Validation of the Japanese Version of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID-J)

Running title: Validation of the Japanese version of DSQIID

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Conflict of Interest

No conflicts of interest have been declared.

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ABSTRACT

Background

Dementia in people with intellectual disabilities (ID) is difficult to detect because of preexisting cognitive deficits. An effective screening method is required. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was developed as an observer rating tool to screen dementia in people with ID. The aim of this study was to verify the screening accuracy of the DSQIID for Japanese people with ID.

Methods

Four-hundred ninety-three subjects with ID participated in this study. Caregivers who had observed the participants for more than two years scored the Japanese version of the DSQIID (DSQIID-J) of the participants. Three doctors examined participants directly and diagnosed dementia using the DSM-5 criteria. To identify the key screening items that predict dementia, the specificities of a single and pairs of items with 100% sensitivity were evaluated relative to the dementia diagnosis.

Results

Of 493 participants, 34 were people with Down syndrome (DS), and 459 were people without DS. Seventeen participants were diagnosed with dementia. The suitable cut-off score of the DSQIID-J was 10/11 (sensitivity 100%, specificity 96.8%) for screening dementia. The inter-rater reliability, test-retest reliability, and internal consistency of the DSQIID-J were excellent. Regarding key items, there was no single item with 100% sensitivity, and the best 2-item combination was the pair of "Cannot dress without help" and " Walks slower" (sensitivity 100%, specificity 93.5%).

Conclusions

We identified several important question items of the DSQIID-J related to the diagnosis of dementia in people with ID. The DSQIID-J is a useful screening tool for dementia in adults with ID.

Keywords

cognitive impairment, dementia, DSQIID, Down syndrome, intellectual disabilities, screening tool

INTRODUCTION

Previous studies have shown that Alzheimer's disease is (AD) more common in people with Down syndrome than in the general population (Bayen *et al*. 2018; Hithersay *et al*. 2019). However, the prevalence of AD or dementia in adults with intellectual disability (ID) who do not have DS has not been well studied because it is difficult to detect AD or dementia in people with ID (Silverman *et al*. 1998; Strydom and Hassiotis 2009; Zigman *et al*. 2004; Takenoshita *et al*. 2020). Screening tests such as the Mini-Mental State Examination (MMSE) or Montreal Cognitive Assessment (MoCA) are commonly used to detect cognitive impairment among the general population (Folstein *et al*. 1975; Nasreddine *et al*. 2005). However, these tests are not suitable for people with ID due to floor effects (Palmer 2006; Deb and McHugh 2010). Additionally, because the level of ID varies among individuals with ID, it is difficult to determine a unified cutoff point of such neuropsychological tests (Deb and Braganza 1999; Strydom and Hassiotis 2003).

In general, there are two methods to screen dementia in people with ID (Strydom and Hassiotis 2003; Zeilinger *et al*. 2013). The first is to evaluate

participants directly. The concepts of this screening method are the same as those of the MMSE or MoCA, in which the participants answer the questions asked by the tester. When test batteries specially designed to avoid floor effects and ceiling effects like the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS) are used, it is necessary to measure the cognitive function during young adulthood as a base point before the cognitive decline appears (Fonseca *et al*. 2008). Re-examination is performed after the onset of cognitive decline, and the presence or absence of cognitive impairment is assessed by comparing current test scores with previous test scores at the base point. Using adequate tests, we can evaluate the people with ID objectively. However, even a test easy for people with mild to moderate ID may be too difficult and inadequate for people with severe ID. Moreover, investigating many cases requires much time and effort because we need to test the subject repeatedly before and after the beginning of cognitive decline.

