Prevalence of dementia in people with intellectual disabilities: cross-sectional study

Running title: Dementia and MCI in intellectual disabilities

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ABSTRACT

Background: There are only a few studies of the prevalence of dementia in people with intellectual disability (ID) without Down syndrome (DS), and there is a large difference in the prevalences between reported studies. Moreover, the prevalence of mild cognitive impairment (MCI) in ID has not been reported. We aimed to evaluate the prevalence of dementia in adults of all ages and the prevalence of MCI in people with ID. Further, we tried to clarify the differences depending on the various diagnostic criteria.

Methods: The survey included 493 adults with intellectual disability at 28 facilities in Japan. The caregivers answered a questionnaire, and physicians directly examined the participants who were suspected of cognitive decline. Dementia and MCI were diagnosed according to ICD-10, DC-LD, and DSM-5 criteria.

Results: The prevalence of dementia was 0.8% for the 45–54 year old group, 3.5% for the 55–64 year old group, and 13.9% for the 65–74 year old group in people with ID wihout DS. The prevalence of MCI was 3.1% for patients 45–54, 3.5% for patients 55–64, and 2.8% for patients 65–74 with ID wihout DS.

DSM-5 was the most inclusive in diagnosing dementia and MCI in people with ID.

Conclusions: People with ID without DS may develop dementia and MCI at an earlier age and higher rate than the general population. Among the diagnostic criteria, DSM-5 was the most useful for diagnosing their cognitive impairment.

KEYWORDS

dementia, intellectual disability, mental retardation, mild cognitive impairment (MCI), prevalence of dementia

Key points

- There are only a few studies of the prevalence of dementia and mild cognitive impairment (MCI) in people with intellectual disability (ID) without Down syndrome (DS).
- This study investigated 493 adults with ID. The physicians directly examined the participants suspected of cognitive decline and diagnosed them according to various diagnostic criteria (ICD-10, DC-LD, and

DSM-5).

- The prevalence of dementia in people with ID without DS may be higher than in the general population. DSM-5 was more inclusive in diagnosing dementia and MCI in people with ID than ICD-10 and DC-LD.

1 INTRODUCTION

The average life expectancy of people with intellectual disability (ID) has increased remarkably in recent years, and not a few people with ID live to over the age of 65.[1] Along with the aging of people with ID, the problem of dementia in people with ID has become important.[2] However, it is not easy to screen for dementia in people with ID. Screening tests for dementia in the general population such as the Mini Mental State Examination (MMSE) are not useful for screening for dementia in the ID population.[3] As one reason, most people with ID have difficulty in achieving more than the cut-off score on the screening tests even before the appearance of cognitive deterioration.[4] In addition, people with ID have various degrees of pre-existing cognitive impairment, and there are generally no reference data or thresholds for screening tests of dementia in ID.[5,6] In order to diagnose dementia in people with ID, it is necessary to compare the present state of their cognitive function and daily living activities with their highest level in the past. Therefore, we need information from family members or care staff who have known the person with ID for a long time.[7,8] Because it takes much time and effort, there are not many epidemiological

studies of dementia in people with ID. From the viewpoint of dementia in people with ID, Down syndrome (DS) has attracted much attention for a long time. DS, trisomy 21, is a genetic disorder, and the most common cause of ID among people whose causes are known.[9] It is well known that people with DS suffer from Alzheimer disease dementia frequently after the age of 40 due to overload of amyloid beta protein.[10] On the other hand, it is not clear whether the frequency of dementia in people with ID who do not have DS is higher than that in the general population. In the past 30 years, only two studies have been done to investigate the prevalence of dementia by age in people with ID without DS, and no unified conclusion on the prevalence of dementia in people with ID without DS has been reached. Zigman *et al*. reported that the prevalence of dementia in people with ID without DS is similar to that in the general population in New York State.[11] On the other hand, Strydom *et al*. showed that the prevalence of dementia in people with ID without DS is higher than that in the general population in London.[12,13] Various differences in the survey methods adopted in those studies are thought to affect the differences in the results.[14] The differences of methods include study design (cross-sectional or longitudinal),

residential environment (community- or institution-based), subjects (inclusion or exclusion of DS), diagnostic method (physician's diagnosis or chart record), and diagnostic criteria (clinical diagnosis or using diagnostic criteria).[11-13,15-17] In order to clarify the prevalence of dementia and mild cognitive impairment (MCI) in an ID population without DS, we conducted an epidemiological survey targeting a large number of adults with ID in Japan.

