MACROJOURNALS

The Journal of **Macro**Trends in
Health and Medicine

Effect of Family Centered Intervention on Copper Level of Children with Wilson's disease

Maha I. Khalif*, Jaklein R. Younis, Mohammed A. Khedr***, Hind M. Emara******

*Prof. of Pediatric Nursing, Paediatric Nursing Department, Faculty of Nursing, Menoufia University, Shebin El-Kom, Egypt

**Assist Prof. of Pediatric Nursing Pediatric Nursing Department, Faculty of Nursing, Menoufia University, Shebin El-Kom, Egypt

***Assist. Prof. of Pediatric Hepatology, National Liver Institute, Menoufia University, Shebin El-Kom, Egypt

****Assist.lecturer. of Pediatric Nursing, Pediatric Nursing Department, Faculty of Nursing, Menoufia University, Shebin El-Kom, Egypt

Abstract

Wilson's disease is the most common cause of defective excretion of copper from the body affecting most commonly children or young adults and running invariably fatal consequences if not adequately managed. Purpose: this study was conducted to evaluate the effect of family centered intervention on copper level of children with Wilson's disease and their family practices. Setting: It was conducted in Pediatric outpatient clinic of the national liver institute Menoufia University at Shebin El_koom town. Design: A quasi experimental design was utilized for this study. Sample: a convenient sample of 25 children with Wilson's disease and their families was included. Instruments: Three instruments were utilized. Instrument one: structured interview questionnaire about the impact of Wilson's disease on child and Family. Instrument two: Structured interview questionnaire about family knowledge of children having Wilson's disease. Instrument three: Laboratory investigations record. Results: The results of this study showed significant improvement in parents' knowledge and practices. There was also, a significant reduction of the copper excretion level in urine in posttest than in pretest. Conclusion: It was concluded that implementation of family centered intervention reduced the level of urinary copper excretion for children with Wilson's disease. Recommendations: Family centered intervention should be urgently planned and utilized for children with chronic health problems to improve families' knowledge, enhance their practices and promote health status of their affected children.

Keywords: *Wilson's disease; Family centered intervention; copper Level.*

1.Introduction:

Wilson's disease (WD) in children is a major health problem of interest. It causes accumulation of copper (CU) in the liver initially progressing to cirrhosis and in the central nervous system resulting in severe neurological complications. Excess CU leads to undesirable life-threatening effects [1].

The prevalence rate of WD in children has been detected in all known ethnic groups worldwide. The incidence was 1 in 55,000 births in the United States and 1 in 30,000 to 1 in 50,000 births worldwide. In Egypt the most common hepatic disease is viral hepatitis C virus (HCV)) with the highest prevalence worldwide. Accordingly, viral hepatitis masks other liver diseases. Therefore, WD can be easily missed during diagnosis in addition to limitations of clinical data on large cohorts. The disease can present at any age from 5 to 50 years or more, but the peak incidence is between 8 and 16 years of age [2] [3]. The younger the patient, the more likely hepatic involvement will be the predominant manifestation. Girls are 3 times more likely than boys to present with acute hepatic failure. Clinically evident liver disease may precede neurologic manifestations by as much as 10 years [3].It can be present clinically as asymptomatic hepatomegaly (with or without splenomegaly), sub-acute or chronic hepatitis, and acute hepatic failure (with or without hemolytic anemia). In addition, cryptogenic cirrhosis, portal hypertension, ascites, edema, variceal bleeding, or other effects of hepatic dysfunction. Also, it can be present as a progressive neurological disorder as intention tremor, dysarthria, rigid dystonia, parkinsonism, choreiform movements, lack of motor coordination, behavioral changes or as psychiatric illness (depression, personality changes, anxiety, or psychosis) [2] &[4].

An available regimen of WD management that gives hope to those predisposed to the disease if diagnosed early is through restriction of CU intake. Besides, there are several chelating agents and zinc salts can be provided. Liver transplantation corrects the underlying pathophysiology and can be lifesaving. Above all, many families do not follow appropriate diet regimen. Therefore, there is progressive and potentially fatal mortality worldwide [4] &[5].

Advanced researches in health care service delivery to children with special health care needs indicates that family-centered practices may enhance a child's health and developmental outcomes, specifically psychological stability, behavioral functioning, and improved health-related quality of life even when the severity of the condition is taken into account [6].

Despite widespread endorsement FCC continues to be insufficiently implemented into clinical practice. FCC demonstrates its best with information sharing, partnering, respect, and negotiation leading to a successful outcome in a difficult clinical scenario. [7].In pediatrics, FCC is based on the understanding that the family is the child's primary source of strength and support and that the children and family's perspectives and information are important in clinical decision making [6].