The second method is to use observer ratings, such as rating by carers. Concretely speaking, carers can evaluate the change of cognitive function and/or

daily function in a person with ID by comparing the current state with the state several years ago. Of course, to detect the decline of a person, the evaluators who must have observed the person for more than a few years to compare the present and past states. Comparison between the present and past clarifies the deterioration of cognitive function and/or daily function by a one-point evaluation. Moreover, these observer ratings can reveal the decline of functions in people with severe ID. However, a drawback of observer rating is that it needs a reliable evaluator who has known the person well for many years. Thus, the Dementia Scale for Down Syndrome (DSDS), Dementia Questionnaire for Persons with Mental Retardation (DMR), and Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) have been created to assess the cognitive decline of people with ID (Gedye 1995; Evenhuis 1992; Deb *et al*., 2007). Although DSDS and DMR are widely used for people with ID, there are problems in sensitivity and specificity (Strydom and Hassiotis 2003). Moreover, DSDS was designed for use by psychologists/psychometrists with university training and is difficult to use (Gedye 1995). On the other hand, it is reported that the DSQIID has relatively high sensitivity and specificity in the study of ID

people with Down syndrome. In addition, the DSQIID can be conducted and evaluated by caregivers or family members who have not received specialised training in psychology (Stanton and Coetzee 2004; Deb *et al*. 2007).

For the purpose of screening many cases, an observer rating instrument is superior to a direct testing instrument because it is simple. For early detection of dementia, the National Task Group on Intellectual Disabilities and Dementia Practice in the United States recommends the use of the NTG-Early Detection Screen for Dementia (NTG-EDSD) based on the DSQIID for Down syndrome and other pople with ID when they are suspected of showing cognitive decline (Esralew *et al*. 2013). However, there have been only a few reports that evaluated the usefulness of the DSQIID in adults with ID without DS. Therefore, we investigated a large group of adults with ID and evaluated the screening utility of the Japanese version of DSQIID (DSQIID-J) to detect dementia in people with ID.

METHODS

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 composed, 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.

Participants

This study reports the results of an analysis based on a cross-sectional study described elsewhere (Takenoshita *et al*. 2020). We recruited participants at the 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. In total, 28 support facilities agreed to participate. Facility residents and home-based residents using a day service at the facilities were included, and the participants 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 (World Health Organization 1993). (ii) There were information providers who had observed the activities of daily living (ADL) of the subject for two or more years, and the information providers agreed to respond to the interview and answer the questionnaire survey. (iii) Informed consent was obtained. (iv) The subject was 20 years or older. Whether the subject had Down syndrome or not was identified from records of chromosomal analysis or by their characteristic features (Strydom *et al*. 2009). The participants of this study are the same as the subjects in our previous report on the prevalence of dementia in ID. The results of the previous epidemiological study have been described (Takenoshita *et al*. 2020).

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Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)

The DSQIID is an observer-rated dementia screening questionnaire that is completed by carers who had known the subject for at least six months (Deb *et al*. 2007). It was developed on the assumption that it would be used for adults with ID. Although the validity of the DSQIID was evaluated only in adults with DS, the authors assert that the DSQIID can be equally useful in ID adults without DS (Deb *et al*. 2007). The DSQIID consists of three parts.

The first part asks about the subject's highest level of function in the past. This part is not added to the score.

The second part contains 43 questions on behaviours and symptoms related to dementia. In order to overcome the floor effect, it is designed to score changes in behaviour rather than current behaviour. Each item is answered by selecting one of four options: 'always has been the case'; 'always, but worse'; 'new symptom'; and 'does not apply'. 'Always, but worse' and 'new symptom' are scored 1, and 'always has been the case' and 'does not apply' are scored 0.

Part 3 consists of 10 questions that evaluate current states compared with past states; for example, ‘speaks (signs) less’ and ‘seems generally more tired’. A ‘yes’ response is scored 1 point and a ‘no’ response is scored 0. The 53 items of the QSQIID comprehensively reflect symptoms of dementia, including cognitive functions such as loss of memory and speech abnormalities, behavioural changes, psychological symptoms, and physical symptoms.

The Japanese translation

The Japanese version of DSQIID was created after translation and back-translation (Kinoshita *et al*. 2010). We used the Japanese version of DSQIID (DSQIID-J) in this study.