2 MATERIALS AND METHODS

2.1 Ethics

This study was approved by the Internal Ethical Committee of the Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences (1708-044) and Asahigawaso Research Institute. This study was registered at The University Hospital Medical Information Network Clinical Trials Registry (UMIN000028708) on 11 November 2017. We provided all participants with simple written explanations of this research, taking into consideration the cognitive impairment of participants. After giving a complete description of the study to the subjects and their relatives, written informed consent was obtained from the subjects who were judged to have the ability to express consent. In addition, written informed consent was obtained from their relatives in all cases.

2.2 Participants

In the studies performed in Europe and the United States, researchers recruited participants using registration data managed by local governments and

conducted a survey on the prevalence of dementia in people with ID.[11,12] On the other hand, people with ID are not registered in Japan. Therefore, we conducted an epidemiological survey at many support facilities that provide services for people with ID in Okayama Prefecture. Okayama is a prefecture of Japan located in the western region of the main island. Pariticipants were recruited from the users of the support facilities in November 2017 according to the following criteria. In total, 28 support facilities agreed to participate. Facility residents and home-based residents using day service at facilities were included, and the subjects fulfilled the following inclusion criteria. (i) The subject was diagnosed with intellectual disbility according to the criteria formulated by ICD-10: a condition of reduced overall level of intelligence (IQ<70) that manifested during the developmental period.[18] (ii) There were information providers who had observed the living condition of the subject for two years or more, and the information providers agreed to respond to the interview and answer the questionnaire survey. (iii) Informed consent was obtained from participants who had the capacity to consent and the relatives of all participants. (iv) The subject was 20 years or older. (v) Down syndrome was identified from

records of chromosomal analysis or by their characteristic features.[14]

2.3 Screening

We designated the service providers who were involved with participants for more than two years and who knew the changes in the daily living activities of participants as ‘informants’. All informants completed the Japanese version of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID).[19] In addition, they completed the Japanese version of the Physical Self-Maintenance Scale (PSMS) and Lawton’s Instrumental Activities of Daily Living Scale (IADL) as performance-based measures of activities of daily living.[8,20] Three professional physicians interviewed all the informants in person, and they recorded detailed information of each participant over the past few years. The three physicians were STa (geriatric psychiatrist), STe (specialist on dementia), and RK (geneticist). The doctors judged participants who satisfied one or more of the two following conditions as screening positive: (i) at least one of the three doctors suspected the possibility that the cognitive function and/or ADL of the participant had deteriorated in the past few years,

and (ii) worsening of the score of PSMS or IADL in the past few years was recognized. The three doctors examined everyone who was positive in the screening. Any disagreement on diagnosis between the three doctors was settled by discussion.

2.3.1 Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)

DSQIID is an observer-rated dementia screening questionnaire that is completed by carers who have known the subject for at least six months.[8] DSQIID was developed on the assumption that it would be used for adults with ID. Although the validity of DSQIID was evaluated only in adults with DS, the authors assert that DSQIID can be equally useful in ID adults without DS.[8] The DSQIID consists of 53 items. The DSQIID comprehensively reflect symptoms of dementia, including cognitive functions such as loss of memory and speech abnormalities, behavioural changes, psychological symptoms, and physical symptoms.

2.3.2 Physical Self-Maintenance Scale (PSMS)

PSMS is an informant-reported measure to evaluate the level of basic activities of daily living. Each of six basic activities of daily living (toileting, feeding, dressing, grooming, ambulation, and bathing) is rated 0–1 point. The maximum PSMS score is 6 points.[20]

2.3.3 Lawton's Instrumental Activities of Daily Living Scale (IADL)

The IADL scale was developed to assess the more complex activities necessary for functioning in community settings. Each of eight activities (e.g. shopping and cooking) is rated 0–1 point.[20] Three tasks, cooking, housekeeping, and laundry, were scored only for females in the original IADL. For this reason, the maximum score of the original IADL was 8 for females and 5 for males. In this study, the IADL scores of males were calculated by multiplying the original IADL score x 1.6. Therefore, in this study, the IADL has a maximum score of 8 for both males and females.

