Effect of Nursing Intervention on Parents Awareness Regarding Health Problems of their Children Suffering from Cystic Fibrosis

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Abstract

Background: Cystic fibrosis (CF) is an inherited disorder that causes severe damage to the lungs, digestive system and other organs in the body. **Aim:** The aim was to evaluate the effect of nursing intervention on parents' awareness regarding health problems of their children suffering from cystic fibrosis. **Research design:** A quasi-experimental study design was used (pre/post). **Sampling:** A purposive sample involved 50 parents (mothers or fathers) of children suffering from cystic fibrosis **Settings:** The outpatient department (OPDs) at Children's Hospital and specialized OPD affiliated to Ain Shams University Hospitals. **Tools:** Data collected through using Tool I: an interviewing questionnaire included parents and children characteristics, parents' knowledge regarding cystic fibrosis, health problems, cystic fibrosis compliance management and reasons for poor cystic fibrosis compliance managements. Tool II: Parents self- reported practice included care of children with CF in relation nebulizer sessions, hand washing, enema care and assessing skin elasticity during dehydration. **Results:** After implementing the nursing intervention most of the studied parents had satisfactory level of knowledge regarding CF disease and satisfactory level of reported practice regarding care of their children. **Conclusion:** Nursing intervention had a positive effect on parents having children with CF in relation to improving their knowledge and reported practice, there was negative correlation between parents' total knowledge and total reported practice pre the intervention compared to positive correlation post the intervention. **Recommendation:** Further studies are needed to assess factors affecting parents' compliance with CF management for their children.

Keywords: Parents, Awareness, Children, Cystic Fibrosis, Health Problems, Nursing Intervention.

Introduction

Cystic fibrosis (CF) is a monogenic disease inherited in an autosomal recessive pattern caused by faulty genes called "cystic fibrosis transmembrane conductance regulator" on a person's chromosome. This leads to disrupted ion transport across epithelial cell membranes, culminating in a complex array of symptoms, including thickened and viscous secretions in the bronchi, intestines, pancreas, biliary tract, reproductive system, and most children have more sodium and chloride in their sweat (**Valdez et al., 2021).**

The most common fatal genetic disorder among Caucasians is CF, afflicting one in 3000 live births, with an estimated 150,000 diagnosed cases globally **(Orenti et al., 2021; Cystic Fibrosis Foundation, 2021).** Moreover, cystic fibrosis is prevalent among the Arab population, particularly in Jordan, with an estimated incidence of 1 case per 2560 live births, while the occurrence of CF has reduced in many countries, and survival rates have increased, with an estimated average lifespan of around 50 years for affected children today **(Scotet et al., 2020).**

Early detection remains paramount in managing CF, with newborn screening programs implemented worldwide to facilitate timely intervention and prevent complications (**Coverstone and Ferkol, 2021**). Nutritional assessment and management are critical components of long-term CF care, particularly challenging in Egyptian CF pediatric patients grappling with failure to thrive and the necessity of meeting their nutritional needs in terms of calories, macronutrients, and micronutrients (**El-Koofy et al., 2020**).

Valdez et al., (2021) mentioned that, "further delineate the clinical manifestations of CF, which can be categorized based on age groups or affected organ systems. Neonates and young infants may present with symptoms like meconium ileus, prolonged jaundice, malnutrition, and frequent vomiting, while older

children may experience chronic respiratory issues, gastrointestinal complications, and bone disease due to vitamin D deficiency. Additionally, insufficient public awareness perpetuates children with CF feel bad about themselves and excluded from society, adversely affecting the self-esteem and standard of life".

Knowledge and comprehension of the disease develop gradually over time through learning experiences, with families and peers affected by cystic fibrosis playing significant roles in this process. Family involvement in educating about dietary requirements and medication is crucial for understanding and adherence, which in turn influences confidence in managing the disease. Focusing on such support during preadolescence may better prepare children to handle the challenges of cystic fibrosis care during adolescence (**Cave and Milnes, 2020).**

The parents should be supported and informed when a child is diagnosed with CF. In order to prevent unnecessary discomfort and complications to their dependents, parents need to be aware of infection control measures early **(Villanueva et al., 2017).**

As stated by **Dogru et al., (2020),** the experiences of parents in managing CF are often overlooked in research. A significant aspect highlighted in their study is the limited knowledge among healthcare providers regarding the CF, how to support both children and their parents, and how to diagnose it. Family involvement in physical activity plays a crucial role in fostering long-term behaviors by enhancing enjoyment and adherence.

Parents contribute significantly to managing CF, including implementing mealtime strategies to promote healthy eating habits. Encouraging children with CF to engage in club activities integrates exercise into their daily routines (**Gruet et al., 2022**). Pancreatic Enzyme Replacement Therapy (PERT) is essential for over 80% of CF pediatric patients due to exocrine pancreatic insufficiency, necessitating precise dosing and timing to address nutrient mal absorption (**Ng et al., 2021**).

