How to Cite:
Mahmoud, N. F., Amin, F. M., & Ayed, M. M. A. (2021). Effectiveness of instructional guidelines implementation on mothers' knowledge and practice regarding their children with methylmalonic acidemia. *International Journal of Health Sciences*, *5*(S2), 912–930. https://doi.org/10.53730/ijhs.v5nS2.13911

Effectiveness of instructional guidelines implementation on mothers' knowledge and practice regarding their children with methylmalonic acidemia

Naglaa Fathi Mahmoud
Assistant Professor, Pediatric Nursing Department, Faculty of Nursing Cairo University, Egypt
Email: naglaa.pedia@gmail.com

Fatma Mohamed Amin, RN, MSN, PhD
Assistant Professor of Pediatric Nursing, Pediatric Nursing Department Faculty of Nursing Mansoura University, Egypt
Email: fatmamohamed@mans.edu.eg
ORCID ID: http://orcid.org/0000-0002-3356-2443

Manal Mohamed Ahmed Ayed
Assistant Professor of Pediatric Nursing, Faculty of Nursing, Sohag University, Egypt
Email: Manal_ayed@yahoo.com
ORCID ID: https://orcid.org/0000-0003-0922-5823

***Abstract*---**Methylmalonic Acidemia (MMA) is a group of inborn errors of metabolism (IEMs), specifically of propionate catabolism characterized by gastrointestinal and neurometabolic manifestations resulting from a deficiency in the function of methyl malonyl-CoA mutase, methylmalonyl-CoA epimerase, and cobalamin metabolism. Aim: To investigate the effectiveness of instructional guidelines implementation on mothers' knowledge and practice regarding their children with methylmalonic academia. Design: A quasi-experimental design was used to achieve the aim of this study. Setting: This study was conducted at the Specialized Genetic Outpatient Clinic at Sohag University Hospital. Subject: A purposive sample of 50 mothers and their children with methylmalonic academia was selected from the previously selected settings. Three tools were used: (1) a structured interviewing questionnaire, (2) Mothers' knowledge regarding methylmalonic academia, and (3) Mothers' Reported Practices format regarding methylmalonic academia. Results: The current study results revealed that two-quarters of the mothers had unsatisfactory

knowledge about methylmalonic acidemia pre-instructional guidelines implementation compared with the majority of them reported satisfactory total knowledge post-instructional guidelines implementation; As well, mothers' total reported practices were significantly improved post-instructional guidelines implementation. Conclusion: It was concluded that instructional guidelines implementation has a highly significant improvement in studied mothers' knowledge and reported practices regarding the care of their children diagnosed with methylmalonic academia. Recommendations: The instructional guidelines implementation is recommended for mothers having children with MMA to improve their knowledge and practice regarding methylmalonic academia, its management, and the prevention of complications.

***Keywords*---**children, instructional guidelines, knowledge, practice, methylmalonic academia, mothers.

Introduction

Methylmalonic Acidemia (MMA) is a genetic condition in which some proteins and lipids cannot be properly metabolized by the body. Disorders associated with methylmalonic acidemia are a diverse category of inborn errors of metabolism (IEMs), including the error of propionate catabolism, which has gastrointestinal and neurometabolic symptoms. A biological trait of MMA is the accumulation of methylmalonic acid (MMA) in bodily fluids and tissues. Methylmalonic Acidemia can be categorized as either an isolated MMA, which is linked to a single propionyl-CoA catabolism deficiency or as a mixed MMA, which occurs in conjunction with other hereditary metabolic deficiencies (Ramsay, et al. 2018).

Except for the Middle East and North Africa, where it is higher, at nearly 6 per 100,000 infants, the detection rate of isolated MMA was less than one per 100,000 births in all locations. Instead, the incidence of MMA (all forms) per 100,000 infants in Asia-Pacific, Europe, and North America is 0.79, 1.12, and 1.22, respectively. Six births per one hundred thousand neonates have been reported to have MMA, which is the region with the highest detection rate (Almasi. et al. 2019).

Following the response to vitamin B12 therapy, methylmalonic acidemia is divided into two categories: vitamin B12 responsive and vitamin B12 non-responsive. There are three different complementation classes: cblA, B, and Dv2. cblA, B, and Dv2 are caused by the MMAA gene (4q31.1-2), MMAB gene (12q24.1), and MMADHC gene mutations, respectively (2q23.2). It was discovered that the previously reported cblH disease was cblDv2. Excessive vomiting, dehydration, muscle weakness (hypotonia), delays in child development, lethargy, liver enlargement, and failure to grow are the main indications and symptoms of MMA (Sutton & El-Hattab, 2018). These symptoms could vary from mild to life-threatening in the affected infant. The complications of MMA, in the long run, include intellectual problems, feeding disorders, and kidney diseases. Coma and death may occur in the affected infant if left untreated (Forny, et al. 2021).

Women with inborn errors of metabolism are getting to reproductive age more regularly and are healthy enough to think about getting pregnant thanks to neonatal screening for early diagnosis and advancements in pediatric care. To achieve the best results, patients with MMA who are pregnant or considering getting pregnant, as well as their healthcare professionals, need information about the potential hazards to the health of the mother and the fetus from the disease state during pregnancy. The use of newborn screening (NBS) enabled early detection and a more accurate calculation of the prevalence of MMA (Almasi. et al. 2019).

