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Effect of Comprehensive Health Promotion Intervention on Quality of Life among Children with Wilson's disease

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Abstract

Background: Wilson's disease, which most frequently affects children or young adults and has a significant impact on children's quality of life, is the most frequent cause of improper excretion of copper from the body. The aim of the study was to evaluate the effect of comprehensive health promotion intervention on the quality of life among children with Wilson's disease. **Methods:** A quasi-experimental design was used. **Setting:** The research was carried out at the outpatient clinic of pediatrics at the National Liver Institute at Shebin Elkoom city. **Sample:** A purposive sample of 44 children with Wilson's disease was selected from the previously mentioned setting. **Tools:** three instruments were used for data **Collection:** a structured interview questionnaire, Precede- proceed model questionnaire, and the Pediatric Quality of Life Inventory (PedsQL) 4.0 Generic Core Scale. **Results:** the mean scores of all quality of life aspects (physical, emotional, social, and school functioning) of studied children had improved on the post and follow-up tests than on the pretest. As well, children with Wilson's disease who received comprehensive health promotion intervention based on the PRECEDE-PROCEED model had stronger predisposing factors, reinforcing and enabling factors on post and follow-up tests than on pretest. **Conclusion:** Comprehensive health promotion intervention was successful in improving the quality of life among children with Wilson's disease. **Recommendation:** This study recommended the integration of a model-based comprehensive health promotion intervention program, PRECEDE-PROCEED, into the outpatient clinic of pediatrics for children with WD and their families.

Keywords: Comprehensive health promotion intervention, Quality of life & Wilson's disease.

Introduction

Wilson's disease is an uncommon autosomal recessive condition where the body accumulates too much copper. It predominantly affects the liver and the brain's basal ganglia, although it can also impact other organ systems **(Hedera, 2019) & (Nagral, et al., 2019)**. The ATP7B gene located on chromosome 13 controls the protein transporter responsible for the secretion of additional copper into the bile and throughout the body. Wilson's disease results from one of several mutations in this gene. The trans-Golgi networks in the liver and brain include the protein transporter. The liver releases the majority of copper (95%). Excess copper collects in the liver and eventually spills into the bloodstream, damaging other organ systems **(Gerosa et al., 2019)**.

According to a recent study, the prevalence of WD in the United States, Europe, and Asia is between 1.5 and 3.5 per 100,000 people. WD affects both males and females in equal numbers, and it may be found in all ethnic groups and ethnicities **(Sandahl et al., 2020)**. Approximately one in every 90 people is heterozygous for the ATP7B illness gene. While WD is most commonly seen in teenagers and young adults, it can affect anyone at any age Within 6 months to 3 years of the onset of symptoms, WD is usually diagnosed. As opposed to those with hepatic consequences, people with WD who have neurologic issues have longer symptom duration before being diagnosed. The two most typical diagnoses for WD are hepatic (nausea, ascites, edema in the legs, jaundice, and itching) and neurological disorders (tremors, muscular weakness, and itchiness). **(Ferenci, & Ott, 2019)**

As WD patients frequently present with hepatic and neuropsychiatric symptoms, treatment should be multidisciplinary. In the last 50 years, no novel therapy alternatives for WD have been presented **(Balijepalli et al., 2021)**. Patients who are diagnosed early and treated appropriately have a life expectancy comparable to the general population. Furthermore, therapy not only extends life but also improves the

quality of life for these people (QOL) **(Balijepalli et al., 2021)**.

The long-term nature of the condition, the possibility of severe symptoms, the daily use of oral medication, frequent medical consultations, and the necessity for regular investigations all influence these patients' day-to-day physical and emotional functioning, as well as their social lives and interpersonal interactions. Analyzing their quality of life is one objective way to assess this influence (Qol) **(Bem et al., 2011)**. According to World Health Organization (WHO), a person's assessment of their place in life in relation to their goals, aspirations, standards, and worries in the context of their culture and value systems constitutes their quality of life. It expresses an individual's contentment and well-being in terms of his or her bodily, psychological, social, economic, and spiritual well-being **(Fouda et al., 2021).**

One of the most comprehensive structures for evaluating health and quality-of-life issues is the PRECEDE-PROCEED model. The PRECEDE-PROCEED planning approach is an educational intervention program that has been effectively utilized in examining QoL in patients with various conditions. The PRECEDE-PROCEED paradigm consists of eight phases or stages; it starts with the results and moves backward to the causes; once the cause is found, an intervention may be developed to enhance the overall well-being. The Clinical Implications Model provides a strong theoretical foundation that makes it easier to develop practical nursing-led therapies. According to this concept, nursing is the process of determining the needs of children, generating suitable nursing care plans, putting those plans into practice while continuously reviewing how they're going, and then measuring the quality of life or children **(Saulle et al., 2020)**.

According to the PRECEDE-PROCEED theory, comprehensive health promotion intervention is designed and developed to improve quality of life. Based on a comprehensive health promotion intervention, nurses investigate risk factors such as children's awareness of their illnesses and attitude toward living a healthy lifestyle (good nutrition, dental care, sleep, exercise, follow-up visits, and the impact of disease on children's self-actualization). Identifying the reinforcing and enabling factors that affect behavior change and, ultimately, quality of life **(Gorina et al., 2019)**. Despite the devastating nature of the disease, limited studies have been conducted to examine the life quality for patients who have been diagnosed with WD. Thus, this study aimed to determine the effect of comprehensive health promotion intervention on the quality of life among children with Wilson's disease.