2.4 Diagnosis

In diagnosing dementia, we used three criteria: ICD-10 Research Diagnostic Criteria (World Health Organization) for dementia, Diagnostic Criteria for Psychiatric Disorders for Use with Adults with Learning Disabilities/Mental Retardation (DC-LD) (Royal College of Psychiatrists) for dementia, and DSM-5 criteria (American Psychiatric Association) for neurocognitive disorder.[7,18,21] We used ICD-10 Research Diagnostic Criteria for mild cognitive disorder and DSM-5 criteria for mild neurocognitive disorder as diagnostic criteria for MCI. We diagnosed a participant who satisfied at least one of these criteria with dementia or MCI. We used the National Institute on Aging-Alzheimer's Association workgroups (NIA-AA) criteria for Alzheimer's disease (AD) dementia, the American Heart Association/American Stroke Association (AHA/ASA) criteria for vascular dementia (VaD), the 2017 Consortium on Dementia with Lewy Bodies (DLB) criteria for DLB, and the International Consensus Criteria for Behavioural Variant FTD (FTDC) for behavioral variant FTD.[22-25]

2.5 Statistical analysis

Statistical analysis was performed using the SPSS 24.0 J software program (SPSS Inc., Chicago, IL). Comparisons between two groups were performed by independent sample *t*-tests. Chi-square tests were used to analyse categorical variables with continuity correction for 2×2 tables. The significance level was set at $P<0.01$ owing to the number of tests.

3 RESULTS

3.1 Demographic and clinical features

There were 909 users of the facilities. Of these, 118 of 909 (13.0%) cases were excluded from the study because they were less than 20 years old or they did not have ID but a mental disorder. The remaining 791 cases were potential participants. Of 791 subjects, 493 (62.3%) agreed to participate. Of 493 participants, 34 (6.9%) were people with ID with DS, and 459 (93.1%) were people with ID without DS. The mean age of all the participants was 46.57 (SD: 11.43 years; range: 19–83 years). The prevalences of current psychiatric disorders were: autism spectrum disorder, 92 (18.7%); psychiatric symptoms related to epilepsy, 9 (1.8%); schizophrenia, 5 (1%); bipolar disorder, 3 (0.6%); obsessive-compulsive disorder, 1 (0.2%); and others, 13 (2.6%). The mean age of 34 participants with DS was 39.41 (SD: 13.93 years; range: 20–65 years).

3.2 Participants of dementia and MCI

Seven of 34 (20.6%) participants with Down syndrome had dementia. All seven patients were classified as probable AD dementia. Cases of dementia have been

seen from the age of 45 years. There were no participants with DS diagnosed with MCI. The mean age of 459 participants without DS was 47.10 (SD: 13.98 years; range: 20–83 years). Ten of 459 (2.2%) participants had dementia. Six out of 10 (60.0%) patients with dementia were classified as probable AD dementia, two (20.0%) as probable VaD, and two (20.0%) as possible AD dementia and possible DLB. Nine of 459 (2.0%) had MCI. The age-specific prevalences of dementia and MCI are shown in Table 1. With regard to the severity of ID, there were no patients with dementia and MCI among the participants who were rated as having mild ID. Two of 135 (1.5%) people with moderate ID and 15 of 298 (5.0%) people with severe ID were diagnosed with dementia. Four of 135 (3.0%) people with moderate ID and 5 of 298 (1.7%) people with severe ID were diagnosed with MCI (Table 2).

3.3 Diagnosis of dementia and MCI, differences by diagnostic criteria

Seventeen of all participants met the criteria for dementia of at least one of DSM-5, ICD-10, or DC-LD. All 17 dementia patients met the DSM-5 criteria, 14 (82.4%) met the DC-LD criteria, and 13 (76.5%) met the ICD-10 criteria.

Seven of 17 dementia patients were people with DS, and they met the criteria for dementia of all three (DSM-5, ICD-10, and DC-LD). Ten of 17 (58.8%) dementia patients were people without DS, and they met the criteria of one or more of DSM-5, ICD-10, or DC-LD for dementia. All 10 patients met the DSM-5 criteria, 7 (70.0%) met the DC-LD criteria, and 6 (60.0%) met the ICD-10 criteria. Nine participants met at least one of the DSM-5 or ICD-10 criteria for MCI. All nine patients diagnosed with MCI were people without DS; they all met the DSM-5 criteria, and five (55.6%) met the ICD-10 criteria.

