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Spanish adaptation and validation of the child- and parent-report cystic fibrosis questionnaire-revised (CFQ-R)

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Abstract

Objective: To evaluate the psychometric properties of the Spanish versions of the child- and parent-report cystic fibrosis questionnaire-revised (CFQ-R).

Methods: A Spanish adaptation of the CFQ-R was performed; 68 children with CF (6–13 years) and their parents completed the child- and parent-report CFQ-R, respectively, and the Revidierter KINDer Lebensqualitätsfragebogen (KINDL) questionnaire. The CFQ-R was completed twice, 7–10 days apart, and its psychometric properties were analyzed.

Results: The internal consistency of both CFQ-R versions was adequate (child-report version, Cronbach's α >.60 for all domains except "Treatment Burden" [α = .42] and "Social Functioning" [α = .57]; parent-report version, α > .60 for all domains except "Social Functioning" [α = .58]). For the child-report version, the lowest measurement error was for "Emotional Functioning" (standard error of measurement [SEM]: 8.3%; minimal detectable change [MDC_{90}]: 19.3%), and the highest was for "Body Image" (SEM: 15%; MDC_{90}: 35%). For the parent-report version, the lowest measurement error was for "Physical Functioning" (SEM: 7.1%; MDC_{90}: 16.5%), and the highest was for "Weight" (SEM: 17.2%; MDC_{90}; 40.1%). The correlation between the versions showed higher agreement for the domains related to observable signs ("Physical Functioning") and lower agreement for "Emotional Functioning." There was a significant correlation between the CFQ-R and KINDL.

Conclusion: Both the child- and parent-report versions of the Spanish CFQ-R have adequate reliability and validity for clinical and research purposes. These versions can be administered before and after starting modulator therapy to assess its effect on daily functioning. The MDC_{90} can help identify, with a high probability, whether real changes have occurred in the quality-of-life subscales in children with CF.

KEYWORDS
child, cystic fibrosis, quality of life, reliability, validity

1 | INTRODUCTION

Cystic fibrosis (CF) is a multisystem disease caused by an alteration in ion channels due to a genetic mutation located on chromosome 7q31.2, which provokes a dysfunction in the CF transmembrane regulator.[1,2] The prevalence of CF is 1:5000 live births in Spain.[3] According to the most recent Spanish CF registry, there are at least 2439 patients with CF, although there could be as many as 3000.[4] The phenotypic features of CF include chronic sinopulmonary disease, gastrointestinal and nutritional abnormalities, salt wasting syndromes, and reproductive system abnormalities in male individuals. The clinical respiratory system manifestations mainly cause morbidity and reduce the quality of life of this population.[5]

To monitor CF progression, clinicians have numerous tools at their disposal, such as pulmonary function, exercise tolerance, and health-related quality-of-life (HRQoL) tests. Recent advances in the development of drugs and therapies for CF have improved the prognosis and quality of life for children with CF[6]; standardized assessment tools are therefore essential for evaluating these changes.[7]

Given that generic questionnaires designed to evaluate HRQoL are insufficiently sensitive in discriminating specific aspects of CF, such as the potential benefits of treatments and the impact of exacerbations or disease progression, there is a need for specific HRQoL questionnaires designed for patients with CF.[8] Recent evidence has shown that the cystic fibrosis questionnaire-revised (CFQ-R)[9–11] has better psychometric properties, in terms of validity and reliability, compared with other HRQoL questionnaires for individuals with CF.[12] The CFQ-R was adapted and revised by Quittner et al.[9,11,13] and Modi et al.[10] from the CFQ created in France in 1998.[14,15] The CFQ-R has been translated into several languages and is widely employed in clinical and research contexts.[16–20] The CF Foundation palliative care guidelines recommend the clinical use of CFQ-R as an optional measure.[7] The CFQ-R has specific versions for children, their parents, adolescents, and adults.[10,11,13] Although the Spanish adult version was adapted by Olveira et al.[19] in 2010, the child version had not yet been adapted in Spain.

In chronic diseases of childhood, such as CF, the differences between adults, adolescents, and school-age children need to be addressed when evaluating HRQoL.[9,10,13] For younger children, it is especially important to assess their parents' perspective together with the child's perspective due to the likely differing perceptions, especially those concerning non-observable symptoms.[11,21] Other Spanish-validated instruments measuring HRQoL have demonstrated that parents can both confirm their children's perceptions and differ in certain aspects.[22] Evaluating the perspectives of children with CF and that of their parents can be of significant utility for implementing specific support measures to improve child-centered assessment and treatment[21] and to enhance parental participation in their children's treatment and care.[23]

The aim of the current study was to adapt and assess the validity and reliability of the Spanish child- and parent-report versions of the CFQ-R.