Significance of the study

Cystic Fibrosis (CF) is a multisystem, terminal disease with an autosomal recessive inheritance pattern **(Rasheed et al., 2019).** It has been referred to as the most prevalent genetic lethal disorder among Caucasians (**Abu-Zahra et al., 2019).**

In the Middle East the incidence of CF ranges from 1:2000 to 1:5800 live births **(Banjar & Angyalosi, 2015).** Caring for a child with CF requires a constant commitment from those responsible, and the burden of treatment is a major concern **(McCray et al., 2023).**

Through earlier diagnosis and palliative treatment, collaborative efforts have extended the lifespan of children with CF from just a few months to 40 years old **(Naehrig et al., 2017).**

Family involvement is crucial in supporting CF pediatric patients, with caregivers playing essential roles in providing information about diet and medication adherence, thereby empowering children to manage their condition effectively (**Dogru et al., 2020**). Furthermore, parental engagement in promoting physical activity and ensuring proper administration of pancreatic enzyme replacement therapy (PERT) is vital for optimizing health outcomes in CF patients (**Gruet et al., 2022**; **Ng et al., 2021**).

Aim of the Study

The aim of this study was to evaluate the effect of nursing intervention on parents' awareness regarding health problems of their children suffering from cystic fibrosis through:

1- Assessing parents' awareness regarding health problems of their children suffering from cystic fibrosis.

2- Designing and implementing a nursing intervention for the parents regarding their awareness of health problems among their children with cystic fibrosis.

3- Evaluating the effect of nursing intervention on parents' awareness regarding health problems of their children suffering from cystic fibrosis.

Operational definition:

Parents' awareness: it means knowledge, reported practice toward cystic fibrosis.

Research hypothesis

H1. Parents' awareness regarding health problems of their Children with CF will be improved after implementation of the nursing intervention through improving their

knowledge regarding CF and their reported practice regarding care of their children with CF.

H2. There will be positive correlations between Parent's knowledge and their reported practice regarding care of their children with CF pre and post nursing intervention.

Subjects and Methods

Research design:

A quasi-experimental design (pre/post).

Settings:

The outpatient department (OPDs) at Children's Hospital and specialized OPD affiliated to Ain Shams University Hospitals.

Subject:

A purposive sample involved 50 Parents (mothers or fathers) of children with cystic fibrosis. The sample size was estimated through using this equation:

$$N = \frac{N \times p\,(1-p)}{\{N-1\times (d^2 \div z^2)\} + p\,(1-P)}$$

N x p(1-p)	= [60*(0.5*(1-0.5))]
N-1	= (60-1)
d^2/z^2	=0.0025 / 2.8561
p(1-p)	=0.5*(1-0.5)
N	= 49.3 = 50

N = The community size / Month
Z = The class standard corresponding to level of significance = 0.95 and 1.96
d = The rate of error = 0.05
p = The ratio provides a neutral property = 0.50

The inclusion criteria are:

- Children suffering from cystic fibrosis at the previously mentioned settings at the time of the data collection, regardless of their age and gender.
- Parents (mothers or fathers) of children suffering from cystic fibrosis at the previously mentioned settings at the time of the data collection, regardless of their age, level of education and job.

Exclusive criteria:

Children suffering from other respiratory diseases.

Tools of data collection:

Data collected through two tools as follows:

Tool I: An Interviewing Questionnaire:

It designed by researchers according to the current related literatures. It was written in an Arabic. It contained 18 questions in the form of closed-open ended and Multiple-Choice Questions (MCQs). It involved three parts:

Part 1: Characteristics of the study subjects:

a- Parents Characteristics included age, level of education, job, residence, consanguinity and presence of other children with CF in family. It contains four questions in the form of MCQs and two questions in the form of closed ended form.

b- Children Characteristics included age, sex, level of education, child's birth order, onset of diagnosis, previous hospitalization and duration of hospital admission by days. It contains five questions in the form of MCQs and one question in the form of closed ended question.

Part 2: Parents' knowledge regarding the following:

a- Cystic fibrosis (Pre/post nursing intervention): It was designed based on the relevant literatures **(Goetz & Clement., 2019 and López et al., 2021)** to assess the parents' knowledge regarding definition of CF, signs & symptoms, etiology, plan of treatment, complications, importance of follow up, parents' role regarding care for their children with CF at home. It included 7 MCQs.

b- Health problems including (respiratory, nutritional, digestive, gastrointestinal, reproductive and liability to infection. It included 6 MCQs.

c- Cystic Fibrosis compliance questionnaire (CFCQ) (Pre/post nursing intervention): It was adopted from **(Llorente et al., 2008)**, to assess the parents' knowledge regarding items of CF compliance management for their children which included physiotherapy, respiratory medications, digestive

medications and nutritional supplements, each item included 5 questions. The total questions 20 (MCQs & closed-open ended).

Scoring system:

Researchers prepared the model answer, correct responses were checked through the following:

- One degree for correct answer meanwhile, zero for incorrect or don't know.
- The total score was 32. The parents' responses summed up, then converted into a percentage to be classified into two categories of knowledge:
- **Satisfactory ≥ 60% (≥ 19 scores).**
- **Unsatisfactory <60% (<19 scores).**

Part 3: Reasons for studied parents' poor compliance with managements of CF for their children. It included 4 MCQs.