The methylmalonic Acidemia treatment plan includes intravenous administration of cobalamin (vitamin B12), carnitine, a high-caloric diet with restricted protein intake, along the administration of specific medications and antibiotics. Some complicated cases require organ transplantation (Gramer & Hoffmann, 2020). It is crucial to perform routine laboratory screening for kids with MMA. To handle MMA effectively, nurses, parents, and medical professionals must work together (Dinchong, 2019). In addition to ensuring that the child receives enough phenylalanine amino acid to maintain the levels necessary for growth and normal bodily processes, it's important to avoid leucine amino acid, which is used as a sweetener in some medications, when caring for a child with MMA. Additionally, it's important to limit your protein intake to avoid consuming too much of the amino acids isoleucine, threonine, methionine, and valine (Dixon, et al., 2020).

In the afflicted pediatric child, amino acids are converted to methylmalonic acid. The afflicted juvenile patient's diet should be specifically designed to ensure the absence of some amino acids while maintaining the presence of other amino acids required for growth. Each pediatric patient needs a treatment plan and diet that are specifically tailored to them (Forny, et al. 2021). Children's MMA can be effectively controlled by mothers. In addition to making sure that their children follow an adequate treatment plan and diet, moms must make daily efforts to deal with the obstacles and limitations that their children experience. Mothers should understand the MMA disease, how to diagnose it, manage it, and give their kids the attention they need (Pillai, et al. 2019).

Nurses are crucial in preventing MMA in kids by preventing natural protein intake (from breast milk, infant formula, and food products that include protein). Rehydration is the mainstay of treatment because MMA is very well excreted through the urine. To avoid catabolism and encourage anabolism, make sure you are getting enough calories. Help with the injection of intravenous glucose, lipids, and for more severe presentations, a condition-specific IV amino acid mixture to reduce ammonia generation from endogenous protein breakdown. On initial presentation with noticeably elevated propionic acid metabolites and elevated ammonia levels, help in hemodialysis or peritoneal dialysis may be advised (Ogier de Baulny et al, 2018).

Administration of carnitine is necessary for the movement of fats across mitochondria, however, when amino acidopathies occur, it is lost in urine attached to organic acids. Limit your intake of odd-chain fatty acids and the natural proteins isoleucine, threonine, methionine, and valine. Free foods, or those that have no protein, include things like fruit, vegetables, sweets, fats, and

specially-made low-protein foods. To aid in the excretion of organic metabolites, large dosages of carnitine are administered either orally or intravenously. Fatty acids are transported into the mitochondria, where they can be used as an energy source. To eliminate the gut bacteria, which produce 40% of propionic acid, metronidazole is used as an antibiotic (Pillai, et al. 2019).

Significance of the study

The majority of Egyptian mothers are uninterested in MMA, diet, and treatment regimens for children. Therefore, the current study emphasizes the importance of moms in providing for their children who are MMA sufferers. For the sake of their children's health, mothers must drastically alter their habits to avoid further issues. To provide for and control MMA-affected children, it is important to evaluate a mother's understanding of and practices related to the disorder. The results of this study need to assist the healthcare professional in developing new strategies for enhancing the effectiveness of MMA case management and treatment in Egypt.

Methylmalonic Acidemia is a burden on Egyptian society, especially in Upper Egypt where consanguineous marriage is common and has several negative impacts such as behavioral problems, fetal abnormalities, and intellectual deficiencies. Lack of understanding of the Methylmalonic Academia's symptoms and diagnosis is the cause of this. In the regions of Asia-Pacific, Europe, North America, and the Middle East and North Africa (MENA), the meta-analysis pooled point estimates of MMA (all forms) detection rates were 0.79, 1.12, 1.22, and 6.04 per 100,000 newborns, respectively. Except for MENA, where it was close to 6 per 100,000 infants, the detection rate of isolated MMA was less than 1 per 100,000 in all areas (Forny, et al. 2021). So, the researchers aimed in the current study to investigate the effectiveness of instructional guidelines implementation on mothers' knowledge and practice regarding their children with methylmalonic academia.

Aim of the study

This study aimed to investigate the effectiveness of instructional guidelines implementation on mothers' knowledge and practice regarding their children with methylmalonic academia through:

- Assess the mothers’ knowledge regarding the care of their children with Methylmalonic Academia.
- Assess the mothers' reported practice regarding the care of their children with Methylmalonic Academia.
- Design, implement and evaluate the effect of instructional guidelines on mothers' knowledge and practice regarding their children with Methylmalonic Academia.

Research Hypotheses

Hypothesis (1): Mothers' knowledge regarding Methylmalonic Academia is expected to improve post-instructional guidelines implementation than before.

Hypothesis (2): Mothers' practice of Methylmalonic Academia is expected to improve post-instructional guidelines implementation than before.

Subject and Methods

Research Design

A quasi-experimental design was used to achieve the aim of this study. Quasi-experimental research is selected into one of some different interventional groups to compare the real effectiveness and safety of non-randomized intervention (Maciejewski, 2020).

Research Setting

This study was conducted at the Specialized Genetic Outpatient Clinic at Sohag University Hospital, which offers services to several kids with MMA and other genetic diseases. Each of the two rooms of the ground-floor Specialized Genetic Outpatient Clinic, which is for children with MMA and other genetic abnormalities, has two beds.

Subjects

A purposive sample of 50 mothers and their children with methylmalonic academia was selected from the previously selected settings. The sample size was calculated with a 5% margin of error and a 95% confidence level from a population size of 60. The calculated sample size was 57. However, data was collected from 50 subjects as they met inclusion and exclusion criteria.

