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Mothers' Knowledge and Practices Regarding Care of their Children Suffering from Maple Syrup Urine Disease

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ABSTRACT

Background: Maple syrup urine disease (MSUD) is a rare inherited autosomal recessive neurometabolic disorder, is brought on by diminished activity of the Branched-chain α-ketoacid dehydrogenase complex (BCKDC), which catalysis the irreversible catabolism of branched-chain amino acids (BCAAs). This disease gets its name because of its burnet sweet odor of the affected infant's urine. **Aim:** To assess the mothers' knowledge and practices regarding care for their children suffering from Maple Syrup Urine Disease. **Design**: A descriptive research design was utilized to fulfill the aim of this study. **Setting**: The study was carried out at Genetic Out Patient Clinic in Mustafa Hassan Pediatric Hospital affiliated to El-Fayoum University Hospitals, and Abo El Rish (El Monira) hospital affiliated to Cairo University Hospitals. **Sample** A purposive sample consisted of 71 mothers who had children suffering from Maple Syrup Urine Disease. **Tools**: consists of two tools were used to conduct this study: **Tool I:** A structured interview questionnaire sheet to assess mothers' knowledge regarding Maple Syrup Urine Disease. **Tool II:** Observational checklist to assess mothers' practices regarding care of their children with Maple Syrup Urine Disease. **Results:** less than three quarter of studied mothers had unsatisfactory knowledge about MSUD and more than two thirds of them had inadequate reported practices regarding care of their children suffering from MSUD. **Conclusion**: Based upon the results the current study, it can be concluded that the studied mothers had unsatisfactory knowledge and practices about maple syrup urine disease. **Recommendation**: Continuous health education programs to mothers regarding Maple Syrup Urine Disease and early screening for children.

Keywords: Maple Syrup Urine Disease, mothers 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 and their toxic products (ketoacids) in blood, urine, and cerebrospinal fluid ***(Amaral & Wajner, 2022).***

Maple syrup urine disease is a genetic disorder. It estimates approximately 1 in 185,000 infants born worldwide, the worldwide prevalence of pediatric MSUD ranged from 1-9 per 1000000. It occurs more frequently in population with an increase frequency of consanguinity, such as the Mennonites in Pennsylvania, where the incidence is high as 1 in 200 births. ***(Ali et al., 2021)***. The incidence of MSUD at Egypt was found to be 1 for each 25,276 healthy newborn and 17 in 3900 high risk newborn ***(Dahpy, et al., 2021).***

Newborn screening for MSUD is performed throughout many countries so that most such infants are detected through these programs. Where such screening is not available, infants with MSUD usually present with advancing neurological signs. Early diagnosis and treatment stabilize the infants and, if well and consistently performed, can largely mitigate against serious long-term complications **(Strauss et al., 2020).**

Effective treatment of MSUD mainly consists of a life-long strict semisynthetic diet to reduce the accumulation of toxic metabolites. Maintenance of normal physical development and nutritional status and preventing catabolism are other goals of disease management ***(Ballikaya et al., 2021)***.

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Mothers are primary health care provider at their home, they spent more time to give protection and care for their children. Therefore, every mother has a MSUD child requires continuous knowledge, skills and support to promote care ***(Yamaguchi et al., 2017).***

Successful management of Maple Syrup Urine Disease (MSUD) requires a team effort from health care providers team, the child and their family. So 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 ***(Maier & Dokoupil, 2022).***

Significance of the study

Maple syrup urine disease is considered one of the metabolic diseases. The screening test should be done within one week after birth. In Egypt, this screening is not routinely done and discovered late after appearance of signs and symptoms, so the numbers of pediatrics with MSUD increasing. 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)***.

AIM OF THE STUDY

This study aimed to assess mothers' knowledge and practices regarding care for their children suffering from maple syrup urine disease through:

1- Assess the mothers' knowledge regarding care of their children suffering from maple syrup urine disease.
2- Assess the mothers' reported practices regarding care of their children suffering from maple syrup urine disease.

Research Questions:

1- What's the mothers' knowledge regarding care of their children suffering from MSUD?
2- What are the mothers' practices regarding care of their children suffering MSUD?
3- Is there relation between mothers' knowledge and practices regarding care of their children suffering from MSUD?

SUBJECTS AND METHODS

This study was conducted under the following four main designs as the following:

I- Technical Design
II-Operational Design
III-Administrative Design
IV-Statistical Design

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 descriptive research design was utilized to fulfill the aim of this study.

