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ORIGINAL ARTICLE

Content validation for the Chilean population of the quality of life assessment instrument in children, adolescents and adults with Cystic Fibrosis: CFQ-R CYSTIC FIBROSIS QUESTIONNAIRE-REVISED version in Spanish, Chile

Validación de contenido del instrumento de evaluación de calidad de vida en niños, adolescentes y adultos con Fibrosis Quística: CFQ-R CYSTIC FIBROSIS QUESTIONNAIRE-REVISED versión en español, Chile

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What do we know about the subject matter of this study?

This is the first study in the Chilean population that validates a quality-of-life questionnaire for people with CF that will support health teams in providing comprehensive care, beyond the usual treatment and clinical follow-up.

What does this study contribute to what is already known?

Quality of life is a multidimensional construct that has been the subject of multiple research and publications, mainly in developed countries. It is frequently used in the comprehensive approach by a multidisciplinary team for people with CF and their family/primary caregivers.

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Abstract

Cystic Fibrosis (CF) is the most frequent chronic hereditary disease in the white race. Although the impact on the quality of life of this disease is significant, there are no validated instruments in the Chilean population to measure it. **Objective:** To carry out a cultural and linguistic adaptation and validate the content and reliability of the CFQ-R Cystic Fibrosis Questionnaire, Spanish version 2.0. **Patients and Method:** The process was carried out in two stages. The first stage consists of an instrumental design to adapt it culturally and linguistically, evaluate content validity by consulting experts, and test the comprehension of the questionnaire in patients and parents through qualitative interviews and a focus group. In the second stage with an observational and cross-sectional design in a sample of 122 people with CF or their caregivers, the behavior of the questionnaire was analyzed using descriptive statistics and Cronbach's alpha for reliability. **Results:** Stage 1: the instrument in its three versions is considered valid with Lynn's index > 0.8 and Validity Coefficient > 0.7. Stage 2: The adolescent/adult and parent/caregiver versions obtain Cronbach's $\alpha > 0.7$ and an average > 3 in most dimensions. **Conclusion:** The questionnaire is adapted and validated in the Chilean population and requires minor modifications. This version is reliable, valid, and allows the assessment of the quality of life in people with CF. It is suggested to increase the sample for the analysis of construct validity with a larger number of patients.

Keywords:
Health-Related Quality of Life;
Chronic Disease;
Evaluation Study;
Cystic Fibrosis

Introduction

Cystic Fibrosis (CF) is the most common inherited disease in Caucasians, caused by genetic variations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene located on the long arm of chromosome 7. This gene encodes a protein involved in the regulation of the chloride, sodium, and bicarbonate channel. Its defect produces dehydration of mucus in epithelial cells with thickening of secretions in all organs, leading to multisystem involvement. Numerous genetic variables have been described, over 2,000, that are associated with diverse manifestations and severity of the clinical picture[1].

Chronic disease such as CF has a significant impact on personal and family life. In Chile, the national program has 574 patients seen in the public health system, with an average diagnostic age of 3.5 years in children without neonatal screening. Currently, in a pilot project in the Metropolitan Region and Valparaíso Region, screening is performed with an average diagnostic age of 6 months.

In the last decades, survival has been increasing progressively and, therefore, the management of people with CF has become significantly more complicated with important repercussions on the quality of life[2,3]. The median age of survival in developed countries is 50 years[4] and 27 years in Chile[1].

The compromise of quality of life is fundamentally related to respiratory infectious exacerbations and deterioration of pulmonary function, which have an impact on daily life and constitute an important public health problem[5,6]. Whatever the method to assess it, it should include at least five dimensions: physical and mental health, social activities, role in daily activities, and perception of well-being7,8.

Several models have been developed to approach quality of life in childhood and adolescence that consider the social context, environment, and socioeconomic conditions in addition to personal and interpersonal aspects in order to develop in adequate health conditions[9].

The objective of this research is to linguistically adapt, validate the content, and evaluate the behavior and reliability of the Cystic Fibrosis Questionnaire-Revised (CFQ-R), Spanish version 2.0[10] for an adequate clinical use that allows its application in Chile.

Patients and Method

In the first stage, an instrumental design was used to validate the content of the questionnaires. In the second one, a cross-sectional observational study was carried out to evaluate the behavior and reliability of the scales of the questionnaire validated in the first stage. The sampling strategy for both stages was by convenience and non-probabilistic. The data collection period was from October 2017 to August 2018 and all patients who agreed to participate were included since CF is a low-prevalence disease.

The questionnaire for validation contemplates three versions according to the age of the patients as children, adolescents/adults, and parents/caregivers. It consists of the following dimensions measured on a Likert scale: 1. Physical capacity; 2. Vitality; 3. Emotional functioning; 4. Diet; 5. Health; 6. Treatment burden; 7. School; 8. Social functioning; 9. Physical appea-

rance; 10. Role (adequate compliance with regulatory obligations); 11. Respiratory symptoms; 12. Digestive symptoms; and 13. Weight. This questionnaire can be self-administered and takes between 20 to 30 minutes to complete. The original version has a computer software for the analysis of the results[10] that automatically reverts the answers of the inverted items in order to maintain the motto: the higher the score, the better the quality of life.

