mental Health Research in Intellectual Disabilities, 15*(4), 322-363.

Abstract

Introduction: Diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D) is complex. Whereas existing dementia screening instruments as a whole are unsuitable for this population, a number of individual items may apply. Therefore, this study aimed to identify applicable items in existing dementia screening instruments.

Methods: Informant interviews about 40 people with SPI(M)D were conducted to identify applicable items in the Dementia Scale for Down Syndrome, Behavioral and Psychological Symptoms of Dementia in Down Syndrome II scale, Dementia Questionnaire for persons with Mental Retardation and Social competence Rating scale for people with Intellectual Disabilities.

Results: Among 193 items, 101 items were found applicable, categorized in 5 domains: behavioral and psychological functioning (60 items), cognitive functioning (25 items), motor functioning (6 items), activities of daily living (5 items), and medical comorbidities (5 items).

Conclusion: Identifying applicable items for people with SPI(M)D is an essential step in developing a dedicated dementia screening instrument for this population.

Introduction

People with intellectual disabilities (ID) grow older, which is driven by improvements in medical and social care (Bittles & Glasson, 2004; Coppus, 2013; E. Evans et al., 2013). Advancing age substantially increases the risk of developing dementia (Alzheimer's Association, 2022). Consequently, dementia is becoming increasingly prevalent among people with ID. Moreover, Down syndrome (DS) is associated with an extremely high genetic risk of developing Alzheimer's disease dementia (Ballard et al., 2016).

Recognizing and diagnosing dementia in people with ID is a major challenge. Dementia is characterized by a decline from an individual's previous level of cognitive functioning, which is sufficient enough to significantly interfere with daily functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). In people with ID, it is complex to differentiate cognitive limitations resulting from the underlying ID from cognitive deficits due to dementia (Ball et al., 2004). Dementia assessment should thus focus on recognizing a deterioration in (cognitive) functioning relative to the premorbid limitations in functioning (Prasher, 2009). The lower the level of baseline functioning, the more difficult the assessment becomes. Therefore, diagnosing dementia is particularly challenging in people with severe/profound intellectual (and multiple) disabilities (abbreviated as SPI(M)D), that is, an estimated intelligence quotient (IQ) of less than 35 points (E. Evans et al., 2013; McKenzie et al., 2018).

In the general population, direct neuropsychological tests are used to identify changes in cognitive functioning associated with dementia (Alzheimer's Association, 2022; Salmon & Bondi, 2009). However, there are hardly any validated and feasible direct neuropsychological tests to aid the diagnosis of dementia for people with SPI(M)D (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). Direct neuropsychological tests are not suitable for people with SPI(M)D, because they require skills such as proper understanding of test instructions and good verbal communication skills, which are very limited in individuals with SPI(M)D (Nieuwenhuis-Mark, 2009; Oliver & Kalsy, 2005). Consequently, floor effects occur when conducting these tests with people having SPI(M)D, making them unsuitable for detecting a decline in cognitive functioning (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher

et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018).

Alternatively, informant-based dementia screening instruments, that is, interviews with or self-administered questionnaires filled out by direct support professionals/caregivers and/or family members, are used to aid the diagnosis of dementia. A number of informant-based instruments are available for people with ID. Recommended and commonly used instruments are, for instance, the Dementia Questionnaire for Learning Difficulties (DLD) previously referred to as the Dementia Questionnaire for Persons with Mental Retardation (DMR; Evenhuis, 1992; Evenhuis et al., 2006; Walker et al., 2015; Zeilinger et al., 2022), the Cambridge Examination for Mental Disorders of Older People with Down's syndrome and Others with Intellectual Disabilities (CAMDEX-DS; Ball et al., 2006; Zeilinger et al., 2022). Nevertheless, various studies indicate that such commonly used scales are not suitable for people with SPI(M)D (Elliott-King et al., 2016; Evenhuis, 1990; Hon et al., 1999; Margallo-Lana et al., 2007).

Today, no standardized dementia screening instruments dedicated to people with SPI(M)D exist. A diagnosis in this population is currently based on multidisciplinary clinical assessment involving observations, informant interviews, and/or screening case notes (Day, 1985; Duggan et al., 1996; Evenhuis, 1990; Määttä et al., 2006; Margallo-Lana et al., 2007; Reid & Aungle, 1974; Sauna-Aho et al., 2018). Improving the diagnostic procedures requires developing a dedicated dementia screening instrument specifically adapted to dementia symptoms observed in people with SPI(M)D. However, literature on dementia in this population is scarce (Wissing, Ulgiati, et al., 2022). Therefore, previous studies have identified dementia symptoms in this population through practice-based observation in order to develop a dedicated instrument (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022). In addition, whereas existing instruments as a whole are considered unsuitable for diagnosing dementia in people with SPI(M)D, specific items within those instruments may still be applicable for this population. Therefore, this study aimed to identify applicable items for people with SPI(M)D in already existing dementia screening instruments available for people with ID.

Methods

Study consortium

This study is part of the research project '*Practice-based questions about dementia in people with severe/profound intellectual (and multiple) disabilities*' (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022), a collaborative effort of University of Groningen, University Medical Center Groningen (UMCG) and Hanze University of Applied Sciences with four care organizations throughout The Netherlands (Alliade, 's Heeren Loo, Ipse de Bruggen, Royal Dutch Visio). These care organizations are representative for the Dutch intellectual disability care sector given the high number of people with SPI(M)D for whom they provide diagnostic work-up, treatments, and deliver care.

Study design

In this explorative study, applicable items for people with SPI(M)D were identified within dementia screening instruments available for people with ID. Four instruments frequently used in The Netherlands were examined 1) adapted Dutch version of the Dementia Scale for Down Syndrome (DSVH; Maaskant & Hoekman, 2011) 2) Behavioral and Psychological Symptoms of Dementia in Down Syndrome evaluation scale version II (BPSD-DS II; Dekker, Sacco, et al., 2018; Dekker, Ulgiati, et al., 2021a), 3) Dutch Dementia Questionnaire for persons with Mental Retardation (DVZ; Evenhuis et al., 1998) and 4) Social competence Rating scale for people with ID (SRZ; Kraijer et al., 2004).

These four instruments are not only in The Netherlands, but also internationally, recommended and widely used to screen for dementia in people with ID. For instance, a recent review of Zeilinger et al., (2022) recommended the usage of the BPSD-DS II and DLD (in Dutch: DVZ). The DLD is one of the most frequently used instrument for dementia assessment in people with ID (among others: Burt et al., 2005; Coppus, 2017; Coppus et al., 2008, 2006, 2009, 2012; Deb & Braganza, 1999; Dekker, Coppus, et al., 2015; Hoekman & Maaskant, 2002; Kirk et al., 2006; Koran et al., 2014; Lott et al., 2012; Mccarron et al., 2014; Prasher, 1997; Rösner et al., 2021; Shultz et al., 2004; Silverman et al., 2004; Startin et al., 2016; Walker et al., 2015; Zigman et al., 2004). Moreover, many studies reported the usage of the Dementia Scale for Down Syndrome (DSDS; in Dutch adapted as DSVH) as instrument to aid diagnosing dementia in people with ID (among others:

Burt et al., 2005; Deb & Braganza, 1999; Devenny et al., 2000; Huxley et al., 2000; Krinsky-McHale et al., 2002; Shultz et al., 2004; Temple et al., 2001). Additionally, various studies have applied the SRZ as part of their dementia screening procedure (Blok et al., 2017; Coppus, 2017; Coppus et al., 2008, 2006, 2009, 2012; De Knegt et al., 2013, 2016; Dekker, Coppus, et al., 2015). Other internationally recommended and widely used dementia screening that – in 2021 – were not (yet) translated/validated/available in Dutch were not examined.

To evaluate whether items in those instruments may be applicable for people with SPI(M)D, it is of essence that people with SPI(M)D are able to display these items at baseline, i.e., the highest level of functioning before decline/dementia occurs. After all, to aid the diagnosis of dementia, identification of change (decline) is essential. The selected four dementia screening instruments in our study were, therefore, completed by conducting interviews with informants of people with SPI(M)D without dementia. For each specific item informants were asked whether that item was applicable for the individual with SPI(M)D. If an item was considered to be not applicable, informants should provide one or more reasons why that item was not applicable.

Dementia screening instruments

DSVH (DSDS)

The DSVH is an adapted, Dutch version of the DSDS, developed in Canada by Gedye, (1995) to aid diagnosing dementia in people with ID. Information about behavioral changes in relation to person's cognitive and activities of daily living (ADL) skills are gathered by interviewing informants. The questions of the original DSDS were translated and studied in 121 persons with ID in The Netherlands (Maaskant & Hoekman, 2011). Similarly, to the DSDS, the DSVH contains a total of 60 items, however the order of items is different. The 60 items are divided into three categories indicating the stage of dementia. Each item is scored as either '*present,*' '*absent,*' '*characteristic,*' or '*not applicable.*' Characteristic indicates that behavior has been present throughout the adult life, whereas present refers to newly developed behavior.

BPSD-DS II

The BPSD-DS II is a recently developed evaluation scale to identify behavioral and psychological symptoms of dementia in people with DS (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a). After initial development, the scale was first studied in 281 people with DS (Dekker, Sacco, et al., 2018). Based on results obtained in this study and clinical experiences, the scale was optimized. The optimized scale was subsequently studied in 524 individuals with DS (Dekker, Ulgiati et al., 2021a). The BPSD-DS II consists of 52 items divided into 11 sections, namely anxious, irritable, obstinate, restless & stereotypic, aggressive, apathetic behavior, depressive, psychotic, disinhibited, eating & drinking behavior and sleeping problems. For every item in the scale, frequency (five-point scale) and severity (four-point scale) are scored for two periods of time, i.e., last 6 months and typical/ characteristic behavior before deterioration occurred, subsequently resulting in a frequency change or severity change score.

DVZ (DMR/DLD)

The DVZ is originally developed in The Netherlands to screen for signs of dementia over time in people with ID (Evenhuis, 1992). Internationally, this dementia screening instrument is known as the DMR and was later renamed as DLD. It encompasses a total of 50 items divided into cognitive skills (i.e., short-term memory, long-term memory and spatial and temporal orientation) and social skills (i.e., speech, practical skills, mood, activity and interest, and behavioral disturbance). Items can be scored as either '*normally yes*,' '*sometimes,*' or '*normally no.*'

SRZ

The SRZ is designed to screen for a decline in social competences over time (Kraijer et al., 2004). It consists of 31 items, which covers aspects regarding ADL skills, effective use of language, social skills and the ability to define and execute tasks. Each item has four answer options, ranging from less to more able to deal with themselves, other people, and everyday situations.

Ethics and consent

The Medical Ethical Committee of the UMCG decided that the Dutch Medical Research Human Subjects Act did not apply to this study (METc 2019/198). The study was registered in the UMCG Research Register (no. 201,900,193) and conducted in accordance with the UMCG Research Code and the EU General Data Protection Regulation. Legal representatives of

people with SPI(M)D provided written informed consent for evaluation of item applicability in the DSVH, BPSD-DS II, DVZ, and SRZ and processing/analyzing coded data for this study.

Participants

Eligible participants were identified within the four participating care organizations based on the following inclusion and exclusion criteria; inclusion criteria: presence of severe/profound ID established according to (medical) records and clinical judgment, aged 25 years to 50 years, stable functioning, thus no changes relative to a person's typical/characteristic functioning, exclusion criteria: mild or moderate ID, (suspected) dementia, functional decline (according to the judgment of involved ID psychologist), long-term admission to hospital in the past 6 months, bedridden or in terminal care, absence of at least one informant able to describe the person's typical/characteristic functioning. Recent life events, e.g., moving home or death of a family member, having long-term impact on the person's functioning (according to clinical judgment) also led to exclusion of an individual. People were eligible to participate regardless of the presence of DS or other disabilities such as visual or motor impairments. Given that people with DS have an extremely high genetic risk of developing dementia due to Alzheimer's disease (Ballard et al., 2016) it was made sure that at least 25% of the participants had a phenotypical diagnosis of DS. After selection, information letters with informed consent forms were sent to legal representatives of eligible participants.

Data collection

A data collection form was constructed in REDCap (Harris et al., 2009), hosted within the secured network of the UMCG. Firstly, demographic data were gathered about age, sex, living situation, attending day care, presence of a syndrome, formal diagnosis of autism spectrum disorder, IQ, social-emotional functioning, verbal communication skills, gross and fine motor function. Gross motor function was according to the judgment of involved ID psychologist categorized into one of the five levels of the Gross Motor Function Classification System (GMFCS): Level I, can walk without limitations; Level II, walk with limitations; Level III, walk with assistive mobility device; Level IV, walking ability severely limited even with assistive devices, use of power wheelchair; Level V, transported by manual wheelchair (Palisano et al., 1997). Similarly, fine motor function was categorized according to the Manual Ability Classification System (MACS) levels: Level

I, handles objects easily and successfully; Level II, handles most objects but with somewhat reduced quality and/or speed of achievement; Level III, handles objects with difficulty, needs help to prepare and/or modify activities; Level IV, handles a limited selection of easily managed objects in adapted situations; V, does not handle objects and has severely limited ability to perform even simple actions (Eliasson et al., 2006). Secondly, data were gathered about the presence of (un) treated comorbidities associated with dementia like symptoms for which the list (part A) in the BPSD-DS II was used (Dekker, Ulgiati et al., 2021a).

Next, the DSVH, BPSD-DS II, DVZ, and SRZ were administered in this sequence. The order of items within these instruments was maintained. The sum of all items of the four instruments was 193 items. For the BPSD-DS II, only the frequency of typical/characteristic behavior was considered, given that in this study the individuals with SPI(M)D had no dementia, i.e., no deterioration in behavior was expected. Regardless of the instrument, for every individual item, the answer option '*not applicable*' was added, if that was not already a possible answer. Not applicable was defined as follows: an individual could impossibly demonstrate the skill/behavior represented in the item, meaning that the skill/behavior cannot occur. Informants were subsequently asked why they answered '*not applicable.*' They could select one or multiple predefined reasons or provide an alternative reason (open answer). Predefined reasons – different depending on the item – based on characteristics of the SPI(M)D group (Nakken & Vlaskamp, 2007) were limited intellectual functioning, limited verbal communication, limited motor functioning, hearing problems, vision problems, ADL dependency complemented with the options limited social-emotional functioning, wheelchair dependent, restrictive measures, and incontinence.

Interviewers

The four instruments were completed by conducting online interviews with informants in Microsoft Teams (due to COVID-19 measures) according to a procedural protocol drawn up in advance. Each interview was performed by an experienced interviewer, such as an ID psychologist (behavioral therapist who studied psychology or special needs education (in Dutch: orthopedagogiek)) or psychological assistant working at the care organizations part of the study consortium. For reasons of uniformity, all interviewers received instructions about the procedure and digital system and were able to practice with system in advance. Interviewers adhered

to the procedural protocol and sequence of items. In total, seven ID psychologists and five psychological assistants alternately conducted the interviews. To improve understanding of items, interviewers shared their screen so that informants could also read items and item explanations. Moreover, a researcher (M.B.G.W.), unacquainted with the individuals with SPI(M)D, was present at each interview to explain the procedure, provide technical assistance, made sure that answers were provided by informants, and keep track of the provided answers (parallel completion of the data collection form) to check afterward for compliance with instructions and protocol. The interviewer and the informant(s) could not see which answer option this researcher selected. Overall, the interviews lasted 60 to 195 minutes.

Informants

Interviews were conducted with at least one key informant of the person with SPI(M)D, such as caregivers working in day-care center/residential facilities or family members. Beforehand, interviewers checked whether informant(s) were able to provide an accurate description of the typical/characteristic functioning. In the case of multiple informants, they were interviewed in a single session. Prior to the interview, informants received information about the procedure by e-mail. Interviews were conducted in absence of the person with SPI(M)D to facilitate honest answering. In line with the procedural protocol, each interview started with welcoming informants, the researcher (M.B.G.W.) introduced the topic, checked if an informed consent form was signed, and explained the procedure and confidentiality. Subsequently, the interviewer ran through the demographic information which was on forehand filled out by the interviewer based on information in clinical records of the individual. Thereafter, in total, 193 questions about item applicability of DSVH, BPSD- DS II, DVZ, and SRZ were asked. Prior to each instrument the scoring system of the instrument was explained to the informants. If necessary, interviewers provided clarification of items and reminded informants to give short and succinct answers. Furthermore, if there was disagreement between informants, the interviewer made sure that consensus was reached during the interview.

Data analysis

Firstly, each completed interview was checked for inclusion/exclusion criteria and compared with the data collection form filled out by the researcher (M.B.G.W.). Provided answers were corrected according to the

protocol if 1) not applicable was unjustifiably scored, the individual was able to show the skill/behavior, or 2) an item was unjustifiably considered to be applicable, the individual could impossibly demonstrate the skill/behavior. The data were analyzed using SPSS Statistics version 25 (IBM Corp). Standard descriptive statistics were used to present results. For each item, the percentage of '*not applicable'* responses were calculated. If one or more times an item was considered to be not applicable, the percentage of a provided '*not applicable*' reason was calculated with respect to the '*not applicable*' score.

To structure the broad range of items, all 193 items were divided according to five domains in line with dementia diagnostic criteria (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022) and literature (Dekker, Ulgiati et al., 2021a; Ries, 2018; Strydom et al., 2010) covering the following: cognitive functioning, ADL, behavioral and psychological functioning, motor functioning, and medical comorbidities. To further improve interpretation, items within each domain were further categorized. Cognitive categories consisted of cognitive functions affected by Alzheimer's disease (Alzheimer's Association, 2022): memory, orientation in time, orientation in place, understanding visual images/spatial relationships, language skills, losing objects, and person recognition complemented with a category other cognitive functions. ADL comprised items of the Barthel Index (Mahoney & Barthel, 1965): feeding, dressing, grooming/bathing, transfers, toilet use and two instrumental ADL: housework and shopping. Behavioral and psychological categories were defined in accordance with the BPSD-DS II scale (Dekker, Ulgiati et al., 2021a): anxious behavior, sleeping problems, irritable behavior, obstinate behavior, restless/stereotypical behavior, aggressive behavior, apathetic behavior, depressive behavior, psychotic behavior, disinhibited behavior, and eating/drinking behavior. The motor domain contained motor skills: walking, balance/fall frequency, movement speed/quality, and fine motor skills (Ries, 2018). The last domain focused on medical comorbidities (Strydom et al., 2010), namely epilepsy, and incontinence complemented with a category other medical comorbidities. Within each category, the calculated percentages of '*not applicable*' responses were ordered from lowest to highest and subsequently divided into four quartiles, namely 0–25% meaning applicable, 26– 50% meaning somewhat applicable, 51–75% meaning hardly applicable and 76–100% meaning not at all applicable.

Lastly, additional analyses were performed for items focusing on verbal communication and gross motor function. In the focus group study of Dekker, Wissing, et al. (2021a) participants already indicated that symptoms like decline in speech and ability to walk cannot be recognized in people who are non-verbal/entirely dependent on a wheelchair. Moreover, results of the study of (Wissing, Fokkens, et al., 2022) indeed showed that the observation of particular symptoms depended on whether individuals had verbal communication or walking skills at baseline. Therefore, for each verbal item, the percentage of '*not applicable*' responses were calculated separately for people with and without verbal communication skills. Similarly, for gross motor items, the percentages of '*not applicable*' responses were calculated for people with (i.e., GMFCS level I, II, and III) and without (i.e., GMFCS level IV, V) independent walking skills. These percentages were also ordered and subsequently divided into four quartiles.

Results

Legal representatives of 99 identified eligible participants received an information letter with informed consent form. Legal representatives of 46 people with SPI(M)D provided written informed consent, 9 did not provide consent, and 44 did not respond. Before planning the interviews, legal representatives of two individuals withdrew their consent without providing a reason. Moreover, four persons were after checking (medical) records and clinical judgment excluded because they had a moderate ID (n=3) or unstable functioning (n=1).

Table 6.1A presents demographic data of the 40 participants. None of these participants had (suspected) dementia, and their functioning was stable, i.e., major life events as well as (un)treated comorbidities did not – according to clinical judgment – result in evident changes of the person's functioning. For none of them an IQ score was determined and reported in their (medical) records. In more than half of the study population the ID was of genetic origin: 11 individuals had DS, and another 11 had other genetic syndromes, namely Rett syndrome (n=2), Fragile X syndrome (n=1), Angelman syndrome (n=1), Cri du chat syndrome (n=1), Kleefstra syndrome (n=1), Edwards syndrome (n=1), Turner syndrome (n = 1), Wolf-Hirschhorn syndrome (n=1), abnormal X chromosome: 46, Y, dup (X) (p22.31 p22.33) (n = 2).

The 40 interviews were conducted with key informants: in 47.5% of cases one informant was interviewed, in 35.0% two informants, and in 17.5% three informants. Key informants were caregivers (54.4%), family members (44.1%), or legal representatives without being a family member (1.5%). Table 6.1B shows the informants' characteristics.

Table 6.1 Participants' and informants' characteristics.

A: Participants' characteristics	**N=40**
Age [years, mean ± SD (min. – max.)]	38.4 ± 5.2 (26.7–46.7)
Sex (% female)	35.0
Living situation: care organization, with family, combination, other (%)	82.5, 2.5, 12.5, 2.5
Attending day-care (%)	100
Intellectual functioning (baseline): severe, profound (%)	60.0, 40.0
Presence of syndrome: DS, other genetic syndrome, no/unknown (%)	27.5, 27.5, 45.0
Formal diagnosis of autism spectrum disorder (%)	20.0
Social-emotional functioning: 0-6 months, 6-18 months, 18-36 months, unknown (%)	12.5, 45.0, 15.0, 27.5
Verbal communication: able, never (%)	35.0, 65.0
Estimated GFMCS: level I, level II, level III, level IV, level V (%)	27.5, 40.0, 15.0, 7.5, 10.0
Estimated MACS: level I, level II, level III, level IV, level V (%)	25.0, 45.0, 7.5, 15.0, 7.5
Vision problems: treated, untreated (%)	20.0, 47.5
Hearing problems: treated, untreated (%)	5.0, 20.0
Depression: treated, untreated (%)	2.5, 0
Epilepsy: treated, untreated (%)	45.0, 5.0
Hypothyroidism: treated, untreated (%)	12.5, 7.5
Vitamin B12 deficiency: treated, untreated (%)	5.0, 0
Sleep apnea: treated, untreated (%)	0, 5.0
Chronic pain: treated, untreated (%)	10.0, 2.5
Swallowing problems (%)	27.5
Dental problems causing eating/drinking problems (%)	7.5
B: Informants' characteristics	**N=68 informants**
Informants per participant (% n = 1, n = 2, n = 3)	47.5, 35.0, 17.5
Sex (% female)	86.8
Role: caregiver, family, no family member but legal representative (%)	54.4, 44.1, 1.5
Years knowing participant (% < 2, 2-10, 10-20, > 20 years)	2.9, 36.8, 14.7, 45.6
Hours per week with participant (% < 10, 10-20, > 20 hours)	29.4, 27.9, 42.6

ID refers to the highest level of functioning (baseline). Gross Motor Function Classification System (GMFCS) levels: Level I, can walk without limitations; Level II, walk with limitations; Level III, walk with assistive mobility device; Level IV, walking ability severely limited even with assistive devices, use of power wheelchair; Level V, transported by manual wheelchair. Manual Ability Classification System (MACS) levels: Level I, handles objects easily and successfully; Level II, handles most objects but with somewhat reduced quality and/or speed of achievement; Level III, handles objects with difficulty, needs help to prepare and/or modify activities; Level IV, handles a limited selection of easily managed objects in adapted situations; V, does not handle objects and has severely limited ability to perform even simple actions. Abbreviations: DS, Down syndrome; ID, intellectual disabilities; max., maximum; min., minimum; SD, standard deviation.

Applicability of items

The 193 items (sum of all items of the four instruments) were completed for all 40 participants. During the data check, 117 of the total 7720 provided answers (1.5%) were corrected in accordance with the protocol. Of these 117 items, 63 were unjustifiably scored as "not applicable," whereas 54 were unjustifiably considered to be applicable. Tables 6.2–6.6 display the calculated percentages of '*not applicable*' responses for cognitive, ADL, behavioral and psychological, motor and medical comorbidities items, respectively.

Cognitive items

In total, 70 items about cognitive functioning were identified within the four existing dementia screening instruments. As shown in Table 6.2, the percentages of '*not applicable*' responses of 25 items fell inside the first quartile (0–25%), meaning that these items were considered to be applicable. Applicable items were identified within different cognitive functioning categories, namely memory (7 items), orientation in place (5), person recognition (3), orientation in time (2), responsiveness (2), understanding visual images/spatial relationships (1), losing objects (1) and other cognitive functions (4), i.e., knowing what to do with objects, attention for the task, expressing wishes and using objects correctly. Only for the category language skills there were no items that fell inside the first quartile. Moreover, 11 items, such as knowing your age/the year, fell inside the fourth quartile (76–100%) and were thus not at all applicable. Not only for these 11 items but also for the other cognitive items, the two most provided reasons why items were not applicable were limited intellectual and verbal functioning.

ADL items

The applicability of the 25 identified ADL items is presented in Table 6.3. In total, five items were found to be applicable given that these items fell inside the first quartile (0–25%). Most of the applicable ADL items focused on feeding (3 items): use of cutlery, everyday support, or extensive assistance with eating. The remaining applicable items were items regarding making transfers (1) and doing housework (1). For the categories dressing, grooming/ bathing, toilet use, and shopping, none of the identified items fell inside the first quartile. Limited intellectual functioning and ADL dependency were within the ADL domain the two most provided reasons why items were not applicable.

Behavioral and psychological items

Table 6.4 presents the applicability of the 81 items categorized within the behavioral and psychological domain. What stands out is that almost three-fourths of the items fell inside the first quartile (0–25%). Accordingly, applicable items were found within 10 of the total 11 behavioral and psychological categories. The apathetic behavior category comprised the most applicable items (13 items). Moreover, applicable items were found for depressive behavior (9), sleeping problems (7), obstinate behavior (6), anxious behavior (5), irritable behavior (5), restless/stereotypic behavior (5), eating/drinking behavior (5), aggressive behavior (4) and disinhibited behavior (1). For psychotic behavior, items fell either in the third (51–75%) or fourth quartile (76–100%), and thus no applicable items were identified within this category. For the behavioral and psychological domain, a variety of reasons why items were not applicable – depending on the item – were provided.

Motor items

As shown in Table 6.5, the percentages of '*not applicable*' responses of all six identified motor items fell inside the first quartile (0–25%). The balance/fall frequency and movement speed/quality category each consisted of two applicable items, namely loss of balance, sitting down, and slowness of movements, slow/clumsy movements, respectively. Moreover, the other two items were motor items regarding walking and fine motor skills. For the few individuals for whom a motor item was not applicable, the main reasons provided were limited motor functioning and wheelchair dependent.

Medical comorbidities items

Eleven items about medical comorbidities were identified in the dementia screening instruments. In Table 6.6, it is apparent that all three items about epilepsy as well as the two items in the category other medical comorbidities fell inside the first quartile (0–25%) and were thus considered to be applicable. In contrast, six items about incontinence were hardly or not at all applicable, primarily because of pre-existing incontinence.

Table 6.2 Applicability of items about cognitive functioning for people with SPI(M)D.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)									
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*ASS*
Memory	DVZ 25	Understanding simple instructions	7.5	7.5	-	2.5	-	-	-	2.5	-	-	-
	DSVH 7	Understanding verbal instructions	10.0	7.5	2.5	2.5	-	-	5.0	2.5	-	-	-
	DVZ 1	Understanding what someone makes clear	10.0	10.0	7.5	2.5	2.5	-	-	-	-	-	-
	DSVH 1	Forgetting daily routines	12.5	10.0	2.5	2.5	7.5	5.0	-	2.5	12.5	-	-
	DSVH 20	Remembering actions	12.5	12.5	-	2.5	-	7.5	-	2.5	-	-	-
	DVZ 5	Remembering family/friends	25.0	25.0	17.5	7.5	-	-	-	-	-	-	-
	DVZ 14	Remembering instructions	25.0	25.0	10.0	7.5	-	-	2.5	-	-	-	-
	DVZ 2	Remembering where you put away something	27.5	25.0	7.5	2.5	10.0	10.0	-	-	-	-	-
	DVZ 33	Remembering something that has been told recently	40.0	40.0	25.0	7.5	-	-	2.5	-	-	-	-
	DSVH 25	Remembering events	52.5	47.5	30.0	12.5	-	-	2.5	-	-	-	-
	DVZ 3	Remembering impressive event	52.5	45.0	40.0	10.0	-	-	5.0	2.5	-	-	-
	DSVH 4	Forgetting names	55.0	42.5	50.0	2.5	-	-	5.0	2.5	-	-	-
	DVZ 10	Remembering events from your youth	62.5	57.5	42.5	10.0	-	-	2.5	2.5	-	-	-
	DSVH 41	Using the wrong name	65.0	52.5	65.0	7.5	-	-	5.0	-	-	-	-
	DVZ 29	Speaking about what you did	70.0	52.5	62.5	7.5	2.5	-	5.0	-	-	-	-
	DVZ 45	Remembering your hometown	70.0	70.0	45.0	7.5	-	-	2.5	5.0	-	-	-
	DVZ 18	Speaking about holiday/trip	72.5	60.0	60.0	27.5	5.0	-	5.0	5.0	-	-	-
	DSVH 26	Speaking about events as if they happened recently	75.0	62.5	65.0	12.5	5.0	-	5.0	2.5	-	-	-
	DVZ 43	Knowing name of home	85.0	85.0	45.0	10.0	-	-	5.0	2.5	-	-	-
	DVZ 47	Knowing name of queen	90.0	90.0	42.5	7.5	-	-	2.5	-	-	-	-
	DVZ 40	Knowing profession of father/ mother	95.0	95.0	40.0	12.5	-	-	5.0	2.5	-	-	-
	DVZ 42	Knowing your age	100	100	47.5	12.5	-	-	2.5	-	-	-	-
Orientation in time	DSVH 45	Day/night confusion	7.5	7.5	5.0	2.5	-	-	-	2.5	-	-	-
	DSVH 5	Changed time perception	15.0	15.0	5.0	2.5	-	-	-	-	-	12.5	-

Table 6.2 Continued.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)									
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*ASS*
Orientation in time	DSVH 6	Confusion about time	30.0	30.0	7.5	7.5	-	-	-	2.5	-	-	-
	DVZ 28	Knowing weekend/weekdays	65.0	65.0	35.0	7.5	-	-	5.0	2.5	-	-	-
	DVZ 6	Knowing the seasons	92.5	92.5	37.5	7.5	-	-	2.5	2.5	-	-	-
	DVZ 35	Knowing the day	97.5	97.5	40.0	10.0	-	-	5.0	5.0	-	-	-
	DVZ 4	Knowing the month	100	100	35.0	10.0	-	-	2.5	2.5	-	-	-
	DVZ 7	Knowing the year	100	100	37.5	7.5	-	-	2.5	2.5	-	-	-
Orientation in place	DSVH 47	Loss of environmental awareness	0	-	-	-	-	-	-	-	-	-	-
	DSVH 8	Decreased orientation in place	5.0	5.0	-	-	2.5	2.5	-	-	2.5	-	-
	DVZ 46	Finding your way about home	12.5	7.5	5.0	2.5	12.5	5.0	-	2.5	10.0	-	-
	DSVH 46	Disoriented in familiar places	17.5	12.5	7.5	2.5	12.5	10.0	-	2.5	-	-	-
	DVZ 15	Finding way to familiar places	25.0	25.0	5.0	2.5	12.5	15.0	-	2.5	-	2.5	-
	SRZ 23	Freedom to move outdoors	45.0	27.5	10.0	20.0	15.0	12.5	-	2.5	17.5	17.5	
Understanding visual images/ spatial relationships	DSVH 36	Disturbed depth perception	10.0	2.5	2.5	-	-	2.5	-	7.5	-	-	-
Language skills	DSVH 29	Less speaking/gestures	40.0	37.5	37.5	5.0	5.0	-	2.5	-	-	-	-
	SRZ 30	Answering questions	40.0	32.5	40.0	5.0	5.0	-	5.0	2.5	-	-	-
	DSVH 9	Reduced frequency/amount of speech	55.0	42.5	55.0		7.5	-	5.0	-	-	-	-
	DVZ 22	Speaking	60.0	40.0	60.0	7.5	2.5	-	5.0	-	-	-	-
	SRZ 24	Use of language	65.0	47.5	65.0	2.5	5.0	-	5.0	-	-	-	-
	SRZ 25	Pronunciation of language	65.0	47.5	65.0	2.5	7.5	-	2.5	-	-	-	-
	SRZ 26	Comprehensibility of language	65.0	47.5	62.5	2.5	7.5	-	5.0	-	-	-	-
	DSVH 30	Speaking more slowly/less intelligible	67.5	55.0	65.0	5.0	5.0	-	2.5	-	-	-	-
	DSVH 31	Mumbling	67.5	55.0	67.5	5.0	7.5	-	5.0	-	-	-	-
	DSVH 56	Loss of speech	67.5	55.0	67.5	7.5	2.5	-	5.0	-	-	-	-
	SRZ 27	Saying your name	67.5	50.0	67.5	2.5	5.0	-	5.0	-	-	-	-
	DSVH 3	Word-finding difficulties	70.0	50.0	70.0	5.0	-	-	7.5	5.0	-	-	-
	DVZ 13	Speaking intelligibly/ comprehensibly	72.5	50.0	70.0	5.0	2.5	-	-	-	-	-	-
	SRZ 28	Repeating words	75.0	55.0	72.5	2.5	7.5	-	7.5	-	-	-	-

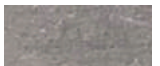


Table 6.2 Continued.

