QUALITY OF LIFE OF CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS**QUALIDADE DE VIDA DE CRIANÇAS E ADOLESCENTES COM FIBROSE CÍSTICA****CALIDAD DE VIDA DE NIÑOS Y ADOLESCENTES CON FIBROSIS QUÍSTICA**

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ABSTRACT

Objective: to evaluate the health-related quality of life (HRQoL) of children and adolescents with cystic fibrosis. **Method:** this is a quantitative and cross-sectional study with children and adolescents. Data was collected in the outpatient clinic weekly with the DISABKIDS® instrument from the perspective of individuals (Self) and their parents or caregivers (Proxy). Statistical analysis was performed using descriptive techniques in SPSS 20 software, and frequency tables were constructed. **Results:** the sample was constituted by 11 subjects. It was recorded that the majority (63.6%) of the companions were mothers with a mean age of 41 years. In the Impact dimension, Self showed an average of 66.4 and, in Proxy, 69.5; however, in the treatment dimension, by the Self version, we obtained an average of 72.4 and, by Proxy, 66.9. The majority of patients with good clinical status (45.4%) were classified according to the Shwachman-Kulczyki score. **Conclusion:** it is concluded that children and adolescents perceive a better HRQoL on treatment and a worse HRQoL on the impact of the disease, when compared to their parents/caregivers. **Descriptors:** Cystic Fibrosis; Chronic disease; Quality of life; Health Care; Child; Teenager.

RESUMO

Objetivo: avaliar a qualidade de vida relacionada à saúde (QVRS) de crianças e adolescentes com fibrose cística. **Método:** trata-se de um estudo quantitativo e transversal, realizado com crianças e adolescentes. Coletaram-se os dados no ambulatório semanalmente com o instrumento DISABKIDS® na perspectiva dos indivíduos (Self) e dos seus pais ou cuidadores (Proxy). Realizou-se a análise estatística por técnicas descritivas no software SPSS 20, sendo construídas tabelas de frequências. **Resultados:** constituiu-se a amostra por 11 sujeitos. Registrou-se que a maioria (63,6%) dos acompanhantes era de mães com idade média de 41 anos. Apontaram-se, na dimensão impacto, pela Self, uma média de 66,4 e, na Proxy, de 69,5; entretanto, na dimensão tratamento, pela versão Self, obteve-se uma média de 72,4 e, pela Proxy, de 66,9. Classificou-se a maioria dos pacientes com boa condição clínica (45,4%) de acordo com o escore de Shwachman-Kulczyki. **Conclusão:** conclui-se que as crianças e adolescentes percebem uma melhor QVRS quanto ao tratamento e uma pior QVRS quanto ao impacto da doença, quando comparados aos seus pais/cuidadores. **Descritores:** Fibrose Cística; Doença Crônica; Qualidade de Vida; Atenção à Saúde; Criança; Adolescente.

RESUMEN

Objetivo: evaluar la calidad de vida relacionada con la salud (QVRS) de niños y adolescentes con fibrosis quística. **Método:** se trata de un estudio cuantitativo y transversal, realizado con niños y adolescentes. Se recogieron los datos en el ambulatorio semanalmente con el instrumento DISABKIDS® en la perspectiva de los individuos (Self) y de sus padres o cuidadores (Proxy). Se realizó el análisis estadístico por técnicas descriptivas en el software SPSS 20, siendo construidas tablas de frecuencias. **Resultados:** se constituyó la muestra por 11 sujetos. Se registró que la mayoría (63,6%) de los acompañantes era de madres con edad promedio de 41 años. Se apunta, en la dimensión impacto, por el Self, una media de 66,4 y, en la Proxy, de 69,5; sin embargo, en la dimensión tratamiento, por la versión Self, se obtuvo un promedio de 72,4 y, por Proxy, de 66,9. Se clasificó la mayoría de los pacientes con buena condición clínica (45,4%) de acuerdo con la puntuación de Shwachman-Kulczyki. **Conclusión:** se concluye que los niños y adolescentes perciben una mejor QVRS en cuanto al tratamiento y una peor QVRS en cuanto al impacto de la enfermedad, cuando comparados a sus padres / cuidadores. **Descriptores:** Fibrosis Quística; Enfermedad Crónica; Calidad de Vida; Atención a la Salud; Niño; Adolescente.