2 | METHODS

2.1 | CFQ-R

Specific versions of the CFQ-R were designed to measure general health, daily well-being, and symptoms.[10,13] This study employed two versions: the CFQ-R for children aged 6–13 years (with a self-administered adaptation for children aged 12–13 years) and the CFQ-R for parents (parent-proxy measurement completed by parents reporting their children's symptoms). The CFQ-R for children includes eight subscales: "Physical Functioning" (6 items), "Emotional Functioning" (8 items), "Social Functioning" (7 items), "Body Image" (3 items), "Eating Disturbances" (3 items), "Treatment Burden" (3 items), "Respiratory Symptoms" (4 items), and "Digestive Symptoms" (1 item). The CFQ-R for parents includes 11 subscales: "Physical Functioning" (9 items), "Vitality" (5 items), "Emotional Functioning" (5 items), "School Functioning" (3 items), "Body Image" (3 items), "Eating Disturbances" (2 items), "Treatment Burden" (3 items), "Health Perceptions" (3 items), "Weight" (1 item), "Respiratory Symptoms" (7 items), and "Digestive Symptoms" (3 items). The questionnaire takes approximately 15 min to complete, and scoring is based on a 4-point Likert scale, with scores ranging from 0 to 100 and higher scores indicating a better quality of life.[10] The CFQ-R is considered an excellent instrument because its adaptations to other languages have demonstrated good psychometric properties.[13,16–20,24]

2.2 | Revidierter KINDer Lebensqualitätsfragebogen

The KINDL is a generic HRQoL questionnaire developed in Germany for healthy and ill children and adolescents. The questionnaire comprises 24 items evaluating the following six dimensions: Physical well-being, emotional well-being, self-esteem, family, friends, and daily life functioning (school).[25] Scoring for each item is based on a 5-point Likert scale, and the score for each dimension can range from 0 to 100, with higher scores indicating a better quality of life. The total KINDL score is obtained from the average of the 6 score dimensions. The Spanish version of the KINDL shows acceptable psychometric properties.[26]

2.3 | Cross-cultural adaptation

Based on the Spanish version of the CFQ-R created for Spanish speakers in the United States,[27,28] an evaluation was conducted with a panel of experts who proposed improvements so that the instrument could be employed in Spain. An expert panel was formed with 15 CF professionals, including a pulmonologist, pediatrician, physiotherapist, occupational therapist, psychologist, social worker, nutritionist, and specialist in therapeutic pedagogy. The selection criteria for the experts were as follows: at least 3 years of experience with individuals with CF, availability, motivation to participate, and impartiality. The members of the expert panel were asked to provide

a qualitative assessment of each item, answering brief questions about the degree of comprehensibility, clarity, and coherence. The experts also completed a 4-point Likert scale (highly relevant/fairly relevant/not very relevant/not relevant) that evaluated each item's relevance. The quantitative ratings for each item's relevance, according to the expert panel, are shown in Supporting Information: Appendix A.

2.4 | Validation process

The validation process was conducted following the Consensus-based Standards for the Selection of Health Measurement Instruments guidelines.[29] The sample was recruited between September 2021 and July 2022 from various hospitals and CF associations. The study was previously approved by each institution's ethics committee. Children were included if they lived in Spain, had a CF diagnosis, were 6–13 years of age, had no exacerbations during the month before study inclusion and could understand the study protocol's questionnaires. These inclusion criteria are the same as those used in other adaptations of this questionnaire.[13,16,17,20] Before the study, the parents of the children younger than 12 years signed the informed consent document, with both agreeing to the child's participation. Children older than 12 years were asked to sign their own assent document, in addition to receiving parental consent.

During the first evaluation, the study collected data on demographics, health status and pulmonary function, and the CFQ-R and KINDL questionnaires were handed out.[22] The demographic data, health status, and pulmonary function were collected from medical records (from tests performed within the past month), while the CFQ-R and KINDL were collected by self-report.[16]

To evaluate test–retest reliability, all children were asked to retake the CFQ-R test 7–10 days after the first assessment,[9] provided they had not experienced an exacerbation.

2.5 | Data analysis

The data were analyzed with SPSS v. 25 software (SPSS Inc.), and the level of significance was set at $p < .05$.

