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The Effect of an Instructional Program on Mothers' Knowledge and Practices Regarding Care of Their Children with Maple Syrup Urine Disease

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Abstract:

Background: Maple syrup urine disease (MSUD) is an uncommon genetic autosomal recessive neurometabolic disorder. This disorder is identified by the burnt sweet scent of the urine of affected infants. **Aim**: the current study aimed to evaluate the effect of an instructional program on mothers' knowledge and practices regarding care for their children with maple syrup urine disease. **Design**: A quasi-experimental study design (pre-posttest) was used to fulfill the aim of the study. **Setting**: The study was conducted at the Genetic Outpatient Clinic in Mustafa Hassan Pediatric Hospital, is affiliated to Fayoum University Hospitals, and Abo El Rish (El Monira) Hospital affiliated to Cairo University Hospitals. **Sample** A purposive sample consisted of 71 of studied mothers who had children with maple syrup urine disease. **Tools**: Two tools were used to collect data in this study: **Tool (I)**: Structured interview questionnaire sheet to assess mothers' knowledge regarding maple syrup urine disease (pre-post). **Tool (II)**: Observational checklist to assess mothers' reported practices regarding care of their children with maple syrup urine disease (pre-post). **Results**: The study result shows that,70.4% of studied mothers had unsatisfactory level of total knowledge pre-instructional program compared to the 88.7% of them who had satisfactory level of total knowledge post instructional program, 67.7% of studied mothers had inadequate self-reported practice pre instructional program , compared to 90.1% of them who had adequate self-reported practice post instructional program and it there was a positive strong statistical significance correlation between studied mothers' knowledge and practices concerning maple syrup urine disease pre and post instructional program as well as. **Conclusion**: Based upon the results of the current study, it can be concluded that after implementation the instructional program mothers' knowledge and practices regarding care of their children with maple syrup urine disease are improved than before. **Recommendation**: Continuous health education programs to mothers' regarding maple syrup urine disease, complications, different aspects of care and management plan.

Keywords: Instructional Program, maple syrup urine disease, mothes knowledge, practices, children.

Introduction

Maple syrup urine disease (MSUD) "also called" Branched Chain Ketoaciduria is a rare inherited autosomal recessive neurometabolic disorder. It is characterized by impaired metabolism of branched-chain amino acids, which is caused by deficiency of branched chain α-ketoacid dehydrogenase enzymes complex. This leads to accumulation of branched chain amino acids includes leucine, isoleucine, and valine as well as toxic products (ketoacids) in blood, urine, and cerebrospinal fluid (**Amaral & Wajner, 2022).**

Maple syrup urine disease is a genetic disorder that affects approximately 1 in 185,000 infants worldwide. The prevalence of pediatric MSUD globally ranges from 1 to 9 cases per 1,000,000 individuals. This disorder is more commonly found in populations with a higher rate of consanguinity, such as the Mennonites in Pennsylvania, where the incidence reaches as high as 1 in 200 births. The prevalence of MSUD in Egypt was determined to be 1 in every 25,276 healthy infants and 17 in 3900 high-risk newborns (**Ali et al., 2021& Dahpy, et al., 2021)**.

Newborn screening is conducted in various countries to detect most infants with MSUD. In places where screening is not accessible, infants with MSUD often show progressing neurological symptoms. Early identification and treatment help to stabilize newborns and, when done effectively and consistently, can greatly reduce the risk of severe long-term issues **(Strauss et., 2020).**

The primary and effective methods for treatment of MSUD consist of a life-long strict semisynthetic diet to prevent the buildup of harmful metabolites. Other objectives of treatment include supporting normal physical growth, maintaining proper nutrition, and avoiding catabolism (**Ballikaya et al., 2021)**.

Mothers can play an important role in management and care child with maple syrup urine disease which needs daily effort to deal with the child needs, child disabilities, and to maintain a special rehabilitation and diet program ***(Harris et al., 2017)***. The pediatric nurses has a vital role toward help parents to gain knowledge and confidence in their abilities to care for their child through giving him appropriate guidance and positive reinforcement and help them to be aware about the dietary requirements, and daily activities should be done for the child ***(Maier& Dokoupil, 2022).***

A program was created to help mothers enhance their knowledge and skills in caring for their children with MSUD. It is crucial for mothers to stay informed about the appropriate diet and treatment advancements for children with MSUD as they play a critical role in managing the condition. Mothers serve as the main healthcare providers in their homes and dedicate a significant amount of time to ensuring the well-being of their children. Hence, every mother with a child affected by MSUD needs ongoing education, expertise, and assistance to provide optimal care **(Yamaguchi et al., 2017).**