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INTRODUCTION

Cystic fibrosis (CF) is considered to be a rare, hereditary, genetic, inherited chronic condition of autosomal recessive transmission, ie inherited from both parents, with a risk of occurrence of one in four if the father and mother are carriers of the gene, being more frequent in Caucasians.¹⁻³ It is known that in Brazil, the incidence of the disease varies according to the region, between 1:6,000, 1:8,000 and 1:10,000.⁴

It is pointed out that the disease generates a significant economic cost related to health and a negative impact on health-related quality of life (HRQL) for both patients and caregivers when compared to the general population.⁵ The median number of deaths is recorded for the pathology between 23 and 29 years, with respiratory failure being the main cause of death.⁶⁻⁷

CF is characterized by multisystemic dysfunction of the exocrine glands, especially affecting the lungs and the pancreas, in an obstructive process caused by increased mucus viscosity. It favors, by this process, the blockade of the airways, propitiating the bacterial proliferation, which leads to the chronic infection; however, in the pancreas, secretion causes blockage of the ducts, leading to pancreatic fibrosis and, consequently, loss of digestive enzymes, resulting in poor nutrition.^{1,8}

It is understood that the pathophysiology of the disease is the result of the absence or dysfunction of a protein called cystic fibrosis transmembrane conductance regulator (CFTR), encoded by the gene located on the long arm of chromosome 7, locus q 31, exon 10. This protein acts as a chlorine channel, regulating the balance between ions and water through the epithelium. It is emphasized that the CFTR protein dysfunction decreases the permeability of the cell membrane to the chloride, causing difficulties for the transport and secretion of this ion.²

In this way, each organ that depends on the CFTR protein - lungs, pancreas, intestines, sweat glands and vas deferens - expresses this dysfunction in a different way, according to the sensitivity of each one to the functional deficit.²

It is observed that the affected individuals may present different clinical manifestations, isolated or combined, in the affected organ, such as chronic cough, recurrent pneumonia, steatorrhea, low weight, pancreatitis, meconium ileus and salty sweat. The disease manifests itself, however, in several other

ways, since it is a disease that affects several systems and organs, and may even remain innocuous for years.^{3,9} The course of the disease is alternated between periods of remission and exacerbation, with increasing severity over the years and with complications such as malnutrition, diabetes, hepatic failure and osteoporosis.¹⁰

It is noted that during childhood and adolescence the individual experiences periods of physical, psychosocial and emotional transformation, constituting moments of self-concept, body image, self-esteem and affective bonds in society. Stigmatizing characteristics are identified and the repercussions of this stigma can have implications in the treatment, in the process of socialization, in the interpersonal relations and in the formation of the individual's identity, directly reflecting their quality of life.^{3,11}

Adolescent carriers of this disease are known to have difficulties in relation to sexuality, independence and adherence to treatment, since they feel different from the general population. It is also evaluated that the dissimilarity among the patients alludes to the heterogeneity of the clinical signs, since its expression corresponds to the type of genetic alteration, which can affect one or more organs, besides demanding the involvement of the parents/caregivers. Due to the seriousness of the therapeutic regimen, it causes a feeling of dependence.¹²

It is traditionally based on the diagnosis of CF in the detection of elevated levels of sodium chloride in the sweat test (> 60 mEq / L for more than three months and > 40 mEq / L for children under three months), in the absence of enzymes pancreatic and radiographic findings, in chronic obstructive pulmonary disease (COPD) and through family history. The diagnosis can now be confirmed by the identification of the deoxyribonucleic acid (DNA) of the mutant genes and by the abnormal measurement of the nasal potential difference.¹

