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Nursing Care and Quality of Life Among Children with Hepatolenticular Degeneration

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Abstract

Hepatolenticular degeneration also known as Wilson's disease (WD), is an autosomal recessive inherited copper metabolic disorder. ***Objective:*** The study aimed to determine the relationship between nursing care and the quality of life among children with hepatolenticular degeneration in Anhui Province, China. ***Methods:*** This research used the descriptive-correlational design. The researcher investigated 45 regular staff nurses and 96 children with hepatolenticular degeneration ages 12 and 18 years old. Two instruments were used. First, the Nursing Care Score Tool was developed by the researcher based on review of related the literature which is used to measure nursing care. Second, the revised PedsQLTM4.0 Generic Core Scales (Pediatric Quality of Life Inventory™ Version 4.0) is used to measure the quality of life among children with hepatolenticular degeneration. The data were analyzed using the following: Frequency, Percentage distribution, Mean, Median scores, IQR (Inter Quartile Range), One-Way Analysis of Variance (ANOVA), t-Test and Pearson Coefficient. ***Results:*** In terms of quality of life, except for age, significant differences were found in terms of children's sex and educational status. Except for knowledge, there were significant differences in terms of physiological care, medication, psychological care, therapeutic communication and functional training as rated by the children and nurses. There was a significant positive correlation ($r = .583$, $p = .000$) between nursing care as rated by the children and quality of life of children with hepatolenticular degeneration. ***Conclusion:*** Nursing care is positively correlated with quality of life. A closer look at the results revealed that the quality of life of female children is generally lower than that of male, hence, more attention is recommended to be given to female children in improving their quality of life. Children with hepatolenticular degeneration should be encouraged to continue to receive education for a better quality of life. It is necessary for nurses to continuously explain to the child the disease process and the signs and symptoms of the disease. Careful observation of the physiological status of the child, the provision of timely care, regular functional training, making accurate judgements in the response to medications guidance in the intake of oral injectable medications, regular provision of psychological care and therapeutic communication to the children with this condition important tasks of the nurses. All these can lead to having these children live a better quality of life.

Keywords: hepatolenticular degeneration, nursing care, quality of life, children

1. Introduction

Hepatolenticular degeneration also known as Wilson's disease (WD), is a rare autosomal recessive inherited copper metabolic disorder which is characterized by excessive deposition of copper in the liver, brain and other tissues. Hepatolenticular degeneration is often fatal if not recognized and treated when asymptomatic. Its occurrence varies in terms of age and clinical symptoms. First, children with this condition are asymptomatic after birth and difficult to diagnose. After 6-8 years of age, for example, fatigue, loss of appetite, vomiting, edema, or ascites occur repeatedly. After the age of 12, there will be symptoms of impaired organ function, such

as difficulty constructing language, difficulty swallowing, awkward movements, involuntary movements, and changes in muscle tone. At older ages, behavioral and mental disorders occur (Sun, 2024).

Epidemiological surveys in Europe and the United States shows that the prevalence of hepatolenticular degeneration is 1 in 100,000 population, the incidence rate is 0.2 per 100,000 population. In China, the preliminary results of the epidemiological study of hepatolenticular degeneration showed that the incidence rate was approximately 2.66/100,000, and the prevalence rate was approximately 6.21/100 000. Twenty-nine thousand patients were admitted in 2015, 21 patients were from the United States, Spain, Italy, and France. The majority of cases were between 6-50 years old (Liu, 2023).

The quality of life of children with hepatolenticular degeneration is significantly lower than that of children of the same age, especially in the physiological and psychological fields, and the factors are multifaceted. Hepatolenticular degeneration patients have physical, psychological and social dysfunction, leading to a decline in their quality of life. For the treatment of children with Wilson's disease, the Institute of Neurology of Anhui University of Traditional Chinese Medicine is the only one in China that focuses on the disease (Liu, 2022). Thus, most of the current researches focus on drug treatment. Consequently, there are few studies that center on the provision of comprehensive nursing care for children with the said disease. The duration of hepatolenticular degeneration is chronically prolonged and treatment takes a long time, which affects the quality of life of children, therefore, paying close attention to the quality of care given by nurses, is of great significance for the overall improvement of children's disease condition. Hence, determining the difference of nursing care on the quality of life among children with hepatolenticular degeneration, improving their quality of life by enhancing nursing care for children with hepatolenticular degeneration as well as upgrading comprehensive nursing care for children with this disease were the reasons that motivated the researcher to undertake the study. Moreover, the experience of the researcher, having been exposed to the said hospital during his undergraduate years gave him insights in terms of care provided to the children. Hence, this finally triggered the researcher to endeavor into this study in the hope that the results would improve the quality of life of the children through the provision of nursing care (Hua, 2023).

