Original Article

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Qulaity of Life in Cystic Fibrosis Children

Hamid-Reza Kianifar*1, MD; Banafsheh Bakhshoodeh[1], MD; Paria Hebrani[2], MD; Fatemeh Behdani[2], MD

1. Allergy Research Center, Ghaem Hospital, Mashhad University of Medical Sciences, Mashhad, Iran
2. Department of Psychiatry, Psychiatry and Behavioral Sciences Research Center, Ibn-e-Sina Hospital, Mashhad University of Medical Sciences, Mashhad, Iran

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Abstract

Objective: Cystic fibrosis (CF) is a chronic, multisystem genetic disease with a wide variability in clinical severity. The measurement of quality of life in CF provides additional information about the impact of this disease. This article tries to assess quality of life (QoL) in children and adolescents with CF and to compare it with control group.

Methods: Patients 2-18 years old with admission diagnosis of cystic fibrosis entered the study. QoL was observed in CF patients and compared with control group.

Findings: Based on children's reports, significant differences between the CF patients and control group were noted for emotional, physical, social, school performance, and total scores (P<0.05). Based on parents' reports, quality of life score in CF patients from the physical point of view as well as social and total scores were decreased (P<0.05).

Conclusion: QoL in CF patients seems to be low, and therapy programs should take into account the suggestive perceived quality of life.

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Key Words: Cystic fibrosis; Quality of life; Child; Adolescence

Introduction

Cystic fibrosis (CF) is an autosomal recessive disease involving various organs. It is mainly defined as the obstruction and infection of respiratory airways as well as indigestion and several systemic complications[1,2]. CF can result in early death, although there has been a significant increase in life expectancy during the past years. Infants affected by severe types usually pass away but children who skip this period may have a normal adolescence[1].

CF has a huge influence on patients' life. Medical insurance, job, family control and life expectancy are major issues. Therefore supporting measurements will have an important role in order to improve psychosocial adaptation[2]. A study conducted in Belgium in 2008 looked at the association between patient reported health-related quality of life (HRQoL), anxiety and depression. Those with depressive symptoms reported lower HRQoL scores for emotional functioning, eating disturbances and body image[3]. However, in a study performed in 2005 on HRQoL, no significant difference was observed between the study and control groups[4].

Although several studies proved that CF lowers quality of life in adults due to limitations in physical function, their psychosocial function did not show a big difference from the control

*** Corresponding Author;**
Address: Department Pediatric Gastroenterology Allergy Research Center Ghaem Hospital Mashhad University of Medical Sciences
E-mail: kianifarhr@mums.ac.ir

group[5,6]. Few studies have been conducted on the quality of life in children and adolescents with CF based on their physical, emotional, social, and school functions in comparison to a control group. Thus in this study we aimed on assessing the quality of life in patients with CF and comparing it with healthy children.

Subjects and Methods

This is a cross-sectional study in CF children who visited the CF clinic of Sheikh Hospital of Mashhad University of Medical Sciences in 2009-2011. Inclusion criteria were 2-18 years old children and adolescents with confirmed CF by a pediatric gastroenterologist based on their clinical manifestations and positive sweat test.

According to the fact that socio-economic status may be a confounding factor that can influence the life quality, controls were age matched siblings from CF families. As the control group was formed by the age-matched normal siblings of the patients, interfering factors such as socio-economical situation and family care were accounted for. Patients with other acute or chronic; internal or neurologic diseases (such as mental retardation) were excluded.

The two groups were then studied with regard to the quality of life using a specific questionnaire (Peds – QL™ 4.0 SF15). The Peds QL™ 4.0 SF15 questionnaire was used for four different age groups: 2-4, 5-7, 8-12 and 13-18 years of age, and included four different fields of children's function: physical function, emotional function, social function and school (and pre-school courses) function. Peds – QL™ had children forms (for 5-7, 8-12, 13-18 year old children) which were filled by children, and parents' forms (for 2-4, 5-7, 8-12, 13-18 year old children) which were filled by their parents. Each question could get one of the five scores of 0 for "never" up to 4 for "almost always"; the total score was calculated by dividing the added score of different sections over the number of answered questions[7] and an ideal score was 100. The pediatrics quality of life questionnaire (Peds QL™) was translated to Persian language and had a validity value of more than 0.7 when filled by the patient or his/her parents. The test and its repetition showed a high reliability in parents' reports (0.68-0.79) and a medium to high reliability in children's reports (0.46-0.73).