Significance of the study

Wilson disease is a rare disorder that affects both sexes equally. The Wilson's disease can found in all races and ethnicity groups. The disorder affects people of all races and ethnicities. Although estimates vary, Wilson's disease is thought to affect one in every 30,000 to 40,000 persons worldwide. One in every 90 individuals may be a carrier of the illness gene. Although there have only been roughly 2,000-3,000 cases reported in the United States, other affected persons may have been misdiagnosed with other neurological, hepatic, or psychological diseases. Wilson's disease may impact up to 9,000 persons in the United States, according to one estimate **(Wallace, & Dooley, 2020)**.

Few kinds of research on WD from Lebanon, Egypt, Saudi Arabia, and Oman have been published in the Arab world. Viral hepatitis, specifically hepatitis C, is the most prevalent liver condition worldwide and in Egypt. **(Abdel Ghaffar, et al., 2011)**. In Egypt's Assiut Liver Center, 42 cases were confirmed, 10 were suspected WD, and 12 had other diagnoses. There are 27 males aged 6 to 35 years old among the 42 confirmed WD cases. Hepatic symptoms predominated in 26/42 patients, neuropsychiatric symptoms in 11/42 patients, and mixed symptoms in 5 cases **(Aboalam et al., 2022)**.

Definition of variables

Comprehensive health promotion intervention: "It is any effort that seeks to move individuals toward a level of better health, including conventional disease preventive initiatives" **(Barnfield et al., 2020).** While **in the current study** a comprehensive health promotion intervention was applied based on Precede- proceed model that focused on empowering of Predisposing, Reinforcing, and Enabling factors that enhance quality of life for children with Wilson disease.

Aim of the study

The study aimed to evaluate the effect of comprehensive health promotion intervention on the quality of life among children with Wilson's disease.

Objectives:

1. Increase children's awareness about Wilson's disease to achieve healthy life style
2. Improve children's attitude for positive coping with the disease for better quality of life.
3. Empower reinforcement and enabling factors of children to follow health promotion intervention for better quality of life.

Research Hypothesis

Children with Wilson's disease who will receive comprehensive health promotion intervention will have a better quality of life post-intervention than pre-intervention.

Methods

Research design

For this study, a quasi-experimental design was used (pre, post, and follow up tests).

Settings

This study was carried out in the outpatient pediatric clinic of the National Liver Institute Menoufia University in Shebin El koom. The pediatric outpatient clinic has two rooms. The first room was designated for children with metabolic liver disorders. Wilson's disease (WD) patients came to the clinic every Sunday between 9 a.m. and 12 p.m. The second area was designated for youngsters suffering from various juvenile liver disorders.

Subjects

A purposive sample of 44 children who were diagnosed with WD from the previously mentioned setting was involved in the study. The sample size was estimated using the G*power program (Heinrich Heine University Düsseldorf, Germany) based on the past review of the literature **(Svetel et al. 2011)** which reported that the total score of Quality of life among patients with Wilson disease was 71.1±24.8. With α error=5%, study power 90%, the sample size was calculated to be 44 patients.

Inclusion criteria

The following criteria were used to select children for the study:

1. Children who have been diagnosed with WD aged from 10 to 16 years old.
2. Free of other mental and chronic disorders to prevent additional stress on children.

Tools

To get the data, the researchers used three instruments. These tools involved:

Tool I: A structured interview questionnaire. It was created by the researcher after reviewing related literature **(Khalif et al., 2017)**. This instrument was split into two parts:

Part 1: Characteristics of studied children. It included questions regarding children's age, sex, and educational level.

Part 2: Medical data of the child with WD. IT included items about the time of diagnosis, regular check-ups, and medications used (D-penicillamine or zinc or both).

Tool II: Precede Model questionnaire (Predisposing, Enabling, and Reinforcing Questionnaires). It was created by researchers after a review of the literature and is based on the preceding model's educational and ecological approach **(Elkholy et al., 2022)**. **It was divided into three parts:**

Part 1: Predisposing Factors Assessment Sheet. It contained 42 questions. The reliability of predisposing factors assessment sheet was established by Cronbach's co-efficiency alpha test. It was r = 0.75. It included two subparts:

Subpart 1: Children's Knowledge about WD disease. It composed of 14 open end questions including the disease definition, reasons, clinical manifestations, treatment, diet, and disease intervention. The scoring system for children's knowledge was as follows; two scores were given for a correct answer, one score for an incomplete answer, and zero for an incorrect answer. The scoring levels were arranged as follows; less than 50% for poor knowledge, from 50% to less than 75% for average knowledge, and ≥ 75% for good children's knowledge.

Subpart 2: Children's attitudes regarding a healthy lifestyle that affect the quality of life. It is composed of 28 items about healthy food (8 questions), dental care (6 questions), sleep (4 items), exercises (5 items), follow-up (3 items), self-actualization (2 items) graded on a five-point scale in which items ranging from (5) strongly agree to (1) strongly disagree. The scoring system included the sum of item scores. The scoring levels were arranged as follows; less than 50% for a negative attitude, from 50% to less than 75% for a fair attitude, and ≥ 75% for a Positive Attitude.

Part 2: Reinforcement factors assessment Sheet. It included five questions about measures of support and encouragement of peers, family, and health care staff. Scores were assigned in the following way: A response of "Yes" to each question was given one score and response of "No" to questions received zero scores. The scoring levels were arranged as follows; less than 50% for weak, from 50% to less than 75% for moderate, and ≥ 75% for strong.