It is recommended, however, that neonatal screening should be carried out in the neonatal period for early detection and timely treatment, which consists of the analysis of immunoreactive trypsinogen performed in a dry blood spot followed by a confirmatory examination of the dosage of chloride in the sweat of all cases suspected of cystic fibrosis, and direct DNA analysis may be required for the presence of the mutation.^{1,8}

After the diagnosis, treatment and follow-up are initiated, which, due to the various clinical manifestations, become complex,

requiring the implementation by specialized services for CF. The patient is assured by the multidisciplinary approach, a total treatment and new therapies are explored that can reduce the rate of progression of the disease and treat its complications in a timely manner, substantially increasing the survival and quality of life of these patients.¹³⁻⁴ The study emphasizes the importance of the nurse as an active member of this multidisciplinary team, ensuring patient follow-up and looking at the basic needs of these subjects.³

The treatment and management of the care started up to two months after the detection of CF in neonates, neonatal screening, less complications related to the disease and better long-term health outcomes.¹⁵⁻⁶

A better life perspective is recorded through correct treatment, surveillance of pulmonary infection and agility in the prescription of effective and specific antibiotics for pathogenic bacteria, as well as the introduction of pancreatic enzymes, offered by the Unified Health System (UHS), allowing the absorption of nutrients in the intestinal tube.⁹

Quality of life (QoL) is defined according to the World Health Organization (WHO) Study Group on Quality of Life as "the individual's perception of their position in life in the context of culture and value system in which he lives and in relation to his goals, expectations, standards and concerns".¹⁷ It reinforces, by the elements about the subject, the organization of the amplified concept of the absolute satisfaction of the individual.

A European project designed to improve the quality of life and independence of children with chronic conditions and their families was created by DISABKIDS, a set of questionnaires for assessing the quality of life from the perspective of the child with regard to their physical, mental and social well-being.¹⁸ HRQoL is understood as multidimensional and as a concept that addresses the physical, emotional, mental, social and behavioral aspects of the subjective well-being and the functional state perceived by the patient and the family member. The two definitions are approximated because they consider the perception of those involved, however, the latter is customized by the subjectivity of the subject and the people close to them.¹⁸

It has been identified in studies about HRQoL that school attendance and improved nutritional status have a positive impact on children with CF;¹⁹⁻²⁰ in contrast, the long

duration of the disease, high levels of dependence, hospitalization, *Pseudomonas aeruginosa* infection and malnutrition are related to a significant and negative impact on HRQoL.^{5,21}

It is argued that the evaluation of HRQoL should be estimated in the course of assistance to the fibrocystic patient, because it allows a better understanding of the multidimensional aspects related to the individual and contribute to the elaboration of clinical protocols and therapeutic guidelines.²²

OBJECTIVE

- To evaluate the quality of life related to health (HRQoL) of children and adolescents with cystic fibrosis.

METHOD

This is a cross-sectional quantitative study carried out at the Júlio Muller University Hospital (JMUH) - Federal University of Mato Grosso (FUMT), a reference in the scope of CF in the State of Mato Grosso, which arose from the implantation and planning for the qualification of Phase III, which includes, within the modalities of the Neonatal Screening Program, cystic fibrosis. At the beginning of the study, 16 patients between the ages of eight and 18 years old were followed, one of whom died, two were absent, and one patient did not follow the service.

All children and adolescents, aged between eight and 18 years, with CF, in the outpatient clinic, who accepted to participate in the study, with the authorization of parents or caregivers, were considered for the study. Participants signed a Term of Assent, concomitant with the Free and Informed Consent Term, signed by their parents or guardians and by the reference health professional, adding up to 12 individuals. One of them was excluded because it did not have an age-compatible cognitive condition, verified through the instrument of the DISABKIDS® group, called "clinical variables for all conditions", applied to the health professional, making a total of 11 subjects.