Research setting:

Children's Hospital affiliated to Fayoum University Hospitals at Genetic Out Patient and Abo El Rish (El Monira) hospital affiliated to Cairo University Hospitals at Genetic Out Patient.

Sampling:

A purposive sample of mothers who had children suffering from maple syrup urine disease, was used in this study within the following inclusive and exclusive criteria:

Inclusion criteria: all available children confirmed diagnosed with maple syrup urine disease and aged from one month to 9 years old from both genders and their accompanying mothers regardless their age, residence and socioeconomic standard.

Exclusion criteria: other children who have physical or mental disorders were excluded from the study.

The study included 71 children who were diagnosed with MSUD, and the sample size was determined using the following formula:

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$$n = \left(\frac{Z_{1-\alpha/2} + Z_{1-\beta}}{ES} \right)^2$$

Tools for data collection:

Two tools were used to collect the data, were designed by the researcher in a simple Arabic language after reviewing the related literature and after reviewing from the researcher' supervisors, they consisted of the following:

Tool (I): A Interviewing Questionnaire Sheet: which consisted of four parts:

Part (I): Characteristics of the studied mothers' include: age, educational level, occupation and marital status.

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

Part (II): Past Medical history and family history of children with MSUD **(14 items)** such as: duration of disease, date of discovery the disease, follow up frequency per month.

Part (III): Physical and psychological assessment of child such as: **(5 Items)** weight, height, head circumference, anxiety level, health need and problem.

Part (IV): Mothers' Knowledge regarding Maple Syrup Urine Disease which consisted of **(6 Questions)** multiple choice about definition, causes, signs and symptoms.
Part (V): Mothers' role in providing care for her child with Maple Syrup Urine Disease which consisted of **(16 Questions)** multiple choice about feeding, diet , medication , 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 0-2 grades, whereas, correct answer had score 2 grade and score zero was given for an incorrect or unknown answer. The total score was 42 grades (equal 100%). The total scores converted 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.

The observational checklist was adapted from ***Bowden& Greenberg (2017)***, ***Hockenberry and Wilson (2021) and Chapman, et al., (2018).***, and modified by the researcher to suit the nature of the study and reviewing from the supervisors to assess mothers' practices regarding care of their children suffering from MSUD. It included nine procedures divided into; NGT feeding (9 steps), diet measuring (6 steps), Dental health care (4 steps), weight measurement (6 steps), physical exercise and physical therapy (6 steps), Urine analysis (6 steps), Daily mother observation (5 items).

Scoring system

The observational checklist assigned a score of two for each item done correctly and 1 score if done incorrectly and zero score for not done items. There were a total of 42 steps in the checklist, resulting in a possible total score of 84, 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.

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Content validity:

To achieve the criteria of trust worthiness of the tool of data collection in this study. Tools of data collection were tested for content validity by three expert's professors of pediatric health nursing in Ain-Shams, Helwan and Fayoum university for clarity, relevance, comprehensiveness, simplicity, and applicability.

Reliability of tool:

The developed tool was tested for reliability on all of each part. The reliability process was assessed by measuring their internal consistency by using the Cronbach alpha coefficients test. it was including the following:

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

Pilot study:

The pilot study was conducted on 10% of the study subjects which consisted of 7 children and their accompanying mothers based on sample criteria. it was conducted to evaluate to the clarity of questions and time needed to complete the study tools. Based on the pilot study findings, no modification was made. The pilot sample was included in the study sample.

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.

Field Work:

The actual field work was carried out for data collection over six months started from November 2022 till end of April 2023. Data collected three days per week during morning shift from 9 am – 12 pm at previously mentioned settings. Initially, the researcher conducted session to assess both of knowledge and practices of mothers.

Mothers interview to assess mothers' knowledge about maple syrup urine disease using questionnaire sheet. Also, data related studied children's as children's characteristics, physical assessment and blood investigation follow up were obtained from medical record of each child. Meanwhile, each mother was assessed regarding to their practices using observation sheet. The time spent to fill the questionnaire ranged between 30 to 40 minutes according to the need explanations.

III- Administration Design:

A written letter should be issued from Dean the Faculty of Nursing Helwan University. Approval should be obtained from the mentioned settings administration.

IV- Statistical Design:

Data collected from the studied sample was analyzed and tabulated using the Statistical Package for Social Science (SPSS) version 20. Qualitative data was presented as numbers and percentages. The statistical tests used the chi-square test, means, slandered deviation, and Correlation test which showed good internal consistency construct validity

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RESULTS

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

Item	N	%
Child age/years		
Birth <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 studied children their age to be less than one year with mean age **2.14±1.97** and about two thirds of studied children are males (63.4%). The same table clarifies that half (50.7%) of studied children ranked as a second child in the family.

