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LIVING WITH CYSTIC FIBROSIS: THE PERSONAL BRAZILIAN ADOLESCENT VIEW[1]

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ABSTRACT. The present study examined the psychological challenges of adolescents with cystic fibrosis (CF) in Brazil. A semi-structured interview with open-ended questions [(i) What is it like for you to have CF?; (ii) Do you have any special needs because you have CF? (iii) How do you envision your future?] was conducted with 42 CF adolescents' patients. We investigated how adolescents faces CF, identified their needs, and how they envision the future. The results show that the adolescents' emotions included fear of death, embarrassment, and anger. Other concerns included the loss of freedom, falling behind at school, a loss of friends, equality and acceptance, and the future perspectives. These feelings and concerns were influenced by the disease and may affect coping with CF. Few studies have examined adolescents' adjustment to living with CF in South America; understanding how to live with CF in adolescence contributes to new psychological interventions for patients and families, stimulate new research, and assist healthcare professionals and others who work and care specific to CF adolescents.

Keywords: Chronic diseases; cystic fibrosis; emotions.

VIVER COM FIBROSE CÍSTICA: A VISÃO PESSOAL DO ADOLESCENTE BRASILEIRO

RESUMO. O presente estudo examinou os desafios psicológicos de adolescentes com fibrose cística (FC) no Brasil, por meio de uma entrevista semiestruturada com perguntas abertas: [(i) Como é ter FC para você?; (ii) Você tem necessidades especiais por ter FC?; (iii) Como você vê o seu futuro?] Foi realizada a entrevista com 42 adolescentes com FC. Foi investigado como os adolescentes lidam com a FC, identificado suas necessidades, e como eles vislumbram o futuro. Os resultados mostram que as emoções dos adolescentes incluíam medo da morte, vergonha e raiva. Outras preocupações incluíam a perda da liberdade, ficando com atraso na escola, perda de amigos, da igualdade e aceitação, e as perspectivas futuras. Esses sentimentos e preocupações foram influenciados pela doença. Há poucos estudos que examinaram ajuste dos adolescentes ao convívio com a FC na América do Sul. Assim, buscamos compreender como é viver com uma doença crônica na adolescência; como isso pode contribuir para novas intervenções psicológicas para pacientes e familiares; estimular novas pesquisas, e auxiliar os profissionais de saúde nos cuidados específicos aos adolescentes com FC.

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Palavras-chave: Doenças crônicas; fibrose cística; emoções.

VIVIENDO CON LA FIBROSIS QUÍSTICA: UNA VISIÓN PERSONAL DE LA ADOLESCENTE BRASILEÑA

RESUMEN. Estudio examinó los retos psicológicos de los adolescentes con fibrosis quística(FQ). Las entrevistas semi-estructuradas con preguntas abiertas [(i) Cómo tienen fibrosis quística para usted? (ii) Tiene necesidad especial de tener la fibrosis quística? (iii) Cómo se imagina su futuro?] se llevó a cabo con 42 adolescentes con FQ. Se investigó cómo los adolescentes tratan FQ, identificas sus necesidades, y cómo ve el futuro. Los resultados muestran que las emociones de los adolescentes incluyen miedo de la muerte, la vergüenza y la ira. Otras preocupaciones incluyen la pérdida de la libertad, quedarse hasta tarde en la escuela, la pérdida de amigos, la igualdad, la aceptación, y las perspectivas futuras. Estos sentimientos y preocupaciones se vieron afectados por la enfermedad. Pocos estudios han examinado el ajuste de los adolescentes que viven con FQ en América del Sur; y comprender sus experiencias pueden conducir al desarrollo de nuevas intervenciones para pacientes y familiares, estimular nuevos profesionales de la investigación y la ayuda de salud en el cuidado de los adolescentes.

Palabras-clave: Enfermedades crónicas; fibrosis quística; emociones.

Introduction

Cystic fibrosis (CF) is a chronic monogenic autosomal recessive disease, with systemic manifestations affecting mailing the respiratory, digestive and reproductive systems. It´s considered the most common lethal congenital disease in Caucasian populations [central Europe, United States of America (USA), and Australia]. CF occurs by *CFTR* (Cystic Fibrosis Transmembrane Regulator) mutations leading with qualitative/quantitative dysfunction or absence of CFTR protein causing highest electrolyte chloride secretion by exocrine glands (Mall & Hartl, 2014). CF incidence varies between countries and ethnic groups (it is particularly unusual in blacks and Asians), ranging from 1:2.000 to 1:5.000 in Caucasian live births in USA, and Canada (Zvereff, Faruki, Edwards, & Friedman, 2013; Mackenzie et al., 2014).

