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A validated WAIS-IV short-form to estimate intellectual functioning in myotonic dystrophy type 1

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ABSTRACT

Currently, no rapid and specific instrument is available to briefly estimate intelligence in patients with myotonic dystrophy type 1 (DM1), a multisystemic disease that involves the CNS and is associated with cognitive deficits and low intellectual functioning. This study aimed to develop a DM1-specific and valid short-form of the Wechsler Adult Intelligence Scale-Fourth Edition (WAIS-IV) to estimate intellectual functioning in this population. Thirty non-congenital DM1 patients (10 female; mean age=46.77; *SD*= 9.76) were assessed with the WAIS-IV. Data were analyzed following two independent strategies: A) multiple linear regression with the aim of maintaining the scale's factorial structure; and B) correlational analyses between scores on all WAIS-IV subtests and Full-Scale IQ (FSIQ). Validity of the resulting short-forms was also analyzed. Three short-forms were developed: Proposal A from strategy A (Vocabulary, Block Design, Arithmetic and Symbol Search), Proposal B1 (Vocabulary, Block Design, Digit Span and Visual Puzzles) and Proposal B2 (Vocabulary and Block Design), from strategy B. All three short-forms showed a strong and significant correlation with the FSIQ and were considered psychometrically acceptable. Arguments in favor of Proposal B1 are discussed. Assessing FSIQ with these short-forms will be useful for avoiding long assessment procedures in a population characterized by high fatigability.

1. Introduction

Myotonic dystrophy type 1 (DM1) is the most common form of muscular dystrophy in adults and is transmitted in an autosomal dominant manner and diagnosed with an expansion length of the trinucleotide CTG (cytosine, thymine, guanine) exceeding 50 repetitions. It is considered a rare disease, with a global prevalence of 1/7400 [1]. DM1 is classified into five phenotypes regarding the age of onset of the disease; congenital (at birth), childhood-onset (1–10 years at the age of onset), juvenile-onset (10–20 years), adult-onset (20–40 years), and late-onset (>40 years), with the adult-onset form being the most common [2]. It is a progressive multisystemic disease (associated with symptoms such as muscular weakness, respiratory, metabolic and cardiac problems, and cataracts) that involves the central nervous system (CNS) [3] as a neurodevelopmental [4] and neurodegenerative disorder [5,6], with a large spectrum of cognitive deficits and other CNS-related symptoms such as excessive daytime sleepiness, fatigue, and apathy [7]. Cognitive deficits are frequently reported in adult-onset DM1 and late-onset DM1, but they are milder than those seen in congenital, juvenile, and childhood-onset DM1 [8]. Previous studies have shown that DM1 patients have significantly lower intellectual functioning than healthy controls and are generally placed one standard deviation below the mean in the classic form of the disease [9–11]. Regarding the pattern

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of cognitive impairment, the results generally indicate a specific executive and visuoconstructive dysfunction [11]. However, other cognitive domains such as visual memory, visuospatial abilities, psychomotor speed, and social cognition, among others, have been reported to be affected [12].

A critical aspect of studies analyzing the cognitive profile in DM1 is an adequate neuropsychological assessment, including an estimation of intellectual functioning. The Wechsler Adult Intelligence Scale (WAIS) is the most widely used instrument for this purpose. The most recent Spanish version of the scale is the WAIS-Fourth Edition (WAIS-IV) [13], with an administration time of approximately 60–70 min for the general population, and longer in the clinical population [14].

In studies on DM1, short-forms to estimate IQ have been used to deal with the symptoms and limitations these patients might face during assessment [5,10,11,15–18]; these forms show considerable variety regarding the subtest composition (see Supplementary Table 1 for additional information). Nevertheless, these short-forms have been developed for general population and might not be valid in a disease associated with cognitive deficits, daytime sleepiness, and fatigue [7]. Thus, the main concern with the use of general population short-forms in DM1 is that they may not fit adequately with the cognitive particularities of the disease. Therefore, the use of these short-forms could lead to biases in Full-Scale Intelligence Quotient (FSIQ) estimations, resulting in systematical over- or under-estimations.