Significance of the study

Maple syrup urine disease is classified as a metabolic disorder. The screening test should be conducted within a week of birth. In Egypt, this screening is not regularly performed and is often discovered late once signs and symptoms appear. As a result, the number of pediatric patients with MSUD is on the rise. Mothers play an important role in the management and care of children with Maple syrup urine disease by providing daily attention to the child's needs, disabilities, and adherence to a specialized rehabilitation and diet plan (**Harris et al., 2017)**.

AIM OF STUDY:

This study aimed to evaluate the effect of an instructional program on mothers' knowledge and practices regarding caring of their children with maple syrup urine disease through:

1. Assess the mothers' knowledge and reported practices regarding care of their children with Maple Syrup Urine Disease.
2. Develop and implement instructional program for mothers' regarding care of their children with Maple Syrup Urine Disease.
3. Evaluate the effect of instructional program for mothers regarding care of their children with Maple Syrup Urine Disease.

Research hypothesis:

The implementation of instructional program will affect positively on knowledge and practices of mothers have children with maple syrup urine disease.

SUBJECTS AND METHODS

I- Technical Design:

Technical design for this study included a description of the research design, setting, subjects, and tools of data collection.

Research design:

A Quasi-experimental research design was utilized to fulfill the aim of this study.

Research setting:

This study conducted at the Genetic Outpatient Clinic in Mustafa Hassan Pediatric Hospital, affiliated to Fayoum University Hospitals and Abo El Rish (El Monira) Hospital, affiliated to Cairo University Hospitals.

Sampling:

A purposive sample composed of 71 of studied mothers who had children with MSUD was used in this study based on the following inclusive and exclusive criteria:

Inclusion criteria: All children who has been officially diagnosed with maple syrup urine disease and are between the ages of one month to 9 years, along with their mothers, regardless the mothers' age, location of residence, or socioeconomic status, are eligible. **Exclusion criteria:** The study did not include children with any other physical or mental disease.

Tools for data collection:

Two tools were used for gathering data, which were created by the researcher in plain Arabic language following a review of relevant literature and approval from the researcher's supervisors. These tools included the following:

Tool (I) Pre-designed Interviewing Questionnaire Sheet: This consisted of four parts:

First Part:

Characteristics of the studied mothers include: age, educational level, occupation, residence and marital status.

Characteristics of the studied children include: age, gender educational grade, and ranking.

Second Part:

Past Medical history and family history of children with MSUD **(12 items)** it included: duration of disease, way of discovery the disease, manifestation appeared on child and time its appeared, previous hospitalization, complication of disease, regular follow up, frequency per month, regular investigation, and family history which contained: family history of MSUD, degree of relation to the child, parents Consanguinity degree of Consanguinity.

Third Part:

Assess child physical growth **(4 items)** weight, height, head circumference and BMI, assess daily needs **(7 items)** need for: attention, encouragement, safe, love, educational qualification, educational programs, and assessment of child lab investigation levels: **(5 items)** leucien, isoluecin, valine, CBC and acetone in urine.

Fourth Part (pre/posttest):

- Studied mothers' knowledge regarding Maple Syrup Urine Disease which consisted of 6 Questions multiple choice about definition, causes, signs and symptoms.
- Mothers' role in providing care for her child with Maple Syrup Urine Disease which consisted of 16 Questions multiple choice about feeding, diet, medication and physical exercise**.**

Scoring system:

Knowledge of mothers was scored and calculated according to their answers, it was evaluated using the model key answers sheet that was prepared by the researcher. Each question had a score ranged from 0-1 grades, whereas, correct answer had score one grade and score zero was given for an incorrect or unknown answer. The total score was 22 grades (equal 100%). The total scores converted in to percentage then categorized as following:

- Satisfactory level of knowledge: ≥ 60% which represent (25 grade and more).
- Unsatisfactory level of knowledge:< 60% which represent less than (25 grade).

Tool (II) Observation checklist pre/posttest.