The reliability of the reinforcement factors assessment sheet was r = 0.80

Part 3: Enabling factors assessment Sheet. It included 11 items about complication avoidance (4 items), interpersonal relations (3 items), access to utilization of training resources (2 items), and stress management (2 items). Scores were given in the following way: A response of "Yes" to each question was given one score and response of "No" to questions received zero scores. The scoring levels were arranged as follows; less than 50% for weak, from 50% to less than 75% for moderate, and ≥ 75% for strong.

The reliability of enabling factors assessment sheet was r = 0.77

Tool III: Pediatric Quality of Life Inventory (PedsQL) 4.0 Generic Core Scale. It was adopted from **James, (1998)**, It included 23 items related to children's physical activities (8 items), feelings (5 items), social functioning (5 items), and school Functioning (5 items) rated on a five-point scale in which items ranging from 0 (Never) to 4 (Almost

always). Scoring system for each item was 0 =100, 1=75, 2= 50, 3= 25 and 4=0. Total scoring system: If the child obtained < 50%, the quality of life was poor. From 50% to less than 75%, the quality of life was fair, and ≥ 75% of the quality of life was good.

It was a valid scale designed to assess the quality of life for children with high test-retest reliability A Pearson at 0.88.

Validity:

To ensure validity, the three instruments were presented to a jury of five specialists, including two professors of pediatric nursing, two professors of medical surgical nursing, and one professor of Pediatric Gastroenterology and Hematology, who were asked to change any required content. The changes were made to ensure their relevance and completeness.

Pilot study

A pilot study was carried out on four children (10% of the population) to ensure that the study's instruments were clear, practicable, and appropriate. There have been no changes. As a result, they were included in the research.

Ethical considerations

The ethics committee approval on the subject of research was taken from the Faculty of Nursing, Menoufia University, Egypt. Also, written consent was obtained from children and their parents who participated in the study after a complete description of the purpose, nature, and confidentiality of the study.

Procedure

Data collection for this study lasted 9 months, from the beginning of April to the end of December 2021. Before data gathering, written consent to complete the study was obtained from the head of the pediatric outpatient clinic after submitting an official letter from the Dean of the Faculty of Nursing at Menoufia University describing the goal of the study and methods of data gathering. Comprehensive health promotion intervention was applied based on the eight steps of preceded proceeded model. The first four steps were preceded steps (assessment phases), and the last four steps proceeded steps (implementation and evaluation phases).

Assessment phase (precede steps) (2 months)

Each child was interviewed individually (for 25-35 minutes). The researchers completed instruments one and two based on the answers of the children with Wilson's disease. Also, a Pre-intervention assessment for children's quality of life was done by the researchers using instrument three (pretest).

Planning phase: (1 month)

The educational intervention components were developed following the model's four-phase assessment. A review of scientific resources was used to design educational objectives, contents, and materials.

The goal of the comprehensive health promotion intervention was to improve predisposing factors (knowledge - attitude), reinforcing factors, and enabling factors to improve QOL for children with WD, which was accomplished.

Implementation and evaluation (Proceed steps) (3 months)

Children received the educational intervention in 4 sessions (every Sunday) based on precede–proceed planning model. Each session lasted 30 minutes using a booklet, pamphlets, questions, and answers to help children better understand of content, in addition to face-to-face small group discussion.

The first session aimed to provide adequate and most important knowledge about Wilson's disease (Predisposing factors): Children received brief explanations with pictures about WD definition, causes, signs, symptoms, complications, treatment, prevention of complications, and how to manage it.

The second session aimed to influence attitudes toward a healthy lifestyle (Predisposing factors): Children received general information supported with pictures of healthy eating habits, including the importance of different nutrients such as vitamin B, C, and folic acid for those children, as well as describing to them the types of drinks and foods that are low in copper. In addition to, dental care and how to prevent tooth decay. Then, discussed activities practice, the importance of exercise practice, and examples of sports that allow them to practice it. Also, they received brief explanations about how to assist them to acquire comfortable and continuous sleep hours, as well as how to deal with insomnia and neuropsychiatric disorders. In addition, the significance of regular follow-up visits. Academic accomplishment and methods for dealing with school absences were also discussed.

The third session aimed to modify reinforcing factors by encouraging the child to become acquainted with healthcare personnel and others who suffer from the same disease, to participate in group activities, to join the Association of Friends of Wilson's Patients, and to advise parents to provide positive reinforcement to their children when they adhere to health behaviors.

The Fourth session aimed to increase enabling factors by offering information on complications prevention, maintenance therapy, hepatitis B and A virus vaccines, stress management, and relaxing techniques and administered educational materials such as pamphlets and booklets.

At the start of each session, feedback on the previous session was provided, and at the end of the session, a summary and conclusion were given.

Evaluation phase

The researchers welcomed all children and thanked them for attending and completing the sessions. Then each child was interviewed individually by a researcher to collect the immediate post-test by using the same instruments. After three months, a reassessment was done using the same instruments.

Statistical analysis

The gathered data was statistically presented and analyzed using the Statistical Package for Social Sciences (SPSS) version 26. Mean and standard deviation (SD) were used to represent quantitative data, whereas frequency and percentage were used to express qualitative data. The Chi-square (χ 2) test was performed to investigate the relationship between categorical variables. The paired t-test was used to compare two groups with normally distributed quantitative variables. A repeated measures ANOVA is used to compare the means of three or more groups with the same individuals in each group. As a measure of the correlation between two continuous variables, Pearson's correlation coefficient was utilized. P values less than 0.05 were considered statistically significant for all tests employed.