2.5.1 | Floor/ceiling effects and reliability

The internal consistency for each subscale of the Spanish CFQ-R child- and parent- report versions was assessed with Cronbach's α, except for the subscales with only 1 item because in these cases Cronbach's α cannot be calculated. A Cronbach's α >.6 is expected for all subscales, which could be considered acceptable for newly developed scales.[30,31] The floor/ceiling effect was calculated for each subscale of these two Spanish versions of the CFQ-R. As with previous questionnaire validation studies,[19,32] the floor/ceiling effect was considered to be present if at least 15% of the child-/parent-report version achieved the minimum/maximum score, respectively.[33]

The test–retest reliability (over a 7 to 10-day period) of both versions of the Spanish CFQ-R was assessed for the absolute value of each subscale separately, using the intraclass correlation coefficient (ICC) as follows: excellent (ICC ≥ 0.90), good (0.90 > ICC ≥ 0.70), fair (0.70 > ICC ≥ 0.40), and poor (ICC < 0.40).[34] The standard error of measurement (SEM) and the minimal detectable change as 90% of the confidence interval (MDC_{90}) were calculated following the same criteria as a previous study.[35] The SEM is the systematic and random error of a participant's score; its score is therefore not attributable to true changes in the construct being measured.[29] The MDC_{90} expresses the minimum change needed between two measures to be 90% confident that the observed change between them reflects a real change and is not a measurement error.[36,37] A 90% confidence interval was used because it is appropriate for decisions regarding the effectiveness of an intervention.[36,37]

2.5.2 | Construct validity

Construct validity was examined by analyzing the concordance between the subscale scores of the child- and parent-report versions of the Spanish CFQ-R using the ICC. In addition, construct validity was also examined by analyzing the relationship between the subscale scores of the child- and parent-report versions of the Spanish CFQ-R and between the KINDL dimension scores separately and between the average of all dimensions (total KINDL score). The relationship between the subscale scores of the child- and parent-report versions of the CFQ-R and the children's clinical outcomes (e.g., lung function, number of hospitalizations, and exacerbations within the past year) was assessed. Pearson's correlation coefficient (r) was employed to establish correlation: low ($r < .30$), moderate ($r = .30–.60$), and strong ($r > .60$).[38]

The test–retest reliability (over a 7 to 10-day period) of both versions of the Spanish CFQ-R was assessed for the absolute value of each subscale separately, using the ICC as follows.

3 | RESULTS

3.1 | Cross-cultural adaptation

The expert panel evaluated the CFQ-R and considered the tool appropriate, clear, simple and correct for 6–13 years age group, and most of the experts rated all items as highly relevant (Supporting Information: Appendix A). Six items were identified (5 in the child-report version and 1 in the parent-report version) that could hinder understanding due to cultural differences in the meaning of the language. Subtle changes were therefore made using adapted vocabulary that is more frequently employed in Spain to facilitate understanding by Spanish children and parents. The modifications were made with the consent of the original

author of the CFQ-R, and the meaning of the items in the original version was maintained. All items from the original version were therefore retained in the final child- and parent-report versions of the Spanish CFQ-R.

3.2 | Clinical characteristics

The total sample consisted of 68 children with CF (who met all the inclusion criteria: mean age 9.26 years, range 6–13 years, 45.7% male) and their parents. Although 72 children were initially recruited, 4 were excluded because they did not complete the entire study protocol. None of the participants were taking highly effective modulators (e.g., ivacaftor or elexacaftor/tezacaftor/ivacaftor [ETI]) at the time of data collection. Table 1 shows the characteristics of the sample.

TABLE 1 Clinical and demographic characteristics of the children included.

Outcomes	*N* (%)	Mean ± SD	Min–Max
Demographic characteristics			
Sex (male/female)	32 (45.7%)/ 36 (51.4%)		
Age (years)		9.26 ± 2.11	6–13
BMI (kg/m^2)		16.63 ± 2.04	12.60–22
Clinical characteristics			
FVC (L)		2.01 ± 0.55	1.07–3.40
FVC (% of predicted)		93.73 ± 10.39	72.7–124
FEV_1 (L)		1.71 ± 0.52	0.95–3.14
FEV_1 (% of predicted)		90.13 ± 13.87	50–137
FEV_1/FVC		87.78 ± 8.84	64–108.5
Exacerbation (past year)		0.60 ± 1.01	0–4
0	45 (66.2%)		
1–2	19 (28%)		
3–4	4 (5.8%)		
Hospitalizations (past year)		0.09 ± 0.33	0–2
0	63 (92.6%)		
1–2	5 (7.4%)		
Colonizations (past year)		0.74 ± 0.66	0–2
0	26 (38.2%)		
1–2	42 (61.8%)		

Abbreviations: BMI, body mass index; FEV1, forced expiratory volume in the 1st second; FVC, forced vital capacity; Max, maximum; Min, minimum; SD, standard deviation.

