
▪ **Basic Research**

Effect of PRECEDE Model Application on Mothers' Care of their Children Diagnosed with Methylmalonic Acidemia

Asmaa Awad Helmy (1), Shadia Abd Elmoniem Syan (2), Manal Mohamed Ahmed Ayed (3)

Pediatric Nursing Department, Faculty of Nursing, Helwan University.

Pediatric Nursing Department, Faculty of Nursing, Sohag University

Pediatric Nursing Department, Faculty of Nursing, Sohag University

E-mail of the corresponding author: Manal_ayed@yahoo.com

Abstract

Introduction: 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 methylmalonyl-CoA mutase, methylmalonyl-CoA epimerase, and cobalamin metabolism. **Aim:** To evaluate the effect of PRECEDE model application on mothers' care of their children diagnosed with Methylmalonic Acidemia (MMA). **Design:** A quasi-experimental design was utilized. **Setting:** This study was conducted at the Specialized Genetic Outpatient Clinic at Sohag University Hospital. **Subject:** A purposive sample of 60 mothers and their children with MMA was recruited from the previously selected settings. **Three tools were used:** (1) structured interviewing questionnaire, (2) PRECEDE model questionnaire, and (3) Mothers' Reported Practices format. **Results:** The mean age of the studied children were 4.8 ± 2.49 years, and the mean age of the studied mothers was 25 ± 6.6 years. More than two-thirds of the mothers had unsatisfactory knowledge about Methylmalonic Acidemia pre- PRECEDE model implementation compared with the majority of them reported satisfactory total knowledge post-implementation; As well, mothers' total reported practices were significantly improved post-implementation of the PRECEDE model. **Conclusion:** It was concluded that PRECEDE model application led to highly significant improvements in studied mothers' knowledge and reported practices regarding the care of their children diagnosed with MMA. **Recommendations:** The PRECEDE model application for mothers having children with MMA should be provided to increase their knowledge and practice regarding the disease, its management, and prevention of complications. Future research should be focused on investigating the relationship between the socioeconomic status of mothers and Methylmalonic Acidemia. Nurses in the Genetic Outpatient Clinic should be trained well and supplied with information and training programs about MMA because the nurses are the main source of mothers' information who are providing their children with needed care. **Keywords:** Methylmalonic Acidemia, PRECEDE model, Mothers, Children.

Introduction

Methylmalonic Acidemia (MMA) is a hereditary disorder in which the body is incapable of metabolizing specific proteins and fats appropriately. Methylmalonic Acidemia disorders are a heterogeneous group of inborn errors of metabolism (IEMs), specifically the error of propionate catabolism characterized by gastrointestinal and neurometabolic manifestations. Accumulation of methylmalonic acid (MMA) in body fluids and tissues is a biochemical characteristic of MMA. Methylmalonic Acidemia can be classified as isolated MMAs and can be related to an isolated deficiency in propionyl-CoA catabolism, or as combined MMAs, in combination with other inherited metabolic defects (Ramsay, et al. 2018).

The detection rate of isolated MMA was less than one per 100,000 newborns in all regions, except the Middle East and North Africa where it is higher, close to 6 per 100,000 newborns. Instead, for Asia-Pacific, Europe, and North America, the incidence of MMA (all types) per 100,000 newborns is 0.79, 1.12, and 1.22, respectively. The highest detection rate of MMA has been reported in the Middle East and North Africa with six newborns per hundred thousand newborns (Almasi. et al. 2019).

Methylmalonic Acidemia is categorized into three types following the responsiveness to vitamin B12 treatment; these are vitamin B12 responsive and vitamin B12 non-responsive. There are three distinct complementation classes, cblA, B and Dv2 which caused by mutations in the MMAA gene (4q31.1-2); cblB by the MMAB gene (12q24.1); and cblDv2 by the MMADHC gene (2q23.2). The previously reported cblH disorder has been shown to be cblDv2.

The predominant signs and symptoms of MMA include excessive vomiting, dehydration, muscle weakness (hypotonia), delays in infant development, lethargy, liver enlargement, and failure to grow (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 to the affected infant if left untreated (Forny, et al. 2021).

With early diagnosis through newborn screening and improved pediatric care, women with inborn errors of metabolism are achieving reproductive age more frequently and are well enough to contemplate pregnancy. Patients with MMA, who are pregnant or interested in becoming pregnant, as well as their health care providers, need information surrounding the risks to maternal and fetal health that may be associated with the disease state during pregnancy and monitoring guidelines to ensure optimal outcomes. Newborn Screening (NBS) allowed for early diagnosis and a better estimation of the incidence of MMA (Almasi. et al. 2019).

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).

Regular laboratory screening for children with MMA is essential. Cooperation between healthcare providers, nurses, and parents is required to achieve proper management of MMA (Dinchong, 2019). Essential measures that should be taken into consideration while caring for a child with MMA include sufficient intake of phenylalanine amino acid as any decrease may affect the levels required for growth and normal body functions, also, leucine amino acid which is found in some drugs as sweetener should be avoided. Along with this, protein intake should be limited to restrict the intake of isoleucine, threonine, methionine, and valine amino acids (Dixon, et al., 2020).

Amino acids are metabolized into methylmalonic acid in the affected pediatric patient. Dietary intake for the affected pediatric patient should be tailored to assure the absence of amino acids while ensuring the presence of other amino acids that are essential for growth. Each pediatric patient needs an individually customized dietary intake and treatment plan (Forny, et al. 2021).