Results

Table (1): Characteristics of Studied Children.

Items	N=44	%
Age		
Mean ± SD	12.55±4.64	
Sex		
Male	32	72.7%
Female	12	27.3%
Time since the diagnosis		
6 months < 2 years	7	15.9%
2years < 4years	11	25.0%
≥4 years	26	59.1%
Current Medications		
Octozinc	16	36.4%
Artamine & octozinc	24	54.5%
Don't take any previous drug	4	9.1%

Table (2): Distribution of children's knowledge about Wilson's disease on pre, post, and follow-up tests (N=44).

The total score of knowledge	Pre		Post		Follow up		X21 P1	X22 P2
	No	%	No	%	No	%		
Poor knowledge	36	81.8	2	4.5	1	2.3	70.29	2.45 NS
Fair knowledge	7	15.9	2	4.5	6	13.6	< 0.0001*	.293
Good knowledge	1	2.3	40	90.9	37	84.1		
Mean ± SD	44.91±5.05		48.09 ± 11.67		44.91±5.05		**Paired T-test** 11.70 < 0.001*	**Paired T-test** 1.46 0.17

N.B: *P < 0.001 means a very highly statistically significant difference. NS: (Not Significance) P> 0.05*

X^2 1 & P1: Comparison between pre and post-intervention.

X^2 2 & P2: Comparison between post-intervention and follow-up.

Table (3): Mean scores of children's attitude on pre, post and follow-up tests (N=44)

Items	Pre-test	Post-test	follow up	Repeated measure ANOVA test	P- value
1. Healthy food	22.96±5.06	35.64±4.14	34.06±3.18	**97.21**	< 0.001*
2. Dental care	13.22±3.83	27.52±6.67	26.25±4.51	**90.80**	< 0.001*
3. Sleep habits	9.72 ± 2.92	15.82 ± 4.36	14.25±3.87	**61.15**	< 0.001*
4. Activity practice	15.38 ± 4.09	22.51± 5.70	20.71±2.51	**52.69**	< 0.001*
5. Follow up schedule visits	9.23 ± 2.44	14.21± 3.63	23.11±6.20	**110.38**	< 0.001*
6. Self -an assertion	6.34± 2.81	10.42± 2.62	13.72±3.24	**79.42**	< 0.001*
7. The total score of Predisposing factors Mean ± SD	16.42±5.27	37.46±12.32	33.19±8.75	**83.92**	<0.000*

N.B: *P < 0.001means very highly statistical significant difference*

Table (4): Distribution of children's reinforcing factors on pre, post, and follow-up tests (N=44)

Reinforcing factors	Pre-test No. (%)	Post-test No. (%)	follow up No. (%)	Chi-square test	P- value
Presence of friends' support					
No	36 (81.8%)	4 (9.1%)	3 (6.8%)	**X1=46.93**	P1= <0.000*
Yes	8 (18.2%)	40 90.9%)	41(93.2%)	**X2=0.15**	P2= 0.69
Satisfaction with friends' support					
No	30 (68.2%)	5 (11.4%)	10(22.7%)	**X1=29.64**	P1=<0.000*
Yes	14 (31.8%)	39(88.6%)	34(77.3%)	**X2=2.01**	P2=0.15
Family encouragement to take medication					
No	11 (25.0%)	4 (9.1%)	1 (2.3%)	**X1=3.93**	P1=0.04*
Yes	33 (75.0%)	40(90.9%)	43(97.7%)	**X2=1.91**	P2=0.16
Family encouragement to follow up					
No	16 (36.4%)	3 (6.8%)	0 (0%)	**X1=11.34**	P1=0.001*
Yes	28 (63.6%)	41 93.2%)	44(100%)	**X2=3.11**	P2=0.07
Availability of care at the treatment facility					
No	10 (22.7%)	8 (18.2%)	5 (11.4%)	**X1=0.27**	P1=0.59
Yes	34 (77.3%)	36 81.8%)	39(88.6%)	**X2=0.81**	P2=0.36
Total score Mean ± SD	2.39±0.98	5.24±1.67	4.79±1.09	**Repeated measure ANOVA** 62.76	**p-value** <0.000*

X^2 1 &P1: Comparison between pre and post intervention.
X^2 2 & P2: Comparison between post intervention and follow-up.

Table (5): Distribution of children's enabling factors on pre, post and follow up test (N=44)