3.3 | Floor/ceiling effects and reliability

The descriptive statistics, floor/ceiling effects, internal consistency, and test–retest reliability of the child- and parent-report versions of the Spanish CFQ-R are shown in Table 2. The subscales "Emotional Functioning," "Social Functioning," and "Respiratory Symptoms" showed no floor/ceiling effect, whereas a ceiling effect was observed for the remaining subscales. The internal consistency of the child version of the Spanish CFQ-R was adequate, with a Cronbach's $\alpha > .60$ in all domains, except for the domains "Treatment Burden" ($\alpha = .42$) and "Social Functioning" ($\alpha = .57$). The test–retest reliability of these last two domains was acceptable/good (ICC ≥ 0.67). All other domains showed good test–retest reliability (ICC > 0.7). Lastly, the SEM of the various domains of the child version of the Spanish CFQ-R ranged from 8.3% to 15%, and the MDC_{90} ranged from 19.3% to 35%. Specifically, the domain with the lowest measurement error (lowest SEM and MDC_{90} values) was "Emotional Functioning," and the domain with the highest measurement error was "Body Image." The mean elapsed time between the first and second CFQ-R measurement to establish test–retest reliability was 8.40 ± 1.19 days (min–max: 7–10). No child presented an exacerbation between measurements. Of the participants, 4 children did not attend the second measurement (2 refused to continue with the study, and 2 did not return our call) and were therefore excluded from the test–retest analysis.

With respect to the parent-report version of the Spanish CFQ-R, all subscales showed a ceiling effect except for "Emotional Functioning," "Vitality," "School Functioning," and "Treatment Burden," with the subscale "Weight" also showing a floor effect. The internal consistency of the parent-report version of the Spanish CFQ-R was adequate, with a Cronbach's $\alpha > .60$ in all domains except for "Social Functioning" ($\alpha = .58$). The test–retest reliability of all the domains was also adequate (acceptable for "School Functioning" [ICC = 0.63] and good for the remaining subscales [ICC ≥ 0.77]). The SEM of the various Spanish CFQ-R parent-report domains ranged from 7.1% to 17.2%, whereas the MDC_{90} ranged from 16.5% to 40.1%. Specifically, the domain with the lowest measurement error (lowest SEM and MDC_{90} values) was "Physical Functioning," and the domain with the highest measurement error was "Weight."

3.4 | Construct validity

The correlations observed between the child-report version of the Spanish CFQ-R and the rest of the self-completed tests and clinical outcomes are shown in Tables 3 and 4. The convergent validity of the child-report version of the Spanish CFQ-R was adequate, given that all domains showed a direct and statistically significant correlation with the total KINDL score. The magnitude of these correlations was moderate in all cases, except for the subscale "Emotional Functioning," which was strong ($r = .67$). In addition, practically all subscales of the child-report version of the Spanish CFQ-R were moderately correlated with the KINDL domains "Physical Well-Being," "Emotional Well-Being," and "Self-Esteem."

TABLE 2 Descriptive statistics, floor/ceiling effects, internal consistency, and test–retest reliability of the child- and parent-report versions of the Spanish CFQ-R.