Tool II: Parents self- reported practice:

It is adapted from **Bowden and Greenberg (2015)** to assess studied parents' reported practice regarding care of their children in relation to four items (nebulizer sessions, technique of hand washing, enema care and assessment of skin elasticity during dehydration. Each item included some steps, one degree for done meanwhile, zero for not done.

The total score was 20, Parents' responses summed up, then converted into a percentage to be classified into two categories of reported practices:

- Satisfactory ≥ 60% (≥ 12 scores).

- Unsatisfactory <60% (<12 scores).

The interviewing questionnaire filled in by the parents and took from 20 to 30 minutes to fill in.

Content validity:

The study tools reviewed by a panel of three experts professors of Pediatric Nursing departments from Faculty of Nursing, Ain Shams University. They tested the face and content validity. All experts reviewed tools for clarity, relevancy, comprehensiveness, appropriateness, and applicability and according to their opinions, modifications through addition or omission for some items according to experts' recommendations.

Reliability:

Testing reliability of the study tools was done by using Cronbach's alpha test. It was (0.779 and 0.877) for the interviewing questionnaire (knowledge and reported practice).

Ethical considerations:

- The research ethical approval was granted from the ethical research committee of the Faculty of Nursing, Ain Shams University.
- The researchers clarified the aim and objectives of the study to the children and their parents
- Oral and written consents were taken from children and their parents before their participation.
- The researchers assured anonymity and data confidentiality of parents and their children.
- Parents informed that they have the right for withdrawing from the study without clarifying any reasons.

Pilot study:

The pilot study was carried out on 10% from the study sample (5 parents, fathers or mothers) having children with CF to evaluate the applicability, clarity and time needed for filling in each tool. In addition, it helps in overcoming problems and barriers that may face the researchers during data collection. The simple modifications were done through rephrasing some items or questions based on the pilot study results. The five children in the pilot study were excluded from the study subjects.

Fieldwork:

The actual fieldwork carried out within four months from October 2023 to January 2024.

A. Assessment phase:

It lasted for 4 weeks. Researchers at this phase were available two days/week (Saturday, Monday) in the morning shift (9 am - 2pm) in study settings. The total number of parents interviewed per week ranged from 5 to 7 parents. The aim of study and steps were clarified for the studied parents. The studied parents filled the study tools through average time for each tool around 10 -15 minutes.

Parents having children with CF interviewed individually for one-hour duration to assess their

knowledge, cystic fibrosis compliance questionnaire through using the study tools as pre assessment prior to implementation nursing intervention.

B. Planning phase:

It lasted for 2 weeks. The researchers designing the nursing intervention sessions through assessing the actual needs of studied parents having children with CF in order to improve their knowledge and reported practice, through using a guiding booklet prepared by the researchers. It designed in simple and clear Arabic language based on the needs assessment of the parents. It is used to provide parents and their children with CF by the important knowledge about the disease as regards; definition of CF; signs & symptoms, etiology, plan of treatment complications, importance of follow up, their role regarding care for their children at home, health problems, cystic fibrosis compliance management and reported practice regarding care of children with CF.

C. Implementation phase:

It lasted for 6 weeks. The researchers started the sessions at outpatient department (OPDs) at New Children's Hospital and specialized OPD affiliated to Ain Shams University Hospitals. The parents classified into 10 groups; each group 5 (father & mother or father or mother) and took 6 sessions (4 theoretical sessions and 2 practical) each session lasted for one hour equal 6 hours for each group and the total hours equal 60 hours for 10 groups through 6 weeks (10 hours each week distributed through two days).

The first session included an introduction and the aim of nursing intervention sessions, performing the pre-test by using the study tools and distributing the booklet.

The second session included the definition, signs & symptoms, etiology, plan of treatment, complications, importance of follow up parents' role regarding care for their children with CF at home and health problems.

The third session included the explanation of cystic fibrosis compliance questionnaire containing the four items of compliance management for CF including physiotherapy, respiratory medications, digestive medications and nutritional supplements. In addition, discussion about reasons for poor compliance with managements for CF.

The fourth session included demonstration and redemonstration of the nebulizer sessions for their children with CF and technique of hand washing for infection control.

The fifth session included demonstration and redemonstration of the enema in case of constipation and assessment of skin elasticity in case of dehydration.

The sixth session the researchers thanked all parents and their children and performing reassessment for their knowledge level, cystic fibrosis compliance post assessment (post the nursing intervention implementation).

At the beginning of each session, the researchers performing feedback for previous session and explained objectives of this session and at the end of every session, answering all parents' questions.

Using different teaching strategies as modified lectures, role-play, small group discussion, and demonstration. Suitable and attractive teaching aids as booklet, colored pictures, posters, videos, doll, and real objects were used by the researchers.

D. Evaluation phase:

At the completion of the sessions, the post-test done immediately for each parent by using the same study tools to evaluate the effect nursing intervention through comparing the results of the pre and post-tests (Post nursing intervention implementation).