Category	Item	Short item description	Not applicable (%)	Reasons why items were not applicable (%) (multiple reasons pp. possible)									
				Limited intellectual functioning	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*ASS*
Losing objects	DSVH 2	Losing objects	20.0	17.5	5.0	2.5	12.5	7.5	2.5	5.0	-	-	-
Person recognition	DSVH 42	Recognizing family/friends	2.5	2.5	-	-	-	-	-		-	-	-
	DVZ 26	Recognizing staff members	5.0	5.0	2.5	-	-	-	-	2.5	-	-	-
	DVZ 23	Recognizing persons	22.5	22.5	7.5	10.0	-	-	-	2.5	-	-	-
Responsiveness	DSVH 16	Being less alert/attentive	0	-	-	-	-	-	-	-	-	-	-
	DSVH 48	Reduced reactions on persons/ stimuli	0	-	-	-	-	-	-	-	-	-	-
Other cognitive functions	DSVH 24	Knowing what to do with objects	2.5	2.5	-	-	2.5	-	-	2.5	2.5	-	-
	SRZ 10	Attention for the task	12.5	12.5	-	5.0	-	-	-	-	-	-	-
	SRZ 29	Expressing wishes	15.0	15.0	7.5	5.0	2.5	-	-	-	-	-	-
	DVZ 49	Using objects correctly	17.5	12.5	-	5.0	15.0	7.5	-	2.5	12.5	-	-
	SRZ 9	Finishing tasks	27.5	25.0	-	10.0	17.5	10.0	-	2.5	17.5	-	-
	SRZ 20	Interacting with others	27.5	20.0	2.5	25.0	10.0	2.5	-	-	-	-	2.5
	DSVH 23	Recognizing difference between breakfast/dinner	30.0	30.0	5.0	2.5	5.0	-	-	2.5	5.0	-	-
	SRZ 31	Naming and establishing links	67.5	57.5	65.0	12.5	-	-	5.0	5.0	-	-	-
	DVZ 24	Being on familiar terms with persons	67.5	45.0	27.5	60.0	10.0	12.5	5.0	5.0	-	2.5	5.0
	SRZ 21	Offering help	77.5	65.0	25.0	55.0	17.5	10.0	5.0	5.0	-	-	-
	DSVH 21	Loss of writing/reading/drawing skills	82.5	82.5	-	5.0	35.0	-	2.5	10.0	-	-	-
	SRZ 19	Borrowing	82.5	75.0	22.5	57.5	15.0	10.0	5.0	5.0	-	2.5	-
	SRZ 18	Sharing	85.0	70.0	22.5	67.5	12.5	10.0	5.0	5.0	-	-	5.0

Within each category, items were ordered from most applicable (i.e., lowest proportion of participants for whom informants answered '*not applicable*') to least applicable (i.e., highest proportion of participants for whom informants answered '*not applicable*'). The percentages of 'not applicable' responses within each category were divided into four quartiles, namely 0 – 25 %, 26 – 50 %, 51 – 75% and 76 – 100%. 0 – 25 % are white meaning applicable, 26 – 50% are light gray meaning somewhat applicable, 51 – 75% are middle gray meaning hardly applicable and 76 – 100% are dark gray meaning not at all applicable. With regard to the not applicable reasons, informants could provide multiple reasons why an item was 'not applicable'. Percentages were calculated based on the total number of participants. Abbreviations: ADL, activities of daily living; ASS, formal diagnosis of autism spectrum disorder; DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; DVZ, Dutch Dementia Questionnaire for persons with Mental Retardation; pp., per person; SRZ, Social competence Rating scale for people with Intellectual Disabilities.

Table 6.3 Applicability of items about ADL for people with SPI(M)D.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)											
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*Incontinence*	*ASS*	*PEG*
Feeding	DSVH 57	Everyday support with eating	5.0	2.5	2.5	-	2.5	-	-	2.5	5.0	-	-	-	2.5
	SRZ 12	Use of cutlery	12.5	10.0	-	-	12.5	5.0	-	2.5	10.0	-	-	-	-
	DSVH 44	Extensive assistance with eating	15.0	7.5	-	-	12.5	2.5	2.5	2.5	12.5	-	-	-	2.5
	SRZ 15	Clearing table	27.5	20.0	-	2.5	20.0	10.0	-	2.5	17.5	-	-	-	-
	SRZ 14	Setting table	27.5	22.5	-	2.5	17.5	12.5	-	2.5	15.0	-	-	-	-
	SRZ 13	Using knife	60.0	40.0	-	2.5	37.5	5.0	-	2.5	47.5	2.5	-	-	-
Dressing	DSVH 19	Ability to (un)dress	30.0	25.0	-	2.5	20.0	12.5	-	2.5	27.5	-	-		-
	DVZ 11	Ability to undress	30.0	17.5	-	2.5	22.5	12.5	2.5	2.5	25.0	-	-	-	-
	DVZ 19	Ability to dress	35.0	22.5	-	5.0	25.0	15.0	2.5	5.0	30.0	-	-	-	-
	SRZ 1	Getting dressed	40.0	30.0	-	5.0	27.5	15.0	-	2.5	30.0	-	-	-	-
	SRZ 4	Getting undressed	40.0	30.0	-	2.5	30.0	7.5	-	2.5	30.0	-	-	-	-
	DSVH 43	Extensive assistance with (un)dressing	35.0	30.0	-	2.5	17.5	10.0	2.5	2.5	30.0	-	-	-	-
	SRZ 17	Hang clothes	45.0	27.5	-	2.5	35.5	12.5		5.0	25.0	2.5	-	-	-
	SRZ 2	Shoe tying	87.5	65.0	-	10.0	55.0	12.5	-	5.0	52.5	-	-	-	-
Grooming/ bathing	SRZ 5	Washing face/hands	52.5	40.0	-	5.0	27.5	7.5	-	2.5	45.0	2.5	-	-	-
	SRZ 6	Brushing teeth/ cleaning denture	57.5	42.5	-	5.0	35.0	7.5	-	2.5	45.0	-	-	-	-
	DVZ 17	Ability to wash	77.5	65.0	-	12.5	35.0	15.0	2.5	2.5	62.5	-	-	-	-
Transfers	DVZ 36	Ability to get in/out bed	25.0	10.0	-	-	17.5	12.5	-	-	15.0	10.0	-	-	-

Table 6.3 Continued.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)											
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*Incontinence*	*ASS*	*PEG*
Toilet use	DVZ 50	Ability to perform acts necessary in toilet	67.5	50.0	10.0	17.5	22.5	12.5	2.5	-	37.5	5.0	30.0	-	-
	SRZ 7	Use of toilet paper	87.5	60.0	-	2.5	45.0	5.0	-	2.5	67.5	7.5	22.5	-	-
Housework	SRZ 16	Tidying stuff	25.0	22.5	-	2.5	20.0	10.0	-	2.5	10.0	-	-	-	-
	SRZ 8	Cleaning up	42.5	35.0	-	12.5	15.0	12.5	-	2.5	30.0	2.5	-	-	-
	DVZ 16	Keeping clothes/ things tidy	50.0	47.5	-	25.0	15.0	10.0	-	5.0	22.5	2.5	-	-	-
	SRZ 11	Bed making	65.0	55.0	-	7.5	35.0	12.5	-	2.5	50.0	-	-	-	-
Shopping	SRZ 22	Doing errands	87.5	85.0	37.5	42.5	30.0	12.5	5.0	5.0	-	5.0	-	2.5	-

Within each category, items were ordered from most applicable (i.e., lowest proportion of participants for whom informants answered *'not applicable'*) to least applicable (i.e., highest proportion of participants for whom informants answered '*not applicable*'). The percentages of 'not applicable' responses within each category were divided into four quartiles, namely 0 – 25 %, 26 – 50 %, 51 – 75% and 76 – 100. 0 – 25 % are white meaning applicable, 26 – 50% are light gray meaning somewhat applicable, 51 – 75% are middle gray meaning hardly applicable and 76 – 100% are dark gray meaning not at all applicable. With regard to the not applicable reasons, informants could provide multiple reasons why an item was 'not applicable'. Percentages were calculated based on the total number of participants. Abbreviations: ADL, activities of daily living; ASS, formal diagnosis of autism spectrum disorder; DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; DVZ, Dutch Dementia Questionnaire for persons with Mental Retardation; PEG, percutaneous endoscopic gastrostomy; pp., per person; SRZ, Social competence Rating scale for people with Intellectual Disabilities.

Table 6.4 Applicability of items about behavioral and psychological functioning for people with SPI(M)D.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)											
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*Incontinence*	*ASS*	*PEG*
Anxious behavior	BPSD-DS II 1.3	Being tense	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 1.6	Being easily panicked	0	-	-	-	-	-	-	-	-	-	-	-	-
	DSVH 33	Being more anxious	2.5	2.5	-	2.5	-	-	-	-	-	-	-	-	-
	DSVH 22	Reassurance seeking	5.0	5.0	2.5	5.0	2.5	2.5	-	-	-	-	-	-	-
	BPSD-DS II 1.5	Being scared to be left alone	5.0	5.0	2.5	5.0	2.5	2.5	-	-	-	-	-	-	-
	BPSD-DS II 1.4	Avoiding situations/ places	40.0	37.5	17.5	15.0	12.5	12.5	2.5	2.5	-	-	-	-	-
	BPSD-DS II 1.1	Worrying about upcoming activities/ events	50.0	50.0	22.5	20.0	-	-	2.5	2.5	-	-	-	-	-
	BPSD-DS II 1.2	Going to toilet too often/long without reason	52.5	30.0	5.0	5.0	20.0	10.0	-	2.5	30.0	5.0	42.5	-	-
Sleeping problems	BPSD-DS II 2.1	Difficulty falling asleep	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 2.2	Waking repeatedly	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 2.4	Waking long before it is time to get up	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 2.6	Being tired/ complaining of fatigue	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 2.7	Daytime sleeping	0	-	-	-	-	-	-	-	-	-	-	-	-
	DVZ 38	Restless/awake at night	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 2.5	Difficulty getting up	2.5	2.5	-	2.5	-	-	-	-	-	-	-	-	-
	BPSD-DS II 2.3	Wandering at night	32.5	7.5	-	-	20.0	15.0	-	2.5		20.0	-	-	-
Irritable behavior	DSVH 11	Being unusual irritable	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 3.1	Being irritable	0	-	-	-	-	-	-	-	-	-	-	-	-
	DVZ 27	Getting angry easily	2.5	2.5	2.5	2.5	-	-	-	-	-	-	-	-	-
	BPSD-DS II 3.2	Being impatient	5.0	5.0	5.0	2.5	-	-	-	2.5	-	-	-	-	-
	BPSD-DS II 3.3	Being short-spoken	10.0	10.0	7.5	5.0	-	-	-	-	-	-	-	-	-
Obstinate behavior	BPSD-DS II 4.4	Sighing/groaning	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 4.3	Not willing to accept necessary help	5.0	2.5	-	-	5.0	2.5	-	-	-	-	-	-	-
	BPSD-DS II 4.1	Being self-willed	7.5	7.5	-	2.5	5.0	2.5	-	-	5.0	-	-	-	-
	DSVH 12	Being uncooperative	10.0	7.5	5.0	10.0	5.0	5.0	-	-	-	-	-	-	-

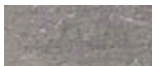


Table 6.4 Continued.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)											
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*Incontinence*	*ASS*	*PEG*
Obstinate behavior	DSVH 53	Uncooperative to walk	15.0	5.0	-	-	15.0	12.5	-	-	-	-	-	-	-
	DSVH 54	Uncooperative to carry one's own weight	15.0	5.0	-	-	15.0	12.5	-	-	-	-	-	-	-
	BPSD-DS II 4.2	Being argumentative/ uncooperative/ obstructive	32.5	32.5	-	15.0	7.5	7.5	2.5	-	-	-	-	-	-
Restless/ stereo-typic behavior	BPSD-DS II 5.3	Stereotypic behavior	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 5.1	General restlessness	5.0	-	-	-	2.5	5.0	-	-	-	2.5	-	-	-
	DSVH 27	Simple repetitive movements	10.0	7.5	-	-	7.5	7.5	2.5	2.5	-	-	-	-	-
	BPSD-DS II 5.6	Compulsive behavior	17.5	15.0	-	7.5	10.0	10.0	-	-	-	-	-	-	-
	BPSD-DS II 5.2	Wandering	25.0	7.5	-	2.5	22.5	20.0	-	2.5	-	5.0	-	-	-
	BPSD-DS II 5.4	Repeatedly (un)dressing	27.5	15.0	-	2.5	20.0	15.0	-	2.5	25.0	-	-	-	-
	BPSD-DS II 5.5	Verbal stereotypy	57.5	40.0	57.5	7.5	2.5	-	5.0	-	-	5.0	-	-	-
Aggres-sive behavior	BPSD-DS II 6.2	Destructive behavior	5.0	2.5	2.5	-	5.0	5.0	-	2.5	-	-	-	-	-
	DSVH 35	Hitting out of frustration	5.0	5.0	-	2.5	2.5	2.5	-	2.5	-	-	-	-	-
	BPSD-DS II 6.3	Physical aggression	7.5	5.0	-	5.0	5.0	5.0	-		-	-	-	-	-
	DVZ 9	Showing aggression	7.5	7.5	-	2.5	7.5	5.0	-	2.5	-	-	-	-	-
	BPSD-DS II 6.1	Verbal aggression	35.0	22.5	32.5	12.5	2.5	-	5.0	-	-	-	-	-	-
	DVZ 31	Threatening by words/gestures	52.5	42.5	30.0	35.0	7.5	10.0	-	5.0	-	-	-	-	-
Apathetic behavior	BPSD-DS II 7.2	Lack of interest in environment	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 7.3	Lack of motivation	2.5	2.5	-	2.5	-	2.5	-	-	2.5	-	-	-	-
	BPSD-DS II 7.8	Jaded emotional responses	2.5	2.5	-	2.5	-	-	-	-	-	-	-	2.5	-
	DSVH 13	Lack of interest in objects/handcraft/ events	5.0	5.0	-	5.0	5.0	-	-	-	-	-	-	-	-
	DSVH 14	Being less occupied	5.0	5.0	-	2.5	2.5	2.5	-	-	-	-	-	-	-

Table 6.4 Continued.

Category	Item	Short item description	Not applicable (%)	Reasons why items were not applicable (%) (multiple reasons pp. possible)										
				Limited intellectual functioning	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*Incontinence*	*ASS*	*PEG*
Apathetic behavior	DVZ 20	Interest in home activities	7.5	7.5	5.0	7.5	-	-	-	-	-	-	-	-	-
	BPSD-DS II 7.1	Lack of initiative	7.5	7.5	-	5.0	7.5	7.5	-	-	-	-	-	-	-
	BPSD-DS II 7.6	Social withdrawal	7.5	7.5	-	7.5	-	-	-	-	-	-	-	2.5	-
	DSVH 34	Undertaking less spontaneous activities	10.0	10.0	-	5.0	5.0	2.5	-	-	-	-	-	-	-
	DSVH 15	Lack of interest in other people's activities	17.5	10.0	2.5	15.0	-	-	2.5	2.5	-	-	-	-	-
	DVZ 8	Interest in indoor activities	17.5	17.5	7.5	12.5	-	-	-	-	-	-	-	-	-
	BPSD-DS II 7.4	Not completing activities/tasks independently	17.5	15.0	-	7.5	10.0	7.5	5.0	5.0	15.0	-	-	-	-
	BPSD-DS II 7.5	Lack of participation in conversation	22.5	22.5	-	15.0	2.5	-	2.5	-	-	-	-	2.5	-
	DVZ 21	Interest in papers/ television	27.5	27.5	12.5	12.5	-	-	2.5	15.0	-	-	-	-	-
	DVZ 30	Keeping yourself busy	35.0	35.0	10.0	27.5	12.5	7.5	2.5	5.0	-	7.5	-	2.5	-
	DVZ 37	Being helpful spontaneously	40.0	37.5	-	27.5	20.0	12.5	-	2.5	15.0	-	-	2.5	-
	SRZ 3	Taking initiative to dress up	52.5	45.0	-	7.5	22.5	5.0	-	2.5	40.0	-	-	-	-
	BPSD-DS II 7.7	Lack of sympathy/ empathy	52.5	47.5	-	47.5	-	-	5.0	5.0	-	-	-	2.5	-
Depressive behavior	DVZ 44	Readily upset	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 8.1	Rapid mood swings	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 8.2	Being sad	0	-	-	-	-	-	-	-	-	-	-	-	-
	BPSD-DS II 8.5	Moving/responding slowly	0	-	-	-	-	-	-	-	-	-	-	-	-

Table 6.4 Continued.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)											
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited verbal communication*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*	*Restrictive measures*	*Incontinence*	*ASS*	*PEG*
Depressive behavior	DSVH 49	Facial masking	0	-	-	-	-	-	-	-	-	-	-	-	-
	DVZ 39	Being gloomy/sad	0	-	-	-	-	-	-	-	-	-	-	-	-
	DSVH 32	Increased sadness	2.5	2.5	2.5	2.5	-	-	-	-	-	-	-	-	-
	DVZ 34	Weeping on slightest provocation	7.5	5.0	-	7.5	-	-	-	-	-	-	-	-	-
	BPSD-DS II 8.4	Physical complaints (no illness)	10.0	10.0	2.5	7.5	-	-	-	-	-	-	-	-	-
	DVZ 48	Utter physical complaints	30.0	30.0	17.5	15.0	-	-	-	-	-	-	-	-	-
	BPSD-DS II 8.3	Being very downhearted	32.5	22.5	-	27.5	2.5	-	5.0	2.5	-	-	-	-	-
Psychotic behavior	BPSD-DS II 9.2	Hallucinations	70.0	60.0	47.5	37.5	-	-	5.0	2.5	-	-	-	-	-
	BPSD-DS II 9.1	Delusions	77.5	70.0	47.5	40.0	-	-	7.5	2.5	-	-	-	-	-
	DVZ 32	Accusing others of harming you	90.0	80.0	57.5	55.0	-	5.0	2.5	-	-	-	-	-	-
Disinhibited behavior	BPSD-DS II 10.1	Behaving impolite/ indecent	15.0	12.5	-	7.5	10.0	7.5	-	-	-	2.5	-	-	-
	BPSD-DS II 10.3	Loss of decorum	57.5	57.5	-	42.5	-	-	2.5	-	-	-	-	-	-
	BPSD-DS II 10.2	Making inappropriate comments	80.0	65.0	67.5	40.0	2.5	-	5.0	-	-	-	-	-	-
Eating/ drinking behavior	BPSD-DS II 11.3	Eating slowly	5.0	-	-	-	2.5	-	-	-	5.0	-	-	-	2.5
	BPSD-DS II 11.4	Being picky about food/drink	5.0	-	-	-	2.5	-	-	2.5	5.0	-	-	-	2.5
	BPSD-DS II 11.5	Pica	5.0	2.5	-	-	5.0	5.0	-	2.5	-	-	-	-	-
	BPSD-DS II 11.1	Drinking poorly	7.5	5.0	-	-	5.0	-	-	-	7.5	-	-	-	2.5
	BPSD-DS II 11.2	Poor appetite	7.5	5.0	-	-	5.0	-	-	-	7.5	-	-	-	2.5

Within each category, items were ordered from most applicable (i.e., lowest proportion of participants for whom informants answered '*not applicable*') to least applicable (i.e., highest proportion of participants for whom informants answered '*not applicable*'). The percentages of 'not applicable' responses within each category were divided into four quartiles, namely 0 – 25 %, 26 – 50 %, 51 – 75% and 76 – 100%. 0 – 25 % are white meaning applicable, 26 – 50% are light gray meaning somewhat applicable, 51 – 75% are middle gray meaning hardly applicable and 76 – 100% are dark gray meaning not at all applicable. With regard to the not applicable reasons, informants could provide multiple reasons why an item was 'not applicable'. Percentages were calculated based on the total number of participants. Abbreviations: ADL, activities of daily living; ASS, formal diagnosis of autism spectrum disorder; BPSD-DS II, Behavioral and Psychological Symptoms of Dementia in Down Syndrome evaluation scale version II; DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; DVZ, Dutch Dementia Questionnaire for persons with Mental Retardation; PEG, percutaneous endoscopic gastrostomy; pp., per person; SRZ, Social competence Rating scale for people with Intellectual Disabilities.

Table 6.5 Applicability of items about motor functioning for people with SPI(M)D.

				Reasons why items were not applicable (%) (multiple reasons pp. possible)						
Category	**Item**	**Short item description**	**Not applicable (%)**	*Limited intellectual functioning*	*Limited social-emotional functioning*	*Limited motor functioning*	*Wheelchair dependent*	*Hearing problems*	*Vision problems*	*ADL dependency*
Walking	DSVH 60	Non-ambulatory	10.0	2.5	-	10.0	7.5	-	-	-
Balance/fall frequency	DSVH 37	Loss of balance	17.5	5.0	-	15.0	17.5	-	2.5	-
	DSVH 10	Sitting down at table	22.5	10.0	2.5	22.5	20.0	2.5	2.5	15.0
Movement speed/quality	DSVH 17	Slowness of movements	0	-	-	-	-	-	-	-
	DSVH 38	Slow/clumsy movements	0	-	-	-	-	-	-	-
Fine motor skills	DSVH 40	Loss of fine motor skills	10.0	7.5	-	10.0	-	-	2.5	-

Within each category, items were ordered from most applicable (i.e., lowest proportion of participants for whom informants answered '*not applicable*') to least applicable (i.e., highest proportion of participants for whom informants answered '*not applicable*'). The percentages of 'not applicable' responses within each category were divided into four quartiles, namely 0 – 25 %, 26 – 50 %, 51 – 75% and 76 – 100%. 0 – 25 % are white meaning applicable, 26 – 50% are light gray meaning somewhat applicable, 51 – 75% are middle gray meaning hardly applicable and 76 – 100% are dark gray meaning not at all applicable. With regard to the not applicable reasons, informants could provide multiple reasons why an item was 'not applicable'. Percentages were calculated based on the total number of participants. Abbreviations: ADL, activities of daily living; DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; pp., per person.

Table 6.5 Applicability of items about medical comorbidities for people with SPI(M)D.

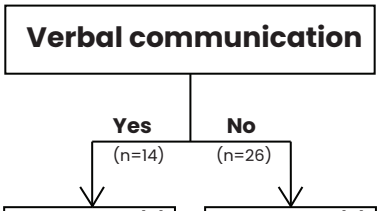
Category	Item	Short item description	Not applicable (%)	Reasons why items were not applicable (%) (multiple reasons pp. possible)	
				Restrictive measures	*Incontinence*
Epilepsy	DSVH 55	Epilepsy	0	-	-
	DSVH 51	Involuntary movements	0	-	-
	DSVH 58	Jerking of limbs	0	-	-
Incontinence	DSVH 39	Fecal incontinence	50.0	-	50.0
	DSVH 59	Urinary/fecal incontinence	52.5	-	52.5
	DSVH 18	Urinary accidents	55.0	-	55.0
	DSVH 52	Urinary incontinence	55.0	-	55.0
	DVZ 12	Incontinence during day	55.0	-	55.0
	DVZ 41	Incontinence during night	65.0	2.5	62.5
Other medical comorbidities	DSVH 50	Droopy eyes	0	-	-
	DSVH 28	Reduced sense of touch	0	-	-

Within each category, items were ordered from most applicable (i.e., lowest proportion of participants for whom informants answered '*not applicable*') to least applicable (i.e., highest proportion of participants for whom informants answered '*not applicable*'). The percentages of 'not applicable' responses within each category were divided into four quartiles, namely 0 – 25 %, 26 – 50 %, 51 – 75% and 76 – 100%. 0 – 25 % are white meaning applicable, 26 – 50% are light gray meaning somewhat applicable, 51 – 75% are middle gray meaning hardly applicable and 76 – 100% are dark gray meaning not at all applicable. With regard to the not applicable reasons, informants could provide multiple reasons why an item was 'not applicable'. Percentages were calculated based on the total number of participants. Abbreviations: DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; DVZ, Dutch Dementia Questionnaire for persons with Mental Retardation; pp., per person.

Verbal communication items

To compare differences in applicability of verbal items for people with and without verbal communication skills at baseline, the percentages of 'not applicable' responses were for verbal items calculated for each subgroup. The study population was divided on the basis of verbal communication skills at baseline: among the 40 participants, 14 had verbal communication skills, whereas 26 had never acquired such skills. Figure 6.1 displays an overview of the percentages of 'not applicable' responses – separately for each subgroup – for the 23 identified verbal items. The initial analysis revealed that none of the 23 items fell inside the first quartile (0–25%; Table 6.2 & 6.4). However, additional analysis in the subgroups showed that 17 of these items were applicable for those with verbal communication skills at baseline. The remaining six verbal items fell for the verbal communication

subgroup within the second quartile (26–50%), meaning that these items were considered to be somewhat applicable. In contrast, in the subgroup without verbal communication skills at baseline, 19 items were not at all applicable, and 3 were hardly applicable. Only the item focusing on verbal aggression was somewhat applicable within this subgroup. Evidently, applicability of verbal-related items depended on pre-existing verbal communication skills.

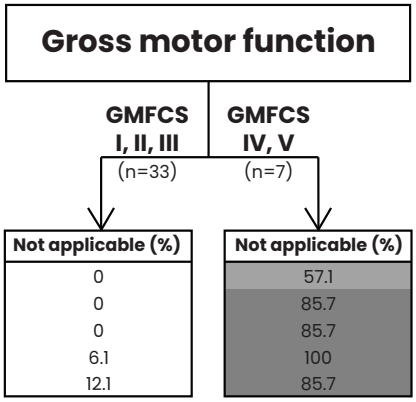


Item	Short item description	Not applicable (%)	Not applicable (%)
SRZ 30	Answering questions	0	61.5
DSVH 9	Reduced frequency/amount of speech	0	84.6
BPSD-DS II 5.5	Verbal stereotypy	0	88.5
SRZ 24	Use of language	0	100
SRZ 26	Comprehensibility of language	0	100
BPSD-DS II 6.1	Verbal aggression	7.1	50.0
DSVH 29	Less speaking/gestures	7.1	57.7
DVZ 22	Speaking	7.1	88.5
DSVH 31	Mumbling	7.1	96.2
DSVH 41	Using the wrong name	7.1	96.2
DSVH 30	Speaking more slowly/less intelligible	7.1	100
DSVH 56	Loss of speech	7.1	100
SRZ 27	Saying your name	7.1	100
SRZ 31	Naming and establishing links	7.1	100
DSVH 3	Word-finding difficulties	14.3	100
SRZ 25	Pronunciation of language	14.3	100
DVZ 13	Speaking intelligibly/comprehensibly	21.4	100
DVZ 29	Speaking about what you did	28.6	92.3
DSVH 26	Speaking about events as if they happened recently	28.6	100
SRZ 28	Repeating words	28.6	100
DVZ 31	Threatening by words/gestures	35.7	61.5
DVZ 18	Speaking about holiday/trip	42.9	88.5
BPSD-DS II 10.2	Making inappropriate comments	42.9	100

Figure 6.1 Applicability of verbal items for people with SPI(M)D with and without verbal communication skills. The not applicable percentages within each subgroup were divided into four quartiles, namely 0–25%, 26–50%, 51–75% and 76–100%. 0–25% are white meaning applicable, 26–50% are light gray meaning somewhat applicable, 51–75% are middle gray meaning hardly applicable and 76–100% are dark gray meaning not at all applicable. Abbreviations: BPSD-DS II, Behavioral and Psychological Symptoms of Dementia in Down Syndrome evaluation scale version II; DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; DVZ, Dutch Dementia Questionnaire for persons with mental retardation; SRZ, Social competence Rating scale for people with intellectual disabilities.

Gross motor function items

Additional analysis was also performed for items about gross motor function. Among the 40 participants, 33 had independent walking skills (i.e., GMFCS I, II, III), and 7 had not acquired walking skills (i.e., GMFCS IV, V). In total, five gross motor function items were identified. What stands out in Figure 6.2 is that all five items fell inside the first quartile (0–25%) for those able to independently walk and thus can consider to be applicable for this subgroup. Conversely, for those not able to independently walk, one item was hardly applicable, whereas the remaining four items were not at all applicable. Evidently, applicability of items about gross motor function depended on the ability to walk independently.



Item	Short item description	Not applicable (%)	Not applicable (%)
DSVH 60	Non-ambulatory	0	57.1
DSVH 53	Uncooperative to walk	0	85.7
DSVH 54	Uncooperative to carry one's own weight	0	85.7
DSVH 10	Sitting down at table	6.1	100
DVZ 36	Ability to get in/out bed	12.1	85.7

Figure 6.2 Applicability of gross motor function items for people with SPI(M)D with (i.e., GMFCS level I, II and III) and without (i.e., GMFCS level IV, V) independent walking skills. The not applicable percentages within each subgroup were divided into four quartiles, namely 0–25%, 26–50%, 51– 75% and 76–100%. 0–25% are white meaning applicable, 26–50% are light gray meaning somewhat applicable, 51–75% are middle gray meaning hardly applicable and 76–100% are dark gray meaning not at all applicable. Gross Motor Function Classification System (GMFCS) levels: Level I, can walk without limitations; Level II, walk with limitations; Level III, walk with assistive mobility device; Level IV, walking ability severely limited even with assistive devices, use of power wheelchair; Level V, transported by manual wheelchair. Abbreviations: DSVH, adapted Dutch version of the Dementia Scale for Down Syndrome; DVZ, Dutch Dementia Questionnaire for persons with mental retardation.

Discussion

In this study, applicable items in existing dementia screening instruments, namely DSVH, BPSD-DS II, DVZ, and SRZ, were identified by interviewing key informants of people with SPI(M)D. Our results demonstrated that 101 of the total 193 items could be considered applicable for individuals with SPI(M)D. Almost two-third of the applicable items focused on behavioral and psychological functioning, namely apathetic (13 items), depressive (9), sleeping problems (7), obstinate (6), anxious (5), irritable (5), restless/stereotypic (5), eating/drinking (5), aggressive (4) and disinhibited behavior (1). Moreover, among the 101 applicable items, 25 items focused on cognitive functioning, i.e., memory (7 items), orientation in place (5), person recognition (3), orientation in time (2), responsiveness (2), understanding visual images/spatial relationships (1), losing objects (1) and other cognitive functions (4). The remaining applicable items were items regarding motor functioning (6), ADL (5), and medical comorbidities (5). Additional analyses revealed that among 23 verbal communication items, 17 were applicable for individuals with verbal communication skills at baseline, but not if a person had never acquired such skills. Similarly, five items concerning gross motor function were found to be only applicable for those able to independently walk (GMFCS levels I, II, III) at baseline.

To diagnose dementia in people with SPI(M)D it is of essence to identify changes (decline). The results of this study indicate which skills/behavior people with SPI(M)D could potentially display before decline/dementia. If someone, at baseline (without decline) is able to show such skills/behavior, these may be of use in the context of dementia as informants may observe changes. Previous studies have focused on identifying observable dementia symptoms in this population (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). Hereafter, we conceptualize our findings about item applicability with reported observable dementia symptoms in previous studies, separately for the five domains: cognitive functioning, ADL, behavioral and psychological functioning, motor functioning, and medical comorbidities (American Psychiatric Association, 2013; Dekker, Ulgiati et al., 2021a; McKhann et al., 2011; Ries, 2018; Strydom et al., 2010; World Health Organization, 2022).

Cognitive functioning

One of the characteristics of the SPI(M)D population is that their cognitive functioning is limited resulting from their underlying ID (American Psychiatric Association, 2013; Nakken & Vlaskamp, 2007). Therefore, in clinical practice, it is commonly believed that it would be very hard to identify applicable cognitive items, because those with more severe ID may be unable to display cognitive skills (Startin, Rodger, et al., 2016). Despite low levels of baseline cognitive functioning, still 25 items focusing on cognitive functioning turned out to be applicable. This is consistent with three SPI(M)D dementia studies that showed that it is possible to observe cognitive dementia symptoms in individuals with SPI(M)D (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). Cognitive symptoms like memory loss, disorientation in place and language problems were in those with more severe ID particularly observed in different contexts, e.g., ADL, communication, leisure activities (Benejam, 2009; Dekker, Wissing, et al., 2021a). Initial analysis in this study revealed indeed applicable items focusing on memory and orientation in place but not on language skills. This finding could be attributed to limited or even absent verbal communication skills (Nakken & Vlaskamp, 2007; Nieuwenhuis-Mark, 2009; Oliver & Kalsy, 2005). Additional analysis revealed that in total 17 items – 15 cognitive and 2 behavioral and psychological items – about verbal communication were applicable for those with verbal communication skills but not for those without such skills. As already addressed in the studies of Dekker, Wissing, et al. (2021a) and Wissing, Fokkens, et al. (2022), observing alterations in language depends on whether at baseline someone has developed such skills.

ADL functioning

Individuals with SPI(M)D often need high levels of support to perform ADL. They might hardly have developed specific skills and therefore are (fully) dependent on others for daily tasks (Dekker, Wissing, et al., 2021a; Nakken & Vlaskamp, 2007). However, in the study of Wissing, Fokkens, et al. (2022), interviewees stressed that despite required assistance, in almost all individuals with SPI(M)D and dementia, they had observed a decline in eating/drinking skills. In line with that, most applicable items about ADL were identified within the feeding category (3 items). Moreover, applicable items were found focusing on transfers (1) and housework (1), whereas no applicable items were identified for the categories dressing, toilet use, grooming/bathing and shopping. Items within these four categories

are thus not applicable for the total SPI(M)D population. In contrast, it was previously reported that dementia symptoms like deterioration in the ability to dress or use the toilet were observed in individuals with SPI(M)D (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022). This may be explained by the fact that also people with SPI(M)D are able to perform small tasks within a larger activity, for example, by putting their arm in the sleeve during dressing. Even performing such small sub-tasks can deteriorate, and were therefore named in previous studies (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022). It is thus important to develop items specifically regarding performing the sub-tasks within larger tasks according to experiences in practice.

Behavioral and psychological functioning

To identify behavioral changes over time one should disentangle behavioral alterations from characteristic/typical behavior of an individual (Dekker, Strydom, et al., 2015). Our results showed that people with SPI(M)D could at baseline display behavior represented in 60 behavioral and psychological items. Such items should be used to screen for dementia in people with SPI(M)D. Behavioral and psychological symptoms of dementia are namely observed in all types of dementia (Finkel, 2000) and also prominent in people with DS (Dekker, Sacco, et al., 2018; Dekker, Strydom, et al., 2015; Dekker, Ulgiati et al., 2021a). Moreover, they are frequently observed dementia symptoms in people with SPI(M)D (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). In fact, behavioral and psychological changes related to dementia are more notable than alterations in cognitive functioning (Ball, Holland, Hon, et al., 2006; Ball et al., 2008; Engelborghs et al., 2005; Nelson et al., 2001), certainly in those with SPI(M)D (Wissing, Fokkens, et al., 2022). In the SPI(M)D population, particularly dementia symptoms like increased irritability, anxiety, apathy and decreased eating/drinking behavior were frequently observed, whereas psychotic symptoms seem less prevalent (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022). In this study, items focusing on psychotic behavior were either hardly or not at all applicable, mainly because of limited intellectual functioning and verbal communication. Previous studies indeed noted that recognizing psychotic symptoms is particularly complex in those with limited verbal communication skills, because they are hardly able to self-report the inner experiences hallucinations and/or delusions (Cooper & Smiley, 2007; Moss et al., 1993; Temple & Konstantareas, 2005).

Motor functioning

Many people with SPI(M)D have to some extent limitations in motor functioning (Houwen et al., 2014; Nakken & Vlaskamp, 2007). However, our results demonstrated that despite pre-existing motor problems, all motor items, namely balance/fall frequency (2 items), movement speed/quality (2), fine motor skills (1) and walking (1) were applicable for people with SPI(M)D. This may seem contradictory, but every individual is – despite limitations in motor functioning – to a certain extent able to move (parts of) their body. Consequently, motor changes can also be observed in individuals with SPI(M)D, for example, decreased movement speed and/or quality. Such motor changes might be related to dementia given that a decline in motor functioning was recognized in individuals with dementia, not only in the general population (Ries, 2018) but also in the SPI(M)D population (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). Moreover, a decline in walking skills in people with SPI(M)D and dementia was only observed in individuals who were able to walk at baseline (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022). Indeed, our additional analysis of gross motor function showed that five items about gross motor function, including the motor item about walking, were only applicable for those able to independently walk at baseline (GMFCS levels I, II, III).