CFQ-R subscales	Trial 1 Mean ± SD	Floor effect (%)[a]	Ceiling effect (%)[a]	Internal consistency (Cronbach's α)	ICC (95% CI)	SEM	MDC_{90}
CFQ-R child-report version							
Physical functioning	81.6 ± 18.5	0	27.5	.71	0.79 (0.63–0.88)	9.8	22.9
Emotional functioning	76.6 ± 15.8	0	4.3	.74	0.72 (0.50–0.84)	8.3	19.3
Eating disturbances	76.0 ± 25.7	1.4	34.8	.74	0.84 (0.73–0.91)	12.6	29.4
Social functioning	65.5 ± 12.6	0	0	.57	0.67 (0.42–0.81)	10.2	23.8
Body image	67.0 ± 27.1	2.9	27.5	.60	0.80 (0.65–0.89)	15.0	35.0
Treatment burden	81.3 ± 17.8	0	34.8	.42	0.69 (0.46–0.82)	13.1	30.6
Respiratory symptoms	76.6 ± 18.3	1.4	11.6	.68	0.83 (0.71–0.90)	8.5	19.9
Digestive symptoms	78.7 ± 24.9	1.4	50.7	–	0.79 (0.63–0.88)	14.9	34.7
CFQ-R parent-report version							
Physical functioning	85.7 ± 17.4	0	32.4	.88	0.89 (0.80–0.94)	7.1	16.5
Emotional functioning	75.6 ± 16.9	0	10.3	.64	0.79 (0.62–0.88)	9.5	22.3
Vitality	65.5 ± 18.4	0	2.9	.71	0.81 (0.66–0.89)	9.5	22.3
Eating disturbances	72.1 ± 31.5	7.4	36.8	.76	0.88 (0.78–0.93)	13.6	31.7
Treatment burden	57.3 ± 28.9	0	10.3	.73	0.85 (0.73–0.92)	15.3	35.7
Body image	76.5 ± 27.4	1.5	39.7	.74	0.82 (0.69–0.90)	15.2	35.5
Health perceptions	78.3 ± 18.0	0	22.1	.72	0.83 (0.70–0.91)	9.4	22.0
School functioning	63.5 ± 17.6	0	2.9	.58	0.63 (0.34–0.79)	14.5	33.8
Weight	60.8 ± 39.9	22.1	41.2	–	0.89 (0.80–0.94)	17.2	40.1
Respiratory symptoms	78.3 ± 18.4	0	19.1	.82	0.85 (0.74–0.92)	8.7	20.3
Digestive symptoms	82.5 ± 13.6	0	26.5	.65	0.77 (0.59–0.87)	10.6	24.8

Abbreviations: CFQ-R, cystic fibrosis questionnaire-revised; CI, confidence interval; ICC, intraclass correlation coefficient; MDC, minimal detectable change; SD, standard deviation; SEM, standard error of measurement.

[a]Floor/ceiling effects: percentage of participants achieving the minimum/maximum score.

TABLE 3 Correlations between the child- and parent-report versions of the Spanish CFQ-R, as well as between the child-report version of the Spanish CFQ-R and the KINDL.

Outcome	CFQ-R child-report version (correlation assessed by the intraclass correlation coefficient [ICC])							
	Physical functioning	Emotional functioning	Eating disturbances	Social functioning	Body image	Treatment burden	Respiratory symptoms	Digestive symptoms
CFQ-R parent-report version								
Physical functioning	0.60	–	–	–	–	–	–	v
Emotional functioning	–	−0.03	–	–	–	–	–	–
Vitality	–	–	–	–	–	–	–	–
Eating disturbances	–	–	0.49	–	–	–	–	–
Treatment burden	–	–	–	–	–	0.26	–	–
Body image	–	–	–	–	0.43	–	–	–
Health perceptions	–	–	–	–	–	–	–	–
School functioning	–	–	–	–	–	–	–	–
Weight	–	–	–	–	–	–	–	–
Respiratory symptoms	–	–	–	–	–	–	0.51	–
Digestive symptoms	–	–	–	–	–	–	–	0.24
Outcome	**CFQ-R child-report version (correlation assessed by the Pearson's correlation coefficient [*r*])**							
	Physical functioning	**Emotional functioning**	**Eating disturbances**	**Social functioning**	**Body image**	**Treatment burden**	**Respiratory symptoms**	**Digestive symptoms**
KINDL child version								
Physical well-being	.52**	.5**	.51**	.48**	.36**	.30*	.41**	.30*
Emotional well-being	.58**	.55**	.31*	.51**	.30*	.13	.35**	.22
Self-esteem	.57**	.66**	.45**	.49**	.40**	.35**	.46**	.41**
Family	−.07	.20	.41**	.19	.14	.22	.12	.11
Friends	.26*	.36**	.17	.22	.3*	.25	.23	.13
Everyday functioning	.21	.26*	.22	.34**	.09	.22	.18	.11
Total score	.55**	.67**	.54**	.58**	.41**	.39**	.46**	.32*

Abbreviations: CFQ-R, cystic fibrosis questionnaire-revised; ICC, intraclass correlation coefficient.

**$p < .01$; *$p < .05$.

TABLE 4 Pearson's correlations between both Spanish CFQ-R versions and children's clinical outcomes.