4 DISCUSSION

4.1 Prevalence of dementia and MCI in people with intellectual disability

When discussing studies on the prevalence of dementia in people with ID, it is necessary to classify them into studies targeting people with ID with DS and studies targeting people with ID without DS. In this study, people with DS over the age of 45 had a high rate of dementia. The prevalence of dementia in people with DS was significantly higher than that in the general population, as reported in many previous studies in Europe, the United States, and Japan.[26-28] There have been a few studies on the prevalence of dementia in people with ID without DS, although the number of those studies is lower than the number of studies on the prevalence of dementia in people with DS. Figure 1 shows the results of the previous studies and this study. For reference, we compared the result of our study (65–74 years, 13.9%) with the results of a large-scale epidemiological study targeting the general elderly population in Japan (65–74 years, 4.2%), and found that the prevalence of dementia in people with ID without DS is high in the young age group.[29] There are large differences between the dementia prevalences in several studies. Strydom *et al.* reported

that the prevalence of dementia in people with ID without DS was 2–3 times higher than that in the general population, whereas Zigman *et al*.[11-13] reported that there was no difference in the prevalence of dementia between the ID population without DS and the general population. The prevalence in our research is roughly in the middle of these two studies. The major difference between the study of Strydom *et al*. and ours is estimated to be due to selection bias.[12-13] Although both studies included facility residents and home-based residents using a day service, Strydom *et al*. recruited the participants from a service for people with ID and a medical service for elderly people. On the other hand, we recruited the participants from a service for people with ID only. Furthermore, in Japan, people who need nursing care and are unable to remain in a facility for people with ID are moved to a facility dedicated to the elderly. Among those who withdraw from facilities for people with ID in a year, 30.9% are due to death and 22.2% are due to movement to hospitals and facilities for elderly patients.[30] Therefore, in this study, there is a possibility that the evaluated prevalence of dementia was lower than the actual, especially in the elderly age band.

Further, in this study, there is the contradiction that the prevalence of dementia is lower in the elderly group than in the younger group from the age of 65 to 84. The prevalence of dementia is lower at 75–84 years (8.3%, n=12) compared to 65–74 years (13.9%, n=36) (Table 1). The cause of this contradiction is presumed to be selection bias as well. The cases of dementia needing nursing care tend to move from a facility for people with ID to a facility for the elderly. In this study, it is presumed that the prevalence of dementia in the age group older than 75 years was lower than in the younger age band for this reason. The major difference between the study of Zigman *et al*. and ours is the methods to detect dementia.[11] Silverman *et al*. said that the cause of the difference between the study of Strydom *et al*. and that of Zigman *et al*. is the different methods used to evaluate and classify cases.[31] In our research, we classified dementia based on defined diagnostic criteria such as ICD-10 and DSM-5, and this method is similar to the study of Strydom *et al*.[12] On the other hand, Zigman *et al*. did not classify dementia based on defined diagnostic criteria, and they limited dementia cases to Alzheimer's disease.[11] The cause of the lower prevalence in the study of Zigman *et al*. than our study

may derive from the difference in diagnostic methods. Unlike previous studies that were limited to the elderly over 65 years, this study investigated all age groups from the age of 20 years. Although the number is small, it was found that dementia and MCI first occurred in the late forties.

4.2 Differences according to diagnostic criteria for dementia and MCI

Because the concept of dementia depends on the criteria, the prevalence of dementia in the general population varies depending on the diagnostic criteria of dementia.[32,33] Some cross-sectional studies have reported that the prevalence of dementia defined by DSM-5 is higher than that by DSM-IV. [34,35] Even in people with ID, the prevalence varies with variations of criteria.[12,36] In a previous study, three diagnostic criteria for dementia (DSM-IV, DC-LD, ICD-10) were used.[12] Strydom *et al*. reported some cases with no duplication of multiple diagnostic criteria, and ICD-10 dementia criteria missed dementia cases of moderate severity in this population.[12] For this reason, we used three diagnostic critera for dementia (DSM-5, DC-LD, ICD-10). As a result, the number of dementia cases covered by each diagnostic criterion differed, and DSM-5 was able to diagnose

the most cases (Figure 2). Memory impairment is essential for diagnosis in ICD-10 or DC-LD, but it was difficult to confirm the function of memory in some cases due to difficulties in conversations and activities. On the other hand, memory impairment is not indispensable in DSM-5. It is possible to diagnose dementia using DSM-5 based on the decline of one of multiple cognitive domains (Table 3). It has been reported that DSM-5 can detect more cases of dementia without memory impairment, language impairment, and decline of instrumental activities of daily living (IADL) than DSM-IV. [34] To extensively diagnose dementia in people with ID, DSM-5 may be suitable. For the diagnosis of MCI, we used DSM-5 and ICD-10. DSM-5 diagnosed more MCI cases than ICD-10. As one reason, impairment of executive function is essential for diagnosis in ICD-10, but it was difficult to confirm in people with severe ID.