Stages of the process (Figure 1)

Stage 1

a) Content validation

Lynn's methodology was used which contemplates evaluation by experienced judges on the topic. There is no agreement on the maximum number of experts, but 10 judges are considered a reasonable representation to reach an agreement[11]. In this study, we invited 10 judges contacted with a formal letter, all were health professionals dedicated to the care of patients with Cystic Fibrosis from the hospitals that participated (Metropolitan, Valparaíso, Del Biobío, and La Araucanía Regions).

The judges received the three versions of the questionnaire, the children's version (7 to 12 years and 11 months) with 35 questions, the adolescents/adults version (over 13 years) with 50 questions, and parents/caregivers of children under 6 years and 11 months with 44 questions. Judges were asked to evaluate each item of the questionnaire regarding its adequacy to the construct in dichotomous questions (1 = adequate and 0 = not adequate). An open-ended question was included to incorporate specific suggestions regarding the language and meaning of the items. Each judge had 30 days to complete the report.

As a criterion to keep an item in the final version, it was considered that at least 80% of the judges rated it as adequate; that is, that the item corresponding to the construct measured[11] and that the Content Validity Coefficient (CV) was greater than 0.7[12]. For the qualitative comments, a content analysis was performed incorporating the modifications in which at least 60% of the judges agreed.

b) Linguistic and cultural adaptation

A member of the research team explained to each of the patients or family members the objectives of the research and they were asked, after signing the Informed Consent or Assent form as appropriate, to participate in the interview or focus group during the scheduled clinical check-up. Once the process was completed, these documents were sent to the research team in a sealed envelope, who systematized the information regarding clarity of language and concepts, writing, adequate understanding of the questions, and possible external factors (reading comprehension, comfort, and calmness) that could affect the data collection in its final application.

Stage 2

A cross-sectional observational study was conducted by applying the questionnaire, previously validated, to patients from seven hospitals: *Hospital Clínico San Borja Arriarán*, *Instituto Nacional de Enfermedades Respiratorias y Cirugía Torácica*; *Hospital Clínico Regional Dr. Guillermo Grant Benavente*; *Hospital Dr. Gustavo Fricke*; *Complejo Asistencial Dr. Víctor Ríos Ruiz*; *Hospital San Juan de Dios*, and *Hospital Padre Hurtado*.

Descriptive statistics (frequencies, means, standard deviation) were used in the analysis of the questionnaires. To characterize the quality of life, each dimension was scored, and the result was described based on descriptive statistics and, consequently, percentiles were constructed.

Regarding reliability, Cronbach's alpha statistical method was used[13,14]. In addition, a sociodemographic questionnaire and the CFQ-R adapted and validated in stage 1 were applied, according to the age of the subjects.

To calculate the scores with a positive meaning (higher score, better quality of life), some of the questions indicated in the "Instructions for scores calculation" (Appendix 1) were recoded. Statistical analyses were performed with SPSS version 23.

This study was approved by the Scientific Ethical Committee of the Central Metropolitan Health Service.

Results

Stage 1

The judges evaluated the different versions, which were considered valid, except for three questions that presented a low Lynn index and CV in question 14 of the child version "Did you feel good about yourself?"; question 11 of the adult/adolescent version "Did you feel exhausted?"; and question 11 of the parent/caregiver version "Does any other family member help care for your child?". All three of these questions presented a 0.75 Lynn index and 0.5 CV. The judges considered them to be reiterative with other questions in the questionnaire, although they were in line with the construct measured, so in the final version, the research team decided to keep them, to make the version under study more comparable with the original.

The suggested changes referred to the terminology used, adapting the questionnaire to the language of the target population, for example: "You had bad

dreams or nightmares" for "You had nightmares", and "You coughed up mucus" for "You coughed up mucus (phlegm)". In addition, they suggested modifying the Likert scale labels from the original concept by replacing "very true" with "always"; "mostly true" with "often"; "somewhat true" with "sometimes"; and "never true" with "never".

With the modifications suggested by the judges, a qualitative evaluation was performed on a group of 10 hospitalized patients who analyzed the questionnaire. Then, the same procedure was done with three focus groups with 16 participants (8 parents/caregivers and 8 CF patients in outpatient management), to ensure that the questions were clear and understandable. This number of patients was defined according to sample saturation, which is considered sufficient when no new ideas or comments appear[15].

All changes were incorporated into the final questionnaire. No questions were eliminated and there were no additional questions but indications regarding writing. No changes were made in the process of linguistic adaptation by hospitalized patients, those in outpatient management, and parents/caregivers, thus the modified questionnaire was available.

Stage 2

The sample consisted of 122 persons distributed among 54 adolescent/adult patients, 56 parents/caregivers of children under 6 years of age (children mean age 3.4 years), and 12 children aged from 7 to 12 years and 11 months with CF who were being treated in the participant hospitals. In the group of 54 adolescents/adults, 40 people were older than 18 years (mean age 28.4 years), with high school or higher education in 93%. Regarding parents/caregivers, 100% were women, and 92% with completed secondary or higher education.

