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Clinical characteristics and outcomes of cystic fibrosis in Palestine: Cross sectional study

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Abstract

Objective: To assess the clinical characteristics and outcomes of cystic fibrosis in Palestine by studying the quality of life (QoL) of participants.

Method: This cross-sectional study involved the application of Cystic Fibrosis Questionnaire-Revised (CFQ-R) to participants attending the pediatric pulmonology clinic at Caritas Baby Hospital between January and May 2017. Health status was assessed by measuring pulmonary function test (FEV_1), body mass index (BMI), age of CF diagnosis, and presence of other affected siblings or deaths in the family.

Results: There were 77 participants from 58 families: 46.8% (36/77) were males, and 53.3% (41/77) were females. The mean age was 10.7 years (range: 0.5–36 years). The participants were divided into three groups by age in years: group I (< 6), II (6–13), and III (≥ 14). The highest and lowest CFQ scores were for the eating domain in group III (55.6 ± 22.5) and the body domain in group II (14.5 ± 17.7), respectively. Mean illness severity was 69.6% (range: 33%–111%). The mean BMI was 15.9 (range: 9.6–23.1). The mean age at the time of diagnosis was 4.2 years (± 6.3). The study showed that 1.7% of the families (1/58) had four affected siblings, and 21% (12/58) had death cases related to CF, of which 58.3% (7/12) were from the Hebron district. Finally, all parameters for CF participants in West Bank, Palestine were noticeably lower than those reported in other countries.

Conclusions: This study illustrates the need for new therapies for CF participants in Palestine to improve QoL, health status, and longevity.

KEYWORDS
cystic fibrosis, quality of life

1 | INTRODUCTION

Cystic fibrosis (CF), also called mucoviscidosis, is a life-threatening inherited disease defined as a recessive genetic disease characterized by dehydration of the airway surface liquid and impaired mucociliary clearance.[1] The discovery of CF can be dated back to the Middle Ages, when people had the saying: "Woe to that child which when kissed on the forehead tastes salty. He is bewitched and soon must die." This is one of the earliest references to CF, recognizing the association between the salt loss in CF and illness, although the condition was unnamed at that time.[2]

Although the number of CF patients is increasing around the world, there is no accurate epidemiological data on CF disease in Palestine. The general impression has been that the disease is rare, but this is most likely the result of under-diagnosis or misdiagnosis due to limited awareness of the condition in the region.

This disease has a high treatment burden. Some of the novel technology used for drug delivery is highly beneficial because it may

ease the patient burden by decreasing administration time and offer improved efficacy and safety, including nutrition combined with the introduction of dornase-alfa, a mucolytic agent, tobramycin inhalation solution by reusable advanced nebulizer solution antibiotic, powder inhaler, and modulator therapies. However, these options are not available for CF patients in Palestine, and they depend on basic traditional therapies only. For example, hypertonic saline is used as a mucolytic, and gentamicin intended for intravenous administrations delivered as an inhalation solution by a simple nebulizer.[3]

The Cystic Fibrosis Questionnaire-Revised (CFQ-R) has been validated as a subjective measure to assess multiple domains of patient quality of life (QoL) and is approved by the FDA as a patient-reported outcome measure to be used as a disease-specific questionnaire.[4]

CFQ-R is currently the most widely used health-related QoL (HRQoL) instrument for CF and was rated "well-established" in recent reviews. The CFQ-R has not only been translated into 34 languages with validation studies in all countries but has demonstrated responsively in several clinical trials of medications with different mechanisms of action (e.g., antibiotics, mucus hydrators, gene potentiators).[4] In this study, we used the Arabic- Hebrew translation of CFQ-R without any modification.[5]

Most CF patient QoL studies have been conducted in developed countries and a few in developing countries; however, no studies were done in Palestine.[6] The objective of the present study was to evaluate CF participants in Palestine using specific comparison tools, including QoL, nutritional and pulmonary outcomes, and treatments used. The second objective was to compare the QoL scores of participants to those reported in developed countries. The hypothesis was that there would be no difference in the QoL domain scores for CF participants in Palestine compared with other populations worldwide.