Although health care professionals agree that FCC is important approach of care they consistently do not regularly provide such care to families [6]. Consequently, we conducted the present study to evaluate the effect of family centered intervention on CU level of children with WD and their family practices. This based on the valuable recommendation of patient and FCC institute in the United States who believes that, family is a constant in the child's life;

facilitating parent professional collaboration at all levels of health care; and adopting policies and practices that enhance child and family developmental needs as part of health care practices. [8].

1.1. Purpose of the study:

This study was carried out to investigate the effect of family centered intervention on CU level of children with WD and their family practices.

1.2. Research hypotheses:

1. Families will have higher level of knowledge and practices related to care of their children on post-test than on pre-test.
2. Families will have more adjustment with disease on post-test than on pre-test.
3. On post-test children will have lower level of CU in the 24-hour urine collection than on pre-test.
4. On post-test children will have improved hepatic function (lower level of Aspartate transaminase (AST) and Alanine transaminase (ALT)) in the blood than on pre-test.

2. Methods:

2.1. Research design:

A quasi-experimental design was utilized for this study (pre and post-test).

2.2. Setting:

This study was conducted at the outpatient clinic of pediatrics in the National Liver Institute Menoufia University at Shebin El_koom city. The outpatient clinic of pediatrics included two rooms. *The first room* was specialized for children with metabolic liver diseases. *The second room* was for children having other pediatric liver diseases. Children who had WD attended the clinic each Sunday between 9 am- 12 pm.

2.3. Sample and Sampling Technique:

All children (25 children) who had WD and and meet the criteria for selection were included.

Inclusion criteria:-

Children had only WD without any other health problem.

2. 4. Instruments:-

Instrument one: Structured Interview questionnaire about the impact of WD on child and Family: It was developed by the researcher. It was used to collect data about children, primary caregivers and siblings in order to examine the effect of WD on them. It included three parts:

Part one: Characteristics of the sample:It included three subparts:

- ***Subpart one: Characteristics of Children with WD***: It includes questions about name, age, sex, duration since diagnosis, medications described to the child (D-pencillamine or zinc or both).

- ***Subpart two***: ***Healthy Child/Sibling Characteristics:*** It includes number of siblings, their sex, their age, and any health problems they may suffer from.
- ***Subpart three***: ***Parental or Family Characteristics****:* It includes primary caregiver's address, telephone number, marital status, and type of family, parent's education and job, any chronic health conditions, other adults in the home and their ages and the family health insurance.

Part two*: Structured questionnaire concerning the effect of WD on children:* It is a multiple choice question developed to assess hepatic, neurological and psychological problems of WD on children. *Firstly*, psychological problems such as social isolation sleep disorders, nervousness, and depression. *Secondly*, problems related to liver such as jaundice, bleeding tendency, abdominal pain, edema and ascites. *Thirdly*, problems related to nervous system such as behavioral changes, tremors and involuntary movements, arthritis and headache, walking and talking disturbances, amnesia, difficulty to concentrate and deterioration of school achievement and others.

Part three*: Structured questionnaire related to the effect of WD on children's family and siblings:* It is multiple choice question developed to assess stresses of families who have children with WD such as absence from work, absence of siblings from school or their work, family disparities or divorce, increased dependency on parents, change of family life style and any other family stressors.

Part four: Illness Adjustment Structured Questionnaire: It includes multiple choice questions to assess to what extent the caregiver's adjustment to WD.

Scoring system for questions:

Parameters	***Score***
Correct	2
Incorrect	1
Others	0

Instrument two: Structured Interview Questionnaire about Family knowledge related to care of children having WD*:*

Part one: knowledge about WD*:* It is a structured interview questionnaire. It is developed by the researcher after a thorough review of related literature. Also, it contains questions about definition, causes, signs and symptoms, medication, care of the disease and diet.

Part two: Family performance checklist. It contains 14 questions answered by yes or no. It is developed to assess family's performance and practices to maintain stability of their child's clinical status and prevent any further complications.

Scoring system for questions 1 - 16:

Parameters	***Score***
Correct	2
In Correct	1
Others	0

Scoring system for questions 17 - 31:

Parameters	***Score***
Yes	2
No	1

Instrument three: Laboratory investigations record*:* This part is developed by the researcher after a thorough review of related literature. It contains three parts.

Part one*:*

It was used to record level of CU in amount of urine collected for 24 hours.