	Exacerbations	Hospitalizations	Colonizations	FEV_1 (% of pred.)	FVC (% of pred.)	FEV_1/FVC
CFQ-R child-report version						
Physical functioning	−.55**	−.32*	−.37**	.06	.01	.04
Emotional functioning	−.42**	−.18	−.36**	.05	.02	.08
Eating disturbances	−.21	−.27*	−.3*	.11	.11	.11
Social functioning	−.09	−.34**	−.34**	−.01	.04	−.2
Body image	−.09	−.12	−.11	.04	−.04	.04
Treatment burden	−.25*	−.20	−.29*	.11	.07	.01
Respiratory symptoms	−.36*	−.24*	−.12	.19	.15	.17
Digestive symptoms	−.13	−.01	−.05	.26*	.2	.15
CFQ-R parent-report version						
Physical functioning	−.58**	−.08	−.43**	.13	.14	.004
Emotion functioning	−.08	−.18	−.15	.16	.16	.16
Vitality	−.25*	−.11	−.27*	.1	.12	.07
Eating disturbances	−.19	.14	−.22	−.02	.004	.06
Treatment burden	−.24*	−.03	−.04	.08	.05	.06
Body image	−.17	−.08	−.13	.04	.01	.08
Health perceptions	−.32**	−.03	−.14	.11	.05	.05
school Functioning	−.23	−.01	−.14	.19	.18	.16
Weight	−.12	−.07	−.21	.03	.03	.07
Respiratory symptoms	−.52**	−.06	−.16	.15	.12	.1
Digestive symptoms	−.1	.02	−.21	.32*	.29*	.24*

Abbreviations: CFQ-R, cystic fibrosis questionnaire-revised; FEV1, forced expiratory volume in the first second; FVC, forced vital capacity; % of pred., percentage of predicted.

**$p < .01$; *$p < .05$.

Overall, the subscales of the child-report version of the Spanish CFQ-R showed fair agreement/correlation with their counterpart subscale in the parent-report version (ICC > 0.40), except for the subscales "Treatment Burden" (ICC = 0.26), "Digestive Symptoms" (ICC = 0.24), and "Emotional Functioning" (ICC = −0.03), where poor agreement/correlation was observed. The subscale "Physical Functioning" showed the highest correlation between the versions (ICC = 0.60).

In terms of the children's clinical outcomes, the two variables that had the largest number of statistically significant inverse correlations with the subscales of both versions of the CFQ-R were the number of exacerbations and colonizations in the past year. Specifically, the number of exacerbations showed the most correlations with the parent-report version, whereas the number of colonizations correlated with the child-report version. In both versions, the subscale "Physical Functioning" showed the highest correlation (moderate; $r = -.37$ to $-.58$) with both the number of exacerbations and the number of colonizations. However, lung function was not related to any of the CFQ-R versions, except for the "Digestive Symptoms" subscale of the parent-report version of the Spanish CFQ-R, which showed a direct and weak correlation with all lung function parameters ($r \leq .32$).

4 | DISCUSSION

The present study aimed to adapt and evaluate the psychometric properties of the child- and parent-report versions of the CFQ-R in the Spanish population. The analyzed sample comprised a broad representation of children with CF aged 6−13 years in Spain, and the study's sample size was larger than that of other validations, such as the Turkish and Iranian studies.[16,20] Our results support the validity and reliability of the child- and parent-report versions of the Spanish CFQ-R, both of which showed acceptable internal consistency and stability over time (test−retest reliability), as well as adequate construct validity due to their relationship with another HRQoL questionnaire (KINDL) and with the participants' health status. Our study establishes the MDC_{90} for each subscale of both versions,

which helps determine the efficacy and potential adverse/secondary effects of various interventions to improve the quality of life of children with CF, such as the recently approved ETI therapy.[6,39] Based on the above, these Spanish versions of the CFQ-R could be applied during routine medical check-ups to monitor the quality of life and respiratory symptoms of children with CF in Spain, allowing changes to be detected and for appropriate early treatment to be applied.[21,23]

4.1 | Reliability

Our results reinforce those obtained in the original CFQ-R validation,[11] showing no floor effect in either version of the questionnaire, except for the "Weight" subscale in the parent-report version. The CFQ-R has therefore been shown to be capable of identifying children with varying degrees of impairment in their quality of life. However, both versions assessed in this study showed a ceiling effect for virtually all subscales, except for "Emotional Functioning" and "Social/School Functioning." Consistent with our findings, the subscales that showed a higher ceiling effect in the child- and parent-report versions were the same as in the original version.[11] The presence of a ceiling effect in this Spanish version and in the original[11] version could be attributed to improvements in the early detection of CF,[5] as well as in the implementation of treatment and healthy behavioral habits, which minimize and delay the negative consequences attributed to the disease.[40,41] The CFQ-R can therefore identify children with varying degrees of impairment in their quality of life, except in those cases in which the impairment is mild. In fact, the only subscales in which a significant number of participants did not achieve the maximum score were those related to emotional/social aspects and respiratory symptoms, which could be the domains least likely to be affected by early CF detection.[42] With the use of new modulator therapy that virtually eliminates the consequences of CF,[6] however, this hypothesis could be tested in future studies.