To the best of our knowledge, the reliability and validity of short-forms have not yet been studied in DM1 patients. For this reason, various studies have highlighted the necessity of validating short-forms in populations with neurological disorders (such as multiple sclerosis, acquired brain injury, epilepsy, and neurofibromatosis, among others) by either analyzing the validity of existing short-forms [19–22] or developing new ones [23].

In line with the DM1 guidelines suggesting a systemic cognitive assessment in DM1 populations [8,24], this study aimed to develop a valid short-form to estimate IQ in DM1.

2. Material and methods

2.1. Participants

In the recruitment phase, all participants attending the outpatient Neurology service of the Donostia University Hospital (Gipuzkoa, Spain) were invited to participate as long as they met the inclusion criteria. All the invited participants accepted, and the recruitment period was concluded when 30 patients had been recruited.

Thirty patients (10 female) with a confirmed diagnosis of DM1 participated in this study. Inclusion criteria included molecular confirmation of the disease, being above 18 years of age, having no other diagnosis of neurological or psychiatric disorders (according to the DSM-V), and the absence of drug or alcohol abuse. In addition, patients with the congenital form of the disease were excluded since the scientific literature recognizes this as a clinically different form [2]. Regarding the phenotype based on the age of onset, the sample was distributed as follows: 17 adult forms (56.7%), nine juvenile forms (30%), three childhood forms (10%), and one late-onset form (3.3%). The inheritance pattern of most of the patients was paternal (83.3%), and the mean age of the patients at neuropsychological assessment was 46.77 years (SD= 9.76). The patients' FSIQ was low average (M = 88.43; SD= 13.59) according to Wechsler's FSIQ classification. A description of the participants is presented in Table 1. Further information on patients' performance on WAIS-IV subtests and indexes is presented in Supplementary Table 2.

Table 1
Demographic and clinical data.

		N/N (%)	*M*	*SD*	Min	Max
Sex	Female	10 (33.3%)				
	Male	20 (66.7%)				
Age (years)		30	46.78	9.76	28	72
Education (years)		30	15.70	4.36	5	24
CTG repeats		29	570.17	371.08	50	1500
MIRS		27	2.93	1.11	0	4
FSIQ		30	88.43	13.59	56	109

Note. M= Mean; SD=Standard Deviation; CTG repeats = CTG expansion size; MIRS = Muscular Impairment Rating Scale; FSIQ = Full-Scale IQ.

All participants were informed about the study, both verbally and by a written document, and they signed the informed consent form. The Ethics Committee for Clinical Investigation of the Health Department of Gipuzkoa (DMRM-2017–01) approved the study.

2.2. Clinical data and neuropsychological assessment

Clinical data (CTG, MIRS, phenotype, and inheritance pattern) were obtained through medical records. The neuropsychological assessment was conducted by two experienced neuropsychologists blind to the patients' clinical condition. The Spanish version of the WAIS-IV [13] was administered, including the optional subtests of the scale. Raw scores of the subtests were converted into scaled scores, following the normative data according to age group.

WAIS-IV is a widely administered intelligence scale, which consists of 15 subtests (10 core subtests and five supplemental subtests): Block Design, Similarities, Digit Span, Matrix Reasoning, Vocabulary, Arithmetic, Symbol Search, Visual Puzzles, Information, Coding, Letter-Number Sequencing, Figure Weights, Comprehension, Cancellation, and Picture Completion. Structurally, WAIS-IV is organized into four indexes: Verbal Comprehension, Perceptual Reasoning, Working Memory, and Processing Speed. FSIQ is calculated based on the scores of these indexes.

2.3. Statistical analysis

The resulting data were analyzed using the SPSS statistical package (Version 24). First, statistical analyses were conducted following two independent strategies to obtain WAIS-IV short-forms. The first strategy was based on multiple linear regression analysis after checking the corresponding assumptions (Strategy A), while the second was based on a simple correlation analysis (Strategy B). IQ estimations of the short-forms were then derived according to the formula described by Tellegen and Briggs [25], where inter-correlations between the subtests and internal consistency reliability were directly estimated from the Spanish DM1 sample, given that clinical samples could potentially differ from general population samples.