Numerous people experience diseases as an attack on his integrity. In addition, this situation experienced in adolescence can trigger an overlapping crisis to another. In other words, sum up the crisis of the disease bodily and psychological changes inherent in the very phase of development of adolescence (Saraiva & Oliveira, 2008).

Adolescence and the stage of psychological development can be difficult and frustrating for young healthy. In addition, for young people with illness, the adolescence and the psychological development can be disappointing, leading to serious frustrations. In adolescence development phase, it is expected that independence begins to arouse more intense and continues, which can often be difficult for CF adolescents. In this phase, the quest for autonomy creates conflict forward dependence on medications and rigid routine treatments in CF patients (Saraiva & Oliveira, 2008).

CF treatment is based in interdisciplinary teams of healthcare professionals including the psychological aspects (Nobili et al., 2011). The disease involves daily treatment that requiring nearly than 120 min/day and include special diets, physiotherapy, vitamin supplementation, inhaled and oral antibiotics (Higham, Ahmed, & Ahmed, 1996). The limitations resulting from CF and its management are a constant source of stress, which can affect the emotional, social life, educational and professional performance of the CF patient. In this context, CF patients can develop psychological problems and to get difficulties to establish long-term goals and to integrate socially (Willians & Mason, 2000). Quittner and Slater (2005) found that psychological and psychiatric changes in CF adolescents include depression, sadness, apathy, being ashamed of their body, being afraid to start new romantic relationships, and uncertainty regarding the future. Therefore, CF adolescents live with the usual difficulties associated with adolescence, combined with emotional issues by a chronic disease, and challenges related to the transition from pediatric to adult health services (Taylor, Gibson, & Franck, 2008; Tuchman & Schwartz, 2013). During this life´s phase, adolescents can have preferences and objectives that differ from those of their parents and healthcare professionals, which can lead to poor adherence to treatment and emotional disruption (Brengnballe, Schiotz, & Lomborg, 2013).

In USA and Europe, high rates of depression and anxiety were reported in CF adolescents. Depression and anxiety symptoms were found in 10% and 22% of CF adolescents, respectively. The anxiety symptom was associated with depression (OR= 14.97) (Quittner et al., 2014). A survey in Brazil investigated the CF effects in the adolescence to understand the perception about puberty, plans and adherence to treatment. They managed to realize that existed in the group studied, difficulties in relation to sexuality, acquisition of autonomy and independence, relationship with peers and adherence to treatment (Rocha, Oliveira, & Moreira, 2004). Studies how to live with a chronic disease in adolescence from an individual viewpoint of emotional aspects are rare in South America. An overview about this phase was done in a reference center in Brazil to enable the understanding about the internal conflicts leading with two aspects – CF disease and emotional aspects related with the disease, as well the adolescence perspective about the future. In this context, the aim of the study was to collect the individual challenges of CF adolescents about how is to live with CF in Brazil.

Method

Type and Location of Study

A qualitative study was carried out with CF patients from reference center at State University of Campinas (Unicamp)/Brazil, by two consecutively years.

Participants

CF adolescents between ten and 19 years of age (n= 52) from the reference center were enrolled.

Procedures

After the phone contact, CF patients are attended in the outpatient clinic. The patients and their caregivers received an explanation of the research objectives and those who agreed, signed the free and informed consent. The interviews were recorded with the agreement of the participants. Caregivers did not attend the interview.

Qualitative study model with a semi-structured interview was performed to access the objectives: understanding the influence of the CF disease on the lives of CF patients. The questions were framed to express an empathic understanding of the significance of the adolescents' comments. The questions performed were: (i) What is it like for you to have CF?; (ii) Do you have any special needs because you have CF?; (iii) How do you envision your future? The categories for each question are shown in **Table 1**.

After the interview, we offered psychological support to CF patients about the problems highlighted. The majority (~75%) of the patients got adhesion on the psychological support.