2 | METHOD

This was a descriptive narrative study conducted for participants from West Bank suffering from CF disease. These participants used basic classic therapies including; oral antibiotic medications, vitamins (K, A, D, E), pancreatic enzymes, hypertonic (3%, 7%) solutions, bronchodilators (salbutamol solution), and gentamicin ampule given as inhalation solution by a nebulizer, and attended the pediatric pulmonology clinic in Caritas Baby Hospital (CBH) from January 2017 until May 2017. The main clinical manifestations were pulmonary infections, malnutrition, and pancreatic insufficiency. In total, 77 participants completed four quantitative assessment measures and provided demographic information. The participants' routine medication was not altered during the study, and potential participants and their parents were approached during routine clinic visits while waiting for their appointments.

Participants were included in the study if they met the following criteria: a proven clinical diagnosis of CF as evidenced by a positive sweat test or the presence of two known CF mutations confirmed by a pediatric pulmonologist based on their clinical manifestations. The principal inclusion criteria were as follows: The caregiver must have a child diagnosed with CF; the disease must have been diagnosed by a specialist doctor; the caregiver must have a reasonable understanding of the study and understand what is expected of them and be competent enough to refuse participation. The status of CF participants was evaluated using the QoL score (CFQ-R) questionnaire individual domain scores range from 0 to 100, with higher scores indicating better health-related quality of life which considered normal with scoring more than 50. The participants were divided into groups by age: those <6 years of age ($CFQ_{<6}$), ≥6, and <12 (CFQ_{6-11}); ≥12 and <14 (CFQ_{12-13}); and ≥14 (CFQ_{14+}). Their health status was screened by measuring different parameters, including the pulmonary function test, body mass index (BMI), age at diagnosis, and mortality rate related to CF disease.

The Statistical Package for the Social Sciences (SPSS) was used for data processing. The scores are expressed as means and standard deviations. The Mann–Whitney test was used for comparing two groups, whereas the Kruskal–Wallis test was used for comparing more than two groups. Pearson's correlation coefficients (r) were calculated between CFQ-R scores and clinical outcomes, and the level of significance was set at $p < 0.05$. Health and socioeconomic status data were analyzed using the SPSS program. Descriptive statistics (means and standard deviations) were used to characterize the demographic variables.

The study was approved by CBH Medical Research Committee/Ethical Review Board (approval number: MRC-21). Written informed consents were obtained from the participants and parents (father or mother) of the children involved in this study. All signed informed consent forms were deposited in the participants' hospital medical charts.

International data for QoL was obtained from the University of Miami/School of Medicine, Miami, Florida, for comparison with our results and to demonstrate the status of our CF participants, the comparison was done by observation of mean score for both without statistical analysis.

3 | RESULTS

A total of 77 participants among all participants who visited CBH (58 families) have participated in this study. There were nine other CF participants that CBH was aware of; however, three of them refused to participate as they did not acknowledge the existence of the disease; three others could not be reached because they traveled to other countries for treatment; the remaining three had not been confirmed as CF.

There were 46.8% (36/77) male and 53.3% (41/77) female participants. The mean age of the participants was 10.7 years (range: 0.5–36 years). Its quartile statistics: first quartile ($Q1$) = 5, second quartile ($Q2$) = 10, third quartile ($Q3$) = 15, interquartile range (IQR) = 10, median = $Q2$ (x~) = 10, minimum (min) = 0.5, maximum (max) = 36, range (R) = 35.5. The majority of the participants were children (6–13 years old).

TABLE 1 Quality of life scores from the questionnaires related to cystic fibrosis participants, by group.[a]

Domain	Groups			
	CFQ_{14+} *n* (22)	CFQ_{6-13} *n* (33)	CFQ $parent_{6-13}$ *n* (33)	$CFQ_{<6}$ *n* (22)
Physical	31.1	26.4	37.6	49.7
Emotion	47.3	48.3	45.9	50.0
Eat	55.6	46.1	51.1	43.2
Treatment	20.2	17.5	41.4	38.4
Body	22.7	14.5	14.8	32.8
Respiratory	28.3	27.5	31.7	42.2
Social	44.9	45.5	-	-
Digest	39.9	23.2	30.6	39.4
Weight	22.7	-	15.1	25.8
Health	26.8	-	46.1	41.4
Role	40.2	-	-	-
Vitality	28.4	-	33.9	36.9
School	-	-	45.9	59.3

Note: (-), set for not applicable. Distributions of scores (0–100) standardized values) were calculated.