Medical comorbidities

People with SPI(M)D frequently experience physical health problems such as vision problems, epilepsy, constipation and incontinence (Nakken & Vlaskamp, 2007; van Timmeren et al., 2017). Particularly, the onset of epilepsy and incontinence are medical comorbidities related to dementia not only in the general (Kurrle et al., 2012) and DS population (Aller-Alvarez et al., 2017; Strydom et al., 2010) but also in the SPI(M)D population (Dekker, Wissing et al., 2021; Wissing, Fokkens et al., 2022, Wissing, Ulgiati et al., 2022). Our results demonstrated that items focusing on epilepsy were applicable for all 40 individuals with SPI(M)D, and thus could be used for this population. Conversely, no applicable items focusing on incontinence were identified, whereas previous studies have shown increased incontinence in people with SPI(M)D and dementia (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). Not identifying applicable items for incontinence is likely to be related to individuals being incontinent at baseline. In fact, the study of van Timmeren et al. (2016) found a prevalence rate for incontinence of 56% for people with SPI(M)D.

Strengths

Existing dementia screening instruments for people with ID as a whole were found to be unsuitable for people with SPI(M)D (Elliott-King et al., 2016; Evenhuis, 1990; Hon et al., 1999; Margallo-Lana et al., 2007). To the best of our knowledge, this study is the first to show that specific items within existing lists are applicable to screen for dementia in individuals with SPI(M)D. Another strength of this study is that we took into account the heterogeneity of the SPI(M)D population. We included people with either a severe or a profound ID and various underlying causes, including DS. We took into account the high genetic risk of developing dementia for people with DS (Ballard et al., 2016), by making sure that at least one-fourth of the total participants had DS. Moreover, we considered the variety of verbal communication and gross motor skills in people with SPI(M)D. Additional analyses allowed to refine results in relation to the presence or absence of these skills.

Limitations

Relatively, a large number of legal representatives, which received an information letter with informed consent form, did either not respond or did not provide consent. This might be explained by the fact that they might not see the added value of filling out dementia screening instruments when the functioning of the person is stable and their relative does not (yet) have dementia. When the information was further clarified, either face-to-face or by a phone call, legal representatives were more willing to provide informed consent. Due to practical difficulties, this was not done within every care organization. Moreover, there are no standardized tests applicable for a valid estimation of the level of ID (Nakken & Vlaskamp, 2007). Therefore, the categorization of severe ID (60%) and profound ID (40%) is based on clinical judgment. There seems to be a slight underrepresentation of those with the most severe ID. As a consequence, items having not applicable percentages around the threshold of quartiles could potentially have been attributed to another quartile when more individuals with profound ID were included. Another possible limitation is the fact that some interviewers were involved in the diagnostic work-up/care for the individual with SPI(M)D. To minimize risk of bias, an independent researcher, unacquainted with the individuals with SPI(M)D, made sure that answers were provided by informants (not the interviewer). Moreover, although care organizations in The Netherlands provide care/support in a variety of residential facilities, ranging from smaller assisted living facilities in communities to larger,

specialized locations, we cannot rule out a potential effect of living situation of individuals on the scoring. Lastly, we only identified applicable items in dementia screening instruments for which a translated/validated Dutch version was available, and thus not for internationally used instruments such as the CAMDEX-DS (Ball, Holland, Huppert, et al., 2006). Nevertheless, the four selected instruments are internationally recommended and widely used to screen for dementia in people with ID (Zeilinger et al., 2022).

Future implications

Timely recognizing and diagnosing dementia in people with SPI(M)D is a major challenge. Today, a clinical diagnosis of dementia in individuals with SPI(M)D is purely based on observations, interviewing informants, and/ or screening case notes (Day, 1985; Duggan et al., 1996; Evenhuis, 1990; Määttä et al., 2006; Margallo-Lana et al., 2007; Reid & Aungle, 1974; Sauna-Aho et al., 2018). Existing dementia screening instruments as a whole are namely unsuitable for this population. This primarily relates to the pre-existing disabilities, which make that not all items within instruments can be scored. In this study, we have shown which skills/behavior individuals with SPI(M)D may – despite pre-existing disabilities – display before decline/ dementia. Based on these results, it cannot yet be determined whether applicable items are indeed relevant to screen for dementia symptoms in those with SPI(M)D. Further research is required to establish whether people with SPI(M)D and dementia indeed show alterations in applicable items. Previous studies already demonstrated which dementia symptoms could potentially be observed in those with SPI(M)D (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022). The authors stress that both aspects: 1) identified applicable items in existing dementia instruments available for people with ID and 2) identified practice-based observation of dementia symptoms in the SPI(M)D population should form the basis for developing a novel dementia screening instrument dedicated to people with SPI(M)D. Moreover, such an instrument should, differently from direct neuropsychological tests, not only focus on a decline in cognitive functioning. Instead, also the ADL, behavioral and psychological, motor, and medical comorbidities domains should be included, because in those with SPI(M)D a decline in cognitive functioning will be observable in all other domains (Benejam, 2009; Dekker, Wissing, et al., 2021a). Additionally, such an instrument should contain a statement that symptoms could be caused by – often treatable – conditions such as depression, delirium, vision or hearing problems, hypothyroidism, sleep

apnea, or vitamin B12 deficiency, which should be ruled out as much as possible before diagnosing dementia (Moriconi et al., 2015; Scott & Barrett, 2007).

Conclusion

This study provided an overview of applicable items for people with SPI(M)D in existing dementia screening instruments available for people with ID. Among 193 items, 101 were found to be applicable for individuals with SPI(M)D. Most applicable items were identified within the behavioral and psychological domain (60 items), followed by cognitive (25), motor (6), ADL (5), and medical comorbidities (5) domains. Moreover, 17 items focusing on verbal communication skills and 5 about gross motor function were specifically found to be applicable for individuals with verbal/ walking skills at baseline. The inventory of applicable items together with the findings of observable dementia symptoms in people with SPI(M)D (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022; Wissing, Ulgiati, et al., 2022) are key elements for developing a new dementia screening instrument dedicated to people with SPI(M)D. Developing a new instrument is essential to be able to timely identify dementia and prevent (too) late diagnosis or no diagnosis at all. This allows to early respond to the person's changing wishes and needs in order to maintain quality of life in people SPI(M)D and dementia.

Chapter 7

Diagnostic aid for dementia in people with severe/profound intellectual (and multiple) disabilities: development and first test in practice

Dutch article with English abstract |
Nederlandstalig artikel met Engelstalige samenvatting
Wissing, M. B. G., Koudenburg, S. D., van der Wal, I. A., Groen, M. R., van Dam, L., Hobbelen, J. S. M., De Deyn, P. P., Waninge, A., & Dekker, A. D. Diagnostisch hulpmiddel dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen: ontwikkeling en eerste praktijktoets. (2023) *NTZ*: *Nederlands Tijdschrift voor de Zorg aan mensen met een verstandelijke beperking*, *50*(3).

Abstract

Introduction: Diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D) is complex. Currently, no standardized dementia screening instrument exists for people with SPI(M)D. Therefore, development of a suitable instrument to aid diagnosis of dementia in this population is desired. The aim of this study was 1) to develop an instrument for psychologists and psychological assistants to identify dementia-related changes in people with SPI(M)D and 2) to examine validity, reliability, discriminative ability and practice-based experiences of this instrument.

Methods: Scientific literature and practice-based experiences guided the development of the instrument. The instrument was tested in clinical practice by conducting structured interviews with informants of people with SPI(M)D without dementia (n=18), with questionable dementia (n=10) and with diagnosed dementia (n=8).

Results: The instrument comprised 45 items, categorized into 7 symptom domains, which were developed through the triangulation of research findings. This ensured face and content validity. Initial findings on reliability demonstrated that the interrater reliability was high. Regarding discriminative ability, people with diagnosed dementia tended to show most changes and people without dementia least based on item scores, domain scores and total scores. Experiences from psychologists, psychological assistants, and informants were predominantly positive.

Conclusion: The first results suggest that this instrument can identify dementia-related changes in people with SPI(M)D. It can be implemented in intellectual disability care organizations, whereby it is recommended to further study reliability and discriminative ability.

Samenvatting

Introductie: Diagnosticeren van dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen ((Z)EV(M)B) is complex. Aangezien reeds bestaande instrumenten niet geschikt zijn voor de (Z)EV(M)B-doelgroep is er in de gehandicaptenzorg grote behoefte aan een passend diagnostisch hulpmiddel. Dit onderzoek richt zich op 1) het ontwikkelen van een diagnostisch hulpmiddel voor gedragskundigen en psychodiagnostisch medewerkers om dementiegerelateerde veranderingen bij mensen met (Z)EV(M)B in kaart te brengen en 2) de validiteit, betrouwbaarheid, het discriminerend vermogen en praktijkervaringen van dit diagnostisch hulpmiddel te bepalen middels een eerste praktijktoets.

Methode: Op basis van wetenschappelijke literatuur en praktijkervaringen is het diagnostisch hulpmiddel ontwikkeld. Vervolgens is dit diagnostisch hulpmiddel onderworpen aan een eerste praktijktoets, waarbij interviews zijn gehouden met informanten van mensen met (Z)EV(M)B zonder dementie (n=18), met twijfelachtige dementie (n=10) en een diagnose dementie (n=8).

Resultaten: Het diagnostisch hulpmiddel bestaat uit 45 items, onderverdeeld in 7 symptoomdomeinen, tot stand gekomen op basis van triangulatie van bevindingen in wetenschappelijke literatuur en praktijkervaringen. Hierdoor zijn de indruks- en inhoudsvaliditeit verzekerd. De eerste verkenning van de betrouwbaarheid liet zien dat de interbeoordelaarsbetrouwbaarheid hoog was. Met betrekking tot het discriminerend vermogen werd zowel voor de item-, domein- als totaalscores een trend gevonden waarbij mensen met een diagnose dementie de meeste veranderingen scoorden en degenen zonder dementie de minste veranderingen. Praktijkervaringen ten aanzien van het diagnostisch hulpmiddel waren overwegend positief.

Conclusie: De resultaten wijzen erop dat het diagnostisch hulpmiddel dementiegerelateerde veranderingen bij mensen met (Z)EV(M)B in kaart kan brengen. Het diagnostisch hulpmiddel kan daarom al in de praktijk ingezet worden. Daarbij is wel het advies om de betrouwbaarheid en het discriminerend vermogen verder te onderzoeken.

Introductie

De levensverwachting van mensen met verstandelijke beperkingen (VB) neemt toe, wat onder meer te relateren is aan betere (medische) zorg (Coppus, 2013; Evans et al., 2013; Torr & Davis, 2007). Aangezien hogere leeftijd de belangrijkste risicofactor is voor dementie (Alzheimer Nederland, 2021), komt dementie ook vaker voor bij mensen met VB, onder wie mensen met (Z)EV(M)B. Dementie is een verzamelnaam voor aandoeningen die gekenmerkt worden door een achteruitgang in het cognitief functioneren t.o.v. een eerder niveau van functioneren dat effect heeft op het dagelijks functioneren (American Psychiatric Association & Nederlandse Vereniging voor Psychiatrie, 2014; Nederlandse Vereniging voor Klinische Geriatrie, 2014; Wereldgezondheidsorganisatie, 2014). De voornaamste oorzaak van dementie is de ziekte van Alzheimer (Alzheimer Nederland, 2021). Mensen met het downsyndroom (DS) hebben een hoog genetisch bepaald risico op dementie door de ziekte van Alzheimer (Dekker & De Deyn, 2018; Wiseman et al., 2015). Van de mensen met DS heeft 20-30% (Z)EV(M)B (Coppus, 2017; Coppus et al., 2006). Dementie komt dus steeds vaker voor bij mensen met (Z)EV(M)B en wordt daardoor een steeds grotere uitdaging voor de hedendaagse verstandelijke gehandicaptenzorg.

Mensen met (Z)EV(M)B hebben een geschat IQ van minder dan 35 punten. Daarnaast hebben ze vaak ernstige gezondheidsproblemen en motorische, zintuiglijke en communicatieve beperkingen (Van der Putten et al., 2017; Van Timmeren et al., 2016). Bij mensen met (Z)EV(M)B is het signaleren en diagnosticeren van dementie ingewikkelder dan in de algemene bevolking of bij mensen met lichte/matige VB. Dit komt onder andere doordat mensen met (Z)EV(M)B al (zeer) ernstige beperkingen hebben waardoor het moeilijker is om achteruitgang van vaardigheden door dementie vast te stellen. Toch is vroegtijdig signaleren en diagnosticeren van dementie belangrijk voor mensen met (Z)EV(M)B en hun omgeving. Uit recent focusgroeponderzoek (Dekker, Wissing, et al., 2021b) bleek dat een diagnose van dementie familieleden en zorgmedewerkers in staat stelt om de veranderingen die iemand laat zien te begrijpen en om geïnformeerde keuzes te kunnen maken. Bij geïnformeerde keuzes kan gedacht worden aan keuzes ten aanzien van begeleiding (bijv. bijstellen van begeleidingsdoelen en -stijl op basis van de veranderende ondersteuningsbehoefte), behandeling (bijv. aanpassen van behandelplan en medicatiegebruik), verwachtingsmanagement

& perspectief (bijv. anticiperen op ziekteverloop) en organisatie (bijv. aanpassen van de dagbesteding en/of woonsituatie) (Dekker, Wissing, et al., 2021b).

Dementie vaststellen bij mensen met (Z)EV(M)B is dus belangrijk, maar ook complex. In de algemene bevolking wordt vaak gebruik gemaakt van neuropsychologische testen om te bepalen of er sprake is van dementiegerelateerde cognitieve achteruitgang (Alzheimer's Association, 2022; Salmon & Bondi, 2009). Mensen met (Z)EV(M)B hebben al (zeer) ernstige beperkingen in het cognitief functioneren waardoor zij testinstructies zeer beperkt begrijpen en zij beschikken veelal over onvoldoende verbale vaardigheden die nodig zijn voor het afnemen van zulke testen (Nieuwenhuis-Mark, 2009; Oliver & Kalsy, 2005). Als gevolg hiervan heeft direct neuropsychologisch onderzoek geen of nauwelijks toegevoegde waarde voor mensen met (Z)EV(M)B (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018).

Voor mensen met VB zijn dementielijsten ontwikkeld die door middel van informantinterviews met familieleden en begeleiders worden ingevuld. Lijsten die veel gebruikt worden in Nederland zijn de *Dementie Vragenlijst voor Verstandelijk Gehandicapten* (DVZ) (Evenhuis et al., 1998), *Dementie Schaal voor mensen met een Verstandelijke Handicap* (DSVH) (Maaskant & Hoekman, 2011). De *Sociale Redzaamheidsschaal voor verstandelijk gehandicapten* (SRZ) (Kraijer & Kema, 1994) wordt, hoewel in steeds mindere mate, ook gebruikt om achteruitgang over de tijd in sociale redzaamheid te meten. Daarnaast wordt vanuit de literatuur het gebruik van de recent ontwikkelde *Gedrags-(Behavioural) en Psychologische Symptomen van Dementie bij DownSyndroom* (BPSD-DS-II) evaluatieschaal (Dekker, Ulgiati, et al., 2021b; Dekker, Vermeiren et al., 2018) aanbevolen (Zeilinger et al. 2022). Recent onderzoek liet zien dat deze lijsten slechts gedeeltelijk toepasbaar zijn voor mensen met (Z)EV(M)B (Wissing, Dijkstra, et al., 2022). Ze bevatten namelijk items waarin wordt gevraagd naar veranderingen in vaardigheden die mensen met (Z)EV(M)B niet hebben ontwikkeld. Daardoor zijn dergelijke items niet van toepassing, d.w.z. niet bruikbaar om een achteruitgang in functioneren door dementie in kaart te brengen (Wissing, Dijkstra, et al., 2022).

Het diagnosticeren van dementie bij mensen met (Z)EV(M)B bestaat, door de afwezigheid van geschikte dementielijsten, vooral uit klinische beoordeling door (ervaren) artsen en gedragskundigen en gesprekken met familieleden en begeleiders (Duggan et al., 1996; Evenhuis, 1990; Margallo-Lana et al., 2007; Reid & Aungle, 1974; Sauna-Aho et al., 2018). Om achteruitgang in vaardigheden veroorzaakt door dementie vast te stellen bij de (Z)EV(M)B-doelgroep is er in de gehandicaptenzorg grote behoefte aan een passend diagnostisch hulpmiddel (Dekker, Wissing, et al., 2021b).

Dit onderzoek heeft als doel om 1) een diagnostisch hulpmiddel te ontwikkelen voor gedragskundigen en psychodiagnostisch medewerkers om bij mensen met (Z)EV(M)B dementiegerelateerde veranderingen in kaart te brengen en 2) de validiteit, betrouwbaarheid, het discriminerend vermogen en praktijkervaringen van dit diagnostisch hulpmiddel te bepalen middels een eerste praktijktoets.

Methode

Samenwerkingsverband
Dit onderzoek is onderdeel van het project '*Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*', een samenwerking tussen Rijksuniversiteit Groningen, Universitair Medisch Centrum Groningen (UMCG), Hanzehogeschool Groningen, Alliade, 's Heeren Loo, Ipse de Bruggen en Koninklijke Visio. De laatste vier zijn zorgorganisaties verspreid over heel Nederland die diagnostiek, begeleiding en behandeling bieden aan mensen met VB. Dit project wordt gesubsidieerd door ZonMW (projectnummer 733050863). Meer informatie: www.vb-dementie.nl.

Studieopzet
Dit onderzoek bestaat uit twee onderdelen: 1) ontwikkeling van een diagnostisch hulpmiddel en 2) eerste praktijktoets daarvan t.a.v. validiteit, betrouwbaarheid, discriminerend vermogen en praktijkervaringen.

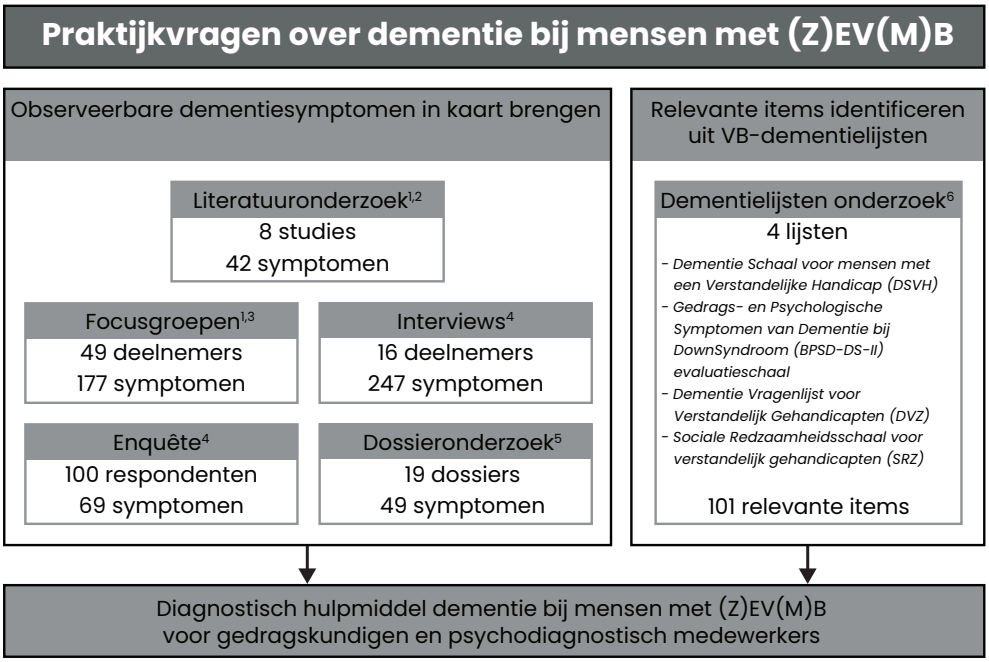
Ethiek en toestemmingsprocedure
De Medisch-Ethische Toetsingscommissie van het UMCG oordeelde dat het onderzoek niet onder de reikwijdte valt van de Wet medisch-wetenschappelijk onderzoek met mensen (METc 2019/198). Het onderzoek is opgenomen in het UMCG Research Register onder nummer 201900193

en is uitgevoerd conform de UMCG Research Code en de Algemene Verordening Gegevensbescherming. Wettelijk vertegenwoordigers van mensen met (Z)EV(M)B gaven schriftelijk toestemming voor het afnemen van het diagnostisch hulpmiddel en analyseren van verzamelde gegevens.

Ontwikkeling diagnostisch hulpmiddel

Schaalontwikkeling

Het diagnostisch hulpmiddel is ontwikkeld op basis van triangulatie van bevindingen (van Staa & Evers, 2010) uit eerdere deelonderzoeken van het project '*Praktijkvragen over dementie bij mensen met (Z)EV(M)B'* (zie Figuur 7.1). Door gebruik te maken van verschillende kwalitatieve en kwantitatieve onderzoeksmethoden zijn observeerbare dementiesymptomen bij mensen met (Z)EV(M)B in kaart gebracht. Allereerst zijn dementiesymptomen opgehaald uit wetenschappelijke literatuur (Dekker, Wissing, et al., 2021b; Wissing, Ulgiati, et al., 2022). Hieruit bleek dat literatuur over de uiting van dementie bij deze groep zeer beperkt was. Daarom is verder gegaan met bevraging van zorgmedewerkers (artsen, verpleegkundig specialisten, gedragskundigen, psychodiagnostisch medewerkers, paramedici en begeleiders) en familieleden die praktijkervaring hebben met mensen met (Z)EV(M)B en de combinatie met dementie. Dementiesymptomen zijn opgehaald middels verschillende onderzoeksmethoden: multidisciplinaire focusgroepen (Dekker, Wissing, et al., 2021b; Wissing, Ulgiati, et al., 2022), enquête onder zorgmedewerkers en familieleden (Wissing, Fokkens, et al., 2022) en verdiepende interviews met zorgmedewerkers met ruime ervaring op het gebied van dementie bij mensen met (Z)EV(M)B (Wissing, Fokkens, et al., 2022). Bovendien is dossieronderzoek verricht om dementiesymptomen te verzamelen die staan gerapporteerd in dossiers van mensen met (Z)EV(M)B (Wissing, Hobbelen et al., 2023). Naast het in kaart brengen van dementiesymptomen zijn relevante items voor mensen met (Z)EV(M)B geïdentificeerd uit vier bestaande dementielijsten voor mensen met VB (Wissing, Dijkstra, et al., 2022). De in totaal 193 items zijn door gedragskundigen/psychodiagnostisch medewerkers tijdens interviews voorgelegd aan informanten (begeleiders en familieleden) van mensen met (Z)EV(M)B zonder dementie. Een item werd als relevant beschouwd wanneer mensen met (Z)EV(M)B in staat waren de vaardigheid/gedrag te tonen.



Figuur 7.1 Schematische weergave van de resultaten verkregen middels verschillende deelonderzoeken binnen het project 'Praktijkvragen over dementie bij mensen met (Z)EV(M)B'. Samenvoegen en analyseren van de resultaten heeft geleid tot de ontwikkeling van items voor het diagnostisch hulpmiddel. Afkortingen: VB, verstandelijke beperkingen; (Z)EV(M)B, (zeer) ernstige verstandelijke (en meervoudige) beperkingen. Referenties: 1, Dekker, Wissing, et al., 2021b; 2, Wissing, Ulgiati, et al., 2022; 3, Dekker, Wissing, et al., 2021a; 4, Wissing, Fokkens, et al., 2022; 5, Wissing, Hobbelen et al., 2023; 6, Wissing, Dijkstra, et al., 2022.

In iedere onderzoeksstap zijn de opgehaalde dementiesymptomen/ relevante items ingedeeld in vijf domeinen, in overeenstemming met de diagnostische criteria voor dementie (American Psychiatric Association & Nederlandse Vereniging voor Psychiatrie, 2014; Nederlandse Vereniging voor Klinische Geriatrie, 2014; Wereldgezondheidsorganisatie, 2014) en wetenschappelijke literatuur (Dekker, Ulgiati, et al., 2021b; Dekker, Vermeiren et al., 2018; Ries, 2018; Strydom et al., 2010), te weten: 1) veranderingen in cognitie, 2) veranderingen in algemene dagelijkse levensverrichtingen (ADL), 3) gedragsveranderingen, 4) veranderingen in motoriek en 5) medische comorbiditeiten. Voor verdere duiding zijn symptomen/ relevante items gebundeld in symptoomcategorieën. Tabel 7.1 toont een overzicht van opgehaalde symptoomcategorieën en geïdentificeerde toepasbare relevante items uit bestaande dementielijsten voor

mensen met VB (triangulatie). Voor symptoomcategorieën die in 4 of 5 onderzoeksmethoden – literatuuronderzoek, focusgroepen, enquête, interviews, dossieronderzoek – zijn gevonden, zijn items ontwikkeld voor in het diagnostisch hulpmiddel. Indien relevante items voor deze symptoomcategorieën waren geïdentificeerd in bestaande dementielijsten, dan zijn deze gebruikt als inspiratie voor de ontwikkeling van de items voor het diagnostisch hulpmiddel.

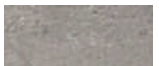
Om de items in het nieuwe diagnostisch hulpmiddel verder aan te scherpen en te ordenen, zijn deze in een overleg voorgelegd aan gedragskundigen en psychodiagnostisch medewerkers werkzaam bij de vier betrokken zorgorganisaties. Verder is een handleiding geschreven over onder andere de procedure, de scoring en het gebruik van het digitale systeem. De conceptversie en handleiding zijn verder geoptimaliseerd door het diagnostisch hulpmiddel in te laten vullen door gedragskundigen en psychodiagnostisch medewerkers aan de hand van drie proefinterviews met familie/begeleiders van mensen met (Z)EV(M)B, met en zonder twijfelachtige dementie of een diagnose dementie. Op basis van opmerkingen van de betrokkenen is de handleiding verbeterd, is een item uit het hulpmiddel verwijderd, zijn items geherformuleerd en aanvullende voorbeelden/uitleg toegevoegd. Hierna is de versie die getoetst ging worden vastgesteld.

Tabel 7.1 Overzicht (triangulatie) van dementiesymptomen bij mensen met (Z)EV(M)B verkregen door vijf onderzoeksmethoden met daarbij geïdentificeerde relevante items uit bestaande dementielijsten voor mensen met VB.

Symptoomdomeinen en -categorieën		Onderzoeksmethoden					
		Literatuur[1,2]	Focus-groepen[1,3]	Enquête[4]	Interviews[4]	Dossiers[5]	VB-dementie-lijsten[6]
Veranderingen in cognitie	↓ Geheugen (amnesie)	✔	✔	✔	✔	✔	✔
	↓ Oriëntatie in plaats	✔	✔	✔	✔	✔	✔
	↓ Uitvoeren handelingen (apraxie)	✔	✔	✔	✔	✔	✔
	↓ Taalvaardigheden (afasie)	✔	✔	✔	✔	✔	✔*
	↑ Spullen kwijtraken	-	✔	✔	✔	✔	✔
	↓ Oriëntatie in tijd	-	✔	✔	✔	✔	✔
	↓ Begrip visuele beelden/ ruimtelijke verhoudingen	-	✔	✔	✔	✔	✔
	↓ Herkenning personen/ objecten/geluiden (agnosie)	-	✔	✔	✔	✔	✔
	↓ Reactievermogen	-	✔	✔	✔	✔	✔
	↓ Voorkeur (geliefde) objecten	-	✔	✔	✔	✔	-
	↓ Volgordebesef	-	✔	✔	✔	✔	-
	↑ Verwardheid	✔	✔	-	-	-	-
	↓ Concentratie	-	-	✔	✔	-	-
	↑ Sensorische gevoeligheid	-	-	✔	✔	-	-
	↓ Sociale vaardigheden	✔	-	-	-	-	-
	↓ Persoonlijke gewoontes	✔	-	-	-	-	-
	↓ Planningsvaardigheden	-	-	✔	-	-	-
	↓ Probleemoplossend vermogen	-	-	✔	-	-	-
	↓ Beoordelingsvermogen	-	-	✔	-	-	-

Tabel 7.1 Vervolg.

Symptoomdomeinen en -categorieën		Onderzoeksmethoden					
		Literatuur[1,2]	Focus-groepen[1,3]	Enquête[4]	Interviews[4]	Dossiers[5]	VB-dementie-lijsten[6]
Veranderingen in ADL	↓ ADL	✔	✔	✔**	✔	✔	-
	↓ Eet- en drinkvaardigheden	✔	✔		✔	✔	✔
	↓ Verzorging	✔	✔		✔	✔	✔
	↓ Vrijetijdsbesteding	-	✔		✔	-	-
	↓ Huishoudelijk werk	-	-		✔	-	✔
	↓ Traplopen	-	-		✔	✔	-
Gedragsveranderingen	↑ Prikkelbaar gedrag	✔	✔	✔	✔	✔	✔
	↑ Slaapproblemen	✔	✔	✔	✔	✔	✔
	↑ Rusteloos en stereotiep gedrag	✔	✔	✔	✔	✔	✔
	↑ Agressief gedrag	✔	✔	✔	✔	✔	✔
	↑ Apathisch gedrag	✔	✔	✔	✔	✔	✔
	↓ Eet- en drinkgedrag	✔	✔	✔	✔	✔	✔
	↑ Angstig gedrag	-	✔	✔	✔	✔	✔
	↑ Koppig gedrag	-	✔	✔	✔	✔	✔
	↑ Depressief gedrag	-	✔	✔	✔	✔	✔
	↑ Ontremd gedrag	✔	✔	-	✔	✔	✔
	↑ Psychotisch gedrag	-	✔	✔	✔	✔	-



Tabel 7.1 Vervolg.

Symptoomdomeinen en -categorieën		Onderzoeksmethoden					
		Literatuur[1,2]	Focus-groepen[1,3]	Enquête[4]	Interviews[4]	Dossiers[5]	VB-dementie-lijsten[6]
Veranderingen in motoriek	↓ Loopvaardigheden	✔	✔	✔	✔	✔	✔
	↑ Rolstoelgebruik	✔	✔	✔	✔	✔	-
	↑ Spierverkrampingen	✔	✔	✔	✔	✔	-
	↓ Transfers/mobiliteit	✔	✔	-	✔	✔	✔
	↓ Balans	-	✔	✔	✔	✔	✔
	↑ Valfrequentie	-	✔	✔	✔	✔	✔
	↓ Kauwen/slikken	-	✔	✔	✔	✔	-
	↑ Stijfheid	-	✔	✔	✔	✔	-
	↓ Spierkracht	-	✔	✔	✔	✔	-
	↓ Bewegingssnelheid/kwaliteit	-	-	✔	✔	✔	✔
Medische comorbiditeiten	↑ Epilepsie	✔	✔	✔	✔	✔	✔
	↑ Incontinentie	-	✔	✔	✔	✔	-
	↓ Gewicht	-	✔	✔	✔	✔	-

Symptoomcategorieën en toepasbare items zijn ingedeeld in vijf domeinen volgens diagnostische dementiecriteria (American Psychiatric Association & Nederlandse Vereniging voor Psychiatrie, 2014; Nederlandse Vereniging voor Klinische Geriatrie, 2014; Wereldgezondheidsorganisatie, 2014) en wetenschappelijke literatuur (Dekker, Ulgiati, et al., 2021b; Dekker, Vermeiren, et al., 2018; Ries, 2018; Strydom et al., 2010). Gedrag kan zowel toe- als afnemen, alleen de meest prominente verandering is weergegeven in de tabel. Een ✔ geeft aan dat een symptoomcategorie opgehaald is met de desbetreffende onderzoeksmethode/ dat één of meerdere toepasbare items in bestaande dementielijsten voor mensen met verstandelijke beperkingen (VB) zijn geïdentificeerd. Wanneer een symptoomcategorie in 4 of 5 van de onderzoeksmethode is gevonden, dan is hiervoor een nieuw item ontwikkeld (wit gekleurde symptoomcategorieën). Symbolen: ↓, afname t.o.v. karakteristieke functioneren; ↑, toename t.o.v. karakteristieke functioneren; *, toepasbare items geïdentificeerd voor personen met verbale vaardigheden (karakteristiek); **, in de enquête zijn veranderingen in algemene dagelijkse veranderingen (ADL) niet uitgesplitst uitgevraagd. Referenties: 1, Dekker, Wissing, et al., 2021b; 2, Wissing, Ulgiati, et al., 2022; 3, Dekker, Wissing, et al., 2021a; 4, Wissing, Fokkens, et al., 2022; 5, Wissing, Hobbelen et al., 2023; 6, Wissing, Dijkstra, et al., 2022.

Scoring

Voor elk item in de schaal wordt gescoord of in de laatste zes maanden verandering zichtbaar is t.o.v. het karakteristieke functioneren/gedrag. Met karakteristiek wordt het functioneren/gedrag bedoeld dat kenmerkend was voor de persoon en hij/zij gedurende het volwassen leven heeft laten zien, dus voordat achteruitgang optrad. Aangezien dementie zich kenmerkt door een achteruitgang in functioneren t.o.v. een eerder hoger niveau van functioneren (American Psychiatric Association & Nederlandse Vereniging voor Psychiatrie, 2014; Nederlandse Vereniging voor Klinische Geriatrie, 2014; Wereldgezondheidsorganisatie, 2014), kunnen items gescoord worden als '*ja, afname*' of '*nee, geen verandering'.* Ook zijn gedragsveranderingen veelvoorkomend bij dementie. Meestal wordt een toename van gedrag gezien, maar afname wordt ook waargenomen (Dekker, Ulgiati, et al., 2021b; Dekker, Vermeiren et al., 2018). Daarom kunnen de items over gedragsveranderingen gescoord worden als '*ja, toename,*' '*ja, afname*' of '*nee, geen verandering'.* Dit geldt ook voor de items over voorkeur voor objecten (item 1.5), eetlust/drinken (item 4.2) en lichaamsgewicht (item 4.5). Aan iedere verandering – afname of toename – wordt een score 1 toegekend en wanneer er geen verandering is, dan wordt een score 0 toegekend.

Mensen met (Z)EV(M)B hebben reeds bestaande (zeer) ernstige beperkingen in functioneren, waardoor zij bepaalde vaardigheden niet hebben ontwikkeld (Nakken & Vlaskamp, 2007). Bij een aantal items, namelijk de tien items over cognitieve veranderingen, eet- en drinkvaardigheden (item 4.1), kauwen (item 4.4) en verzorging (item 5.1) is daarom de optie '*niet van toepassing, vaardigheid nooit ontwikkeld*' toegevoegd. Bij het item over incontinentie (item 7.2) kan *'niet van toepassing*' gescoord worden wanneer de persoon altijd al incontinent was. Wanneer *'niet van toepassing*' wordt gescoord, dan wordt geen score toegekend aan dat item.