The final results were calculated using SPSS software, version 11.5 and also chi-square or Fisher's exact test for qualitative data and independent t test for quantitative data. A *P*-value <0.05 was considered as significant.

This research was approved by ethical committee of Mashhad University of Medical Sciences. Patients and/or their parents gave informed consent.

Findings

Thirty six children and adolescents with CF and 39 controls along with their parents participated in the study with the age range from 2 to 18 years and the mean age of 5.47±3.4 yrs. The control group age ranged from 2 to 17 years with a mean age of 4.84±2.43 yrs.

The study group consisted of 20 (55.6%) males and 16 (44.4%) females. The same figures were 21 (53.8%) and 18 (46.2%) in the control group in which according to the χ^2-test no significant relation existed.

Table 1 shows the difference in mean and standard deviation of the quality of life (QoL) test in CF patients with the control group at Dr Sheikh Hospital in the years 2009-2011 based on their parents' reports and in different age groups. The comparison was also done on a small scale in different age groups based on the parents' reports. In the 2-4 years age group QoL showed no significant difference between the two groups, whereas in the 5-7 years age group it did show a significant difference in the physical and general subgroups but not in the other aspects.

Table 2 shows the differences in mean and standard deviation of the QoL test in CF patients and controls in Dr Sheikh Hospital in the years 2009-2011 based on their own report and in different age groups. Comparing the mean score of the study and control group in the physical sub-group revealed a significant difference (*P*=0.001) which showed a lower level of quality of life in the CF group from this aspect. Significant difference

Table 1: The difference in mean (SD) of the quality of life test in CF patients with the control group based on their parents' reports and in different age groups (Mashhad 2009-2011)

Age group	Groups	General score		Physical		Emotional		Social		Schooling	
		Mean (SD)	*P* Value	Mean (SD)	*P* Value	Mean (SD)	*P* Value	Mean (SD)	*P* Value	Mean (SD)	*P* Value
2-4 Years	CF	76.9(26.1)	0.07	89.6(25.8)	0.6	88.3(15.1)	0.2	88(29.8)	0.9	–	
	Control	90.4(11)		93(9)		81.6(15.9)		96.6(8.3)			
5-7 Years	CF	73.3(28.2)	0.01	72.7(28.1)	0.003	79.0(25.2)	0.3	86.5(22.5)	0.08	83.5(25.5)	0.8
	Control	92.9(9.3)		97.3(6.3)		86.9(16.41)		97.2(7.7)		89.5(15.5)	
8-12 Years	CF	53.5(3.5)	0.3	39(7.1)	0.04	67.5(45.9)	0.8	90(0.0)	0.9	56(0.0)	0.6
	Control	81.3(27.2)		86.6(18.1)		76.6(32.2)		91.6(14.4)		91.6(7.6)	
13-18 Years	CF	57(8.5)	0.2	41.0(0.0)	0.2	65(14.1)	0.3	67.5(31.8)	0.6	70(7.1)	0.2
	Control	91		75		100		100		100	
Total	CF	72.8(26.1)	<0.001	76.1(29.4)	0.001	81.5(22.3)	0.6	86.2(25.2)	0.02	80.1(24.2)	0.1
	Control	90.7(11.7)		93.8(9.4)		83.8(17.3)		96.5(8.4)		90.7(13.6)	

CF: Cystic Fibrosis; SD: Standard Deviation

was seen between the mean scores (*P*=0.006) in terms of emotion by children's report. In the social sub-group the difference in mean scores was statistically significant (*P*=0.01) and in the schooling sub-group the mean scores also showed a significant difference (*P*=0.01). In general, the difference in the mean scores between the two groups was statistically significant (*P*<0.001) which proves a lower general QoL in CF patients in comparison to the controls.

In the latter age groups and based on the children's report, the two groups were statistically different from the physical, emotional, social and general point of view, but not in schooling.

In the 8-12 years age group and based on parents' reports the QoL solely differed in the physical subgroup; while the children reports showed no difference at all. In the 13-18 years age group, based on the parental reports, a difference in QoL of the two groups was only seen in the physical sub-group.

Discussion

CF is a rather common disease in Caucasians and is an important cause of chronic pulmonary involvement and a life-shortening disease among children.