Inclusion criteria

Mothers having children suffering from MMA, their children's age ranged from birth to 8 years old.

Exclusion criteria included

Children suffering from mental and chronic diseases

Tools for data collection

Data were collected using the following tools: (pre/post)

Tool I: A structured interview questionnaire

It was developed and modified by the researchers depending on Dixon, et al., (2020) and was written in Arabic language, it consisted of four parts:

Part (1): It included four items regarding demographic data of the studied mothers as age, educational level, occupation, residence, and item regarding attending any type of formal education regarding MMA.

Part (2): It included three items' questions regarding the demographic data of the studied children as gender, age, and educational level.
Part (3): It included items regarding the disease history of the child with MMA as the duration of the disease, how to discover the disease, disease manifestations that appeared on the child and time of manifestations appearance, previous hospitalization, complications of the disease, regular follow-ups, regular investigation, and family history, which consisted of (10) questions.
Part (4): It included five items with (41) questions. It was used to assess children and their health needs and problems such as weight, height, and body mass index, child health needs, and problems.

Tool II: Mothers' knowledge regarding methylmalonic academia

It was developed by researchers after reviewing the literature (Pillai, et al. 2019 & Forny, et al. 2021), it was developed by the researchers after an extensive review of the related literature to assess the level of mothers' knowledge regarding MMA. It included 35 close-ended question items to assess mothers' knowledge about MMA children as a meaning, causes, signs, and symptoms, diagnosis of MMA, treatments, prevention and management, complications, proper nutrition, and mother's knowledge regarding their role in caring for their children with MMA.

Scoring system

The studied mothers' knowledge was scored as each question was given zero if the answer was incorrect or don't know, while a score of one was given to the correct answer. The total score was 35 grades equal to 100%. Accordingly, the total mothers' knowledge was classified as the following:

- Satisfactory ≥ 60%
- Unsatisfactory < 60%

Tool III: Mothers' Reported Practices format regarding methylmalonic academia

It was adapted from Pillai, et al. (2019) to assess the studied mothers' reported practices regarding the care of their children suffering from MMA. It included main 7 domains that consisted of 47 items. It was distributed as follows; measurement of diet balance (4 items), physical sports (4 items), dental care (8 items), follow-up of weight at home (5 items), food given by Ryle, and care taken at home (10 items), and daily mothers observation (7 items), and blood sugar measurement at home (9 items).

Scoring system

Each statement was given two scores if the action was reported as complete or correctly done, one score if the action was reported as incomplete or not correctly done was scored zero. The totals score of 94 grades is equal to 100% classified as the following:

- Adequate reported practices ≥ 60%

- Inadequately reported practices < 60%

The procedure

Preparatory Phase

To become familiar with different facets of the research issue and to build the study tools, a review of the relevant national and international literature was conducted during this phase. To build the instruments that were used and create the booklet that was used to execute the mothers' instructional guidelines, it was necessary to conduct a literature review, review numerous studies, and acquire theoretical knowledge about various parts of the research topic. Based on the adoption of instructional requirements for MMA, the educational session's content was created. There were prepared videos, lovely images, and a brochure. Printed out booklet covered all of the session's content and was written in a simplified version of Arabic.

Validity of the tools

The tools were tested for their content validity by a group of five experts in pediatric nursing and pediatric medicine. No modifications were carried out accordingly.

Reliability of the tools

Testing reliability of the study tools was done by Cronbach alpha, the result was α= 0. 89 for the first tool, α= 0. 88 for the second tool, and α=0.79 for the third tool.

Ethical Consideration

Before any data was collected, mothers' informed consent was obtained. The study's goal and anticipated results were explained to the mothers, who were also informed that their participation was optional and that they might withdraw from the study at any moment without providing a reason. They were also given the assurance that the data would only be utilized for research purposes and that anonymity and confidentiality would be protected.

Pilot study

A pilot study was carried out on 10% of the studied mothers (5 mothers and their children), to test the applicability and feasibility of the study tools. There was no modification done in the tools of data collection, so the mothers included in the pilot study were included in the total study sample.

Field Work

From March 2019 to August 2019, data were gathered for six months. The researchers visited the aforementioned location twice a week during the morning shift (Sunday and Monday), from 9 AM to 1 PM. The researchers introduced

themselves to each mother individually in the waiting area of the outpatient clinic before going over the purpose of the study. The surveys took between 25 and 35 minutes to complete or respond to, thus the researchers chose to conduct face-to-face interviews with the moms to help them fill out their knowledge and reported practices.

Assessment phase

In this stage, the researchers were gathering information about mothers' knowledge and practices regarding MMA using the tools they had developed (pre-test). Before beginning the interviews and data collecting, the researchers described the study's goals and expectations to the mothers who would be participating in it.

Planning phase

After establishing the goal of the execution of the instructional guidelines, mothers were consulted to discuss the contents, instructional strategies, and evaluation. The researchers considered the results of the literature review while creating the instructional guidelines for the Arabic language. It was updated, structured, and the materials were created with mothers' needs in mind. Selecting the most effective teaching strategies and mediums for delivering these topics was done after creating the instructional guidelines implementation contents. The researchers created a booklet with photos that are illustrated.