2. Methods

2.1 Research Design

This research design used in the study was the descriptive correlational design. This study aimed to describe the variables of the study and at the same time to determine the relationship of these two quantitative variables. A descriptive study is an investigation that occurs in a completely natural condition where the researcher does not intervene and control the independent variable. In this study, the researcher sought to describe and determine relationship between the nursing care and quality of life among children with hepatolenticular degeneration.

2.2 Population and Sampling

There were two sets of population in the study, namely the children with hepatolenticular degeneration and nurses who cared for the children with the said condition. There are approximately 900 children with hepatolenticular degeneration, ages from 12 to 18 years old every year. There are more hospitalized children summer vacation than in other months in China. The summer vacation is from July to September, and all schools have no classes. Most children with hepatolenticular degeneration choose to be hospitalized at this time. A purposive sample of 96 children and 45 regular staff nurses were recruited to participate in the study. Purposive sampling is a non-probability sampling method and it occurs when elements selected for the sample are chosen by the judgment of the researcher. In the present study, children were chosen on the basis of the eligibility of potential participants for the study. The researcher recruited children with the following inclusion criteria.

(1) All hospitalized children diagnosed with hepatolenticular degeneration;

(2) Hospital stay is within 32 days or more;

(3) Age ranges from 12 to 18 years old;

(4) Children and their parents agreed to participate. The researchers explained the assent form to the child and the child understood and signed it.

(5) Admitted from July to September 2018. So that by data collection day, they had been admitted for at least 32 days.

As for nurses, the researcher recruited nurses using the inclusion criteria of:

(1) Regular staff nurses handling children with hepatolenticular degeneration employed in the hospital;

(2) Have a professional qualification certificate.

2.3 Research Locale

The researcher conducted the study at Affiliated Hospital of Institute of Neurology, Anhui University of Traditional Chinese Medicine (China Hepatolenticular Degeneration Center). Since 1973, the hospital has been the exclusive clinical treatment for patient with hepatolenticular degeneration and is the only specialist hospital to treat the disease. There are currently 400 beds that accept patients with Wilson's disease from all over China. The hospital experts have developed a relatively complete diagnosis and treatment system for hepatolenticular degeneration, in the 40-year in-depth study of hepatolenticular degeneration, including the pathogenesis, diagnosis and treatment of the disease. Patients with hepatolenticular degeneration receive long-term treatment at this hospital. The standard hospitalization day is 32 days or more, 8 days for 1 course of treatment times four cycle, 6 days of copper discharge treatment, 2 days of intermittent period with no treatment.

2.4 Instrument

Three (3) instruments were used in this study. First was the children's profile which was obtained through a Demographic Data Sheet. To measure the main variables of the study the Nursing Care Score Tool and the The PedsQL™ 4.0 Generic Core Scales (Pediatric Quality of Life Inventory™ Version 4.0) were used.

2.5 Demographic Data Sheet

The demographic data sheet was used to collect the information about children, includes age, sex and educational status.

2.6 The Nursing Care Score Tool

The Nursing Care Score Tool was used to measure nursing care. This was developed by the researcher based on the review of related literature. This is used to measure nursing care. This is a 42-item scale that consists of five (5) dimensions namely: 1) Knowledge (4 items), 2) Physiological care (8 items), 3) Medication (9 items), 4) Psychological care (9 items), 5) Therapeutic communication (5 items), 6) Functional training (7 items). A 5-point Likert response scale was utilized across all the nurses (0 = never; 1 = hardly; 2 = sometimes; 3 = often; 4 = always). The item is scored positively and converted to 0-4 points (0 = 0, 1 =1, 2 = 2, 3 = 3, 4 = 4). The total scoring ranges from 0 to 168. The higher scores indicate better nursing care. Scale Scores are computed as the sum of the items divided by the number of items answered. If more than 30% of the items in the scale are missing, the Scale Score is not computed.

2.7 Statistical Treatment

All the information obtained from the surveys was entered into SPSS for analysis. To answer question number one, the researcher used Frequency and Percentage distribution. To answer question number two, three and four, the researcher used Median scores, Inter Quartile Range and Percentage. To answer question number five, six and seven, the researcher used Mean scores, One-Way Analysis of Variance (ANOVA) or t-Test. To answer question number eight, the researcher used t-Test and Pearson Coefficient.

3. Results

Table 1. Distribution of Children according to demographic profile

Profile Variable	Frequency (n=96)	Percentage
Age		
12 – 14	39	41
15 – 16	27	28
17 – 18	30	31
Total	96	100
Sex		
Male	60	62.5
Female	36	37.5
Total	96	100
Educational Status		
Enrolled	70	73
Drop-out	26	27
Total	96	100

In Table 1, the profile distribution of the 96 children according to age, sex, and educational status is presented using frequency and percentage. In terms of age, most of the respondents (41%) have ages from 12 years old to 14 years old. There were 27 children (28%) whose ages are from 15 years old to 16 years old while the age group 17-18 years old have frequency 30 or 31% of the total.