In some developed countries due to the high prevalence of the disease and the importance of its early diagnosis, screening tests are performed at the time of birth; with the help of supportive treatments they have lessened the clinical signs and symptoms, improved growth indicators and increased life's quality and duration.

Studying the quality of life in patients with CF has led to paradoxical results; in Koscik RL et al study similar levels of quality of life were observed in CF neonates and their healthy peers[8], whereas in Schmitz study a lower level of health-related life satisfaction was seen in CF patients[9].

Comparing the mean results obtained by QoL test in CF children and the controls based on their

Table 2: The difference in mean (SD) of the quality of life test in CF children and controls, based on their own report and in different age groups (Mashhad 2009-2011)

Age group	Groups	General score		Physical		Emotional		Social		Schooling	
		Mean (SD)	*P* Value	Mean (SD)	*P* Value	Mean (SD)	*P* Value	Mean (SD)	*P* Value	Mean (SD)	*P* Value
5-7 Years	CF	76.8(19.8)	0.02	81.5(17.8)	0.006	78(26.2)	0.01	86.6(19.9)	0.04	72(30.8)	0.08
	Control	95.4(5.4)		96.8(6.4)		97.1(4.4)		98.4(3.5)		92(13.1)	
8-12 Years	CF	55(7.1)	0.05	48.5(10.6)	0.1	65(49.5)	0.7	82.5(24.8)	0.5	55(0.0)	0.1
	Control	87.3(12.9)		87.3(21.9)		78.3(22.5)		98.3(2.9)		88.3(10.4)	
13-18 Years	CF	57(8.5)	0.2	41.0(0.0)	0.2	65(14.1)	0.3	67.5(31.8)	0.6	70(7.1)	0.2
	Control	91		75		100		100		100	
Total	CF	71.6(20.6)	<0.001	72.7(23.9)	0.001	73.9(27.9)	0.006	84.8(21.1)	0.01	67.3(29.5)	0.01
	Control	93.9(7.1)		94.2(10.7)		94.6(10.9)		98.5(3.3)		91.8(12.0)	

CF: Cystic Fibrosis; SD: Standard Deviation

own reports, the difference in quality of life in CF patients from the emotional point of view as well as physical, social, schooling and the general sup-groups, was statistically significant. However, based on the parents' reports the quality of life of CF patients in the emotional and schooling sub-groups was not significantly different from the controls whereas in the physical, social and general sub-groups the difference was significant.

It is worth mentioning that very few studies have been conducted on evaluating QoL in CF children and adolescents from the physical, emotional, social and schooling aspects, especially on the basis of comparison with the control group. Most available studies have focused on assessing the prevalence of depression and its negative effect on treatment and QoL[3,10], its influence on QoL of neonates with CF[8], estimating health-related life satisfaction degree in CF patients[9], evaluating the effect of disease severity, medical factors and social situation on QoL in such cases[6,11,12].

In 2005, Thomas et al studied QoL in children receiving local CF treatment; they concluded that CF children living in suburb areas in England have an acceptable health-related quality of life[4]. It differs from our results proving a lower QoL in CF cases.

In 1993, QoL of CF adolescents were studied based on their schooling functions which showed a good QoL in this aspect[13]. In our study children aged 7 to 18 years also showed good QoL in schooling based on both the children's and their parents' reports. This could be due to good support from families, giving the essential information of the disease especially at the time of diagnosis and subsequently preventing psycho-social issues such as anxiety and depression due to irregular presence in school.

In another study by de Jong et al in 1997 on QoL of adults with CF, it was concluded that QoL of CF patients on the basis of physical function was lower in comparison to the control group whereas their emotional and social functions were almost similar to the controls[5].

Our results were similar to those of this study; the 8-18 year-olds showed a higher in comparison to 5-7 year-olds which could mean that those children who have reached older ages, probably had a lower disease severity and thus have suffered less from psycho-social aspects. Moreover 2-7 years old CF children reported a lower QoL.

However, in order to confirm this issue, children QoL should be simultaneously compared with their physiological improvement criteria.

Conclusion

According to the results of the current study and those reviewed by other researches, a low level of quality in physical, social, schooling and general aspects of life is the result of neglecting the other effects of this disease by pediatricians on children lives including psychological, cognitive and social aspects. Therefore, treating such cases as a teamwork consisting of pediatricians, pediatric psychologists and social service worker would be of incredible value which is usually ignored.

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Conflict of Interest: None

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