Implementation phase

This phase lasted 12 weeks, during which time the instructional guidelines were put into practice. The subject material was broken down into 4 sessions (two for theory and two for practice), with each session lasting 30 to 45 minutes. Mothers who participated in the study were split into ten groups for a total of 3 hours each. Five mothers were present in each group. Each session started with a summary of the previous session's feedback after a brief introduction to the content of the instructional guidelines was given at the beginning of the first session. There were lectures, demonstrations, and re-demonstrations, among other educational techniques. Posters and a brochure were used as appropriate media.

The first and second session was concerned with the mothers' knowledge regarding MMA. In the 1st session, the concept, causes, signs, and symptoms, diagnosis of MMA were discussed. In the 2nd session treatments, complications, prevention, proper nutrition, and mothers' knowledge regarding their role in caring for their children with MMA were discussed. While the third session focused on the discussion of mothers' stated practices for the care of their children who suffer from MMA, it also included instruction on measurement nutrition balance (4 things), physical sports (4 items), dental care (8 items), and weight monitoring at home (5 items). The discussion of mothers' stated practices around the care of their children who suffer from MMA, including meals provided by Ryle, care given at home (10 items), daily mother observation (7 items), and blood sugar measurement at home (9 items) was covered in the fourth session.

Prepared videos and lovely photographs were displayed. The crucial points were reviewed after each session. The sessions were repeated for each group of mothers. Each mother was provided with the educational booklet at the end of the 1st session as a guide.

Evaluation phase

The effect of the implementation of the instructional guidelines about MMA was reassessed post three months of the implementation phase using the same tools II, and III to determine the level of improvement.

Administrative Design

Before starting the research, approval was obtained from the Dean of the Faculty of Nursing and the directors of the previously selected setting at Sohag University Hospital to carry out this study to clarify the aim of the study and take their approval.

Statistical Design

Qualitative data were presented as frequencies and percentages. McNemar's test and Wilcoxon's signed-rank test were used to comparing the results of pre-and post-instructional guidelines implementation. The significance level was set at $P \leq 0.05$. Statistical analysis was performed with IBM SPSS Statistics for Windows, Version 23.0. Armonk, NY: IBM Corp.

Results

Table 1 showed that 60% of the mothers in the study were between the ages of 20 and 30 at a mean age of 24 6.4 years, 56% had a secondary education, and 70% were housewives. 60% of the moms who participated in the study resided in rural areas.

Figure (1) showed that the majority (98%) of the studied mothers didn't attend any educational training regarding MMA and only 2% had attended educational training regarding MMA.

Table (2) showed that 64% of the children were girls and about one-third (36%) of them were aged from 1 to 3 years. As regards educational level; nearly half of the children (48%) were in nursery school, while (36%) of them were at the primary school level.

Table (3) illustrated that 60% of children were diagnosed with MMA at ages less than one year. Concerning discovering the MMA disease (48% and 32%) of children were diagnosed by chance and were diagnosed by symptoms respectively. Approximately 70% and 62% of mothers reported that they follow up at a frequency of once monthly, 20% of them reported that there were other affected members in the family; and 40% of them were third-degree members. The same table also, showed that 52% of the children have complications such as delayed

mental and social skills 58% being the major complication among the studied children.

Table (4) revealed that almost half (48%) of the studied children their weight ranged from 10 – 20 Kg and 60% of them had a height was<100 cm. Approximately half (44%) of the studied children were under-weight, while children with a normal range of BMI constituted (30%) of participants. Regarding health problems among children with MMA, table (4) also illustrated that 44% of children had poor appetite, 56% had vomiting, 62% had extreme sleepiness or lack of energy, 38% had low muscle tone, 36% had delays in walking and motor skills, and 24% of children had enlarged liver.

Table (5) demonstrated that there was an improvement with a highly statistically significant difference between all items of mothers' knowledge pre/post three months of instructional guidelines implementation about MMA (P<0.001).

Figure (1): Most of the studied mothers (90%) had unsatisfactory total knowledge scores pre-instructional guidelines implementation about MMA, while 85 % of them had total satisfactory knowledge scores after the implementation of the instructional guidelines about MMA.

Figure (2) clarified the total practices score of the mothers' pre and three-month post-instructional guidelines implementation regarding MMA. It observed that most of the mothers (88%) had inadequate practices toward MMA care pre-instructional guidelines implementation and decreased to become 14% after three-month post- instructional guidelines implementation. Reversely, 12% of the mothers had adequate practices toward MMA care pre-instructional guidelines implementation in comparison to 86 % after three-month post- instructional guidelines implementation.

Table (6): Portrayed that there was a highly statistically significant relationship between knowledge and practice scores of the studied mothers regarding MMA pre and post-three months of instructional guidelines implementation at p<0.001.