The data was collected in the outpatient clinic weekly, on Mondays, from September 11 to December 18, 2017, according to the follow-up schedule of the patients, and the approaches were performed in the order in which the subjects arrived for the your queries. It should be noted that all children and adolescents responded to the instruments separately and alone, and that the

instruments used for data collection have a high internal consistency index.²²

Specific instruments, developed and validated by the DISABKIDS® group, were used to measure HRQoL in children and adolescents aged 8 to 18 years with CF from the perspective of individuals (self version) and their parents or caregivers (proxy version) . It is pointed out that the items are the same, differing only in relation to the essay, with a focus on children and adolescents. Ten items are presented in the instrument, exposing the two dimensions to be evaluated, called impact and treatment: a) Impact (four items) - describes the feeling of tiredness and exhaustion; b) Treatment (six items) - describes the emotional impact caused by the treatment.²²

It is observed that the instrument uses the Likert scale to collect the answers, with five graduation points: never, almost never, sometimes, often and always (the score of each item varies from one to five). We recoded the items for the calculation of the final scores: 1 = 5, 2 = 4, 3 = 3, 4 = 2 and 5 = 1; therefore, the higher the scores, the lower the negative impact of the chronic condition on children's HRQoL or adolescents.²²

There are three more items on the symptoms of the disease that, although not computed in the scores of the instrument, are related to the severity of the condition perceived by children and adolescents.²²

Another instrument from the DISABKIDS® group, called "clinical variables for all conditions", was applied to the health professional responsible for the child and adolescent with CF, presenting questions about their underlying disease, cognitive aspects and the value of the score of Shwachman-Kulcziki to know the severity of the disease.²³

The HRQoL is highlighted as the variable of interest, and the other variables are sex (female and male), age, date of birth, number of siblings, time elapsed since the patient attends school, series school level, school level (Elementary, Middle, Higher and other), the severity of CF during the last year (none, few, moderate, very and extreme), the amount of bad moments caused by CF symptoms during the last year (never, once, twice, three times and more than three times), the last time you had hemoptysis (never, last year, the last six months, last month and last week), the person responsible for questionnaire "clinical variables for all conditions" (child pediatrician, hospital specialist, community specialist, physician or school nurse, other clinician and hospital

documentation), the child's basic diagnosis, ICD 10, cognitive skills cognitive impairment, developmental delay, learning disability, mental retardation and unknown), Shwachman-Kulczyki score and severity of baseline disease (excellent, good, medium, moderate and severe).

The database was structured through the spreadsheet editor of Microsoft Office Excel, with double independent typing. Data from both banks were compared using the Data Compare tool to identify and correct possible inconsistencies by querying the original collection documents. Statistical analysis was performed using descriptive techniques in SPSS 20 software, and frequency tables were constructed.

This study was authorized by the Head of the Unit of Attention to the Health of Children and Adolescents of the Department of Pediatrics and by the Management of Teaching and Research in the Sector of Management of Research and Technological Innovation - GEP / SGPIT / HUJM - UFMT / EBSERH, under the Opinion No 032017.

The project was submitted and approved, after authorization from the respective sectors, by the Research Ethics Committee involving human beings, under protocol No. 2,191,112 and CAAE 71483517.3.0000.5541, in July 2017, following all the prerogatives of Resolution 466/2012.

RESULTS

It was recorded that of the 11 subjects who participated in the research, six (54.5%) were male. The other sociodemographic characteristics of children, adolescents and caregivers are shown in Table 1.

Table 2 shows the standardized mean values, standard deviation, medians, maximum and minimum for the scores of the instrument that evaluates the Health-Related Quality of Life of children / adolescents with cystic fibrosis, according to patients and parents or caregivers. The scores of the impact dimension and the treatment dimension were calculated separately, and all items of both dimensions were answered. The scores were standardized for the presentation of the results, on a scale from zero to 100.

As regards the child's perception of the severity of their cystic fibrosis during the last year, it is pointed out that four (36.4%) reported no severity, while three (27.2%) assessed little severity, two (18, 2%), very serious and two others (18.2%), moderate severity; however, five (45.4%) had low severity, three (27.3%), moderate severity,

two (18.2%), no severity and one (9.1%), very serious.