Voor de items over taal en spraak (item 2.1 en 2.2) en loopvaardigheden (item 6.8 en 6.9) geldt dat wanneer de persoon deze vaardigheden niet heeft ontwikkeld, er geen achteruitgang geobserveerd kan worden. Deze vragen worden daarom via een selectievraag alleen aangeboden en ingevuld wanneer de persoon karakteristiek deze vaardigheden heeft getoond, dus voordat er sprake was van achteruitgang. Alleen in dat geval worden er scores toegekend aan deze items.

Afhankelijk van de woonsituatie is er niet altijd zicht op het slaapgedrag van de persoon. Daarom is er bij het item 's nachts wakker (item 3.6) de extra scoringsoptie *'onbekend, geen zicht op het slaapgedrag'* toegevoegd. Wanneer deze optie wordt gescoord, dan wordt er net als bij *'niet van toepassing'* geen score toegekend aan dit item.

Digitalisering
Voor dit onderzoek is een digitale versie van het diagnostisch hulpmiddel gemaakt in REDCap (Harris et al., 2009) dat wordt gehost via het beveiligde netwerk van het UMCG. Interviewers werden stapsgewijs door het diagnostisch hulpmiddel geleid. De volledigheid van de ingevoerde gegevens is gewaarborgd doordat een melding verschijnt wanneer items niet worden gescoord. Items over taal en spraak en loopvaardigheden worden automatisch wel of niet aangeboden aan de hand van de selectievraag hierover. Voor data-analyse werd een gepseudonimiseerde SPSS-exportbestand gedownload uit REDCap.

Praktijktoets diagnostisch hulpmiddel

Deelnemers
Potentiële deelnemers met en zonder twijfelachtige dementie of een diagnose dementie zijn geselecteerd voor de praktijktoets door gedragskundigen en psychodiagnostisch medewerkers werkzaam binnen de vier betrokken zorgorganisaties. Inclusiecriteria waren: (zeer) ernstige VB (oorspronkelijke mate VB, voordat eventuele achteruitgang optrad), vastgesteld op basis van gegevens uit het cliëntdossier en klinisch oordeel, en leeftijd van 30 jaar of ouder. Mensen konden deelnemen ongeacht de aanwezigheid van DS of bijkomende beperkingen (bijv. motorisch of visueel). Exclusiecriteria waren: lichte of matige VB, niet-aangeboren hersenletsel, langdurige ziekenhuisopname in de afgelopen zes maanden en terminaal zijn. Daarnaast zijn personen die in de laatste zes maanden veranderingen toonden die – naar klinisch oordeel – te wijten zijn aan recente levensgebeurtenissen (bijv. verhuizing of overlijden) en/of (on)behandelde comorbiditeiten (bijv. epilepsie, hypothyreoïdie, vitamine-B12-tekort, gehoorproblemen, visusproblemen, slaapapneu) en/of recente wijzigingen in psychofarmacagebruik uitgesloten. Ook de afwezigheid van een informant die in staat was om veranderingen in de laatste zes maanden t.o.v. het karakteristieke functioneren/gedrag van de persoon met (Z)EV(M)B te beschrijven, was een reden voor exclusie. Na selectie

werden de informatiebrief en het toestemmingsformulier verstuurd naar wettelijk vertegenwoordigers van potentiële deelnemers.

Dementiediagnostiek
Voorafgaand aan dit onderzoek zijn deelnemers ingedeeld in drie groepen, namelijk 1) geen dementie, 2) twijfelachtige dementie, d.w.z. dat er sprake was van achteruitgang, maar nog geen klinische diagnose was gesteld en 3) klinisch gediagnosticeerde dementie. De indeling was gebaseerd op multidisciplinaire klinische beoordeling van ervaren artsen en gedragskundigen en informatie uit het cliëntdossier. Bij deelnemers is vooraf geen nieuw dementieonderzoek afgenomen. De diagnose en indeling bij een van de drie groepen vond van tevoren plaats en was niet gebaseerd op de uitkomsten van het diagnostisch hulpmiddel.

Interviewers
Het diagnostisch hulpmiddel is door middel van een interview met informanten ingevuld door een gedragskundige (orthopedagoog/psycholoog) of psychodiagnostisch medewerker, met ervaring in het afnemen van evaluatieschalen en werkzaam binnen de vier betrokken zorgorganisaties. Alle interviewers ontvingen voorafgaand aan de praktijktoets de handleiding met uitleg over de procedure, de scoring en het gebruik van het digitale systeem. Hierna volgden de interviewers een instructiemoment waarin de in de handleiding beschreven stappen nader zijn toegelicht en onduidelijkheden zijn opgehelderd. Ook werd de mogelijkheid geboden om het digitaal invullen van het diagnostisch hulpmiddel te oefenen.

Informanten
Interviews zijn afgenomen met ten minste één, maar bij voorkeur twee informanten, zoals een familielid of begeleider van de woon- of dagbestedingslocatie. Bij het plannen van de afname checkte de interviewer of informanten in staat waren om veranderingen in de laatste zes maanden t.o.v. van het karakteristieke functioneren/gedrag van de persoon met (Z)EV(M)B te beschrijven. In de week voor het interview ontvingen informanten per mail informatie over de procedure. Interviews zijn afgenomen in afwezigheid van de persoon met (Z)EV(M)B. Voorafgaand aan ieder interview zijn het doel, de opzet en het scoringssysteem door de interviewer uitgelegd aan de informant(en). Vervolgens zijn de vooraf op basis van het cliëntdossier ingevulde algemene gegevens doorgenomen

om te bepalen of deze klopten. Daarna is ieder item voorgelegd aan de informant(en), waarbij de symptoomdomeinen en itemvolgorde is gevolgd. Wanneer informanten een verschillend antwoord gaven, dan zorgde de interviewer ervoor dat de informanten tijdens het interview tot overeenstemming kwamen. Ook zijn items nader toegelicht door de interviewer wanneer deze onduidelijk waren voor de informant(en). Aan het eind van het interview is aan informanten gevraagd wat hun algemene indruk was van het diagnostisch hulpmiddel, of de uitleg/voorbeelden duidelijk waren, of zij uitleg/voorbeelden misten en of zij suggesties voor aanpassingen hadden. Deze vragen zijn na het interview ook ingevuld door de interviewer.

Kwaliteit van de data

Alle ingevulde interviews zijn gecontroleerd op inclusie-/exclusiecriteria en naleving van de instructies in de handleiding, waaronder de scoring. Scores zijn zo nodig in overeenstemming met de handleiding gecorrigeerd. Bij onduidelijkheden is navraag gedaan bij de interviewer. Bij blijvende onduidelijkheden is het desbetreffende item voor de deelnemer geëxcludeerd.

Validiteit

De indruks- en inhoudsvaliditeit zijn gewaarborgd in het ontwikkelproces. De items in het diagnostisch hulpmiddel zijn ontwikkeld op basis van triangulatie van resultaten uit gedegen onderzoek naar dementie bij mensen met (Z)EV(M)B waarbij verschillende kwalitatieve en kwantitatieve onderzoeksmethodes zijn gebruikt (Dekker, Wissing, et al., 2021b, 2021a; Wissing, Dijkstra, et al., 2022; Wissing, Fokkens, et al., 2022; Wissing, Hobbelen et al., 2023; Wissing, Ulgiati, et al., 2022). Niet alleen achteruitgang in cognitie, de voornaamste indicator voor dementie in de algemene bevolking (American Psychiatric Association & Nederlandse Vereniging voor Psychiatrie, 2014; Nederlandse Vereniging voor Klinische Geriatrie, 2014; Wereldgezondheidsorganisatie, 2014), maar ook veranderingen in andere domeinen, namelijk ADL, gedrag, motoriek en bijkomende gezondheidsproblemen, zijn meegenomen in de ontwikkeling van het diagnostisch hulpmiddel. Items zijn ontwikkeld op basis van zowel wetenschappelijke literatuur als praktijkervaringen. De validiteit is verder versterkt door de uitkomsten van het diagnostisch hulpmiddel te vergelijken met het klinische oordeel (gouden standaard). Dit geeft inzicht in het discriminerend vermogen van het nieuwe diagnostisch hulpmiddel.

Betrouwbaarheid
De interbeoordelaarsbetrouwbaarheid is verkend bij een subgroep van de deelnemers: twee zonder dementie en drie met klinische diagnose dementie. Bij vijf interviews met ieder een andere interviewer was aanvullend ook een onderzoeker (MBGW) aanwezig die de deelnemer niet kende en geen actieve rol in het interview vervulde. Zowel de interviewer als de onderzoeker vulde het diagnostisch hulpmiddel in aan de hand van de antwoorden van de informant(en), waarbij ze elkaars scores niet konden zien. De interbeoordelaarsbetrouwbaarheid is berekend door het overeenkomstpercentage (aantal overeenkomstig gescoorde items/totaal aantal gescoorde items x 100%) te bepalen voor zowel de afzonderlijke symptoomdomeinen als het gehele diagnostisch hulpmiddel (Gisev et al., 2013).

Verder is de interne consistentie bepaald door Cronbachs alfa te berekenen voor alle items gezamenlijk. Ook is gekeken naar veranderingen in Cronbachs alfa na verwijdering van individuele items. Deelnemers met geëxcludeerde itemscores (n=3) zijn niet meegenomen in de bepaling van Cronbachs alfa. Items die gescoord zijn als *'niet van toepassing'* of '*onbekend'* zijn behandeld als ontbrekende waardes, zonder daarbij de persoon te excluderen in de berekeningen (Holman et al., 2004). Cronbachs alfa-coëfficiënten zijn als volgt geïnterpreteerd: <0,50 (onacceptabel), 0,50-0,60 (slecht), 0,60-0,70 (twijfelachtig), 0,70-0,80 (acceptabel) en >0,80 (goed) (Nunnally, 1978). Berekeningen voor zowel de interbeoordelaarsbetrouwbaarheid als Cronbachs alfa zijn uitgevoerd in Microsoft Excel.

Data-analyse
Data zijn aan de hand van de handleiding gecontroleerd en gecorrigeerd en vervolgens geanalyseerd in SPSS Statistics versie 28 (IBM, Corp). Ten aanzien van karakteristieken van deelnemers en informanten zijn verschillen tussen groepen (Tabel 7.3) voor de niet-normaal verdeelde continue data (leeftijd) bepaald met de Kruskal-Wallistoets en voor categorische data met chikwadraattoetsen.

Met betrekking tot het discriminerend vermogen is gekeken naar de mate waarin veranderingen werden gescoord in relatie tot de dementiestatus. Gezien het exploratieve karakter van de eerste praktijktoets is gefocust op trends. Ter aanvulling zijn er statistische toetsen uitgevoerd. Allereerst

berekende we per item voor iedere groep het percentage deelnemers dat een verandering (afname of toename) scoorde (score=1) en het percentage deelnemers dat geen verandering scoorde (score=0). Om itemscores tussen groepen te vergelijken, zijn chikwadraattoetsen gebruikt. De antwoorden *'niet van toepassing,' 'onbekend'* en geëxcludeerde itemscores zijn in de analyses behandeld als ontbrekende waardes. In het kader relevantie van items in de praktijk zijn items geïdentificeerd die niet of nauwelijks veranderen in relatie tot dementie. Dit zijn items waarvoor ≥85% van de deelnemers in de groepen twijfelachtige dementie én diagnose dementie geen verandering scoorde (Dekker, Ulgiati, et al., 2021b; Dekker, Vermeiren et al., 2018). Vervolgens zijn het aantal veranderingen per domein, het totale aantal veranderingen en de totale procentuele veranderingsscore (totale veranderingsscore/totaal aantal items waarop verandering gescoord had kunnen worden x 100%) berekend. Voor het vergelijken van de domein- en totaalscores tussen de groepen zijn Kruskal-Wallistoetsen gebruikt. Wanneer een statistisch significant verschil tussen groepen gevonden werd, dan zijn Bonferroni-Dunn's post-hoc toetsen voor meervoudige vergelijkingen uitgevoerd om nader te bepalen welke groepen van elkaar verschilden. Voor alle statistische testen zijn *p*-waardes lager dan 0,05 als statistisch significant beschouwd.

Tot slot zijn ervaringen van interviewers en informanten in kaart gebracht door antwoorden op de vragen over de algemene indruk van het diagnostisch hulpmiddel, duidelijkheid van uitleg/voorbeelden, suggesties voor aanvullende uitleg en verdere aanpassingen te analyseren. Illustratieve citaten zijn aan de resultatensectie toegevoegd om ervaringen van interviewers en informanten te verduidelijken (Elo & Kyngäs, 2008).

Resultaten

Ontwikkeling diagnostisch hulpmiddel

Het diagnostisch hulpmiddel bestaat uit 45 items, onderverdeeld in 7 symptoomdomeinen: 1) veranderingen in cognitie, 2) veranderingen in taal en spraak, 3) gedragsveranderingen, 4) veranderingen in eten en drinken, 5) veranderingen in persoonlijke verzorging, 6) veranderingen in motoriek en 7) bijkomende gezondheidsproblemen (Tabel 7.2). De eerder gehanteerde indeling in vijf symptoomdomeinen is naar aanleiding van onderzoeksbevindingen en feedback van gedragskundigen en

psychodiagnostisch medewerkers aangepast. De wijze waarop mensen met (Z)EV(M)B communiceren, is voornamelijk non-verbaal (van der Putten et al., 2017). Om die reden vormen items over veranderingen in taal en spraak een apart domein, dat kan worden overgeslagen indien een persoon niet verbale vaardigheden heeft getoond. Voor items over loopvaardigheden zou hetzelfde kunnen gelden, maar gezien de samenhang met andere motorische veranderingen, zoals verminderde balans en vaker vallen (Ries, 2018), zijn deze items ingedeeld in het motorische domein. Tot slot heeft zowel het advies van gedragskundigen en psychodiagnostisch medewerkers als de indeling van symptomen in de contexten verzorging en eten/drinken in het focusgroeponderzoek (Dekker, Wissing, et al., 2021b) ertoe geleid dat het ADL-domein is opgesplitst in twee domeinen: veranderingen in eten en drinken en veranderingen in persoonlijke verzorging.

Tabel 7.2 Getoetste versie van het diagnostisch hulpmiddel dementie bij mensen met (Z)EV(M)B voor gedragskundigen en psychodiagnostisch medewerkers.

Symptoomdomeinen en items	
Veranderingen in cognitie	
1.1	Dagelijkse handelingen herkennen
1.2	Eenvoudige keuzes maken
1.3	Personen herkennen
1.4	Objecten herkennen
1.5	Voorkeur voor objecten
1.6	Objecten kwijtraken
1.7	Afstanden waarnemen
1.8	Weg vinden
1.9	Dagritme herkennen
1.10	Dag en nacht herkennen
Veranderingen in taal en spraak	
2.1	Woordgebruik
2.2	Verstaanbaar spreken
Gedragsveranderingen	
3.1	Angst
3.2	Paniek
3.3	Verdriet
3.4	Interesse voor directe leefomgeving
3.5	Zichzelf terugtrekken
3.6	's Nachts wakker worden
3.7	Overdag slapen
3.8	Prikkelbaar gedrag
3.9	Verzetten tegen benodigde hulp
3.10	Fysieke agressie
3.11	Rusteloos gedrag
3.12	Stereotiep gedrag
3.13	Dwangmatig gedrag
3.14	Ontremd gedrag
3.15	Stemmingswisselingen
3.16	Hallucinaties/wanen
Veranderingen in eten en drinken	
4.1	Eet-/drinkvaardigheden
4.2	Eetlust/drinken
4.3	Verslikken
4.4	Kauwen
4.5	Lichaamsgewicht
Veranderingen in persoonlijke verzorging	
5.1	Verzorging
Veranderingen in motoriek	
6.1	Mobiliteit/transfers
6.2	Balans
6.3	Valfrequentie
6.4	Rolstoelgebruik
6.5	Stijfheid
6.6	Spierkracht
6.7	Spierverkrampingen
6.8	Looppatroon
6.9	Loopafstand
Bijkomende gezondheidsproblemen	
7.1	Epilepsie
7.2	Incontinentie

Items over taal en spraak (item 2.1 en 2.2) en loopvaardigheden (item 6.8 en 6.9) worden alleen aangeboden en ingevuld wanneer de persoon deze vaardigheden karakteristiek heeft getoond, dus voordat er sprake was van achteruitgang.

Praktijktoets diagnostisch hulpmiddel

Beschrijving studiepopulatie
Wettelijk vertegenwoordigers van 56 mensen met (Z)EV(M)B hebben een informatiebrief met toestemmingsformulier ontvangen. Hieruit volgde toestemming voor deelname van 39 mensen en geen toestemming voor 6 mensen. Voor 11 mensen is geen reactie ontvangen. Op basis van exclusiecriteria zijn naderhand 3 interviews uitgesloten: 2 vanwege onbehandelde comorbiditeiten met effect op het functioneren, 1 vanwege de afwezigheid van ten minste één informant die in staat was om de veranderingen in de laatste zes maanden t.o.v. het karakteristieke functioneren/gedrag van de persoon te beschrijven.

Tabel 7.3A toont de beschrijvende gegevens van de 36 geïncludeerde mensen met (Z)EV(M)B apart voor de drie groepen: geen dementie, twijfelachtige dementie en diagnose dementie. Met betrekking tot aanwezigheid van een syndroom is tussen de groepen een statistisch significant verschil gevonden (p=0,021). Van de mensen zonder dementie had 11,1% DS, terwijl dit respectievelijk 40,0% en 62,5% was voor de groepen met twijfelachtige en diagnose dementie. Verder bezochten significant minder mensen met twijfelachtige (60,0%) en diagnose dementie dagbesteding (62,5%) in vergelijking met degenen zonder dementie (100,0%) (p=0,003). Tussen de groepen werd een significant verschil gevonden in het aantal aanwezige informanten (p=0,039, zie Tabel 7.3B). Voor mensen met twijfelachtige of diagnose dementie waren dat er een of twee; voor mensen zonder dementie in enkele gevallen drie.

Betrouwbaarheid
Aan de hand van vijf interviews (13,9% van totaal aantal interviews) is de interbeoordelaarsbetrouwbaarheid verkend. Deze bleek hoog voor de symptoomdomeinen: 96,0% voor cognitie, 100,0% voor taal en spraak, 96,3% voor gedrag, 100,0% voor eten en drinken, 80,0% voor persoonlijke verzorging, 84,4% voor motoriek en 100,0% voor bijkomende gezondheidsproblemen. Voor het gehele diagnostisch hulpmiddel kwam de interbeoordelaarsbetrouwbaarheid uit op 94,0%.

De samenhang tussen items (interne consistentie) was goed, aangezien de Cronbachs alfa voor alle items tezamen 0,892 was (Nunnally, 1978). Bij verwijdering van individuele items uit de lijst varieerde de alfa tussen

0,884 en 0,896. Verwijdering van individuele items zou niet tot een noemenswaardige verbetering van de interne consistentie leiden.

Tabel 7.3 Kenmerken van deelnemers en informanten.

A: Kenmerken van deelnemers	Geen dementie n=18	Twijfelachtige dementie n=10	Diagnose dementie n=8	p
Leeftijd (jaren, mediaan (IQR), min. – max.)	61,5 (17,8), 39,0 – 84,0	63,5 (28,0), 53,0 – 81,0	58,5 (10,8), 46,0 – 65,0	0,124
Geslacht (% vrouw)	55,6	50,0	50,0	0,946
Mate VB (baseline): ernstig; zeer ernstig (%)	50,0; 50,0	30,0; 70,0	37,5; 62,5	0,564
Aanwezigheid syndroom: DS; ander genetisch syndroom; nee (%)	11,1; 0,0; 88,9	40,0; 0,0; 60,0	62,5; 0,0; 37,5	0,021*
IQ-score beschikbaar (%)	0,0	10,0	0,0	0,267
Sociaal-emotioneel functioneren 0-6; 6-18; 18-36 maanden; onbekend (%)	16,7; 27,8; 0,0; 55,6	10,0; 30,0; 0,0; 60,0	25,0; 12,5; 12,5; 50,0	0,616
Verbale vaardigheden (karakteristiek): ja (%)	33,3	50,0	37,5	0,687
Loopvaardigheden (karakteristiek): ja (%)	83,3	90,0	100,0	0,302
Wonend bij zorgorganisatie (%)	100,0	100,0	100,0	-
Dagbesteding (%)	100,0	60,0	62,5	0,003*
B: Kenmerken van informanten	**n=34**	**n=14**	**n=12**	
Informanten per participant (% n=1; n=2; n=3)	44,4; 22,2; 33,3	60,0; 40,0; 0,0	50,0; 50,0; 0,0	0,039*
Geslacht (% vrouw)	79,4	92,9	83,3	0,478
Rol: begeleider wonen; begeleider dagbesteding; familielid; mentor maar geen familie (%)	52,9; 2,9; 41,2; 2,9	85,7; 7,1; 7,1; 0,0	58,3; 16,7; 25,0; 0,0	0,132
Aantal jaar bekend met deelnemer (% <2; 2-10; 10-20; >20 jaar; onbekend)	8,8; 35,3; 8,8; 32,4; 14,7	14,3; 57,1; 7,1; 21,4; 0,0	16,7; 25,0; 16,7; 41,7	0,738
Uren per week bij deelnemer (% <10; 10-20; >20 uur; onbekend)	32,4; 26,5; 26,5; 14,7	14,3; 21,4; 64,3; 0,0	25,0; 50,0; 16,7; 8,3	0,096

Baseline betreft het hoogste niveau van functioneren voordat er sprake was van eventuele achteruitgang. Om verschillen tussen groepen te vergelijken, is de Kruskal-Wallistoets gebruikt voor niet normaal verdeelde continue data (leeftijd) en zijn chikwadraattoetsen gebruikt voor categorische data. Symbool: *, p<0,05. Afkortingen: DS, downsyndroom; IQ, intelligentiequotiënt; VB, verstandelijke beperking.

Discriminerend vermogen: itemscores

Figuren 7.2-7.6 tonen voor ieder item per groep (geen dementie, twijfelachtige dementie en diagnose dementie) het percentage deelnemers dat een afname, toename of geen verandering liet zien. Bovendien is het percentage deelnemers weergeven voor wie *'niet van toepassing'/'onbekend'* is gescoord of een itemscore is geëxcludeerd. De

figuren laten voor een aanzienlijke deel van de items een duidelijke trend zien: het percentage deelnemers dat een verandering scoorde (afname, dan wel toename) was hoger in de groep met twijfelachtige dementie en het hoogst in de groep met een diagnose dementie. Voor een aantal items verschilde de itemscores bovendien statistisch significant tussen de drie groepen. Deze zijn hieronder per domein toegelicht. Er werden geen significante verschillen gevonden voor de items in het domein bijkomende gezondheidsproblemen (Figuur 7.6).

Veranderingen in cognitie
Statistisch significante verschillen werden gevonden voor de volgende cognitieve items: dagelijkse handelingen herkennen (1.1; p=0,004), voorkeur voor objecten (1.5; p=0,030), weg vinden (1.8; p=0,039) en dagritme herkennen (1.9; p=0,007). Meer moeite met het herkennen van dagelijkse handelingen en verminderde voorkeur voor objecten werden het meest gescoord voor de diagnose dementie-groep en het minst in de groep zonder dementie. Zowel voor item 1.8 als 1.9 werd een achteruitgang gescoord voor deelnemers met twijfelachtige dementie, in mindere mate ook voor degenen met een diagnose dementie, maar niet wanneer er geen sprake van dementie was (Figuur 7.2).

Veranderingen in taal en spraak
De items over taal en spraak waren binnen iedere groep voor (meer dan) de helft van de deelnemers niet ingevuld aangezien zij niet verbale vaardigheden hebben gehad. Desalniettemin werd een statistisch significant verschil tussen de drie groepen gevonden voor verstaanbaar spreken (2.2; p=0,045). Met name deelnemers met een diagnose dementie, maar ook degenen met twijfelachtige dementie spraken minder verstaanbaar, terwijl dit niet het geval was in de groep zonder dementie (Figuur 7.2).

Gedragsveranderingen
Groepen verschilden statistisch significant voor de items: 's nachts wakker worden (3.6; p=0,004), prikkelbaar gedrag (3.8; p=0,029), rusteloos gedrag (3.11; p=0,014) en ontremd gedrag (3.14; p=0,047). Veranderingen in 's nachts wakker, prikkelbaar gedrag en ontremd gedrag werden met name gescoord voor deelnemers met een diagnose dementie, gevolgd door deelnemers met twijfelachtige dementie en beperkt bij degenen zonder dementie. Een toename in rusteloosheid werd hoofdzakelijk gescoord voor

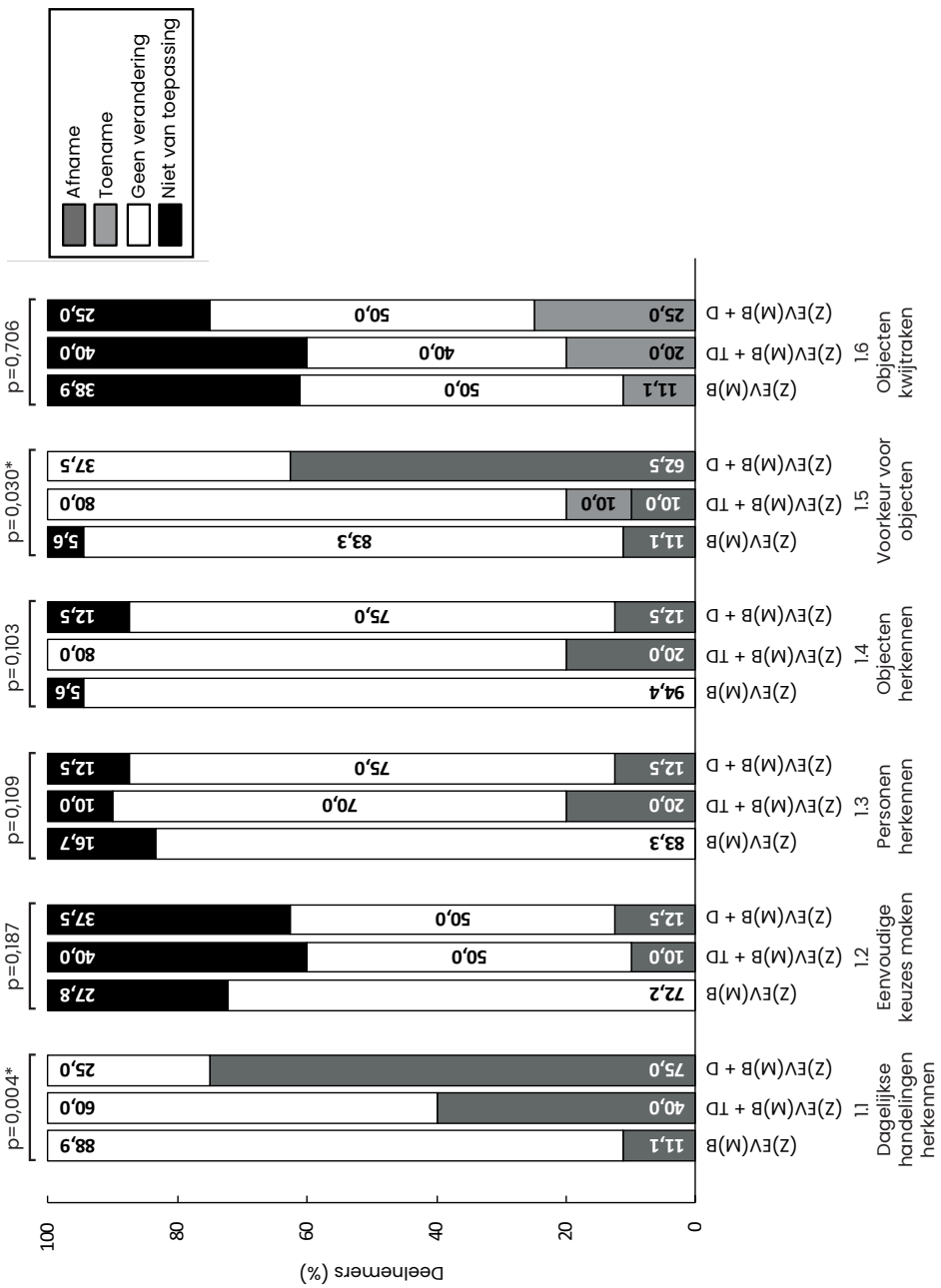
de groep met een diagnose dementie en beperkt voor de groepen met twijfelachtige of geen dementie (Figuur 7.3).

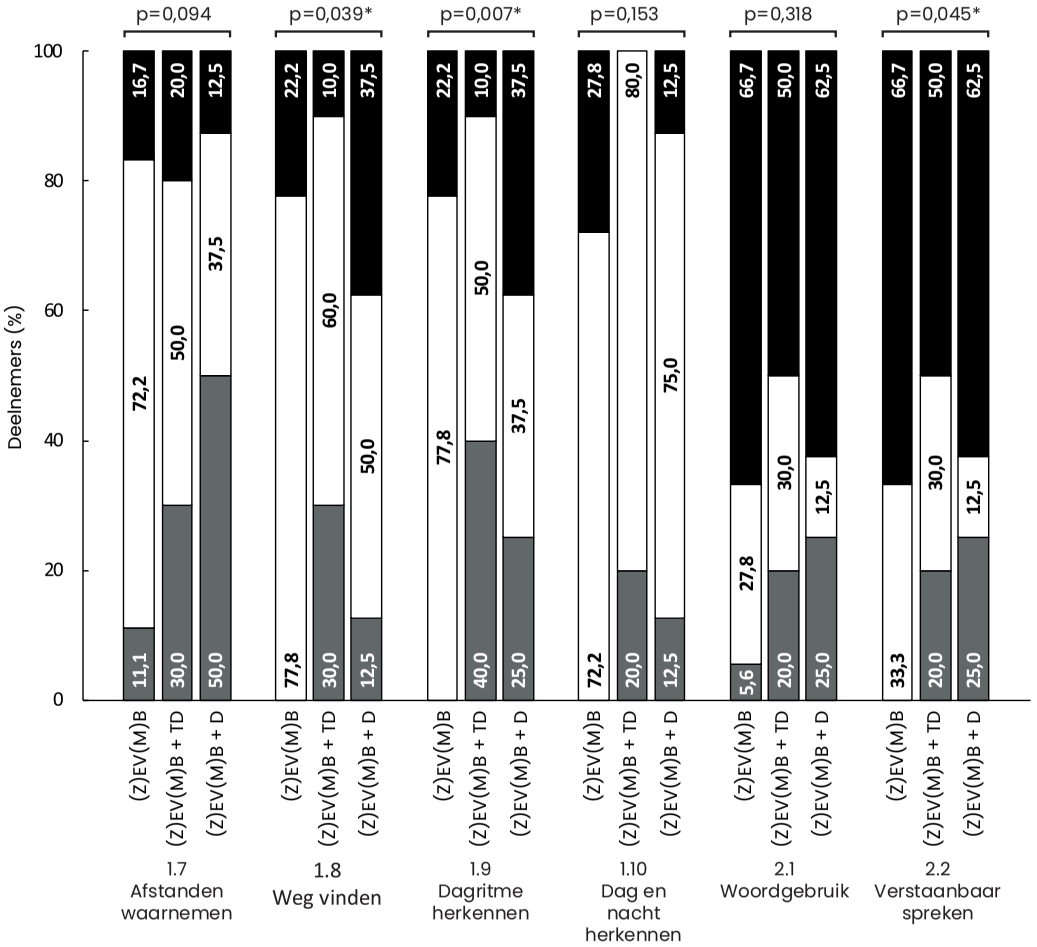
Veranderingen in eten en drinken
Binnen dit domein zijn statistisch significante verschillen gevonden voor eet- en drinkvaardigheden (4.1; p=0,022) en kauwen (4.4; p=0,019). Achteruitgang werd met name gescoord bij deelnemers met twijfelachtige of een diagnose dementie en beperkt bij degenen zonder dementie (Figuur 7.4).

Veranderingen in persoonlijke verzorging
Groepen verschilden statistisch significant met betrekking tot het item verzorging (5.1; p=0,012). Ook hier werd een achteruitgang met name bij degenen met twijfelachtige en een diagnose dementie gescoord en nauwelijks bij de groep zonder dementie (Figuur 7.4).

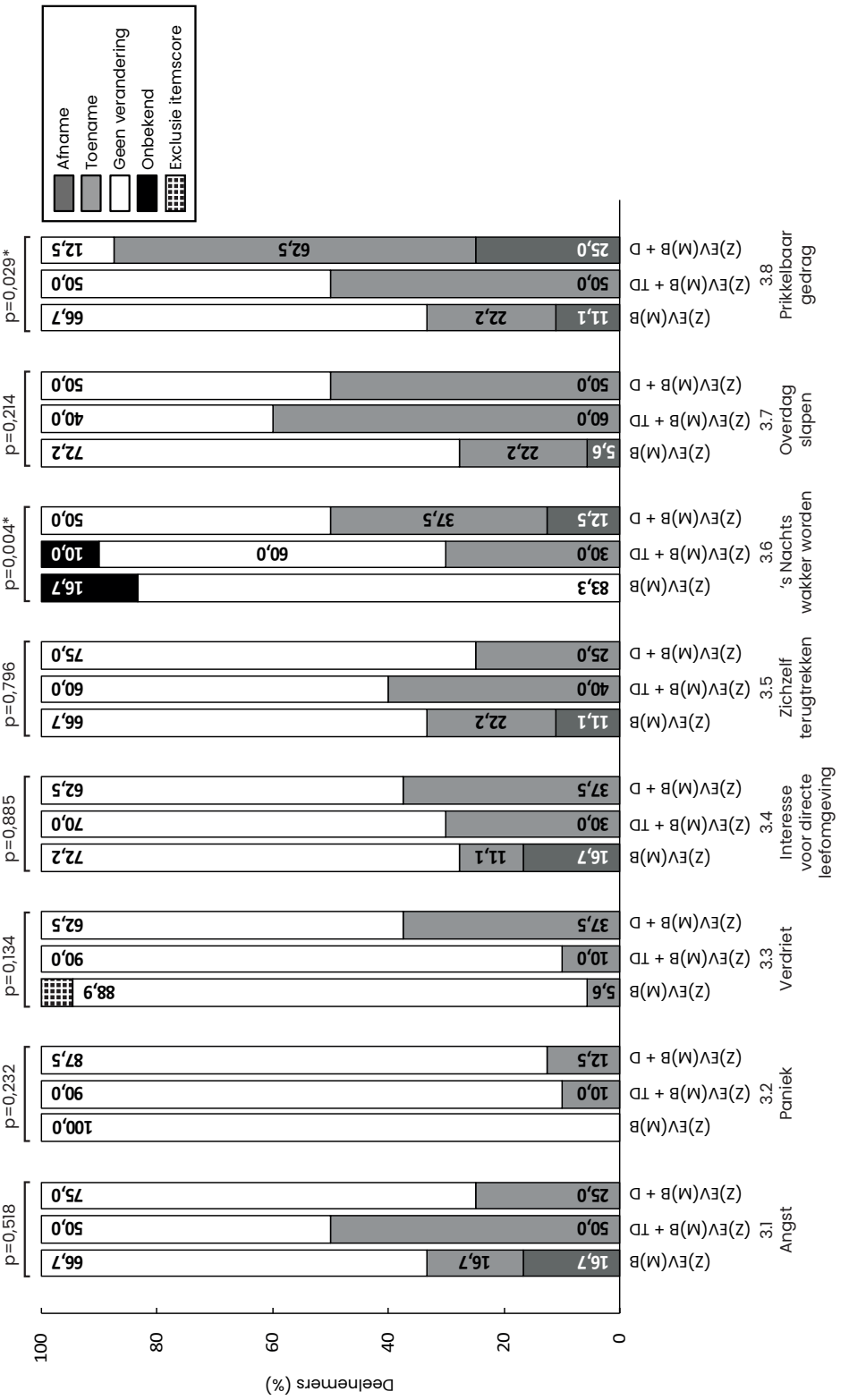
Veranderingen in motoriek
De volgende motorische items verschilden significant tussen de drie groepen: valfrequentie (6.3; p=0,029) en looppatroon (6.8; p=0,033). Voor beide items gold dat een verandering met name gescoord werd voor deelnemers met diagnose dementie, gevolgd door degenen met twijfelachtige dementie en beperkt voor degenen zonder dementie (Figuur 7.5).