Scoring system for copper level

Parameters	***Score***
More than 70	1
Less than15	2
15-70	3

Part two*:*

It was used to record levels of Aspartate transaminase (AST) in the blood

Scoring system for levels of AST

Parameters	***Score***
More than 34	1
Less than10	2
10-34	3

Part three*:*

It was used to record levels of Alanine transaminase (ALT) in the blood

Scoring system for levels of ALT

Parameters	***Score***
More than 44	1
Less than10	2
10-44	3

2.4.1.Validity:

Three instruments were provided to a jury of three experts. One pediatric nursing professor, one pediatric nursing assistant professor and one assistant professors in pediatrics.

2.4.2. Reliability:

For ***Family knowledge related to care of children having WD ,***the reliability of the tool was done to determine the extent to which items in the tool were related to each other by Cronbach's co-efficiency Alpha (a=74).

For ***Structured questionnaire about the impact of WD on child and Family,*** the reliability of the tool was done to determine the extent to which items in the tool were related to each other by Cronbach's co-efficiency Alpha (a=61).

3. Data collection methods:

Ethical Consideration:

For protection of human rights an official letter was sent to the previously mentioned setting to inform about the purpose of study and request their assistance to facilitate the work. A seminar was conducted in the Pediatric Department National Liver Institute to assess adherence to the rights of patients. Families were informed about the privacy of their information, nature of the study and their right to withdraw. An oral and written consent were obtained from parents or care givers to share in the study. Therefore, the objectives of the study, its importance, safety and confidentiality were obtained.

3.1. Pilot study:

It was carried out on four parents and their children (10% of the sample) after the tools were developed and before starting the data collection. It was done to test the practicability, applicability and to estimate the needed time to fill the tools. Some modifications were done. Therefore, the pilot study was excluded from the total sample.

3.2. Procedure of data collection:

1- A written permission to carry out the study was obtained from the director of the setting after submitting an official letter from the dean of the faculty of Nursing at Menoufia University explaining the purpose of the study and methods of data collection.

2- ***Data collection procedure,*** data collection started at April 2016 and lasted until the end of December 2016. Data collected one day per week.

3- The researcher introduced herself to the parents of children, explained the purpose of study and methods of data collection.

4- Family centered intervention was provided to parents and their children for three months.

5- Instruments one and two were filled by the researcher according to answers of primary caregivers who accepted to share in the study. The researcher provided full explanation of each item and they were encouraged to ask questions.

6- Collected data was used to analyze and determine areas of knowledge deficit and formulate a plan for health education and family support.

7- Each primary caregiver was instructed about 24 hours urine collection to assess CU level. They were provided two laboratory experimental tubes (each one contained 5 ml of preservative HCL). Caregivers were instructed to collect the urine passed within 24 hours for each child. Instructions were provided about time, technique of urine collection (e.g. urination in cup then pouring in bottle). Also, caregivers were instructed to protect urine from light (through covering bottle with a black plastic bag) and hair dropping (e.g. covering the bottle after each urine collection) (pretest).

8- The researcher telephoned each caregiver to answer their required questions, provide required guidance, and discuss their concerns.

9- A Health education plan was planned according to the individual needs for each child. Health education was individually provided to parents of each child.

10- Caregivers of each child received one individual health education session. The session contained Health education about WD, its management and required dietary modifications.

11- The researcher used colored brochures for low educated parents. It lasted for one hour. At the end caregivers received a full booklet containing WD definition, causes, pathophysiology, signs and symptoms, medical treatment, diet therapy complications, high CU and low CU diet and family role to adjust to the disease and maintain stable child health status.

12- The researcher enhances communication with the primary caregivers, sharing experiences and provided the required guidance.

13- At the end of the session, the researcher answered their questions, provided needed guidance and planned for future meetings.

14- The researchers telephoned caregivers of each child one time per week. Necessary guidance and health education were provided according to the condition of each child and parent's needs. Researcher provided them psychological support and empowerment. Some parents needed more frequent telephone calls to ensure their adherence to the family centered intervention, provide empowerment and promote their coping (listening to complaints and helping to solve accompanying problems such as lack of medications, emotional and personal stresses).

15- Reassessment was done for their knowledge and practices about WD, management and dietary modifications (posttest).

16- Investigation of CU level in the urine was obtained from the laboratory records (posttest).

3.3. Data analysis:-

Data was coded and transformed into specially designed form to be suitable for computer entry process. Data was entered and analyzed by using SPSS (Statistical Package for Social Science) statistical package version 21. Graphics were done using Excel program.