The PRECEDE–PROCEED model is a cost–benefit evaluation framework proposed in 1974 by Lawrence W. Green that can help health program planners, policy makers and other evaluators analyze situations and design health programs efficiently. It provides a comprehensive structure for assessing health and quality of life needs, and for designing, implementing and evaluating health promotion and other public health programs to meet those needs. One purpose and guiding principle of the PRECEDE–PROCEED model is to direct initial attention to outcomes, rather than inputs. It guides planners through a process that starts with desired outcomes and then works backwards in the causal chain to identify a mix of strategies for achieving those objectives. A fundamental assumption of the model is the active participation of its intended audience that is, that the participants ("consumers") will take an active part in defining their own problems, establishing their goals and developing their solutions (Glanz, K. and Rimer, 2011)

In this framework, health behavior is regarded as being influenced by both individual and environmental factors, and hence has two distinct parts. First is an "educational diagnosis" PRECEDE, an acronym for Predisposing, Reinforcing and Enabling Constructs in Educational Diagnosis and Evaluation. Second is an "ecological diagnosis" PROCEED, for Policy, Regulatory, and Organizational Constructs in Educational and Environmental Development. The model is multidimensional and is founded in the social/behavioral sciences, epidemiology, administration, and education. The systematic

use of the framework in a series of clinical and field trials confirmed the utility and predictive validity of the model as a planning tool (Green et al., 1975).

The meaning of PRECEDE is contained Predisposing, Reinforcing, and Enabling Constructs in Educational/Environmental Diagnosis and Evaluation, and PROCEED stands for Policy, Regulatory and Organizational Constructs in Educational and Environmental. Using the PRECEDE-PROCEED model can help in behavior change that examines the probable outcome of health programs (**Didehvar, et al. 2016**).

Educational intervention based on the PRECEDE model increased the average knowledge, attitude, enabling and reinforcing factors, and behavior of the individuals. The PRECEDE model provided an appropriate conceptual framework for designing an educational program (Pillai, et al. 2019). It can be used to design and evaluate a health promotion plan. The PRECEDE component allows the educational results to be reported and approved by (**Didehvar, et al. 2016**)

Mothers play an effective role in controlling MMA in children. Daily efforts are required from the mothers to face their children's challenges and disabilities along with assuring appropriate dietary intake and treatment plan. Mothers should be aware of the MMA disorder, its diagnosis, how to manage it, and provide suitable care for their children (Pillai, et al. 2019).

Nurses play an important role in controlling MMA in children as stop natural protein intake (infant formula / breast feeding / food products which are sources of protein). Rehydration - MMA is very well cleared by urinary excretion; hydration is thus the mainstay of treatment. Ensure adequate calorie intake to prevent catabolism and promote anabolism. Help in administration of intravenous glucose, lipids and for more severe presentations, condition specific IV amino acid mixture - to minimize ammonia production from endogenous protein breakdown. Help in hemodialysis or Peritoneal dialysis may be recommended on initial presentation with markedly elevated propionic acid metabolites and elevated ammonia levels (Pillai, et al. 2019).

Administration of Carnitine – this is essential for the transport of fats across the mitochondria, but is lost in urine bound to organic acids when amino acidopathies occur. Restrict intake of isoleucine, threonine, methionine and valine (natural protein) and odd-chain fatty acids. Free foods (i.e. non-protein) e.g. fruit, vegetables, sugars, fats and specially manufactured low-protein foods. Large doses of Carnitine are given orally or intravenously to assist the excretion of organic metabolites. It transports fatty acid into the mitochondria where they can be utilised for energy. Metronidazole (antibiotic) is used to clear the gut bacteria, which is responsible for 40% of propionic acid production (Ogier de Baulny et al, 2018).

Significance of the study:

Methylmalonic Acidemia represents an encumbrance on Egyptian society, particularly in Upper Egypt due to the prevalence of consanguineous marriage that occurs among family members triggering many adverse effects such as behavioral issues, congenital anomalies, and intellectual disabilities. Insufficient awareness of the Methylmalonic Acidemia rose due to poor knowledge about MMA diagnosis, and symptoms. Meta-analysis pooled point estimates of MMA (all types) detection rates were 0.79, 1.12, 1.22 and 6.04 per 100,000 newborns in Asia-Pacific, Europe, North America and the Middle East and North Africa (MENA) regions, respectively. The detection rate of isolated MMA was < 1 per 100,000 newborns in all regions with the exception of MENA where it approached 6 per 100,000 newborns (Forny, et al. 2021).

Mothers in Egypt are most prominently non-oriented about caring for children with MMA, dietary intake, and treatment plans. So, the current study highlights the role of mothers in caring for their children suffering from MMA. Mothers are required to completely change their lifestyles to prevent further complications and serious health problems for their children. Therefore, assessing a mother's knowledge and practices towards MMA disorder helps in achieving good care and control of children suffering from MMA. This study should also help the healthcare provider to plan further measures to improve the output of treatment and management of MMA cases in Egypt.

Operational definitions:

PRECEDE: It is meaning to Predisposing, Reinforcing, and Enabling Constructs in Educational Diagnosis and Evaluation.

Predisposing factors: It included some characteristics or socioeconomic situations that put an individual at risk of developing a disease or disorder. Educational intervention can affect some of the predisposing factors which include knowledge, attitude, self-efficacy, and self-care.

Enabling factors: It means the availability and accessibility of the skills and the resources.

Reinforcing factors:

It included the attitude of influential people (family, friends, peers, healthcare providers, media, teachers, community leaders, policymakers, etc.) who influence adopting healthy behaviors. An intervention may aim the influential people to successfully reach the actual target group.