There were more males than females with frequency of male at 60 and female at 36. In terms of educational status, 70 out of 96 (73%) are enrolled while 20 (or 27%) are drop-out. In the process of the research, the researchers found that dropouts are often older, and of course there are a few younger ones. Due to serious illness, they had to withdraw from school (GAO, 2023).

Table 2.

Overall Perceived Nursing Care	Mean	df	Stat Value	p-Value (Test Used)	Decision
Age					
12 – 14	2.628	2	1.808	.170	Do not reject null hypothesis; There is no significant difference between groups.
15 – 16	2.376				
17 – 18	2.397				
Sex					
Female	2.442	1	1.115	.294	Do not reject null hypothesis; There is no significant difference between groups.
Male	2.465				
Education Status		1	1.050	.308	Do not reject null hypothesis; There is no significant difference between groups.

In Table 2, the comparison between age groups yielded a p-value of 0.170, which is greater than 0.05. Thus, the null hypothesis was not rejected and the concluded that there was no significant difference in the overall quality of nursing care as rated by children grouped according to age.

Moreover, comparison between females and males yielded a p-value of 0.294, which is greater than 0.05. Thus, the null hypothesis was not rejected and the researcher concluded that there is no significant difference in the overall quality of nursing care as rated by children grouped according to sex.

Finally, the comparison between education status yielded a p-value of 0. 308. Thus, the null hypothesis was not rejected and we concluded that there was no significant difference in the overall quality of nursing care as rated by children grouped according to education status.

There was no significant difference in the perceived nursing care as rated by the children with hepatolenticular degeneration when grouped according to their profile variables in terms of age, sex and educational status. There was no significant difference in nursing care for children in this age group, and the quality of nursing care received between male and female children is the same. There was no significant difference between the children who are enrolled in school and drop-out of school. It can be seen that during the nursing care process, the nurses treat all the children equally, and there was no significant difference.

Table 3.

Nursing Care	Mean	df	Stat Value	p-Value	Decision
Knowledge					Do not reject null hypothesis; there is no significant difference.
Children	2.3672	70.979	-1.682	.097	
Nurses	2.5944				
Physiological care					Reject null hypothesis; there is a significant difference.
Children	2.5052	111.827	-5.098	.000	
Nurses	2.8489				
Medication					Reject null hypothesis; there is a significant difference.
Children	2.8419	114.122	-9.472	.000	
Nurses	3.3720				

Psychological care					Reject null; there is a significant
Children	2.2293	96.768	-12.304	.000	difference.
Nurses	3.2344				
Therapeutic					Reject null hypothesis; there is a
communication		79.786	-8.013	.000	significant difference.
Children	2.0417				
Nurses	2.8978				
Functional training					Reject null hypothesis; there is a
Children	2.3782	58.746	2.741	.008	significant difference.
Nurses	2.6919				
Overall					Reject null hypothesis; there is a
		113.370	-10.669	.000	significant difference.

Note that in Table 3, the comparison between children and nurses showed significance in terms of physiological care (p-value<0.005), medication (p-value<0.005), psychological care (p-value<0.005), therapeutic communication (p-value<0.005), and functional training (p-value=0.008). The overall nursing care rating comparison also yielded significant difference between nurses and children (p-value<0.005). Only the comparison in terms of knowledge showed no significant difference (p-value=0.097).

It is noted that in all the different aspects of nursing care, including the overall ratings, nurses rated themselves higher compared to the ratings given by the children.

Hepatolenticular degeneration is an autosomal recessive inherited copper metabolic disorder, the pathological mechanism is very complicated, and even if the nurse explains the child and his parents, it is difficult to understand the disease. Therefore, when the nurse explains the disease, the child is more likely to accept it, agreeing with the nurse, there is no significant difference. There are obvious differences in the physical care, drug guidance, psychological care, therapeutic communication, and functional training provided by nurses. This will explain the difference in the quality of care to a certain extent. When the nurses are taking care, they pay attention to whether the children are concerned. Can be fully accepted, can the implementation of the care to be complete, consistent with the real situation of the child.

Table 4.