Quantitative data was expressed as mean & standard deviation (X ±SD) and analyzed by using Friedman test and ANOVA test for comparison between means.

Qualitative data was expressed in the form of number and percentage. It was analyzed by using chi-square test (X^2).Pearson correlation was used for explaining relationship between normally distributed quantitative variable.

A statistical significant difference was considered if P < 0.05

Results:

Table (1):Characteristics of studied children. It was obvious from this table that more than two thirds of studied children (68.0%) were in the age group of 12-18 years. Regarding children's sex, more than two thirds of studied children (68.0%) were males. In relation to time elapsed since diagnosis, more than half of studied children were diagnosed 4yearsago or more. For present medications, about half of studied children (52.0%) were taking Artamine and Octozinc.

Table (2): Mean and standard deviation of Total knowledge of studied Parents about Wilson disease in pre and post-tests

shows mean and standard deviation of Total knowledge of studied parents about WD in pre and post-tests. As indicated in the table, the mean of total knowledge of studied Parent in pre and post intervention was 17.63±9.28 compared to 44.43±0.82 respectively. There were obvious highly significant statistical differences between parents knowledge at 1% level of statistical significance.

Table (3): Mean of level of family role in the care of their affected children in pre and post intervention Table 6 represents mean of total family role with their affected children in pre and post intervention. Findings showed that the mean of total families' role regarding care of their children with WD in pre and post intervention were 20.26 ±3.25 and24.78 ± 1.91respectively. There were highly significant statistical differences between families' roles in pre and post intervention at 1% level of statistical significance.

Table (4): Mean of total children's adherence to diet regimen on pre and post intervention. Table 8 showed level of children's adherence to diet regimen on pre and post intervention. Findings illustrated that the mean of total diet regimen taken by studied children on pre and post intervention were 15.82 ±3.46and 21.35 ±2.85respectively. There were obvious highly significant statistical differences between diet regimen taken by studied child pre and post intervention at 1% level of statistical significance.

Table (5): Adjustment of families for their children having Wilson's disease. Table 10 showed adjustment of families for their children having WD, there were highly significant statistical differences between studied sample in pre and post intervention regarding daily diet recall and suggestions to other parents having the same conditions at 1% level of statistical significance($P<0.001$). However, no significant statistical difference were found between studied sample in pre and post intervention concerning family stressors due to WD and things help family accommodate to WD.

Table (6): Means and standard deviation of Urinary CU level, ALT and AST on pre and post-test.

Table 11 represented means and standard deviation of Urinary CU level, ALT and AST on pre and post-test. This table indicated that, level of Urinary CU was reduced after intervention (118.84 ± 82.66 Vs 245.35± 203.93). There was a significant statistical difference between mean Urinary CU level on pre and post intervention at 5 % level of statistical significance. Unlike urinary CU level, there were no statistical significance differences between means of AST and ALT on pre and post-test.

Table (7): Pearson Correlation between parent's total knowledge and total family role, total diet regimen, Urinary CU excretion, ALT and AST Table 12 illustrated the Pearson correlation between parent's total knowledge and total family role, diet regimen total diet regimen, Urinary CU excretion, ALT and AST of studied sample. It reflected that there were a positive correlation between parent's total knowledge and total family role and total diet regimen at 1% level of statistical significance and there were a negative correlation between parent's total knowledge and urinary CU excretion at 5% level of statistical significance.

On the other hand, there were no correlation between parent's total knowledge and AST and ALT.

Table (8): Pearson Correlation between total diet regimen and urinary CU excretion, ALT and AST

Table 18 showed Pearson correlation between total diet regimen and urinary CU excretion level, ALT and AST. There was a negative correlation between total diet regimen score and urinary CU excretion level at 5% level of statistical significance.

Meanwhile, there was no correlation between total diet regimen, ALT and AST

Figure (1): Levels of Urinary CU exertion, ALT and AST in pre and post intervention. This figure showed levels of Urinary CU exertion, ALT and AST in pre and post intervention. As illustrated from this figure, level of Urinary CU exertion was reduced on post-test (245.3, 60.87, 53.93), than on pre-test (118.84, 50.81, 49.5)**.**

Table (1):Characteristics of studied children.