It was adapted from various sources, including **Bowden & Greenberg (2017), Hockenberry and Wilson (2021), Chapman et al., (2018) and the National Center for Inherited Metabolic Disorders (2014)** to suit the nature of study and was reviewed by supervisors to evaluate mothers' reported practices in caring their children with MSUD. It consisted of nine procedures, which were categorized as follows: NGT feeding (10 steps), diet measurement (6 steps), dental health care (4 steps), weight measurement (6 steps), physical exercise and therapy (6 steps), urine analysis (6 steps), and daily mother observation (5 items).

Scoring system

The observational checklist assigned a score of one for each item done correctly and zero for not done items. There were a total of 42 steps in the checklist, resulting in a possible total score of 42, equivalent to 100%. The scoring system for mothers' practice was categorized as follows:

- Adequately level of practice: ≥ 60% which represent (50 grade and more).
- Inadequately level of practice : < 60% which represent less than (50 grade).

II- Operational Design:

The Operational design included the preparatory phase, validity, reliability, ethical consideration, pilot study, and fieldwork.

Preparatory phase:

An extensive review of recent, current, national and international related literature in various aspects of the problems was done to design the study tools and to be acquainted with various aspects of the problems.

Content validity:

In order to ensure the trustworthiness of the data collection tool used in this study, the tools were assessed for content validity by three professors specializing in pediatric health nursing from Ain-Shams, Helwan, and Fayoum universities. They evaluated the tools for clarity, relevance, comprehensiveness, simplicity, and applicability.

Reliability of tool:

The reliability of the tool was assessed across all its components. The determination of internal consistency was carried out using the Cronbach's alpha coefficient test. This assessment comprised the following elements:

Items	Cronbach's alpha coefficients
Reliability for knowledge	0.845
Reliability for practices	0.813

Ethical Considerations:

Ethical approval was obtained from the Scientific Ethical Committee Research, Faculty of Nursing, - Helwan University, after submitting a proposal for the research and examining all papers by the concerned committee. Then the purpose and nature of the study were explained to the participants and oral permission were taken from the mothers and informed that each study subject is free to withdraw at any time through the study without giving any reasons.

Pilot study:

The pilot study was carried out on 10% of the participants in the research, which included 7 children and their mothers who met the sample criteria. The aim was to assess the clarity of questions and the time required to complete the study tools. After analyzing the results of the pilot study, no changes were made. The participants in the pilot study were also included in the overall study sample.

Field Work:

The data collection process took place over a period of eight months, from November 2022 to June 2023, with the researcher being present during morning shifts. The researcher was available three days a week, from 9 am to 12 pm, at previously mentioned settings. Initially the researcher conducted a pre-test to assess both of knowledge and reported practices followed by delivering an instructional program for mothers, consisting of seven sessions lasting about an hour each. Prior to each session, there was an open discussion to address any questions and provide a brief summary of the previous session's content. At the conclusion of each session, the researcher summarized key points and ensured that the mothers understood the information presented.

III- Administration Design:

A formal written communication letter sent from Dean the Faculty of Nursing at Helwan University to the administrators of the educational institutions where the study was conducted. The letter was intended to facilitate the implementation of the research.

IV- Statistical Design:

The data obtained from the sample under study was meticulously analyzed and organized using Statistical Package for the Social Sciences (SPSS) version 20. Quantitative data was shown using numbers and percentages. The statistical analyses included chi-square tests, mean calculations, standard deviation measurements, and correlation tests, all revealing high internal consistency and construct validity.

RESULTS

Table (1): Distribution of studied children according to their characteristics **(n=71)**.

Item	N	%
Child age/years		
1 month <1 year	34	**47.9**
1<3 years	22	31.0
3<6 years	10	14.1
6<9 years	5	7.0
Mean ±SD	**3.84±2.65**	
Gender		
Male	45	**63.4**
Female	26	36.6
Ranking		
First	20	28.2
Second	36	**50.7**
third and more	15	21.1
Level of education		
Not attending school	53	74.6
Preschool	11	15.5
primary	7	9.9

Table (1): shows that less than half (47.9%) of the studied children were under one year old, with an average age of 3.84±2.65 years, and approximately two-thirds of them were male (63.4%). The data also indicates that about half (50.7%) of the studied children were the second child in their family.