Table (1): Frequency and percentage distribution of the studied mothers regarding their demographic data (n = 50)

	Mothers (n = 50)	
Variables	No	%
Age		
<20 year	10	20
20 – 30 years	30	60
30 – 40 years	8	16
>40 years	2	4
M±SD	24 ± 6.4 years	
Educational level		
Illiterate	4	8

Read and write	10	20
Secondary education	38	56
University degree	8	16
Occupation		
Working	15	30
Housewife	35	70
Residence		
Urban	20	40
Rural	30	60

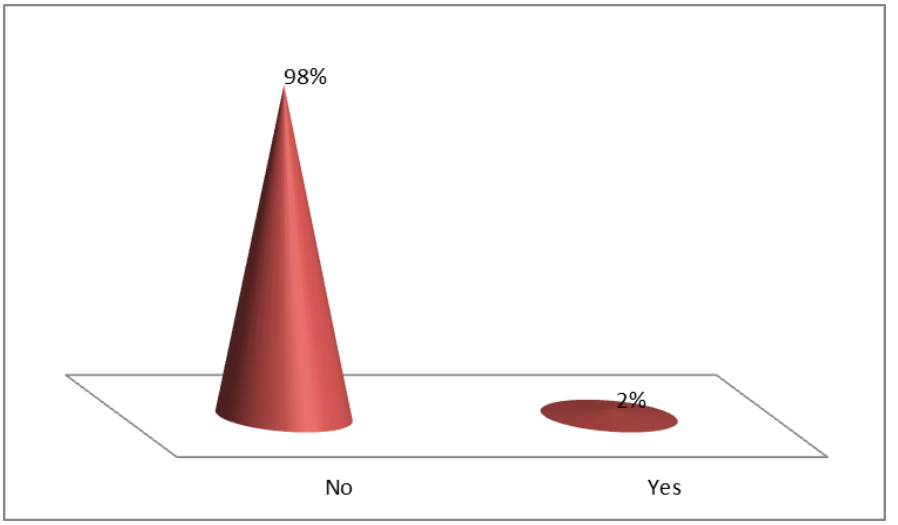

Figure (1): Percentage distribution of the studied mothers according to their attendance to educational training regarding MMA (n=50)

Table (2): Frequency and percentage distribution of the studied children regarding their demographic data (n = 50)

Variables	Children (n = 50)	
	No	%
Gender		
Boy	18	36
Girl	32	64
Age		
<1 year	10	20
1 – 3 years	18	36
3 – 6 years	5	10
6 – 9 years	17	34
M±SD	4.8 ± 2.55 years	
Educational level		
Baby class	24	48

Nursery school	8	16
Primary school	18	36

Table (3): Distribution of the studied children regarding their disease history (n = 50)

Disease History		Children (n = 50)	
		No.	%
Age at the time of MMA diagnosis	<1 year	30	60
	1 – <3 years	17	34
	3 – <6 years	2	4
	6 – 9 years	1	2
Discovering the disease	By chance	24	48
	Symptoms	16	32
	Complications	10	20
Periodic follow-up	Yes	35	70
	No	15	30
Frequency of follow-up	Once weekly	4	8
	Once monthly	31	62
	Twice per month	10	20
Compliance with follow-up	Always regular	30	70
	Sometimes regular	15	30
Other affected family members	Yes	10	20
	No	40	80
Relation with a family member	First degree	20	40
	Second degree	10	20
	Third degree	20	40
Presence of complications	Yes	24	48
	No	26	52
Type of complications	Delayed mental and social skills	29	58
	Behavioral problems	10	20
	Convulsions	6	12
	Mental retardation	5	10

Table (4): Frequency and percentage distribution of the studied children regarding their measurements and health problems (n = 50)

Items	Children (n = 50)	
	No	%
Weight		
<5 Kg	1	2
5 – 10 Kg	10	20
10 – 20 Kg	24	48

20 – 30 Kg	22	22
>30 Kg	4	8
Height		
<100 cm	30	60
100 – 150 cm	17	34
>150 cm	3	6
BMI		
Under-weight	22	44
Normal	15	30
Over-weight	10	20
Obese	3	6
Health problems		
Poor appetite	22	44
Vomiting	28	56
Extreme sleepiness or lack of energy	31	62
Low muscle tone (floppy muscles and joints	19	38
Breathing problems	14	28
Seizures	11	22
Learning problems or intellectual disabilities	12	24
Vision loss due to problems with the nerves in the eye	3	6
Delays in walking and motor skills	18	36
Enlarged liver	12	24

Table (5) Comparison of the mothers' knowledge, pre and post-three months of instructional guidelines implementation about MMA (n=50)

Mothers 'knowledge	No =(50)		P-value
	Pre (No/%)	Post (No/%)	
Definition of MMA	21 (42.0)	43 (86.0)	<0.001*
Causes	17 (34.0)	44 (88.0)	<0.001*
signs and symptoms	19 (38.0)	43 (86.0)	<0.001*
diagnosis of MMA	13 (26.0)	42 (84.0)	<0.001*
Treatments	20.0) (10	90.0) (45	<0.001*
prevention and management	17 (34.0)	43 (86.0)	<0.001*
Complications	14 (28.0)	43 (86.0)	<0.001*
proper nutrition	24.0) (12	94.0) (47	<0.001*
mother's knowledge regarding their role in caring for their children with MMA.	18 (38.0)	45 (90.0)	<0.001*