Quality of Life	Nursing Care	Pearson Coefficient	Interpretation	p-Value	Decision
Physiological Function	Knowledge	0.089	Small	0.388	There is no a significant correlation
	Physiological Care	0.375	Moderate	0.000	There is a significant correlation
	Medication	0.274	Small	0.007	There is a significant correlation
	Psychological Care	0.409	Moderate	0.000	There is a significant correlation
	Therapeutic Communication	0.393	Moderate	0.000	There is a significant correlation
	Functional Training	0.493	Moderate	0.000	There is a significant correlation

In Table 4, all the nursing care dimensions, except for knowledge, showed a significant positive linear relationship with physiological function. This means that as the level of nursing care in terms of physiological care, medication, psychological care and therapeutic communication functional training increases, the level of quality of life in terms of physiological function also increases.

Explain to the child what is hepatolenticular degeneration, more is the psychological comfort of the child, the disease can be restored early treatment, is conducive to psychological function. Give children more physiological care and functional training, can improve the body movements of children and increase physiological functions.

Medication guidance can effectively eliminate copper in children and improve physiological function. Give children more psychological care and therapeutic communication, can increase the clarity of treatment goals and confidence in recovery (Huang, 2023).

Studies have shown that aerobic exercise and muscle training can improve children's mobility and promote recovery of limb function, thereby improving quality of life.

Table 5.

Quality of Life	Nursing Care	Pearson Coefficient	Interpretation	p-Value	Decision
Emotional Function	Knowledge	0.491	Moderate	0.000	There is a significant correlation
	Physiological Care	0.394	Moderate	0.000	There is a significant correlation
	Medication	0.291	Small	0.004	There is a significant correlation
	Psychological Care	0.407	Moderate	0.000	There is a significant correlation
	Therapeutic Communication	0.367	Moderate	0.000	There is a significant correlation
	Functional Training	0.096	Small	0.350	There is no a significant correlation

In Table 5, all the nursing care dimensions, except for functional training, showed a significant positive linear relationship with emotional function. This means that as the level of nursing care in terms of knowledge, physiological care, medication, psychological care and therapeutic communication increases, the level of quality of life in terms of emotional function also increases.

Functional training begins in the afternoon, when some children receive drug treatment, they often show impatience with treatment and reduce their emotional function. Give children more knowledge, physiological care, psychological care and therapeutic communication, can improve the body movements of children and increase physiological functions. Being able to increase the child's understanding of the disease can be cured, build confidence to restore health, and benefit emotional function (Yao, 2024).

Table 6.

Quality of Life	Nursing Care	Pearson Coefficient	Interpretation	p-Value	Decision
Social Function	Knowledge	0.484	Moderate	0.000	There is a significant correlation
	Physiological Care	0.392	Moderate	0.000	There is a significant correlation
	Medication	0.303	Moderate	0.003	There is a significant correlation
	Psychological Care	0.404	Moderate	0.000	There is a significant correlation
	Therapeutic Communication	0.34	Moderate	0.001	There is a significant correlation
	Functional Training	0.082	Small	0.429	There is no a significant correlation

In Table 6, all the nursing care dimensions, except for functional training, showed a significant positive linear relationship with social function. This means that as the level of nursing care in terms of knowledge, physiological care, medication, psychological care and therapeutic communication increases, the level of quality of life in terms of social function also increases.

Give children more knowledge, physiological care, medication, psychological care and therapeutic communication, can improve the body movements of children and increase hope for disease recovery, benefit for social function.

The decrease in social support and the estrangement of the surrounding environment will often increase the psychological burden on patients with movement disorders, and thus affect their quality of life (Hua, 2024). The

mental health status of hepatolenticular degeneration patients is related to the social support of patients; while strengthening the material support for patients, it is more important to pay attention to the emotional support of patients (Yan, 2024).

Table 7.

	Pearson Coefficient	Interpretation	p-Value	Decision
Nursing Care	0.583	Strong	0.000	There is a significant correlation

The overall correlation between quality of life and nursing care is presented in Table 8.4. It is noted that the p-value is found to be less than 0.005. Thus, there was a significant positive correlation between quality of life and nursing care. This means that as the level of nursing care increases, the level of quality of life also increases.

4. Conclusion

Nursing care provided to children with hepatolenticular degeneration is correlated with their quality of life. There is a strong positive relationship between nursing care and quality of life. Hence, when nursing care is improved or increased the quality of life also increases.

Nursing care is positively correlated with quality of life. A closer look at the results revealed that the quality of life of female children is generally lower than that of male, hence, more attention is recommended to be given to female children in improving their quality of life. Children with hepatolenticular degeneration should be encouraged to continue to receive education for a better quality of life. It is necessary for nurses to continuously explain to the child the disease process and the signs and symptoms of the disease. Careful observation of the physiological status of the child, the provision of timely care, regular functional training, making accurate judgements in the response to medications guidance in the intake of oral injectable medications, regular provision of psychological care and therapeutic communication to the children with this condition important tasks of the nurses. All these can lead to having these children live a better quality of life.

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