

# COMMON CONTENT BETWEEN QUALITY OF LIFE QUESTIONNAIRES FOR CHILDREN WITH CYSTIC FIBROSIS AND THE INTERNATIONAL CLASSIFICATION OF FUNCTIONING, DISABILITY AND HEALTH

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**Objective:** To identify the most common quality of life instruments for children with cystic fibrosis and link the content with the International Classification of Functioning, Disability and Health (ICF).

**Methods:** The study was conducted in 2 stages. The first stage involved a review of the literature to select quality of life questionnaires. In the second stage 2 independent reviewers identified questionnaire items and categories corresponding to the ICF, according to approved methodology. The degree of agreement was calculated using the kappa coefficient.

**Results:** Two questionnaires were selected: the Cystic Fibrosis Questionnaire and DISABKIDS®. A total of 130 concepts were identified from the 112 items. Forty-seven different ICF categories were linked ( $k > 0.62$  for all questionnaires), 21 (44.7%) were related to the “body function” domain, 20 (42.6%) to “activity and participation” and 6 (12.8%) to “environmental factors”. Thirteen items (10%) could not be linked because they represent personal factors or are not covered by the ICF.

**Conclusion:** Body functions were the category most linked to the ICF. Environmental factors were poorly described, and no items were related to body structures in any of the instruments.

**Key words:** cystic fibrosis; child; quality of life; International Classification of Functioning, Disability and Health.

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Cystic fibrosis (CF), a chronic progressive condition that involves several systems, leads to numerous clinical manifestations, the most common being pancreatic insufficiency and pulmonary alterations with chronic airway infection. These alterations impose a heavy treatment burden on the quality of life and psychological well-being of individuals with CF (1–3).

In recent years, several instruments have been developed to evaluate health-related quality of life (HRQoL) (3–6). The HRQoL indicator provides information on the impact of a disease and its treatment in different

## LAY ABSTRACT

Cystic fibrosis imposes a heavy treatment burden on the quality of life and psychological well-being of children. Since much of the daily management of cystic fibrosis occurs at home, observing the child’s ability to participate in school-based and leisure activities is of paramount importance, as participation in these activities is considered essential for child development, in addition to contributing to health-related quality of life. This study identified the quality of life questionnaires for cystic fibrosis and linked their items to the International Classification of Functioning, Disability and Health (ICF). In the 2 questionnaires presented here, the categories primarily represented body functions and, although environmental factors play an important role in children’s participation in home, school and community activities, and can act as a support or barrier, they were the categories least covered by the 2 questionnaires and no items were related to body structures in any of the instruments.

life situations, which are measured by specific or generic questionnaires aimed at assessing the individual in terms of physical, emotional and social impacts. However, these instruments may focus differently on the components of functionality and health, and analysing their scope could result in better clinical practice applications (7, 8).

In recent years, numerous studies on the breadth of HRQoL instruments have proposed using the International Classification of Functioning Disability and Health (ICF) to provide relevant information on how they can be used in the assessment, planning and care of children with CF (7–10). This is because the ICF is used to monitor the functionality of individuals and populations on a broader scale in order to consider the health and biopsychosocial aspects involved in the health-disease process. It includes a number of categories describing aspects of human functionality that interfere with daily activities, as well as environmental factors that may be facilitators or barriers (9).

The common content between ICF and generic instruments that evaluate HRQoL in children has already been identified (7, 8), but the points of convergence and/or divergence between questionnaires in order to assess

the biopsychosocial impacts of CF and ICF have yet to be established. As such, the objectives of this study were to identify the main HRQoL questionnaires specifically designed for children with CF and to examine how their content is represented in the ICF categories.

## METHODS

The study was conducted in 2 stages. The first stage involved a review of the literature to identify and select specific quality of life (QoL) questionnaires for children with CF, and the second consisted of linking the items of the questionnaires to the ICF according to the methodology proposed by Cieza et al. (11).