4.3 Difference in prevalence of dementia and MCI due to severity of ID

Regarding the severity of mental retardation and the risk of developing dementia, Strydom *et al*. has said the prevalence of dementia was not influenced by ID level.[14] On the other hand, many of the cases of dementia were found in

patients with severe ID, followed by many cases with moderate ID, but dementia was not observed in patients with mild ID in this study. As in the study of Strydom *et al*., there is no significant difference in the median age between the three groups with severe, moderate, and mild ID in this study. Strydom *et al*. mentioned that potential cases that did not have sufficient information for diagnosis were more common in participants with severe ID compared to mild or moderate ID.[14] The more severe the ID, the more difficult it is to evaluate the objective cognitive function. Strydom *et al*. used ICD-10 and DSM-IV as diagnostic criteria.[14] In diagnosing dementia using these criteria, unlike DSM-5, confirmation of memory impairment is indispensable. Even in the study by Strydom *et al*., it may have been possible to diagnose dementia in more cases using DSM-5.[14] The cognitive reserve hypothesis that people with lower brain reserve are more likely to develop dementia has been long proposed.[37] Therefore, it is not surprising that the prevalence of dementia increases as the severity of ID increases.

4.4 Limitations

There are two limitations to this study. Firstly, there is possibility that this sample does not reflect the general population of people with ID precisely. Since there was no other way to recruit a large group of people with ID, we recruited in facilities for ID in this study. However, if there is a database in which all people with ID in the area are registered, it is best to conduct surveys based on those databases. There may be some people with ID do not use this type of social service. The reason why we did not have a participant over the age of 85 in this study may be because they had been moved to facilities for the elderly with dementia. In Japan, which service is used by elderly people with ID is decided case by case. Second, this study is a cross-sectional assessment, which is less reliable than a longitudinal assessment. It is desirable to evaluate sequentially cognitive function by test batteries for people with ID such as CAMDEX-DS.[38]

4.5 Conclusions

In summary, this is the first study of people with ID without DS to investigate the prevalence of dementia and to report the prevalence of MCI by age. People

with ID without DS may develop dementia and MCI at an earlier age and higher rate than the general population. DSM-5 is thought to be the most useful among ICD-10, DC-LD, and DSM-5 in diagnosing dementia in people with ID, and DSM-5 is more useful than ICD-10 for diagnosing MCI. ICD-10 and DC-LD require a decline of specific cognitive domains such as memory and executive function, but DSM-5 can diagnose dementia and MCI based on a single decline in multiple cognitive domains and thus can detect more cases.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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Table 1. Demographic details of participants