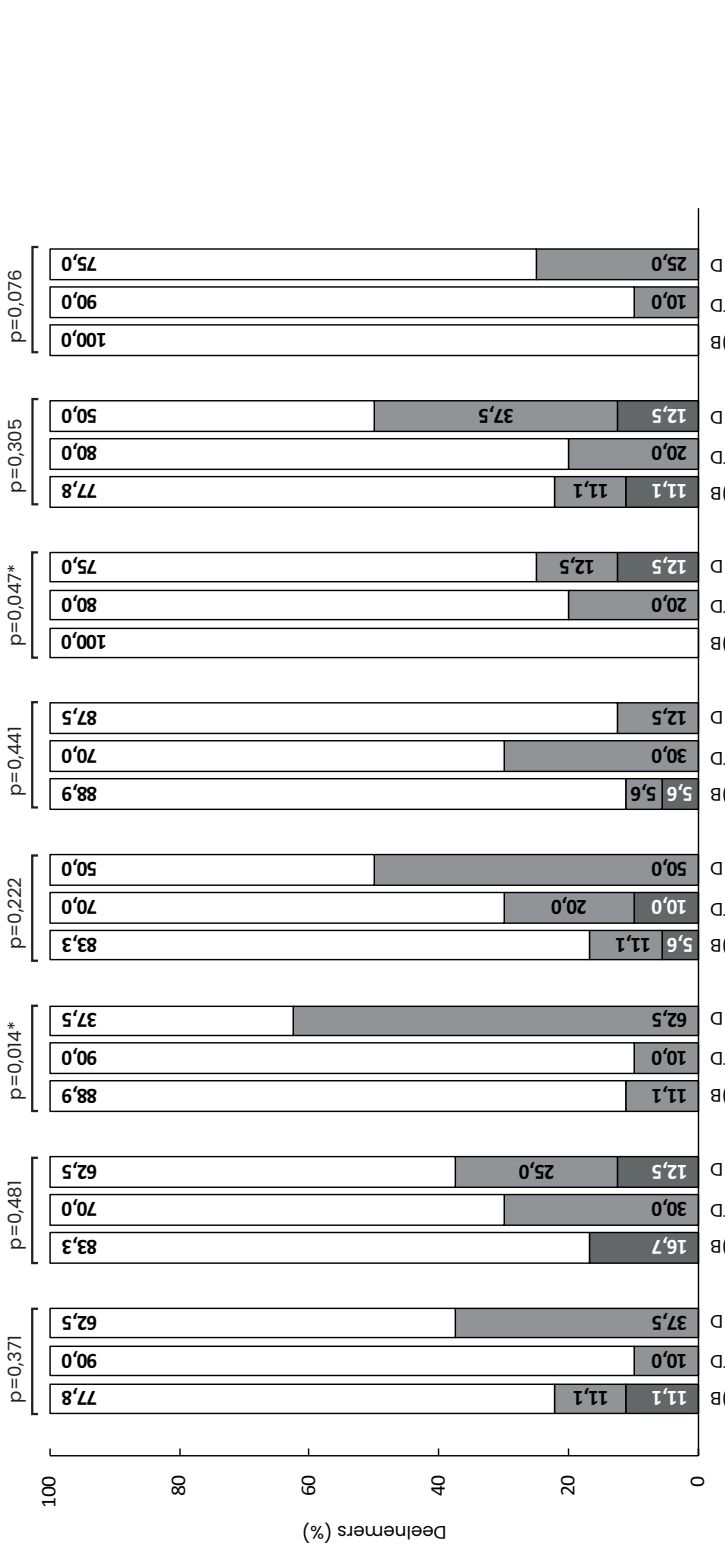
Discriminerend vermogen: minder relevante items
Drie items voldeden aan het criterium dat ≥85% van de deelnemers in de groepen met twijfelachtige én diagnose dementie geen verandering vertoonden. Voor item 3.2 paniek was een verandering afwezig bij 90,0% in de twijfelachtige dementie-groep en bij 87,5% in de diagnose dementie-groep (Figuur 7.3). Een verandering was afwezig bij 100,0% met twijfelachtige dementie en bij 87,5% met diagnose dementie ten aanzien van item 6.7 spierverkrampingen (Figuur 7.5). Voor item 7.1 epilepsie gold dat bij 90,0% met twijfelachtige dementie en 100,0% met diagnose dementie verandering afwezig was (Figuur 7.6).


Afname
Toename
Geen verandering
Niet van toepassing
Deelnemers (%)
0
20
40
60
80
100
1.1 Dagelijkse handelingen herkennen
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
11,1
40,0
75,0
88,9
60,0
25,0
p=0,004*
1.2 Eenvoudige keuzes maken
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
72,2
10,0
12,5
27,8
40,0
37,5
p=0,187
1.3 Personen herkennen
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
83,3
20,0
12,5
16,7
10,0
12,5
p=0,109
1.4 Objecten herkennen
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
94,4
20,0
12,5
5,6
80,0
75,0
p=0,103
1.5 Voorkeur voor objecten
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
11,1
10,0
62,5
83,3
10,0
37,5
5,6
80,0
p=0,030*
1.6 Objecten kwijtraken
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
11,1
20,0
25,0
50,0
40,0
50,0
38,9
40,0
25,0
p=0,706

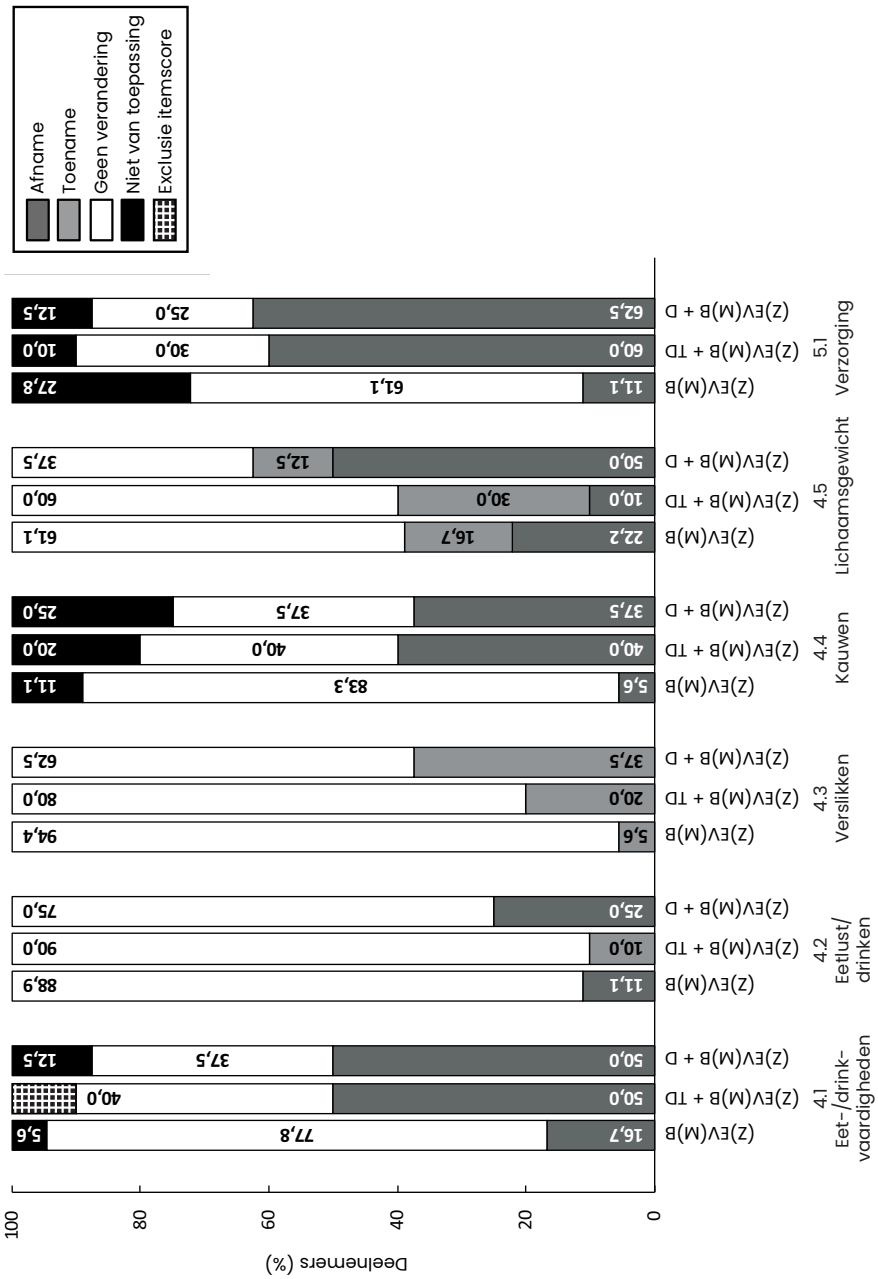


Figuur 7.2 Veranderingen in 1. cognitie en 2. taal en spraak per groep: geen dementie ((Z)EV(M)B), twijfelachtige dementie ((Z)EV(M)B + TD), diagnose dementie ((Z)EV(M)B + D). Chikwadraattoetsen zijn gebruikt voor het identificeren van verschillen tussen groepen. Symbool: *, p<0,05. Afkorting: (Z)EV(M)B, (zeer) ernstige verstandelijke (en meervoudige) beperkingen.

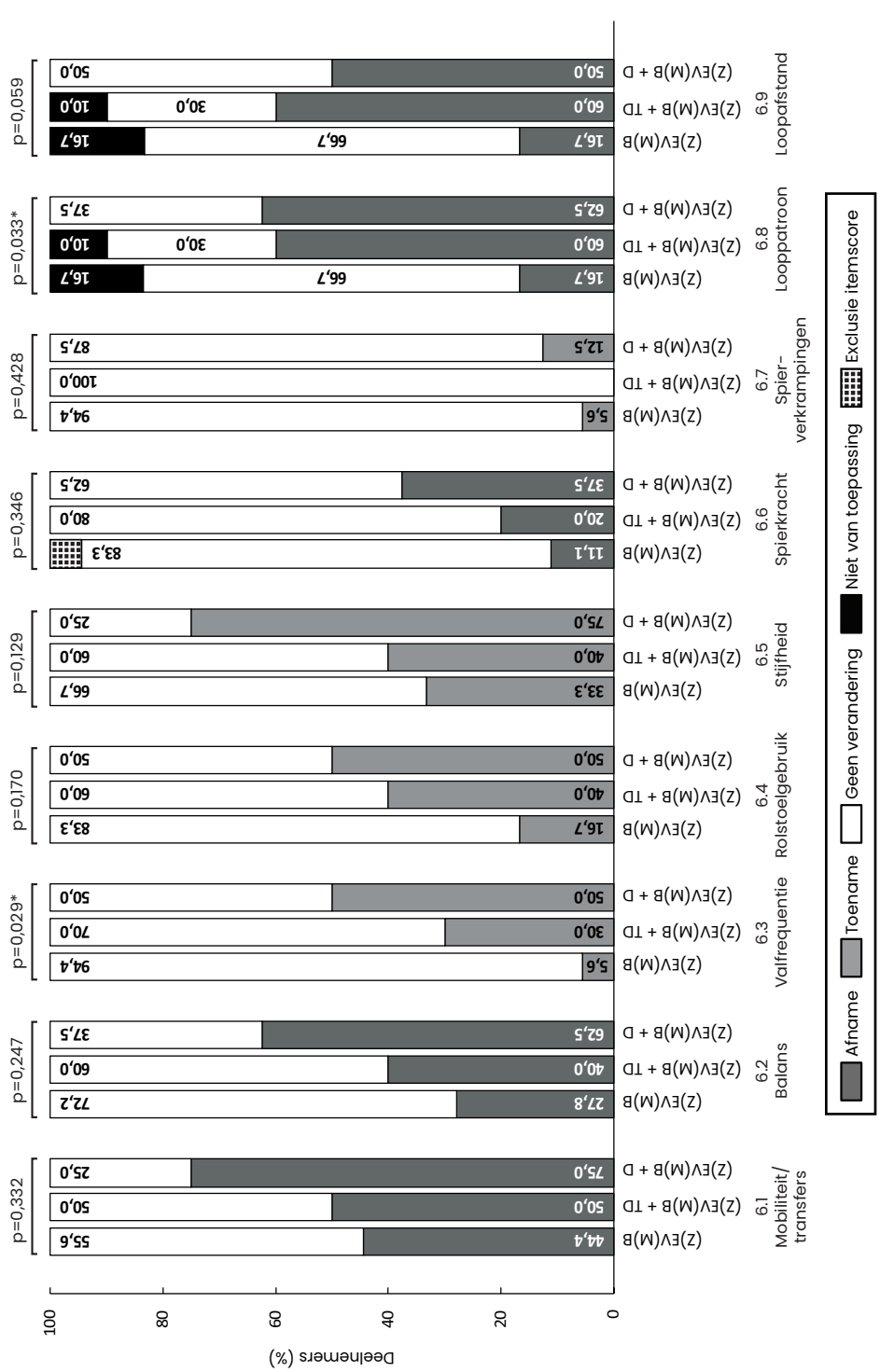

Afname
Toename
Geen verandering
Onbekend
Exclusie itemscore
Deelnemers (%)
0
20
40
60
80
100
3.1 Angst
3.2 Paniek
3.3 Verdriet
3.4 Interesse voor directe leefomgeving
3.5 Zichzelf terugtrekken
3.6 's Nachts wakker worden
3.7 Overdag slapen
3.8 Prikkelbaar gedrag
(Z)EV(M)B
(Z)EV(M)B + TD
(Z)EV(M)B + D
p=0,518
p=0,232
p=0,134
p=0,885
p=0,796
p=0,004*
p=0,214
p=0,029*
16,7
16,7
66,7
50,0
50,0
75,0
100,0
10,0
90,0
87,5
5,6
88,9
10,0
90,0
62,5
16,7
11,1
72,2
30,0
70,0
62,5
11,1
22,2
66,7
40,0
60,0
75,0
83,3
16,7
30,0
60,0
10,0
12,5
37,5
50,0
5,6
22,2
72,2
60,0
40,0
50,0
11,1
22,2
66,7
50,0
50,0
25,0
62,5
12,5



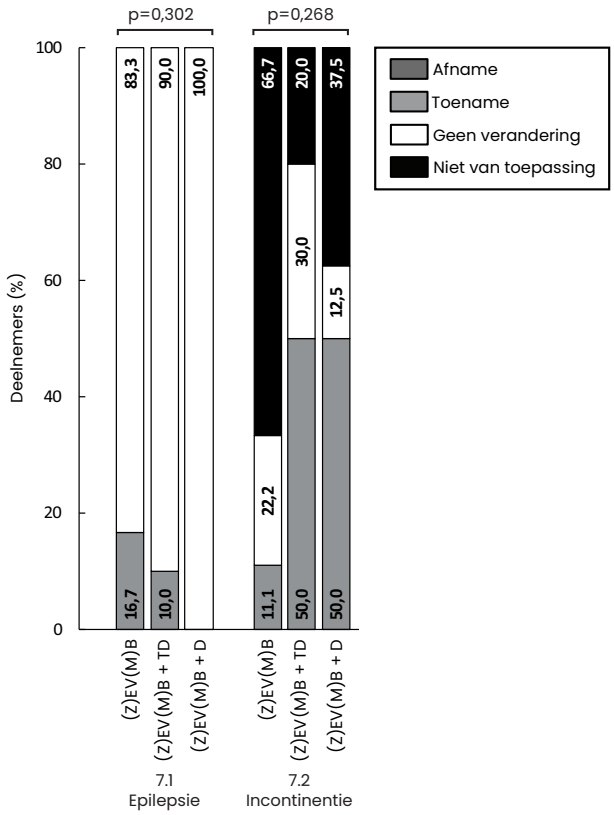
Figuur 7.3 Gedragsveranderingen per groep: geen dementie ((Z)EV(M)B), twijfelachtige dementie ((Z)EV(M)B + TD), diagnose dementie ((Z)EV(M)B + D). Chikwadraattoetsen zijn gebruikt voor het identificeren van verschillen tussen groepen. Symbool: *, p<0,05. Afkorting: (Z)EV(M)B, (zeer) ernstige verstandelijke (en meervoudige) beperkingen.



Figuur 7.4 Veranderingen in 4. eten en drinken en 5. persoonlijke verzorging per groep: geen dementie ((Z)EV(M)B), twijfelachtige dementie ((Z)EV(M)B + TD), diagnose dementie ((Z)EV(M)B + D). Chikwadraattoetsen zijn gebruikt voor het identificeren van verschillen tussen groepen. Symbool: *, p<0,05. Afkorting: (Z)EV(M)B, (zeer) ernstige verstandelijke (en meervoudige) beperkingen.



Figuur 7.5 Veranderingen in motoriek per groep: geen dementie ((Z)EV(M)B), twijfelachtige dementie ((Z)EV(M)B + TD), diagnose dementie ((Z)EV(M)B + D). Chikwadraattoetsen zijn gebruikt voor het identificeren van verschillen tussen groepen. Symbool: *, p<0,05. Afkorting: (Z)EV(M)B, (zeer) ernstige verstandelijke (en meervoudige) beperkingen.



Figuur 7.6 Bijkomende gezondheidsproblemen per groep: geen dementie ((Z)EV(M)B), twijfelachtige dementie ((Z)EV(M)B + TD), diagnose dementie ((Z)EV(M)B + D). Chikwadraattoetsen zijn gebruikt voor het identificeren van verschillen tussen groepen. Afkorting: (Z)EV(M)B, (zeer) ernstige verstandelijke (en meervoudige) beperkingen.

Discriminerend vermogen: domein- en totaalscores

Het aantal veranderingen per domein en de totaalscores zijn apart voor iedere groep weergegeven in Tabel 7.4. Statistisch significante verschillen tussen groepen werden gevonden voor veranderingsscores van vijf domeinen: cognitie, gedrag, eten en drinken, persoonlijke verzorging en motoriek. Post-hoc toetsen toonden dat de groepen met twijfelachtige en een diagnose dementie statistisch significant hogere cognitieve en persoonlijke verzorging domeinscores hadden dan de groep zonder dementie (alle p-waardes kleiner dan 0,05). Ten aanzien van de domeinen gedrag en motoriek scoorde alleen de groep met een diagnose dementie statistisch significant hoger dan de groep zonder dementie (p=0,016 en

p=0,024, respectievelijk). Voor het domein eten en drinken waren geen van de groepsvergelijkingen statistisch significant, al lag de p-waarde voor de vergelijking tussen geen dementie en diagnose dementie vlak boven 0,05 (p=0,059).

Met betrekking tot de totaalscores werd zowel voor de totale veranderingsscore (p=0,001) als de totale procentuele veranderingsscore (p=0,002) een statistisch significant verschil gevonden. Post- hoc toetsen lieten zien dat de totale veranderingsscore statistisch significant hoger was voor de groepen met twijfelachtige dementie en een diagnose dementie dan de groep zonder dementie. (p=0,032 en p=0,002, respectievelijk). De procentuele veranderingsscore was alleen statistisch significant hoger voor de groep met een diagnose dementie in vergelijking tot de groep zonder dementie. Het verschil tussen geen dementie en twijfelachtige dementie was net niet significant (p=0,054).

Tabel 7.4 Veranderingen per domein en totaalscores

Domein	Geen dementie n=18	Twijfelachtige dementie n=10	Diagnose dementie n=8	p
Cognitie	0 (1), 0 – 2	3 (3), 0 – 5	2 (3,5), 0 – 8	<0,001*
Taal en spraak	0 (0), 0 –1	0 (1), 0 – 2	0 (1,5), 0 – 2	0,186
Gedrag	2 (3,25), 0 – 7	5 (3,75), 0 – 10	6,5 (5,5), 3 – 10	0,015*
Eten en drinken	0 (1), 0 – 4	1,5 (3), 0 – 3	2 (2,75), 0 – 5	0,048*
Persoonlijke verzorging	0 (0), 0 – 1	1 (1), 0 – 1	1 (1), 0 – 1	0,009*
Motoriek	2 (3), 0 – 7	4 (6), 0 – 6	4,5 (4,75), 1 – 9	0,021*
Bijkomende gezondheidsproblemen	0 (0,25), 0 – 2	1 (1), 0 – 1	0,5 (1), 0 – 1	0,175
Totale veranderingsscore	5 (6,5), 0 – 13	14 (10,25), 0 – 26	19 (12,75), 5 – 33	0,001*
Totale procentuele veranderingsscore	12,2 (18,7), 0,0 – 32,4	31,4 (21,4), 0,0 – 61,9	44,4 (37,0), 11,1 – 75,0	0,002*

Aantal veranderingen per symptoomdomein, totale veranderingsscore en totale procentuele veranderingsscore (mediaan (interkwartielafstand), min.–max.) zijn weergegeven per groep. Kruskal-Wallistesten zijn gebruikt om verschillen tussen groepen te vergelijken. Symbool: *, p<0,05.

Ervaringen van interviewers en informanten

Interviewers gaven aan dat het diagnostisch hulpmiddel duidelijk, overzichtelijk en compleet is. Het omvat verschillende gebieden met betrekking tot dementie. Het diagnostisch hulpmiddel sluit volgens de interviewers aan bij de mensen met (Z)EV(M)B.

> ***Psychodiagnostisch medewerker:*** *''[Het diagnostisch hulpmiddel] sluit aan bij de doelgroep. Door uitvragen en langslopen van de hele lijst krijg je veel informatie die volgens mij dekkend is wat betreft dementieproblematiek.''*

Interviewers gaven aan dat er nog een aantal verbeterpunten zijn. Zo mogen er meer voorbeelden toegevoegd worden, met name voorbeelden van kleine observeerbare veranderingen. Verder werd gesuggereerd om eventueel aandacht te besteden aan hoe vaak de verandering zich voordoet.

> ***Gedragskundige:*** *''Een meerwaarde zou zijn om [in het diagnostisch hulpmiddel] meer aandacht te besteden aan hoe frequent iemand veranderingen laat zien.''*

Ook informanten waren overwegend positief over het diagnostisch hulpmiddel. Degenen die eerder informant waren geweest bij interviews met andere dementielijsten voor mensen met VB, gaven aan dat het diagnostisch hulpmiddel duidelijker is en beter aansluit bij mensen met (Z)EV(M)B.

> ***Informant 1:*** *''Sluit beter aan dan de [reeds] bestaande dementielijsten zoals de DVZ en DSVH. [Dit diagnostisch hulpmiddel] past beter bij zijn niveau van functioneren.''*

Verder bevat het diagnostisch hulpmiddel volgens informanten relevante vragen die aanzetten tot denken over mogelijke veranderingen op verschillende gebieden rondom dementie. Informanten gaven aan dat toelichting van de gedragskundige/psychodiagnostisch medewerker op vragen nodig was om deze te kunnen beantwoorden. Daarom vonden ze het goed dat de lijst in interviewvorm werd ingevuld.

> ***Informant 2:*** *''Passend [diagnostisch hulpmiddel], geeft globaal een goed beeld. Uitgebreide vragen maar voldoende specifiek en alles zit erin waar ik aan dacht bij de vraag of er sprake kan zijn van dementie.''*

De uitleg en voorbeelden waren volgens informanten duidelijk, maar een aantal gaf ook aan dat deze minder toegespitst zijn op personen met zeer ernstige verstandelijke en meervoudige beperkingenniveau. De ervaring was dat met name voor mensen met zeer ernstige verstandelijke en meervoudige beperkingen, die een zeer beperkt aantal vaardigheden hebben ontwikkeld, het lastiger blijft om het hulpmiddel in te vullen. De suggestie was om de voorbeelden nog verder aan te scherpen aan de hand van kleine veranderingen die zichtbaar zijn bij deze subgroep.

> ***Informant 3***: *''De voorbeelden mogen wat uitgebreider met betrekking tot personen die al bijna geen vaardigheden hebben.''*

Discussie

Dit artikel beschrijft de ontwikkeling en eerste praktijktoets van het diagnostisch hulpmiddel dementie bij mensen met (Z)EV(M)B voor gedragskundigen en psychodiagnostisch medewerkers. In totaal omvat de getoetste versie 45 items, onderverdeeld in 7 symptoomdomeinen, tot stand gekomen op basis van triangulatie van bevindingen in wetenschappelijke literatuur en praktijkervaringen. Hierdoor zijn de indruks- en inhoudsvaliditeit verzekerd. Vervolgens is het diagnostisch hulpmiddel in interviewvorm ingevuld voor 36 personen met (Z)EV(M)B met en zonder twijfelachtige dementie of diagnose dementie. De resultaten wijzen erop het diagnostisch hulpmiddel betrouwbaar is. In het kader van discriminerend vermogen zijn item-, domein-, en totaalscores vergeleken tussen de groepen zonder dementie, met twijfelachtige dementie en met een diagnose dementie. Voor een aanzienlijk deel van de items werd een verandering (afname, dan wel toename) het vaakst gescoord in de groep met een diagnose dementie en het minst in de groep zonder dementie. Verschillen in domeinscores werden gevonden voor vijf van de zeven domeinen, namelijk cognitie, gedrag, eten en drinken, persoonlijke verzorging en motoriek. Ook werden verschillen tussen groepen gevonden ten aanzien van de totaalscores. Voor zowel domein- als totaalscores hadden met name mensen met een diagnose dementie hogere scores in vergelijking tot degenen zonder dementie. Tezamen geeft dit een eerste indicatie dat het diagnostisch hulpmiddel voldoende discriminerend vermogen heeft. Tot slot zijn praktijkervaringen met het diagnostisch hulpmiddel in kaart gebracht: zowel interviewers (gedragskundigen

en psychodiagnostisch medewerkers) als informanten (begeleiders, familieleden en mentor maar geen familie) bleken overwegend positief. Zij gaven aan dat het diagnostisch hulpmiddel duidelijk is en aansluit bij mensen met (Z)EV(M)B.

Sterktes van het onderzoek

Dementie herkennen en diagnosticeren bij mensen met (Z)EV(M)B is een van de grote uitdagingen in de hedendaagse gehandicaptenzorg. De vraag naar een passend diagnostisch hulpmiddel om dementiegerelateerde achteruitgang bij mensen met (Z)EV(M)B vast te stellen is groot, aangezien reeds bestaande instrumenten slechts gedeeltelijk toepasbaar zijn voor deze doelgroep (Dekker, Wissing, et al., 2021b; Wissing, Dijkstra, et al., 2022). Dit onderzoek geeft naar ons weten voor het eerst gehoor aan deze praktijkvraag door specifiek voor mensen met (Z)EV(M)B een nieuw diagnostisch hulpmiddel voor dementie te ontwikkelen. Dit hulpmiddel richt zich op vaardigheden/gedrag die mensen met (Z)EV(M)B kunnen (ver)tonen en die bij dementie kunnen veranderen.

Een belangrijke sterkte van het nieuwe diagnostisch hulpmiddel is dat rekening gehouden is met de aanzienlijke heterogeniteit binnen de (Z)EV(M)B-doelgroep. Er is namelijk grote variatie in de ontwikkeling van vaardigheden; zo is de een in staat om te spreken en lopen, terwijl de ander non-verbaal communiceert en gebruikmaakt van een rolstoel. In beide gevallen kan het diagnostisch hulpmiddel ingevuld worden, aangezien items over veranderingen in taal en spraak en loopvaardigheden wel of niet aangeboden worden op basis van de karakteristieke aan-/afwezigheid van deze vaardigheden. Ook is rekening gehouden met de reeds bestaande (zeer) ernstige beperkingen in functioneren door bij een aantal items de score optie *'niet van toepassing'* aan te bieden als antwoordmogelijkheid. In de berekening van de totale procentuele veranderingsscore wordt gecorrigeerd voor items waarop *'niet van toepassing'* is gescoord. Immers, niet ontwikkelde vaardigheden kunnen niet achteruitgaan en kunnen dus niet dienen als symptoom voor dementie (Llewellyn, 2011; Sheehan et al., 2015).

Een ander sterk punt is dat items ontwikkeld zijn op basis van triangulatie van bevindingen uit eerdere deelonderzoeken binnen het project '*Praktijkvragen over dementie bij mensen met (Z)EV(M)B*' (Dekker, Wissing, et al., 2021b, 2021a; Wissing, Dijkstra, et al., 2022; Wissing, Fokkens, et al.,

2022; Wissing, Hobbelen et al., 2023; Wissing, Ulgiati, et al., 2022). In de deelonderzoeken is gebruikgemaakt van verschillende kwalitatieve en kwantitatieve onderzoeksmethoden (methodentriangulatie), waarbij verschillende analysetechnieken zijn toegepast (analysetriangulatie) (van Staa & Evers, 2010). Tijdens de analyse is niet alleen gefocust op het identificeren van cognitieve achteruitgang, die de voornaamste indicator is voor dementie in de algemene bevolking en mensen met lichte VB (Jamieson-Craig et al., 2010; Wereldgezondheidsorganisatie, 2014), maar ook dementiegerelateerde veranderingen binnen andere domeinen zijn meegenomen. Dit is een belangrijk pluspunt, aangezien met name de achteruitgang in cognitie bij mensen met (Z)EV(M)B door de al aanwezige (zeer) ernstige VB lastig te herkennen is.

Het diagnostisch hulpmiddel is ontwikkeld voor gebruik in de praktijk en niet puur als wetenschappelijk onderzoeksinstrument. Gedurende het ontwikkelingsproces was er daarom een nauwe samenwerking tussen onderzoekers en deskundigen uit de praktijk. De uiteindelijke versie is vervolgens onderworpen aan een eerste praktijktoets. De resultaten hiervan geven eerste inzichten in de validiteit, de betrouwbaarheid en het discriminerend vermogen van het hulpmiddel. Door tijdens de praktijktoets ook te vragen naar praktijkervaringen kan de uiteindelijke praktische toepasbaarheid van het hulpmiddel beter gewaarborgd worden.

Beperkingen van het onderzoek

Hoewel het uitvoeren van een eerste praktijktoets als sterkte van het onderzoek gezien kan worden, was de omvang van steekproef beperkt. In deze steekproef was er sprake van een onderrepresentatie van mensen zonder loopvaardigheden (karakteristiek). Verder was het opvallend dat juist in de groepen twijfelachtige dementie en een diagnose dementie het merendeel een zeer ernstige verstandelijke beperking heeft. In het diagnostisch hulpmiddel is gevraagd naar de baseline mate van VB, d.w.z. het hoogste functioneren voordat er sprake was van achteruitgang. In de praktijk komt het echter veel voor dat over de tijd de mate van VB aangepast wordt. Dit terwijl de richtlijnen voor het vaststellen van VB stellen dat de VB ontstaan is voor de leeftijd van 22 jaar (Schalock et al., 2021). Hoewel wij hier geen informatie over hebben, zou het kunnen dat dit fenomeen zich ook heeft voorgedaan binnen de steekproef van de praktijktoets.

Een ander mogelijke beperking van dit onderzoek is dat interviewers vaak betrokken zijn (geweest) bij de multidisciplinaire dementiediagnostiek. Hoewel zij door hun betrokkenheid mogelijk bevooroordeeld zouden kunnen zijn geweest, zijn zij uit praktische overwegingen niet uitgesloten als interviewers. Het diagnostisch hulpmiddel werd bovendien niet ingevuld op basis van de kennis van de interviewer, maar op basis van de antwoorden van informanten, waardoor mogelijke invloed van eerdere betrokkenheid van de interviewer bij diagnostiek beperkt wordt.

De beantwoording door informanten bevat een bepaalde mate van subjectiviteit. Informantinterviews in het kader van dementiediagnostiek bij mensen met (Z)EV(M)B zijn echter het beste wat er momenteel is door de afwezigheid van objectievere bloed- en hersenbiomarkers voor dementie en de onbruikbaarheid van direct neuropsychologisch onderzoek. (Elliott-King et al., 2016; Esbensen et al., 2017; Fletcher et al., 2016; Hon et al., 1999; Keller et al., 2016; McKenzie et al., 2018). De gouden standaard voor het diagnosticeren van dementie bij mensen met (Z)EV(M)B is dus het klinische oordeel. Doordat er geen valide referentieschaal bestaat voor dementie bij deze doelgroep, kon de concurrente validiteit niet bepaald worden. Desalniettemin is de validiteit wel versterkt door de uitkomsten van het diagnostisch hulpmiddel te vergelijken met het klinische oordeel (geen dementie, twijfelachtige dementie en diagnose dementie).

Toekomst

Het signaleren en diagnosticeren van dementie bij mensen met (Z)EV(M)B is complex en de vraag naar een passend instrument is groot (Dekker, Wissing, et al., 2021b). Daarom was het doel van dit onderzoek om specifiek voor de (Z)EV(M)B-doelgroep een nieuw diagnostisch hulpmiddel voor dementie te ontwikkelen. De resultaten betreffende de validiteit en de mate waarin het instrument onderscheid kan maken tussen mensen zonder dementie en zij die twijfelachtige dementie of een diagnose dementie hebben zijn veelbelovend. Om die reden en gezien het feit dat er momenteel nog geen hulpmiddelen beschikbaar zijn voor mensen met (Z)EV(M)B en dementie kan het diagnostisch hulpmiddel ingezet gaan worden in de praktijk.

