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Original Article

Psychometric Properties of the Persian Version of Cystic Fibrosis Questionnaire: Revised in Children with Cystic Fibrosis

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Abstract

Background: Cystic fibrosis is a progressive, fatal disease affecting the quality of life. The cystic fibrosis questionnaire-revised (CFQ-R) is an efficient tool to monitor health-related quality of life in patients. The aim of this study was to explore the psychometric properties of the child and parent versions of the Persian version of the CFQ-R in the Iranian population. **Methods:** Fifty children with cystic fibrosis (6–11 years) and their parents were allocated in this methodological study to examine convergent validity, discriminant validity, test-retest reliability (n = 30), internal consistency, ceiling and floor effects, and agreement between two versions of the CFQ-R. **Results:** Convergent validity was confirmed for parent proxy ($P < 0.05$). CFQ-R discriminated patients among stages of disease severity based on lung function, age, and BMI ($P < 0.05$). Test-retest analysis revealed good to excellent reliability (inter-class correlation coefficient (ICC) = 0.78–0.97). In most domains, lower quality of life scores was obtained in the parent proxy compared to the child version ($P < 0.05$). Domain-specific correlations were found between the child version and parent proxy ($P < 0.05$). Internal consistency was generally confirmed (α = 0.13–0.83 in child version and α = 0.25–0.87 in parent proxy). There were no floor effects. Ceiling effects were mostly seen for physical, digestion, and body image domains in the child version and for eating, weight, and school domains in the parent proxy. **Conclusions:** The child version and parent proxy of the Persian CFQ-R are valid and reliable measures and can be applied in clinical trials to monitor the quality of life in children with cystic fibrosis. It is recommended to use both versions in conjunction to better interpret the quality of life aspects of children with cystic fibrosis.

Keywords: *Cystic fibrosis, reliability, validity*

Introduction

Cystic fibrosis (CF) is one of the life-limiting genetic diseases among children, affecting multiple organs such as digestive and pulmonary systems.[1,2] The prevalence of CF is about 1 in 3000 live births.[1,2] Pulmonary involvement as a major cause of death in patients with CF is identified by bronchial infection, excessive inflammation, progressive airway obstruction, bronchiectasis, and eventual respiratory failure.[3]

Because of the chronic and disabling nature of CF, evaluating the quality of life (QoL) of these patients is crucial.[4] World Health Organization defines QoL as the person's feeling about his/her current status of beliefs, expectations, standards, and interests in the framework of values and cultural context. QoL encompasses variety of physical, psychological, and social elements.[5] The assessment of QoL should be performed by standard measures which are disease specific. These measures are applicable to evaluate the efficacy of clinical trials in chronic and progressive diseases.[6,7] In this regard, different questionnaires were introduced for CF diseases, such as cystic fibrosis questionnaire-revised (CFQ-R),[3] Cystic Fibrosis Quality of Life (CFQoL),[8] and disease-specific scale of the questions on life satisfaction for adolescents and adults with CF (FLZM-CF)[9] among which, the CFQ-R is the most validated and utilized questionnaire.[10] Indeed, CFQ-R is the only disease-specific health-related measure of the CF disease that consists of children and adult versions and, therefore, can monitor health-related QoL over the life span in these patients.[10] The psychometric properties of the CFQ-R were reported in many languages, such as French,[7]

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English,[11] German,[12] Dutch,[13] and Turkish,[14] but there is no evidence in Persian language or Persian-spoken countries. Therefore, this study aimed to translate the CFQ-R to the Persian language and explore the psychometric properties of the Persian version of CFQ-R in the Iranian population.

Methods

Study design and participants

Fifty children with confirmed CF disease and their parents volunteered to participate in this methodological and cross-sectional study.[14] Children aged 6–11 in a stable disease condition and able to read and write in Persian were included in this study. The children with language problems, pulmonary exacerbations, hospitalization in the previous six weeks[15] and who were receiving oral and/or venal antibiotics were excluded. Also, the children who were not interested in participating in this study were excluded at any time they desired. Thirty children and their parents also agreed to participate in the reliability (test-retest) phase of the study. Children with unstable health and disease complications in the period of ten days to two weeks based on the parents' affirmation were not included in this phase.[8] A written informed consent was obtained from parents. This study was approved by the local ethics committee of Tabriz University of Medical Sciences (IR.TBZMED.REC.1397.895).