Demographic	All participants	DS	without DS	*p*
Total (n)	493	34	459	
Age (mean years ± SD)	46.57±14.09	39.41±13.93	47.10±13.98	<0.010
20-44	212	21	191	
45-54	141	7	134	
55-64	90	4	86	
65-74	38	2	36	
75-84	12	0	12	
Sex (n) (male/female)	311/182	18/16	293/166	0.204
Education (mean years ± SD)	10.10±2.52	10.36±3.36	10.08±2.45	0.241
Type of residence				
Independent or group home (n) (%)	201 (40.7)	19 (55.9)	182 (39.7)	0.063
Facility residents (n) (%)	292 (59.3)	15 (44.1)	277 (60.3)	
Severity of ID				
Mild ID (n) (%)	60 (12.2)	2 (5.9)	58 (12.6)	0.245
Moderate ID (n) (%)	135 (27.4)	12 (35.3)	123 (26.8)	0.284
Severe ID (n) (%)	298 (60.4)	20 (58.8)	278 (60.6)	0.841
DSQIID (mean ± SD)	2.50±5.89	7.68±13.29	2.11±4.71	<0.010
PSMS (mean ± SD)	2.48±1.91	2.18±1.69	2.51±1.92	0.390
IADL (mean ± SD)	1.87±1.77	2.07±1.68	1.85±1.78	0.560
Psychiatric disorders (n) (%)	124 (25.2)	1 (2.9)	123 (26.8)	<0.010
Epilepsy (n) (%)	175 (35.5)	4 (11.8)	171 (37.3)	<0.010
Visual probrems (n) (%)	19 (3.9)	0 (0)	19 (4.1)	0.633
Hearing problems (n) (%)	12 (2.4)	4 (11.8)	8 (1.7)	<0.010
Mobidity problems (n) (%)	59 (12.0)	3 (8.9)	56 (12.2)	0.785
Dementia (n) (%)				
20-44	0 (0)	0 (0)	0(0)	-
45-54	4 (2.8)	3 (42.9)	1 (0.8)	<0.010
55-64	5 (5.6)	2 (50.0)	3 (3.5)	<0.010
65-74	7 (18.4)	2 (100.0)	5 (13.9)	<0.010
75-84	1 (8.3)	-	1 (8.3)	-
MCI (n) (%)				
20-44	0 (0)	0 (0)	0 (0)	-
45-54	4 (2.8)	0 (0)	4 (3.1)	-
55-64	3 (3.3)	0 (0)	3 (3.5)	-
65-74	1 (2.6)	0 (0)	1 (2.8)	-
75-84	1 (8.3)	-	1 (8.3)	-

SD, standard deviation; ID, intellectual disability; DS/without DS, subjects with Down syndrome and without Down syndrome; DSQIID, the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; PSMS, Physical Self-Maintenance Scale; IADL, Lawton's Instrumental Activities of Daily Living
P-value is comparison between DS and without DS.

Table 2. Difference in prevalence of cognitive impairment due to severity of intellectual disability

	mild ID	moderate ID	severe ID
Total (n)	60	135	298
DS/without DS	2/58	12/123	20/278
Age (mean years ± SD)	46.85±15.82	49.25±14.19	45.30±13.47
dementia	0	2	15
MCI	0	4	5

SD, standard deviation; ID, intellectual disability; DS/without DS, subjects with Down syndrome and without Down syndrome

Table 3. Differentiation of diagnostic criteria for dementia

	ICD-10	DC-LD	DSM-IV	DSM-5†
Memory impairment	+	+	+	○
Higher cortical functions				
Executive function			○	○
Thinking	○	○		
Judgment	○	○		
Other cognitive skills		○		
Information processing	○			
Aphasia, Language skills			○	○
Apraxia			○	
Agnosia			○	
Complex attention				○
Perceptual-motor				○
Social cognition				○
Behavioural and emotional function				
Emotional lability	△	△		
Irritability	△	△		
Apathy	△	△		
Coarsening of social behaviour	△	△		
Other criteria				
Change from premorbid state/decline in level of functioning		+	+	
Duration of at least 6 months	+			
Cognitive deficits interfere with independence				+
Exclusions				
Not caused by delirium	+	+	+	+
Not caused by mental illness or physical illness		+	+	+

†major neurocognitive disorder

+, required for diagnosis

Figure Legends

Figure 1. Studies of prevalences of dementia in people with intellectual disability without Down syndrome

N/M, not mentioned

Figure 2. Venn diagram of differences by diagnostic criteria

DSM-5 was able to diagnose the most cases of dementia and MCI.