Op basis van de resultaten van dit onderzoek blijkt dat de items 3.2 (paniek), 6.7 (spierverkrampingen) en 7.1 (epilepsie) minder relevant zijn en verwijderd kunnen worden. Verder gaven zowel gedragskundigen en

psychodiagnostisch medewerkers als begeleiders en familieleden aan dat het wenselijk is om meer voorbeelden van kleine observeerbare veranderingen bij mensen met zeer ernstige verstandelijk en meervoudige beperkingen toe te voegen. Juist bij deze subgroep is het nog complexer om dementie te herkennen en diagnosticeren. Bepaalde vaardigheden zijn niet ontwikkeld, waardoor deze dus ook niet achteruit kunnen gaan. Als vaardigheden achteruitgaan, dan is voldoende kennis nodig om deze, vaak kleine veranderingen adequaat te observeren (Dekker, Wissing, et al., 2021a; Wissing, Fokkens, et al., 2022). Zorgmedewerkers en familieleden missen hierin vaak de nodige achtergrondkennis (Cleary & Doody, 2017; Iacono et al., 2014), met name doordat informatie over subtiele dementiesymptomen bij mensen met (Z)EV(M)B beperkt was (Wissing, Ulgiati, et al., 2022). Om informanten op interactieve wijze te voorzien van informatie over dementie bij mensen met (Z)EV(M)B zijn vrij toegankelijke kennismodules ontwikkeld in het project *'Praktijkvragen over dementie bij menen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen'*. Verder is het van belang dat gedragskundigen en psychodiagnostisch medewerkers die het diagnostisch hulpmiddel afnemen, bij de items (aanvullende) voorbeelden geven die passend zijn bij de persoon voor wie het diagnostisch hulpmiddel wordt ingevuld.

Het diagnostisch hulpmiddel helpt gedragskundigen en psychodiagnostisch medewerkers om veranderingen in vaardigheden/ gedrag in kaart te brengen. Op basis van alleen de resultaten van het diagnostisch hulpmiddel kan de diagnose dementie niet gesteld worden. Het is namelijk van belang om andere aandoeningen die dementie-achtige symptomen kunnen veroorzaken uit te sluiten (differentiaaldiagnoses). Hierbij kan ook gekeken worden naar de domeinscores van het diagnostisch hulpmiddel. Stel dat iemand met (Z)EV(M)B zowel gedragsveranderingen als veranderingen in motoriek laat zien, maar geen veranderingen in cognitie, dan is er waarschijnlijk geen sprake van dementie. Mede door het gebruik van het diagnostisch hulpmiddel kan onder- en overdiagnostiek (bijv. bij mensen met (Z)EV(M)B en DS) van dementie mogelijk worden voorkomen.

Het diagnostisch hulpmiddel kan al in de praktijk ingezet worden. De eerste resultaten wijzen op een toegevoegde waarde, zeker gezien het (inter)nationale gemis van een instrument dat specifiek gericht is op dementie bij (Z)EV(M)B. Vervolgonderzoek wordt geadviseerd om de

betrouwbaarheid verder te onderzoeken, inclusief het bepalen van de test-hertestbetrouwbaarheid. Ook is het van belang om in een grotere steekproef opnieuw te kijken naar het discriminerend vermogen van het diagnostisch hulpmiddel door het vergelijken van de drie vooraf ingedeelde diagnostische groepen: geen dementie, twijfelachtige dementie en klinisch gediagnosticeerde dementie. Hierbij kunnen sensitiviteit, specificiteit en positieve/negatieve voorspellende waardes en mogelijke afkapwaardes geanalyseerd worden. Verder kan het waardevol zijn het diagnostisch hulpmiddel meermaals voor een individu met (Z)EV(M)B in te vullen om hiermee veranderingen over de tijd te monitoren. Een dergelijk longitudinaal onderzoek draagt bij aan meer inzicht in zowel vroege signalen als het verloop van dementie.

Conclusie

Om dementiegerelateerde achteruitgang bij mensen met (Z)EV(M)B in kaart te brengen is in dit onderzoek het diagnostisch hulpmiddel dementie bij mensen met (Z)EV(M)B voor gedragskundigen en psychodiagnostisch medewerkers ontwikkeld. Het diagnostisch hulpmiddel bestaat uit 45 items, onderverdeeld in 7 symptoomcategorieën, waarbij veranderingen in de laatste zes maanden t.o.v. het karakteristieke functioneren/gedrag in kaart worden gebracht. De indruks- en inhoudsvaliditeit zijn gewaarborgd in het ontwikkelproces door triangulatie van bevindingen in wetenschappelijke literatuur en praktijkervaringen. Vanuit de praktijktoets waarin de betrouwbaarheid is verkend blijkt dat de interbeoordelaarsbetrouwbaarheid hoog was. Ten aanzien van het discriminerend vermogen lieten de groepsvergelijkingen een trend zien waarbij mensen met een diagnose dementie de meeste veranderingen scoorden en degenen zonder dementie het minst. Dit gold zowel voor item-, domein- als totaalscores. Dit suggereert dat het discriminerend vermogen van het diagnostisch hulpmiddel voldoende is. Praktijkervaringen ten aanzien het gebruik van het diagnostisch hulpmiddel zijn overwegend positief. Tezamen geeft dit een eerste indicatie dat het diagnostisch hulpmiddel een veelbelovend instrument is om dementiegerelateerde achteruitgang vast te stellen bij mensen met (Z)EV(M)B. Aangezien er op dit moment voor de (Z)EV(M)B-doelgroep niets beschikbaar is qua diagnostische hulpmiddelen, kan het al in de praktijk ingezet worden. Vervolgonderzoek wordt geadviseerd om de betrouwbaarheid en het discriminerend vermogen verder te onderzoeken.

Noot

Het diagnostisch hulpmiddel en de bijhorende handleiding zijn gratis te downloaden via www.vb-dementie.nl/diagnostisch-hulpmiddel/.

De interactieve kennismodules over dementie bij mensen met (Z)EV(M)B zijn gratis beschikbaar via www.vb-dementie.nl/kennismodules/.

Chapter 8

Summary & General discussion

Summary

Dementia is increasingly prevalent among people with severe/profound intellectual (and multiple) disabilities (SPI(M)D) than before as their life expectancy has increased. A major challenge in intellectual disability care is to observe symptoms and diagnose dementia in this population. Care professionals and family members are in great need of knowledge about the symptoms and course of dementia as well as dementia screening instruments dedicated to people with SPI(M)D. To fulfill these needs this dissertation aimed to gain insight into dementia in people with SPI(M)D. Firstly, we focused on the relevance of a dementia diagnosis. Secondly, training/information needs regarding dementia people in with SPI(M)D were identified. Thirdly, an inventory was made of observable dementia symptoms in this population. Lastly, we developed a new instrument to aid the dementia diagnosis in people with SPI(M)D. The following paragraphs summarize the main findings of the chapters in this dissertation that report on these four steps.

The relevance of dementia diagnosis

Chapter 2 described the outcomes from multidisciplinary focus groups with care professionals (n=41) and family members (n=8) having experience with people with SPI(M)D showing decline/dementia. They were asked for their opinion on the relevance of dementia diagnosis for people with SPI(M)D. Qualitative inductive content analysis (also known as thematic analysis) showed that participants wanted to know whether an individual with SPI(M)D has dementia for a better understanding. Awareness that someone has dementia, for example, enabled participants to understand 'problematic' behavioral symptoms of dementia. Moreover, participants indicated that a dementia diagnosis is important for making informed choices. First of all, it allowed them to adopt support aims, the way support is provided, and the way of contact and interaction. Secondly, participants were better able to make choices about (medical) treatment, e.g., adjusting treatment plan and medication use. Thirdly, they could better anticipate the future by making, for example, choices about palliative care and the end of life when dementia is diagnosed. Lastly, organizational choices were mentioned, meaning that a diagnosis could contribute to expanding the number/hours of staff or changing daytime activities or living situation if needed. Some participants wondered whether a dementia diagnosis would actually change support or treatment for those with the most

severe disabilities. Nevertheless, the majority stressed that they want to know whether people with SPI(M)D, including those with the most severe disabilities, have dementia.

Training/information needs

In **Chapter 2**, we identified training/information needs regarding dementia in people with SPI(M)D. According to focus group participants, (preliminary) training does not focus (enough) on dementia-related knowledge. Therefore, (preliminary) training should be enhanced by, for example, offering in-depth modules about dementia in people with SPI(M)D. However, participants stated that information about dementia in this population is lacking, and thus more research is needed to develop new knowledge about this topic. Furthermore, it is desired to translate information about dementia in the general population and people with mild/moderate intellectual disabilities (ID) to people with SPI(M)D, if possible. Participants would also like to see available knowledge made more accessible through, for example, collaborations between care organizations. Care organizations should allocate more resources (time, finances) to allow staff to follow (obligatory) training courses. According to participants, such courses should be practice-oriented, include experience-based exercises and will enable them to learn from others' experiences. Besides opportunities to increase knowledge, participants would like to see that (standardized) suitable dementia screening instruments for people with SPI(M)D are developed. This would allow participants to systematically monitor individuals with SPI(M)D. Moreover, such an instrument would also improve the transferability of information. Lastly, participants mentioned that loss of knowledge and experience could be prevented by improving multidisciplinary collaboration and reducing staff changes.

Observable dementia symptoms

An inventory of observable dementia symptoms in people with SPI(M)D was obtained using quantitative and qualitative research methods: a systematic literature review (**Chapter 3**), focus groups (**Chapter 2**), survey & interviews (**Chapter 4**), and clinical records review (**Chapter 5**). In each step, dementia symptoms were according to diagnostic dementia criteria (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022) and literature (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a; Nieuwenhuis-Mark, 2009; Ries, 2018; Strydom et al., 2010) categorized into symptom domains, namely 1) cognitive symptoms,

2) activity of daily living (ADL) symptoms, 3) behavioral and psychological symptoms, 4) motor symptoms, and 5) medical comorbidities.

In **Chapter 3**, we systematically identified observable dementia symptoms reported in existing literature. Only eight studies focused – in part – on dementia in people with SPI(M)D. Symptoms were reported for 25 persons with either severe (n=13) or profound (n=12) ID, among whom 20 had Down syndrome (DS). The total number of identified dementia symptoms was 42, most of which were behavioral and psychological changes (e.g., increased aggression, withdrawal, irritability, and daytime sleepiness). All eight studies reported behavioral and psychological symptoms in those with severe or profound ID and dementia. Cognitive (e.g., memory loss and speech deterioration) and ADL symptoms were found in seven and five studies, respectively. Furthermore, four studies reported that individuals with dementia also showed motor changes (e.g., decline in mobility and gait disturbances). Medical comorbidities (e.g., the onset of epilepsy, incontinence, and weight changes) were described in six studies. Despite the limited number of studies, a first overview of observable dementia symptoms in people with SPI(M)D was obtained with this systematic literature review.

Since literature only provided, to a very limited extent, an answer to the question which dementia symptoms can be observed in people with SPI(M)D, we undertook an explorative approach to collect dementia symptoms based on experiences of intellectual disability care professionals and family members. In **Chapter 2**, we asked focus group participants which dementia symptoms they recognized in people with SPI(M)D. Participants mentioned a total of 177 symptoms, which were categorized using a symptom matrix. The matrix rows were deductively designed, consisting of the five symptom domains described above. To further enhance interpretation, symptoms within domains were grouped into symptom categories. Matrix columns consisted of daily contexts in which symptoms were seen according to participants, resulting from the inductive content analysis. The categorization matrix revealed that cognitive and behavioral changes were most prominently observed dementia symptoms in people with SPI(M)D. A decline in cognitive functioning in this population was observed during ADL, such as nursing, eating/drinking, and mobility/transfers.

The systematic literature review and focus group study results have led to an inventory of dementia symptoms in people with SPI(M)D. In **Chapter 4**, we further identified and deepened observable dementia symptoms in this population through survey and interviews. Firstly, care professionals (n=87) and family members (n=13) of people with SPI(M)D and questionable dementia or diagnosed dementia filled out a survey about which symptoms they had recognized since the onset of questionable dementia or diagnosed dementia. Symptoms included in the survey were those observed in general populations, those identified with focus groups (**Chapter 2**) and those obtained from the literature (**Chapter 3**). The most frequently recognized symptoms concerned: a decline in ADL functioning, behavioral and psychological changes (e.g., increase/decrease in irritable and eating/drinking behavior), and a decline in walking skills. Cognitive symptoms and medical comorbidities were, to a lesser extent, also observed in individuals with SPI(M)D and questionable dementia or diagnosed dementia. Secondly, to collect a richer and more in-depth perspective on symptoms covered in the survey we interviewed care professionals (n=16) having vast experience in recognizing/diagnosing dementia in people with SPI(M)D. Similar to symptoms obtained with focus groups, the total number of 247 symptoms mentioned by interviewees were structured using a symptom matrix. Deductively designed rows comprised the five symptom domains and symptom categories addressed in the survey. Inductive content analysis used to design columns revealed that the observation of certain symptoms depended on whether individuals with SPI(M)D could communicate verbally or could walk. Particularly, cognitive changes were (more easily) observed when individuals had verbal communication and/or walking skills. Nevertheless, some cognitive changes and most behavioral and psychological changes were observed regardless of having such skills. Moreover, motor changes were particularly observed when people were at baseline able to walk. Lastly, changes in ADL functioning and medical comorbidities were observed in people with and people without walking skills at baseline.

Next, in **Chapter 5** we further characterized the natural history of dementia in people with SPI(MD) by determining prevalence and time of onset of symptoms. We identified whether symptoms obtained in previous steps (**Chapters 2-4**) were over time reported in clinical records of people with SPI(M)D without dementia (n=103), with questionable dementia (n=19), and with diagnosed dementia (n=19). Differences in prevalence of symptoms

were found between those with and without questionable dementia or diagnosed dementia. Differences in the prevalence of symptoms were found between those with and without questionable dementia or diagnosed dementia. Most symptoms were more common in people with questionable dementia and most prevalent in those with diagnosed dementia. Except for medical comorbidities, the total number of symptoms in each domain (i.e., cognitive, ADL, behavioral and psychological, and motor domain) was significantly higher for people with questionable dementia or diagnosed dementia compared to those without dementia. Further analysis showed that the number of cognitive symptoms, behavioral and psychological symptoms, and motor symptoms were predictors for questionable dementia or diagnosed dementia. Regarding time of onset of symptoms, results demonstrated that most prevalent early symptoms in those with diagnosed dementia were memory loss, declined walking skills, increased anxious, apathetic and irritable behavior.

In short, the systematic literature review evidently showed that dementia in people with SPI(M)D had received very little attention so far. The symptoms reported in the limited number of identified studies have led to a first inventory of observable dementia symptoms in people with SPI(M)D (**Chapter 3**). To further enhance understanding, we adopted an explorative research approach to collect dementia symptoms based on experiences in practice. Firstly, focus groups revealed that dementia symptoms, particularly cognitive changes, were observed in several specific daily contexts (**Chapter 2**). Secondly, survey results indicated that the most frequently observed symptoms were a decline in ADL functioning, behavioral and psychological changes, and a decline in walking skills (**Chapter 4**). Thirdly, interviews showed that symptoms, particularly cognitive changes, were (more easily) observed when people with SPI(M)D have verbal communication or walking skills (**Chapter 4**). Lastly, from the clinical records review it became apparent that in people with questionable dementia or diagnosed dementia more cognitive, ADL, behavioral and psychological as well as motor symptoms were observed than in those without dementia. Memory loss, declined walking skills, increased anxious, apathetic and irritable behavior were the most prevalent early symptoms in people with SPI(M)D and diagnosed dementia. The number of cognitive, behavioral and psychological, and motor symptoms were predictors for dementia (**Chapter 5**).

Instrument to aid the dementia diagnosis

Until now, no standardized dementia screening instrument was available for people with SPI(M)D. Existing dementia screening instruments available for people with ID (all levels of severity) as a whole are namely unsuitable for this population. However, specific items within those instruments may still apply to this population. Therefore, in **Chapter 6**, we identified applicable items for people with SPI(M)D in four frequently used dementia screening instruments: adapted Dutch version of the Dementia Scale for Down Syndrome (DSVH), Behavioral and Psychological Symptoms of Dementia in Down Syndrome evaluation scale version II (BPSD-DS II), Dutch Dementia Questionnaire for persons with Mental Retardation (DVZ) and Social competence Rating scale for people with ID (SRZ). We conducted interviews with informants of people with SPI(M)D without dementia (n=40) to determine which items are applicable at baseline, i.e., the highest level of functioning before decline/dementia occurs. Results demonstrated that of the 193 items, 101 were applicable for people with SPI(M)D. Most applicable items were those about behavioral and psychological changes (60 items). Furthermore, items about cognitive changes (25 items), motor changes (6 items), changes in ADL (5 items), and medical comorbidities (5 items) were found applicable. Additional analysis revealed that items about verbal communication (17 items) and gross motor function (5 items) were only applicable for the subgroup of individuals who were able to communicate verbally/independently walk at baseline.

In **Chapter 7**, we developed a novel instrument to aid the dementia diagnosis in people with SPI(M)D based on identified observable dementia symptoms (**Chapters 2-5**) and identified applicable items (**Chapter 6**) (Figure 8.1). Firstly, we summarized and assimilated results of our previous research steps. There was a great resemblance in dementia symptoms identified with various research methods. Therefore, items for the new diagnostic aid were developed for symptoms found with four or five research methods, i.e., systematic literature review, focus groups, survey, interviews and clinical records review. Identified applicable items were used as inspiration for new items. Face and content validity were ensured by developing items based on triangulation, i.e., results obtained with various research methods and analyzed using various techniques. In total, we developed 45 items, and categorized them based on research findings and feedback from psychologists and psychological assistants into 7 symptom domains, namely 1) cognitive changes, 2) changes in language

and speech, 3) behavioral changes, 4) changes in eating/drinking, 5) changes in personal care, 6) motor changes, and 7) medical comorbidities.

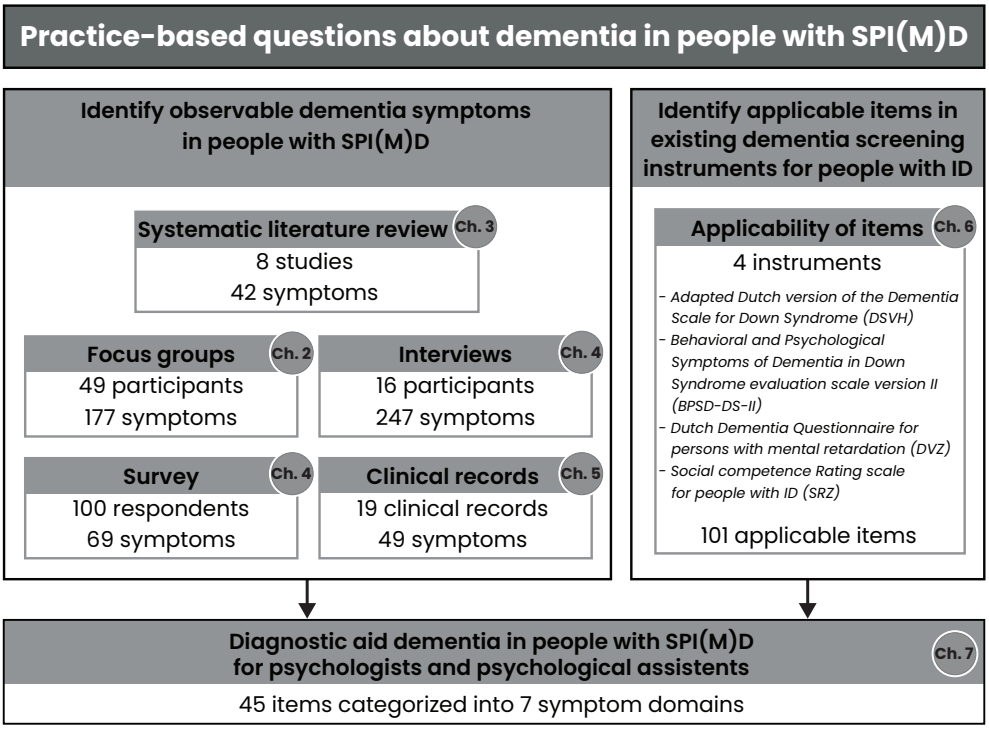


Figure 8.1 Schematic overview of the main findings of chapters that report on observable dementia symptoms in people with SPI(M)D and applicable items for this population. These findings guided the development of the diagnostic aid for dementia in people with SPI(M)D. Abbreviations: Ch., Chapter; ID, intellectual disabilities; SPI(M)D, severe/profound intellectual (and multiple) disabilities.

We tested the diagnostic aid in practice by conducting interviews with informants of people with SPI(M)D without dementia (n=18), with questionable dementia (n=10), and with diagnosed dementia (n=10). A psychologist/psychologic assistant asked for every item whether the informant(s) had observed that the person with SPI(M)D showed a change compared to their characteristic functioning/behavior, i.e., baseline functioning/behavior before any deterioration occurred. Every item answered with either *'yes, decreased'* or *'yes, increased'* was scored as 1 point. Items for which there was *'no change'* observed were scored as 0 points. Some items had the extra scorings option *'not applicable'* or *'unknown'* for which no score was assigned. Additionally, the two items

about changes in language and speech and two items about walking skills were only asked and scored if individuals were at baseline able to verbally communicate/walk.

Based on the first test in practice the reliability, discriminative ability, and practice-based experiences were examined. Interrater reliability was established for five interviews: scored by a psychologist/psychological assistant as well as a researcher. Interrater reliability for each domain (80.0-100.0%) and overall interrater reliability (94.0%) were high. Furthermore, internal consistency among all items was good (Cronbach's alpha=0.892). Regarding discriminative ability, we found for the majority of items a trend that people with diagnosed dementia showed the most change (either decrease or increase) and people without dementia the least. Scores on fourteen items differed significantly between the three groups. Three items (panic, muscle cramps, and epilepsy) were identified as being irrelevant, given that ≥85% of the people with questionable dementia and people with diagnosed dementia showed no change on these items. Furthermore, not only item scores but also domain and total scores were compared between groups. Between groups, significant differences were found for five domain scores (sum of individual item scores within that domain), i.e., cognitive changes, behavioral changes, changes in eating/drinking, changes in personal care, and motor changes, as well as total scores. People with questionable dementia or diagnosed dementia had higher domain and total scores compared to people without dementia. These results provide a first indication that the instrument has sufficient discriminative ability. Lastly, we also asked informants and interviewees about their experiences with the diagnostic aid. Their experiences were mainly positive because it was clear and covered all areas where change can occur in people with dementia.

Overall, the results suggest that the instrument can identify dementia-related changes in people with SPI(M)D. Future studies are required to examine reliability and discriminative ability further, i.e., cut-off scores with corresponding sensitivity, specificity, and predictive values. Given that until now no dementia screening instrument dedicated to this population existed, it is recommended to implement the diagnostic aid – with the removal of three identified irrelevant items – in intellectual disability care organizations.

General discussion

We might shut our eyes when we think that things are getting too complicated. Could that also be the case when recognizing and diagnosing dementia in people with SPI(M)D? What if we take our time to open our eyes and observe again? This will in all likelihood result in new insights. That is exactly what is needed to be better able to recognize and diagnose dementia in people with SPI(M)D.

The above description applies to our research about dementia in people with SPI(M)D. Instead of neglecting the topic, we have taken up the challenge to unravel the complex puzzle of dementia in people with SPI(M)D. This dissertation provides the first stepping stones to lay this puzzle by addressing:

1. Why should we observe once again?
2. What is needed to observe once again?
3. Which dementia symptoms can be observed?
4. How can we observe again?

The main findings for each of these questions will be discussed in the following paragraphs.

Why should we observe once again?

People with SPI(M)D have, just like anyone else, the right to the highest attainable standard of health (UN General Assembly, 2006; World Health Organization, 1946). Because of this right, one could argue that we should do all that is needed to establish a dementia diagnosis, but is a dementia diagnosis really that relevant for people with SPI(M)D? In the general population, an early dementia diagnosis helps individuals to understand the symptoms they are experiencing (Rasmussen & Langerman, 2019). It also allows people to live longer independently and plan ahead while they still have the capacity, i.e., make choices and express wishes for (future) support, care and treatment (Rasmussen & Langerman, 2019). Because of the pre-existing severe/profound limitations in intellectual functioning, people with SPI(M)D probably have little to no understanding of the meaning of a dementia diagnosis. Moreover, people with SPI(M)D need high levels of support for daily tasks, and thus already (totally) depend on caregivers and family members (American Psychiatric Association, 2013; Schalock et al., 2021). Hence, the positive benefits of receiving an (early) dementia diagnosis, as presented above, do not seem to apply to people

with SPI(M)D. Nevertheless, a diagnosis provides an explanation for the person's health problem and informs decisions, not only for the individual him-/herself, but also for care professionals and family members (Diagnostic Error in Health Care, 2015; Rasmussen & Langerman, 2019). Our research showed that family members and care professionals want to know if an individual with SPI(M)D has dementia as it allows them to better understand and sympathize with dementia-related changes an individual displays (**Chapter 2**). Moreover, a dementia diagnosis enables family members and care professionals to (early) make informed choices (**Chapter 2**). For example, the way of being supported can be adjusted from an activating, developmental-oriented approach to an approach aimed at comfort and maintaining skills. Therefore, a dementia diagnosis is an important means for maintaining quality of life of individuals with SPI(M)D as quality of life in this population depends on quality of support/ care provided by others (Nieuwenhuijse, 2023; van der Putten et al., 2017).

The question remains how far we should go to establish a dementia diagnosis in people with SPI(M)D. When an individual with SPI(M)D starts showing potential dementia signs it is important to try to unravel the underlying cause. Symptoms can namely be caused by – often treatable – conditions such as depression, delirium, vision or hearing problem, hypothyroidism, sleep apnea, or vitamin B12 deficiency (Moriconi et al., 2015; Scott & Barrett, 2007). If such conditions remain undiagnosed, treatment might be withheld, and therefore treatable symptoms persist or even aggravate. Dementia cannot (yet) be prevented or cured, and symptoms will – even with treatment – worsen progressively. Therefore, over time it will become more apparent that an individual has dementia (Alzheimer's Association, 2022). Establishing a diagnosis of dementia is thus a process of clinical assessment and reassessment (Scott & Barrett, 2007). Because of the complexity of diagnosing dementia in people with SPI(M)D, clinicians might – particularly in early stages of dementia – be hesitant to diagnose dementia in this population. This probably has to do with the binary classification of diseases, i.e., someone either has a particular disease or not (Vickers et al., 2008). To diagnose dementia, a person should meet the diagnostic criteria for dementia (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). Given that dementia is a progressive disease, it might be beneficial to classify symptoms as they progress over time, i.e., from no symptoms to very severe symptoms (Rasmussen & Langerman, 2019; Reisberg et al., 1982). Diagnosing dementia

will then become an even more iterative process, in which clinicians regularly observe once again, i.e., reassess to gather information, integrate and interpret information and adapt the classification if necessary. In that way, more emphasis is placed on obtaining information that enables care professionals and family members to make informed choices instead of focusing on establishing an actual dementia diagnosis.

What is needed to observe once again?

To be able to observe dementia symptoms and establish a diagnosis knowledge about about symptoms and course of dementia in people with SPI(M)D is needed. Noteworthy, our research showed that (preliminary) training of care professionals hardly focuses on dementia-related knowledge (**Chapter 2**), which was also reported in scientific literature (Iacono et al., 2014; Whitehouse et al., 2000). To increase care professionals' knowledge about dementia in people with SPI(M)D, (preliminary) training needs to be enhanced. To contribute to this, we developed an online training product consisting of eight modules developed based on the knowledge presented in this dissertation. Each module consists of a theoretical part, a quiz, and an example from practice. In this way, we have complied with participants' wishes for a practice-oriented training product about dementia in people with SPI(M)D with experience-base exercises (**Chapter 2**). In intellectual disability care, training products like e-learnings are often offered to care professionals working within care organizations, and are therefore usually not accessible for, for example, family members. Not only by participants (**Chapter 2**) but also in literature it was reported that knowledge should be made more accessible (Heller et al., 2018; Iacono et al., 2014). Therefore, the training modules and thus knowledge about dementia in people with SPI(M)D is freely accessible via www.vb-dementie.nl/kennismodules. This allows care professionals and family members to individually acquire knowledge about dementia in people with SPI(M)D. Moreover, it is recommended that care organizations implement the newly developed training product within their organization, preferably by offering them in group sessions to stimulate a discussion, making it possible to learn from others' experiences. To facilitate such training sessions and presentations, we offer a set of Powerpoint sheets for each module to offer more background information (www.vb-dementie.nl/downloads). Additionally, these sheets can be included in preliminary training, i.e., intermediate vocational education (mbo), higher vocational education (hbo) as well as higher education (wo). Overall, this will lead to increased

awareness among (future) care professionals about the possibility that people with SPI(M)D will or already have developed dementia.

Which dementia symptoms can be observed?

In people with SPI(M)D, dementia manifests as a set of cognitive symptoms, ADL symptoms, behavioral and psychological symptoms, motor symptoms, and medical comorbidities (**Chapters 2-5 & 7**). The same applies to the general population (Alzheimer's Association, 2022), indicating that dementia symptom domains do not differ between the general and SPI(M)D population. While the domains do not differ, we demonstrated that the way symptoms are to be observed in this population does differ. One needs to observe in detail whether the person shows (subtle) alterations (**Chapters 2 & 4**). Despite pre-existing cognitive deficits, cognitive symptoms were widely recognized in people with SPI(M)D and dementia (**Chapters 2-5 & 7**). These symptoms were mainly apparent during ADL, such as nursing, eating/drinking, and mobility/transfer (**Chapter 2**). Memory loss was the most frequent early cognitive symptom in people with SPI(M)D (**Chapter 5**). Our results thus showed a decline in cognitive functioning is – just like in the general population (Alzheimer's Association, 2022; Stern et al., 1993) – an early indicator for dementia in people with SPI(M)D. Some cognitive symptoms like decreased planning, problem solving, and judgment were hardly recognized in people with SPI(M)D (**Chapter 4**). If cognitive functions have not been developed, such skills cannot decline, and therefore cannot serve as symptoms indicative of dementia (Llewellyn, 2011; Sheehan, Sinai, et al., 2015). The presence and presentation of cognitive symptoms, but also ADL and motor symptoms, thus depends on the baseline functioning of the individual with SPI(M)D. Changes are less noticeable when someone does not verbally communicate or walk independently (**Chapter 4**). In fact, the more severe the pre-existing disabilities, the smaller the set of symptoms becomes and the more subtle changes are (e.g., no longer able to put the arm into a sleeve (**Chapter 2**), making no eye contact anymore (**Chapter 4**)). Therefore, dementia signs may easily go unnoticed, particularly in people with profound intellectual and multiple disabilities.

In people with SPI(M)D, behavioral and psychological symptoms are most observable for caregivers and family members (**Chapters 2-7**). People with SPI(M)D could, for whatever underlying reason, display different behavior compared to their (life-long) characteristic behavior. Our results demonstrated that behavioral and psychological changes were

more frequently observed in individuals with dementia (**Chapters 5 & 7**). Particularly, changes in anxious, apathetic, irritable behavior were common (**Chapters 2, 4 & 5**) and seem to be early 'alarm signals' for dementia in people with SPI(M)D (**Chapter 5**). These results are similar to findings in the general population (Eikelboom et al., 2021) and people with DS (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a). In these studies, depressive behavior was also considered to be an early symptom (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a; Eikelboom et al., 2021; Jost & Grossberg, 1996). In most clinical records of individuals with SPI(M)D and dementia, increased depressive behavior was reported in the years before the diagnosis (**Chapter 5**). However, changes in depressive behavior were less often recognized by care professionals and family members filling out the survey (**Chapter 4**). A possible explanation for this might be that apathy and depression have overlapping symptoms (Dekker, Sacco, et al., 2018). Therefore, differentiating between apathetic and depressive symptoms of dementia is rather complex in people with SPI(M)D. Another explanation for this is that people with SPI(M)D hardly or not at all communicate verbally, and therefore cannot report their mood and even do not have the cognitive level for specific symptoms that classically characterize depression, such as doom-mongering or being tired of life (Dekker, Sacco, et al., 2018; Dekker, Strydom, et al., 2015; K. M. Evans et al., 1999). According to care professionals and family members, depressive symptoms in people with SPI(M)D include, for example, increased mood changes and crying (**Chapters 2 & 4**). The presentation of behavioral and psychological symptoms of dementia is thus somewhat different than in the general population.

Besides cognitive and behavioral and psychological changes, motor changes are frequently observed in people with SPI(M)D and dementia (**Chapters 2-7**). Similarly to the general population (Kueper et al., 2017; Ramakers et al., 2007), a deterioration in gait (e.g., decline in gait speed, increased gait unsteadiness) was observed in those able to walk independently (**Chapters 4, 5 & 7**). Some individuals with SPI(M)D have severe limitations in motor functioning (Houwen et al., 2014; Nakken & Vlaskamp, 2007). The ability to move (parts of) their body may be very limited, but changes can still be observed (**Chapter 6**). Given the pre-existing disabilities in people with SPI(M)D, it might not always be clear what the underlying problem of changes is. For example, caregivers have observed that an individual no longer takes food from a spoon. This may be due to loss of oral motor skills, forgetting the spoon (amnesia), no

longer recognizing the spoon (agnosia), less focus on eating (apathetic behavior), or having less appetite (eating/drinking behavior) (**Chapter 4**). The fact remains that individual shows a decline compared to his/her characteristic functioning. If multiple changes are observed over time, the person with SPI(M)D might have developed dementia. Our results namely showed that people with SPI(M)D had in total more cognitive, behavioral and psychological, motor, and ADL symptoms than those without dementia (**Chapter 5**). Moreover, the number of cognitive, behavioral and psychological, and motor changes were predictors for dementia in people with SPI(M)D.

How can we observe once again?

A diagnostic process of dementia should start way before an individual with SPI(M)D potentially develops dementia. A dementia diagnosis namely requires the presence of cognitive/behavioral symptoms which represent a decline from an individual's previous higher level of functioning, which is sufficient enough to interfere with daily functioning (American Psychiatric Association, 2013; McKhann et al., 2011; World Health Organization, 2022). Information about the baseline level of functioning/behavior is thus needed to establish a diagnosis. Performing a timely baseline measurement is essential to gather information about abilities/skills/behavior someone with SPI(M)D has displayed throughout adult life, and which in the future may decline/change due to the development of dementia (Keller et al., 2016). In The Netherlands, it is recommended that psychologists/psychological assistants perform such a baseline measurement for people with SPI(M)D at the age between 30 and 35 years (Uijl & Weijer, 2022). For the baseline measurement as well as monitoring of changes over time in people with ID, informant-based dementia screening instruments are used (Zeilinger et al., 2022). Our results, as presented in **Chapter 6**, demonstrated that four dementia screening instruments used in The Netherlands, but also internationally recommended and widely used instruments – DSVH (Dutch adaptation of DSDS, (Maaskant & Hoekman, 2011), BPSD-DS II (Dekker, Sacco, et al., 2018; Dekker, Ulgiati et al., 2021a); DVZ (DMR/DLD; Evenhuis et al., 1998) and SRZ (Kraijer et al., 2004) – were as a whole unsuitable for people with SPI(M)D. For people with SPI(M)D, the newly developed instrument to aid the dementia diagnosis (**Chapter 7**) can – with the removal of three identified irrelevant items – be implemented in practice, and can thus be used for baseline measurement. In doing so, it is important to not only score items, but also to report in detail what the individual is capable of

doing and which (characteristic) behavior they are displaying.