Abbreviations: CFQ, cystic fibrosis questionnaire; $CFQ_{<6}$: participants <6 years of age; CFQ_{6-13}, participants ≥6 and <14 years of age; CFQ_{14+}, participants ≥14 years of age.

[a]Values expressed as mean score.

The participants were distributed throughout the different regions of the West Bank (Jerusalem, Bethlehem, Hebron, Ramallah, Nablus, Jenin, Qalqilia, Tulkarem).

The overall scoring for participants in this study was less than 50%, which indicated poor QoL (Table 1). The data appeared to show the lowest score for treatment, body, and respiration. The highest score appeared to be for eating and emotion.

In the CFQ_{14+} group, the mean scores ranged from 20.2 for the treatment domain to 55.6 for the eating domain. The lowest mean score was for question 10, "You felt energetic?" with a value of 13.6 for the vitality domain, and the highest score was for question 25, "I think I look different from others my age?" with a value of 83.3 for the body domain.

In the CFQ_{6-13} group (Table 1), the mean scores ranged from 14.5 for the body domain to 48.2 for the emotion domain. The lowest mean score was for question 30, "Doing your treatments bothered you?" with a value of 9.1 for the treatment domain, and the highest score was for question 23, "You felt left out?" with a value of 91.92 for the social domain.

Although the CFQ < 6 parent version (Table 1), the mean scores ranged from 25.8 for the weight domain to 59.3 for the school domain. The lowest mean score was for question 30 "My child spends a lot of time on his/her treatments every day?" with a value of 15.2 for treatment domain and the highest score for question 14 "The extent to which your child participated in sports and other physical activities, such as gym class?" with a value of 90.9 for physical domain.

Table 1 shows the data regarding the questionnaire completed by the parents for children 6–13 years of age. In the CFQ $parents_{6-13}$, the mean scores ranged from 14.8 for the body domain to 51.1 in the eating domain. The lowest mean score was for question 21, "My child thinks that he/she is too thin?" with 11.1 for the body domain, and the highest score was for question 31, "How difficult is it for your child to do his/her treatments each day?" with a value of 73.7 for the treatment domain.

When we compared the mean scores of the CFQ_{14+} group with those of the CFQ_{6-13} group, we observed no statistically significant differences (Table 2) except for the eating domain. The scores determined for adult participants (14+) were slightly higher than those for children (6–13), except for the emotion and social domains. The scores for participants less than 6 years of age were much higher compared with both groups.

When we compared the mean scores of the CFQ $parent_{6-13}$ group with those of the CFQ_{6-13} group, we observed no statistically significant differences (Table 3) except in the physical and treatment domains.

3.1 | Health profile section

Screening the participant medications revealed that all participants took vitamin A&D (Adol®) drops, tocopherol (Evitol®) tablet, vitamin K 2 mg ampoule, hypertonic saline (3%, 7%) solutions, pancreatic Creon 10000 IU tablet, salbutamol 5 mg/ml (Ventolin®) nebulized solution, gentamicin 80 mg ampoule (every other month colonized with *Pseudomonas aeruginosa*), and other oral antibiotics.

Illness severity measured by FEV_1 was available for only 26 participants (Table 4) and ranged from 33% to 111%, with a predicted mean value of 69.6%. The majority of the participants (53.8% from the available data) fell into the moderate category ($40 \le FEV_1 < 70\%$, predicted). Of the 26 participants evaluated, 2 had a predicted $FEV1_1 < 40\%$ (mean = 36), 14 had $40 \le FEV_1 < 70\%$ (mean = 61.3), and 10 had $FEV1_1 \ge 70$ (mean = 88).