Table (2): Number and percentage distribution of studied mothers according to their characteristics **(N=71).**

Item	N	%
Age /years		
< 20	4	5.6
20 < 25	12	16.9
25 < 30	28	**39.4**
30 < 35	17	23.9
≥35	10	14.1
Mean ±SD	**33.24±5.16**	
Education level		
Illiterate	16	22.5
Primary	21	29.6
Preparatory	11	15.5
Secondary	14	19.7
University	9	12.7
Occupation		
House wife	47	**66.2**
Working	24	33.8
Marital Status		
Married	61	85.9
Divorced\ Widow	10	14.1
Place of residence		
Rural	55	**77.5**
Urban	16	22.5

Table (2): clarifies that more than one third (39.4%) of studied mothers their age ranged from 25 < 30 years with mean age **33.24±5.16** years and about two thirds (66.2%) of studied mothers were housewives. The same table illustrate that more than three quarter (77.5%) of studied mothers lived in rural areas.

Table (3): Distribution of the studied mothers total level of knowledge regarding care of their children with maple syrup urine disease (pre-post instructional program implementation) n=(71).

Total knowledge	Pre		Post		Chi-square	
	N	%	N	%	X^2	P-value
Satisfactory	21	29.6	63	**88.7**	51.414	<0.001*
Unsatisfactory	50	**70.4**	8	11.3		
Mean ± SD	**8.78±4.22**		**18.6±3.45**			

Fig (1): Distribution of the studied mother's total level of knowledge regarding Maple Syrup Urine Disease (pre-post instructional program implementation) n=(71).

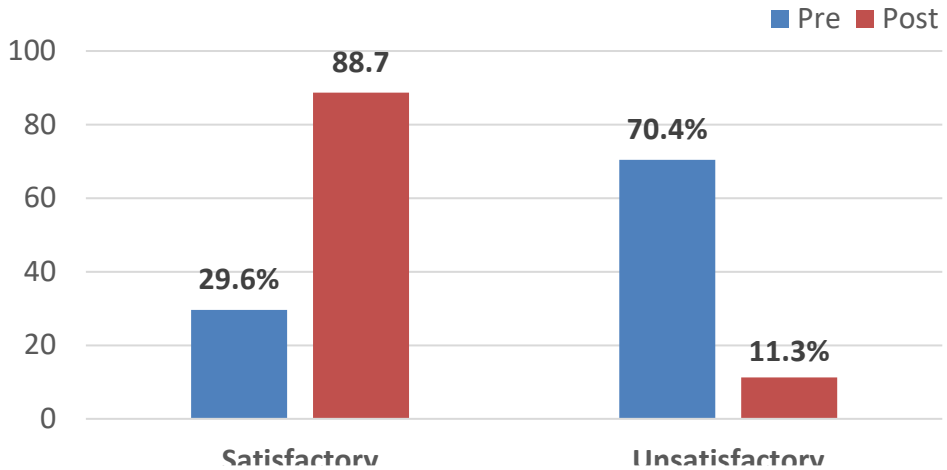

Table (3) fig (1): Reveals that less than three quarter (70.4%) of studied mothers had unsatisfactory level of total knowledge regarding MSUD in pre instructional program compared to the majority (88.7%) of them who had satisfactory level of total knowledge post instructional program implementation.

Table (4): Distribution of the studied mothers regarding total level of self-reported practices regarding care of their children with maple syrup urine disease (pre-post instructional program implementation) N=**(71).**

Total practice	Pre		Post		Chi-square	
	N	%	N	%	X^2	P-value
Adequately done	23	32.4	64	**90.1**	49.885	**<0.001***
Inadequately done	48	**67.6**	7	9.86		
Mean ± SD	**14.06±4.31**		**32.67±5.34**			

Fig (2): Distribution of the studied mothers regarding total level of self-reported practices regarding care of their children with maple syrup urine disease (pre-post instructional program implementation) n=(71).