When changes are observed in people with SPI(M)D, the newly developed diagnostic aid (**Chapter 7**) could be used in the diagnostic process. It namely allows psychologists/psychological assistants to identify, based on observations of informants, if and in which areas the individual has shown changes in the last six months. If a baseline measurement is performed with the diagnostic aid, scores could be compared to determine whether there is a decline from an individual's higher level of functioning. Moreover, repeatedly using the diagnostic aid allows to monitor the progression of changes over time. Based solely on the results of the diagnostic aid, dementia cannot and should not be diagnosed. Dementia is namely a diagnosis of exclusions, meaning that dementia-like changes can be caused by recent (life) events (e.g., moving home, death of a family member, new caregiver/fellow residents, etc.) or conditions with symptoms resembling those of dementia (Scott & Barrett, 2007). Conditions such as cerebrovascular accident, chronic pain, delirium, depression, epilepsy, hearing impairments, hypothyroidism, medication side effects and intoxication, sleep apnea, visual impairments, and vitamin B12 deficiency should be ruled out (Alzheimer's Association, 2022; Moriconi et al., 2015; Scott & Barrett, 2007). In line with research reporting on the prevalence of physical health problems in people with SPI(M)D (Evenhuis et al., 2001; Rousseau et al., 2019; van Timmeren et al., 2016), we found in clinical records a high prevalence of vision impairments, hearing impairments and epilepsy in people with SPI(M)D (**Chapter 5**). These conditions appear to be common in people with SPI(M)D. Therefore, they should be extensively screened for in individuals suspected of having dementia, considering that they could be the underlying cause of dementia-like changes. Remarkably, in clinical records of people with questionable or diagnosed dementia, often nothing was reported about the presence/absence of conditions that mimic dementia (**Chapter 5**). It is important to report more systematically on steps undertaken in the diagnostic process as it helps clinicians to integrate/interpret gathered information and to make a (working) diagnosis.

Recognizing and diagnosing dementia in people with SPI(M)D requires close multidisciplinary collaboration between clinicians (e.g., physicians, psychologists), caregivers, family members, and allied health care professionals (e.g., speech-language therapists, and physiotherapists).

This is in intellectual disability care, but also in studies presented in this dissertation (**Chapters 2 & 4-7**) referred to as multidisciplinary collaboration. Multidisciplinary implies that people from different professions each work on their own discipline-specific plan or care, but work together with others by sharing their findings (Choi & Pak, 2006). Given the complexity of recognizing and diagnosing dementia in people with SPI(M)D (**Chapter 1**), the aim should be to collaborate interprofessional. This goes beyond multidisciplinary collaboration in that people from various professions actively work together with each other as well as the people themselves and their families to deliver the highest quality of care across settings (World Health Organization, 2010). With regard to the diagnostic process in people with SPI(M)D, it means that care professionals and family members collaborate more intensively, i.e., repeatedly discuss which changes are observed, jointly decide on and plan steps to establish what (potentially) causes the changes an individual with SPI(M)D is displaying.

Overall, the gold standard to diagnose dementia in people with SPI(M)D is currently still clinical assessment involving informant interviews, physical examination, observations, screening case notes, brain imaging and/or blood tests to identify conditions that mimic dementia. There is ongoing research on biomarkers for dementia to improve the detection, diagnosis, and treatment of dementia (Ahmed et al., 2014; de França Bram et al., 2019; Fortea et al., 2021; Teunissen et al., 2022). In the general population, brain imaging has become a widely used technique in diagnostic work-up for people suspected of having dementia (Scott & Barrett, 2007; Young et al., 2020). In people with ID, the added value of brain imaging is being questioned. People with ID are likely to already have atypical brain development, and therefore brain images may be misinterpreted (Sheehan et al., 2014; Sullivan et al., 2012). Moreover, in people with DS aged 40 years and over, positron emission tomography (PET) with amyloid tracers will be positive regardless of having clinical symptoms of dementia due to Alzheimer's disease (Abrahamson et al., 2019). Therefore, but also because of the potential burden and risks, clinicians may be hesitant to let people with SPI(M)D suspected of dementia undergo brain imaging. Therefore, for example, in The Netherlands, only in a few cases brain imaging is part of the diagnostic process of dementia in people with SPI(M)D. Potential alternative biomarkers are cerebrospinal fluid (CSF) and blood-based biomarkers as reported in studies in the general population (Ahmed et al., 2014; Teunissen et al., 2022), but also DS population (de França Bram et al.,

2019; Dekker et al., 2017; Fortea et al., 2020, 2021; Montoliu-Gaya et al., 2023, 2021). Particularly blood examination is a relatively easy, less burdensome/ invasive procedure, which is often already part of the diagnostic work-up, i.e., to test for hypothyroidism or vitamin B12 deficiency, and therefore the preferred option for people with ID (Montoliu-Gaya et al., 2021). Despite promising results in research, blood biomarkers for dementia in ID are not yet widely available for clinical practice. It would be beneficial for people with SPI(M)D if blood-based biomarkers for dementia detection become standard part of the diagnostic process as it might assist (early) diagnosis in this population.

Methodological reflection

Recognizing and diagnosing dementia in people with SPI(M)D is highly complex (**Chapter 1**), partly because it has largely been neglected in literature (**Chapter 3**). To the best of our knowledge, we are the first to specifically address dementia in this population. Given the lack of scientific data, we undertook an explorative approach to obtain practice-based observations and experiences of care professionals and family members. People with SPI(M)D namely rely on their observations to recognize dementia (McKenzie et al., 2018). A potential risk of obtaining practice-based observation and experiences is that it may result in the same, already available knowledge. It is namely likely that care professionals and family members lack necessary background knowledge (Furniss et al., 2011; Herron & Priest, 2013; Whitehouse et al., 2000), partly because information about dementia in people with SPI(M)D has been scarce (**Chapter 3**). Consequently, dementia symptoms might not be recognized or may wrongly be attributed to the ID, and thus not reported by family members and care professionals. On the other hand, obtaining practice-based observations and experiences can help break the vicious circle. Care professionals and family members having (vast) experience with dementia in people with SPI(M)D are able to give concrete examples of subtle signs of decline that they have observed. Summarizing and analyzing practice-based observations and experiences obtained using different research methods, i.e., triangulation, has led to a more rich and more in-depth perspective on the presentation and course of dementia in people with SPI(M)D (**Chapters 2, 4-7**). Therefore, these quantitative and qualitative studies were important first steps in generating knowledge in an area where information is scarce.

Obtaining data based on observations and experiences in practice also entailed some challenges. Care professionals participating in either the focus groups (**Chapter 2**), the survey, or interviews (**Chapter 4**) often support/provide care to people with different levels of functioning. Consequently, they might – despite clearly emphasizing the focus on people with SPI(M)D – have referred to dementia symptoms observed in people with mild or moderate ID. If this became apparent from the data, those specific parts in the transcripts of focus groups and interviews and we excluded the survey respondent. Furthermore, we also excluded participants having a mild or moderate ID (**Chapters 5-7**). It still might be the case that some symptoms observed among people with a baseline mild or moderate ID were included. Existing literature (**Chapter 3**; Burt et al., 1998) and clinical record review (**Chapter 5**) namely revealed that in intellectual disability care, they tend to change the level of ID over time. With the progression of dementia, someone with a baseline mild or moderate ID is now referred to as someone having severe or profound ID. The ID, which originates before the age of 22 (Schalock et al., 2021), has a different underlying cause than dementia. Therefore, the development of dementia does not change the level of ID.

Another methodological challenge in research with people with SPI(M)D is the small sample size (Maes et al., 2021). It is already complex to recruit large samples of participants with SPI(M)D, let alone those with SPI(M)D and dementia. Our strategy to improve research participation was to recruit participants via intellectual disability care organizations within the study consortium. What stands out is that included participants with either questionable dementia or diagnosed dementia were almost all able to walk at baseline, while it was emphasized that also those with severe motor disabilities could participate (**Chapters 5 & 7**). This most likely relates to the fact that the more severe the pre-existing disabilities, the smaller the set of symptoms and the more subtle the signs are, and therefore the more complex recognizing and diagnosing dementia becomes. That is why an enhanced understanding of dementia in people with SPI(M)D is so important. Knowledge about the presentation and course of dementia in people with SPI(M)D namely allows caregivers and family members – the people with SPI(M)D rely on for observing symptoms (McKenzie et al., 2018) – to more timely recognize signs based on which steps can be undertaken to determine the underlying cause.

A challenge is the lack of instruments to screen for dementia in people with SPI(M)D. This is a complicating factor not only for establishing a dementia diagnosis in clinical practice, but also for conducting research. Dementia symptoms could not be identified with a standardized instrument. Direct neuropsychological tests could, for example, not be used because of floor effects (Elliott-King et al., 2016; Esbensen et al., 2017; McKenzie et al., 2018). We applied various quantitative and qualitative research methods to identify symptoms and applicable items for people with SPI(M)D in already existing dementia screening instruments for people with ID (**Chapters 2-6**). Triangulation – multiple quantitative/quantitative methods, data sources, analyzing techniques as well as collaborative research (van Staa & Evers, 2010) – proved to be essential for developing new items for a novel instrument to aid the dementia diagnosis in people with SPI(M)D as it allowed to ensure face and content validity (**Chapter 7**). Validity was strengthened by letting care psychologists and psychologists assistants involved in diagnostic work-up review developed items. Thereafter, they conducted interviews with informants of people with SPI(M)D with and without questionable dementia or diagnosed dementia to test the instrument in practice. This may be a risk of bias, because many interviewers were already involved in the diagnostic process. To minimize that risk, informants provided answers, and thus not by interviewers. Moreover, a researcher, unacquainted with the individuals with SPI(M)D, was present at a subset of interviews to also score informants' responses. Based on this, interrater reliability was established, but further assessment of reliability (test-retest reliability) is required. Furthermore, concurrent validity could not be established, because results of the first test in practice could not be compared to a (gold) standard instrument as currently no such instrument exists. Discriminative ability has been assessed in relation to the status of dementia. Thereby, it is important to note that diagnosis of dementia, and thus division into groups, was established on forehand without considering the results of the diagnostic aid. Overall, results of the first test in practice are promising, and therefore will benefit practice, but also pave the way for future research.

Future directions for research

The need in intellectual disability care for a dedicated instrument to aid the dementia diagnosis dementia in people with SPI(M)D was high. Based on the results of the first test in practice, we recommend implementing

the newly developed diagnostic aid for dementia in people with SPI(M)D in intellectual disability care organizations.. Further assessment of its reliability and discriminative ability is still required. To also meet the international need for a suitable instrument (Esbensen et al., 2017; McKenzie et al., 2018), it is recommended to translate the diagnostic aid into English and other languages. This would also allow to examine reliability and discriminative ability in a larger international SPI(M)D study population. Regarding discriminative ability, sensitivity, specificity and predictive values should be determined for different cut-off scores for domain as well as total scores. Based on the scores, a dementia diagnosis cannot be established, but cut-off scores can serve as an indicator for clinicians that an individual with SPI(M)D may have dementia. Implementation in practice should go along with further research. For example, further development of a digital version of the diagnostic aid would enhance feasibility and enable easy (international) data collection – either with consent or anonymized – for further research. The digital version can be optimized based on research findings, and changes can directly be implemented in practice.

Furthermore, it is recommended to use the diagnostic aid for dementia in people with SPI(M)D in longitudinal studies as well as cross-sectional studies with larger sample sizes to further enhance the understanding of the natural history of dementia in people with SPI(M)D. Longitudinal studies would enable to assess the progression of dementia in individuals over time in those with DS, but also in those without DS. In the studies presented in this dissertation, the majority of the participants with either questionable dementia or diagnosed dementia had DS (**Chapters 5 & 7**). Given that people with DS have an extremely high genetic risk of developing dementia due to Alzheimer's disease (Ballard et al., 2016), most identified symptoms are likely to be symptoms of Alzheimer's disease. Cross-sectional studies with larger sample sizes would allow to compare symptoms observed in people with and without DS. If underlying etiology is established – which is still rather difficult in people with SPI(M)D – symptoms observed in individuals with different dementia subtypes (e.g., AD, dementia with Lewy bodies, vascular dementia, or frontotemporal dementia) should be examined. Biomarker research in people with SPI(M)D could also enhance understanding of dementia and its underlying etiology. Ideally, biomarkers and the diagnostic aid for dementia in people with SPI(M)D should be used in future research to study the relationship between biomarkers and the clinical presentation of symptoms. Thereby, the diagnostic aid can be

further validated based on biomarkers, and the diagnostic performance of newly identified biomarkers can be validated with scores on the diagnostic aid for dementia in people with SPI(M)D.

In some intellectual disability care organizations, video observations are part of the diagnostic process. Video observations allow to identify subtle changes that may else go unnoticed by family members and caregivers. It would be relevant to further develop a standardized video protocol for diagnosing dementia in people with SPI(M)D. We found that symptoms were generally observed in specific daily contexts (i.e., nursing, eating/drinking, mobility/transfers, communication, and leisure activities), and therefore these should be the moments to record videos (**Chapter 2**). To analyze the videos, recordings could be scored using the diagnostic aid for dementia in people with SPI(M)D. Combining video and diagnostic aid enables researchers to further examine early subtle symptoms as well as the presentation of symptoms in those with the most severe disabilities. Most importantly, it would benefit practice as it further enhances the diagnostic process of dementia in people with SPI(M)D.

In this dissertation, the main focus has been on recognizing dementia symptoms and establishing a dementia diagnosis. Further research is required to assess management strategies for dementia in people with SPI(M)D. Dementia cannot (yet) be prevented or cured, but people with SPI(M)D and dementia may benefit from pharmacological and non-pharmacological interventions. Pharmacological options are, for example, antidementia medication to slow the progression of symptoms (Arvanitakis et al., 2019). None of the participants in our clinical records review used antidementia medication (**Chapter 5**). This might be related to the fact that little research has been carried out to establish the efficacy, safety, and tolerability of this medication in people with ID (de Oliveira & Faria, 2022; Keller et al., 2016). Moreover, various non-pharmacological interventions are available, e.g., physical exercise, music therapy, snoezelen, and much more (Arvanitakis et al., 2019; Solé et al., 2022; Tisher & Salardini, 2019). The effects of (non)-pharmacological interventions could, in practice as well as in research, be determined and monitored by using the diagnostic aid for dementia in people with SPI(M)D.

Recommendations for practice

Figure 8.2 summarizes recommendations for practice, which are based on results and experiences acquired within the project *'Practice-based questions about dementia in people with severe/profound intellectual (and multiple) disabilities'* of which this dissertation is part. The recommendations are explained in more detail on the next two pages, separately for each group: 1) intellectual disability care organizations, 2) clinicians and allied health care professionals, 3) caregivers and family members, and 4) people with SPI(M)D.

Figure 8.2 Recommendations for practice regarding recognizing and diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D).

Intellectual disability care organizations

- Actively spread knowledge about dementia in people with SPI(M)D to increase awareness among staff as well as family members.
- Train staff about dementia in people with SPI(M)D by implementing and offering the training product which is freely available on www.vb-dementie.nl/kennismodules.
- Implement the diagnostic aid for dementia in people with SPI(M)D which is freely available on www.vb-dementie.nl/diagnostisch-hulpmiddel, and make it part of standard regular screening for dementia in people with SPI(M)D, particularly for those with DS.
- Create a culture of interprofessional collaboration where care professionals and family members actively work together to recognize and diagnose dementia in people with SPI(M)D.
- Minimize staff turnover as much as possible to make sure that there are caregivers who have knowledge about an individual's characteristic functioning/behavior.
- Optimize clinical records systems so that it allows to report observed changes at a central and uncluttered location and train staff in how and where to report changes in clinical records.
- Collaborate with research centers on projects about dementia in people with SPI(M)D.

Clinicians & allied health care professionals who support the diagnostic process

- Increase your level of knowledge about dementia in people with SPI(M)D to be better able to recognize and diagnose dementia. Visit our project website www.vb-dementie.nl.
- Use the diagnostic aid to perform a timely baseline measurement for individuals with SPI(M)D, and thereby focus on obtaining information about what someone is capable of doing and which (characteristic) behavior he/she is displaying.
- Make the diagnostic aid standard part of the diagnostic process to identify and monitor changes over time.
- Observe regularly once again, not only by repeatedly filling out the diagnostic aid but by observing the individual yourself.
- Ensure short communication lines that allow caregivers and family members to easily contact you if they observe changes.

- Do not be too hesitant to share your suspicions of dementia with caregivers and family members. This can namely already help them to understand what potentially is going and it also allows to earlier make informed choices together.
- Make a shared and well-considered decision about which steps (not) to undertake in the diagnostic process and systematically report on them.

Caregivers and family members

- Increase your level of knowledge about dementia in people with SPI(M)D to be better able to recognize dementia symptoms. Visit our project website www.vb-dementie.nl.
- Make time to observe regularly once again and discuss the (potential dementia) changes you observe with other caregivers and family members.
- Systematically report on the changes you have observed.
- Contact clinicians if you observe (potential dementia) changes and have the feeling that something might be wrong.

People with SP(I(M)D

It might be very difficult for you to understand the symptoms you are experiencing due to the development of dementia. It is up to us family members, care professionals, and researchers to take our time to observe again. Only then can we better understand what is going on with you and how we can support you best to maintain your quality of life.

Appendices

Summary in Dutch	Samenvatting
Acknowledgements	Dankbetuiging
List of abbreviations	Lijst met afkortingen
References	Literatuurlijst

Summary in Dutch | Samenvatting

Introductie

Iedere persoon met (zeer) ernstige verstandelijke (en meervoudige) beperkingen (afgekort tot (Z)EV(M)B) heeft unieke vaardigheden en gedragingen. Over de tijd kunnen vaardigheden achteruitgaan en gedragingen veranderen, bijvoorbeeld door leeftijdsgerelateerde aandoeningen zoals dementie. Dementie kenmerkt zich door verschillende symptomen, waarbij er sprake is van een achteruitgang in het cognitief functioneren vergeleken met een eerder hoger niveau van functioneren. Deze achteruitgang moet invloed hebben op het dagelijks functioneren. Naast de achteruitgang in cognitie en algemene dagelijkse levensverrichtingen (ADL) kunnen er ook gedragsveranderingen, achteruitgang in motorische vaardigheden en/of bijkomende gezondheidsproblemen ontstaan. Voorheen kwam dementie nauwelijks voor bij mensen met (Z)EV(M)B, omdat zij niet de leeftijd bereikten waarop dementie kon ontwikkelen. Leeftijd is immers de belangrijkste risicofactor voor dementie. In de afgelopen decennia is de levensverwachting van mensen met (Z)EV(M)B sterk toegenomen door betere (medische) zorg en leefomstandigheden. Doordat mensen met (Z)EV(M)B steeds ouder worden, komt dementie steeds vaker voor.

Met name mensen met downsyndroom hebben een enorm groot risico op dementie door de ziekte van Alzheimer. Van de mensen met downsyndroom heeft zo'n 20-30% een (zeer) ernstige verstandelijke beperking. Downsyndroom wordt veroorzaakt door een derde exemplaar van chromosoom 21, vandaar dat het ook wel trisomie 21 wordt genoemd. Op dit chromosoom bevindt zich het gen voor de productie van het amyloïd-eiwit. Het hebben van een extra kopie van het chromosoom 21 zorgt voor een overmatige aanmaak van het amyloïd-eiwit dat langzamerhand 'samenklontert' en zich ophoopt tussen de hersencellen in zogenaamde amyloïd-plaques. Deze plaques zijn kenmerkend voor de ziekte van Alzheimer. Rond de leeftijd van 40 jaar hebben bijna alle mensen met downsyndroom een behoorlijke hoeveelheid amyloïd-plaques in de hersenen. Opvallend genoeg betekent dit niet dat iedere persoon met downsyndroom op die leeftijd ook al klinische symptomen van dementie vertoont. Op 40-jarige leeftijd is de klinische diagnose dementie gesteld bij ongeveer 9%, dit percentage stijgt naar ongeveer 75% bij een leeftijd van

65 jaar. Dit percentage is aanzienlijk hoger dan in de algemene bevolking, waar zo'n 5% tussen de 65 en 74 jaar dementie ontwikkelt. Voor mensen met verstandelijke beperkingen met een andere onder liggende oorzaak dan downsyndroom is er weinig onderzoek gedaan naar hoe vaak dementie voorkomt. Het beperkte aantal studies dat gedaan is rapporteert ook nog eens wisselende percentages die lager (4.2% bij leeftijd van 65^{+}) of hoger (13,9% & 18,3% bij leeftijd van 65^{+}) zijn dan in de algemene bevolking. Wat deze studies wel lieten zien is dat dementie dus óók voorkomt bij mensen met verstandelijke beperkingen.

Om dementie bij mensen met (Z)EV(M)B te herkennen, het liefst vroegtijdig, en te diagnosticeren is kennis nodig over de uiting en het verloop van dementie bij deze groep mensen. Dit bleek slechts beperkt bekend doordat er bijna geen onderzoek was gedaan naar dementie bij mensen met (Z)EV(M)B. Mede daardoor gaven behandelaren (artsen/verpleegkundig specialisten, gedragskundigen, psychodiagnostisch medewerkers en paramedici), begeleiders en familieleden aan dat zij te weinig kennis hebben over dementie bij mensen met (Z)EV(M)B. Zij merkte wel op dat het functioneren van een aantal mensen met (Z)EV(M)B achteruitging, maar konden niet precies vaststellen wat er aan de hand was en of deze achteruitgang veroorzaakt werd door dementie. Door zowel te weinig kennis over dementie en omdat het complex is om dementie te herkennen bij mensen met (Z)EV(M)B is het zeer waarschijnlijk dat dementie vaak niet wordt herkend en dus ook niet gediagnosticeerd wordt. Verschillende factoren zorgen er namelijk voor dat het moeilijk is voor begeleiders en familieleden om symptomen te herkennen en voor behandelaren om een diagnose te stellen. In Figuur A.1 zijn de complicerende factoren voor het herkennen en diagnosticeren van dementie bij mensen met (Z)EV(M)B samengevat. Deze factoren zijn ingedeeld in drie niveaus: 1) karakteristieken van mensen met (Z)EV(M)B, 2) herkennen van symptomen en 3) diagnostisch onderzoek.

Figuur A.1 Complicerende factoren voor het herkennen en diagnosticeren van dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen ((Z)EV(M)B).

Gezien het grote aantal complicerende factoren kun je jezelf afvragen of het wel mogelijk en zelfs noodzakelijk is om een achteruitgang veroorzaakt door dementie te vast te stellen. De complicerende factoren met betrekking tot de karakteristieken van mensen met (Z)EV(M)B zullen niet veranderen. Wel kan het vergroten van de kennis van begeleiders en familieleden ervoor zorgen dat symptomen beter en eerder herkend worden. Om dementie te diagnosticeren hebben behandelaren niet alleen meer kennis nodig, maar zij hebben ook behoefte aan een diagnostisch hulpmiddel voor dementie dat geschikt is voor mensen met (Z)EV(M)B. Bestaande instrumenten blijken namelijk niet geschikt te zijn voor mensen met (Z)EV(M)B. Directe neuropsychologische testen zijn bij mensen met (Z)EV(M)B door de (zeer) ernstige verstandelijke beperking en beperkte verbale vaardigheden lastig of onmogelijk. Daarnaast blijken ook dementielijsten ontwikkeld voor mensen met verstandelijke beperkingen in z'n geheel ongeschikt voor mensen met (Z)EV(M)B. Deze lijsten bevatten namelijk items waarin gevraagd wordt naar veranderingen in vaardigheden die mensen met (Z)EV(M)B nooit ontwikkeld hebben. Het diagnosticeren van dementie bij mensen met (Z)EV(M)B bestaat, door de afwezigheid van geschikte dementielijsten, vooral uit klinische beoordeling door (ervaren) artsen en gedragskundigen en gesprekken met begeleiders en familieleden. Juist zij vragen zich af hoe dementie zich uit bij mensen met (Z)EV(M)B en hoe de diagnose dementie gesteld kan worden.

Om antwoord te kunnen geven op de bovenstaande praktijkvragen richt dit proefschrift zich op het verkrijgen van meer kennis en inzichten over dementie bij mensen met (Z)EV(M)B door te focussen op het:

1) In kaart brengen van de relevantie van de diagnose dementie
2) Identificeren van scholings- en kennisbehoefte van zorgmedewerkers en familieleden
3) Identificeren van observeerbare dementiesymptomen bij mensen met (Z)EV(M)B
4) Ontwikkelen van een diagnostisch hulpmiddel voor dementie bij mensen met (Z)EV(M)B

Om dit te bereiken is er praktijkgericht onderzoek uitgevoerd in het project '*Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*'. Dit project is een samenwerking tussen drie kennisinstellingen (Rijksuniversiteit Groningen, Universitair Medisch Centrum Groningen en Hanzehogeschool Groningen) en vier zorgorganisaties (Alliade, 's Heeren Loo, Ipse de Bruggen en Koninklijke Visio) gesubsidieerd door ZonMW. De belangrijkste resultaten van het onderzoek worden hieronder samengevat.

Relevantie van de diagnose dementie bij mensen met (Z)EV(M)B

In **Hoofdstuk 2** zijn zorgmedewerkers (41 personen) en familieleden (8 personen) van oudere mensen met (Z)EV(M)B bij wie sprake is van achteruitgang/dementie in focusgroepen (d.w.z. groepsinterviews) bevraagd over waarom het belangrijk is om te weten of bij iemand met (Z)EV(M)B sprake is van dementie. Uit de thematische analyse, blijkt dat focusgroep deelnemers behoefte hebben aan een diagnose dementie om (probleem)gedrag te kunnen verklaren. Verder gaven deelnemers aan dat een diagnose zorgmedewerkers en familieleden in staat stelt om geïnformeerde keuzes te maken. Zo kunnen de begeleidingsdoelen en begeleidingsstijl worden aangepast wanneer bekend is dat iemand met (Z)EV(M)B dementie heeft. Verder kunnen er keuzes gemaakt worden over het aanpassen van het behandelplan en medicatiegebruik. Ook gaven deelnemers aan dat de diagnose het mogelijk maakt om te anticiperen op de toekomst, zoals het anticiperen op het ziekteverloop, familieleden tijdig voorbereiden op wat er mogelijk gaat komen en keuzes maken over palliatieve zorg en het levenseinde. Een diagnose kan ook bijdragen aan de besluitvorming over het inzetten van meer personeel en het aanpassen van de dagbesteding- en/of woonsituatie. Sommige deelnemers vroegen zich af of een diagnose daadwerkelijk zou leiden tot aanpassingen van

de zorg/behandeling van de mensen met de ernstigste beperkingen. Toch benadrukten de meeste deelnemers dat ze ook bij deze mensen willen weten of er sprake is van dementie. Het is daarom belangrijk dat er bij mensen met (Z)EV(M)B gedegen (differentiaal) diagnostisch onderzoek wordt uitgevoerd, om te voorkomen dat er een verkeerde diagnose gesteld wordt en mensen daardoor geen of een verkeerde behandeling krijgen. Dementie zelf kan (nog) niet behandeld worden, maar ook voor mensen met (Z)EV(M)B is het mogelijk om farmacologische (bijv. gedragsregulerende medicatie) en niet-farmacologische interventies (bijv. muziektherapie en snoezelen) in te zetten. Het is dus belangrijk voor zorgmedewerkers en familieleden om te weten of iemand met (Z)EV(M)B dementie heeft voor begrip en om geïnformeerde keuzes te kunnen maken. Daarmee kan de kwaliteit van bestaan van mensen met (Z)EV(M)B en dementie behouden worden, aangezien de kwaliteit van bestaan bepaald wordt door de kwaliteit van ondersteuning/zorg die geboden wordt door zorgmedewerkers en familieleden.

Scholings- en kennisbehoefte van zorgmedewerkers en familieleden

In **Hoofdstuk 2** is ook de scholings- en kennisbehoefte van zorgmedewerkers en familieleden ten aanzien van dementie bij mensen met (Z)EV(M)B in kaart gebracht. Focusgroep deelnemers gaven aan dat kennis over dementie niet of nauwelijks aan bod is gekomen tijdens hun vooropleiding. Dit wordt mede veroorzaakt doordat er tot voor kort nauwelijks kennis was over dementie bij mensen met (Z)EV(M)B op basis waarvan scholing ontwikkeld kon worden. Deelnemers benadrukten dat er meer onderzoek nodig is naar dementie bij mensen met (Z)EV(M)B. Bestaande kennis over dementie in de algemene bevolking en mensen met lichte/matige verstandelijke beperking zou beter vertaald moeten worden naar de (Z)EV(M)B doelgroep. Op basis van opgedane kennis kunnen zowel opleidingen als na/-bijscholing verbeterd worden, bijvoorbeeld door de ontwikkeling van een module over dementie bij mensen met (Z)EV(M)B. Samenwerking tussen zorgorganisaties is daarbij belangrijk om te voorkomen dat iedere organisatie zelf iets ontwikkeld. Zo'n module moet goed aansluiten bij de praktijk (concreet, kort en bondig) en moet de mogelijkheid bieden om te leren van de ervaringen van collega's. Verder is het van belang dat zorgorganisaties tijd en geld beschikbaar stellen om personeel (verplichte) scholing te laten volgen. Naast mogelijkheden tot kennisverbreding, gaven deelnemers ook aan dat er grote behoefte is aan de ontwikkeling van een diagnostisch hulpmiddel specifiek voor

dementie bij mensen met (Z)EV(M)B. Dit stelt behandelaren in staat om veranderingen bij mensen met (Z)EV(M)B systematisch te monitoren. Dit maakt informatie ook beter overdraagbaar. Ten slotte benadrukten deelnemers dat multidisciplinaire samenwerking en samenwerken met familieleden belangrijk is. Personeelswisselingen moeten zoveel mogelijk beperkt worden, zodat kennis en ervaringen niet verloren gaan.

Om tegemoet te komen aan de scholings- en kennisbehoefte van zorgmedewerkers en familieleden is binnen het samenwerkingsverband een online scholingsproduct over dementie bij mensen met (Z)EV(M)B ontwikkeld. Dit scholingsproduct is ontwikkeld op basis van de kennis die beschreven staat in dit proefschrift. De invulling en vormgeving is afgestemd met mensen werkzaam in de gehandicaptenzorg. Het scholingsproduct bestaat uit acht interactieve kennismodules waarin onderwerpen zoals het belang van het (h)erkennen van dementie bij mensen met (Z)EV(M)B worden behandeld. Elke module bestaat uit een theorie gedeelte, vijf quizvragen om je kennis te testen en een praktijkvoorbeeld (casus). Vaak worden scholingsproducten aangeboden binnen zorgorganisaties (achter een inlogmuur) waardoor ze niet toegankelijk zijn voor familieleden. De kennismodules over dementie bij mensen met (Z)EV(M)B zijn vrij toegankelijk via www.vb-dementie.nl/kennismodules. Hierdoor kunnen zowel zorgmedewerkers als familieleden zelf kennis opdoen over dementie bij mensen met (Z)EV(M)B. Bovendien kunnen de kennismodules binnen zorgorganisaties in groepsbijeenkomsten aangeboden worden om van elkaars kennis en ervaringen te kunnen leren. Ter ondersteuning van dergelijke bijeenkomsten hebben we voor iedere module een Powerpoint presentatie met aanvullende achtergrondinformatie gemaakt (www.vb-dementie.nl/downloads). Deze presentaties kunnen ook ingezet worden in mbo-, hbo- en universitaire opleidingen. Door de kennismodules en presentaties kan het kennisniveau van (toekomstige) zorgmedewerkers en familieleden verhoogd worden. Dit bevordert (vroeg) signalering en diagnostisering van dementie bij mensen met (Z)EV(M)B.

Observeerbare dementiesymptomen bij mensen met (Z)EV(M)B

In **Hoofdstuk 3** staan de resultaten van het systematisch literatuuronderzoek beschreven. Er is gezocht naar dementiesymptomen bij mensen met (Z)EV(M)B die gerapporteerd stonden in bestaande Engelstalige wetenschappelijke artikelen. Slechts acht studies richtten zich gedeeltelijk op dementie bij mensen met (Z)EV(M)B. Symptomen stonden

beschreven voor 25 personen met ernstige (13 personen) of zeer ernstige (12 personen) verstandelijke beperkingen. Het totaal aantal gevonden symptomen was 42, waarvan de meeste gedragssymptomen waren (bijv. toename in agressie, zichzelf terugtrekken, prikkelbaarheid, overdag slapen). Alle acht studies beschreven gedragssymptomen. Cognitieve symptomen (bijv. geheugenverlies en achteruitgang in taalvaardigheden) en ADL symptomen (bijv. achteruitgang in persoonlijke verzorging) zijn in respectievelijk acht en vijf studies gevonden. Bovendien rapporteerden vier studies dat personen met (Z)EV(M)B en dementie motorische veranderingen (bijv. achteruitgang in mobiliteit en veranderingen in looppatroon) lieten zien. Bijkomende gezondheidsproblemen (bijv. ontstaan van epilepsie, incontinentie en gewichtsveranderingen) stonden beschreven in zes studies.

Aangezien literatuur over dementie bij mensen met (Z)EV(M)B beperkt bleek, was er heel weinig bestaande informatie om op af te gaan. Daarom hebben we in dit onderzoek gebruik gemaakt van verschillende kwalitatieve en kwantitatieve onderzoeksmethoden om observeerbare dementiesymptomen bij mensen met (Z)EV(M)B in kaart te brengen. Praktijkervaringen zijn opgehaald middels focusgroepen, een enquête en interviews en er werd dossieronderzoek verricht. Door gebruik te maken van zogenaamde triangulatie van onderzoeksmethoden werd voorkomen dat individuele ervaringen het beeld te veel verkleuren en zijn de grote gemene delers geïdentificeerd, d.w.z. symptomen die uit meerdere methoden naar voren komen. In iedere stap zijn symptomen ingedeeld in de symptoomdomeinen die bekend zijn uit de literatuur: 1) cognitieve symptomen, 2) ADL symptomen, 3) gedragssymptomen, 4) motorische symptomen en 5) bijkomende gezondheidsproblemen.

In **Hoofdstuk 2** is aan focusgroep deelnemers gevraagd welke dementiesymptomen zij bij mensen met (Z)EV(M)B en dementie herkennen. Deelnemers noemden in totaal 177 symptomen, die gecategoriseerd zijn door middel van een symptoommatrix. De rijen van de matrix bestaan uit de vijf eerder beschreven symptoomdomeinen. Voor verdere duiding zijn symptomen binnen elk domein ingedeeld in symptoomcategorieën. De kolommen van de matrix bestaan uit dagelijkse situaties waarin symptomen in de praktijk vaak worden gezien. Uit de symptoommatrix blijkt dat cognitieve veranderingen en gedragsveranderingen het meest geobserveerd werden voor mensen met (Z)EV(M)B en dementie. Een

achteruitgang in het cognitief functioneren werd in deze groep mensen met name herkend tijdens ADL zoals verzorging, eten/drinken en mobiliteit/transfers. Specifieke symptomen staan beschreven in **Hoofdstuk 2** (Engels) en in het Nederlandstalige artikel: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*: *onderzoek naar observeerbare symptomen, relevantie van diagnose, en scholingsbehoefte* (Dekker, Wissing et al., 2021b).