Diagnosis

Diagnosis of dementia was performed at the same time this validation study of the DSQIID-J was done. Three physicians directly interviewed the informants of all the participants to obtain information on the course of their cognitive functioning and ADL over the past few years. The three physicians were STa

(geriatric psychiatrist), STe (dementia specialist), and RK (geneticist in dementia). Three physicians determined the suspicion of cognitive decline by comparing the cognitive function and the ADL at the best time known to the informant with at the present time and directly examined the participants who were suspected of cognitive decline. The screening and diagnostic procedures are described in detail in other report (Takenoshita *et al*. 2020). In diagnosing dementia, we used the DSM-5 criteria defined as “major neurocognitive disorder” (American Psychiatric Association 2013). PSMS and IADL were used for quantitative clinical evaluation (Lawton and Brody *et al*. 1969). In the DSM-5, "cognitive deficits interfere with independence in everyday activities" is listed as a requirement to diagnose major neurocognitive disorder. However, many people with ID who use facilities are already dependent in their daily lives, regardless of whether they have dementia or not. Therefore, it was determined that this criterion was met when the activities of daily living that were previously partially possible were no longer possible due to cognitive impairment. Three physicians were blinded to the DSQIID-J score at the time of diagnosis.

Reliability

Inter-rater reliability was measured by determining the intraclass correlation coefficient (ICC) of 44 consecutive participants. Two raters assessed participants at the same time, and they were blind to each other's scores. We evaluated test-retest reliability using the ICC of 44 consecutive patients. The second session for test-retest reliability was done four to eight weeks after the first session. We evaluated the internal consistency reliability of the DSQIID-J using Cronbach's coefficient α (Cronbach and Meehl 1955).

Key question items

The DSQIID-J has a relatively large number of questions. In order to find a more efficient screening method for dementia, we searched for critically important items that are particularly relevant to the diagnosis of dementia. For all 53 items, we investigated the sensitivity and specificity of each item to screen dementia. Then, we investigated the sensitivity and specificity of all combinations of two items extracted from the 53 items. In the case of the combination of two items, if one or both of the two items were positive, we judged the combination of the two

items positive for screening dementia.

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 the categorical variables with continuity correction for 2×2 tables. The significance level was set at $P<0.01$ owing to the number of tests. We determined the sensitivity and specificity of the DSQIID-J using a receiver operating characteristic (ROC) curve. We used the area under the curve (AUC) as a scale of the test's ability to differentiate between groups of participants (dementia vs. nondementia). The most suitable cut-off scores for identifying dementia were determined to be the scores that led to the maximal accuracy of classification. Subsequently, positive predictive values (PPV) and negative predictive values (NPV) were estimated at different prevalence rates (5%, 10%, 20%, and 40%) for each optimal cutoff score.

RESULTS

Clinical characteristics of participants

There were 791 adults with ID using the target facilities. Of 791 potential participants, 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 participants was 46.57 (SD: 11.43 years; range: 20–83 years). Forty-seven of all participants were suspected of having dementia and directly examined by three physicians. Of 47 directly examined participants, 7 (14.9%) were people with ID with DS, and 40 (85.1%) were people with ID without DS. The mean age of 47 directly examined participants was 60.28 (SD: 9.31 years; range: 41–83 years). Seventeen of all participants met the DSM-5 criteria for dementia. An outline of the demographic data is shown in Table 1.

Normative data

The mean total score of the DSQIID-J was 1.69 ± 2.30 (mean ± SD) for all nondementia participants. The mean total score of nondementia participants with DS was 1.81 ± 2.39 (mean ± SD), and the mean total socore of nondementia

participants without DS was 1.68±2.29 (mean ± SD).

Diagnostic interpretation

We used the receiver operating characteristic (ROC) method to calculate the best fit between specificity and sensitivity. The ROC curves of the DSQIID-J for diagnosing dementia are shown in Figure 1 (for all participants in Figure 1-A, for participants with mild to moderate ID in Figure 1-B, and for participants with severe ID in Figure 1-C). The most suitable cut-off score of the DSQIID-J for discriminating participants with dementia from participants without dementia was 10/11 (sensitivity 100% and specificity 96.8%). When limited to participants without DS, the optimal cut-off score of the DSQIID-J for discriminating participants with dementia from participants without dementia was 10/11 (sensitivity 100% and specificity 96.9%). When limited to participants with DS, the most suitable cut-off score of the DSQIID-J for discriminating participants with dementia from participants without dementia was 13/14 (sensitivity 100%, specificity 96.3%). The PPV and NPV of the DSQIID-J for identifying dementia at different prevalence rates are shown in Table 2.