Enabling factors	Pre-test No. (%)	Post test No. (%)	follow up No. (%)	Chi square test	P- value
Prevention of the complication					
No	21 (47.7%)	7(15.9%)	2 (4.5%)	**X1=10.26**	P1= 0.001*
Yes	23 (52.3%)	37(84.1%)	42(95.5%)	**X2=3.09**	P2= 0.078
Maintain normalcy					
No	30 (68.2%)	5 (11.4%)	10 (22.7%)	**X1=29.64**	P1=<0.000*
Yes	14 (31.8%)	39 (88.6%)	34 (77.3%)	**X2=2.01**	P2=0.15
Lifelong decoppering					
No	17 (38.6%)	0 (0%)	2 (4.5%)	**X1=19.37**	P1=<0.000*
Yes	27 (61.4%)	44 (100%)	42 (95.5%)	**X2=1.86**	P2=0.17
Compliance with medication					
No	28 (63.6%)	8 (18.2%)	3 (6.8%)	**X1=18.80**	P1=<0.000*
Yes	16 (36.4%)	6 (81.8%)	41(93.2%)	**X2=2.59**	P2=0.11
Getting rid of stress resulting from Wilson's disease					
No	26 (59.1%)	10(22.7%)	5(11.4%)	**X1=12.03**	P1=0.001*
Yes	18(40.9%)	34(77.3%)	39(88.6%)	**X2=2.01**	P2=0.15
Wilson's disease effects on relationships with others					
No	13 (29.5%)	35 79.5%)	37(84.1%)	**X1=22.18**	P1=<0.000*
Yes	31 (70.5%)	9 (20.1%)	7 (15.9%)	**X2=0.31**	P2=0.58
Self-isolation					
No	23 (52.3%)	40(90.9%)	42(95.5%)	**X1=15.24**	P1=**<0.000***
Yes	21 (47.7%)	4 (9.1%)	2 (4.5%)	**X2=0.71**	P2=0.39
Bulling by others when Wilson's disease symptoms shown in a child					
No	19 (43.2%)	37(84.1%)	35(79.5%)	**X1=15.91**	P1=<0.000*
Yes	25 (56.8%)	7(15.9%)	9 (20.1%)	**X2=0.31**	P2=0.58

Enabling factors	Pre-test No. (%)	Post test No. (%)	follow up No. (%)	Chi square test	P- value
Participation in Wilson's disease awareness groups **No** **Yes**	 38 (86.4%) 6 (13.6%)	 11(25.0%) 33(75.5%)	 15(34.1%) 29(65.9%)	 **X1=33.56** **X2=0.87**	 P1=<0.000* P2=0.35
Participation in psychological support groups to fight Wilson's disease **No** **Yes**	 40 (90.9%) 4 (9.1%)	 12(27.3%) 32(72.7%)	 10(22.7%) 34(77.3%)	 **X1=36.85** **X2=0.24**	 P1=<0.000* P2=0.62
The hospital accessibility **No** **Yes**	 23 (52.3%) 21 (47.7%)	 14(31.8%) 30 68.2%)	 11(25.0%) 33(75.0%)	 **X1=3.77** **X2=0.50**	 P1=0.05 P2=0.47
Total score Mean ± SD	4.74±1.55	8.07±3.14	9.88±2.97	**Repeated measure ANOVA** 54.56	<0.000*

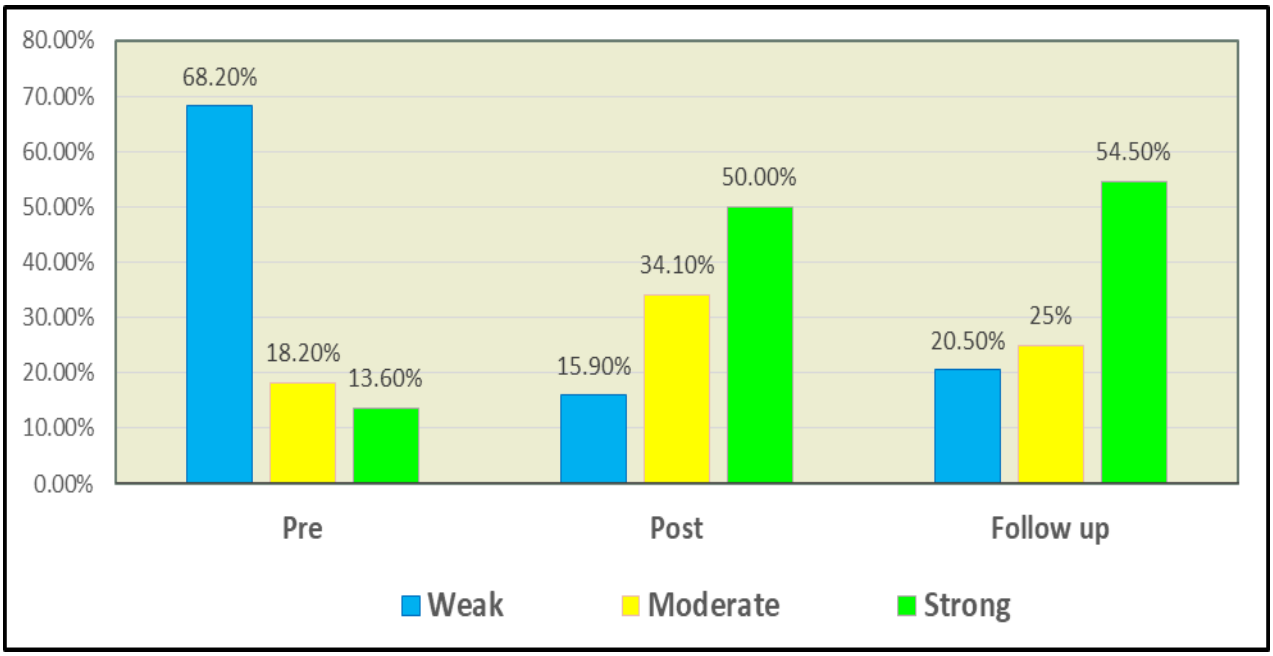

Figure (1): Distribution of total children's predisposing factors, reinforcing and enabling factors scores on pre, post, and follow-up tests.