The first step was performed on the PubMed, SciELO and LILACS electronic databases in June 2017 using the descriptors Quality of Life Questionnaires AND Cystic Fibrosis AND Child. The search found 176 studies in English, Portuguese and Spanish between 1980 and 2017, sorted by title and abstract. In the studies that evaluated the QoL of children with CF, we sought to identify the most commonly used instruments in the literature. Two questionnaires were selected: the Cystic Fibrosis Questionnaire (CFQ) and DISABKIDS® – Cystic Fibrosis Module (DISABKIDS® – CFM). The former, which has been translated and validated in different languages (4, 12–14), is widely used in the literature and evaluates individuals from childhood to adulthood through 4 versions: CFQ<sub>6-11</sub>, CFQ<sub>12-13</sub>, CFQ<sub>14+</sub> and CFQ<sub>parents6-13</sub>. Based on the criteria of the present study, the CFQ<sub>6-11</sub> and CFQ<sub>parents6-13</sub> versions, which contain 35 and 44 questions, respectively, were included in addition to the demographic sections.

The DISABKIDS® – CFM is 1 of 7 modules developed by the DISABKIDS group for specific health conditions. This short 10-item questionnaire is divided into 2 scales (impact and treatment). The former (4 items) describes the feelings of tiredness and exhaustion, and the latter (6 items) the emotional impact of the treatment. There is a version for children and adolescents from 8 to 18 years (DISABKIDS<sub>self</sub>) and another, with parallel items, for parents or caregivers (DISABKIDS<sub>proxy</sub>).

In the second stage of the study, the methodology used to link the QoL instruments to the ICF (9) was based on the rules proposed by Cieza et al., in 2002 (15). This methodology has been updated twice (11, 16) and is widely disseminated in the

literature. The link between the items of the QoL instruments and the ICF was determined by 2 independent physical therapists with theoretical knowledge and previous practical ICF experience in health services. In case of disagreement, a third senior physical therapist with broad ICF experience was consulted in order to reach a consensus.

Absolute and relative frequencies were used in descriptive statistical analysis. The degree of agreement between the 2 professionals who established the link between the items and the ICF was calculated using the kappa coefficient, and the classification suggested by Landis & Koch (17), in which values less than zero are poor, between 0 and 0.2 slight, between 0.21 and 0.4 fair, between 0.41 and 0.6 moderate, between 0.61 and 0.8 substantial and between 0.81 and 1.0 almost perfect. In all analyses, a 95% confidence interval (95% CI) and the Statistical Package for the Social Sciences (SPSS), version 20.0 were used.

## RESULTS

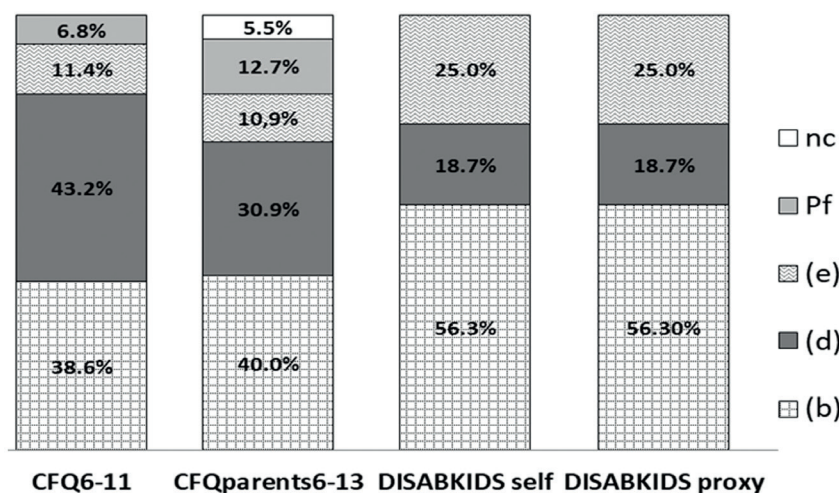
A total of 130 concepts were identified from the 112 items and 47 different ICF categories were linked (QFC<sub>6-11</sub> k=0.646; CFQ<sub>parents6-13</sub> k=0.670; DISABKIDS<sub>self</sub> and DISABKIDS<sub>proxy</sub> k=0.622).