PRECEDE-PROCEED model:

PRECEDE-PROCEED model is a comprehensive structure for assessing health needs for designing, implementing, and evaluating health promotion and other public health programs to meet those needs. PRECEDE stands for Predisposing, Reinforcing, and Enabling Constructs in Educational Diagnosis and Evaluation. According to this model, there are two categories of enabling resources that affect the use of health services which include community and personal enabling resources.

Aim of the study:

This study aimed to evaluate the effect of PRECEDE model application on mothers' care of their children diagnosed with Methylmalonic Academia (MMA) through:

-Assess the mothers' knowledge and reported practice regarding care of their children with Methylmalonic Academia.

-Design, implement and evaluate the effect of PRECEDE model for mothers to improve their care of their children with Methylmalonic Academia.

Research Hypotheses:

Hypothesis (1): Mothers' knowledge regarding Methylmalonic Academia will be improved after applying the PRECEDE model than before.

Hypothesis (2): Mothers' practice of Methylmalonic Academia will be improved after applying the PRECEDE model than before.

Hypothesis (3): The PRECEDE model will reflect a positive effect on mothers' care of their children suffering from Methylmalonic Acidemia.

Subject and methods:**Research Design:**

A quasi-experimental design was utilized to achieve the aim of this study. Quasi-experimental research is a prospective or retrospective study in which participants self-select or are 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 provides services for many children suffering from MMA and other genetic disorders. The Specialized Genetic Outpatient Clinic is on the ground floor and consists of two rooms every room contains two beds to receive children suffering from MMA and other genetic disorders.

Subjects:

A purposive sample of 60 mothers and their children suffering from MMA were selected from the previously selected settings within six months.

The sample size was calculated through Raosoft (2004), with a 5% margin of error and 95% confidence level from a population size of 70. The calculated sample size was 67. However, data was collected from 60 subjects as they met inclusion and exclusion criteria.

Inclusion criteria:

Mothers having children suffering from MMA, their children's age was ranged from birth to 9 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 depend on Dixon, et al., (2020) and was written in Arabic language, it has 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 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 as weight, height, body mass index, child health needs, and problems.

Tool II: Predisposing, Enabling, and Reinforcing Questionnaires (Precede Model questionnaire) Green & Kreuter, (2005):

It was developed by researchers after reviewing the literature and based on the educational and ecological approach of precede model. It included the following parts:

Part (1): Predisposing factor which included mothers' knowledge regarding MMA: it was developed by the researchers after an extensive reviewing of the related literature to assess the level of mothers' knowledge regarding MMA and their source of information. It included 35 close-ended question items to assess mothers' knowledge about MMA children as a concept, causes, signs and symptoms, diagnosis of MMA, treatments, complications, proper nutrition, and mother's knowledge regarding their role in caring for their children with MMA.

Scoring system: The studied mothers' knowledge was checked with a model key answer by the researchers, each question was given zero if the answer was incorrect or don't know, while a score 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 $\geq 60\%$

Unsatisfactory $< 60\%$

Part (2): Enabling factors assessment: It included 3 questions: Do you know available resources and facilities for MMA prevention and management? And Do you have easy access to healthcare facilities? Scores were given in the following way: A response of "Yes" to each question was given one score and response of "No" to questions received zero scores.

Part (3): Reinforcing factors assessment: It included three questions: Do you have emotional support from your family? Do you have a support group (friends, relatives, and doctors) related to their children's health nutritional recommendation? And Do you visit health centers regularly? Scores were given in the following way: A response of "Yes" to each question was given one score and response of "No" to questions received zero scores.

Part (4): To assess mothers' attitudes regarding their children's cause of MMA and other recommendations, it contains six items; Do you think that children's of MMA causes stress for you? Do you think that a high protein diet may increase risk MMA? Do you think that sports may improve MMA? Do you think weight to control obesity may increase risk of MMA? Do you think that regular medical follow-up is important?. And Do you think that regular blood sugar measurement is important. Response divided into 5 score categories were strongly agree, agree, no opinion, disagree, and strongly disagree. A total score equal 24. Total attitude scores ≥ 12 considered positive attitude, score less than 12 considered negative attitudes.

Tool III: Mothers' Reported Practices format:

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 the follows; measurement 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 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 $\geq 60\%$

Inadequate reported practices $< 60\%$

II. Operational Design:

Preparatory Phase

During this phase, a review of the literature was done through reviewing the available national and international related literature to be oriented with various aspects of the research topic and to develop the study tools. It involved a study of the literature, different studies, and theoretical part of knowledge about various aspects of the research subject by using books, journals, the internet, periodicals, and magazines which in the development of the used tools and designing the booklet that was used for the mothers' PRECEDE model application. Contents of the educational sessions based on PRECEDE model about MMA were designed. Videos, attractive pictures, and booklet were prepared. Booklet was written

in simplified Arabic language, covered all contents of the sessions, printed out regarding the sample size, and given to them.

Validity of the tools:

The tools were tested for their content validity by a group of three 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 $\alpha= 0.87$ for the first tool, $\alpha= 0.89$ for the second tool, and $\alpha=0.79$ for the fourth tool.

Ethical Consideration

Informed consent was obtained from mothers before data collection, the studied mothers were informed about the purpose and the expected outcomes of the study, and they were assured that the study was harmless and their participation was voluntary and they have the right to withdraw from the study at any time without giving any reason. They were also assured that anonymity and confidentiality will be guaranteed, as well the gathered data will be used for the research purpose only.