BMI recorded for 72 participants ranged between 9.6–23.1 with a mean value of 15.9, as shown in Table 4. The quartile statistics for adult participant was first quartile $q_1 = 17.305$, second quartile $q_2 = 17.85$, third quartile $q_3 = 20.68$, IQR = 3.375, median = $q2$ ($x\sim$) = 17.85 minimum (min = 14.00, maximum (max) = 23.12, range (r) = 9.12. To measure the health status of five participants under 2 years of age, we used the weight per length percentile. The results showed that they were in less than the 5th percentile; thus, they were in severe status (2.9 kg/51 cm, 9.7 kg/79 cm, 9 kg/75 cm, 11 kg/88 cm, 2.7 kg/49 cm). Age at the time of CF diagnosis was determined (Table 4); the overall mean age of diagnosis for participants in our sample was 4.2 years of age. For adult quartile Statistics analysis was first quartile $q1 = 6$, second quartile $q2 = 10.5$, third quartile $q3 = 20$, Interquartile range IQR = 14, median = $q2$ ($x\sim$) = 10.5, min = 1, max = 30, r = 29.

TABLE 2 Compression between mean scores of the CFQ_{14+} group with CFQ_{6-13} group by which the Eat domain was significant.

Ranks					Level of significance	
		N	Mean rank	Sum of ranks	Mann–Whitney U	Asymp. Sig. (two-tailed) *$p < 0.05$
Physical	Child	33	27.0	891.0	330.0	0.569
	Adults	22	29.5	649.0		
	Total	55				
Emotion	Child	33	27.9	921.0	360.0	0.959
	Adults	22	28.1	619.0		
	Total	55				
Eat	Child	33	24.4	806.0	245.0	0.041*
	Adults	22	33.4	734.0		
	Total	55				
Body	Child	33	26.9	886.0	325.0	0.492
	Adults	22	29.7	654.0		
	Total	55				
Treatment	Child	33	28.8	951.0	336.0	0.638
	Adults	22	26.8	589.0		
	Total	55				
Respiratory	Child	33	27.5	906.0	345.0	0.756
	Adults	22	28.8	634.0		
	Total	55				
Digest	Child	33	24.9	822.0	261.0	0.070
	Adults	22	32.6	718.0		
	Total	55				
Social	Child	33	27.9	921.0	360.0	0.959
	Adults	22	28.1	619.0		
	Total	55				

3.2 | Percentage of death cases due to CF disease in families and their geographic distribution

The study showed that 21% of families (12/58) had death cases related to CF disease; 58.3% (7/12) from the Hebron district, two from Ramallah (16.7%), two from Jenin (16.7%), and one from Nablus (8.3%), as described in Table 5.

3.3 | Comparison between QoL domain scores between our CF participants and normative CF international data

All parameters for participants from West Bank, Palestine appeared to be noticeably lower than those reported in other countries[7,8] (Figure 1).

There was a high difference between our CF QoL domain scores and the US international data scoring,[9] with the scores of our participants being less than 50% in all domains for all participant groups, which indicated the severe status of our participants compared to the data for the US sample.

QoL assessment in participants with CF by means of the CFQ was carried out at the State University at Campinas School of Medical Sciences, Campinas, Brazil.[7] This study indicated that our participants with CF disease had poor QoL compared with the participants followed at the Brazilian center, as described.

4 | DISCUSSION

We described the clinical characteristics and outcomes of the 77 participants in the CBH who visited the pediatric pulmonology clinic. Our data indicate that the participants with CF followed under this study had poor QoL. There were two death cases; 1 month after we finished the study, a boy died at 16 years of age, and a girl did not complete the 7 months of her life. Both patients died from

respiratory complications. Our sample was the population sample for CF disease, as we included all the available CF participants in West Bank.

Regarding participants' characteristics, the mean age at diagnosis was 4.2 years of age, ranging from <1 to 30 years, as in other studies. As already known, late diagnosis results in a worse prognosis of CF lung disease. Fortunately, improvements in age at diagnosis have been observed in recent years.[10] In this study, CF participants were divided into age groups according to the original protocol of the CFQ-R.[11] The formal inclusion of CFQ-R questionnaires as a clinical outcome was essential, as it detected the impact of treatment over QoL from the perspective of participants and their families. It allowed the identification of baseline reference values for QoL in different age groups and the progression of the QoL domain scores over time. The present study was an attempt to rescue the outcomes of distinct age groups. Thus, it would be possible to make comparisons between these groups, to evaluate individual participants in relation to the baseline characteristics of their respective age groups, and to plan future strategies.