Procedures

The English version of CFQ-R for child and parent proxy was first translated into Persian, and face and content validity of them were confirmed. Then, the validation processes, including convergent validity (as construct validity), discriminant validity (as construct validity), test-retest reliability (with almost two weeks interval between two test occasions), internal consistency, calculating ceiling and floor effect and also agreement between two versions of the CFQ-R, was conducted. For this purpose, demographic information, such as age, gender, body mass index as a child's nutritional state (weight/height expressed as kg/m^2), was recorded during routine visits of CF children. Then, the questionnaires including CFQ-R and Pediatric Quality of Life Inventory™ 4.0 (PedsQL™) were completed by children and the children's primary caregivers under the supervision of a specialized physiotherapist in a quiet room. After the fulfillment of the questionnaires, children underwent pulmonary function tests. All processes were conducted in the Iranian outpatient CF center in Tehran which lasted for 18 months October (from 2018 to 2019). A pediatrician supervised these procedures.

Translation process

The translation process is applied in a forward and backward fashion by four native Persian speakers based on international guidelines. Backward translation was approved by an additional authorized translator and an assistant professor of linguistics sciences. We used forward and back translation by selecting translators, employing a review team, and pilot testing the target population. Selected translators were academic members who were proficient forward translators and were fluent in both the source and target language and knowledgeable in the purpose and intent of the instrument. The back translators were equally qualified, but they were not familiar with the original version of the instrument. The translators were blind to each other.[16] The final version was prepared under the supervision of the corresponding author and the coauthor was Pediatrician. The permission of the CFQ-R developer was obtained for backwardly translated versions. Minor revisions were made in the Persian version because of the linguistic differences to improve the simplicity and clarity of the content.

Content validity

Fifteen academic members (six physiotherapists with a PhD and six pediatricians, one psychologist with a PhD, one occupational therapist with a PhD, and one speech therapist with a PhD) participated in an expert panel in multiple sessions to qualify the content validity of the CFQ-R questionnaires. The quantity of the content validity was examined by calculating the content validity ratio (CVR) and content validity index (CVI).

Face validity

Face validity was also examined by an expert panel qualitatively and quantitatively. Items based on difficulty, irrelevancy, and ambiguousness were examined qualitatively and minor corrections were made. Then, the impact score was calculated on a five-point Likert scale for each item.

Instruments

Cystic fibrosis-Revised questionnaire

CFQ-R is a disease-specific measure to report health-related QoL in children with CF. It is presented in four versions. CFQ-R child version is designed for children aged 6–11 years and consists of 35 items (The number of items in each domain is presented in parenthesis after each domain) in 8 domains, including physical symptoms (6), emotional functioning (8), social functioning (7), body image (3), eating disturbances (3), treatment burden (3), respiratory symptoms (4), and digestive symptoms (1). Children were interviewed and the best response was chosen using special answering cards. This procedure was conducted to ensure that the children understood the items well. The parent proxy is a self-report questionnaire to demonstrate the child's health-related QoL from the parent's point of view. The proxy comprises 43 items (The number of items in each domain is presented in parenthesis after each domain) in 11 domains consisting

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of physical symptoms (9), emotional functioning (5), school functioning (3), body image (3), eating disturbances (2), treatment burden (3), respiratory symptoms (6), digestive symptoms (3), vitality (5), overall health perceptions (3), and weight (1). These questionnaires lack a general score. The QoL for each domain is computed based on the sum of the scores of subscales. Questions of each questionnaire are scored on a four-point scale. Raw scores for each domain are converted to standard scores ranging from 0 (low) to 100 (high). Higher scores indicate better QoL. The time needed to complete each questionnaire is about 15 min.[17]

Pediatric Quality of Life Inventory™ 4.0

The PedsQL is a generic questionnaire representing health-related QoL in children. The validated Persian PedsQL is available and it contains age-appropriate children and parents. PedsQL consists of 23 questions exploring QoL in 4 domains, including physical, emotional, social, and school dimensions. In PedsQL, each item was rated on a four-point Likert scale, and the total score of each domain was calculated for analysis. A lower score indicates better QoL. The time needed to complete the questionnaire is approximately 15 min.[18]

Pulmonary function test

In the present study, forced expiratory volume in the 1 second (FEV_1) was measured to represent the lung function. The percentage of the predicted value was calculated.[19] The amount of FEV_1 was implied to classify disease severity into two groups with mild involvement (≥70%) (n = 34) and moderate/severe involvement (<69%) (n = 16).[20] We classified patients into only two groups because the majority of the children meeting our criteria had mild and moderate lung involvement and only five patients had severe involvement.