Table (6): Mean scores of children's initiative generic core quality of life scale (version 4.0) on pre, post, and follow-up tests (N=44)

Items	Pre-test	Post-test	Follow-up test	Repeated measure ANOVA	P- value
1.Physical functioning	36.24 ±11.5	64.71 ±20.47	61.28 ±14.36	52.39	< 0.001*
2.Emotional functioning	27.28±9.33	79.30±22.09	76.92 ±20.74	123.28	< 0.001*
3.Social functioning	41.54 ±15.16	58.78 ±17.84	63.36 ± 22.79	26.37	< 0.001*
4.School functioning	33.82 ±10.8	66.95 ±24.20	69.66 ±21.72	53.22	< 0.001*
Total score	34.58±11.91	67.43±18.62	68.05±10.43	92.02	< 0.001*

N.B: *P < 0.001 (*) means very highly statistical significant difference*

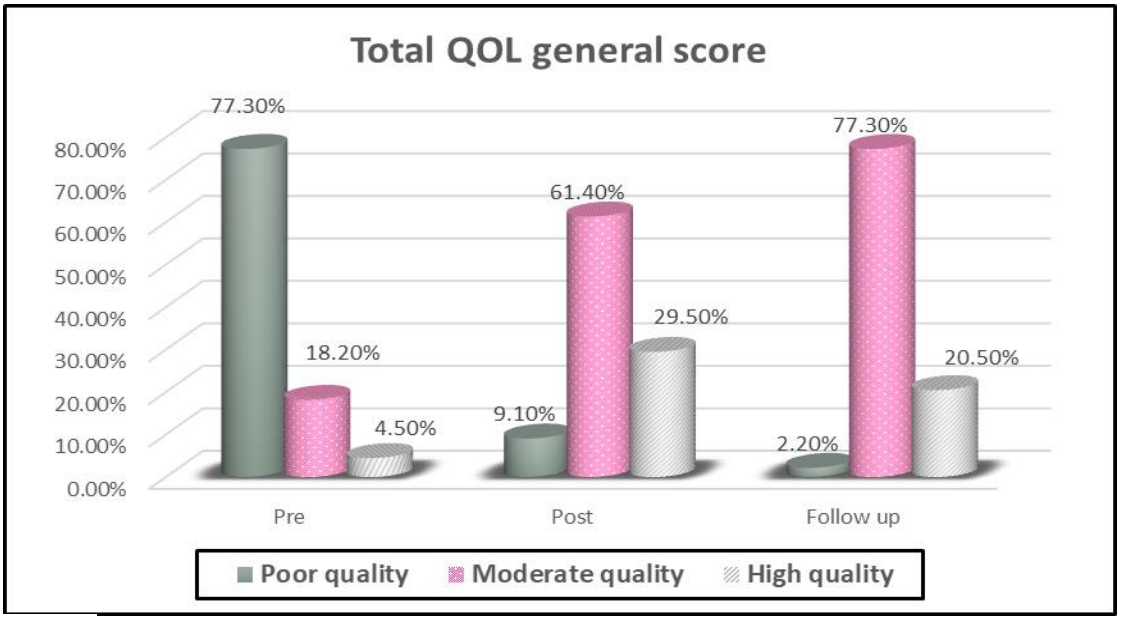

Figure (2): Distribution of children's initiative generic core scale quality of life on pre, post, and follow-up tests.

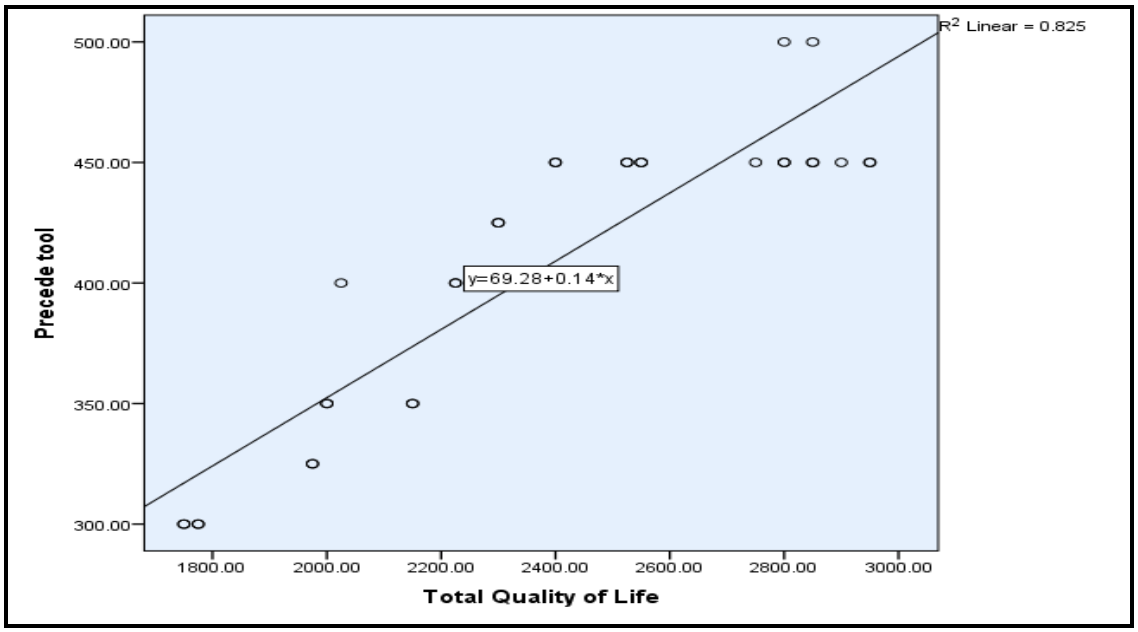

Figure (3): Pearson correlation between the total score of Precede tool and great total score of quality of Life among studied children

Table (1): Clearly showed that the average age of the children studied was 12.55±4.64. In terms of gender, more than two-thirds of the children studied (72.7%) were males. In terms of time since diagnosis, more than half of the children studied (59.1%) were diagnosed four years or more ago. Approximately half of the children evaluated (54.5%) were taking Artamine and Octozinc as current treatments.