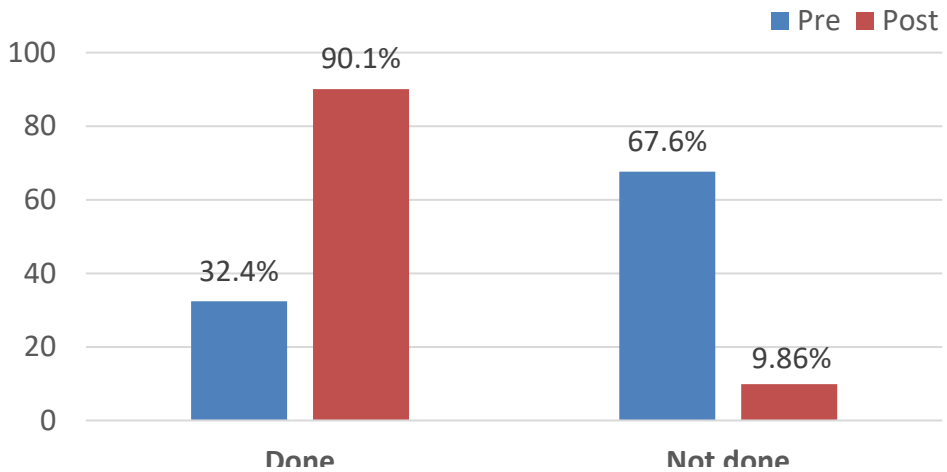

Table (4), fig (2): illustrates that more than two thirds (67.7%) of studied mothers had inadequate self-reported practice pre instructional program, compared to the most (90.1%) of them who had adequate self-reported practices post instructional program implementation.

Table (5): Relationship between studied mothers total level of knowledge and their characteristics pre and post instructional program implementation (n=71).

Items	Knowledge score							
	Pre		Tests		Post		Tests	
	Mean	SD	t/f	P-value	Mean	SD	t/f	P-value
Age/years								
< 20	5.00	2.00	30.410	<0.001*	10.75	2.87	8.404	<0.001*
20 < 25	4.75	1.06			12.50	2.20		
25 < 30	7.22	2.39			12.33	2.22		
30 < 35	11.12	3.79			14.47	3.87		
≥35	14.82	1.94			17.64	3.29		
Education level								
Illiterate	4.81	1.28	45.937	<0.001*	12.06	2.41	11.651	<0.001*
Primary	6.67	2.33			12.24	2.12		
Preparatory	8.55	1.75			12.00	2.28		
Secondary	12.57	3.25			15.93	3.60		
University	15.22	1.92			17.89	3.41		
Occupation								
House wife	6.45	2.35	10.371	<0.001*	12.11	2.23	6.413	<0.001*
Working	13.38	3.20			16.54	3.59		
Marital Status								
Married	14.20	1.87	5.081	<0.001*	17.20	3.29	3.887	<0.001*
Divorced\ Widow	7.90	3.83			13.02	3.13		

Items	Knowledge score							
	Pre		Tests		Post		Tests	
	Mean	SD	t/f	P-value	Mean	SD	t/f	P-value
Place of residence								
Rural	4.81	1.28	4.935	<0.001*	12.06	2.41	2.076	0.042*
Urban	9.95	4.08			14.05	3.60		

Table (5): This table demonstrates a significant relationship between the studied mothers total level of knowledge and their demographic characteristics pre and post instructional program implementation. These characteristics include age groups, level of education, occupation, marital status, and place of residence.

Table (6): Relationship between studied mothers total level of reported practices and their characteristics pre and post instructional program implementation (n=71).

Items	Practice score							
	Pre		Tests		Post		Tests	
	Mean	SD	t/f	P-value	Mean	SD	t/f	P-value
Age								
< 20	9.75	0.50	13.432	<0.001*	22.25	0.96	14.946	<0.001*
20 < 25	10.25	2.49			24.50	2.81		
25 < 30	11.56	3.46			25.41	3.89		
30 < 35	14.65	4.47			30.71	5.35		
35 and more	18.64	1.36			33.82	3.28		
Education level								
Illiterate	10.13	2.16	14.816	<0.001*	23.94	2.64	14.389	<0.001*
Primary	11.90	3.45			25.24	3.88		
Preparatory	11.27	3.07			27.18	4.92		
Secondary	15.86	4.55			32.14	4.97		
University	18.89	1.36			33.44	3.47		
Occupation								
House wife	11.06	2.97	7.211	<0.001*	25.28	3.94	6.665	<0.001*
Working	17.00	3.82			32.29	4.66		
Marital Status								
Married	18.80	1.03	5.352	<0.001*	33.30	3.23	3.974	<0.001*
Divorced\ Widow	12.13	3.90			26.72	5.05		
Place of residence								
Rural	10.13	2.16	3.317	<0.001*	23.94	2.64	3.385	<0.001*
Urban	13.93	4.42			28.73	5.46		

Table (6): This table illustrates statistically significant relationship between the studied mothers total level of self-reported practices and demographic characteristics pre and post instructional program implementation. These characteristics include age groups, level of education, occupation, marital status, and place of residence.

Table (7): Correlation between studied mothers' total level of knowledge and reported practices pre and post instructional program implementation (n=71).