Statistical analysis

Statistical analysis was done using Statistical Package for Social Sciences (SPSS software version 25 Chicago IL, USA). P values less than 0.05 were considered significant. The content validity of the Persian versions of CFQ-R was obtained by calculating CVR and CVI. Because of the number of experts participating in this study (n = 15), CVR >0.49 were acceptable. Also, CVI values >0.79 were accepted.[21] Item impact method was applied to quantify the face validity of each item, and the quantities greater than 1.5 were considered acceptable.[22] Correlations between CFQ-R and PedsQL questionnaires in child and parent proxy were measured using Spearman's correlation coefficient to obtain convergent validity. Spearman's correlation coefficient was also applied to assess the agreement between the child version and parent proxy. Further, the Spearman's correlation coefficient was used to explore discriminant validity by examining the relationships between the domain scores and disease severity (FEV_1), BMI, and child's age. The relationships were classified as strong (>0.5), moderate (0.35–0.5), weak (0.2–0.35), and very weak (<0.2).[23] A comparison of QoL between two groups of children with mild and moderate/severe disease was made by Mann-Whitney U test. ICC (Model: two-way mixed, Type: single rater, Definition: absolute agreement)[24] was used to analyze the test-retest reliability and also to interpret the agreement between two versions of the CFQ-R.[25] The ICCs were interpreted according to the Landis and Koch criteria (0–0.2 as poor, 0.21–0.4 as fair, 0.41–0.7 as moderate, 0.71–0.80 as substantial, and 0.81–1 as almost perfect).[26] The standard error of measurement (SEM) was calculated using the ICC with formulae (SEM = $Z\alpha * SD * \sqrt{1 - ICC}$), where $Z\alpha$ is the Z-score for the 95% CI of the true score about the observed score ($Z\alpha$ = 1.96) and SD = is the test's SD.[27] Wilcoxon test was also used to compare the QoL between the child and parent versions. Internal consistency was evaluated using Cronbach's alpha. Alpha coefficients >0.6 were satisfactory for all domains.[28] Ceiling and floor effects were calculated for each item. The percentages of the respondents who were rated lower than 5% and the percentages of the respondent who rated equal or higher than 95% were considered the floor and ceiling effects, respectively.[17]

Results

The demographic characteristics of children with CF are presented in Table 1. In that, 18.4% of the children were at preschool level. Only 2% of them were from rural areas. Among respondents as caregivers, 68% were mothers, 30% were fathers, and 2% were grandmothers. Thirty percent of parents were undergraduates (had a primary school degree) and 22.5% had a university degree.

The CVR and CVI results are presented in Table 2. Item impact scores also are presented in Table 3.

There were relationships between CFQ-R and PedsQL questionnaires. The strong relationships were found between parent proxy and PedsQL in physical and emotional symptoms. (R = −0.537, and R = −0.561, respectively). There was no relationship between CFQ-R and PedsQL in the children's version [Table 4].

Table 1: Demographic and clinical characteristics of children with CF

Characteristics	Child
All patients *N*,	50
Child's age (year) at CFQ-R, mean±SD	8.47±1.78
Gender *n*, (%)	
Male	25 (50)
Female	25 (50)
BMI (Kg/m^2), mean±SD	14.37±1.65
FEV_1 (%), mean±SD	88.34±34.13

BMI: body mass index; CFQ-R: cystic fibrosis questionnaire-revised; FEV1: forced expiratory volume in 1 second; SD: standard deviation

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Table 2: CVR and CVI values for CFQ-R questionnaire (child version and parent-proxy)