Since the study is focused on analyzing the reliability of the questionnaire, at least 50 participants are needed to conduct these analyses[16], therefore, it was decided not to include the questionnaire for children aged 7 to 12 years and 11 months, due to the small sample size (12 people).

For both questionnaires, most dimensions presented an adequate reliability level ($\alpha > 0.7$), except for the dimensions of the adolescent/adult version of treatment burden ($\alpha = 0.631$), digestive symptoms ($\alpha = 0.681$), and social functioning ($\alpha = 0.470$); and those of the parent/caregiver version of health ($\alpha = 0.510$), digestive symptoms ($\alpha = 0.379$), and school, which presented a very low Cronbach's alpha ($\alpha = 0.018$) (Table 1).

The average for most of the dimensions of both versions (Table 2) was higher than 3, which, out of a maximum of 4, ranks the sample at a medium-high level. The exceptions in the questionnaire for adolescents/adults were physical capacity, treatment burden, social functioning, respiratory symptoms, and weight; and in the questionnaire for parents/caregivers were treatment burden, school, and weight. The dispersion for the various dimensions is similar between them except for weight, which is higher.

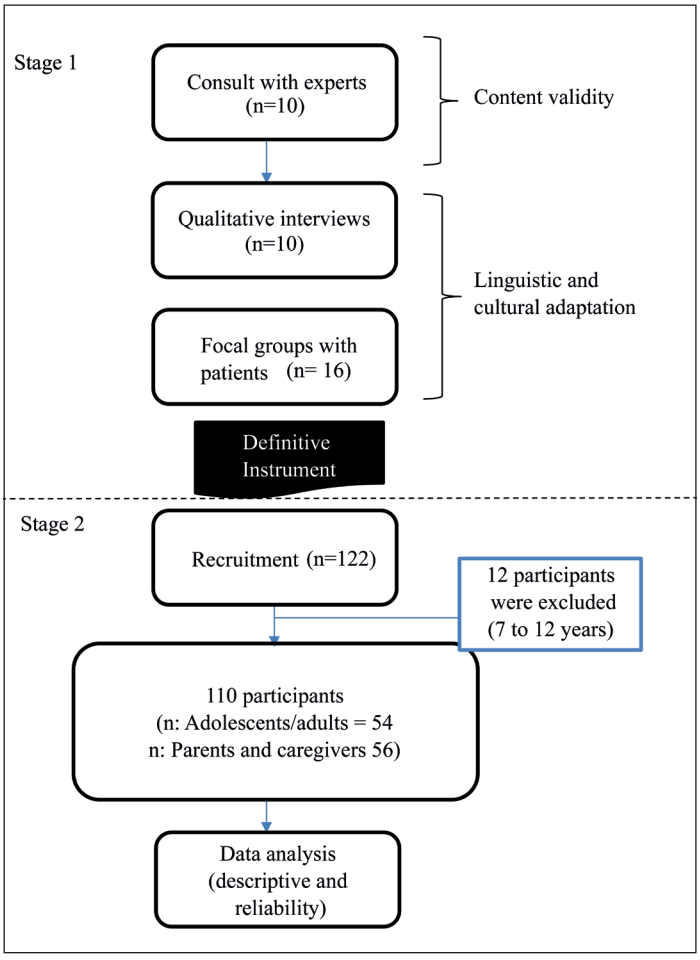

Figure 1. Flowchart.

Tables 3, 4, and 5 show the calculation and interpretation of percentiles for the different dimensions.

Regarding the ceiling and floor effects (Table 6), to evaluate the discrimination of the test, ≤ 15% was used as a quality criterion[17]. It was observed that in both versions, the weight dimension is the one that presents the highest indicators of ceiling and floor effect because it is composed of only one item. However, when analyzing the ceiling effect in the adolescent/adult version, an effect > 15% was found in the domains of food (53.7%), health (22.2%), physical appearance (25.9%), role (27.8%), and digestive symptoms (24.1%); and in the parent/caregiver version in the dimensions of food (40.3%), physical appearance (45.8%), role (27.8%), and digestive symptoms (20.9%).

Table 1. Cronbach's α statistic for the dimensions of the adolescent and adult version questionnaires, parents and caregivers

	Adolescent / Adult		Parents / Caregivers	
	Nº ítems	α	Nº ítems	α
Physical capacity	8	0.9	9	0.897
Vitality	4	0.752	5	0.704
Emotional functioning	5	0.758	5	0.615
Diet	3	0.864	2	0.874
Health	3	0.812	3	0.51
Treatment burden	3	0.631	3	0.7
School	-	-	3	0.018
Social functioning	6	0.47	-	-
Physical appearance	3	0.767	3	0.733
Role	4	0.846	-	-
Respiratory symptoms	7	0.823	7	0.838
Digestive symptoms	3	0.681	3	0.379
Weight	1	-	1	-

α = alfa de Cronbach.

Discussion

In the last 20 years, the measurement of quality of life has received progressively more attention[7,18]. This study is the first in Chile to validate the content and make a linguistic adaptation of the CFQ-R questionnaire in Spanish for this population.