The data collected in the interviews were analyzed through content analysis. Content analysis is a set of communication analysis techniques, using systematic procedures and description of the objectives of message content. It is a method widely used in the analysis of qualitative data, with a set of techniques for the purpose of search of a document senses (Campos, 2004).

Ethical Considerations

The study was approved by the Ethics Committee on Research of the Faculty of Medical Sciences at Unicamp according to the Brazilian Resolution 196/96 (#1155/2009).

Data Analysis

Transcriptions were completed following the content analysis criteria. This process consists of recording, transcribing, and repeatedly listening to the patient interviews to allow the representative parts of their experiences to emerge. The interview records were transcribed in chronological order and patients number (PN) were used to ensure patients anonymity. The interviewer listened to the answers at least three times with a psychologist in order to decide the categories and subcategories. These categories were identified following the criteria of repetition and relevance to the study. In the next step, another psychologist, blind to the previous coding, listened to the same set of responses and independently categorized the responses. In the case of disagreement, the researchers listened to that specific answer and categorized after reaching consensus. The transcriptions were performed without language correction.

Results

Table 1. Semi-structured questions and the associated categories and subcategories identified including numerical data

Questions	Categories	Patients (%)
Question 1: What is it like for you to have CF?	Perception that all is well	12 (29)
	Emotional perspective:	
	- Fear of death	24 (57)
	- Embarrassment	30 (71)
	- Anger	32 (76)
	Perception of losses suffered:	
	- Loss of freedom	40 (95)
	- Falling behind at school	35 (83)
	- Loss of friends	32 (76)
	- Loss of self-confidence	30 (71)
Question 2: Do you have any special needs because you have CF?	Objective needs:	
	- Needs related to recognizing the consequences of CF	40 (95)
	Subjective needs:	
	- Equality	30 (71)
	- Acceptance	34 (81)
Question 3: How do you envision your future?	Negative views	28 (67)
	Positive views	12 (29)
	Expectations about a cure for CF	40 (95)

CF, cystic fibrosis; %, percentage.

Of the 52 CF adolescents invited, 42 (80.7%) accepted to participate [23 (54.8%) women]. Ten patients (19.3%) did not attend a consultation or did not participate in the study due to unavailability.

The patient's average age was 15.85 (± 2.8) years. The CF diagnosis was performed with an average of 2.58 (± 3.32) years. The CF population enrolled showed an average of 13.3 (± 3.29) years.

Of those interviewed, 39 (92.8%) were from the state where the center was located from the country. The interviewed attended regular school with their age group. Ten patients (24%) reported having their school performance adversely affected by CF. According to religion, 27 (64.3%) were Catholic, seven (16.7%) were Evangelic, five (11.9%) did not belong to any religion, one (2.4%) was Buddhist, one (2.4%) Spirits and one (2.4%) Jehovah's Witness.

The length of the interviews was 60 (± 20) minutes. The analysis of the replies of the adolescents to the questions given in **Table 1** demonstrated that they experienced lifestyle changes due to hospitalization, limits imposed by the disease, treatment, anguish, alterations in the rhythm of life, and conflicting feelings of hope for life and fear of death.

Question 1

What is it like for you to have CF? The categories identified in the responses to the first question were: (i) perception that all is well, (ii) emotional perspectives, (iii) perception of losses suffered.

Perception that all is well. This expression is commonly used by adolescents to avoid facing something that generates conflicts or emotions that are difficult to verbalize (Fonaggy & Allison, 2015). It also represent a refusal to deal with the disease.

> *It is something, which tires you, it is a drag, I do not like to talk… (silence), put later that I said it was alright, really alright… (PN1, 11 years old)*
>
> *(…) to have CF, I do not know, I think it is alright, I do not know, my mother is the one who remembers when I need to take my medicine. (PN2, 11 years old)*

Emotional perspectives. The chronic disease on adolescent can cause adverse emotional effects (Willians & Maso, 2000). These effects were grouped in three subcategories: (i) fear of death, (ii) embarrassment, (iii) anger.

Fear of death. Among healthy adolescents, the death is rarely mentioned because they are more concerned with the "here and now" (Higham, Ahmed, & Ahmed, 2013; Bates & Kearney, 2014).