Pilot study:

A pilot study was carried out on 10% of the studied mothers (6 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

Data were collected within six months, from March 2019 up to August 2019. The researchers were visiting the previously mentioned setting 2 days/week at morning shift (Sunday and Monday) from 9 Am to 1 pm. The researchers met each mother individually at the waiting area present at Out-patient Clinic to explain the aim of the study after introducing themselves to mothers. The researchers used face-to-face interviews as the researchers read the questions and possible answers to the mothers to help them fill their responses in the knowledge and reported practices as it took about 25-35 minutes to fill or answer the questionnaires.

1- Assessment phase: (1 month)

In this phase, the researchers were using the designed tools in collecting the data about mothers' knowledge and practices regarding MMA (pre-test). The purpose of the study and its expectations were explained by the researchers to the mothers involved in the study before starting interviewing and data collection. During this phase, mothers' predisposing factors, enabling factors, reinforcing factors, the behavior practice regarding MMA, and the attitude towards MMA were assessed based on PRECEDE Model questionnaire as a pretest.

2- Planning phase: (1 month)

After determining the objective of the PRECEDE model application, a discussion with mothers about contents, teaching methods, and evaluation was carried out. The PRECEDE model application was designed by the researchers in the Arabic language considering the literature review. It was revised, organized and the contents were prepared according to mothers' needs. Constructing the PRECEDE model application contents was then followed by selecting the suitable teaching methods and appropriate media for teaching these contents. A booklet was developed by the researchers including illustrated pictures.

3- Implementation phase: (3 months)

This phase included 12 weeks, to implement the PRECEDE model application, the subject contents have been sequenced through 4 sessions (2 sessions for theory and 2 sessions for practices), and each session took 30-45 minutes. The total time was 3 hours for each group; mothers involved in the study were divided into seven groups. Each group included nine to ten mothers. At the beginning of the first session, an introduction about the PRECEDE model was given and each session started with summary feedback about the previous session. Different methods of teaching were used as lectures, demonstrations, and re-demonstration. Suitable media was used such as posters and a booklet.

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 was discussed. In the 2nd session treatments, complications, proper nutrition, and mother's knowledge regarding their role in caring for their children with MMA were discussed.

While the 3rd session was concerned with the discussion of mothers' reported practices regarding the care of their children suffering from MMA as measurement diet balance (4 items), physical sports (4 items), dental care (8 items), and follow up of weight at home (5 items) was taught. In the 4th session was concerned with the discussion of mothers' reported practices regarding the care of their children suffering from MMA as 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) was discussed. Prepared videos and attractive pictures were presented. At the end of each session, the important points were reviewed. The sessions were repeated to each group of mothers. Each mother was provided with the educational booklet at the end of the 1st session as a guide.

4- Evaluation phase:

The effect of the PRECEDE model application about MMA was reassessed after and evaluated immediately and after three months of implementation phase using the same tools I, II, and III to determine the level of improvement.

III. Administrative Design:

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

IV. Statistical Design:

Qualitative data were presented as frequencies and percentages. McNemar's test and Wilcoxon signed-rank test were used to comparing results pre- and post-PRECEDE model application. 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) illustrated that 64% of the studied mothers aged from 20 to 30 years old at mean age 25 ± 6.6 years, 58% of them had secondary education and less than two-thirds (63%) of them were housewives. Regarding residence 60% of the studied mothers were living in rural areas.

Figure (1) showed that the majority (93%) of the studied mothers didn't attend any type of formal education regarding MMA and only 7% had attended formal education regarding MMA.

Table (2) showed that 65% of the children were girls, about one third (35%) of them aged from 1 to 3 years. As regards educational level; nearly half of children (47%) were in nursery school, while more than one third (37%) of them were at primary school level.

Table (3) illustrated that 61% of children were diagnosed with MMA at ages less than one year. Concerning discovering the MMA disease (45% and 35%) of children were diagnosed by chance and were diagnosed by symptoms respectively. Approximately 71% and 70% of

mothers reported that they follow up at a frequency of once monthly, 22% of them reported that there were other affected members in the family; and 50% of them were third degree members. The same table also, showed that 55% of the children have complications such as delayed mental and social skills (60%) being the major complication among the studied children.

Table (4) revealed that almost half (46%) of the studied children their weight was ranged from 10 – 20 Kg and 62% of them their height was <100 cm. Approximately half (45%) of the studied children were under-weight, while children with a normal range of BMI constituted more than one-quarter (29%) of participants. Regarding health problems among children with MMA, table (4) also illustrated that 45% of children were had poor appetite, 55% had vomiting, 62% had extreme sleepiness or lack of energy, 38% had low muscle tone, 35% had delays in walking and motor skills, and 25% of children had enlarged liver.

Table (5): Revealed that mothers' knowledge, enabling, and reinforcing factors regarding MMA were decreased pre- PRECEDE model application. While there were significant changes and increases in the knowledge, enabling and reinforcing factors have occurred (89%, 97%, 87%) respectively compared to (37%, 63%, 60%) pre-implementation with statistical significance pre and after three months of PRECEDE model application (P=0.000)

Figure (2) clarified the MMA care total practices score of the mothers' pre and after three-month post-intervention. It observed that most of the mothers (92%) had inadequate practices toward MMA care pre-intervention and decreased to become 13% after three-month post-intervention. Reversely, 8% of the mothers had adequate practices toward MMA care pre-intervention in comparison to 87 % after three-month post-intervention.

Figure (3) clarified the total attitude scores of the mothers' regarding MMA pre and three months post-intervention. It observed that 71% of mothers had a negative attitude toward MMA pre-intervention and decreased to become 16% three-month post-intervention. Reversely, 29% of the mothers had a positive attitude toward MMA pre-intervention in comparison to 84 % three months post-intervention.