The purpose of instrument validation[18] is to evaluate the evidence regarding the psychometric quality of the instruments to ensure that their application contributes adequately to the development of the discipline to which it belongs. It also allows to compare care centers and measure the effectiveness of interventions.

Using instruments that do not have a cultural or linguistic validation or with an inadequate process results in inaccurate measurements, especially if these instruments are used to make decisions in the clinical practice[19,20].

The CFQ-R questionnaire applied in this study has a good standard in the context of origin, is widely used, and is translated into different languages[21-23]. It has three versions which make its validation difficult, however, maintaining them has clinical importance because they consider the stage of the life cycle of the person with CF.

In general, the validation process is sequential, using various methodological and statistical techniques. It begins with an adaptation, translation, and counter-translation[24,20], although this was not the case in this study since the Spanish version was used[21]. Secondly, content validity is carried out by experts[8] and then tested on a sample of the target population, as we did in this study.

The evaluation of construct validity in this research was not performed since a factor analysis is required with a minimum of 200 subjects for each questionnaire[25]. Therefore, it is necessary to incorporate a larger sample in the future in order to make this determination[12.]

To measure reliability, internal consistency is evaluated by Cronbach's alpha[13], which is a reliability indicator in the dimensions. This study shows similar results in several of them compared with other publications, with Cronbach's alpha > 0.7[21,24,26]. The social dimension in this study (Cronbach = 0.47) needs further qualitative research, including other questions more relevant to Chilean idiosyncrasy. It is important to use with caution the dimensions that present low internal consistency.

To improve the adolescent/adult questionnaire, in the digestive symptom dimension, question 48 could be eliminated as well as question 8 in the emotional dimension, thus Cronbach's α would increase to 0.731 and 0.515, respectively. The team decided to wait for a larger sample to see the behavior of these questions. It was not necessary to eliminate questions for parents/caregivers. For the weight dimension, both questionnaires have only one question, therefore, it is not possible to assess internal consistency (Table 1).

In the evaluation of reproducibility, the test-retest

Table 2. Means for adolescent and adult quality of life dimensions, parents and caregivers

	Adolescent / Adult					Parents / Caregivers				
	Nº	Min	Max	P	SD	Nº	Min	Max	P	SD
Physical capacity	54	1.25	4	2.98	0.74	38	1	4	3.06	0.82
Vitality	54	1.75	4	3.05	0.55	38	1.8	4	3.16	0.52
Emotional functioning	54	2.2	4.6	3.4	0.55	38	2.4	4	3.47	0.49
Diet	54	1.33	4	3.52	0.68	56	1	4	3.17	0.88
Health	54	1.67	4	3.04	0.68	56	1.67	4	3.08	0.55
Treatment burden	54	1	3.7	2.32	0.62	56	1	4	2.79	0.79
School	-	-	-	-	-	37	1	4	2.9	0.66
Social functioning	54	1.5	4	2.94	0.52	-	-	-	-	-
Physical appearance	54	1	4	3.19	0.84	37	1	4	3.32	0.85
Role	54	1.5	5	3.38	0.69	-	-	-	-	-
Respiratory symptoms	54	1.29	4	2.95	0.68	56	2.14	4	3.27	0.59
Digestive symptoms	54	1.67	4.3	3.37	0.56	56	2	4	3.4	0.44
Weight	54	1	4	2.67	1.24	56	1	4	2.96	1.15

Nº= number of adolescents and adults, parents and caregivers; Min= minimum; Max= maximum; P= average; SD= standard deviation.

Table 3. Percentiles for adolescent/adult quality of life dimensions

	Percentiles						
Dimensions	5	10	25	50	75	90	95
Physical capacity	1.62	1.94	2.38	3.06	3.63	4	4
Vitality	2	2.38	2.75	3	3.5	3.88	4
Emotional functioning	2.2	2.6	3	3.5	3.8	4	4
Diet	2.17	2.5	3	4	4	4	4
Treatment burden	1.92	2.33	2.58	3	3.67	4	4
School	1.25	1.33	2	2.33	2.75	3	3.33
Social functioning	2.13	2.33	2.5	3	3.21	3.73	3.83
Physical appearance	1.33	1.67	2.67	3.5	4	4	4
Role	1.94	2.5	3	3.5	4	4	4
Respiratory symptoms	1.89	2	2.43	2.93	3.45	3.93	4
Digestive symptoms	2.25	2.67	3	3.33	3.75	4	4
Weight	1	1	1	3	4	4	4

Table 4. Percentiles for Parent/Caregiver Quality of Life Dimensions

	Percentiles						
Dimensions	5	10	25	50	75	90	95
Physical capacity	1.49	1.82	2.61	3.33	3.78	3.89	4
Vitality	2.16	2.36	2.8	3.2	3.6	3.84	4
Emotional functioning	2.4	2.6	3	3.6	3.8	4	4
Diet	1	1.4	2.75	3.5	4	4	4
Treatment burden	2.27	2.33	2.67	3	3.67	3.67	4
School	1.3	1.6	2.33	2.67	3.67	3.73	4
Social functioning	1.6	2	2.58	3	3.42	3.67	4
Physical appearance	1.6	1.93	2.67	3.67	4	4	4
Respiratory symptoms	2.14	2.26	2.71	3.14	3.71	4	4
Digestive symptoms	2.67	2.97	3	3.33	3.67	4	4
Weight	1	1	1	1	3	4	4