There were practically no differences between the children's reports about how CF affects their quality of life and their parents' perceptions of the effect. This absence of differences was expected based on the results from previous validations, given that most of the subscales focus on observable behaviors.[9] Surprisingly, however, the burden of treatment was considered higher by the parents than by the children; the ceiling effect in the included children was 34.8%, who did not consider the treatment burden excessive. This finding could be attributed to the high levels of stress that parents of children with CF develop due to perceiving their child's increased vulnerability. The parents' anxiety and sadness can persist and intensify periodically throughout their child's life, especially in relation to medical events such as hospital admissions.[42] Similarly, such a discrepancy reflects the relevance of applying the CFQ-R to both the child- and parent-report versions, because for cognitive processing and for environmental reasons, the perspective on HRQoL can differ.[9,21,42] On the other hand, the greater perception by parents that the treatment regimen is very burdensome could also be explained by the fact that most of the burden of scheduling treatments, setting up the equipment and cleaning it falls on the parents, not on the children in this age range. However, this situation is likely to change with the introduction of the new modulator therapy. Future studies are needed to help clarify this issue and determine the influence of modulator therapy on the treatment burden by reducing the need for other therapies.

The internal consistency of both Spanish versions of the CFQ-R was similar to that of previous validated versions,[11] showing an acceptable/good internal consistency for all subscales ($\alpha \geq .60$). The only notable discrepancy was in the "Treatment Burden" subscale of the child-report version, in which the internal consistency ($\alpha = .42$) was markedly lower than the cut-off established as acceptable for the new scales ($\alpha = .60$). These findings again highlight how a child's perspective of the treatment burden can fluctuate significantly over short periods, due in part to their still developing cognitive processes.[42] The subscales with the lowest test–retest reliability in our study (although in both cases the reliability was fair) were "Social Functioning" and "School Functioning" for the child- and parent-report versions, respectively. Based on these results, it is possible to hypothesize that the psychological and physical burden attributable to experiencing a chronic illness is partially responsible for the variability observed in context-related subscales.[21,23,42,43] Reinforcing this hypothesis, the reliability for the emotional state subscale obtained values highly similar to those obtained for "Social Functioning."

Overall, the test–retest reliability of the parent-report version of the Spanish CFQ-R proved to be superior to that of the child-report version for all counterpart subscales. The higher reproducibility in the parents' version was attributed to their strong commitment to maintaining their children's health,[23] paying close attention to many of the aspects addressed in the CFQ-R in relation to their children's quality of life.[13,21,42] A higher recall ability compared with their children cannot be ruled out, although this difference should be minimized by the time interval between the measurements. The German version[17] of the CFQ-R obtained almost identical results to ours in test–retest reliability for both versions, except for the subscales "Weight" (parent-report version) and "Digestive Symptoms" (child-report version). The lack of other studies assessing the test–retest reliability of the CFQ-R makes further comparisons impossible, and more studies assessing the stability of the questionnaire over time are needed.

This is the first study to assess the SEM and MDC_{90} of all CFQ-R subscales, and knowledge of these values is essential in the clinical setting[37] because it helps determine with a high probability of accuracy whether there is a real improvement or deterioration in a child's HRQoL. SEM values <10% indicate small variability of the measure and adequate reliability.[44] The subscales "Body Image" (15%), "Digestive Symptoms" (14.9%), and "Treatment Burden" (13.1%) had the highest SEM for the child-report version, and "Weight" (17.2%), "Treatment Burden" (15.3%), and "Body Image" (15.2%) had the highest for the parent-report version, which were the

least accurate subscales of these versions. Due to their close relationship, these subscales had an MDC_{90} > 30%, which could be considered too demanding, whereas a number of authors have considered that a change must exceed 30% to be clinically relevant.[36,37] With certain exceptions, both versions showed highly similar SEM and MDC_{90} values between the counterpart subscales, which reinforces their measurability. In any case, further studies are needed to corroborate or contradict the results of the present study.