Table (6): Portrayed that there was a highly statistically significant relationship between knowledge, enabling factors, and reinforcing factors score and practices scores of the mothers throughout the intervention phases regarding MMA at $p < 0.001$.

**Table (1): Distribution of the studied mothers regarding their demographic data
(n = 60)**

Variables	Mothers (n = 60)	
	No	%
Age		
<20 year	10	16
20 – 30 years	38	64
30 – 40 years	10	16
>40 years	2	4
M±SD	25 ± 6.6 years	
Educational level		
Illiterate	4	6
Can read and write	13	22
Secondary education	35	58
University degree	8	14
Occupation		
Not working	4	7
Working	18	30
Housewife	38	63
Residence		
Urban	24	40
Rural	36	60

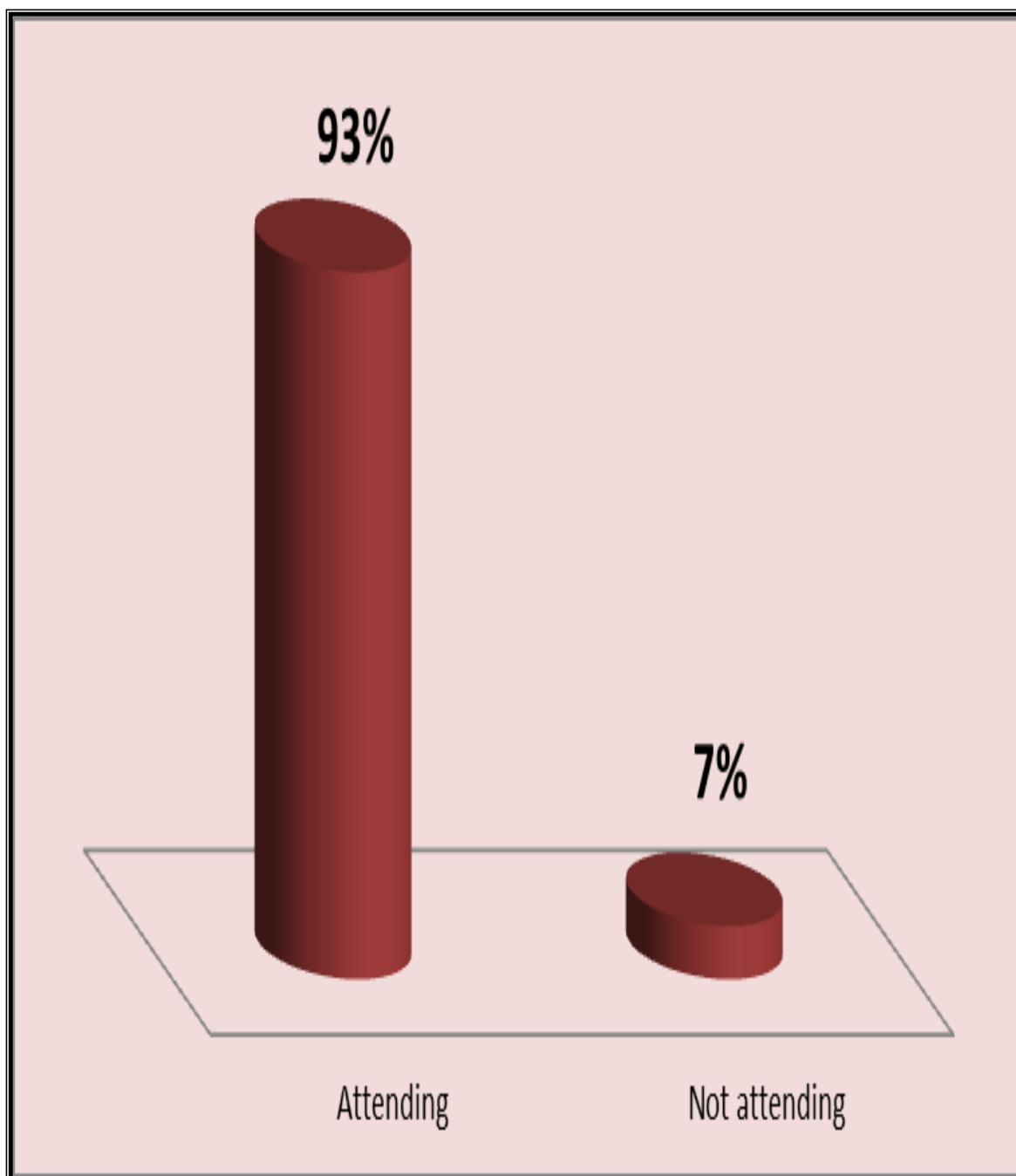


Figure (1): Percentage distribution of the studied mothers according to their attendance to any type of formal education regarding MMA (n=60).