*highly significance at 0.001 levels -Chi-square test

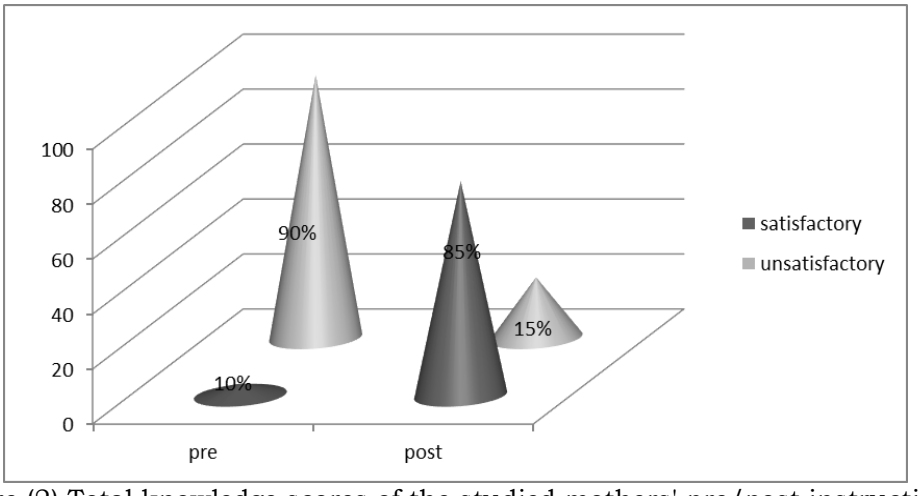

Figure (2) Total knowledge scores of the studied mothers' pre/post instructional guidelines implementation about MMA (n=50).

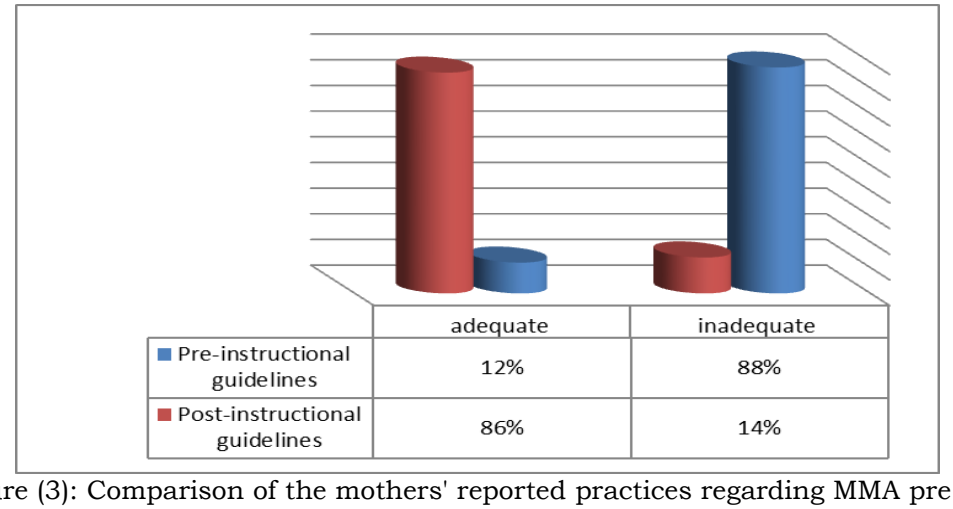

Figure (3): Comparison of the mothers' reported practices regarding MMA pre and post-three months of instructional guidelines implementation (n=50).

Table (6): Correlation between total knowledge and practices scores of the studied mothers regarding MMA pre and post-three months of instructional guidelines implementation (n=50)

Knowledge	Practice			
	Pre		Post	
	R	P	r	P
Pre	0.42	0.48*		
Post			0.86	0.001*

Highly statistically significant at p<0.001

Discussion

Methylmalonic acidemia (MMA) is regarded as a disorder with numerous variants, each of which has a unique origin and course of therapy. Cobalamin diseases A and B-related MMA is only one kind of MMA. To find out more about further MMA kinds brought on by cobalamin diseases C, D, and F. A disorder known as MMA prevents the body from properly metabolizing some fats and proteins. Because it can result in dangerous levels of organic acids and toxins in the body, it is regarded as an organic acid condition. When a child has this syndrome, they struggle to produce cobalamin enzymes A and B. The body needs cobalamin enzymes to digest specific meals (Zwickler et al., 2018).

The current study aimed to investigate the effect of instructional guidelines implementation on mothers' care of their children diagnosed with Methylmalonic Academia (MMA). The finding of the current study supported the stated hypothesis and indicates that the implementation of the instructional guidelines has a good impact on mothers' understanding and care of kids with methylmalonic acidemia since improper MMA regulation causes unwanted vomiting, metabolic acidemia, and poor feeding.

The current study showed that the mean age of the mothers it surveyed was 24 6.4 years, with two-thirds of them being between 20 and 30 years old. These results are consistent with those of a study by Ramadan et al., (2020), titled "Assessment of Women's Knowledge towards Care of Their Children Suffering from Methylmalonic Acidemia (MMA)," which discovered that the mean age of the mothers who were subjected to the study was 26.9 4.96 years. Additionally, these findings supported a study by Shahin et al., (2012) that examined "Nurses Knowledge and Practices Regarding Enteral Nutrition at the Critical Care Department of Al-Manial University Hospital" and discovered that the mean age of the analyzed mothers was 27.4 5.8 years.

According to the study's findings, more than two-thirds of the mothers who participated in it were stay-at-home mothers who came from rural areas roughly the same proportion. These results are consistent with those from a study by Ramadan et al. (2020), who also reported the same outcomes. According to the study's findings, the majority of the mothers who were investigated had not taken any MMA-related classes. This demonstrates the mothers who were tested needed this study.

Less than two-thirds of the children, according to the current study, were female. This finding contradicts Ramadan et al., (2020) claim that roughly two-thirds of the investigated children were male. The findings of the present study are consistent with those of Almási, et al., (2019) study, which was entitled "Systematic Literature Review and Meta-Analysis on The Epidemiology of Methylmalonic Acidemia (MMA) with a Focus on MMA Caused by Methylmalonyl-CoA Mutase (Mut) Deficiency" and revealed that two-thirds of the children were female. These findings differ from those of Tejada-Ortigosa, et al. (2019), who conducted a study on the "Health and Socio-Educational Needs of The Families and Children with Rare Metabolic Diseases" and discovered that about three-quarters of the participating children had one or more rare metabolic diseases.