Characteristics of studied children	*n=25*	*%*
Age		
<6 years	2	8.0%
6-12 years	6	24.0%
12-18 years	17	68.0%
Sex		
Male	17	68.0%
Female	8	32.0%
Time elapsed since diagnosis		
6 months <2 years	5	20.0%
2years < 4years	5	20.0%
4 years or more	15	60.0%
Present Medications		
Octozinc	11	44.0%
Artamine and octozinc	13	52.0%
Don't take previous medications	1	4.0%
Associated chronic diseases		
Yes	6	24.0%
No	19	76.0%

Table (2): Mean and standard deviation of Total knowledge of studied Parents about Wilson disease in pre and post-tests

Total knowledge of studied parents about WD	Pre	Post	t. test	P -value
Mean ± SD	17.63 ±9.28	44.43 ± 0.82	-23.303-	<0.001

Table (3): Mean of level of family role in the care of their affected children in pre and post intervention

Mean & SD of level of family role	Pre	Post	Wilcoxon test	P -value
Mean ± SD	20.26 ±3.25	24.78 ± 1.91	-7.05-	<0.001

Table (4): Mean of total children's adherence to diet regimen on pre and post intervention.

Mean & SD of total adherence to diet regimen	Pre	Post	t-test	P -value
Mean ± SD	15.82 ±3.46	21.35 ±2.85	-6.95-	<0.001

Table (5): Adjustment of families for their children having Wilson's disease

Adjustment of families for their children having WD	Pre		Post		X^2	P -value
	n	%	n	%		
Family stressors due to WD					4.03	0.672
Absence from job	7	28.00%	5	20.00%		
Siblings absence from the school and faculty	1	4.00%	0	0.00%		
Family disparities as divorce	2	8.00%	3	12.00%		
Increase dependency on parents	2	8.00%	2	8.00%		
Change in daily living activities and life style	4	16.00%	7	28.00%		
All of the above	5	20.00%	2	8.00%		
Others	4	16.00%	6	24.00%		
Your suggestions to parents having the same conditions					21.42	<0.001
Strictly follow recommended diet for WD	1	4.00%	0	0.00%		
Administer medication on time	2	8.00%	0	0.00%		
follow up regularly at clinic	3	12.00%	0	0.00%		
All of the above	10	40.00%	25	100.00%		
Others	9	36.00%	0	0.00%		
Things help family accommodate to WD					3.98	0.263
Financial support	1	4.00%	1	4.00%		
Psychological support	9	36.00%	4	16.00%		
All of the above	14	56.00%	20	80.00%		
Others	1	4.00%	0	0.00%		
Daily diet recall					26.29	<0.001
High CU diet	23	92.00%	5	20.00%		
Low CU diet	2	8.00%	20	80.00%		

Table (6): Means and standard deviation of Urinary CU level, ALT and AST on pre and post-test.

Diagnostic tests	Pre	Post	t-test	P -value
	Mean & SD X ± SD	Mean & SD X ± SD		
Urinary CU level	245.35± 203.93	118.84 ± 82.66	2.56*	0.021
ALT (n=25)	60.87 ± 51.50	50.81±36.51	1.22	0.238
AST (n=25)	53.93 ± 37.04	49.50 ± 36.86	1.29	0.214

Table (7): Pearson Correlation between parent's total knowledge and total family role, total diet regimen, Urinary CU excretion, ALT and AST

Items	*Correlation with Knowledge*	
	r	*P -value*
Total family role	0.674**	<0.001
Total diet regimen	656**	<0.001
Urinary CU excretion	-0.471**	0.004
ALT	-0.152-	0.407
AST	-0.091-	0.622

** Correlation is significant at the 0.01 level (2-tailed).

Table (8): Pearson Correlation between total diet regimen and urinary CU excretion, ALT and AST

Diet regimen and urinary CU excretion, ALT and AST	**Total Diet regimen score**	
	r	**P –value**
Urinary CU excretion	-0.386*	0.022
ALT	0.173	0.293
AST	0.277	0.088

* Correlation is significant at the 0.05 level (2-tailed).