Table (2): Distribution of the studied children regarding their demographic data (n = 60)

Variables	Children (n = 60)	
	No	%
Gender		
Boy	21	35
Girl	39	65
Age		
<1 year	13	21
1 – 3 years	21	35
3 – 6 years	6	11
6 – 9 years	20	33
M±SD	4.8 ± 2.49 years	
Educational level		
Baby class	28	47
Nursery school	9	15
Primary school	23	37

Table (3): Distribution of the studied children regarding their disease history (n = 60)

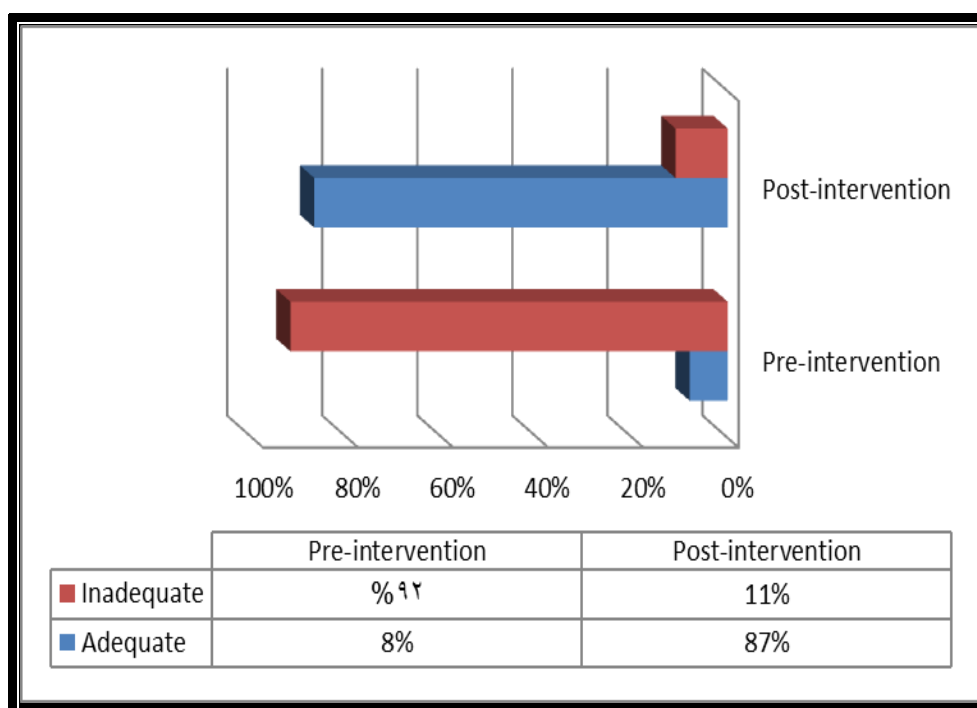
Disease History		Children (n = 60)	
		No.	%
Age at time of MMA diagnosis	<1 year	37	61
	1 – <3 years	20	33
	3 – <6 years	2	4
	6 – 9 years	1	2
Discovering the disease	By chance	27	45
	Symptoms	21	35
	Complications	12	20
Periodic follow-up	Yes	43	71
	No	17	29
Frequency of follow-up	Once weekly	3	7.4
	Once monthly	30	70
	Twice per month	10	22.6
Compliance with follow-up	Always regular	30	70
	Sometimes regular	13	30
Other affected family members	Yes	13	22
	No	47	78
Relation with family member	First degree	26	43
	Second degree	4	7
	Third degree	30	50
Presence of complications	Yes	27	45
	No	33	55
Type of complications	Delayed mental and social skills	16	60
	Behavioral problems	5	17.7
	Convulsions	3	12.2
	Mental retardation	3	10

Table (4): Distribution of the studied children regarding their measurements and health problems (n = 60)

Items	Children (n = 60)	
	No	%
Weight		
<5 Kg	1	2
5 – 10 Kg	13	21
10 – 20 Kg	28	46
20 – 30 Kg	15	25
>30 Kg	4	6
Height		
<100 cm	37	62
100 – 150 cm	21	35
>150 cm	2	3
BMI		
Under-weight	27	45
Normal	17	29
Over-weight	13	21
Obese	3	5
Health problems		
Poor appetite	27	45
Vomiting	33	55
Extreme sleepiness or lack of energy	37	62
Low muscle tone (floppy muscles and joints)	23	38
Breathing problems	17	29
Seizures	13	21
Learning problems or intellectual disabilities	15	25
Vision loss due to problems with the nerves in the eye	3	5
Delays in walking and motor skills	21	35
Enlarged liver	15	25

Table (5) Comparison of the mothers' knowledge, enabling factor, reinforcing factor pre and post-three months of PRECEDE model application (n=60).

Variables	Pre- PRECEDE model application		After three months post- PRECEDE model application		X ²	P- value
	No	%	No	%		
Knowledge (Predisposing Factor)	22	37	53	89	0.79	0.000
Enabling factor	38	63	58	97	0.37	0.000
Reinforcing factor	36	60	52	87	0.39	0.000

**Figure (2): Comparison of the mothers' practices of MMA care of the studied mothers' pre and post three months of PRECEDE model application (n=60).**

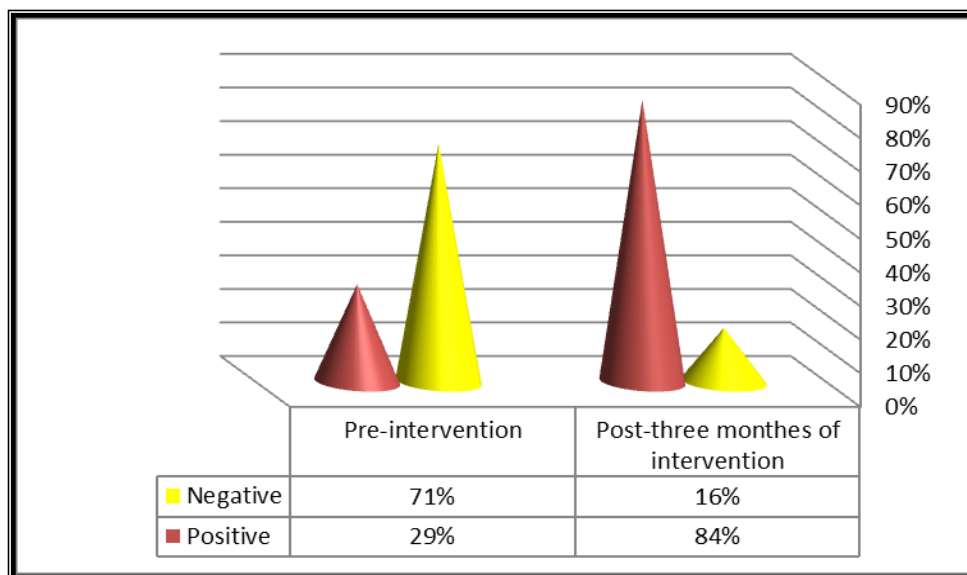


Figure 3: Attitude of the studied mothers regarding controlling of MMA pre and post three months of PRECEDE model application (n=60).