According to the current study, three-fifths of children with MMA were diagnosed between the ages of one and three years old, with a mean age of less than a year. Ma, et al., (2018) provided evidence in support of the findings when they wrote in their study entitled "Shunt Surgery for Early-Onset Severe Hydrocephalus in Methylmalonic Acidemia" that the majority of the analyzed children had received a diagnosis at a young age. This was linked by the researcher to the MMA disease's unavoidable hazard and lethality. In a study regarding "Maple Syrup Urine Disease (MSUD): A Case with Long-Term Follow-Up after Liver Transplantation," McLaughlin et al., (2013) found that the children were diagnosed during their first ten months of life. On the other hand, Keyfi, et al., (2016), who researched "Methylmalonic Acidemia Diagnosis by Laboratory Methods," reported that almost half of the children were diagnosed at old ages, which runs counter to this result.

According to the study's findings, approximately half of the pediatric diagnoses were made by accident. According to the experts, this might be because mothers aren't aware of the sickness or don't know much about it. A recent study discovered that more than half of the children have issues, with delayed mental and social development being the main issue among the children who were the subject of the study. These findings are consistent with research by Evans et al., (2012) entitled "Home Enteral Tube Feeding in Children with Inherited Metabolic Disorders," which discovered that the majority of the children had social and mental difficulties. This illustrates how MMA harms children's growth, development, and health.

The findings of this study showed that mothers' knowledge of MMA was unsatisfactory before the implementation of instructional guidelines among the majority of mothers and became satisfactory among the majority of them after the implementation of instructional guidelines, with a statistically significant difference occurring after three months of the implementation of instructional guidelines. Ramadan et al., (2020), who reported that fewer than two-thirds of the investigated mothers had insufficient total knowledge, support the findings. This enhancement of the existing study demonstrated the beneficial impact of implementing instructional guidelines. It is also evidence that a thorough explanation of MMA for mothers contributes to and enhances MMA management, counseling, and education.

The findings of the current study showed that most mothers' practices for MMA care were deficient before intervention and became adequate among most of the examined moms three months after the adoption of instructional guidelines. These results are in line with those of Ramadan et al. (2020), who found that more than two-thirds of mothers reported using insufficient procedures overall to care for their children who had MMA. Additionally, this outcome was consistent with a study by Tejada-Ortigosa, et al. (2019), which found that the majority of the analyzed moms had poor habits. The outcomes demonstrated the value of the approach in enhancing mothers' habits and knowledge when caring for kids with methylmalonic acidemia.

The results of the current study showed that there was a highly statistically significant correlation between the mothers' knowledge and practice scores regarding the use of the MMA instructional guidelines. These findings support the

findings of Ramadan et al., (2020), who found a link between mothers' overall reporting behaviors and their knowledge of mixed martial arts (MMA). According to the researchers, this demonstrated how advances in understanding influenced better practices. Dizaj et al., (2014) report that employing instructional guidelines implementation could boost enabling and reinforcing variables supports the findings of the current study.

Additionally, findings are in line with those of Solhi et al., (2016), who investigated "educational intervention of quality of life, According to Ranjbaran et al. (2016), employing instructional guidelines to administer an intervention can considerably boost pre-intervention between the study group and control group. Similar findings were made by Mazloomy, et al., (2014) who discovered that applying an intervention based on an educational program enhances the quality of life. Another study by Gielen et al., (2018) demonstrated that the program may be successful in enhancing patient knowledge. Additionally, Oruoj et al., (2012) reported that the educational program was successful in encouraging people to adopt preventative practices.

Conclusion

Depending on the results of the current study, aim, and hypotheses, it was concluded that the majority of mothers had unsatisfactory knowledge and inadequate practices regarding MMA among their children. Instructional guidelines implementation has a highly significant improvement in studied mothers' knowledge and reported practices regarding the care of their children diagnosed with methylmalonic academia.

Recommendations

In light of the study's findings, the researchers recommended that:

1. The instructional guidelines implementation is recommended for mothers having children with MMA to improve their knowledge and practice regarding methylmalonic academia, its management, and the prevention of complications.
2. To attain generalizability, additional research should concentrate on replicating the current work on a larger probability sample.
3. Mothers who are taking care of kids with methylmalonic acidemia should have access to a reference handbook about the condition that is simplified and illustrated.

References

Almasi T, Guey LT, Lukacs C, Csetneki K, Voko Z, Zelei T. Systematic literature review and meta-analysis on the epidemiology of methylmalonic acidemia (MMA) with a focus on MMA caused by methylmalonyl-CoA mutase (mut) deficiency. Orphanet. J. Rare Dis. 2019; 14, 84.

Dinchong R. Reducing the psychosocial impact of a false positive newborn screen for inborn errors of metabolism. Faculty of Graduate Studies - FGS - (Electronic Theses & Dissertations) (Public) [24382]. Master of Science,

Manitoba Heritage Theses [5726], 2019; https://mspace.lib.umanitoba.ca/handle/1993/34019.