Item	Child version		Parent version	
	CVR	CVI	CVR	CVI
1	0.87	0.96	1	1
2	1	1	1	1
3	1	1	1	1
4	1	1	1	0.98
5	1	1	1	0.98
6	1	1	1	1
7	1	0.96	1	1
8	0.87	0.96	1	1
9	0.87	0.87	1	0.93
10	1	0.87	1	1
11	1	0.93	1	0.98
12	0.87	1	1	1
13	0.87	1	1	1
14	1	1	1	0.93
15	1	1	1	0.93
16	1	1	1	1
17	1	0.91	1	1
18	0.87	0.98	1	1
19	1	0.98	1	0.93
20	1	0.98	1	0.93
21	1	1	1	0.93
22	0.73	0.93	1	0.93
23	0.73	0.80	1	1
24	1	0.80	0.87	1
25	1	1	1	0.89
26	1	0.89	1	0.96
27	1	1	0.87	0.98
28	1	1	0.87	1
29	1	1	1	1
30	1	0.93	1	1
31	1	1	1	1
32	1	1	0.87	1
33	1	0.89	1	1
34	1	1	1	0.93
35	1	1	1	1
36	-	-	1	1
37	-	-	1	1
38	-	-	1	1
39	-	-	1	1
40	-	-	1	1
41	-	-	1	1
42	-	-	1	1
43	-	-	1	1
44	-	-	1	1

Table 3: Item impact scores for two versions of the CFQ-R

Item	Child version-impact score	Parent version-impact score
1	4.49	5
2	4.62	4.58
3	4.70	4.69
4	4.12	4.72
5	4.49	4.27
6	4.47	4.89
7	4.63	4.80
8	4.25	5
9	4.30	4.80
10	4.63	4.89
11	4.67	3.70
12	4.49	4.89
13	4.12	4.89
14	4.69	4.72
15	4.80	4.72
16	4.72	4.67
17	3.39	3.39
18	4.43	4.72
19	4.69	4.70
20	4.63	4.80
21	4.80	4.82
22	2.96	4.80
23	4.03	4.80
24	4.23	4.80
25	4.69	4.34
26	4.03	4.32
27	4.47	4.80
28	4.66	4.72
29	4.67	4.35
30	4.89	4.70
31	4.89	4.80
32	4.89	4.89
33	4.80	4.89
34	4.89	5
35	4.80	5
36	-	4.89
37	-	4.83
38	-	5
39	-	4.82
40	-	5
41	-	4.72
42	-	4.89
43	-	4.32
44	-	4.80

The relationships between CFQ-R scores and age, disease severity, and BMI are presented in Table 5. A comparison of group differences (mild vs. moderate/severe) revealed no difference in the child's version ($P > 0.05$). In parent proxy, the QoL of children with moderate/severe disease was significantly lower than children with mild disease in emotional (67.50 ± 17.13 in moderate/severe compared to 78.53 ± 15.20 in mild; $P = 0.025$ as mean ± SDs), body image (48.44 ± 16.45 in moderate/severe compared to 67.40 ± 27.32 in mild; $P = 0.019$ as mean ± SDs), eating (61.72 ± 23.92 in moderate/severe compared to 77.94 ± 18.22 in mild; $P = 0.022$ as mean ± SDs), treatment burden (67.19 ± 16.80 in moderate/severe compared to 77.45 ± 15.56 in mild; $P = 0.048$ as mean ± SDs),

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Table 4: The relationships between CFQ-R questionnaires and PedsQL questionnaires for each version using Spearman's correlation coefficients at specific domains

Domain	Child version		Parent version	
	Correlation coefficient	*P*	Correlation coefficient	*P*
Physical symptoms	−0.207	0.460	−0.537	0.039*
Emotional functioning	−0.419	0.120	−0.561	0.009*
Social/school functioning	0.14	0.961	0.011	0.970

*Indicates significance (lesser than 0.05)

and respiratory domains (50.67 ± 12.32 in moderate/severe compared to 70.80 ± 13.58 in mild; $P < 0.001$ as mean ± SDs).

The results of test-retest reliability are demonstrated in Table 5. Each domain has reached almost perfect reliability except for the treatment burden in parent proxy (ICC = 0.78) [Table 6].