Table (6): Correlation between total knowledge, enabling factors, and reinforcing factors and practices scores of the studied mothers regarding MMA pre and post three months of PRECEDE model application (n=60).

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 considered a condition with many different forms, which all have different causes and treatments. MMA caused by cobalamin disorders A and B is just one type of MMA. To learn about other types of MMA which are caused by cobalamin disorders C, D, and F. MMA is considered an inherited condition in which the body is unable to break down certain fats and proteins. It is considered an organic acid condition because it can cause a harmful amount of organic acids and toxins in the body. Children with this form of the condition have trouble producing cobalamin enzymes A and

B. Cobalamin enzymes are necessary for the body to break down certain foods (**Zwickler et al., 2018**).

The current study aimed to evaluate the effect of PRECEDE model application on mothers' care of their children diagnosed with Methylmalonic Acidemia (MMA). The finding of the current study supported the stated hypothesis, and indicates that the PRECEDE model intervention reflected positively effect on mothers' knowledge, practice and attitude for care of their children suffering from Methylmalonic Acidemia because inappropriate control of MMA results in undesired vomiting, metabolic acidemia, and poor feeding.

The present study illustrated that less than two-thirds of the studied mothers aged from 20 to 30 years old and mean age of them was 25 ± 6.6 years. These findings match with results in a study conducted by **Ramadan et al., (2020)** titled "Assessment of Mothers' Knowledge towards Care of Their Children Suffering from Methylmalonic Acidemia (MMA)" and found that the mean age of studied mothers was 26.9 ± 4.96 years. Also, these results agreed with a study conducted by **Shahin, et al., (2012)** who studied "Nurses Knowledge and Practices Regarding Enteral Nutrition at the Critical Care Department of Al-Manial University Hospital" and found that the mean age of studied mothers was 27.4 ± 5.8 years.

The results of the present study revealed that less than two-thirds of the studied mothers were housewives and relatively the same proportion of the studied mothers were from rural areas. These findings are in the same line with results in a study conducted by **Ramadan et al., (2020)** who reported the same results.

The results of the present study indicated that most of the studied mothers didn't attend any type of formal education regarding MMA. This indicates the actual need of the studied mothers for this study.

The present study showed that more than two-thirds of the children were girls. This result is not following **Ramadan et al., (2020)** who stated that about two-thirds of the studied children were male. The result of the current study is similar to a study done by **Almási, et al. (2019)** who performed a study titled " 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 reported that two-third of the children were females. While these results are not similar to **Tejada-Ortigosa, et al. (2019)** who done a study about "Health and Socio-Educational Needs of The Families and Children with Rare Metabolic Diseases" and found that around three-quarters of the participated children were males.

The present study revealed that one third of children were diagnosed with MMA aged from one to three years and the mean age of them was 4.8 ± 2.49 years. The results are supported by **Ma, et al. (2018)** who stated in their study about "Shunt Surgery for Early-Onset Severe Hydrocephalus in Methylmalonic Acidemia" that most of the studied children were diagnosed in their early life⁹⁹. The researcher attributed this to the danger and fatality of the MMA disease that cannot be neglected. In the same line **McLaughlin, et al., (2013)** conducted a study about "Maple Syrup Urine Disease (MSUD): A Case with Long-Term Follow-Up after Liver Transplantation" and reported that the children were diagnosed throughout the first ten months of their life. On the other hand, this result is contradicted with **Keyfi, et al., (2016)** who studied "Methylmalonic Acidemia Diagnosis by Laboratory Methods" and found that approximately half of the children were diagnosed at old ages.

Results of the present study indicated that almost half of children were diagnosed by chance. From the researchers' point of view, this may be due to a lack of mothers' awareness and knowledge deficit regarding the disease.

The current study found that more than half of the children have complications as delayed mental and social skills being the major complication among the studied children. These results are in the same line with **Evans et al., (2012)** who done a study about "Home Enteral Tube Feeding in Children with Inherited Metabolic Disorders" and found that most of the children had mental and social complications. This reflects the negative effects of MMA on children growth, development and health status.

The results of the present study indicated that mothers' knowledge, enabling, and reinforcing factors regarding MMA were unsatisfactory pre- PRECEDE model application. While a statistically significant difference occurred after three months of PRECEDE model application. The results are supported by **Ramadan et al., (2020)** who stated that less than two-thirds of the studied mothers had unsatisfactory total knowledge. This improvement of the current study reflected the positive effect of PRECEDE model application. It is also, confirms that a comprehensive description of MMA for mothers helps to provide and improve understanding, management, and counseling of MMA.