> *It is hell to have this, I am afraid of dying every time I get ill and I have crises, it is a pain…I feel like I am about to die, the only person I knew who had this died…so I think I am also going to die soon too. (PN3, 15 years old)*
>
> *(…) I speak of dying, but I think I fear what it is like to die, sometimes I say that I would rather die than take so many medicines, but I think what I really want is to live. (PN4, 14 years old)*
>
> *(…) I can not stand this anymore, if it were only the CF, but I have all these things as I said, diabetes, to have a finger pricked, have one injection a day, medicines, well, I don't know, sometimes I think death is kind of a good thing, but only sometimes (…) (PN4, 14 years old)*

Embarrassment. Adolescents do not welcome comments regarding their physical appearance and health problems.

> *I do not like it when my mother tells people about my disease. I am embarrassed, not even my best friend knows. (P5, 10 years old)*
>
> *(…) it is awful, I feel embarrassed, this cough never stops, it is awful, really awful, sometimes I try to hide the fact that I am coughing, everyone looks at points and me. (PN3, 15 years old)*

Anger. Anger forms part of the defense mechanism to deal with the fear about chronic disease.

> *(…) that is all I have to say, I do not like having this, it is awful, it is terrible to have this, a drag, and I feel like shouting that I do not want to have this. (P3, 15 years old)*

Perception of losses suffered. For the CF adolescents who participated in this research, the losses were reported as an interruption to the everyday life, with changes in the daily routine and a feeling of incompetence and failure. This category was grouped in four subcategories, as follow: (i) loss of freedom, (ii) failing behind at school, (iii) loss of friends, (iv) loss of self-confidence.

Loss of freedom.

> *(…) I ca not accompany the others, I ca not go out at night, I have to stop to have my treatment and go to hospital. I have to take medicines every day. I ca not go out at night because of this. (PN6, 13 years old)*

Falling behind at school.

> *(…) having CF is a drag, because it messes me up at school. When I go to school and get sick, it messes me up. I have already flunked many times. I have been in the hospital many times. In addition, I have to have surgery next week and I will be left behind at school (…) (PN7, 17 years old)*

Loss of friends.

> *I do not have many friends in my class, only A, who is older than me, because those of my age are already in a more advanced class. I have lost many friends because I did not progress to the next year with my class and they do not want to hang out with me anymore. (PN7, 17 years old).*

Loss of self-confidence.

> *(…) I was afraid that he would not accept my disease and would leave me. (PN8, 18 years old)*
>
> *(…) I want to date and I do not date, because I am afraid to tell him about the CF in case he does not want to be with me anymore (…) (PN9, 14 years old)*

Question 2

Do you have any special needs because you have CF? This question sought to understand the everyday needs of the CF patients; we noted both objective and subjective needs.

Objective needs

Needs related to recognizing the consequences of CF. The adolescents had to deal with the loss of social relationships, financial losses, and reduced physical capacity for activities, mainly leisure activities, because they felt uncomfortable with their physical appearance and threatened by the disease.

> *I need time to get into physiotherapy; if I did not have to do it, things would be easier. When I go to someone's home, I have to return early to take my medicines. Now, at this age I have more responsibilities, more is demanded of me, so it becomes more difficult. (PN10, 15 years old)*
>
> *(...) sometimes it is a bit of a drag having to take the medicine, I have to eat every 20 minutes, if not, I have to take medicine again. Sometimes I want to do something and I have to keep stopping, it is a pain and takes time. (PN11, 11 years old)*

Subjective needs

Equality. The search for a personal identity occurs early in adolescence; to succeed, adolescents need to feel accepted by and equal to their peers.

> *I would like to be just like my friends, they do not need to take medicines every day or get into physiotherapy; they eat and do not need to take loads of medicines. (PN3, 15 years old).*
>
> *I would like to be the same as everyone else, run, run and not be sick afterwards and everyone wanting to know what is wrong with you. (PN12, 14 years old).*

Acceptance. Within this need for existential acceptance is the need to deal with body image.