Table 5. Interpretation of percentiles

Danger zone	percentil < 25
Poor quality of life	percentil ≥ 25 y < 50
Good life quality	percentil ≥ 50 y < 75
Very good quality of life	percentil ≥ 75

Table 6. Ceiling and floor effect CF

Dimensions	Floor effect	Ceiling effect
Adolescents / Adults		
Physical capacity	0%	11.10%
Vitality	0%	9.30%
Emotional functioning	0%	14.90%
Diet	0%	53.70%
Health	0%	22.20%
Treatment burden	3.70%	0%
School	0%	1.90%
Social functioning	1.90%	25.90%
Physical appearance	0%	27.80%
Respiratory symptoms	0%	9.30%
Digestive symptoms	0%	24.10%
Weight	25.90%	38.90%
Parents / Caregivers		
Physical capacity	2%	10.20%
Vitality	0 %	8.20%
Emotional functioning	0%	14.30%
Diet	4.50%	40.30%
Health	0%	13.40%
Treatment burden	3.00%	10.40%
School	2.10%	8.30%
Social functioning	2.10%	45.80%
Physical appearance	0%	27.80%
Respiratory symptoms	0%	11.90%
Digestive symptoms	0%	20.90%
Weight	50.70%	13.40%

technique and correlations (intraclass, Pearson's, or Spearman's) were used. The literature shows a low response rate within a short period after the first measurement[21]. The applied instrument asks for the last two weeks, so it is expected that reproducibility can be observed in that period. This study did not evaluate this characteristic since there were some dimensions in which a longer period of stability could be expected, such as social function and school, while in others the response may change depending on clinical variables, such as treatment burden and respiratory symptoms, which question the usefulness of the retest.

The self-application of the questionnaire generates difficulties in the group of parents/caregivers who have a lower response rate, so this procedure should be accompanied by a professional who can help in the understanding of the questions.

The elaboration of percentiles in this study, not done in other publications, facilitates the interpretation and gives a clinical sense to the measurement. An instruction for interpretation was developed to enable the professional to label the subjet's quality of life at a given moment and to guide timely and specific interventions (Appendix 1).

The use of a validated scale allows for monitoring the difficulties presented by patients and their families and improves the relationship with the health team during clinical management, especially in communication[27]. It is essential to have good records to evaluate the evolution of the quality of life in the disease process and to program interventions that could improve or mitigate the effects derived from it[28]. The Chilean Consensus for the comprehensive care of children and adults with CF[1] does not include the quality-of-life survey and the importance of this variable for the clinical management of patients[29].

In published versions, there is little discriminative power in the dimensions of physical appearance, nutrition, and weight[21], as occurred in this research expressed in the ceiling-floor results. In addition, the dimension role, digestive symptoms, and health were added in this study.

The sampling strategy used was non-probabilistic, so it is not possible to rule out selection biases when analyzing the questionnaire; however, the use of convenience sampling is the standard in the studies reviewed[6,21,22,24,26].

The literature review did not provide information regarding the frequency of application, so the research team, according to experience, proposed a measurement twice a year as appropriate, which will be variable, depending on the condition of the person with CF.

It is advisable to incorporate assessment in people with CF pain, sleep quality, anxiety, and depression symptoms, all dimensions present in other studies[30-34].

The major limitation of this study is the small sample in the population studied because CF in Chile is a disease with an incidence of approximately 1 per 8,000 to 10,000 live newborns[22].

The sample size of the group aged 7 to 12 years and 11 months was not enough to perform the quantitative analysis; therefore, it is essential to increase the number of people to draw conclusions.

Regarding the reliability of the questionnaires, in general, the dimensions are reliable; however, there

are some with low internal consistency indexes, which means that they do not behave in a coordinated manner among themselves, raising the question of whether they are measuring the same dimension. In this sense, future analysis of the construct validity of the instrument is necessary to carry out, for which the current sample was insufficient.

As this is a multicenter study, it incorporates various teams and hospitals, with differences among them (geographical areas, number and type of professionals, distance between the research team and the clinical centers), making it difficult to ensure that the data collection mechanisms are similar, which constitutes a possible information bias.

It is important to use with caution the dimensions that present low internal consistency such as treatment burden, digestive symptoms, and social functioning in the adolescent/adult version, and health, digestive symptoms, and school dimensions in the version for parents/caregivers.

Conclusions

The validation of the document is the first published in Chile and allows the evaluation of quality of life, enabling the development of interventions. The experts consulted consider that the questionnaires are adequate to the quality-of-life construct and suggest minor changes in the writing of the questions and can be used in the Chilean population.

It is recommended that they be applied twice a year with the modified version depending on clinical criteria.