The amount of agreement between the child version and parent proxy ranged from poor to moderate based on the ICC analysis. The results of Spearman's correlation analysis are demonstrated in Table 6. Comparing the domains between two versions revealed that parents rated lower in physical ($P < 0.001$), emotional ($P = 0.003$), body image ($P = 0.011$), eating ($P = 0.006$), and respiratory domains ($P < 0.001$) compared to children. There was no difference between parent and child versions in treatment burden ($P = 0.708$) and digestion ($P = 0.769$) [Table 7].

The findings of internal consistencies using Cronbach's alpha are demonstrated in Table 8.

Analyzing the ceiling and floor effects showed that: in the child version, ceiling effects ranged from 4% to 48% which were mostly seen for physical, digestion, and body image domains (48%, 38%, and 34%, respectively). In parent version, ceiling effect ranged from 0% to 24% which is most obvious for eating, weight, and school domains (24%, 24%, and 20%, respectively). There were no floor effects.

Discussion

This study aimed to explore the psychometric properties of the Persian CFQ-R (children 6–11 years old and parent proxy). Findings revealed satisfactory content and face validity for both versions.

Convergent validity

In the present study, there was no significant correlation between children's CFQ-R and PedsQL. But, there were strong relationships for parent proxy in physical and emotional domains. PedsQL is a generic questionnaire and can be adapted with corresponding items in CFQ-R. Of course, only four domains are adaptable (physical, emotional, social, and school). Compared with our results, Yuksel *et al*. (2013) reported good convergent validity for Turkish children CFQ-R (using KINDL questionnaire) in most domains except the school domain.[14] Because of different questions designed for KINDL and PedsQL, comparing the results is not possible. Significant correlations are also demonstrated between adult CFQ-R and SF-36.[8,11] Absence of a relationship between CFQ-R child version and PedsQL may cause uncertainty about responses. Indeed, considering agreements between the child version and parent proxy in the Persian language, it is likely that relying only on children's opinions about their QoL is not logical and it is a need to consult about their general health with their parents to better understand the child needs and to better conduct the clinical plan. Also, the generic nature of PedsQL is not comparable with specific items designed for CFQ-R completely because of different questions applied.[4]

Discriminant validity

Based on the results, in both versions, age is not correlated to the QoL domains except for a weak relationship between age and social functioning in the child version (R = 0.282). The findings are in accordance with the literature.[14] Examining the wider age range may reveal different results. Because of the progressive nature of the CF disease, it is expected that the patient's health-related complications worsen with increasing age and consequently, patients do not feel well at older ages.[8,11,29]

Disease severity in CF is commonly expressed by changes in $FEV_{1.}$[17] In the child version, social and respiratory domains are moderately correlated to the FEV_1. In parent proxy, physical, emotional, body image, eating, and respiratory domains are correlated to the FEV_1 with similar trends where the highest amount of correlation was obtained for respiratory symptoms. Our results reveal good discriminant validity compared to the previous findings in child and parent versions.[12,17] In this area, Modi and Quittner (2003) did not find a meaningful association between FEV_1 and QoL domains which were attributed to the limited range of disease severity in their study population.[20] Generally, better pulmonary functioning is expected in non-hospitalized CF children in this age group compared to older adults and/or hospitalized patients.[17]

According to the results, BMI is correlated modestly with body image and weight in parent proxy (R = 0.453, R = 0.421, respectively). Quittner *et al*. (2012) also found poor correlations between eating and BMI in the children and parent versions (n = 2068 children, R = 0.22 and R = 0.24, respectively). In the parent proxy, a similar relationship was found between weight and BMI.[17] Similar findings are demonstrated in adults for body image and weight.[29]

Overall, a possible explanation of the disparity between the results of the studies may be due to the methodological

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Table 5: The relationships between CFQ-R scores and age, disease severity, and BMI