Dixon M, MacDonald A, White F J. Disorders of amino acid metabolism, organic acidaemias and urea cycle disorders. Clinical Paediatric Dietetics, 2020; 513-598.

Dizaji M, Taghdisi M H, Solhi M, Hoseini M, Shafieyan Z, Qorbani M, Rezapoor A. Effects of Educational Intervention Based on PRECEDE Model on Self-Care Behaviors and Control in Patients with Type 2 Diabetes, *Journal of Diabetes & Metabolic Disorders*; 2014; 13(1), 72.

Evans S, Preston F, Daly A, Ashmore C, Holden C, MacDonald A. Home enteral tube feeding in children with inherited metabolic disorders: a review of long-term carer knowledge and technique. Journal of Human Nutrition and Dietetics, 2012; 25(6), 520-525.

Forny P, Hörster F, Ballhausen D, Chakrapani A, Chapman K A, Dionisi-Vici, C, Baumgartner M R. Guidelines for the diagnosis and management of methylmalonic acidaemia and propionic acidaemia: first revision. Journal of Inherited - Metabolic Disease, Wiley O online Library, 2021; https://orcid.org/0000-0003-1877-2976.

Gielen A C, McDonald E M, Gary T L, Bone L R. Using the Precede-PROCEED Model to Apply Health Behavior Theories, Health Behavior and Health Education: Theory, Research, and Practice, 2018; 4:407–29.

Gramer G, Hoffmann G. F. Vitamin B 12 Deficiency in Newborns and their Mothers—Novel Approaches to Early Detection, Treatment, and Prevention of a Global Health Issue. Current Medical Science, 2020; 40(5), 801-809.

Keyfi F, Talebi S, Varasteh A. R. Methylmalonic acidemia diagnosis by laboratory methods. Reports of biochemistry & molecular biology, 2016; 5(1), 1.

Ma M, Wu M, Li Y, Wu D, & Zhang B. Shunt surgery for early-onset severe hydrocephalus in methylmalonic acidemia: report on two cases and review of the literature. Child's Nervous System, 2018; 34(7), 1417-1421.

Maciejewski, M. Quasi-Experimental design. Biostatistics & Epidemiology; 2020; 4 (1): 38-47.

Mazloomy S, Masoudy G, Fallahzadeh H, & Jalili Z. Education based on precede-proceed on quality of life in elderly. Glob J Health Sci; 2014; 6(6):178–84. Doi: 10.5539/gets.v6n6p178.

McLaughlin P M, Hinshaw J, Stringer A Y. Maple syrup urine disease (MSUD): a case with long-term follow-up after liver transplantation. The Clinical Neuropsychologist, 2013; 27(7), 1199-1217.

Ogier de Baulny H, Dionisi-Visi C, Wendel U. (2018) Branched-chain Organic Acidurias/Acidaemias. In Inborn Metabolic Diseases: Diagnosis and Treatment (Saudubray J.M., van den Berge G. & Walter J.H. eds). 5th edition, Springer, Germany, pp. 277-296.

Oruoji MA, Hashemi J, Hazavehei M, Charkazi A, Javaheri J, Moazeni M. The positive impact of an educational intervention program on preventive behaviors to reduce brucellosis in the rural people of Khomeini. Journal of Research Development in Nursing & Midwifery; 2012; 9 (1):51–60.

Pillai N R, Stroup B M, Poliner A, Rossetti L, Rawls B, Shayota B J, Burrage L C. Liver transplantation in propionic and methylmalonic acidemia: A single-center study with literature review, Molecular genetics, and metabolism, 2019; 128(4), 431-443.

Ramadan Z M, Ibrahim I A, Osama K Z. Assessment of Mothers' Knowledge towards Care of Their Children Suffering from Methylmalonic Acidemia (MMA), faculty of nursing, Ain Shams University, 2020; 1-218.

Ramsay J, Morton J, Norris M, Kanungo S. (2018): Organic acid disorders, Ann. Transl. Med; 6, 472.

Ranjbaran S, Dehdari T, Sadeghniiat-Haghighi K, Majdabadi MM. Poor Sleep Quality in Patients after Coronary Artery Bypass Graft Surgery: An Intervention Study Using the PRECEDE Model. J Tehran Heart Cent; 2015; 10(1):1–8.

Shahin M, Mohamed W, Sayed M. Nurses Knowledge and Practices Regarding Enteral Nutrition at the Critical Care Department of Al-Manial University Hospital in Egypt: Impact of a Designed Instructional Program. Journal of American Science, 2012; 8(11), 397-404.

Solhi M, Shabani M, Salehi M. Educational Intervention In Quality of Life of Women-Headed Households in Iran. Medical Journal Islam Repub Iran; 2016; 30:417.

Sutton VR, El-Hattab A W. Inborn Errors of Metabolism, an Issue of Pediatric Clinics of North America, E-Book, 2018; Vol. 65, No. 2.

Tejada-Ortigosa E M, Flores-Rojas K, Moreno-Quintana L, Muñoz-Villanueva M C, Pérez-Navero J L, Gil-Campos M. Health and socio-educational needs of the families and children with rare metabolic diseases: Qualitative study in a tertiary hospital, Anales de Pediatría (English Edition), 2019; 90(1), 42-50.

Zwickler M, Tamaris A, Ariane S. (2018): Metabolic decompensation in methylmalonic aciduria: which biochemical parameters are discriminative? M.D. University Children's Hospital Heidelberg, GERMANY; Journal of Inherited Metabolic Disease.