It is necessary to continue incorporating a larger sample, especially in children between 7 and 13 years of age.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

Apéndice 1: Cuestionarios e interpretación

Cuestionario para evaluación de calidad de vida en Fibrosis Quística: versión adolescentes y adultos (basado en el instrumento CFQ-R CYSTIC FIBROSIS QUESTIONNAIRE-REVISED con validación cultural y de contenido para población chilena)

Instrumento: CFQ-R CYSTIC FIBROSIS QUESTIONNAIRE-REVISED versión adolescentes y adultos.

Durante las dos últimas semanas, cuanta dificultad ha tenido:

Ítem	Mucha dificultad	Mediana dificultad	Poca de dificultad	Ninguna dificultad
1. Participando en actividades agotadoras como correr o practicar algún deporte.	1	2	3	4
2. Caminando tan rápido como los demás.	1	2	3	4
3. Cargando o levantando cosas pesadas como libros o mochilas.	1	2	3	4
4. Subiendo escaleras.	1	2	3	4
5. Subiendo escaleras tan rápido como los demás.	1	2	3	4

Y en las dos últimas semanas, indique cuán frecuentemente:

Ítem	Siempre	Casi siempre	A veces	Nunca
6. Se sintió bien.	4	3	2	1
7. Se sintió preocupado(a).	1	2	3	4
8. Se sintió inútil en actividades diarias.	1	2	3	4
9. Se sintió cansado(a).	1	2	3	4
10. Se sintió con mucha energía.	4	3	2	1
11. Se sintió agotado(a).	1	2	3	4
12. Se sintió triste.	1	2	3	4

Ítems de respuesta múltiple 13 al 18, marque con un círculo, UNA de las alternativas para las siguientes preguntas:
Pensando en su estado de salud en las últimas dos semanas:

13. ¿Hasta que punto tiene dificultad al caminar?
    4. Pudo caminar por mucho tiempo sin cansarse.
    3 Pudo caminar por mucho tiempo pero se cansa.
    2. No pudo caminar por mucho tiempo porque se cansa rápidamente.
    1. Evita caminar cuando le es posible porque se cansa mucho.

14. ¿Cómo se siente con respecto al comer?
    1. Sólo pensar en comida le causa malestar.
    2. No disfruta al comer.
    3. Algunas veces disfruta al comer.
    4. Siempre disfruta al comer.

15. ¿Hasta qué punto los tratamientos le hacen su vida diaria más difícil?
    4. Nada en lo absoluto.
    3. Un poco.
    2. Moderadamente.
    1. Mucho.

16. ¿Cuánto tiempo dedica cada día a sus tratamientos?
    1. Mucho tiempo.
    2. Algo.
    3. Poco.
    4. Casi nada.

17. ¿Cuán desgastante es para usted hacer los tratamientos (incluyendo medicamentos) cada día?
    4. Nada en lo absoluto.
    3. Un poco.
    2. Moderadamente.
    1. Mucho.

18. ¿Cómo piensa que está su salud hoy?
    4. Excelente.
    3. Buena.
    2. Más o menos.
    1. Mala.

Alternativas de respuesta a las preguntas desde la 19 a 34.
Pensando en su salud durante las dos últimas semanas, responda:

Ítem	Siempre	Casi siempre	A veces	Nunca
19. Tengo dificultad recuperándome después del esfuerzo físico.	1	2	3	4
20. Tengo que limitar mis actividades físicas como correr o practicar deportes.	1	2	3	4
21. Tengo que obligarme a comer.	1	2	3	4
22. Tengo que quedarme en casa más de lo que quisiera.	1	2	3	4
23. Me siento cómodo hablando de mi enfermedad con otros.	4	3	2	1
24. Pienso que estoy muy delgado(a).	1	2	3	4
25. Pienso que me veo diferente en comparación con otros(as) de mi edad.	1	2	3	4
26. Me siento mal con respecto a mi apariencia física.	1	2	3	4
27. La gente teme a contagiarse conmigo.	1	2	3	4
28. Me reúno con mis amigos a menudo.	4	3	2	1
29. Pienso que mi tos molesta a los demás.	1	2	3	4
30. Me siento con ánimo para salir en la noche ("carretear").	4	3	2	1
31. Me siento solo (a).	1	2	3	4
32. Me siento saludable.	4	3	2	1
33. Es difícil hacer planes para el futuro (por ejemplo, ir a la universidad, matrimonio, etc.).	1	2	3	4
34. Llevo una vida normal.	4	3	2	1

Ítems de respuestas de la 35 a 38, marque con un circulo, UNA de las alternativas para las siguientes preguntas:

35. Durante las dos últimas semanas, ¿hasta qué punto tuvo dificultad manteniéndose al día en su trabajo escolar, profesional o en otras actividades diarias?
    4. No he tenido dificultad para mantenerme al día.
    3. He podido mantenerme al día, aunque se me ha hecho difícil.
    2. Me he atrasado.
    1. No he podido hacer estas actividades en lo absoluto.

36. Durante las últimas dos semanas, ¿con qué frecuencia estuvo ausente de la escuela, trabajo, o no pudo completar sus actividades diarias debido a su enfermedad o sus tratamientos?
    1. Siempre.
    2. Casi siempre.
    3. Algunas veces.
    4. Nunca.