Twenty-one (44.7%) concepts were related to the body function domain, 20 (42.6%) to the activity and participation domain, and 6 (12.8%) to environmental factors. Thirteen items (10%) could not be linked to the ICF because they represent personal factors or are not covered by the ICF. All of these unrelated items were contained in the CFQ – Revised (CFQ<sub>parents6-13</sub> and CFQ<sub>6-11</sub>) (Fig. 1).

The absolute and relative frequencies of the categories identified in the questionnaires are presented in Table I, and divided by ICF domains, chapters, and categories.

Half (15) of CFQ<sub>parents6-13</sub> categories were related to body function (b), 40% (12) to activities and participation (d) and 10% (3) to environmental factors (e). Four of the 8 body function domain chapters were linked to these items, the most cited was (b1) – mental functions, and the most detailed categories were the 4-level categories b1522 – range of emotion and b1801 – body image.

However, most of the linked categories in CFQ<sub>6-11</sub> were related to the activities and participation domain (14=51.3%), body function domain (9=33.3%) and environmental factors (4=14.3%). Six of the 9 activities and participation domain



**Fig. 1.** Distribution of the content of the questionnaires in relation to the domains of the ICF. (b) body function; (d) Activity and participation; (e) environmental factors; Pf: personal factors; nc: not covered; CFQ: Cystic Fibrosis Questionnaire

**Table I.** Absolute and relative frequency of the categories identified in the questionnaires by component and chapter of the International Classification of Functioning, Disability and Health