Domain	Child version			Parent version		
	Age (year)	FEV_1 (%)	BMI (kg/m^2)	Age (year)	FEV_1 (%)	BMI (kg/m^2)
Physical symptoms	R=0.164	R=0.045	R=0.192	R=0.077	R=0.380	R=0.181
	P=0.256	*P*=0.756	*P*=0.181	*P*=0.593	*P*=0.030*	*P*=0.210
Emotional functioning	R=−0.073	R=−0.020	R=−0.170	R=−0.072	R=0.286	R=0.160
	P=0.616	*P*=0.893	*P*=0.237	*P*=0.620	*P*=0.044*	*P*=0.267
Social/school functioning	R= −0.282	R=−0.283	R=−0.228	R=0.068	R=0.091	R=0.175
	P=0.047*	*P*=0.047*	*P*=0.112	*P*=0.640	*P*=0.532	*P*=0.225
Body image	R=0.062	R=0.084	R=0.223	R=−0.199	R=0.349	R=0.435
	P=0.670	*P*=0.562	*P*=0.120	*P*=0.165	*P*=0.013*	*P*=0.002*
Eating disturbances	R=0.063	R=0.229	R=0.098	R=0.064	R=0.332	R=0.038
	P=0.662	*P*=0.109	*P*=0.497	*P*=0.660	*P*=0.018*	*P*=0.795
Treatment burden	R=0.243	R= −0.030	R=−0.208	R=−0.136	R=0.268	R=0.201
	P=0.089	*P*=0.837	*P*=0.147	*P*=0.347	*P*=0.060	*P*=0.161
Respiratory symptoms	R=0.174	R=0.302	R=0.212	R=−0.249	R=0.545	R=0.216
	P=0.226	*P*=0.033*	*P*=0.140	*P*=0.081	*P*<0.001*	*P*=0.133
Digestive symptoms	R= −0.066	R=−0.204	R=−0.136	R=−0.029	R=095	R=0.139
	P=0.649	*P*=0.156	*P*=0.345	*P*=0.842	*P*=0.511	*P*=0.337
Vitality	–	–	–	R=0.092	R=0.128	R=0.107
	–	–	–	*P*=0.527	*P*=0.377	*P*=0.459
Overall health perception	–	–	–	R=−0.027	R=0.278	R=0.035
	–	–	–	*P*=0.853	*P*=0.051	*P*=0.807
Weight	–	–	–	R=−0.250	R=0.142	R=0.421
	–	–	–	*P*=0.080	*P*=0.324	*P*=0.002*

R: Spearman's correlation coefficient; * indicates significance (lesser than 0.05)

Table 6: The results of test-retest analysis for CFQ-R versions

Domain	Child version			Parent version		
	ICC	95% CI	SEM	ICC	95% CI	SEM
Physical symptoms	0.92	0.83–0.96	0.646	0.96	0.93–0.98	1.176
Emotional functioning	0.88	0.75–0.94	0.514	0.91	0.82–0.96	0.546
Social/school functioning	0.88	0.76–0.94	0.636	0.92	0.83–0.96	0.521
Body image	0.92	0.83–0.96	0.606	0.97	0.94–0.98	0.556
Eating disturbances	0.88	0.75–0.94	0.259	0.85	0.68–0.92	0.267
Treatment burden	0.93	0.86–0.96	0.374	0.78	0.55–0.89	0.317
Respiratory symptoms	0.91	0.81–0.95	0.436	0.93	0.86–0.96	0.733
Digestive symptoms	0.84	0.66–0.92	0.129	0.81	0.60–0.91	0.316
Vitality	–	–	–	0.95	0.91–0.98	0.431
Overall health perception	–	–	–	0.91	0.82–0.95	0.354
Weight	–	–	–	0.85	0.69–0.93	0.200

CI: confidence interval; SEM: standard error of measurement; ICC: intra-class correlation coefficient

differences in the inclusion criteria. For example, sick and well-visited children classification at the time of the questionnaire admission was considered in one survey[17] but, similar to the others,[12,20] children with stable conditions who were not hospitalized were included in our study. Hospitalization and exacerbation are major factors affecting health outcomes in children with CF.[14,17,30] So, different clinical outcomes might be seen in different patient groups, and consequently, the QoL measures could be affected. The other explanation may be the cultural differences affecting disagreements between the child and parents about the child's health status. In this regard, parents in our study believed that children with moderate/severe disease (grouping by the FEV_1) were in a lower health condition in some aspects, including emotional, body image, eating, treatment, and respiratory issues. However, children did not believe in being different.