Table 1. Sociodemographic characteristics of children, adolescents and caregivers. Cuiabá (MT), Brazil, 2018

Children and adolescents		
Age (years) (Average/SD)		12/3.6
Number of brothers/sisters (Average/SD)		1/1.3
Years of schooling (Average/SD)		6/3.5
Shcooling level	Elementary (n/%)	9/81.8
	Highschool (n/%)	2/18.2
Parents or caregivers		
Age (years) (Average/SD)		41/5.7
Respondents	Mothers (n/%)	7/63.6
	Fathers (n/%)	3/27.3
	Grandmother (n/%)	1/9.1
Marital status	Married (n/%)	8/72.7
	Divorced (n/%)	1/9.1
	Single (n/%)	1/9.1
	Widow (n/%)	1/9.1

Table 2. Standardized mean values, standard deviation, medians, maximum and minimum for the scores of the instrument according to the children or adolescents (Self) and parents or caregivers (Proxy). Cuiabá (MT), Brazil, 2018.

Dimension	Average (Self/Proxy)	SD (Self/Proxy)	Median (Self/Proxy)	Maximum (Self/Proxy)	Minimum (Self/Proxy)
Impact (0-100)	66.4/69.5	18.4/16.0	70.0/65.0	90.0/90.0	30.0/45.0
Treatment (0-100)	72.4/66.9	22.8/21.8	73.3/80.0	100.0/100.0	40.0/33.3

In relation to the occurrence of bad moments because of CF symptoms during the last year, three children / adolescents (27.3%) answered that they never happened or only once, two (18.2%), twice (18.2%), more than three times and one (9.0%), three times. In the perception of parents / caregivers, three (27.3%) had one episode, three (27.3%), two episodes, two (18.2%), three episodes, two (18.2%), never one (9.0%), more than three times.

It was observed that, when asked about the last time they had hemoptysis, eight children / adolescents (72.7%) said they never had one, two (18.2%) said they had hemoptysis last year and one (9.1%) in the last six months; however, nine (81.8%) parents / caregivers

indicated that patients never had hemoptysis, one (9.1%) last year and one (9.1%) in the last six months.

Table 3 shows the distribution of children and adolescents according to the severity of cystic fibrosis, in which five (45.4%) classified it as good, according to the Shwachman-Kulczyki score.

Table 3. Distribution of children and adolescents according to the severity of cystic fibrosis classified according to the Shwachman-Kulczyki score. Cuiabá (MT), Brazil, 2018.

Gravity	n	%
Severe	1	9.1
Moderate	4	36.4
Average	0	0.0
Good	5	45.4
Excellent	1	9.1
Total	11	100.0

DISCUSSION

The HRQoL measure is defined as essential to know the perspective of the patient and the parents/caregivers regarding the subjective aspects of health, especially when it comes to a patient with chronic conditions.²² This study evaluates the impact of the disease and the treatment on the HRQoL of the children and adolescents investigated, contributing to guide the future clinical interventions. Through the combination of evaluations, a broader view of the HRQoL is possible, that is, in a comparative analysis of the two self/proxy instruments, and a more reliable perception of the subject's actual health situation is portrayed.

It should be pointed out that there is no consensus in the literature regarding the distribution of CF between the sexes in children and adolescents,^{22,24-5} but in relation to the adult child/adolescent reference, the mother appears in evidence in the proxy version, as well as in other studies.^{20,22}

It is verified that mothers present themselves as the main caregivers of the children and feel satisfaction in doing so, understanding the task as a delivery to the learning process that they understand to be motherhood, in the sense of being competent mothers, who love, care and are always present.²⁶