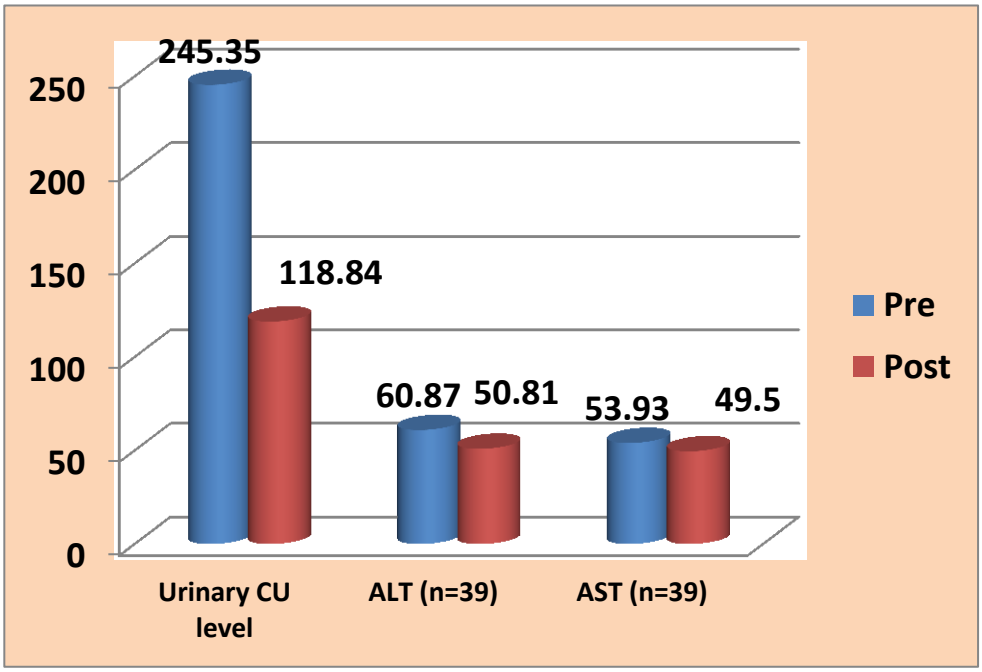

Figure (1): Levels of Urinary CU exertion, ALT and AST in pre and post intervention.

Discussion:

Wilson disease is one of the most dangerous genetic disorders in children. Impaired CU excretion causes deposition of CU in the liver and brain with significant disability or death if left untreated. WD causes extensive suffering for both children and their families [9]. An available regimen of treatment gives hope to those predisposed to the disease if diagnosed early [10].

Studies show that appropriate FCC for children with significant health problems which focuses on increasing parents' awareness regarding disease process and its better management. will lead to improved health and functional status of these children[11].

Accordingly, the current study hypothesized that, firstly: family centered intervention will improve knowledge and practices of families related to care of their children.Secondly: Families will have more adjustment with disease process. Thirdly:Children will have lower level of CU in 24-hour urine collection and lower level of AST and ALT in the blood. Fortunately, the present study is one of the pioneer's research studies in investigating the effect of family centered intervention on urinary CU level and family practices of children with WD.

Regarding characteristics of studied children, the present study illustrated that two of the twenty five studied children (8.0%) were less than six years. They were two boys (2.8 and 5.5 years old) respectively. They were screened after death of their older female sibling due to complications of WD. This finding agreed with [12]. whom conducted a research about " WD in Children: Analysis of Fifty Seven Cases " They found that the youngest patients were two girls. Their ages were four months and twenty three months respectively. They were screened after the diagnosis of WD in their older siblings. However, this finding contradicted with [13]. whose study entitled "Frame shift and nonsense mutations in the gene for ATPase7B are associated with severe impairment of CU metabolism and with an early clinical manifestation of WD" and stated that the mean age of patients at initial disease manifestation was 26 ± 9 years. According, [12]. this finding also reflected that WD could start at early ages when CU accumulated in large amounts in the body. Therefore, it is imperative that WD should be considered in the differential diagnosis of established severe liver disease in any young child even below three years old. This could reveal that WD could be discovered before six years of age. Early diagnosis could lead to better prognosis.

For sex of studied children, the present study identified that approximately two thirds of the sample (68.0%) were males and 32.0% were females respectively. This result was consistent with [14]. whom conducted a research about "Gender Differences in WD" and they observed that there was a male predominance in their studied population (e.g. 627 males vs. only 290 females). However, this finding comes in contrast with Wong et al., 2011they conducted a research about "A Clinical Assessment of WD in Patients with Concurrent Liver Disease" and found that forty two WD patients (eighteen male and twenty four female) were only identified.

Regarding characteristics of twenty five studied families the present study indicated that two parents (8%) had two dead children and seven parents (28%) had one dead child from complications of WD. In the researcher opinion, this result may be due to late diagnosis and lack of knowledge about appropriate management of the diseased children at home. Also, it can be due to lack of follow up which may be attributed to high illiteracy level among parents (64% and

76% of mothers and fathers were illiterate respectively). Furthermore, 32% of fathers were workers and 24% were farmers so that this low socioeconomic status leads to inability of the family to follow up regularly at the pediatric liver clinic and do all required checkups for their diseased children. Also, 4% of parents were divorced because of having children with WD.