In **Hoofdstuk 4** zijn eerder geïdentificeerde symptomen onderzocht en nieuwe symptomen in kaart gebracht middels een enquête en interviews. Allereerst vulden zorgmedewerkers (87 personen) en familieleden (13 personen) van mensen met (Z)EV(M)B en (vermoedens van) dementie een enquête in over welke symptomen zij gezien hadden sinds het ontstaan van (vermoedens van) dementie. Symptomen waarnaar gevraagd werd waren symptomen gevonden met de focusgroepen en het literatuuronderzoek (**Hoofdstukken 2 & 3**) en symptomen die bekend zijn van dementie in de algemene bevolking en mensen met lichte/matige verstandelijke beperking. Achteruitgang in ADL, gedragsveranderingen (bijv. toename/afname in prikkelbaar en eet-/drinkgedrag) en een afname in loopvaardigheden zijn het vaakst genoemd. Cognitieve symptomen en bijkomende gezondheidsproblemen werden in mindere mate herkend bij mensen met (Z)EV(M)B en (vermoedens van) dementie. Vervolgens zijn zorgmedewerkers (16 personen) met een rijke ervaring in het herkennen en diagnosticeren van dementie bij mensen met (Z)EV(M)B geïnterviewd. De in totaal 247 gevonden symptomen zijn, net als de symptomen uit de focusgroepen, gecategoriseerd door middel van een symptoommatrix. De resultaten laten zien dat cognitieve symptomen met name worden herkend bij mensen met (Z)EV(M)B die verbale en/of loopvaardigheden hebben. Sommige cognitieve symptomen en de meeste gedragsveranderingen werden gezien ongeacht de aan-/afwezigheid van deze vaardigheden. Veranderingen in motoriek werden voornamelijk opgemerkt bij mensen met loopvaardigheden. Tot slot werden veranderingen in ADL en bijkomende gezondheidsproblemen genoemd voor mensen met en zonder loopvaardigheden.

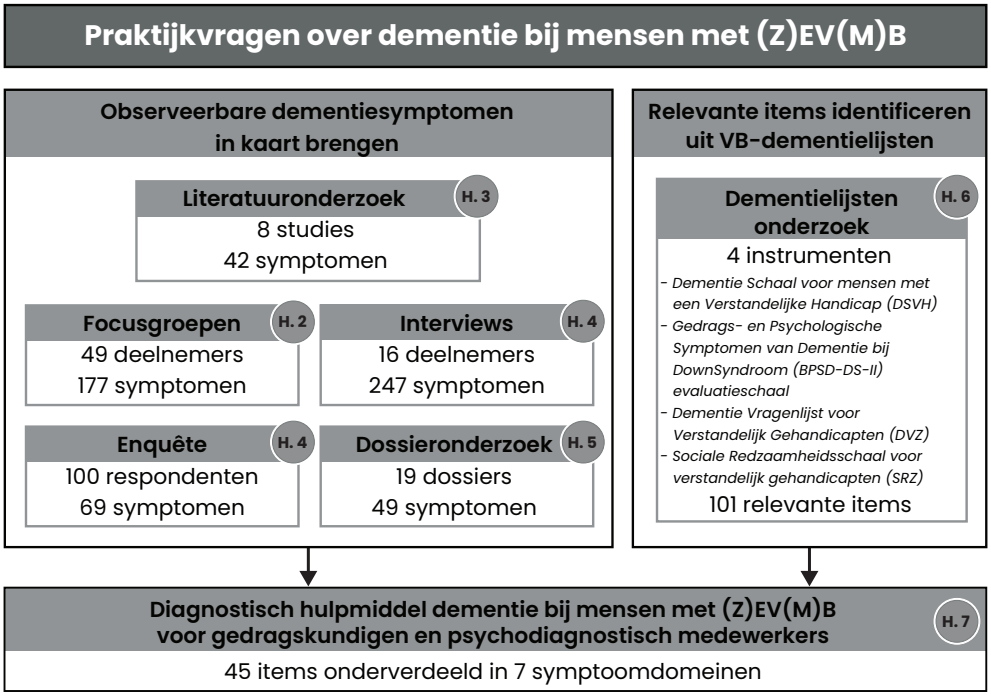
In **Hoofdstuk 5** is het verloop van dementie bij mensen met (Z)EV(M)B onderzocht door te bepalen hoeveel mensen symptomen vertoonden en op welk moment symptomen ontstaan. Hiervoor zijn dossiers van mensen met (Z)EV(M)B zonder dementie (103 personen),

met twijfelachtige dementie (19 personen) – d.w.z. dat er vermoedens van dementie zijn, maar nog geen klinische diagnose is gesteld – en klinisch gediagnosticeerde dementie (19 personen) onderzocht. In deze dossiers is gekeken of de in de **Hoofstukken 2-4** gevonden symptomen over de tijd gerapporteerd waren. De meeste symptomen stonden vaker gerapporteerd in dossiers van mensen met twijfelachtige dementie en het vaakst bij mensen met de diagnose dementie. Met uitzondering van bijkomende gezondheidsproblemen was het totaal aantal symptomen in ieder domein (cognitieve, ADL, gedrags- en motorische domein) hoger voor mensen met twijfelachtige dementie of diagnose dementie, in vergelijking met mensen zonder dementie. Bovendien bleek dat het aantal cognitieve symptomen, gedragssymptomen en motorische symptomen voorspellers zijn voor twijfelachtige dementie of een diagnose dementie. Wat betreft het moment van ontstaan van symptomen bleken geheugenverlies, achteruitgang in loopvaardigheden, toename in angstig gedrag, apathisch gedrag en prikkelbaar gedrag de meest voorkomende vroege signalen te zijn van dementie bij mensen met (Z)EV(M)B.

Observeerbare dementiesymptomen bij mensen met (Z)EV(M)B, geïdentificeerd door gebruik te maken van verschillende onderzoeksmethoden (methodentriangulatie) en analysetechnieken (analysetriangulatie) (**Hoofdstukken 2-5**), komen sterk overeen. Gezamenlijk laten deze resultaten zien dat dementie bij mensen met (Z)EV(M)B zich, net als in de algemene bevolking, uit als een groep van cognitieve symptomen, ADL symptomen, gedragssymptomen, motorische symptomen en bijkomende gezondheidsproblemen. Echter, bij mensen met (Z)EV(M)B is de groep van (mogelijke) dementiesymptomen kleiner in vergelijking met de algemene bevolking. Dit is te wijten aan het feit dat mensen met (Z)EV(M)B bepaalde vaardigheden, zoals het kunnen plannen, oplossen van problemen en beoordelen van situaties, nooit ontwikkeld hebben. Deze vaardigheden kunnen dan ook niet achteruitgaan, waardoor deze niet kunnen dienen als signalen van dementie. Bovendien laten de resultaten zien dat je bij mensen met (Z)EV(M)B klein moet kijken om (subtiele) dementiesymptomen te herkennen. Dit vraagt van begeleiders en familieleden dat zij weten hoe het karakteristieke functioneren/gedrag van iemand met (Z)EV(M)B is en welke veranderingen kunnen wijzen op dementie.

Diagnostisch hulpmiddel voor dementie bij mensen met (Z)EV(M)B
Zoals eerder vermeld, bestond er geen geschikt diagnostisch instrument voor dementie bij mensen met (Z)EV(M)B. Bestaande dementielijsten ontwikkeld voor mensen met verstandelijke beperkingen zijn in z'n geheel ongeschikt voor mensen met (Z)EV(M)B. Echter, specifieke items kunnen wel van toepassing zijn voor deze groep mensen. Daarom zijn in **Hoofdstuk 6** relevante items voor mensen met (Z)EV(M)B geïdentificeerd uit vier dementielijsten die in de Nederlandse gehandicaptenzorg (veel) gebruikt worden: 1) Dementie Schaal voor mensen met een Verstandelijke Handicap (DSVH), 2) Gedrags- (Behavioural) en Psychologische Symptomen van Dementie bij Down Syndroom (BPSD-DS-II) evaluatieschaal, 3) Dementie Vragenlijst voor Verstandelijk Gehandicapten (DVZ) en 4) Sociale Redzaamheidsschaal voor verstandelijk gehandicapten (SRZ). Er zijn interviews gehouden met begeleiders en familieleden van mensen met (Z)EV(M)B zonder dementie (40 personen) om te bepalen welke items van toepassing zijn voordat er sprake van achteruitgang/dementie is. Van de in totaal 193 items bleken er 101 relevant te zijn voor mensen met (Z)EV(M)B. De meeste van deze relevante items gingen over gedragsveranderingen (60 items). Bovendien bleken items over cognitieve veranderingen (25 items), motorische veranderingen (6 items), veranderingen in ADL (5 items) en bijkomende gezondheidsproblemen relevant te zijn. Verdere analyses toonden dat 17 items over verbale communicatie alleen relevant waren voor mensen met (Z)EV(M)B die zich verbaal kunnen uitten. Tot slot bleken 5 items met betrekking tot grove motoriek alleen relevant te zijn voor mensen die in staat zijn om te lopen.

In **Hoofdstuk 7** is een nieuw diagnostisch hulpmiddel voor dementie bij mensen met (Z)EV(M)B ontwikkeld op basis van geïdentificeerde dementiesymptomen (**Hoofdstukken 2-5**) en relevante items (**Hoofdstuk 6**) (Figuur A.2). Hierdoor zijn de indruks- en inhoudsvaliditeit verzekerd. In totaal zijn er 45 items ontwikkeld die zijn onderverdeeld in 7 symptoomdomeinen, namelijk 1) veranderingen in cognitie, 2) veranderingen in taal en spraak, 3) gedragsveranderingen, 4) veranderingen in eten en drinken, 5) veranderingen in persoonlijke verzorging, 6) veranderingen in motoriek en 7) bijkomende gezondheidsproblemen. De indruks- en inhoudsvaliditeit zijn gewaarborgd in het ontwikkelproces door triangulatie van bevindingen in wetenschappelijke literatuur en praktijkervaringen.



Figuur A.2 Schematische weergave van de belangrijkste resultaten beschreven in de hoofdstukken die ingaan op observeerbare dementiesymptomen bij mensen met (Z)EV(M)B en relevante items voor deze doelgroep. Deze resultaten vormden de basis voor het ontwikkelen van een passend diagnostisch hulpmiddel om dementiegerelateerde veranderingen bij mensen met (Z)EV(M)B in kaart te brengen. Afkortingen: H., hoofdstuk; VB, verstandelijke beperkingen; (Z)EV(M)B; (zeer) ernstige verstandelijke (en meervoudige) beperkingen.

Het diagnostisch hulpmiddel is onderworpen aan een eerste praktijktoets, waarbij gedragskundigen en psychodiagnostisch medewerkers interviews hebben gehouden met begeleiders en familieleden van mensen met (Z)EV(M)B zonder dementie (18 personen), met twijfelachtige dementie (10 personen) en een diagnose dementie (8 personen). Vanuit de praktijktoets waarin de betrouwbaarheid is verkend blijkt dat de interbeoordelaarsbetrouwbaarheid hoog was. Wat betreft het discriminerend vermogen – d.w.z. de mate waarin het diagnostisch hulpmiddel onderscheid kan maken tussen mensen zonder dementie en mensen met twijfelachtige dementie en mensen met een diagnose dementie – blijkt dat bij de meeste items veranderingen vaker gescoord werden voor mensen met twijfelachtige dementie en het vaakst voor degenen met een diagnose dementie. De drie items over paniek,

spierverkrampingen en epilepsie bleken irrelevant te zijn, omdat ≥85% van de mensen met twijfelachtige dementie en de mensen met een diagnose dementie geen verandering lieten zien. Verschillen tussen de groepen zijn niet alleen gevonden voor itemscores, maar ook voor de domein- en totaalscores. Voor vijf van de zeven domeinscores, namelijk veranderingen in cognitie, gedragsveranderingen, veranderingen in eten en drinken, veranderingen in persoonlijke verzorging en veranderingen in motoriek zijn er net als voor de totaalscores statistisch significante verschillen gevonden tussen de groepen. Mensen met twijfelachtige dementie of een diagnose dementie hadden hogere domein- en totaalscores in vergelijking met de mensen zonder dementie. Tezamen geeft dit een eerste indicatie dat het diagnostisch hulpmiddel voldoende discriminerend vermogen heeft. Tot slot hebben we zowel gedragskundige en psychodiagnostisch medewerkers als begeleiders en familieleden gevraagd naar hun ervaringen met het diagnostisch hulpmiddel. Zij gaven aan dat het diagnostisch hulpmiddel duidelijk is en goed aansluit bij mensen met (Z)EV(M)B.

Gezien de veelbelovende resultaten en het feit dat er momenteel geen geschikt diagnostisch instrument voor dementie bij mensen met (Z)EV(M)B bestaat, kan het diagnostisch hulpmiddel ingezet gaan worden in de praktijk. Het diagnostisch hulpmiddel met bijbehorende handleiding is beschikbaar via www.vb-dementie.nl/diagnostisch-hulpmiddel. Dit diagnostisch hulpmiddel kan gebruikt worden om een zogenaamde nulmeting te doen, waarbij rond de leeftijd van 30-35 jaar het karakteristiek functioneren/gedrag in kaart gebracht worden. Wanneer bij iemand met (Z)EV(M)B veranderingen geobserveerd worden kan het diagnostisch hulpmiddel opnieuw ingevuld worden om te bepalen op welke domeinen iemand in de laatste zes maanden veranderingen vertoond. Wanneer er een nulmeting is gedaan kunnen scores met elkaar vergeleken worden om vast te stellen of er sprake is van een achteruitgang ten opzichte van een eerder hoger niveau van functioneren. Door het diagnostisch hulpmiddel over de tijd regelmatig in te vullen kunnen veranderingen over tijd gemonitord worden. Op basis van alleen de scores van het diagnostisch hulpmiddel kan en mag de diagnose dementie niet gesteld worden. Veranderingen kunnen namelijk ook veroorzaakt worden door recente (levens)gebeurtenissen (bijv. verhuizen) of door aandoeningen (bijv. traag werkende schildklier of vitamine-B12-tekort) die dementie-achtige symptomen kunnen veroorzaken. Deze differentiaaldiagnoses dienen eerst uitgesloten te worden, zoals ook duidelijk vermeld staat in de handleiding van het diagnostisch hulpmiddel.

Wel wordt geadviseerd om de betrouwbaarheid en het discriminerend vermogen van het diagnostisch hulpmiddel verder te onderzoeken. Met betrekking tot het discriminerend vermogen kunnen mogelijk afkapwaardes voor zowel domein- als totaalscores bepaald worden. Deze afkapwaardes kunnen dan in de praktijk worden gebruikt als indicatie dat een persoon met (Z)EV(M)B mogelijk dementie heeft. Om ook tegemoet te komen aan de internationale vraag naar een geschikt diagnostisch instrument, is het wenselijk om het diagnostisch hulpmiddel op termijn te vertalen naar andere talen.

Figuur A.3 Aanbevelingen voor de praktijk betreffende het herkennen en diagnosticeren van dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen ((Z)EV(M)B).

Aanbevelingen voor de praktijk

Gebaseerd op de onderzoeksresultaten zoals beschreven in dit proefschrift, samen met de opgedane ervaringen binnen het onderzoeksproject '*Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*' zijn aanbevelingen voor de praktijk geformuleerd. Deze aanbevelingen zijn apart weergegeven in Figuur A.3 voor de drie niveaus: 1) zorgorganisaties voor mensen met verstandelijke beperkingen, 2) behandelaren, en 3) begeleiders en familieleden. gen dat er begeleiders zijn die kennis hebben over het karakteristieke functioneren/ gedrag van de persoon met (Z)EV(M)B.

Zorgorganisaties voor mensen met verstandelijke beperkingen

- Verspreid kennis over dementie bij mensen met (Z)EV(M)B om bewustwording bij personeel en familieleden te vergroten.
- Bied het vrij beschikbare scholingsproduct aan binnen de organisatie om personeel te scholen over dementie bij mensen met (Z)EV(M)B.
  www.vb-dementie.nl/kennismodules.
- Implementeer het diagnostisch hulpmiddel dementie bij mensen met (Z)EV(M)B in de organisatie. Zorg dat het een standaard onderdeel wordt van regelmatige screening, met name voor mensen met downsyndroom.
  www.vb-dementie.nl/diagnostisch-hulpmiddel.
- Creëer een cultuur van interprofessioneel samenwerken waarin behandelaren, begeleiders en familieleden actief samenwerken met als gezamenlijke doel het kunnen herkennen en diagnosticeren van dementie bij mensen met (Z)EV(M)B.
- Minimaliseer personeelsverloop zoveel mogelijk (ondanks hoog ziekteverzuim en personele krapte) om ervoor te zorgen dat er begeleiders zijn die kennis hebben over het karakteristieke functioneren/gedrag van de persoon met (Z)EV(M)B.
- Optimaliseer dossiersystemen zodat geobserveerde veranderingen op een centrale en overzichtelijke plek gerapporteerd kunnen worden. Bied personeel training aan over hoe en waar ze veranderingen moeten rapporteren in het dossier.
- Bevorder onderzoek over dementie bij mensen met (Z)EV(M)B door samen te werken met kennisinstellingen.

Behandelaren

- Vergroot uw kennis over dementie bij mensen met (Z)EV(M)B om dementie beter te kunnen herkennen en diagnosticeren. Bezoek onze website: www.vb-dementie.nl.
- Gebruik het diagnostisch hulpmiddel voor dementie bij mensen met (Z)EV(M)B om tijdig een nulmeting uit te voeren. Focus daarbij op de vaardigheden en het (karakteristieke) gedrag dat iemand met (Z)EV(M)B vertoont. www.vb-dementie.nl/diagnostisch-hulpmiddel.
- Gebruik het diagnostisch hulpmiddel dementie voor mensen met (Z)EV(M)B als standaard onderdeel in het diagnostisch proces om dementiegerelateerde achteruitgang te identificeren en te monitoren over de tijd.
- Observeer regelmatig nogmaals, niet alleen door het herhaaldelijk invullen van het diagnostisch hulpmiddel, maar ook door daadwerkelijk zelf de persoon met (Z)EV(M)B te observeren.
- Zorg voor korte communicatielijnen waardoor begeleiders en familieleden makkelijk contact kunnen opnemen wanneer zij veranderingen observeren.
- Wees niet te terughoudend om vermoedens van dementie te delen met begeleiders en familieleden. Dit kan namelijk al bijdragen aan een beter begrip van wat iemand met (Z)EV(M)B laat zien. Verder kunnen op basis hiervan eerder geïnformeerde keuzes gemaakt worden.
- Maak gezamenlijk weloverwogen beslissingen over de stappen die wel of niet worden ondernomen in het diagnostisch onderzoek en rapporteer dit systematisch.

Begeleiders en familieleden

- Vergroot uw kennis over dementie bij mensen met (Z)EV(M)B om dementiesymptomen beter te kunnen herkennen. Bekijk onze website: www.vb-dementie.nl.
- Maak tijd om regelmatig te observeren en bespreek waargenomen (mogelijke dementie) veranderingen met andere begeleiders en familieleden.
- Rapporteer veranderingen in het dossier.
- Neem contact op met behandelaren wanneer u (mogelijke dementie) symptomen observeert en het gevoel heeft dat er iets niet klopt.

Conclusie

Het kan voorkomen dat we onze ogen sluiten wanneer dingen te complex worden. Dit leek ook het geval te zijn voor het onderzoek naar dementie bij mensen met (Z)EV(M)B. In plaats van het negeren van dit onderwerp, zijn wij in het project '*Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*' de uitdaging aangegaan om de complexe puzzel van dementie bij deze groep mensen te gaan ontrafelen. Hierbij hebben wij de tijd genomen om onze ogen te openen en nogmaals te observeren (**Observe once again**, de titel van dit proefschrift). Dit heeft geleid tot nieuwe kennis en inzichten over dementie bij mensen met (Z)EV(M)B, waardoor dementie beter en eerder herkend en gediagnosticeerd kan worden. Dit is belangrijk omdat een diagnose zorgt voor beter begrip en in staat stelt om geïnformeerde keuzes te kunnen maken. Zo kan de kwaliteit van bestaan van mensen met (Z)EV(M)B behouden worden.

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List with abbreviations | Lijst met afkortingen

A

A	Capacity/skill absent at baseline
AAMD	American Association on Mental Deficiency
Aβ	Amyloid-β
ADL	Activities of daily living | Algemene dagelijkse levensverrichtingen
AD	Alzheimer's disease
APP	Amyloid precursor protein
ASS	Formal diagnosis of Autism Spectrum Disorder

B

BHI	Behaviour History Inventory
BPSLD	British Present Psychiatric State-Learning Disabilities assessment
BPSD	Behavioral and Psychological Symptoms of Dementia
BPSD-DS	Behavioral and Psychological Symptoms of Dementia in Down Syndrome evaluation scale
BPSD-DS II	Behavioral and Psychological Symptoms of Dementia in Down Syndrome evaluation scale version II | Gedrags- en Psychologische Symptomen van Dementie bij DownSyndroom evaluatieschaal versie II

C

Ch.	Chapter
CAMDEX-DS	Cambridge Examination for Mental Disorders of Older People with Down's syndrome and Others with Intellectual Disabilities
CI	Confidence intervals
COREQ	Consolidated Criteria for Reporting Qualitative Research
CVA	Cerebrovascular accident

D

D	Daily

DASH	Diagnostic Assessment for the Severely Handicapped
DF & SR	Digits Forward and Sentence Recall subscales
DLD	Dementia Questionnaire for Learning Difficulties
DMR	Dementia Questionnaire for Persons with Mental Retardation
DS	Down syndrome | Downsyndroom
DSDS	Dementia Scale for Down Syndrome
DSI	Depression Status Inventory
DSVH	Adapted Dutch version of the Dementia Scale for Down Syndrome | Dementie Schaal voor mensen met een Verstandelijke Handicap
DTVMI	Developmental Test of Visual-Motor Integration
DQMRP	Dementia Questionnaire for Mentally Retarded Persons
DVZ	Dutch Dementia Questionnaire for persons with Mental Retardation | Dementie Vragenlijst voor Verstandelijk Gehandicapten

E

EU	European Union

F

FxS	Fragile X syndrome

G

GRAMMS	Good Reporting of A Mixed Methods Study
GMFCS	Gross Motor Function Classification System
GMP	Glossary of Mental Disorder
GPT	Grooved Pegboard Test
GQ	Gerontological Questionnaire subscales

H

H.	Hoofdstuk
hbo	Higher vocational education

I

ICD	International Classification of Diseases and Related Health Problems

ID	Intellectual disabilities
IQ	Intelligence quotient | Intelligentiequotiënt
IQR	Interquartile range | Interkwartielafstand

L

LIPS	Leiter International Performance Scale

M

M	Monthly
MACS	Manual Ability Classification System
MAS	Mood Assessment Scale for mental retardation
Max.	Maximum
mbo	Intermediate vocational education
METc	Medical Ethical Committee | Medisch Ethische Toetsingscommissie
Min.	Minimum

N

N/A	Not applicable

O

OR	Odds ratio

P

P	Capacity/skill present at baseline
PD	Picture Description test
PEG	Percutaneous endoscopic gastrostomy
PID	Profound intellectual disability
pp.	Per person
PPVT-R	Peabody Picture Vocabulary Test-Revised
PRISMA	Preferred Reporting Items for Systematic and Meta-Analyses

Q

Q	Quarterly

R

REDCap	Research Electronic Data Capture
RSMB	Reiss Screen for Maladaptive Behavior

S

SD	Standard deviation
SP(M)D	Severe/profound intellectual (and multiple) disabilities
SPI(M)D + D	People with severe/profound intellectual (and multiple) disabilities and diagnosed dementia
SPI(M)D + TD	People with severe/profound intellectual (and multiple) disabilities and questionable dementia
SPSS	Statistical Package for the Social Sciences
SRZ	Social competence Rating scale for people with intellectual disabilities | Sociale Redzaamheidsschaal voor verstandelijk gehandicapten

V

VABS	Vineland Adaptive Behavior Scale
VB	Verstandelijke beperking

W

W	Weekly
wo	Higher education
WS	Williams syndrome

U

UMCG	University Medical Centre Groningen | Universitair Medisch Centrum Groningen

Y

yrs	Years

Z

(Z)EV(M)B	(Zeer) ernstige verstandelijke (en meervoudige) beperkingen
(ZE)V(M)B + D	(Zeer) ernstige verstandelijke (en meervoudige) beperkingen + diagnose dementie
(Z)EV(M)B + TD	(Zeer) ernstige verstandelijke (en meervoudige) beperkingen + twijfelachtige dementie

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About the author

From an early age, the author **Maureen B.G. Wissing** was looking into the eyes of people with severe/profound (intellectual) and multiple disabilities. As she grew older, she became more curious to get to know these people. Since September 2019, her mission has been to improve the early recognition and diagnosis of dementia in people with severe/profound intellectual (and multiple) disabilities. Maureen is adept at bridging the gap between research, intellectual disability care, and education. Her collaboration with intellectual disability care organizations has enabled her to amass a wealth of knowledge and practical tools for recognizing and diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities. She has a knack for making these resources accessible to family members, students, and researchers in an engaging way. As a postdoctoral researcher, Maureen is committed to maintaining her focus on people with intellectual disabilities, persistently exploring and advocating for their needs through ongoing research projects.

Over de auteur

Van kleins af aan stond de auteur **Maureen B.G. Wissing** oog in oog met mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen. De nieuwsgierigheid om deze mensen beter te leren kennen groeide naarmate ze ouder werd. Vanaf september 2019 zet zij zich in om dementie beter te kunnen herkennen en diagnosticeren bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen. Maureen is een gedreven onderzoeker die wetenschap, de gehandicaptenzorg en onderwijs met elkaar weet te verbinden. Door nauw samen te werken met zorgorganisaties voor mensen met verstandelijke beperkingen heeft zij kennis en praktische handvatten ontwikkeld voor dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen. Ontwikkelde kennis en handvatten worden door haar op een creatieve manier onder de aandacht gebracht bij familieleden, zorgmedewerkers, studenten en onderzoekers. Maureen zal zich als postdoctoraal onderzoeker blijven inzetten voor meer kennis over mensen met verstandelijke beperkingen en het verbeteren van de gehandicaptenzorg.

Curriculum vitae

Education | Opleidingen

2019-2023 **PhD Dementia in people with severe/profound intellectual (and multiple) disabilities**
University of Groningen, University Medical Centre Groningen & Hanze University of applied sciences, Groningen

2017-2019 **MSc Human Movement Sciences, specialization in rehabilitation**
University of Groningen, Groningen

2014-2017 **BSc Human Movement Sciences**
University of Groningen, Groningen

2014-2017 **Pre-university education**
Almende College Isala, Silvolde

Professional appointments | Werkervaring

2023-present **Postdoctoral researcher**
Academic Collaborative Centre GOUD, Erasmus University Medical Center, Rotterdam

International presentations | Internationale presentaties

2022 **Presentation:** *Dementia in people with severe/profound intellectual (and multiple) disabilities*. (25 October). Dementia special interest group MacIntyre, online.

Presentation: *Motor symptoms of dementia in people with intellectual disabilities*. (30 March). Invitational conference International joint research group Move in Age, Groningen, The Netherlands.

2021 **Symposium:** *Dementia in people with severe/profound intellectual disabilities.* (8 July). IASSIDD Europe Congress 2021 | Amsterdam, online.

National presentations | Nationale presentaties

2023 **Presentation:** *Observeer nogmaals*: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (30 June & 1 July). Kennisdagen Alliade, Wolvega.

Presentation: *Observeer als begeleider/familielid nogmaals om dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen te herkennen.* (13 June). Koninklijke Visio, online.

Presentation: *Observeer nogmaals*: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (12 June). 's Heeren Loo, online.

Presentation: *Observeer nogmaals*: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (25 May). Ipse de Bruggen, online.

Presentation: *Observeer als gedragskundige/ psychodiagnostisch medewerker nogmaals om dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen te herkennen.* (11 May). Koninklijke Visio, online.

Presentation: *Observeer nogmaals*: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (11 May). Koninklijke Visio, online.

Presentation: *Diagnostisch hulpmiddel dementie bij mensen met (Z)EV(M)B voor gedragskundigen en psychodiagnostisch medewerkers.* (18 April). Verstandelijke beperking & dementie, online.

2022 **Lecture:** *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (15 September). Kenniskring ergotherapie, online.

Pitch: *Breinbreker. Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (19 May). Vlootschouw Alliade, online.

Poster: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (15 November). Lustrum lectoraat Healthy Ageing, Allied Health Care and Nursing, Groningen.

Presentations: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (11 November). Verstandelijke beperking & dementie: Eindcongres dementie bij mensen met (Z)EV(M)B, Wolvega.

Presentation: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (11 October). Dementietafel Groningen-Drenthe, Roden.

Presentation: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (4 October). Nederlandse Vereniging voor Fysiotherapie voor Verstandelijk Gehandicapten: Fysiotherapeut in de VG, een wereld te winnen, Nijkerk.

Presentation: *Klein kijken*: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (2 June). Kennisdag Alliade, Wolvega.

Workshop: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (5 July). Hart voor Zorg congres: Masterclass Gehandicaptenzorg, Zwolle.

2021 **Lecture:** *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*. (16 November). Kringbijeenkomst logopedisten, online.

Lecture: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*. (6 April). Landelijke dag Netwerk Gedragsdeskundigen Ouderen, online.

Lecture: *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*. (24 March). Regioavond artsen verstandelijk gehandicapten, online.

Pitch: *Onderzoek: Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen* (25 March). Vlootschouw Alliade, online.

Presentation: *Dossieronderzoek dementie & verstandelijke beperking*. (15 March). (H)erken jij dementie bij mensen met een verstandelijke beperking?, online.

Workshop: *Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*. (17 November). Vilans 'Zoek het uit! Praktijk en wetenschap dichter bij elkaar', online.

Workshop: *Wie ziet dat ik dingen vergeet? Over dementie!* (19 November). Academische werkplaats EMB: Perspectieven op samenwerking bij EMB, online.

2020 **Workshops:** *Herkent u dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen? & (H)erkennen van dementie: wat is het belang?* (27 February). Pronkjewailtjes uut Noorden, Groningen.

Conference organization | Congresorganisatie

2022 *Eindcongres dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* Verstandelijke beperking & dementie, Wolvega, The Netherlands.

2020 *Pronkjewailtjes uut Noorden.* University of Groningen, University Medical Centre Groningen & Hanze University of Applied Sciences, Groningen, The Netherlands.

Webinars

2022 *Dementie bij mensen met (Z)EV(M)B.* Hart voor Zorg.
www.hartvoorzorg.nl/videos/dementie-bij-mensen-met-zevmb/

Inleiding in dementie bij mensen met VB + dossieronderzoek. (H)erken jij dementie bij mensen met een verstandelijke beperking.
www.youtube.com/watch?v=YFjhFoQiQP0&list=PLXR0MWZt3YIgoTcPdfJilRlFq-H3HWhUF&index=3

Praktijkkennis symptomen van dementie bij mensen met (Z)EV(M)B. (H)erken jij dementie bij mensen met een verstandelijke beperking.
www.youtube.com/watch?v=yZTSrQx9NDw

Media interviews

2022 *Kennismodules dementie bij (Z)EV(M)B.* Kennisplein gehandicaptensector.
www.kennispleingehandicaptensector.nl/tips-tools/tools/kennismodules-dementie-z-ev-m-b

Hoe ernstiger de beperking, hoe moeilijker je dementie herkent. Hart voor Zorg.
www.hartvoorzorg.nl/nieuws/hoe-ernstiger-de-beperking-hoe-moeilijker-je-dementie-kan-herkennen/

*Dementie vaststellen bij mensen met een verstandelijke beperking: 4 tips. K*ennisplein gehandicaptensector.
www.kennispleingehandicaptensector.nl/actueel/verhalen/dementie-bij-mensen-verstandelijke-beperking

Bij mensen met (Z)EV(M)B is het 'kleine kijken' belangrijk. Kennisplein gehandicaptensector.
www.kennispleingehandicaptensector.nl/actueel/verhalen/bij-mensen-met-z-ev-m-b-is-het-kleine-kijken-belangrijk

2021 *Dementie (h)erkennen bij mensen met (Z)EV(M)B.* Kennisplein gehandicaptensector. www.kennispleingehandicaptensector.nl/actueel/verhalen/dementie-h-erkennen-bij-mensen-met-z-ev-m-b

Newsletters | Nieuwsbrieven

2019-2022 *Project status updates: Praktijkvragen over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* Verstandelijke beperking & dementie. www.vb-dementie.nl/project-updates/

Product development | Productontwikkeling

2019 - 2023 **Diagnostic aid:** *Diagnostisch hulpmiddel dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen voor gedragskundigen en psychodiagnostisch medewerkers.* www.vb-dementie.nl/diagnostisch-hulpmiddel

Factsheet: *Eindcongres dementie bij mensen met (Z)EV(M)B – Projectoverzicht.* www.vb-dementie.nl/downloads

Online training product: *Kennismodules over dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* www.vb-dementie.nl/kennismodules

Powerpoint sheets: *Diareeks onderwijs modules 1-8.* www.vb-dementie.nl/downloads

Website: *Verstandelijke beperking & dementie* www.vb-dementie.nl

Teaching | Onderwijservaring

2022 **Guest lecture:** *Klein kijken: dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen*. (1 December) Minor Healthy Ageing mbo-hbo-wo, Groningen, The Netherlands.

Workshop: *Developing a new dementia screening instrument for people with severe/profound intellectual (and multiple) disabilities.* (14 June). Faith Summerschool, Leeuwarden, The Netherlands.

2021 **Guest lecture:** *Dementie bij mensen met (zeer) ernstige verstandelijke (en meervoudige) beperkingen.* (2 December). Minor Healthy Ageing mbo-hbo-wo, Groningen, The Netherlands.

Student supervision | Studentbegeleiding

2021 **Master project nursing science**: *Prevalence of pseudodementia in older adults with a severe or profound intellectual disability: a systematic review.* Utrecht University.

2020 **Bachelor thesis nursing**: *Dementie cormorbiditeiten en psycho-farmacagebruik bij mensen met een (zeer) ernstige verstandelijke (en meervoudige) beperking.* Hanze University of applied sciences.

Bachelor thesis physiotherapy: *Welke motorische veranderingen zijn zichtbaar bij mensen een (zeer) ernstige verstandelijke (en meervoudige) beperking met (een vermoeden van) dementie?*. Hanze University of applied sciences.

Master thesis physician assistant: *Brainwave, epilepsie bij een verstandelijke beperking en dementie*. Hanze University of applied sciences.

2019 **Bachelor project medicine**: *Psychoactive medication use in people with severe/profound intellectual disability.* University of Groningen.

List of publications | Lijst met publicaties

International publications | Internationale publicaties

Wissing, M. B. G., Hobbelen, J. S. M., De Deyn, P. P., Waninge, A., & Dekker, A. D. (2023). Dementia in people with severe/profound intellectual (and multiple) disabilities, and its natural history. *Journal of Mental Health Research in Intellectual Disabilities*. https://doi.org/10.1080/19315864.2023.2240734

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