4.2 | Construct validity

The convergent validity of the child-report version of the Spanish CFQ-R was adequate, given that all subscales of this version correlated moderately with the total KINDL score (except for "Treatment Burden"), results similar to those of the Turkish version.[16] Other validations did not analyze its correlation with other HRQoL measurements,[11,17] except for the Iranian version, which correlated CFQ-R with Pediatric Quality of Life Inventory version 4.0 and obtained adequate results with lower correlations in "Social Functioning."[20] The subscales most related to quality of life were those addressing emotional and social factors, reinforcing the ample evidence that improving psychosocial factors is fundamental to increasing the quality of life of children with CF.[42]

Similar to the results obtained in CFQ-R validity studies,[11,17,45] there were adequate agreement between counterpart subscales of the child- and parent-report versions. Thus, the correlation between the child- and parent-report items for the Spanish version showed a stronger relationship for subscales measuring more observable signs (e.g., "Physical Functioning") and a weak relationship for subscales measuring symptoms/perceptions (subjective measures). The lack of an association in the subscale "Emotional Functioning" between parents and children can be explained by differing perceptions of the social and personal context.[9,21] In fact, the influence of experiencing exacerbations and colonizations was related to emotional state in children but not in parents, showing different ways of experiencing the same context.[23,42] Another possible explanation could be that parents are not aware of their children's emotional functioning, as children often do not disclose their sadness or anxiety as much as they can feel these emotions to protect their parents. Thus, it is important therefore, not to use only the parents' report and to use the child's report of "Emotional Functioning" given that it more closely reflects the child's emotions. In addition, physical ability was directly related to children's emotional state, probably because impaired physical ability limits activities and participation.[42] The International Classification of Functioning, Disability and Health points out the need to improve the psychosocial aspects, such as limits in participation, which act as barriers to improving the quality of life of children with CF.[46,47]

In contrast to the Spanish version of the CFQ-R for patients 14 years and older[19] and the English[11] and Iranian[20] versions, this study found no correlation between lung function and subscales scores. In recent years, there has been a qualitative leap in the early detection of CF in Spain,[48] as well as in the treatments and lifestyle guidelines recommended to minimize the adverse effects of this disease.[3] Thus, almost all of the children assessed in the study had normal lung function (FVC and $FEV_1 \geq 90\%$ of predicted). In addition, the subscale "Respiratory Symptoms" focuses mainly on coughing, given that these children are intimately familiar with everyday physiotherapy techniques aimed at promoting expectoration to reduce the risk of infection.[23] Many children might therefore consider that they cough regularly and that it does not reflect a negative aspect of their quality of life, which could explain why no relationship was observed in our study between lung function and the CFQ-R, in contrast to other validations.[13,20] However, the subscale "Respiratory Symptoms" correlated significantly with exacerbations–showing that for those children with symptoms and lung complications. Thus, our findings may suggest that the subscale CFQ-R respiratory symptoms is associated with the lung function outcomes, but no significant correlation was found because the children had normal lung function.

4.3 | Limitations

As a main limitation, the study sample could have been larger; however, Spain has a low prevalence of children whose age is within the study's age range. Efforts were made to include children and families from different territories. However, similar results have been achieved in previous validations of this scale.[11,16,17,20] In addition, this is a cross-cultural adaptation to Spanish that only included children who live in Spain; the results cannot therefore be extrapolated to children from other Spanish-speaking countries.

5 | CONCLUSION

The child- and parent-report versions of the Spanish CFQ-R have been demonstrated to be valid and reliable for use in the Spanish population for clinical and research purposes. The CFQ-R can therefore be administered both before and after starting modulator therapy to assess whether it is affecting daily functioning. The MDC_{90} established in the present study will help identify with a high probability whether actual changes have occurred in the various quality of life domains of children with CF.

AUTHOR CONTRIBUTIONS

Paula Blanco-Orive: Writing—original draft; investigation; methodology; validation; writing—review and editing; data curation. **Tamara del Corral**: Conceptualization; funding acquisition; writing—original draft; writing—review and editing; supervision; project administration. **Patricia Martín-Casas**: Conceptualization; funding acquisition; writing—original draft; writing—review and editing; supervision; project administration. **Maria Àngels Cebrià I Iranzo**: Investigation; methodology; data curation; validation; resources; writing—review and editing. **Cristina Godoy-Nieto**: Investigation; methodology; validation; writing—review and editing; data curation; resources. **Ibai**

López-de-Uralde-Villanueva: Conceptualization; investigation; writing—original draft; methodology; validation; writing—review and editing; data curation; software; formal analysis.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

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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