| Chapters by domains                                                                         | CFQ 6–11 |      |                                                                                                                                                                                            | CFQ parents 6–13 |      |                                                                                                                                                                                                                                | DISABKIDSself |      |                                                                                                                                            | DISABKIDSproxy |      |                                                                                                                                            |
|---------------------------------------------------------------------------------------------|----------|------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|------------------|------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------|------|--------------------------------------------------------------------------------------------------------------------------------------------|----------------|------|--------------------------------------------------------------------------------------------------------------------------------------------|
|                                                                                             | n        | %    | Linked categories                                                                                                                                                                          | n                | %    | Linked categories                                                                                                                                                                                                              | n             | %    | Linked categories                                                                                                                          | n              | %    | Linked categories                                                                                                                          |
| <b>b: Body function</b>                                                                     |          |      |                                                                                                                                                                                            |                  |      |                                                                                                                                                                                                                                |               |      |                                                                                                                                            |                |      |                                                                                                                                            |
| (b1) Mental functions                                                                       | 5        | 26.8 | b1265 – Optimism<br>b1341 – Onset of sleep<br>b1344 – Functions involving the sleep cycle<br>b1522 – Range of emotion (5)*<br>b1801 – Body image (3)*                                      | 7                | 24.4 | b126 – Temperament and personality functions<br>b1300 – Energy level<br>b134 – Sleep functions<br>b152 – Emotional functions<br>b1520 – Appropriateness of emotion<br>b1522 – Range of emotion (3)*<br>b1801 – Body image (3)* | 2             | 37.5 | b1300 –<br>b152 – Emotional functions (5)*                                                                                                 | 2              | 37.5 | b1300 – Power level<br>b152 – Emotional functions (5)*                                                                                     |
| (b2) Sensory functions and Pain                                                             | 1        | 2.4  | b28012 – Pain in stomach or abdomen                                                                                                                                                        | 1                | 2.2  | b28012 – Pain in stomach or abdomen                                                                                                                                                                                            | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (b4) Functions of the cardiovascular, haematological, immunological and respiratory system. | 3        | 12.2 | b440 – Respiration functions<br>b450 – Additional functions of the respiratory system(3)*<br>b4552 – Fatiguability                                                                         | 4                | 15.6 | b440 – Respiration functions (2)*<br>b450 – Additional functions of the respiratory system<br>b4552 – Fatiguability (2)*<br>b498 – (2)*                                                                                        | 3             | 18.8 | b455 – Exercise tolerance functions<br>b4552 – Fatiguability<br>b460 – Sensations associated with cardiovascular and respiratory functions | 3              | 18.8 | b455 – Exercise tolerance functions<br>b4552 – Fatiguability<br>b460 – Sensations associated with cardiovascular and respiratory functions |
| (b5) Functions of the digestive, metabolic and endocrine systems                            | --       | --   | --                                                                                                                                                                                         | 3                | 6.7  | b525 – Defecation functions<br>b5254 – Flatulence<br>b530 – Weight maintenance functions                                                                                                                                       | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| <b>d: Activity and Participation</b>                                                        |          |      |                                                                                                                                                                                            |                  |      |                                                                                                                                                                                                                                |               |      |                                                                                                                                            |                |      |                                                                                                                                            |
| (d1) Learning and applying knowledge                                                        | --       | --   | --                                                                                                                                                                                         | 1                | 2.2  | d160 – Focusing attention                                                                                                                                                                                                      | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (d2) General Tasks and Demands                                                              | --       | --   | --                                                                                                                                                                                         | 1                | 2.2  | d2101 –                                                                                                                                                                                                                        | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (d4) Mobility                                                                               | 6        | 17.1 | d4303 – Carrying on shoulders, hip and back<br>d450 – Walking<br>d4551 – Climbing<br>d4552 – Running (2)*<br>d4553 – Jumping<br>d4602 – Moving around outside the home and other buildings | 4                | 13.3 | d4303 – Carrying on shoulders, hip and back<br>d450 – Walking (2)*<br>d4551 – Climbing<br>d4552 – Running                                                                                                                      | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (d5) Self -care                                                                             | 2        | 9.8  | d550 – Eating (3)*<br>d5700 – Ensuring one's own physical comfort                                                                                                                          | 1                | 2.2  | d550 – Eating                                                                                                                                                                                                                  | 1             | 6.3  | d5701 – Managing diet and fitness                                                                                                          | 1              | 6.3  | d5701 – Managing diet and fitness                                                                                                          |
| (d7)                                                                                        | 1        | 2.4  | d7101 – Appreciation in relationships                                                                                                                                                      | 1                | 2.2  | d760 – Family Relationships                                                                                                                                                                                                    | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (d8) Major life areas                                                                       | 1        | 2.4  | d820 –                                                                                                                                                                                     | 2                | 8.9  | d820 – (3)*<br>d845 –                                                                                                                                                                                                          | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (d9) Community, social and civic life                                                       | 4        | 14.6 | d920 – Recreation and Leisure (3)*<br>d9201 – Sports<br>d9202 – Arts and Culture<br>d9205 – Socializing                                                                                    | 2                | 6.7  | d920 – Recreation and Leisure (2)*<br>d9201 – Sports                                                                                                                                                                           | 1             | 12.5 | d920 – Recreation and Leisure (2)*                                                                                                         | 1              | 12.5 | d920 – Recreation and Leisure (2)*                                                                                                         |
| <b>e: Environmental factors</b>                                                             |          |      |                                                                                                                                                                                            |                  |      |                                                                                                                                                                                                                                |               |      |                                                                                                                                            |                |      |                                                                                                                                            |
| (e1) Products and technology                                                                | --       | --   | --                                                                                                                                                                                         | --               | --   | --                                                                                                                                                                                                                             | 1             | 6.3  | e1101 – Drugs                                                                                                                              | 1              | 6.3  | e1101 – Drugs                                                                                                                              |
| (e3) Support and relationships                                                              | --       | --   | --                                                                                                                                                                                         | 1                | 2.2  | e325 – Acquaintances, peers, colleagues, neighbours and community members                                                                                                                                                      | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (e4) Attitudes                                                                              | 2        | 4.9  | e425 – Individual attitudes of acquaintances, peers, colleagues, neighbours and community members<br>e460 – Societal attitudes                                                             | --               | --   | --                                                                                                                                                                                                                             | --            | --   | --                                                                                                                                         | --             | --   | --                                                                                                                                         |
| (e5) Services, systems and policies                                                         | 2        | 7.3  | e5800 – Health services (2)*<br>e5850 – Education and training services                                                                                                                    | 2                | 11.1 | e5800 – Health services (4)*<br>e5850 – Education and training services                                                                                                                                                        | 1             | 18.8 | e5800 – Health services (3)*                                                                                                               | 1              | 18.8 | e5800 – Health services (3)*                                                                                                               |