Table (2): Demonstrated that the majority of studied children (81.8 %) had poor knowledge of pre-test meanwhile the majority of them (90.9% & 84.1%) had good knowledge of post and follow-up tests respectively. Furthermore, the mean knowledge scores of studied children improved post-test and follow-up compared to the pretest (48.09 ± 11.67, 44.91±5.05 &44.91±5.05 respectively). For this reason, there were highly statistically significant

differences between children`s knowledge on pre, post, and follow-up tests (P<0.001).

Table (3): Revealed that the mean score of all aspects of children's attitudes had been improved on the post and follow-up tests compared to the pretest. Also, the total mean scores on the post and follow-up tests had been improved than the pretest (37.46±12.32, 33.19±8.75 &16.42±5.27 respectively). Therefore, there were highly statistically significant differences between children's attitudes toward activity practice on the pre, post, and follow-up tests (P<0.001).

Table (4): Showed that the majority of children had strong reinforcing factors on the post and follow-up tests than on the pretest. In addition, the total mean score was increased from 2.39±0.98 on the pretest to 5.24±1.67 & 4.79±1.09 on the posttest and follow-up. For this reason, there were highly statistically significant differences between children`s 'reinforcing factors on the pre, post, and follow-up tests (P<0.001).

Table (5): Clarified that the most of children had strong enabling factors on the post and follow-up tests than on the pretest. In addition, the mean of the total score improved on the posttest and follow-up compared to the pretest (8.07 ±3.14, 9.88± 2.97&4.74±1.55 respectively). For this reason, there were highly statistically significant differences between children`s enabling factors on the pre, post, and follow-up tests (P<0.001).

Figure (1): Revealed that two-thirds of studied children (68.20%) had weak factors on the pretest meanwhile half of them (50.00% & 54.50% respectively) had strong factors on the post and follow-up tests.

Table (6): Showed that the mean scores of all quality of life aspects (physical, emotional, social, and school functioning) of studied children had been improved on the post and follow-up tests than on the pretest. In addition, the total score improved from (34.58±11.91) on the pretest to (67.43±18.62 & 68.05±10.43) on the posttest and follow-up. Therefore, there were highly statistically significant differences between children's quality of life on the pre, post, and follow-up tests (P<0.001).

Figure (2): Showed that three-quarters of studied children (77.30%) had Poor quality of life on the pretest meanwhile around two-thirds of them (61.40%) had a moderate quality of life on the post-test and three-quarters of them (77.30%) on follow up test.

Figure (3): Reflected that there was a positive highly statistically significant correlation between the total score of the total precedes tool and the total score of quality of life at the 0.01 level.

Discussion

One of the most serious genetic illnesses in children is Wilson's disease (WD), characterized by an excess of copper in the brain, liver, and other major organs, as well as several organs and tissues. It can cause complications with the liver, neurological, psychiatric, ocular, renal, and muscular systems **(Tremmel, Merle, & Weiskirchen, 2019)**. Nurses play an crucial role in enhancing quality of life (QoL), which measures how well an individual can cope with the burden of disease and treatment in daily life **(Balijepalli, 2021)**. To establish appropriate nursing interventions for patients, it is critical to understand their behaviors and the factors that influence them using the PRECEDE-PROCEED paradigm **(Simeon, 2020)**. Therefore this study aimed to evaluate the effect of comprehensive health promotion intervention on the quality of life among children with Wilson's disease

Regarding the distribution of children's knowledge on the pre, post, and follow-up tests, the present study showed that the majority of studied children had poor knowledge of the pre-test meanwhile the majority of them had good knowledge of the post and follow-up tests. Also, the mean scores for knowledge of studied children improved post-test and follow-up compared to the pretest with a statistically significant difference. This confirmed the positive effect of providing comprehensive health promotion intervention based on precede model. This finding was in line with **(Khalifa, et al., 2017)** who found that the mean of total knowledge of studied parents increased in post-intervention compared to pre-intervention with highly significant statistical differences. This was because children were interested in instructions and nursing interventions provided about the disease, which is considered rare, to gain more information about it.

Concerning the mean scores of children's attitudes regarding healthy food, dental care, Sleep habits, Activity practice, Follow up schedule visits, and Self -assertion on the pre, post, and follow-up tests, the findings clarified that the mean attitude score of children had been improved on the post and follow up tests compared to pretest. Also, the total mean scores on the post and follow-up tests had been improved than the pretest with highly statistically significant differences. From the researchers' point of view, this may be related to the positive effect of comprehensive health promotion interventions on children's knowledge, skills, and attitudes as education improved the self-care of children with Wilson's disease and its management as the essential role of health care providers.

Also, this finding was congruent with **(Nejad et al., 2017)** who discovered that the mean scores of patients' attitudes in both groups before the training

intervention did not differ significantly (p< 0.89), but that 2 months later, the mean score of attitude in the case group was significantly higher than in the control group (p < 0.001). This result could be related to children and adolescents needing to change their behaviors to be able to cope with the disease and avoid complications.