Table (2): 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 illustrates that more than three quarter (77.5%) of studied mothers lived in rural areas.

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Table (3): Distribution of the studied mothers level of knowledge regarding maple syrup urine disease **(n=71).**

Total knowledge	N	%
Satisfactory	21	**29.6**
Unsatisfactory	50	**70.4**
Mean±SD	**8.78±4.22**	

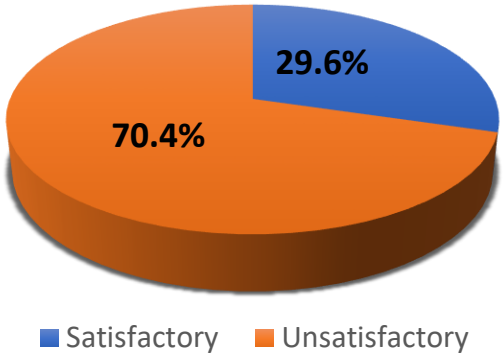

Fig:(1): Distribution of the studied mothers level of knowledge regarding maple syrup urine disease **(n=71**

Table (3), Fig (1): As regards to the total knowledge of the studied mothers about maple syrup urine disease this table and figure reveals that **70.4%** of studied mothers had unsatisfactory total level of knowledge and **29.6%** of them had satisfactory knowledge.

Table (4): Distribution of the studied mothers regarding level of practices regarding maple syrup urine disease **(n=71).**

Total practice	N	%
Done	23	**32.4**
Not done	48	**67.6**
Mean±SD	**14.06±4.31**	

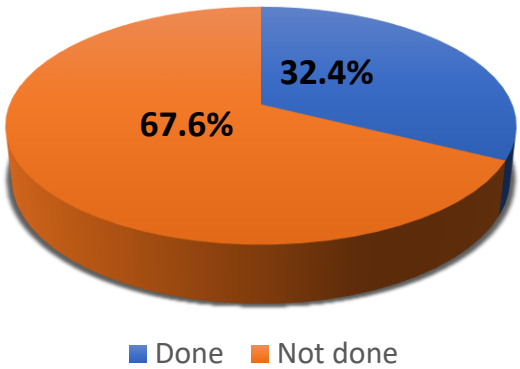

Fig (2): Distribution of the studied mothers regardin total level of self reported practices regarding maple syrup urine disease N= (71).

Table (4), Fig (2): As regards the mothers' practices about care of their children with maple syrup urine disease, this table and figure shows that **32.4%** of studied mothers had adequate self-reported practices, while **67.6%** of them had inadequate self-reported practices.

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Table(5): Relationship between characteristics of studied mothers and their knowledge level **(n= 71).**

Items	Knowledge score			
	Mean	SD	Tests	
			t/f	P-value
Age				
< 20	5.00	2.00	30.410	<0.001*
20 < 25	4.75	1.06		
25 < 30	7.22	2.39		
30 < 35	11.12	3.79		
35 and more	14.82	1.94		
Education level				
Illiterate	4.81	1.28	45.937	<0.001*
Primary	6.67	2.33		
Preparatory	8.55	1.75		
Secondary	12.57	3.25		
University	15.22	1.92		
Occupation				
House wife	6.45	2.35	10.371	<0.001*
Working	13.38	3.20		
Marital Status				
Married	14.20	1.87	5.081	<0.001*
Divorced\ Widow	7.90	3.83		
Place of residence				
Rural	4.81	1.28	4.935	<0.001*
Urban	9.95	4.08		

Table(5): Regarding the relation between characteristics of studied mothers and their knowledge this table demonstrates statically significant relationship in knowledge scores based on various demographic factors notably age groups, education levels, occupations, marital status and places of residence showed significant variations in knowledge scores.

Table(6): Relationship between characteristics of studied mothers and their practices level **(n= 71).**

Items	Practice score			
	Mean	SD	Tests	
			t/f	P-value
Age				
< 20	9.75	0.50	13.432	<0.001*
20 < 25	10.25	2.49		
25 < 30	11.56	3.46		
30 < 35	14.65	4.47		
35 and more	18.64	1.36		
Education level				
Illiterate	10.13	2.16	14.816	<0.001*
Primary	11.90	3.45		
Preparatory	11.27	3.07		
Secondary	15.86	4.55		
University	18.89	1.36		
Occupation				
House wife	11.06	2.97	7.211	<0.001*
Working	17.00	3.82		
Marital Status				
Married	18.80	1.03	5.352	<0.001*
Divorced\ Widow	12.13	3.90		
Place of residence				
Rural	10.13	2.16	3.317	<0.001*
Urban	13.93	4.42		

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Table (6): Concerning the relation between characteristics of studied mothers and their total self-reported practices this table illustrates statistically significant relationship in practice scores across various demographic characteristics as: age groups, education level, occupations, marital status and places of residence all exhibited significant variations in practice scores.