( ) \* Frequency; FQ: Cystic Fibrosis Questionnaire.

chapters were represented in the linked categories, chapter d4 (mobility) and categories d4551 (climbing) and d920 (recreation and leisure) being the most cited.

Only 9 of the 47 categories identified were linked to the items of DISABKIDS® (DISABKIDS<sub>self</sub> and DISABKIDS<sub>proxy</sub> version), most pertaining to the body function domain (5 = 55.6%) in both versions, representing 2 of the 8 chapters that make up this domain, namely (b1) – mental functions, with category b152 (emotional functions) being the most cited; and (b4) – functions of the cardiovascular, haematological, immunological and respiratory systems. Category frequency was 22.2% (2) in the activities and participation and environmental factors domains in both versions of the questionnaires.

## DISCUSSION

Comparing HRQoL measures with the ICF may reveal its uniqueness, demonstrating the nature and scope of its content. In most cases, instruments focus on different aspects of ICF domains, with “body functions” and “activities and participation” components usually predominating (7, 8). In the 4 versions presented here (CFQ<sub>parents6–13</sub>, CFQ<sub>6–11</sub>, DISABKIDS<sub>self</sub> and DISABKIDS<sub>proxy</sub>), the categories primarily represented body functions, except for the CFQ<sub>6–11</sub> version, where the activities and participation component was the most frequent. By contrast, environmental factors were poorly described, and none of the categories were related to body structures.

Although chapters b1 to b5 have been linked to the items identified, the only chapters that contained categories cited in all 4 versions were Mental functions (b1) and Functions of the cardiovascular, haematological, immunological and respiratory systems (b4). Category b152 (emotional functions) has often been linked in the different instruments (20 times), because a number of feelings/sensations, including “happy”, “worried”, “angry”, “annoyed”, “sad”, “upset”, and “bothered”, were represented in this category. The high attachment index of this category to different feelings has been addressed in other studies (8, 18).

Because of the clinical manifestations present in the scope of body functions resulting from the multisystemic nature of CF, the questionnaires can be used to address a large number of aspects related to this domain (7, 8, 19). Studies that seek to identify clinical factors associated with HRQoL in individuals with CF have concluded that variables assessing exacerbation and pulmonary function have the greatest impact on HRQoL (4, 20, 21). Multidirectional relationships between ICF domains demonstrate that body functions are only 1 aspect for practitioners to consider. Other domains should also be incorporated (22).

The complex treatment these children are subjected to, including ingested and inhaled medication, nebulization, antibiotics, pancreatic enzymes, nutritional supplements, respiratory physical therapy techniques, and hospitalization, is time-consuming and a burdensome daily routine (3, 13). These factors, together with body function limitations, may interfere with the performance of daily activities and community participation.

The CFQ<sub>6–11</sub> was the only instrument where most items were related to the activities and participation domain, although the categories linked CFQ<sub>parents6–13</sub> to chapters d1 (learning and applying knowledge) and d2 (general tasks and demands), a finding not observed in the children’s version. The most representative chapters were d4 (mobility) and d9 (community, social and civic life), the latter addressed in all the questionnaires, and d920 (recreation and leisure) the most frequent category. Since much of the daily management of CF occurs at home, observing the child’s ability to perform age-appropriate activities,

and participate in typical school-based and leisure activities is of paramount importance, since participation in the activities of daily living is considered essential to child development, in addition to contributing to HRQoL, and is the ultimate goal of rehabilitation programmes (22–24).