Statistical design:

- Study data collected, organized, coded and then analyzed by using the software version 23 of Statistical Package of Social Science (SPSS). Quantitative data were expressed as mean± standard deviation (SD). Qualitative data were expressed as frequency and percentage. Data were presented in tables and figures. The Comparison between groups with qualitative data was done by using Chi-square test. Pearson's correlation coefficient (r) test was used to assess the degree of association between two sets of variables.

Results

Table 1 clarified that the mean age of the studied parents was 33.36±6.34 and 39.88±7.58 years for mothers and fathers respectively, as well 40% of mothers and 44 % of fathers have high level of education in addition to 56% & 40% of studied parents have positive consanguinity and another child with cystic fibrosis in the family respectively.

Table 2 illustrated that 60% of studied children aged<3 years at the mean age 3.10±0.59, and 60% of them were the second child & their onset of diagnosis was < 1 year. Also, all of them had previous hospitalization at the mean 8.50±1.62 for duration of hospital admission by days.

Table 3 demonstrated that 70%, 66% & 80% of the studied parents had unsatisfactory knowledge regarding definition & importance of follow up, signs & symptoms, complications & Parent's role regarding care of their children with C.F at home pre-nursing intervention respectively; meanwhile 100%, 90% and 94% of them had satisfactory knowledge regarding CF post implementation of the nursing intervention with statistically significance difference.

Table 4 reflected that 70% %, 80% & 90% of the studied parents had unsatisfactory knowledge regarding digestive, liable to infection & reproductive health problems of CF pre-nursing intervention respectively while 100%, 90% & 94% of them had satisfactory knowledge regarding health problems of CF post implementation of the nursing intervention with statistically significance difference.

Table 5 revealed that 70 % &78% of the studied parents had unsatisfactory knowledge regarding cystic fibrosis compliance including Physiotherapy & nutritional supplements CF pre-nursing intervention respectively compared to 80% & 90% of them had satisfactory knowledge regarding physiotherapy & digestive medication post implementation of the nursing intervention respectively with statistically significance difference.

Figure1 illustrated that there was statistically significance difference between total parent's knowledge regarding cystic fibrosis, health problems and cystic fibrosis compliance pre &post nursing intervention.

Table 6 showed that 54%, 70%, and 84% of the studied parents mentioned the reasons for their poor compliance for physiotherapy management of CF as there is not enough time, it interferes with the child social life and children transplantation respectively. Also, 40% &100% of them mentioned that they forgot the respiratory medication & digestive medication for their children respectively. Moreover, 68% of the studied parents mentioned their children don't like the taste or texture of nutritional supplements. The number is not exclusive for the mentioned reasons for poor compliance for CF management.

Table 7 illustrated that 70% &100% of parents had unsatisfactory reported practice regarding hand washing and assessing skin elasticity during dehydration pre-nursing intervention. Meanwhile, 90% of them had satisfactory reported practice regarding nebulizer post nursing intervention with statistically significance difference.

Figure2 illustrated that there was statistically significance difference between total parents' reported practice regarding care of their children with cystic fibrosis pre &post nursing intervention.

Table 8 revealed that, there was negative correlations between the total levels of studied parent knowledge and their total reported practice pre nursing intervention compared by positive correlations post nursing intervention ($r = 0.572$, at p-value <0.001 .

Table (1): Distribution of the Studied Parents according to their Characteristics (n=50).

Parents' characteristics	Mothers		Fathers	
	No	%	No	%
Age (Years)				
< 20	4	8	2	4
20- < 30	6	12	2	4
30- < 40	35	70	16	32
≥ 40	5	10	30	60
$\bar{X}$ ±SD	33.36±6.34		39.88±7.58	
Level of education				
Read and write	6	12	6	12
Primary education	4	8	6	12
Secondary education	20	40	16	32
High education	20	40	22	44
Job				
Work	22	44	100	100
Not work	28	56	0	0
Residence				
Urban	40	80	40	80
Rural	10	20	10	20
Consanguinity				
Yes:	28	56	28	56
Presence of another Child with cystic Fibrosis in the Family				
Yes:	20	40	20	40

Table (2): Distribution of the Studied Children according to their Characteristics (n=50).

Children' characteristics	No	%
Age (in years)		
<3	30	60
3-< 6	10	20
≥6	10	20
$\bar{X}$ ±SD	3.10±0.59	
Sex		
Male	28	56
Female	22	44
Level of education		
Nursery	30	60
Primary	20	40
Child birth order		
1st	10	20
2nd	30	60
3rd or more	10	20
Onset of diagnosis (year)		
< 1	30	60
1 - < 3	20	40
Previous hospitalization		
Yes	50	100
Duration of hospital admission (days)		
< 5	10	20
5 - < 10	25	50
10 - < 15	10	20
≥ 15	5	10
$\bar{X}$ ±SD	8.50±1.62	

Table (3): Distribution of the Studied Parents according to their Knowledge regarding Cystic Fibrosis Pre & Post Nursing Intervention (n=50).