Concerning hypothesis one, it was obvious that the majority of parents had higher level of knowledge on post-test than on pre-test. This finding was in agreement with Ernst et al., (2017) they conducted a research about "Development and Evaluation of a Generic Education Program for Chronic Diseases in Childhood". They found that the parents demonstrated improved disease-specific knowledge.

In addition, the mean total knowledge of studied Parent about WD and its related care improved on post-test than on pre-test. This finding came in agreement with [15]. they carried out a research entitled "a study to assess the effectiveness of structured teaching program regarding knowledge on management of febrile convulsion among mothers of fewer than five children in Rajarajeswari medical college and hospital, Bangalore". Their results revealed that mean post-test knowledge of mothers of fewer than five children regarding management of febrile convulsion was significantly greater than their mean pre-test knowledge. This could be attributed to the continuity of health education and phone calls asking about children's health and providing needed instructions.

In relation to the family practices regarding care of their children on pre and post intervention, the current study showed that the majority of families had more adequate practices related to the care of their affected children in post-test than in pre-test. This could be related to the positive correlation between total parent's knowledge and total family role. These results agreed with [16].they run a research about" Developmental education for parents of delayed infants: Effects on Parental Motivation and Children's Development" Results revealed that Parents gained better knowledge and had better participation in home care program . This result came in agreement with [17]. they conducted a study about" The Evolution of FCC" and stated that FCC enabled parents to be influential in improving care of their children.

In addition,[18]. they conducted a study about "Parenting a child with chronic illness as they transition into adulthood: A systematic Review and Thematic Synthesis of Parents' Experiences ". They found that Parents require clarification on their role and support from service providers to be the key facilitators of their child's healthcare transition supporting them to become experts in their own condition and care. Consequently, parents of children with WD needed continuous psychological support to enhance their role in improving the health status of their children. This finding could be attributed to brochures distributed by the researcher, booklets and frequent phone calls with parents.

Besides, the previous results were supported by [19]. they studied "a randomized trial of a nursing intervention to promote the adjustment of children with chronic physical disorders". This showed improvements in performance of their roles.

However, this finding was contradicted by [20]. whom conducted a research about "Knowledge, attitudes, and practices among mothers of children with epilepsy: A study in a

teaching hospital" They showed that in spite of demonstrating good knowledge scores, the majority of mothers felt the need for further training in epilepsy management.

For hypothesis two, the current study revealed that the majority of children had better adherence to the recommended diet regimen in post-test than in pre-test (52% and 76% respectively).Moreover, 80% of children received low CU diet in posttest compared to 92% who received high CU diet in pre-test. There was a positive correlation between total parent's knowledge and adherence to diet regimen. These results were confirmed by the previous experience of [21]. they conducted a study about "Nutrition knowledge and Mediterranean Diet Adherence in the Southeast United States: Validation of a Field-based Survey Instrument". They revealed that greater Mediterranean Diet adherence was found with increasing formal nutrition education in university students and farmers market participants.

This finding was in line with a study conducted by [22]. about" The effect of short message system (SMS) reminder on adherence to a healthy diet, medication, and cessation of smoking among adult patients with cardiovascular diseases" Study group participants showed more adherence to healthy diet than control group. Adherence to healthy diet reduced complications of diseases. Therefore, children on post-test didn't develop further complications.

On the contrary, [23]. in his study about "The Impact of Family Functioning on Treatment Adherence and Metabolic Control for Adolescents with Poorly Controlled Type 1 Diabetes" found that there were no differences between study and control group in relation to adherence to diet regimen. Moreover, [24]. who conducted a research about" Understanding The Role of Self-Efficacy and Social Support on Diet and Exercise Adherence to a Lifestyle Change Program" stated that compliance to diet regimen was problematic. It is not well understood why individuals fail to maintain these regimens.

Concerning distribution of children according to complications of WD, the present study revealed that there were no changes in complications of children in post-test. This could be attributed to the long lasting history of liver cirrhosis before conducting the family centered intervention. Earlier start of family centered intervention is expected to reduce further complications and stabilize serious deteriorations in actually symptomatic children. Perhaps, it could prevent the occurrence of complications in pre-symptomatic children.

For family stressors due to WD, there were no significant statistical differences between pre and post-test. The family faced the following stressors absence from job, siblings' absence from the school and faculty, family disparities as divorce, increased dependency on parents and Change in daily living activities and life style. Stressors weren't reduced in this study due to their need for financial aid and social problems such as divorce.