Reliability

The inter-rater reliability of the DSQIID-J was excellent (n = 44, ICCs = 0.880, p <0.01, >80% power). Analysis of the inter-rater reliability of the DSQIID-J according to the severity of ID revealed that the test-retest reliability of the DSQIID-J was fairly good for those with mild to moderate ID (n = 16, ICCs = 0.706, p = 0.013, >80% power) and excellent for those with severe ID (n = 28, ICCs = 0.900, p <0.01, >80% power). The test-retest reliability of DSQIID-J was also good (n = 44, ICCs = 0.842, p <0.01, >80% power). Analysis of the test-retest reliability of the DSQIID-J according to the severity of ID revealed that it was good for those with mild to moderate ID (n = 16, ICCs = 0.894, p = 0.013, >80% power) and good for those with severe ID (n = 28, ICCs = 0.839, p <0.01, >80% power). Two raters were involved for both studies of test–retest and inter-rater reliabilities. During the first and second tests, no participant experienced a large stressful incident that could strongly affect his or her ADL. Cronbach's coefficient α for all 53 items of DSQIID-J was high, 0.951. Analysis of Cronbach's coefficient α according to the severity of intellectual disability showed that it was

high (n = 195, Cronbach's α = 0.916) in mild to moderate intellectual disability and it was also high (n = 298, Cronbach's α = 0.960) in severe intellectual disability.

Key question items

First, a single screening item with 100% sensitivity was sought in the 53 items of the DSQIID-J. However, no single item was positive in all dementia cases. Second, combinations of two items with 100% sensitivity was sought in the 53 items of the DSQIID-J (total 1378 pairs). The best two-item combination with 100% sensitivity for dementia was the combination of PART2-2 "Cannot dress without help" and PART3-9 "Generally appears more forgetful" with a specificity 93.5%. The other good two-item combinations with the best three specificities are shown in Table 3.

DISCUSSION

DSQIID-J questions are simple and can be answered by caregivers who are not specially trained in psychology. Also, the DSQIID-J takes only about 10–15 minutes to complete, like the original edition, so it is easy to use. The reliability of the DSQIID-J was excellent. The DSQIID-J was found to be a sensitive and specific screening test to diagnose dementia in adults with ID of a Japanese sample. These results suggest that the DSQIID-J is a good screening instrument of cognitive impairment that has excellent validity and reliability.

Comparison with other language versions

In addition to the original English version, Chinese and Italian versions have been published, and validation studies on those versions were conducted (Deb *et al*. 2007; Li *et al*. 2015; Gomiero *et al*. 2017). The validation study of the original English version reported that the optimal cut-off score for identiying dementia was 20/21, and the study of the Chinese version reported 22/23 (Deb *et al*. 2007; Li *et al*. 2015; Gomiero *et al*. 2017). On the other hand, the optimal cut-off score of this study (10/11) was lower compared with that of the original and Chinese

versions. There are two pssible causes. Firstly, it may be due selection bias. There was a difference in the severity of ID and proportion of DS among participants in previous studies and our study. Although the validation study of the original English version does not show the severity of ID of the subjects, the Chinese version study included a relatively small proportion, 25.5%, of severe ID cases (Deb *et al*. 2007; Li *et al*. 2015; Gomiero *et al*. 2017). On the other hand, this study included a relatively large proportion, 60.4%, of severe ID cases (Table 4). In addition, the original English version included only cases with DS, but this study and the Chinese and Italian versions included cases both with and without DS (Deb *et al*. 2007; Li *et al*. 2015; Gomiero *et al*. 2017). Deb et al. reported that mild-to-moderate ID cases in DS tend to present the loss of recent memory as the early prominent symptom of dementia (Deb *et al*. 2007), but these symptoms may be difficult to detect in cases with severe ID. Instead of memory impairment, loss of skills and some behavioural problems seem to be the early indicators of dementia in cases with severe ID. The second cause is the difference in diagnostic methods. The original English version used ICD-10 criteria to diagnose dementia, the Italian version did not use specific diagnostic criteria, and the Chinese version

used DC-LD criteria (Royal College of Psychiatrists 2001). On the other hand, this study used DSM-5 criteria. In the general population, it has been reported that the prevalence of dementia varies depending on the criteria of dementia (Erkinjuntti *et al*. 1997; Stokin *et al*. 2015). We have reported that dementia in people with ID is diagnosed most inclusively using DSM-5 (Takenoshita *et al*. 2020). Differences in the diagnostic criteria used can lead to differences in the prevalence of dementia, which can result in differences in cut-off scores between studies.