Parent's Knowledge	Pre-Intervention		Post= Intervention		X2	p-value
	Satisfactory	Unsatisfactory	Satisfactory	Unsatisfactory		
	%	%	%	%		
Definition	30	70	86	14	29.926	<0.001**
Signs & Symptoms	34	66	100	0	46.314	<0.001**
Etiology	40	60	90	10	25.319	<0.001**
plan of treatment	50	50	94	6	21.875	<0.001**
Complications	20	80	80	20	33.640	<0.001**
importance of follow up	30	70	90	10	35.042	<0.001**
Parent's role regarding care of their children with C.F at home	20	80	84	16	38.502	<0.001**
Total	**32**	**68**	**88**	**12**	**30.375**	<0.001**

Using: Chi-square test
*p-value >0.05 is insignificant; *p-value <0.05 is significant; **p-value <0.001 is highly significant*

Table (4): Distribution of the Studied Parents according to their Knowledge regarding Health problems of Cystic Fibrosis Pre & Post Nursing Intervention (n=50)

Health problems	Pre- Intervention		Post –Intervention		X2	p-value
	Satisfactory	Unsatisfactory	Satisfactory	Unsatisfactory		
	%	%	%	%		
Respiratory	54	46	100	0	27.329	<0.001**
Nutritional	36	64	100	0	44.164	<0.001**
Digestive	30	70	100	0	50.813	<0.001**
Gastrointestinal (GIT)	60	40	100	0	22.562	<0.001**
Liable to Infection	20	80	90	10	46.707	<0.001**
Reproductive	10	90	94	6	67.348	<0.001**
Total	**34**	**66**	**96**	**4**	**39.560**	<0.001**

Using: Chi-square test
*p-value >0.05 is insignificant; *p-value <0.05 is significant; **p-value <0.001 is highly significant*

Table (5): Distribution of the Studied Parents according to their Knowledge regarding Cystic Fibrosis Compliance Questionnaire Pre & Post Nursing Intervention (n=50)

Items of Cystic Fibrosis compliance	Pre- Intervention		Post –Intervention		X2	p-value
	Satisfactory	Unsatisfactory	Satisfactory	Unsatisfactory		
	%	%	%	%		
Physiotherapy	30	70	80	20	23.273	<0.001**
Respiratory medication	54	46	78	22	5.392	0.020*
Digestive medication	60	40	90	10	10.453	<0.001**
Nutritional Supplements	22	78	74	26	25.040	<0.001**
Total	**40**	**60**	**80**	**20**	**15.042**	<0.001**

Using: Chi-square test
*p-value >0.05 is insignificant; *p-value <0.05 is significant; **p-value <0.001 is highly significant*

Figure (1): Percentage Distribution of the Studied Parents' Total knowledge regarding Cystic Fibrosis, Health Problems and Cystic Fibrosis Compliance Questionnaire(n=50)

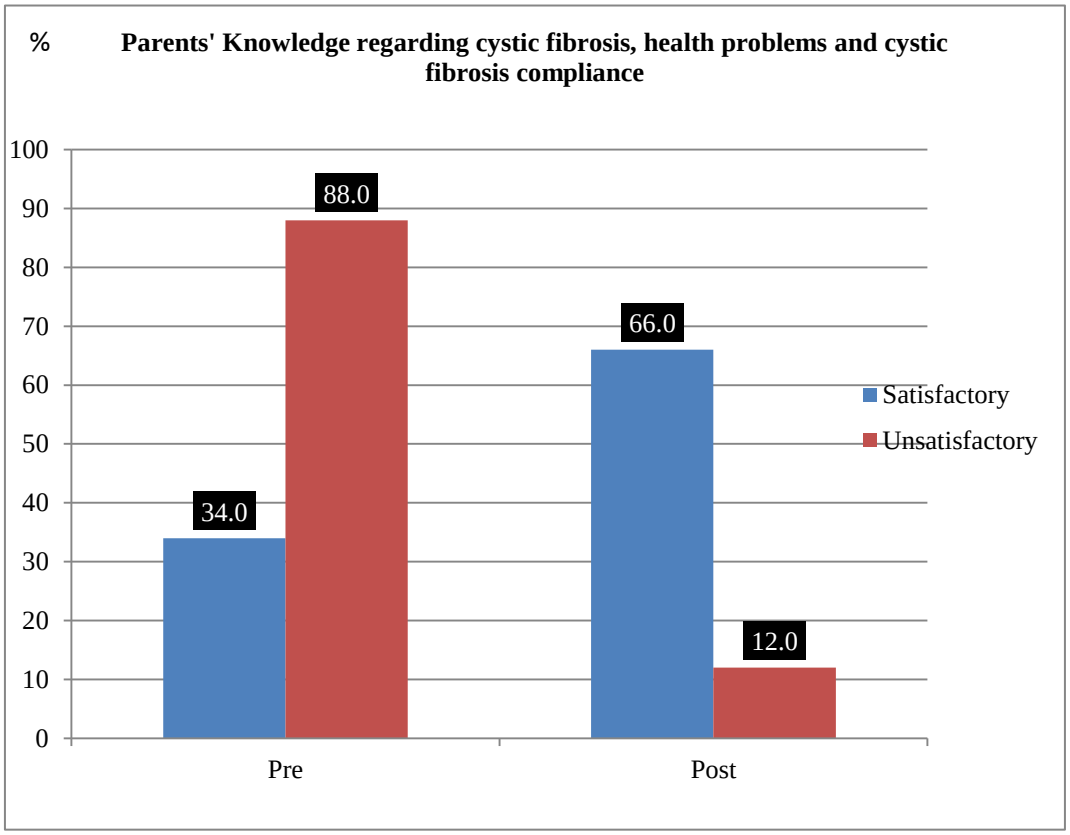

Table (6): Distribution of the Studied Parents according to their Reasons for Poor Compliance with Items of treatment for their children with Cystic Fibrosis (n=50).