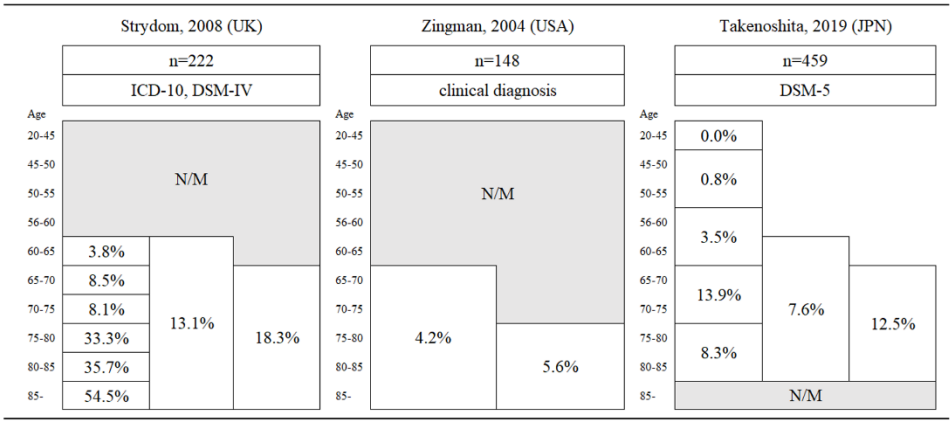
Strydom, 2008 (UK)
n=222
ICD-10, DSM-IV
Age
20-45
45-50
50-55
56-60
60-65
65-70
70-75
75-80
80-85
85-
N/M
3.8%
8.5%
8.1%
33.3%
35.7%
54.5%
13.1%
18.3%
Zingman, 2004 (USA)
n=148
clinical diagnosis
Age
20-45
45-50
50-55
56-60
60-65
65-70
70-75
75-80
80-85
85-
N/M
4.2%
5.6%
Takenoshita, 2019 (JPN)
n=459
DSM-5
Age
20-45
45-50
50-55
56-60
60-65
65-70
70-75
75-80
80-85
85-
0.0%
0.8%
3.5%
13.9%
8.3%
N/M
7.6%
12.5%

Figure 1

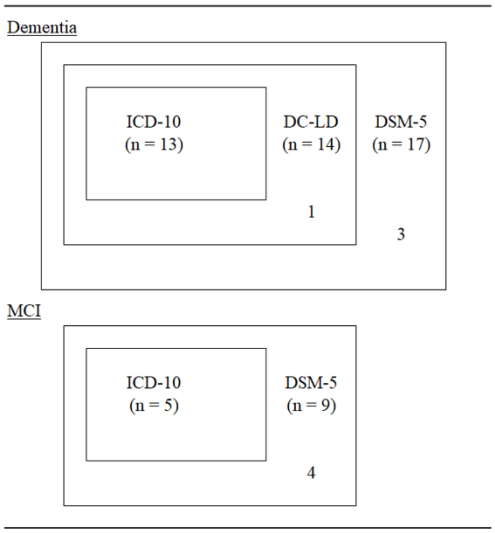

Figure 2

Table 1. Demographic details of participants

Demographic	All participants	DS	without DS	*p*
Total (n)	493	34	459	
Age (mean years ± SD)	46.57±14.09	39.41±13.93	47.10±13.98	<0.010
20-44	212	21	191	
45-54	141	7	134	
55-64	90	4	86	
65-74	38	2	36	
75-84	12	0	12	
Sex (n) (male/female)	311/182	18/16	293/166	0.204
Education (mean years ± SD)	10.10±2.52	10.36±3.36	10.08±2.45	0.241
Type of residence				
Independent or group home (n) (%)	201 (40.7)	19 (55.9)	182 (39.7)	0.063
Facility residents (n) (%)	292 (59.3)	15 (44.1)	277 (60.3)	
Severity of ID				
Mild ID (n) (%)	60 (12.2)	2 (5.9)	58 (12.6)	0.245
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45-54	4 (2.8)	3 (42.9)	1 (0.8)	<0.010
55-64	5 (5.6)	2 (50.0)	3 (3.5)	<0.010
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75-84	1 (8.3)	-	1 (8.3)	-
MCI (n) (%)				
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45-54	4 (2.8)	0 (0)	4 (3.1)	-
55-64	3 (3.3)	0 (0)	3 (3.5)	-
65-74	1 (2.6)	0 (0)	1 (2.8)	-
75-84	1 (8.3)	-	1 (8.3)	-

SD, standard deviation; ID, intellectual disability; DS/without DS, subjects with Down syndrome and without Down syndrome; DSQIID, the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; PSMS, Physical Self-Maintenance Scale; IADL, Lawton's Instrumental Activities of Daily Living
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Information processing	○			
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Apraxia			○	
Agnosia			○	
Complex attention				○
Perceptual-motor				○
Social cognition				○
Behavioural and emotional function				
Emotional lability	△	△		
Irritability	△	△		
Apathy	△	△		
Coarsening of social behaviour	△	△		
Other criteria				
Change from premobid state/decline in level of functioning		+	+	
Duration of at least 6 months	+			
The cognitive deficits interfere with independence				+
Exclusions				
Not caused by delirium	+	+	+	+
Not caused by mental illness or physical illness		+	+	+

†major neurocognitive disorder

+, required for diagnosis

At least one circle and one triangle is required