> *I know G, who has to take medicines and use soaps for her skin, but she is like this, she takes medicines but her treatment is finishing and mine is not, it never ends. (PN9, 14 years old).*

> *CF hinders my normal growth …, growing takes longer than other people's, I think it will take a long time for me to be the same size as my friends. (PN6, 13 years old)*

> *(…) my friends say that I am strange, that I am a little fat here (pointing to arms) and thin here (point to legs) (…) (PN9, 14 years old)*

> *(...) it is only my height, they make jokes. (PN13, 13 years old)*

Question 3

How do you envision your future? The aim of this question was discover the expectations and life projects of the CF adolescents, in an attempt to understand whether from their prospective CF changes their plans and projects for the future. The categories identified in the responses to this question were: (i) negative view of the future, (ii) positive view of the future, (iii) expectations about a cure for CF.

Negative views. Adolescents with chronic diseases, such as CF, are faced with different and difficult questions regarding school routine, work, family relationships, and how they think about and plan the future due to the disease.

> *I think about doing a degree in mechanical engineering, but sometimes I ask myself: how long can I live as a CF patient? (PN14, 17 years old)*

> *I would like to do many things, but I do not know if I can, sometimes I think that I am always going to depend on my mother and my grandmother. (PN4, 14 years old)*

Positive views. For some individuals, physical problems do not alter their expectations about their projects and perspectives of fulfillment.

> *I think about living as normally as possible, within the limitations, which I have, to go to a good university, have a good job and live a long time. (PN10, 15 years old)*

> *I want to study, work, have children, and I know that CF is not going to mess up my plans. (PN15, 17 years old)*

Expectations about a cure for CF. Adolescents expressed hope for a cure as a way to lessen the discomfort and changes forced on their everyday lives.

> *For my future (…) sometimes it is awful to have to be always coming here to the hospital and if there was a cure I would not have to come so often. (PN16, 13 years old)*

> *If in my future they have already developed a cure for CF it will be much better because sometimes it is a drag having to keep taking the medicine, then everything has to be eaten within 20 minutes, if not the medicine has to be taken again. (PN11, 11 years old)*

> *If there is a cure before I get older it will be better, I will be more self-assured, I dream of a cure and hope that they find it soon. (PN12, 14 years old)*

Discussion

Adolescence is a period of high physical and emotional conflicts (Bee, 1997). Associated with a chronic illness, such conflicts can be more intense, hindering the natural development (Kyngas, 2000).

Adolescent's answers focused on the lifestyle changes due to hospitalization, limits imposed by the CF disease, treatment, anguish, alterations in the rhythm of life, and conflicting feelings of hope for life and fear of death.

For the first question - *"What is it like for you to have CF?"* - Participants held an attitude that "everything is fine". Everything is fine is an expression commonly used by adolescents to avoid facing something that generates conflicts or emotions that are difficult to verbalize (Fonaggy & Allison, 2015). It also represents a refusal to deal with the disease. The major part of CF patients with "everything is fine" responses were in the beginning of adolescence (ten - 12 years old). Maybe, in the beginning of adolescence, the patients do not fully understand the disease and rely on the support and aid of their families. The fact that an adolescent has a chronic disease can cause adverse emotional effects, which can affect their treatment adherence, disease acceptance, and the way in which they face life (Zindani, Streetman, Streetman, & Nasr, 2006). Emotions expressed by our participants included "fear of death", among healthy adolescents. The death was rarely mentioned because they are more concerned with the "here and now" (Kyngas, 2000). Health adolescent´s show a whole libido that revolves around the construction of their world and, thus, there is little "space" for thinking of death (Kyngas, 2000; Higham et al., 2013; Bates & Kearney, 2014). For adolescents who live with chronic disease, fear of death is often present. Chronic diseases affect the development of the individual and can intensify the problems associated with passing from one developmental stage to another. The defense and confrontation mechanisms used to deal with a chronic disease, particularly CF, vary according to the length of time the patient has the disease, its severity, the presence of comorbidities, the prognosis, the degree of limitation, and the way in which the individual perceives the death (Kyngas, 2000). It is not death but rather dying which is feared.

CF adolescents want to be seen as health subjects, without the stigma of being ill. For the adolescents in this study, being ill was negative and meant being unwholesome, undesirable, and socially undervalued. To say that "I have CF disease", was associated with embarrassment by the patients in our casuistic. It is usual for the adolescent express anger. Anger forms part of the defense mechanism to deal with the fear associated with chronic disease and tends to appear as the denial mechanism. The individual begins to experience anger, which is aimed at doctors, nurses, caregivers, God, and others (Kyngas, 2000).