Regarding the HRQoL assessment, in relation to the two dimensions - impact and treatment - weighted from the perspective of children and adolescents (self) and parents / caretakers (proxy), few differences were found between the mean impact of the disease on the HRQoL; however, when the averages of the treatment dimension are analyzed, it is verified that children and adolescents tend to qualify their HRQoL better when compared to the perception of their parents / caregivers, which coincides with other findings.²⁷⁻⁸ It is suggested that this may occur due to the better coping of the disease by the children/adolescents.²⁹

It is observed, through researchers, through the discourses of the mothers of patients with CF, a difficulty to live with the daily treatment of the disease. It is a tiring routine of schedules, medications, respiratory physiotherapy and returns to the consultations; In addition, hospitalizations are frequent, causing them to move away from their everyday activities.¹¹

It is pointed out that, when comparing the results presented with those of another study,

carried out in three Brazilian states, with 113 patients, the mean impact size in the self version was 72.71, and there was a lower negative impact of the condition in the HRQoL than in this study. It should be noted, however, that in the treatment dimension the opposite was recorded, since an average of 67.70 was obtained, lower than the one found in this study, that is, the higher the score, the lower the negative impact of the condition in the child / adolescent's HRQoL.²²

Differences are observed between the severity perceived by the patients and the parents / caregivers. It is noted that the patient tends to disdain the severity of his disease while it is mild and, with the advancement of CF, subjective perception ceases to discriminate the progressive impairment of health, especially when the disease progresses from the moderate stage to the serious stage. It was identified by the authors that the perception of the severity of the disease by the CF patient correlates with the objective measures of the severity of the disease, such as the clinical score of SK.¹⁰

It should be noted that, in the case of the SK score, applied in a study carried out in Hungary, the highest proportion of individuals whose clinical condition was classified as good (55.9%) was also identified, however, only 3.4% were severe.²¹ The SK score was developed with the purpose of evaluating the severity of the disease through the clinical and radiological study, divided into four categories: general activity; physical exam; nutrition and radiological findings, each with five possible scores. The severity of CF is indicated as the single most important factor, since it affects the survival of these patients.³⁰

A moderate relationship between the HRQoL and the disease-specific SK score is understood, according to the perspective of patients and parents/caregivers;²¹ therefore the SK score can provide information about the clinical severity of the CF and can also act, as an indicator of HRQoL.

Studies indicate that school attendance and nutritional status have a positive impact on the HRQoL of children and adolescents with CF;¹⁹⁻²⁰ however, the long duration of the disease, the advancement of age, high levels of dependence, hospitalization, *Pseudomonas aeruginosa* infection and malnutrition are related to a significant and negative impact on HRQoL. It is known that, in patients with CF, the HRQoL declines slowly over time - approximately 1% per year.^{5,21,25}

Early diagnosis is considered during neonatal screening as the best tool to ensure

timely treatment and a better prognosis for the patient¹⁹. It is pointed out that health professionals should be able to identify the needs of these patients and their families, in order to achieve a better understanding and adaptation of the transition process between health and disease.³

It is suggested that the multidisciplinary approach, evidenced at the study site, can be an important benefit already established in patients' HRQoL, especially when providing a humanized care, reducing the negative impact that HR causes on the patient's life and family.

This study identifies some limitations, such as insufficient sample to observe the relationship between the studied variables, therefore, no conclusion about the associations could be drawn. Further research is needed to advance more and more towards a better understanding of the HRQoL of children and adolescents with CF.

CONCLUSION

It is concluded that children and adolescents perceive a better HRQoL in treatment and a worse HRQoL in relation to the impact of the disease, when compared to the perception of their parents/caregivers. The dimensions of the HRQoL that are most affected in this context are favored by the planning of Nursing care and, consequently, the improvement in the quality of care. It is also pointed out that the use of specific questionnaires in routine clinical practice could expand the usefulness of HRQoL measures, in which aspects that have been influenced by CF in the life of these patients are detected, allowing individualized care and leading to in consideration of the patient's own perception. It is evaluated that, in order to understand the influence of the independent variables of the HRQoL, further studies are necessary, with an increase in the sample.

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