Reasons for Poor Compliance	No.	%
Physiotherapy		
• Not enough time	27	54
• I don't think my child needs it, I feel well without treatment	5	10
• Exercise instead	18	36
• I believe that doing it my child don't feel better	5	10
• It interferes with my child social life	35	70
• I and my child simply forget it	13	26
• Transplantation	42	84
Respiratory medication		
• I don't think my child need it	10	20
• Not enough time	30	60
• Only when my child feels worse	17	34
• It interferes with my child social life	19	38
• I simply forget it	20	40
Digestive medication		
• I and my child simply forget it	50	100
Nutritional supplements		
• I don't think my child need it	19	38
• My child doesn't like the taste or texture	34	68
• I and my child simply forget it	28	56
• I and my child don't believe in it	19	38

**Number is not exclusive*

Table (7): Distribution of the Studied Parents according to their Reported Practice regarding Care of their Children with Cystic Fibrosis. (n=50)

Items of Reported Practice	Pre- Intervention		Post- Intervention		X2	p-value
	Satisfactory	unsatisfactory	Satisfactory	unsatisfactory		
	%	%	%	%		
Nebulizer sessions	34	66	90	10	30.942	<0.001**
Hand Washing	30	70	84	16	25.454	<0.001**
Enema care	60	40	90	10	10.453	<0.001**
Assessing skin elasticity during dehydration	0	100	82	18	66.143	<0.001**
Total	**30**	**70**	**86**	**14**	**29.926**	<0.001**

Using: Chi-square test
*p-value >0.05 is insignificant; *p-value <0.05 is significant; **p-value <0.001 is highly significant*

Figure (2): Percentage Distribution of the Studied Parents according to their Total Reported Practice regarding Care for their Children with Cystic Fibrosis. (n=50)

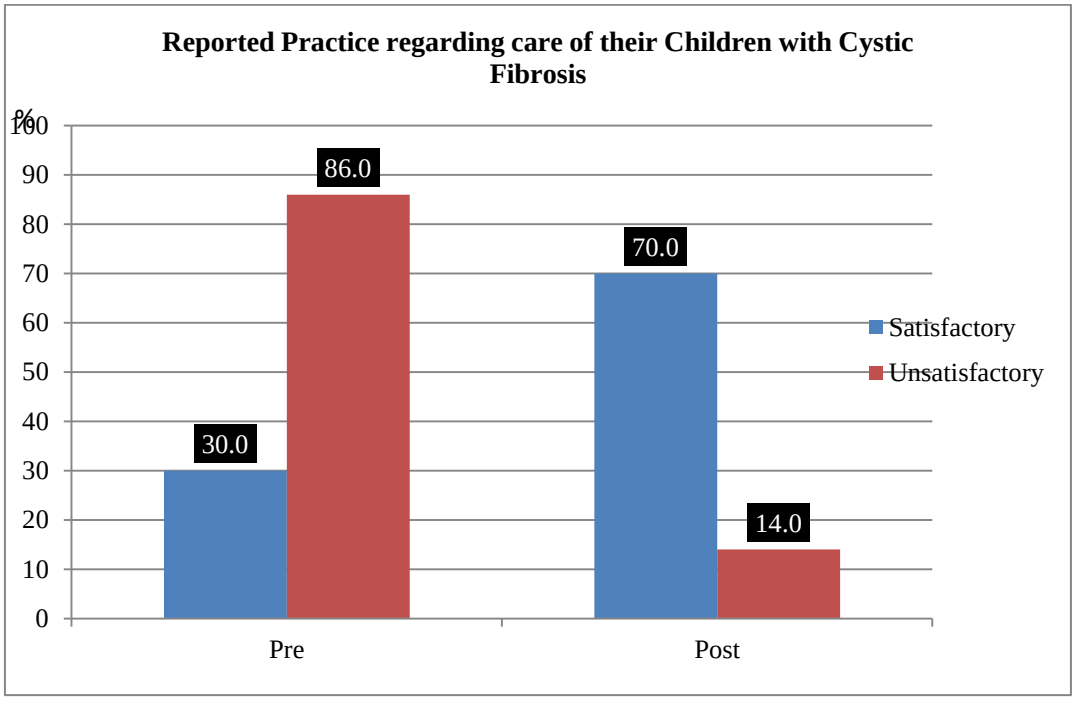

Table (8): Correlation between the studied parents' total knowledge and total reported practice regarding cystic fibrosis. (n=50)

Correlation(r)		Total Knowledge			
		Pre-intervention		Post- intervention	
		r-value	p-value	r-value	p-value
Total reported practice	Pre- intervention	0.186	0.672		
	Post- intervention			0.572	<0.001**

r-Pearson Correlation Coefficient;
*p-value >0.05 is insignificant; *p-value <0.05 is significant; **p-value <0.001 is highly significant*

Discussion

Children living with CF are required to manage disease progression through daily physiotherapy, oral medications, nutritional supplements and increased caloric intake **(Richards et al., 2022).** Parents should communicate closely with healthcare professionals especially at home Dykowska et al., (2023).