Key question items

Because the original DSQIID-J requires 53 answers, we examined the performances of a single item and combinations of two items in screening for dementia. Remarkably, dementia can be determined when one or both of two specific items is positive. If the combination of items shown in Table 3 is used, dementia in people with ID can be efficiently screened with 100% sensitivity and >90% specificity. These "excellent" items are suggested to be particularly important symptoms that are related to the symptoms of dementia in people with

ID. The items shown in Table 3 include only items related to adaptive ability and behaviour, not items related to memory impairment. As reported in several studies, the initial symptoms of dementia in people with ID may not be memory impairment but deterioration of executive function and general functioning (Strydom *et al*. 2007; Ball *et al*. 2008; Nieuwenhuis-Mark 2009). Otherwise, people with ID can exhibit memory impairment in the early stages of dementia like the general population, as Block et al. reported, but it is difficult to detect memory impairment because of preexisting cognitive disabilities (Block *et al*. 2017). It is important to note that these two-item combinations are not diagnostic tools, and their performances are not guaranteed. Therefore, they should not be used alone. These are considerations from the results of this exploratory research, and other confirmatory studies are necessary in the future.

Limitation

Firstly, there is possibility that this sample does not reflect the general population of people with ID precisely. Because the participants of this study were day-service users and residents of facilities for people with ID, the general population

with ID may have less severe ID and have a different age distribution. Secondly, there were relatively few cases of dementia in this study population. Care must be taken in interpreting the results, particularly in subgroup analysis. We consider this a preliminary study to validate the DSQIID-J for dementia, and we are planning a confirmatory study in a large population to obtain more accurate sensitivity and specificity. Thirdly, the reliability of the test-retest was adequate, but the interval between the test (4–8 weeks) may have been too long for participants with dementia.

Conclusions

Although a relatively small number of patients were included in the analysis, this study indicated that DSQIID-J is a valid and reliable rating scale for dementia screening in adults with ID both in DS and non-DS.

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Table 1.

Table 1. Comparison of demographic data nondementia and dementia groups.

Demographic	Non-dementia	Dementia	*P*
Total (n) (%)	476 (100)	17 (100)	
Age (mean years ± SD)	46.02±11.29	61.94±8.75	<0.01
20-44 (n) (%)	212 (44.5)	0 (0)	
45-54 (n) (%)	137 (28.8)	4 (23.5)	
55-64 (n) (%)	85 (17.9)	5 (29.4)	
65-74 (n) (%)	31 (6.5)	7 (41.2)	
75-84 (n) (%)	11 (2.3)	1 (5.9)	
Sex (n) (%)			
Male	306 (64.3)	5 (29.4)	<0.01
Female	170 (35.7)	12 (70.6)	
DS (n) (%)			
With DS	27 (5.7)	7 (41.2)	<0.01
Without DS	449 (94.3)	10 (58.8)	
Severity of ID (n) (%)			
Mild ID	60 (12.6)	0 (0)	
Moderate ID	132 (27.7)	3 (17.6)	0.06†
Severe ID	284 (59.7)	14 (82.4)	
DSQIID (mean ± SD)	1.69±2.30	25.12±10.92	

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

P-value is comparison between participants with and without dementia.

Comparison of sex ratio, DS, and severity of ID between two groups was done using $\chi 2$ test.

†Comparisons of ID severity were analyzed in two groups: mild to moderate ID and severe ID.

Table 2.