Subsequently, the results were interpreted considering the following validation criteria established by Donders and Axelrod [26]: a) reliability coefficient $\geq$ 0.90 of the short-form; b) a corrected correlation $\geq$ 0.82 between FSIQ and short-form IQ; c) a proportion $\geq$ 0.81, of short-form estimates that fell within the 90% confidence interval (CI) of the FSIQ (i.e., within $\pm$ 7 FSIQ points). The strategies mentioned above are described in further detail below.

Based on the factorial structure of the scale, the first strategy (Strategy A) aimed to maintain the structure of the scale by selecting one subtest from each of the four indexes which provide a better fit with the FSIQ. To this end, multiple linear regressions were conducted to determine the subtest with the greatest predictive capacity for each index. The following assumptions were checked: linearity, curvilinear relationship, homocedasticity,

Table 2
Linear regressions between short-forms and FSIQ.

Variable	β	*SE*	*t*	*p*	*95% CI*
Proposal A					
(Constant)	39.94	4.16	9.61	0.000	[31.38–48.50]
Vocabulary	1.68	0.31	5.48	0.000	[1.05–2.31]
Block Design	1.76	0.40	4.38	0.000	[0.93–2.59]
Arithmetic	0.99	0.33	3.01	0.006	[0.31–1.67]
Symbol Search	1.19	0.43	2.77	0.011	[0.30–2.07]
Proposal B1					
(Constant)	38.45	3.16	12.19	0.000	[31.96–44.95]
Vocabulary	1.60	0.25	6.47	0.002	[1.09–2.11]
Block Design	1.21	0.34	3.54	0.002	[0.51–1.92]
Digit Span	1.40	0.40	6.47	0.000	[1.09–2.11]
Visual Puzzles	1.37	0.31	4.43	0.000	[0.73–2.01]
Proposal B2					
(Constant)	50.75	3.45	14.73	0.000	[43.68–57.82]
Vocabulary	1.96	0.35	5.57	0.000	[1.24–2.68]
Block Design	2.41	0.42	5.72	0.000	[1.55–3.28]

Note. SE= Standard error.

normality of residuals, multicollinearity, auto-correlation, and influential cases (See Supplementary Table 3). Once the most representative subtest of each index was selected, a multiple linear regression was carried out, with these four subtests as predictors and the FSIQ as the criterion variable. Finally, the percentage of explained variance of the model and the model's predictive capacity was calculated for each subtest.

The second strategy (Strategy B) focused on identifying the subtests closely related to the scale's total score without considering the index to which they belong. For this purpose, Spearman's correlation analyses were conducted between all WAIS-IV subtests and the FSIQ. To select only subtests with a high correlation, a cut-off point was set at rho= 0.7, representing a very strong association [27]. Once the subtests were selected (rho≥0.7), a multiple linear regression was conducted between the combination of these subtests and the FSIQ. Finally, the subtests with the highest correlations were selected with the additional objective of developing a very brief short-form. A regression analysis was conducted between the combination of these subtests and the FSIQ. In other words, the aim was to analyze whether a very short-form still showed acceptable validity. In all cases, we excluded the subtests whose coefficients did not reach statistical significance.

3. Results

Three WAIS-IV short-form proposals were developed; one short-form using Strategy A (Proposal A) and two using Strategy B (Proposal B1 and Proposal B2). The proposals are described as follows.

3.1. Proposal a

The subtests with the highest standardized β coefficient in each of the regressions were Vocabulary subtest (standardized β coefficient = 0.47) in the Verbal Comprehension Index; Block Design subtest (standardized β coefficient = 0.40) in the Perceptual Reasoning Index; Arithmetic subtest (standardized β coefficient = 0.68) in the Working Memory Index; and Symbol Search subtest (standardized β coefficient = 0.58) in the Processing Speed Index. The linear regression model conducted with the combination of Vocabulary, Block Design, Arithmetic, and Symbol Search explained 88% of the variance of the FSIQ. All selected subtests were statistically significant predictors of the FSIQ (p<0.05) (see Table 2).