Regarding parents' characteristics **(table 1),** the mean age of the mothers was 33.36±6.34 years and 39.88±7.58 years for fathers. These findings were parallel with Continisio et al., (2020), who reported that, mean age of studied parents was 38.87 years.

Concerning to parents' Consanguinity, the current study proved that, two fifths of the studied parents have positive Consanguinity, this finding was completely consistent with Khatami et al., (2010), who reported in a similar study that, "Consanguinity of parents was discovered in two fifths of studied patients." This similarity may be because, both studies are conducted in the middle east in which there are no laws to prevent consanguineous marriage.

As regards the parents' educational level & employment, the findings of the current study proved that two fifths of them are highly educated & employed. These findings are contradicted with Continisio et al., (2020), who reported that "less than fifth of the studied parents were highly educated & about half of them were employed." These differences may be due to difference of study setting & cultures of the studied parents.

Concerning to children's characteristics **(table 2),** the mean age of them was 3.10±0.59 years and more than half of them were boys. These findings were parallel with Vahedi et al., (2016), who proved that, "the mean age of the studied children was 2.58 years & more than half of them were boys." While These results were contradicted with findings of Goodfellow et al., (2015), who mentioned that, "the age range of studied children was 10.1 years while around 3 fifths of them were females. "This difference may be due to the difference in aim & settings of studies.

In relation to the studied children birth order & age of the diagnosis with CF, the current study proved that, three fifths of them were the second child for the parents with age of the diagnosis of the disease at age of less than one year. These findings were parallel with Continisio et al., (2020), who found that, "about two fifths of the studied children were the second child for their parents with mean age at diagnosis of CF at 1.59 ± 1.35" this may be because of the early manifestations of cystic fibrosis which are affecting the neonates & infants that necessitate seeking medical consultation that permits the early diagnosis in infancy period.

In relation to the effect of nursing intervention on parents' knowledge **(table 3)**, the current findings noticed that, most of the studied parents had satisfactory knowledge regarding C.F with highly statistically significant difference that indicating positive effects of nursing intervention. From the researchers' point of view parent' knowledge gape is improved after nursing intervention.

These findings were highly supported with Reisinho & Gomes (2022), who emphasized in a similar study that, "nurses play a pivotal role in caring for adolescents with CF, so they need extended knowledge to recognize the different phases of the disease and understand how children with CF and their parents/family cope with this health-threatening situation. Nursing clinical practice indicators are a key element to this process, allowing targeted interventions that meet emergent needs".

In the same line, it was mentioned by ICN, (2017) that, "the nurse should consider the family as a group interacting according to its own rules and expectations, capable of fulfilling its wishes and needs to support and encourage patients and family members".

As regards the parents' knowledge regarding digestive, liability to infection & reproductive health problems of CF pre/ post nursing intervention **(Table 4),** there was significant improvement detected of parents' knowledge post implementation of the nursing intervention which noticed in the total score of satisfactory parents' knowledge about the all items of health problems.

These findings were parallel with Lonabaugha et al., (2018), who emphasized that "information is important to CF patients and families because knowledge and understanding have been tied to a variety of positive outcomes. Several studies have tied lower rates of adherence with decreased understanding of a provider's recommendations to the children & their parents." On the other hand, Dykowska et al., (2023), added in a similar study that, "CF is a high-maintenance disease that requires both the parents and the child to make everyday life compatible with the treatment and to take the necessary steps to prevent exacerbations of the disease and subsequent hospitalizations".

On investigating the studied parents' knowledge regarding compliance of the management plan including physiotherapy, nutritional supplements of children with CF pre/ post nursing intervention **(Table 5),** findings of the current study reflected that, most of the studied parents had satisfactory knowledge regarding the mentioned items post implementation of the nursing intervention compared with pre implementation with statistically significance difference. This may be due to low health awareness regarding long term management plan for daily handling of the child's related health problems which obviously improved after the nursing intervention.

In the same direction Goodfellow et al., (2015), stated that, "adherence in cystic fibrosis is an important research area and more research is needed for better understanding of family barriers to adherence for clinicians to provide appropriate intervention. Adherence to therapy is therefore essential to prevent worsening of the disease and future adherence interventions addressing parental beliefs about enzyme supplements and chest physiotherapy may improve child adherence to treatment. There is a larger scope to improve adherence if interventions are targeted to older children".