TABLE 3 Comparison between CFQ parent$_{6-13}$ scores and CFQ$_{6-13}$ child scores with level of significance in physical and treatment domains.

		N	Mean	Standard deviation	Standard error mean	Sig. (two-tailed) $p < 0.05$
Physical	Parents	33	37.6	14.3	2.5	0.010*
	Child	33	26.4	19.6	3.4	
Emotion	Parents	33	45.9	22.1	3.8	0.612
	Child	33	48.2	15.1	2.6	
Eat	Parents	33	51.0	28.5	5.0	0.478
	Child	33	46.1	26.9	4.7	
Body	Parents	33	14.8	16.4	2.8	0.936
	Child	33	14.5	17.7	3.1	
Treatment	Parents	33	41.4	8.5	1.5	0.000*
	Child	33	17.5	13.6	2.4	
Respiratory	Parents	33	31.7	14.4	2.5	0.328
	Child	33	27.5	19.9	3.5	
Digest	Parents	33	30.6	22.1	3.8	0.227
	Child	33	23.2	27.0	4.7	

TABLE 5 CF Death cases geographic distribution.

Regions	Number of participants (families no.)/region	Number (%) of deaths
Jerusalem	1	0%
Bethlehem	4	0%
Hebron	38	14 (73.7%) from 7 families
Ramallah	4	2 (10.5%) from 2 families
Nablus	4	1 (5.3%) from 1 family
Jenin	5	2 (10.5%) from 2 families
Qalqilia	1	0%
Tulkarem	1	0%
Total	58	19 from 12 families (21%)

TABLE 4 Health profile results, by group.

Health profile	Parameter	Group CFQ$_{14+}$ *n* (=22)	CFQ$_{6-13}$ *n* (=33)	CFQ$_{<6}$ *n* (=22)[a]	Total sample *N* (=77)	Mean of total sample
FEV$_1$	FEV$_1$ ≥ 70	85.2	90.3		10 (13%)	88
	70 > FEV$_1$ ≥ 40	68.5	55.3		14 (18.2%)	61.3
	FEV$_1$ < 40	36			2 (2.6%)	36
BMI	Minimum	14.00	9.60	13.3	*n* (=72)[b]	
	Maximum	23.12	16.70	20.9	*n* (=72)[b]	
	Mean value	18.32	14.42	16.1		15.9
Age at time of diagnosis (years)	Minimum	1	1	<1	*n* (=77)	
	Maximum	30	11	2	*n* (=77)	
	Mean value	9.68	2.48	1.14		4.2

Note: Values expressed as mean values.

Abbreviations: BMI, body mass index; CFQ, cystic fibrosis questionnaire; CFQ$_{<6}$: participants <6 years of age; CFQ$_{6-13}$, participants ≥6 and <14 years of age; CFQ$_{14+}$, participants ≥14 years of age; n, number of participants.

[a]Twenty-two participants in the CFQ$_{<6}$ group did not undergo spirometry test.

[b]Five participants less than 2 years cannot measured BMI and have weight/length absolute of percentile (2.9 kg/51 cm, 9.7 kg/79 cm, 9 kg/75 cm, 11 kg/88 cm, 2.65 kg/49 cm) were less than 5th%.