Generally, multiple aspects of health-related QoL in patients with CF are dependent on disease severity.[12,20,29]

Test-retest reliability

Reliability analysis provides basic information about health measures to monitor the disease change in a specific time span precisely or examine the effects of a specific treatment on a disease. Test-retest reliability analysis revealed very

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Table 7: Agreement between two versions of the CFQ-R questionnaire (child version and parent proxy) and the comparison between two versions in common domains

Domain	ICC	Correlation between child and parent versions (Spearman test)		Comparison between child and parent versions (Wilcoxon test)	
		R	*P*	(Mean±SD)	*P*
Physical symptoms	0.522	0.393	<0.001*	Child (87.75±13.92)	0.000**
				Parent (74.11±18.21)	
Emotional functioning	0.163	0.083	0.567	Child (84.06±10.03)	0.001*
				Parent (75.00±16.51)	
Body image	0.481	0.311	0.029*	Child (73.33±27.46)	0.011*
				Parent (61.33±25.80)	
Eating disturbances	0.252	0.159	0.271	Child (82.67±13.02)	0.006**
				Parent (72.75±21.38)	
Treatment burden	−0.278	−0.166	0.421	Child (73.33±14.68)	0.708
				Parent (74.17±16.52)	
Respiratory symptoms	0.637	0.443	0.001*	Child (78.38±16.90)	0.000*
				Parent (64.36±16.14)	
Digestive symptoms	0.560	0.307	0.030*	Child (79.00±20.43)	0.796
				Parent (77.50±16.52)	

Spearman's correlation was applied to explore agreement between child and parent versions. Also, ICC values referring intra-class correlation coefficient are used to explore agreement between child and parent version in each domain; Wilcoxon test was run to compare each domain scores between child and parent versions. * denotes significance (<0.05); ** significant which is computed using Wilcoxon test

Table 8: Internal consistency of domains of each CFQ-R version using Cronbach's alpha

Domain	Child version-Cronbach's alpha	Parent version-Cronbach's alpha
Physical symptoms	0.66	0.87
Emotional functioning	0.51	0.61
Social/school functioning	0.21	0.72
Body image	0.83	0.83
Eating disturbances	0.23	0.73
Treatment burden	0.13	0.50
Respiratory symptoms	0.66	0.85
Digestive symptoms	–	0.72
Vitality	–	0.47
Overall health perception	–	0.25
Weight	–	–

good reproducibility for both versions even in domains in which the internal consistency gained low values (only the ICC for treatment burden was equal to 0.78). These findings are in agreement with the previous reports for CFQ-R.[12,13,29,31,32] Of course, different analysis methods were used in the literature to examine the reproducibility, including computing ICCs and Spearmen's correlation coefficients.[12,13,29,31,32] Therefore, comparing the results is not possible directly. In the present study, test-retest was performed in an almost stable health condition (two weeks interval) in children with CF, and further analysis of different health conditions based on the disease severity is recommended.[13]

Agreement between child version and parent proxy

Our results showed poor to moderate agreements between the child version and parent proxy in common domains, with the most in respiration (ICC = 0.637) and the least in emotions (ICC = 0.163). Also, correlation analysis revealed weak to moderate correlations between the two versions in respiration, physical, body image, and digestion. Generally, our results are aligned with German and English reports in most domains (children and parents meaningfully agreed about respiratory, digestion, physical, and body image).[12,17,20] Findings on body image and respiratory domains are in line with the English version.[19] We observed low agreement in eating; however, moderate agreement were reported in English and German which are in contrast to our findings.[12,17,20] Further, parents in our study believe more problems in eating; whereas in the English version, parents and children did not have different beliefs about eating.[17,20] Conflicting results were also obtained for treatment burden and digestion. In English, parents believed more problems in treatment issues compared to children[17,20] and conversely, they estimated digestion higher than children[20] although we did not find any difference in these domains between parents and children. This disparity may be due to differences in international health care systems. Controversy was seen in physical and emotional domains. Our results reveal higher scores for the child version while lower scores were reported in the English version.[17,20] This finding may disclose cultural issues in self-rating systems. Indeed, in the present study, the amount of agreement is highest for most relevant clinical features of the CF disease which

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is in line with the literature using specific health-related QoL measures.[25]