On the other hand, Hommel et al., (2019), emphasized that, "the challenges of adhering to the multicomponent treatment recommendations for CF necessitate understanding of patient adherence to each component, identification of patients who may have difficulty adhering to dietary interventions should result in better treatment-to patient matching, improved adherence promotion efforts, and cost savings." Additionally, Jones et al., (2020), mentioned that, poor adherence to all aspects of CF treatment has been associated with an increased number of infective pulmonary exacerbations and the need for intravenous (IV) antibiotic therapy

In relation to total studied parents' knowledge **(figure 1)** regarding cystic fibrosis, health problems and cystic fibrosis compliance pre & post nursing intervention. There was a clear improvement in the parents' total knowledge after application of nursing intervention compared to before intervention. This may be related to low health awareness & knowledge gap pre intervention which successfully overcome after intervention. These findings were parallel with Richards et al., (2022), who mentioned that "lessons related to CF can be learnt from observing and evaluating their implementation within the outpatient setting. The adherence protocols are considered appropriate and acceptable to both clinicians and consumers."

Concerning the studied parents' reasons for poor compliance to all items of treatment for their children with Cystic Fibrosis **(Table 6),** finding of the current study reflected that, the majority of the studied parents' mentioned reasons for their poor compliance for physiotherapy management of CF because they have no enough time or it interferes with the child social life and difficulties in children transplantation. Also, two fifths of them mentioned that they forgot the respiratory medication while all of them forget digestive medication for their children. Moreover, more than two thirds of the studied parents mentioned that their children don't like the taste or texture of nutritional supplements.

These findings were highly supported with Jones et al., (2020), who mentioned that "poor adherence to medication and health advice is a global problem and is particularly important in chronic diseases. The WHO estimates that around a third to half of medications prescribed are not taken as directed. In CF adherence rates to nebulized treatments are less than two fifths in adults and only two thirds in children. The adherence rate for pancreatic enzymes in children is only two fifths when measured electronically. Adherence to chest physiotherapy has been recorded at only less than one third."

In the same line Tiwari et al., (2022), added that, "in children with CF, the most common reason for not doing physiotherapy and exercises was lack of time. For vitamins and enzyme supplementations children with CF reported that they simply forget to take medications. Most children felt right about physiotherapy, vitamin, and enzyme supplementations. More than half of the participants felt that their exercise level was not enough. Most CF children were having positive perception regarding treatment adherence (86.66%). Treatment adherence is a common problem among CF children that needs regular reinforcement by healthcare team members on vital components of treatment.

Furthermore Jones et al., (2020), mentioned that ‘‘Suboptimal adherence to treatment attributed to the complexity of the treatment regimen and the large burden it therefore places upon the individual", and added "Factors leading to suboptimal adherence include poor levels of self-efficacy, poor understanding of the condition or treatment, and the presence of negative cognitions, and this can be overcome by providing education in several different ways’’.

In investigating studied parents according to their reported practice regarding care of their children with CF. **(Table 7),** finding of the current study revealed that, most parents had unsatisfactory reported practice regarding hand washing and assessing skin integrity during dehydration pre-nursing intervention. Meanwhile, majority of them had satisfactory reported practice regarding nebulizer post nursing intervention with statistically significance difference. From the researchers' point of view this result may be due to poor compliance with the treatment plan due to insufficient knowledge of the parents regarding its importance which already improved after application of the nursing intervention.

These findings were supported with (Jacob et al., (2020), who mentioned that," after adaptation period, CF parents described better treatment adherence which stemmed from their motivation to stay healthy for their children and to be able to care for them as long as possible". Furthermore, Al Adaileh et al., (2021), added that, "Parents of children with CF suffer from difficulties and challenges that limit their daily lives. Education is needed for health care providers, society, and parents about CF; and parents need support to improve the quality of life of their child."

In relation to the total studied parents' knowledge regarding cystic fibrosis, health problems and cystic fibrosis compliance management pre &post nursing intervention. There was a clear improvement after implementation of nursing intervention compared to before intervention that reflects the positive effect of nursing intervention of the current study.

In this regard Dykowska et al., (2023), mentioned that, "there is a need to raise community awareness about CF through education. These educational efforts need to focus on society and health care professionals in supporting parents with CF children. Pediatric nurses and social workers play a major role in guiding the community by making people aware of the financial and moral burdens resulting from the presence of a child with cystic fibrosis to provide financial and moral support to parents who have CF children. Pediatric nurses must be sensitive to the children and their families. It seems that the occurrence of CF in a child requiring long-term care and parental involvement in treatment is tantamount to the parent's classification of CF as a chronic disease."

Regarding correlations between the parents' total knowledge and their total reported practice **(Table 8),** current study revealed that, there was negative correlations between the parents' total knowledge and their total reported practice pre nursing intervention compared by positive correlations post nursing intervention. This may be due to the positive effect of the nursing intervention that result in encouragement of the

parent to follow the treatment plan after covering knowledge gap compared to pre intervention.

Conclusion

Nursing intervention had a positive effect on parents having children with CF disease in relation to improving their knowledge and reported practice. there was negative correlation between parent's total knowledge and total reported practice pre the intervention compared to positive correlation post the intervention.

Recommendations

The following recommendations were suggested based on current study results:

- Early detection of the complications among children with CF through regular follow up and evaluation for their health conditions.
- Performing educational program for parents having children newly diagnosed with CF to increase their awareness regarding care for their children in other study settings.
- Further studies needed to assess factors affecting parents' compliance with CF management for their children.

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