Regarding the fulfillment of validation criteria, a) the reliability coefficient is 0.96; b) the corrected correlation between FSIQ and the IQ of Proposal A is 0.90, and c) the proportion of estimations within the CI 90% is 0.80.

3.2. Proposal B1

The results of the Spearman's correlation analysis between the FSIQ and all the scale subtests are shown in Table 3. The subtests Vocabulary, Block Design, Digit Span, Visual Puzzles and Comprehension, yielded correlations >0.7, and were thus selected to be included in the model to conduct the linear regression analysis.

The comprehension subtest was rejected because although it showed a correlation > 0.7, it did not reach a statistically significant value in the regression model. Thus, the regression model composed of Vocabulary, Block Design, Digit Span, and Visual Puzzles subtests explained 92% of the FSIQ variance. Furthermore, all the subtests were statistically significant predictors of the FSIQ (p<0.05), as shown in Table 2.

Regarding validation criteria fulfillment, a) the reliability coefficient of Proposal B1 is 0.98; b) the corrected correlation between FSIQ and the IQ of Proposal B1 is 0.87, and c) the proportion of estimations within the 90% CI is 0.77.

3.3. Proposal B2

As shown in Table 3, Vocabulary and Block Design were the subtests with the highest correlations. The short-form composed of these two subtests explained 83% of the FSIQ variance, with these subtests being statistically significant predictors of the FSIQ (see Table 2).

Regarding validation criteria fulfillment, a) the reliability coefficient of Proposal B2 is 0.96; b) the corrected correlation between FSIQ and the IQ of Proposal B1 is 0.69, and c) the proportion of estimations within the 90% CI is 0.53.

4. Discussion

To the best of our knowledge, this is the first attempt to propose a DM1-specific and validated short-form for estimating intellectual functioning. We emphasize the importance of reducing long assessment protocols, given that DM1 patients frequently deal with symptoms such as fatigue, daytime sleepiness, and apathy. Thus, under such circumstances and, always considering the inherent limitations of short-forms, the use of these brief tools containing acceptable psychometrical properties can ensure an adequate estimation of intellectual functioning in a relatively short time [20,28]. Indeed, the short-forms proposed in this study provide an estimation of the general intellectual functioning in less than half of the time taken to complete the full scale (see Supplementary Table 4 for more information). Moreover, short-forms developed specifically for DM1 adjust better to the cognitive and neurological characteristics of the disease than other short-forms developed for the general population.

This study yielded three WAIS-IV short-form proposals; two were composed of four subtests (Proposal A and Proposal B1) and one was composed of two subtests (Proposal B2). Proposals A and B1 satisfied the reliability and corrected correlation criteria of Donders and Axelrod [26], but not the last criterion (proportion of estimations within 90% CI). Proposal B2 (Vocabulary and Block Design) only fulfilled the first criterion. Although neither of the proposals fulfilled the third criterion, the mean discrepancy between the FSIQ and the IQ obtained from the short-form is small, in accordance with Van Duijvenbode et al. [22]. Indeed, the results showed that Proposals A and B1 contain a high percentage of agreement (>75%). It should be mentioned that the last criterion is based on the recommendation of Kaufman [29] for

Table 3
Spearman's Correlations between FSIQ and the subtests of the scales.

	BD	SI	DS	MR	VC	AR	SS	VP	IN	CD	LN	FW	CO	CA	PCm
Rho	0.81**	0.68**	0.77**	0.55**	0.82**	0.54**	0.55**	0.74**	0.58**	0.34	0.54**	0.54**	0.77**	0.28	0.50**
N	30	30	30	30	30	30	30	30	30	29	29	29	30	29	30

Note. BD = Block Design; SI = Similarities; DS = Digit Span; MR = Matrix Reasoning; VC = Vocabulary; AR = Arithmetic; SS = Symbol Search; VP = Visual Puzzles; IN = Information; CD = Coding; LN = Letter-Number Sequencing; FW = Figure Weights; CO = Comprehension; CA = Cancellation; PCm = Picture Completion. The shaded areas in the table illustrate the subtests with rho≥0.7. **. Statistically significant $p<0.01$ (two-tailed). *. Statistically significant $p<0.05$ (two-tailed).

the clinical application of short-forms and that various studies have found difficulties in satisfying this criterion in their short-forms [21,22,26,30].