Table (7): Correlation between level of mothers' knowledge and their practices **(n=71).**

Knowledge score	Practice score	
	r	P-value
Total score knowledge about MSID	0.712	<0.001*
Total score knowledge about care of children with MSUD	0.518	<0.001*
Total score of knowledge	0.733	<0.001*

Table (7): According to correlation between Knowledge of studied mothers and their practices, this table shows that there was a positive correlation between total score of mothers knowledge and total score of their practices at the level with (r=0.733).

DISCUSSION

Maple syrup urine disease (MSUD) is a metabolic disorder characterized by difficulty digesting and processing proteins necessary for growth. To monitor and maintain ideal growth for children with MSUD, caregivers must carefully control consumption of harmful branched-chain amino acids (BCAAs). Treatment for MSUD primarily consists of a lifelong strict semi-synthetic diet to reduce the accumulation of toxic metabolites. Other goals of disease treatment include maintaining normal physical development and nutritional status and preventing catabolism (**Alrige et al., 2023 & Ballikaya et al., 2021)**.

Part I: Demographic characteristics of the studied children.

Regarding the characteristics of the study children, the results of the present study showed that the age of less than half of the studied children was between birth and less than one year, with the mean age being 2.14 ± 1.97 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" which examined a total of 25 patients, with two thirds of them being male.

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.

Regarding studied mothers according to their characteristics 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. The same table illustrated that the majority of mothers lived in rural areas. 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 to assess knowledge and practice of neonatal care among 290 postnatal mothers 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: Mothers' knowledge about MSUD:

As regards the total knowledge of studied mothers about MSUD the current study revealed that less than three quarter of them had unsatisfactory knowledge about MSUD this may be lack of mothers awareness about MSUD and they didn't receive any educational programs about disease. This findings was supported with **Abi – Warde et al., (2017)** in study about " Long-term metabolic follow-up and clinical outcome of 35 patients with branched chain keto-aciduria" they found that the most caregivers accomplished poor level of knowledge about BCKA.

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Part III: Mothers' practices about MSUD:

Regarding the studied mothers about total reported practices the current study revealed that more than two thirds of them had inadequate total level of reported practices. These results similar to the result of the study performed by **Blackburn et al., (2017)** about " Maple Syrup Urine Disease: Mechanism and Management" they found that most of the studied caregivers had inadequate practice level for children with MSUD.

Part IV: Correlation between mothers knowledge and their practices:

Concerning the correlation between the mothers' total knowledge and their total reported practices the present study revealed that there was a positive correlation between total score of knowledge and total score of practices. This result was supported by the study performed by **McGill et al., (2019)** about "children and young people's understanding of inherited conditions and their attitudes toward genetic testing" and stated that there was a very high statically significance correlation between mothers' knowledge and their practices. And 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.

Part V: Relationships between mothers' knowledge and their characteristics:

Regarding relationship between knowledge score of studied mothers and their characteristics the current study revealed that had a significant relationship between knowledge scores and various demographic factors, these factors include age groups. 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. In contrast, these results were not supported by **Friedman et al., (2016)** in study about " A neonatal presentation of co-morbid maple syrup urine disease and cystic fibrosis" and reported that no statically significant relation between the caregivers' personal characteristics and their total level knowledge.

Concerning relationship between practice score of studied mothers and their characteristics. This result illustrates statistically significant relationship in practice scores across various demographic characteristics as: age groups, education level, occupations, marital status and places of residence. These result supported by study performed by **Gallo et al., (2010)** in study about " family information and practice management pattern in childhood genetic condition" and found that a high statically relation between the total knowledge and practices scores

CONCUSIONS

The current study concluded that the studied mothers had un satisfactory knowledge and practices regarding care of children suffering from maple syrup urine disease.

RECOMMENDATIONS

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 conducted periodically for mothers' to update their knowledge and improve their practices.

3- Early screening for all newborns that help to discover disease early, early management and prevent complication.

4- awareness of mothers about prevention methods such as genetic counseling, premarital screening, prenatal screening to reduce the incidence of MSUD.

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