A Brazilian study conducted with adolescents, showed that respondents also reported feeling anger. In adolescence, it is natural that they want to feel equal in pairs, belonging to the same group. Having a chronic disease, such as CF, where often the physical development takes place more slowly, begin to feel different from their peers, rising anger in response (Rock, Oliveira, & Moreira, 2004). Often this feeling can also occur by the physical changes expected at this stage of development, where there are hormonal changes, occurring mood swings (Bee, 1997). These changes occur so quickly that often not even the teenagers recognize their attitudes and feelings (Knobel, 1978).

The adolescents who participated in this research related losses, as interruption to everyday life, with changes in the daily routine and feeling of incompetence and failure. Loss of freedom, one of the losses reported by our participants, has been described in previous studies (Kyngas, 2000). Adolescents confronting a chronic disease feel different from others without the disease. Patients have a routine associated with the treatment. The treatment is not a reality for the healthy adolescents. In this way, feelings of social isolation can be present. Studies with adolescents and chronic disease have shown that they have difficulties at school because they feel embarrassed about changes in their appearance and a lack of physical abilities, which leads to feelings of social isolation (Rhee, Wenzel, & Steeves, 2007) and loss of friendships. Adolescents reported problems adjusting to changes caused by their illness, particularly in adolescence, when they spent most of their time with friends (Modi & Quittner, 2006). Losses and the chronic nature of the disease can lead to a fear of ineffective friendship; this fact can get a loss of self-confidence.

For the second question - *"Do you have any special needs because you have CF?"* - The subjects had objective and subjective needs. CF changes the everyday rhythm, while the priority before was to play; there are now the restrictions to adolescent interests owing to their chronic disease. The priority now for these adolescents is often the disease and they need to adapt to the limitations related to physical conditions, food, and changes in their everyday life, associated with the presence of chronic disease. We found that the time spent on CF treatment routines nearly than 120 min/day, which included physiotherapy, doctor care, taking medicines, administering inhalants, accordingly the disease evolution, mainly for the lung disease (Higham, Ahmed, & Ahmed, 2013). A study in Brazil showed the difficult for CF patients considering the adherence to treatment, especially regarding diet and physical activity. This difficulty is not associated with financial support, because the medication is provided by the government, but by the difficulty in getting time for all required tasks (Dalcin et al., 2007). The adolescents in this study had to deal with the social relationships and financial losses, and reduced physical capacity for activities, mainly leisure activities, because they felt uncomfortable with their physical appearance and threatened by the disease.

Subjective needs included the desire for equality and acceptance. The search for a personal identity occurs in the beginning of adolescence; to succeed, adolescents need to feel accepted by and equal to their peers. This feeling of having an identity can have two forms: the first is to see oneself as the same and continuous in time and space - recognize oneself as a part of a group -, and the second is to see that others recognize this similarity and continuity - the group recognize you as a part of a group. The development of an identity and individual values often requires comparisons of one's similarities and differences from others and the recognition of limitations and abilities. During identity formation, an individual needs external support and outside opinions to evaluate themselves (Sestito & Sica, 2014; Lannegrand & Barbot, 2015). Part of the need for equality is the desire to be accepted, which requires dealing with body image. Adolescents do not welcome comments regarding their physical appearance and health problems; CF adolescents want to be seen as health subjects, without the stigma of being ill. For adolescents in this study, being ill was negative and meant being unwholesome, undesirable, and socially devalued. In general, in this situation, comparisons, which may cause lower self-esteem and feelings of discrimination, inhibit the acceptance of their reality. In general, adolescents are concerned with the transformation of their body, which requires the acceptance of an altered body image, making one feel different. Deviations from the "idealized body," which may result from therapy, are not tolerated, and neither are the limitations of the disease that modifies your appearance and restricts your independence and abilities (Rocha, Oliveira, & Moreira, 2004). In other study, the concern of teenagers with body self-image problems seems to be a recurring issue for CF adolescents. At this stage of life is important they see themselves as similar to peers. In this disease, physical development can happen later. Being able to generate some feelings for these adolescents, such as depression and anxiety in relation to the idealized body and reality (Rocha, Oliveira, & Moreira, 2004; Quittner et al., 2014).