Environmental factors play an important role in children’s participation in home, school and community activities, and can act as a support or barrier (25). Despite their relevance, environmental factors were the categories least covered by the 4 versions. In this component, the most cited category was e5800 (health services).

Other categories addressed in this component were education and training services (e5850); acquaintances, peers, colleagues, neighbours and community members (e325); societal attitudes (e460); individual attitudes of acquaintances, peers, colleagues, neighbours and community members (e425); and drugs (e1101).

According to the model presented by the ICF, the functionality of an individual with a specific health condition depends on body-related aspects, as well as personal and environmental factors (9). The weakness of the instruments in failing to sufficiently address these factors limits an extended evaluation, as proposed by the ICF, and makes it difficult to identify possible facilitators or barriers that the environment imposes on children with CF and their impact on QoL, in addition to showing the need to refine paediatric HRQoL instruments. This recurrent weakness has been demonstrated in other studies (7, 8).

In the context faced by children with CF, categories such as: e310 (immediate family); e355 (health professionals); e410 (individual attitudes of immediate family members); and e450 (individual attitudes of health professionals) could have been addressed. The environment, access to healthcare and drugs, and support from the spouse, family, friends, teachers, classmates and colleagues are important aspects that influence the HRQoL reported by individuals with CF (26).

All items that could not be linked to the ICF belonged to the CFQ – Revised versions, most of which referred to personal factors contained in the demographic sections that make up these instruments. Age, sex, body mass index (BMI), and other factors seem to have an impact on HRQoL (27–29). Identifying these factors in the linking process may reveal a number of relevant personal factors to add to the ICF if classification of personal factors is desired in the future (18, 30). The items classified as not covered (nc) were contained only in the CFQ<sub>parents6–13</sub> version, exhibiting comprehensive content, including “my child leads a normal life” (item 24), and “my child has a feeding problem” (item 44), or very specific items such as “my child’s phlegm has been predominant” (item 37).

This study sought to link the items of 2 specific instruments that evaluate the HRQoL of children with CF. The included questionnaires display good psychometric properties. DISABKIDS® contains few items, thereby optimizing application and assessment time; however, more than half of its items are linked to body function categories. The versions of the CFQ were linked to a larger number of categories that addressed the 4 ICF domains. The CFQ<sub>6–11</sub> was the only version in which the activity and participation domain exhibited the highest frequency, but did not address some of the chapters related to the functions of the digestive, metabolic and endocrine systems (b5), learning and applying knowledge (d1), and general tasks and demands (d2) that were covered in the parents’ version. Thus, we suggest the simultaneous use of the CFQ<sub>6–11</sub> and CFQ<sub>parents6–13</sub> questionnaires.

In conclusion, the main HRQoL questionnaires specifically designed for children with CF were CFQ and DISABKIDS®. The link between the items of the instruments and the ICF domains made it possible to acquire information on the scope of the human

functionality components in each questionnaire. More than half of the content of the DISABKIDS® (DISABKIDS<sub>self</sub> and DISABKIDS<sub>proxy</sub>) is linked to the categories related to body function. The CFQ versions (CFQ<sub>parents6-13</sub>; CFQ<sub>6-11</sub>) were linked to a larger number of categories that reached the 4 domains that make up the ICF, namely body function (b), body structures (s), activities and participation (d), and environmental factors (e), allowing a broader assessment of health status. However, both questionnaires (CFQ and DISABKIDS®) exhibit disadvantages in relation to the biopsychosocial evaluation proposed by ICF, since they barely address factors of the physical, social and attitudinal environment in the HRQoL assessment process of children with CF.

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