Practice score	Knowledge score	
	R	P-value
Pre	0.733	<0.001*
Post	0.652	

Fig (3-4): Correlation between studied mothers' total level of knowledge and reported practices pre and post instructional program implementation (n=71).

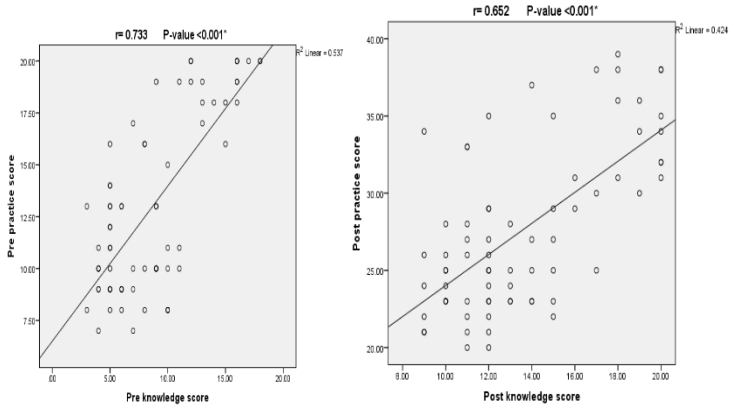

Table (7) fig (3-4): This table and figures clarifies that there was a strong positive correlations between the scores for studied mothers' knowledge and practices, with a correlation coefficient of 0.733 for pre-test and 0.652 for post-test, both of which have a p-value less than 0.001.

DISCUSSION

Maple syrup urine disease (MSUD) is a metabolic condition marked by challenges in metabolizing proteins essential for proper growth and development. The management of children with MSUD necessitates meticulous regulation of the intake of detrimental branched-chain amino acids (BCAAs) to ensure optimal growth. The primary approach to treating MSUD involves adhering to a lifelong strict semi-synthetic diet to mitigate the buildup of harmful metabolites. Additional objectives in managing this condition encompass preserving typical

physical growth and nutritional well-being, as well as preventing catabolism (**Alrige et al., 2023 & Ballikaya et al., 2021)**.

Part I: Characteristics of the studied children.

Regarding the characteristics of the study children, the results of the present study showed that less than half of the studied children were under one year old, with an average age of 3.84±2.65 years. These results were contrasted by **Mostafa and Mohammed (2017**), who examined the "Educational Program for Nurses Regarding Management of Children with Maple Syrup Urine Disease during Acute Intermittent Late-Onset" and reported that the studied children were between the ages of less than one year and were less than 10 years old with a mean of 6.98+2.28 years.

Regarding the gender of the children under study, the results showed that around two thirds of the children were male. This finding was consistent with the finding of **Ballikaya et al., (2021)** who found in a study titled "Oral health status of children and young adults with maple syrup disease in Turkey" that, two thirds of children under study were males. From the researcher's point of view, it is important to note that MSUD is inherited in an autosomal recessive manner, impacting both male and female equally. From the researcher's perspective, MSUD is inherited in an autosomal recessive manner, affecting males and females equally.

Regarding characteristics of studied mothers, the finding of current study clarified that, more than one third of studied mothers were aged between 25 < 30 with an average age of **33.24±5.16** years, and two thirds of them were housewives. This finding is corresponding with **Madhu et al., (2017),** who conducted study about "breastfeeding practices and newborn care in rural areas in Indian" and found that the majority of the mothers were between the ages of 21 and 25 years old, and more than half of them were housewives. The findings of the current study were consistent with **Hadush et al., (2016),** who carried out a study about assessment knowledge and practice of neonatal care among postnatal mothers attending in Ayder and Mekelle Hospital in Tigray, Ethiopia, and found that, the mean age of studied mothers was **27.04±5.9** years.

Part II: Studied mothers' knowledge about MSUD:

Concerning distribution of the studied mothers regarding total level of knowledge about Maple Syrup Urine Disease pre-post instructional program, the present study revealed that less than three quarter of studied mothers achieved unsatisfactory level of total knowledge regarding MSUD pre-instructional program compared to the majority of them who had achieved

satisfactory level of total knowledge post-instructional program. These findings were similar to **Ramadan et al., (2023)** who studied "Effect of Online Education on Mothers' Knowledge and Practice regarding Caring for Children with Phenylketonuria" and reported that majority of the studied mothers had unsatisfactory level of total knowledge pre-online education while the majority of them had satisfactory knowledge post-online education with significant improvement.