37. ¿Con qué frecuencia le impide a usted la Fibrosis Quística alcanzar sus metas de escuela, trabajo o metas personales?
    1. Siempre.
    2. Casi siempre.
    3. Algunas veces.
    4. Nunca.

38. ¿Con qué frecuencia le impide la Fibrosis Quística salir de su casa para hacer actividades como ir de compras o ir al banco?
    1. Siempre.
    2. Casi siempre.
    3. Algunas veces.
    4. Nunca.

Alternativas de respuestas a las preguntas desde la 39 a 41.
Durante las dos últimas semanas:

Ítem	Bastante	Moderado	Poco	Nada
39. Ha tenido dificultad para aumentar de peso.	1	2	3	4
40. Ha tenido congestión nasal.	1	2	3	4
41. Ha estado tosiendo durante el día.	1	2	3	4
42. Ha tenido tos con secreciones ("flemas").	1	2	3	4

Ítem de respuesta múltiple.
Por favor seleccione y encierre en un circulo la alternativa correspondiente a su respuesta:

43. Su secreción ("flema") ha sido mayormente:
    4. Transparente.
    3. Amarilla-verde.
    2. Verde con sangre.
    1. No me he fijado.

Alternativas de respuesta a las preguntas desde la 44 a 50.
Indique cuán frecuentemente en las dos últimas semanas:

Ítem	**Siempre**	**Casi siempre**	**A veces**	**Nunca**
44. Ha estado con silbido al respirar.	1	2	3	4
45. Ha tenido dificultad al respirar.	1	2	3	4
46. Se ha despertado durante la noche porque estaba tosiendo.	1	2	3	4
47. Se ha sentido "hinchado" con gases.	1	2	3	4
48. Ha tenido diarrea.	1	2	3	4
49. Ha tenido dolor abdominal.	1	2	3	4
50. Tuvo dificultad con su apetito.	1	2	3	4

Desea Ud. realizar otras observaciones importantes que influyen en su calidad de vida relacionado con la Fibrosis Quística

_____

_____

Instructivo para el cálculo de puntajes (aplicar en paciente clínicamente estable).
Versión para adolescentes y adultos.

Esta versión consta de 50 preguntas, todas en la escala de Likert con puntajes de 1 a 4. Para el análisis del cuestionario, sume y calcule un promedio de las respuestas por dimensión.

a) Con los promedios utilice la tabla 3 de percentiles.
b) Para evaluar al paciente según percentil, utilice tabla 5.

Dimensión	Preguntas	Promedio	Percentil	Evaluación
Capacidad física	1,2,3,4,5,13,19, 20.			
Vitalidad	6,9,10,11.			
Emoción	7,8,12,31,33.			
Alimentación	14, 21,50.			
Peso del tratamiento	15,16,17.			
Salud	18,32,34.			
Social	22,23,27,28,29,30.			
Apariencia física	24,25,26.			
Rol	35,36,37,38.			
Peso	39.			
Síntomas respiratorios	40,41, 42. 43,44,45,46.			
Síntomas digestivos	47,48,49.			

Cuestionario para evaluación de calidad de vida en Fibrosis Quística: versión padres y cuidadores.
(basado en el instrumento CFQ-R CYSTIC FIBROSIS QUESTIONNAIRE-REVISED
con validación cultural y de contenido para población chilena)

Si su niño tiene menos de tres años, pase a la pregunta 30.
Alternativas de respuesta desde la pregunta 1 a 5.
¿Cuánta dificultad ha tenido su niño(a)?

Ítem	Mucha dificultad	Mediana dificultad	Poca de dificultad	Ninguna dificultad
1. Participando en actividades agotadoras como correr o practicar algún deporte.	1	2	3	4
2. Para caminar tan rápido como los otros niños.	1	2	3	4
3. Subiendo escaleras tan rápido como los demás.	1	2	3	4
4. Cargando o levantando cosas pesadas como libros o una mochila.	1	2	3	4
5. Subiendo muchos escalones.	1	2	3	4

Alternativas de respuesta desde las preguntas 6 a 13.
Durante las dos últimas semanas, indique cuán a menudo su niño(a):

Ítem	Siempre	Casi siempre	A veces	Nunca
6. Pareció estar contento(a).	4	3	2	1
7. Pareció estar preocupado(a) (no aplica a menor de 5 años).	1	2	3	4
8. Pareció estar cansado(a).	1	2	3	4
9. Pareció estar de mal humor.	1	2	3	4
10. Pareció estar bien.	4	3	2	1
11. Pareció estar enojado.	1	2	3	4
12. Pareció tener mucha energía.	4	3	2	1
13. Estuvo ausente o llegó tarde a la escuela, jardín infantil u otra actividad debido a su enfermedad o tratamientos.	1	2	3	4

Ítems de respuesta de las preguntas 14 y 15.
Por favor seleccione y encierre en un circulo la alternativa correspondiente a su respuesta.
Con respecto al estado de salud de su niño(a) durante las dos semanas pasadas, indique:

14. ¿Cuánto participó su niño(a) en deportes en la escuela, incluyendo durante el recreo y la clase de educación física? 1. No ha participado en deportes en la escuela. 2. Ha participado menos de lo usual en deportes en la escuela. 3. Ha participado igual que siempre, pero con alguna dificultad. 4. Ha podido participar en deportes sin ninguna dificultad.
15. ¿Cuán difícil le resulta caminar a su niño(a)? 4. Puede caminar por mucho tiempo sin cansarse. 3. Puede caminar por mucho tiempo pero se cansa. 2. No puede caminar mucho porque se cansa rápidamente. 1. Evita caminar siempre que puede, porque se cansa mucho.