Based on the results, it seems that parents rated less to the more externalized behaviors by children. These findings indicate that parents and children have different concerns about QoL issues related to the CF disease. These differences may be due to emotional and psychological status of parents,[25] different perceptions about QoL (among the child and his/her caregiver),[25] and/or the child's inability to express his/her own feelings and emotions. Therefore, it seems that gathering information from both children and their primary caregivers in subjective instruments is necessary to get a better picture of the child's QoL.[17,33]

Internal consistency

In the present study, higher α values were obtained for the body image in both versions, and for physical and respiratory symptoms in parent proxy (α > 0.8). Also, the α values for emotional, school, eating, and digestive domains in parent proxy were higher than 0.6. The treatment burden obtained α = 0.50 and overall, only two domains (vitality and overall health perception) gained lower α values. The analysis was not performed for weight domain because it was consisting only one item. Cronbach's α in the child version for physical and respiratory domains were >0.6 and for emotional was also equal to 0.51. Because of consisting of a single item, α was not calculated for digestion. Moreover, only two domains in the child version did not gain satisfactory internal consistency (e.g., eating and treatment burden). Therefore, the internal consistency of the Persian CFQ-R is confirmed for the child version and the parent proxy. Our findings are consistent with previous studies.[12,14,20] However, the parent proxy in German obtained internal consistency (α > 0.6) in all domains.[12] Besides, the body image in the child version and school functioning in the parent version in our study gained better reliability compared to others.[12,14,17,20] In English, internal consistency of the child version is lowest for the treatment burden[17,20] which is in agreement with our findings for the treatment burden domain. Therefore, it seems that the treatment burden and its related items should be revised in the original version, and additional revision could also be implemented in other languages. However, adaptation to the CF disease because of its progressive nature and consequent adaptation to the treatment regimen over time may justify the low internal consistency for treatment burden in the child version to some extent.[13] In addition, Modi and Quittner (2003) showed that internal consistency was increased in older ages.[20] Therefore, the effect of age on the reliability indicators should be considered in the future studies on the Persian version. Finally, the limited sample size of the present study compared to others[17,20] and also cultural differences between developed and developing countries may partly explain the results.

Ceiling and floor effects

Floor effect was not seen in the two versions which is in accordance with previous studies.[17,20] This finding reveals that CFQ-R is a valid instrument to apply in CF patients with notably lower health outcomes. The ceiling effect is mostly observed for physical (48%), digestive (38%), and body image (34%) domains in the child version and for eating (24%), weight (including only one item) (24%), and school (20%) in the parent proxy. These findings demonstrate the patients' and their parents' tendency to rate higher scores on some domains. Others also found ceiling effects in three domains, including eating, digestive, and body image for English child version of the CFQ-R.[17,20] However, the limits of ceiling-floor effect were set at 20% in their study.[20] The ceiling effect of parent proxy in the English version was in body image, eating, school, and weight domains[17] which are comparable with our findings. However, in comparison to the English version, the amount of ceiling effects in our study is minimal.[17,20] Indeed, we observed lower ceiling effect for eating in the child version (14%) which is in contrast to the English version.[17,20] The only difference between our study and English child version is the existence of higher ceiling effect for physical domain in our study (48%). The limited range of disease severity in the present study population may partly explain this finding. Overall, our findings are comparable with the literature for adolescents and adults.[29,31]

Limitations

Our study has some limitations. CF database at Children's Medical Center in Tehran is the only available CF database in Iran. Because of the limited referrals and adherence issues, only a limited number of patients were approached in this study (n = 50). Evaluating psychometric properties of questionnaires is highly dependent on the number of participants and some analysis, such as confirmatory factor analysis, is not applicable in studies with low sample size. In some situations, children did not cooperate well with the examiner. Therefore the reliance on children's responses is difficult.

To conclude, the findings of the present study reveal that the Persian CFQ-R child version and parent proxy is a valid and reliable measure to assess the specific health-related QoL in children with CF. The results of our study have implications for conducting comparative studies and clinical trials in Iranian children with CF. It is recommended to survey the psychometric properties in hospitalized children in the future studies. Also, reporting the responsiveness of the questionnaires has useful implications in the clinical setting. Further, the validation of the teen/adult version of the CFQ-R is under consideration in Persian.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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