Part III: Studied mothers' reported practices about MSUD:

As regard distribution of the studied mothers towards total level of self-reported practices about Maple Syrup Urine Disease, the current study result illustrated that more than two thirds of studied mothers had inadequate self-reported practices pre- instructional program, compared to the most of them who had adequate self-reported practice post-instructional program with a highly statistical significant relationship pre/post-instructional program. The result of this study could be attributed to a lack of comprehensive understanding among mothers regarding the condition of their children, resulting in substandard caregiving practices for children with Maple Syrup Urine Disease and a heightened focus on their nurturing responsibilities.

This finding of this study was in agreement with that of **Khalil et al., (2023)** who studied "Effect of Educational Program on Mothers' Practices towards Caring for their Children Having Phenylketonuria" and noticed that high percentage of the studied mothers had poor practice level pretest, meanwhile level of reported practice improved posttest. Additionally, there was a high statistically significant relation in the total level of practice among studied mothers pre-posttest.

Part V: Relationships between studied mothers' total level of knowledge, practices and their characteristics

Regarding relationship between knowledge score of studied mothers and their characteristics pre and post instructional program, demonstrates a significant relationship between knowledge scores and various demographic factors before and after the instructional program. These factors include age groups, level of education, occupation, marital status, and place of residence. This result aligns with **Maheen et al., (2018)**, who studied "Assessing Parental Knowledge bout Thalassemia in a Thalassemia Center of Karachi, Pakistan" and observed a positive relationship between mothers' knowledge about splenomegaly and their age as well as it was found in study done by **Kumar and Pujari (2020),** who noted in their study about" Knowledge, attitude,

practices among parents of β thalassemia children regarding thalassemia a that there was a positive relationship between mothers' knowledge about splenomegaly and their education level. Concerning relationship between practice score of studied mothers and their characteristics pre and post instructional program. This result illustrates statistically significant relationship in practice scores pre and post the instructional program across various demographic characteristics as: age groups, education level, occupations, marital status and places of residence. This result was in accordance with **Khalil et al., (2023)** who studied "Effect of Educational Program on Mothers' Practices towards Caring for their Children Having Phenylketonuria" and noticed that there were statistically significance relations in practice scores and their characteristics mothers' high education and working had high practice score.

Part IV: Correlation between studied mothers' total level of Knowledge and their level of Practices:

Regarding correlation between practices level and knowledge level pre and post-instructional program, the findings of current study revealed a significance positive correlation between participants practice and knowledge scores. This result supported by **Adel Hussien et al., (2019)** in a study entitled "Mothers' knowledge and Practices toward Their Children Suffering from Juvenile Diabetes: an Assessment Study" and revealed that there was a positive correlation between total knowledge of the studied mothers and their total reported practices.

CONCULSION

Based upon the results of the current study, it can be concluded that after implementation the an instructional program mothers' knowledge and practices regarding care of their children with maple syrup urine disease are improved than before.

RECOMMENDATION

Based on the findings of the current study, the following recommendations can be suggested:

1- Continuous health education to mothers regarding Maple Syrup Urine Disease, complications, different aspects of care and management plan.

2- Developing a training program should be carried out so that mothers can update their knowledge and improve their practices.

3- Early screening for all newborns which helps to detect disease early, treat early and prevent complications.

4- Increase awareness of mothers about preventive strategies such as genetic counseling, premarital screening and prenatal screening to reduce the incidence of MSUD.

5- Providing a rehabilitation programs for children with MSUD to improve health status and prevent disabilities.