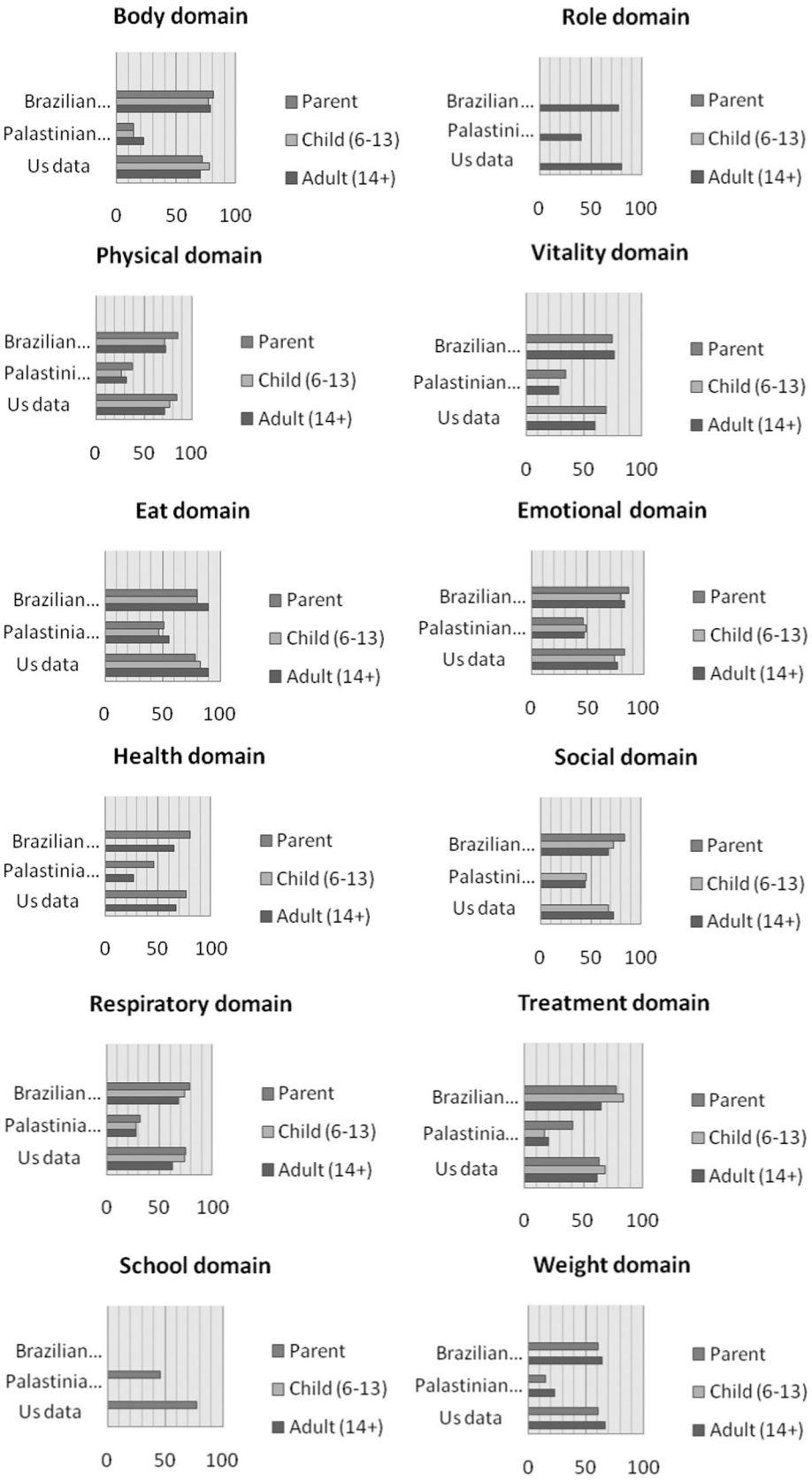

FIGURE 1 Data labels for Brazil, Palestine, and US.

Comparison of the mean scores between the CFQ_{14+} and CFQ_{6-13} groups revealed no statistically significant differences;. There is a significant statistical difference between adults and children in the eating domain, as adults care about the nature of their eating more than children. Additionally, the score for participants less than 6 years of age was much higher than both groups, as parents take care of their food needs.

Symptoms of anxiety and depression in children, adolescents, and adults correlate with low scores on instruments designed to quantify QoL and have been described as risk factors for poor treatment compliance, increased morbidity, and increased health care use in chronic diseases in many studies.[12]

Regarding the questionnaires completed by the parents, we observed good agreement between the mean scores on the CFQ $parents_{6-13}$ and the information obtained by means of the questionnaires applied to the participants. As determined in the score results of the parents, the lowest scores were observed for the weight and body domains, suggesting parental dissatisfaction with the gastrointestinal functioning of their children. Statistical significance was found in the physical and treatment domains only.