Negative and positive views were included in the responses to our third question - "How do you envision your future?" Adolescents with chronic diseases, such as CF, are faced with different and difficult questions regarding school life, work, family relationships, and how they think about and plan the future due to the disease. They are often physically, emotionally, and financially dependent on their family, which has implications regarding the transition to independent living and from school to work. It is important that adolescents manage to maintain their self-esteem and confidence (Rhee, Wenzel, & Steeves, 2007). However, for some individuals, physical problems do not alter their expectations about their projects and perspectives of fulfillment. Some participants reported efforts to reach normality and, despite the effects of the disease and treatment on their lives, they believed in the future (McEwan, Espie, Metcalfe, Brodie, & Wilson, 2004). We found similar positive views of the future among our participants.

The question about the CF future can be associated with the knowledge about new personalized therapy, which is a plausible treatment to correct the basis of the CF genetic disease (Marson, Bertuzzo, & Ribeiro, 2015). To know about the therapies can bring hope for the CF patients. Another aspect related with the disease is the new life expectancy, which improve considerably in the last decades (Kuehn, 2014; Stephenson et al., 2015). Nowadays, CF patients can have a normal life style, considering the limitations about the disease treatment. Each new medicine and treatment available bring a better clinical outcome and quality of life, then, step by step we are achieving a long time dream for the CF disease, the "normal" life. For our patients enrolled, the lung disease was present before the onset study, then we did not have the special condition that we have known, but the positive expectations about a cure for CF were important to improve the future for our CF patients. We have the bad issues related with the disease, although the hope is present yet. A study conducted in Brazil showed that new treatments can improve the quality of life and life expectancy of CF patients and consequently their vision about the disease in the emotional and social spheres (Rozov et al., 2010).

Considering the expectations and emotions about a cure for CF, the adolescents and the expressed hope for a cure as a way to lessen the discomfort and changes forced on their everyday lives. This feeling of hope, which can involve the search for a cure, is different from denial because it does not affect the adjustment to the disease process, and it is compatible with acceptance of the reality and adhering to treatment (Stephenson et al., 2015).

Final considerations

The results obtained from the interviews showed that CF has emotional repercussions for adolescents. The magnitude of the disease effect on their lives can be observed through their answers and is experienced differently by each individual. Chronic disease brings changes to the lives of adolescents. Therefore, it is of important that they had a place to voice their concerns and reactions to the disease. Assistance given to these adolescents should the physical and emotional challenges.

The methods of leading with CF are closely related to internal resources, which each of these adolescents manifested, and social and family support. Their acceptance of the disease and adherence to treatment is dependent on their perception of CF, having needs met, and expectations of the future.

This study suggests new avenues for future research and concrete actions to benefit and improve the quality of life of these individuals, who pass through their adolescence in a paradox: desire to live and fear of death. Also gave us an opportunity to reflect on methods for improving the quality of life of the patients, family, and healthcare professionals who care for these patients.

We suggest the following actions to provide for CF adolescent patients living in South America better knowledge, enabling that the patients to feel welcomed and supported.

1. Inclusion of mental health professionals in the interdisciplinary healthcare teams;
2. Studies that address the emotional aspects related to coping with a chronic disease and comparisons with control groups;
3. Studies on the social and sexual evolution and quality of life of CF adolescents;
4. Opportunities for patients to express their feelings about living with a chronic disease;
5. Further research and education by the interdisciplinary healthcare teams so they are better prepared to deal with the emotional pain of the patients;
6. Research on different phases of adolescence and youth, for instance, pubescent and non-pubescent adolescents;
7. Multidisciplinary studies at different reference centers;
8. Evaluation of the psychological aspects of CF patients by the caregivers (parents, guardians and healthcare professionals);
9. Design of questionnaires that evaluate the psychological issues of patients and consider the differences in realities and cultures;
10. CF awareness programs in schools for students and teachers to prevent discrimination based on a lack of knowledge.

Considering the adolescence a complex and peculiar period of life is important that all those involved parts seek to understand this step and assist the adolescents in this delicate moment. The adolescence is part of a natural process of development that shows numerous degree of difficulty or defensive mechanisms to deal with chronic disease. Family members and health care staff involved must support, guide and encourage the adolescents looking for healthier possible way to deal with this transition of human development.

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