Alternativas de respuesta de las preguntas 16 a 30.
Con respecto al estado de salud de su niño(a) durante las dos últimas semanas, responda:

Ítem	Siempre	Casi siempre	A veces	Nunca
16. Mi niño(a) tiene dificultad para recuperarse después de un esfuerzo físico.	1	2	3	4
17. Da problemas para comer.	1	2	3	4
18. Los tratamientos de mi niño(a) interfieren con sus actividades.	1	2	3	4
19. Mi niño(a) se siente pequeño(a) de estatura comparado con otros niños(as) de su misma edad.	1	2	3	4
20. Mi niño(a) se siente físicamente diferente a otros niños(as) de su misma edad.	1	2	3	4
21. Mi niño(a) piensa que es muy flaco(a).	1	2	3	4
22. Mi niño(a) se siente saludable.	4	3	2	1
23. Mi niño(a) tiende a ser retraído(a).	1	2	3	4
24. Mi niño(a) lleva una vida normal.	4	3	2	1
25. Mi niño(a) se divierte menos de lo usual.	1	2	3	4
26. Mi niño(a) tiene dificultad para llevarse bien con los demás.	1	2	3	4
27. Mi niño(a) tiene dificultad para concentrarse.	1	2	3	4
28. Mi niño(a) es capaz de mantenerse al día con sus tareas de la escuela o actividades de verano.	4	3	2	1
29. A mi niño(a) no le va tan bien como antes en la escuela o en sus actividades del verano (ejemplo: en el campamento de verano).	1	2	3	4
30. Mi niño gasta mucho tiempo en sus tratamientos todos los días.	1	2	3	4

Ítems de respuesta de las preguntas 31 y 32.
Por favor seleccione y encierre en un circulo la alternativa correspondiente a su respuesta:

31. ¿Cuán difícil es para su niño(a) hacer sus tratamientos (incluyendo los medicamentos) cada día? 4. Nada en lo absoluto. 3. Un poco. 2. Moderadamente. 1. Mucho.
32. ¿Cómo cree que está la salud de su niño(a) en este momento? 4. Excelente. 3. Buena. 2. Regular. 1. Mala.

Alternativas de respuesta a las preguntas 33 a 36.
Por favor indique como su niño(a) ha estado en las dos últimas semanas:

Ítem	Bastante	Moderado	Poco	Nada
33. Mi niño(a) ha tenido dificultad para aumentar de peso.	1	2	3	4
34. Mi niño(a) tuvo congestión nasal.	1	2	3	4
35. Mi niño(a) tosió durante el día.	1	2	3	4
36. Mi niño(a) tosió secreciones ("flemas").	1	2	3	4

Ítem de respuesta 37.
Por favor seleccione y encierre en un círculo la alternativa correspondiente a su respuesta:

Ítem
La mucosidad de mi niño(a) ha sido mayormente: 4. Transparente. 3. Amarilla – verde. 2. Verde con muestras de sangre. 1. No me he fijado.

Alternativas de respuesta a las preguntas 38 a 44.
Durante las dos últimas semanas:

Ítem	Siempre	Casi siempre	A veces	Nunca
38. Mi niño(a) estuvo con silbido al pecho al respirar.	1	2	3	4
39. Mi niño(a) tuvo dificultad para respirar.	1	2	3	4
40. Mi niño(a) se despertó durante la noche porque estaba tosiendo.	1	2	3	4
41. Mi niño(a) tuvo gases.	1	2	3	4
42. Mi niño(a) tuvo diarrea.	1	2	3	4
43. Mi niño(a) tuvo dolor abdominal ("dolor de guatita").	1	2	3	4
44. Mi niño(a) tuvo problemas para comer.	1	2	3	4

Comentarios

Instructivo para el cálculo de puntajes (aplicar en paciente clínicamente estable)
Versión para padres/cuidadores
Esta versión consta de 44 preguntas, todas en la escala de Likert con puntajes de 1 a 4.
Para el análisis del cuestionario sume y calcule un promedio de las respuestas por dimensión.

a) Con los promedios utilice la tabla 4 de percentiles.
b) Para evaluar al paciente según percentil utilice tabla 5.

Dimensión	Preguntas	Promedio	Percentil	Evaluación
Capacidad física	1,2,3,4,5,13,14,15,16.			
Vitalidad	8,9,10,11,12.			
Emoción	6,7,23,25,26.			
Alimentación	17,44.			
Peso del tratamiento	18,30,31.			
Salud	22,24,32.			
Escuela	27,28,29.			
Apariencia física	19,20,21.			
Peso	33.			
Síntomas respiratorios	34,35,36,37,38,39,40.			
Síntomas digestivos	41,42,43.			

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