Some studies have shown differences between parent and child reporting's of QoL, especially regarding the physical and emotional aspects.[13] This is probably due to the high level of emotional distress experienced by parents. The health problems of children with CF limit their participation in physical activities, as well as in school and family activities, causing caregivers anxiety and depression.[14] In addition, concerns about life expectancy, together with the expensive and demanding treatment regimens, contribute to the onset of depressive symptoms in caregivers, directly affecting the activities of daily living and treatment compliance.[14]

Illness severity as measured by FEV_1 ranged from 33% to 111% predicted with a mean value of 69.6%; this was done for only 26 participants, as the rest of the participants (29) were unable to pay for the high test price (~$200) due to poverty, and 22 participants were less than 6 years of age and could not do the test. For child and adult groups, there was no correlation between the QoL domain score and the FEV_1level measured. As shown in many studies, FEV_1 percent predicted may not improve substantially in participants whose lung function is >75% predicted; however, these participants may report improvement in respiratory symptoms after treatment with antibiotics. Thus, in some cases, the participants' reported outcomes may be more sensitive to changes in symptoms than traditional pulmonary function indexes.[15–18]

The assessment of QoL in individuals with CF is important because it reveals the participants' perception of what it is like to live with a fatal chronic illness and can improve treatment compliance.[7,19] In addition, it provides information for economic planning and makes it possible to determine the impact of new treatments, as shown in a recent study.[20]

With the geographic distribution of participants results as described for 58 families, Hebron was the district with most participants suffering from CF disease (50 participants), and many families had more than one participant (eight families had two participants). The reason for this is the high rate of marriage between relatives; as indicated in many studies, the rate of consanguineous marriages in Palestine was found to be 45% in 2004.[21] The family with the highest number of participants (four participants) was from the Nablus district.

In our study, 21% of families (12/58) reported death cases related to CF disease. One family from Hebron had four CF death cases; this high rate of diagnosed CF death cases was related to poor management strategies, therapies, and lack of earlier diagnostic procedures.

In our study, we compared our CF participants' QoL scores to normative CF international data, the US sample and the Brazilian sample, which were similar to our research study.[7,8] As we observed, Palestinian CF participants have poor QoL, which is related to our treatment regimen for CF being highly complex and time-consuming, requiring 2–4 h of treatment every day. The treatment regimen includes multiple inhaled therapies, airway clearance two times per day, oral medications, and boosting calories to 110%–200% of the recommended daily allowance. The challenges of adhering to this regimen include the time required, the complexity of using and cleaning the equipment, and its considerable cost. In addition, participants experience frequent pulmonary exacerbations, hospitalizations, and segregation from peers due to multi-resistant bacteria. High rates of depression and anxiety have also been reported by both participants and caregivers.

These results should be interpreted in light of some limitations. First, as a result of the lack of specialized centers that monitor and deal with CF participants and clarify many clinical aspects and outcomes of CF participants, we had difficulty in obtaining the full number of participants in the West Bank and Gaza strip. As well, the sample size, although reasonable for this rare disease, was relatively small for the analyses that were conducted. Thus, this study was likely underpowered to detect some of the predictable relationships.

Second, the lack of a comparison group treated with new medications such as dornase-alpha or tobramycin inhalation solution is a limitation that impairs a complete investigation about the benefits of these new drugs in the West Bank. In view of the current recommendations regarding the use of dornase alpha or tobramycin, it was decided to base our data on the literature studies due to the high-cost price for these medications and lack of financial and human resources to apply these drugs in the West Bank. Nevertheless, in our study, subjects served as their own controls, as clinical information for our CF participants were compared to normative CF international data.

5 | CONCLUSION

Overall, our participants had significant disease morbidity and mortality despite the routine therapies they receive. There was a significant decrease in the CFQ-R scores of the respiratory symptoms, emotional functioning, body image, and treatment burden domains in the subgroup of participants with CF. The QoL for

participants with CF was poor compared with the international standards. The medications used, including hypertonic saline and gentamicin, are not the only therapies around the world, and participants and their families demand better treatment to improve their QoL, health status, and longevity.

AUTHOR CONTRIBUTIONS

Samya Salah: Conceptualization (equal); writing—original draft (equal). **Nisreen Rumman**: Project administration (equal); supervision (lead). **Amal Nassar**: Data curation (equal). **Maher Khdour**: Methodology (equal). **Hussein Hallak**: Conceptualization (equal); supervision (lead); writing—review & editing (equal).

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

Data available on request from the authors.

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