Regarding parents' suggestions or advice to other parents having children with the same disease, the present study demonstrated that parents provided suggestions about following recommended diet for WD. Strictly, administration of medication on time and following up regularly at clinic. The present study demonstrated that families showed more adjustment with challenges of WD that was represented in their suggestions about followingrecommended diet forWD Strictly, administration of medication on time and following up regularly at clinic. This

finding comes in line with Pelentsov, [23]. they conducted a study about "The Supportive Care Needs of Parents Caring for a Child with a Rare Disease: A scoping Review". Additionally, this finding was supported with [24]. they conducted a study about "Coping when a Child has a Disability: Exploring the Impact of Parent to Parent Support". This could be attributed to parents' talking with other parents would enable them to better adjust and have more positive outlook for their lives.

In relation to family need for psychological support, the needs were reduced in post-test than on pre-test. However, there was no significant statistical difference between pre and posttest. This result agreed with [21]. they run a study about "Experience of Support for Parents of Adolescents with Heart Defects—Supported to be Supportive". However, psychological support that was provided through family centered intervention could help parents to be supportive for children. Family centered intervention was expected to raise the sense of gratification and contentment of parents for fulfilling supportive functions. This could help parents to provide better care and support for their children.

In relation to hypothesis three and four as regards to the level of urinary CU, AST and ALT , the present study revealed that level of urinary CU was reduced post intervention (118.84 ± 82.66) compared to (245.35± 203.93) on pre intervention. The current result came in agreement with Brewer & Askari, (2005) they conducted a research, about " WD: Clinical Management and Therapy".

Also, this finding came in line with [25]. they conducted a study about "Low-CU diet as a preventive strategy for Alzheimer's disease" Stated that a high-CU intake regimen declines the efficiency of CU absorption, but more CU is absorbed and retained. As well as, Excess CU can also lead to specific biochemical changes and thus undesirable and life-threatening effects.

This study was consistent with [26]. they studied" Metabolism of CU in Hepatolenticular degeneration "He found that low CU diet could lead to lower amounts of CU retained in the body.

This was attributed to their consumption of low level of CU in food. Unlike urinary CU level, levels of AST and ALT weren't changed in pre and post-test. This could be due to the long lasting history of liver cirrhosis.

From the researcher perspective, the lower CU intake regimen, the less CU is absorbed resulting in reduction in urinary CU excretion level and the fewer serious effects of CU accumulation or intoxication.

Regarding the correlation between parent's total knowledge and urinary CU level, the present study found that the increment of parent's knowledge was associated with reduced CU level. This could be attributed to parents' adherence to educational content about the importance of low CU intake in children's' food.

Conclusion:

Implementation of a family centered intervention increased parents' knowledge and practices regarding care of children with WD, decreased the level of CU in the 24 hour urine collection on post-test and increased parent adjustment with disease on post-test than on pre-test. However, there was no improvement in hepatic function (AST and ALT) on post-test than on pre-test.

Recommendations:

In the light of the findings obtained from the current study, the following recommendations are suggested:

A. Recommendations for Clinical Nursing Practice:-

1. Ongoing in-service education programs should be designed and implemented at pediatric clinics to provide pediatric nurses knowledge about the benefits and practices of the key elements of FCC for children with WD.
2. All principles of FCC should be taken in consideration when caring for children with WD and their families to improve their outcomes.
3. Wilson's disease management protocol of care should be well utilized for all pediatric nurses, children with WD and their families.

B. Recommendations for Education:-

1. Continuing education and service training programs regarding FCC of children with WD and their families should be performed continuously for pediatric nurses to improve their competency and care delivery.
2. Family centered nursing interventions for children with WD and their families should be urgently integrated into pediatric nursing curriculum.

C. Recommendations for Hospital polices:-

1. All service providers in the hospitals (e.g. nurses and physicians), regardless of the frequency of their interactions with the child and family or what role they play, need to be aware of the key elements of family-centered service.
2. Programs to disseminate FCC practices are needed to improve children and family outcomes in the National Liver Institute.
3. Specialized units are urgently required for developing parent's knowledge and practices regarding care of their diseased children.
4. Advanced scientific booklets for children and their parents regarding WD management should be available at pediatric liver clinics.

D. Recommendations for Research:-

1. Suggested further family centered interventional studies on children with WD should be conducted to address family stressors and enhances coping.

2. Replication of this study on a larger probability sample from different locations in Egypt is amenable.

Limitation of the study:

Small sample size due to the unavailability of other pediatric care setting for children with WD.

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