The results of the present study revealed that most of the mothers had inadequate practices toward MMA care pre-intervention and become adequate among the majority of the studied mothers after three-month post-model application. These findings agree with **Ramadan et al., (2020)** who reported that more than two-thirds of mothers had inadequate total reported practices about the care of their children suffering from MMA. Also, this result was similar to a study performed by **Tejada-Ortigosa, et al. (2019)** who observed that most of the studied mothers had inadequate practices. The results reflected the importance of the model in improving both knowledge levels and practices of mothers caring for children with Methylmalonic Acidemia.

The findings of the present study revealed that there was a highly statistically significant relationship between knowledge, enabling factors, and reinforcing factors score and practices scores of the mothers throughout the intervention phases regarding MMA. These results are in accord with Ramadan et al., (2020) who stated that there was a positive correlation between mother's knowledge about MMA and their total reporting practices. From the researchers' point of view, this reflected that improving in the knowledge led to improve practices.

Findings of the current study are supported by Dizaj et al., (2014) who reported that implementing intervention using the PRECEDE model could increase enabling and reinforcing factors.

Also, results are consistent with the results of Solhi, et al., (2016) in Iran who studied " A PRECEDE PROCEED depending educational intervention of quality of life, **Ranjbaran, et al., (2016)** are reporting that applying intervention using the PRECEDE PROCEED model can increase reinforcing factors and significantly different from the before intervention between the study group and control group.

Similary, Mazloomy, et al., (2014) found that implementing intervention based on the PRECEDE model improves the quality of life. Another study done by Gielen et al., (2018) showed that the PRECEDE model can be effective in improving knowledge among patients. Also, Oruoij et al., (2012) reported that the PRECEDE model was effective in promoting preventive behaviors among individuals.

Conclusion:

Depending on the results of the current study, aim, and hypotheses, it was concluded that the majority of mothers had unsatisfactory knowledge, inadequate practices, and a negative attitude toward MMA among their children. While improved after PRECEDE model application which was effective in improving their knowledge and practice and attitude regarding the care of their children diagnosed with MMA. A highly statistically significant relationship between knowledge, enabling factors, and reinforcing factors score and practices scores of the mothers throughout the intervention phases regarding MMA.

Recommendations:

The PRECEDE model application regarding MMA should be integrated into the care for children in another study setting.

Continuous health education programs to mothers regarding MMA can help in providing continuous support, controlling, preventing MMA, and early detection of MMA.

Further research should focus on replication of the current study on a larger probability sample is recommended to achieve generalizability.

A simplified illustrated booklet regarding Methylmalonic Acidemia should be available to mothers who are caring for children with Methylmalonic Acidemia as a reference.

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البحث العربي

تأثير تطبيق نموذج PRECEDE على رعاية الأمهات لأطفالهن الذين تم تشخيص إصابتهم بحمض الميثيل مالونيك

مقدمة: حمض الميثيل مالونيك (MMA) هو مجموعة من الأخطاء الوراثية في التمثيل الغذائي (IEMs) ، وتحديدًا من تقويض البروبيونات الذي يتميز بمظاهر معدية معوية وعصبية استقلابية ناتجة عن نقص في وظيفة ميثيل مالونيل-CoA ، إبيميراز ميثيل مالونيل-CoA ، واستقلاب الكوبالامين. **الهدف:** تقييم تأثير تطبيق نموذج PRECEDE على رعاية الأمهات لأطفالهن الذين تم تشخيصهم بـ (Methylmalonic Academia (MMA). **التصميم:** تم استخدام تصميم شبه تجريبي. **اماكن جمع العينة:** أجريت هذه الدراسة في العيادة الخارجية التخصصية للوراثة بمستشفى جامعة سوهاج.

عينة الدراسة: تم استخدام عينة هادفة من 60 من الأمهات وأطفالهن مع MMA من الإعدادات المحددة مسبقًا. تم استخدام ثلاث أدوات: (1) استبيان المقابلة، (2) استبيان نموذج PRECEDE ، و (3) تنسيق ممارسات الأمهات المبلغ عنها.

النتائج: كان متوسط عمر الأطفال المدروسين 2.49 ± 4.8 سنة ، ومتوسط عمر الأمهات المدروسات 25 ± 6.6 سنة. ، أكثر من ثلثي الأمهات لديهن معرفة غير مرضية حول حمض الميثيل مالونيك قبل تنفيذ نموذج PRECEDE مقارنة مع أغلبية الأمهات أبلغن عن إجمال المعرفة المرضية بعد التنفيذ ؛ كذلك ، تم تحسين الممارسات بين اجمالي الأمهات بشكل ملحوظ بعد تنفيذ نموذج PRECEDE.

الخلاصة: استنتج أن تطبيق نموذج PRECEDE أدى إلى تحسن في معرفة الأمهات والممارسات فيما يتعلق برعاية أطفالهن الذين تم تشخيصهم بـ MMA. **التوصيات:** يجب تقديم تطبيق نموذج PRECEDE للأمهات اللاتي لديهن أطفال مصابون بـ MMA لزيادة معرفتهم وممارستهم فيما يتعلق بالمرض والوقاية من المضاعفات. يجب أن تركز الأبحاث المستقبلية على تحليل العلاقة بين الحالة الاجتماعية والاقتصادية للأمهات وحمض الدم الميثيل مالونيك. يجب تدريب الممرضات في عيادة العيادات الخارجية الوراثية جيدًا وتزويدهم بالمعلومات وبرامج التدريب حول MMA لأن الممرضات هم المصدر الرئيسي لمعلومات الأمهات اللاتي يقدمن لأطفالهن الرعاية اللازمة.

الكلمات المفتاحية: حمض ميثيل مالونيك ، نموذج PRECEDE ، أمهات ، أطفال.