Table 2. Sensitivity, specificity and positive predictive values (PPV) at different prevalence rates of optimal cut off scores for identifying dementia

SUBJECT	CUT-OFF	DEMENTIA		PPV at different prevalence rates			
	SCORE	SENSITIVITY	SPECIFICITY	5%	10%	20%	40%
All ID	10/11	100%	96.8%	62.2%	77.6%	88.7%	95.4%
ID without DS	10/11	100%	96.9%	62.9%	78.2%	89.0%	95.6%
ID with DS	13/14	100%	96.3%	58.7%	75.1%	87.1%	94.7%

For all items in table 2, negative predictive values (NPV) at each prevalence rates is all 1.000 because the sensitivity at the optimal cutoff is all 1.000.

Table 3.

Table 3. Top three 2 items and 3 items screening for dementia

Combinations of 2 items (Positive if 1 or more)			sensitivity	specificity
1	PART2-2	Cannot dress without help	100%	93.5%
	PART3-9	Walks slower		
2	PART2-10	Cannot follow simple instructions	100%	93.3%
	PART3-1	Lost some skills (e.g. Brushing teeth)		
3	PART3-9	Walks slower	100%	91.6%
	PART3-10	Generally appears more confused		

Table 4.

Table 4. Comparison between other validation studies

		n (dementia)	range of age (mean years ± SD)	DS+/DS-	severity of ID (mild/ moderate/ severe or more)	cut-off to identify dementia (sensitivity/ specificity)
English [a)]	Deb *et al*. 2007	193 (49)	23 to 77 (55 ± 7.6)	193/0	N/M	20/21 (0.92/ 0.97)
Chinise	Li *et al*. 2015	200 (13)	40 to 73 (51 ± 7.34)	36/164	(31/ 118/ 51)	22/23 (0.995/ 0.923)
Italian	Gomiero *et al*. 2017	200 (5)	40 to 80 (55.2 ± 7.51	58/142	(25/ 69/ 48)	N/M
Japanese	Takenoshita *et al*. 2019	493 (17)	20 to 83 (46.57 ± 14.09)	34/459	(60/ 135/ 298)	10/11 (1.00/ 0.96)

a) original

N/M, not mentioned

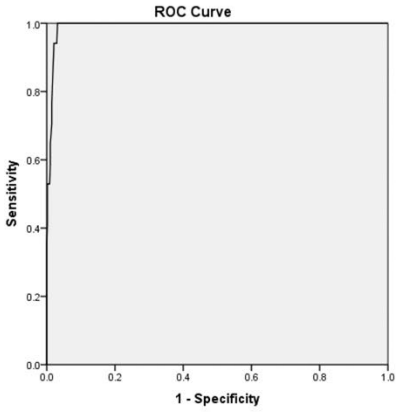
Figure 1. The AUROCs of the DSQIID-J for diagnosis of each condition.

1-A (upper). ROC curves for discriminating participants with dementia from nondementia participants among all participants. The AUC of the DSQIID-J for diagnosing dementia was 0.992 (0.985–0.999).

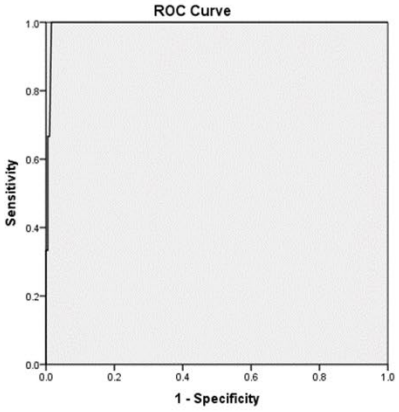
1-B (middle). ROC curves among participants with mild to moderate ID. The AUC was 0.994 (0.983–1.000).

1-C (lower). ROC curves about participants with severe ID. The AUC was 0.991 (0.982–1.000).

A. All participants



B. Participants with mild to moderate ID



C. Participants with severe ID

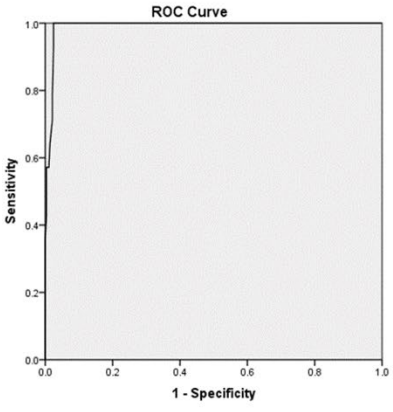


Figure 1.