Additionally, these results were consistent with **(Gholampour et al., 2021)** who stated that compared to the control group, the study group shown a significantly higher level of knowledge, attitude, self-efficacy, enabling factors, social support, and health promotion behaviors six months after the intervention. The present study demonstrated the efficacy of a PRECEDE constructs-based intervention in the mentioned factors 6 months after the intervention. This may be due to the concentration of children and adolescents on educational program steps to achieve a better quality of life.

Regarding the distribution of children's reinforcing factors on the pre, post, and follow-up tests, The findings revealed that most children had stronger reinforcing factors on the post and follow-up tests than on the pretest with a statistically significant difference. This result was consistent with **(Bazpour et al., 2019),** who reported that at baseline, the subjects' mean scores for enabling and reinforcing factors were at a low level, but immediately after and a month after intervention, they increased from a moderate level to a high level with highly statistically significant differences (P < 0.001).This may be attributed to the appropriate comprehensive health promotion interventions that tend to be beneficial in boosting positive reinforcement and minimizing negative reinforcement targeted behaviors.

Furthermore, these findings show that treatments based on this model can successfully enhance empowering characteristics that are important for the development of goal-directed learning behaviors and can be accomplished by removing barriers. Also, how families and peers perceived the severity of the disease and therefore provided support and encouragement for their children and adolescents to accept the disease and cope with it.

Regarding the distribution of children's enabling factors on the pre, post, and follow-up tests, the findings revealed that the majority of children had strong enabling factors on the post and follow-up tests than on the pretest. In addition, the mean of the total score improved on the posttest and follow-up compared to the pretest with highly statistically significant differences. This result was in line with the findings of **(Khani et al., 2021)**, who found there was no statistical significant difference between the both groups regarding knowledge, attitude, self-efficacy, reinforcing and enabling factors before the intervention; however, four months later, the study group demonstrated a significant improvement. This result could be related to whether children and adolescents were able to follow the comprehensive health promotion interventions carefully for how to manage stress and prevent complications of the disease this is reflected in the good effect of introducing a comprehensive health promotion intervention regarding to precede proceed model.

Concerning the distribution of total children's predisposing factors, reinforcing and enabling factors scores on the pre, post, and follow-up test. The current study discovered that more than half of the studied children had weak factors on the pretest meanwhile about half of them had strong factors on the post and follow-up tests. This finding was congruent with **(Elkholy et al., 2022)** who revealed that almost all studied children had weak factors on the pretest while most of them had strong factors on the post and follow-up tests. This indicated how much more effective the comprehensive health promotion intervention based on the proceed-proceed model was in increasing factors for children and adolescents to manage and cope with the rare disease in their lives.

Regarding mean scores of the total children's initiative generic core quality of life scale (version 4.0) on the pre, post, and follow-up test, the findings demonstrated that the mean scores of all quality of life components (physical, emotional, social, and school functioning) of studied children had been enhanced on the post and follow up test than on pretest. Furthermore, the total score improved from the pretest to on posttest and follow up with a very highly statistically significant difference. Nonetheless, our research offers a new perspective on the evaluation of WD outcomes through integrated health promotion strategies. It emphasizes the significance of frequent and life-long medicine in this population. To evaluate how WD and its therapy have affected these patients' lives, it is necessary to include measures such as QoL in addition to established scales of impairment. This result was consistent with **(Azar et al., 2020)**, who discovered that in the intervention group, the mean score of quality of life in the physical domain, psychological domain, social domain, and environmental domain improved with a statistically significant difference after one month and three months of intervention.

Concerning the distribution of children’s initiative generic core scale quality of life on the pre, post ,and follow-up test**.** The present study illustrated that three-quarters of studied children had Poor quality of life on the pretest meanwhile more than half of them had a moderate quality of life on the post and follow-up test. This finding was congruent with **(Elkholy et al., 2022)** who reported that all children had poor

quality of life on the pretest while two-thirds of children had a moderate quality of life on the post and follow-up tests with very highly statistically significant differences (P<0.001). This result could be related to comprehensive care achieved by family support, nurse role, and good self-efficacy of children to follow positive behavior to cope with the disease and avoid complications.

Concerning the Pearson correlation between the total score of Precede tool and the total score of Quality of Life among studied children, it reflected that there was a positive highly statistically significant correlation between the total score of the total precedes tool and the total score of quality of life at the 0.01 level. This finding was in line with **(Azar et al., 2017)** who reported that Pearson's correlation coefficients (r) indicated that quality of life was significantly correlated with attitude (r=0. 2, p<0.001), self-management(r=0.665, p<0.001), enabling factors (r= −0.8, p<0.001) and self-efficiency (r=0.21, p<0.001).

Moreover, this finding was the same with **(Maheri et al., 2020)** who reported that associations of QoL with all of the PRECEDE model concepts, including anxiety–depression, self-efficacy, perceived barriers, knowledge, enabling factors, and reinforcing factors were positive and significant correlation (all $p <$ 0.001). This was due to factors of the PRECEDE-PROCEED model affecting children's behavior positively or negatively and consequently affecting their quality of life. The more positive factors, the better the quality of life.

Conclusion

Comprehensive health promotion intervention was successful in improving the quality of life among children with Wilson's disease.

Recommendation

1- Integrating of model-based comprehensive health promotion intervention program PRECEDE-PROCEED in the outpatient clinic of pediatrics for children with WD and their caregivers to improve their quality of life outcomes.
2- Continuous health education programs for children with WD can help in providing continuous support and early detection of any complications.
3- Further studies
   Large future studies in children with WD with a larger sample probability are needed to increase the accuracy of the results.
   In Egypt, the development of a national program with regular education about Wilson's disease seminars for health center nurses.

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