Nevertheless, we interpreted our results using these criteria, because as far as we know, there are no specific validity criteria for research purposes. Data on the validity and reliability of short-forms for the DM1 population are provided in this work, while the short-forms administered in previous DM1 studies do not include such data. Furthermore, the reliability coefficients of the short-forms proposed in this study are based on our DM1 sample instead of using data provided by the manual.

Taken together, both four-subtest short-forms (Proposal A and B1) developed in this work are considered psychometrically acceptable. Proposal B1 has the advantage of not containing a graphomotor component, which could be beneficial for a population with a frequent distal affection that could hinder adequate performance of the test. Further, the employed analysis strategy in B1 is not conditioned by the factorial structure of the scale, since this short-form is data-driven, and thus adjustable to the specific cognitive characteristics of the disease.

Concerning the two-subtest short-form (Proposal B2), although psychometrically weaker than the others, its use could be justified when a very brief test is required. Finally, it is worth mentioning that this short-form has already been proposed for the non-clinical population and is considered one of the best dyadic combinations for estimating FSIQ [31,32].

Nevertheless, short-forms present certain limitations. Some authors, such as Kaufman et al. [33], warn that estimations derived from short-forms should always be used with caution and never for diagnostic or specific classification purposes [23].

IQ estimations are often required both in DM1 investigations and in clinical trials, given the fundamental role of the CNS regarding the severity and natural course of the disease [34]. To this end, these short-forms could rapidly estimate intellectual functioning and be used as a marker, together with other relevant measures (e.g., MIRS, CTG length, specific cognitive measures, and other extracerebral biomarkers). However, this latter application has not yet been tested. Finally, these short-forms can be suitable for use in clinical settings, screening purposes, or a brief assessment of the patients' intellectual status [20,23,35,36]. This would provide clinicians and researchers with the time needed to assess other key cognitive domains in depth (e.g., visuospatial abilities, executive functions, and social cognition), as well as other areas implicated in patients' daily functioning.

Regarding the scoring procedure of the proposed short-forms, although the Tellegen and Briggs formula (1967) was used for calculating IQ values of the short-forms, prorating can be used by clinicians to simplify IQ acquisition, given the high correlation between the IQ estimations obtained by both methods in this study in all short-form proposals (Proposal A r: 0.94; Proposal B1 r: 1.00; Proposal B2 r: 1.00). Prorated IQ can be obtained by summing the scaled scores of the short-form subtests and by multiplying by 10/4 for Proposal A and B1, and 10/2 for Proposal B2 (number of subtests required for calculating FSIQ/ number of subtests of the short-form). The obtained score needs to be converted to FSIQ according to the WAIS-IV normative data. The prorating formula can be found in Supplementary Data 1.

In agreement with Axford and Pearson [37], a precise and DM1-specific set of standardized tests would provide a more accurate description of the deficits in this population while allowing for more feasible comparability between studies. Thus, it is expected that these data will help advance this systematization process of neuropsychological assessment in DM1.

Despite the value of this study, it is not free of limitations. The main constraint is the reduced sample size. In addition, the sample included heterogeneous phenotype conditions. Although this is a limitation because each phenotype shows variability in cognitive profile and intellectual functioning [2], phenotype heterogeneity also provided a better representativeness of the DM1 population. These findings, however, may not generalize to a broader population or other diseases, given the specificity of the sample. Therefore, further research should replicate this study in other DM1 samples, ideally with a larger sample that includes a better representation of the phenotypes, or even analyzing each phenotype separately.

In conclusion, we have proposed three WAIS-IV short-forms, of which, based on the reasons outlined above (statistical strategy employed, absence of graphomotor load, and psychometric properties) proposal B1 is of particular relevance. DM1-specific and validated short-forms might provide better reliability, considering the characteristics of the disease, such as fatigue and daytime sleepiness. Additionally, this work could contribute to a more adequate systematization of neuropsychological assessment in DM1, favoring the comparison between studies and scientific consensus.

Declaration of Competing Interest

None.

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.nmd.2022.06.012.

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