REFERENCES

1) Adel Hussien, H., saleh, S., R. Tantawi, H. (2019). 'Mothers ' knowledge and Practices toward Their Children Suffering from Juvenile Diabetes: an Assessment Study', Egyptian Journal of Health Care, 10(2), pp. 190-200. doi: 10.21608/ejhc.2019.36696
2) Ali, H., Elsharkay, S., Nassar, A. and Zaki, O. (2021) :Assessment of Mothers Care Towards Their Children Suffering from Branched Chain Keto- Acidurua. Un published Master degree, faculty of nursing, Ain Shams University. Pp (135).
3) Alrige, M., Banjar, H., Shuaib, T., Ahmed, A. and Gharbawi, R. (2023): Knowledge-Based Dietary Intake Recommendations of Nutrients for Pediatric Patients with Maple Syrup Urine Disease. Healthcare, 11(3):301
4) Amaral, U. and Wajner, M. (2022): Pathophysiology of Maple Syrup urine disease: Focus on the Neurotoxic Role of The accumulated Branched-chain Amino Acids and Branched-chain α-Keto Acids. Neurochemistry International, doi: 10.1016/j.neuint.2022.105360.
5) Ballikaya, E., Yildiz, Y., Koç, N., Tokatli, A., Uzamis, M. and Sivri H. 2021. Oral Health Status of Children and Young Adults with Maple Syrup Urine Disease in Turkey. BMC Oral Health. 21(1):1-9
6) Bowden, V. and Greenberg, C. (2017): Pediatric Nursing Procedure. 3rded Wolter Kluwer and Lippincott Willams & Wilkins Comp, Philadelphia. Pp. 612-618.
7) Chapman, K., Gramer, G., Viall, S. and Summar, M. (2018). Incidence of maple syrup urine disease, propionic acidemia, and methylmalonic aciduria from newborn screening data. Molecular genetics and metabolism reports, Vol.15, doi: 10.1016/j.ymgmr.2018.03.011. PP:106-109.

8) Dahpy, M., Saleem, T., El-Asheer, O., Abd EL-Rasoul, A. and Elgeit, A. (2021). Clinical, Biochemical, Molecular, and Therapeutic Analysis of Maple Syrup Urine Disease in Upper Egypt. Journal of Pediatric Genetics, 10(2):116-125.

9) Elsayed, A., Mohamed, I., AlRafay, S. and Khalifa, O. (2020): Assessment of Mothers Care Toward their Children Having Phenylketonuria. Egyptian Journal of Health Care, 11(2):241-254.

10) Hadush, A., Mekuria, K., Kalayou, K. and Weyzer, T. (2016): Assessment of Knowledge and Practice of Neonatal Care among Post Natal Mothers Attending in Ayder and Mekelle Hospital in Mekelle, Tigray. Ethiopia SMU Medical journal, 3(1):509–524.

11) Harris-Haman, P., Brown, L., Massey, S. and Ramamoorthy, S. (2017). Implications of Maple Syrup Urine Disease in Newborns. Nursing For Women's Health, 21(3):196-206.

12) Hockenberry, M. and Wilson, D. (2021). Wong's Clinical Manual of Pediatric Nursing. 10th ed., Mosby Comp, London: PP 652-658.

13) Hussien, A., Saleh, S. and Tantawi, H. (2019): Mothers' knowledge and Practices toward Their Children Suffering from Juvenile Diabetes: an Assessment Study, Egyptian Journal of Health Care, 10(2):190-200

14) Khalil, A., Amin, E., Salah Eldin, S. and Ali, O. 2023. Effect of Educational Program on Mothers' Practices towards Caring for their Children Having Phenylketonuria. Egyptian Journal of Health Care, 14(2):172-182

15) Madhu, K., Chowdary, S. and Masthi, R. (2017): Breastfeeding practices and newborn care in rural areas: a descriptive cross-sectional study. Indian J Community Med. 34(3):243–246.

16) Maier, E. M., & Dokoupil, K. (2022). 3.19 Inborn Errors of Metabolism. Pediatric Nutrition in Practice, 124, 351-361.

17) Magor, N., Shalabi, S., Mohamed, Y., Attia, R. and Ayed, M. (2023): Effect of Online Education on Mothers' Knowledge and Practice regarding Caring for Children with Phenylketonuria. Egyptian Journal of Health Care, 14(1):1057-1068

18) Maheen H., Malik F., Siddique, B. and Qidwai, A. (2018): Assessing Parental Knowledge bout Thalassemia in a Thalassemia Center of Karachi, Pakistan. J Genet Counsel, 24(6):945–951

19) Mostafa, S. and Mohammed, K. (2017): Educational Program for Nurses Regarding Management of Children with Maple Syrup Urine Disease during Acute Intermittent Late- Onset. 6(2):28-41.

20) Strauss, K., Puffenberger, E. and Carson, V. (2020): Maple Syrup Urine Disease. GeneReviews, ISSN: 2372-0697.Available at: https://www.ncbi.nlm.nih.gov/books

21) Yamaguchi, K., Wakimizu, R. and Kubota, M. (2017): Difficulties in Daily Life and Associated Factors, and QoL of Children with Inherited Metabolic Disease and Their Parents in Japan: a literature review. JIMD Reports, Vol. 33, doi: 10.1007/8904_2016_573 1-10. PP:1-10