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RESEARCH ARTICLE

EFFECT OF UNCOOKED CORNSTARCH TREATMENT ON METABOLIC CONTROL IN PATIENTS WITH HEPATIC GLYCOGEN STORAGE DISEASES

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ABSTRACT

Background and Purpose: Glycogen storage diseases (GSD) are a group of rare inherited metabolic disorders. Hypoglycemia, the most severe clinical manifestation of hepatic GSD, can be effectively prevented with the supplementation of uncooked cornstarch. This study aims to clarify the nutritional condition of individuals with hepatic GSD and to examine the impact of dietary therapy using uncooked cornstarch on metabolic regulation.
Methods: This study was a single-center, prospective cohort study involving 8 patients with hepatic GSD, aged between 0 and 18 years. Demographic characteristics, anthropometric measurements, biochemical markers, and energy and macronutrient intake of patients were assessed over a period of six months.
Results: The mean age of the patients was 8.0 ± 5.9 years. Twenty-five percent of the patients were female, while 75% were male. A significant difference was observed in plasma fasting glucose and alanine aminotransferase (ALT) levels among patients consuming uncooked cornstarch (p=0.024; p=0.044, respectively) and those with extended overnight fasting durations. The patients' body weight, height, and abdominal circumference showed significant changes during the 6-month follow-up period (p=0.038; p=0.001; p=0.008, respectively).
Conclusions: An uncooked cornstarch-based diet in individuals with hepatic glycogen storage diseases effectively maintains normoglycemia and mitigates possible metabolic consequences. Its appropriate consumption might facilitate adequate growth and development in patients and plays a vital role in metabolic regulation.

Keywords: Diet, Glycogen Storage Disease, Metabolism, Nutritional Status, Starch

INTRODUCTION

Glycogen storage diseases (GSD) are hereditary metabolic illnesses caused by a congenital enzyme deficit in glycogen metabolism and encompass many groupings (1). Hepatic GSD is a condition that affects the mechanisms of glycogen synthesis or breakdown in the liver. It is marked by the disruption of glucose homeostasis and is compounded by hypoglycemia, hyperlipidemia, hyperlactatemia, growth retardation, and hepatomegaly in untreated individuals (2). Long-

term consequences, including hepatic adenomas, renal disease, and osteoporosis, arise from inadequate metabolic regulation (3,4). Nutritional treatment serves as the principal intervention for the condition, facilitating plasma glucose management, averting metabolic problems, and supplying the energy and nutrients essential for the proper growth and development of children (5). In addition to supplying sufficient energy and nutrients suitable for the condition, uncooked cornstarch and/or continuous nocturnal dietary support are advised to maintain normoglycemia (2). Until the 1980s, patients received constant nutritional assistance based on clinical results; thereafter, uncooked cornstarch with a low glycemic index was incorporated into the therapy regimen (3,6). Cornstarch therapy may begin as early as six months of age, once pancreatic amylase activity has developed sufficiently. However, intestinal immaturity can lead to diarrhea, which may limit its use. A starch-to-water ratio of 1:2 is used to prepare cornstarch suspensions in sugar-free liquids for clinical administration (1, 2). The conventional care for individuals with glycogen storage diseases includes uncooked cornstarch therapy, strict diet control, and monitoring of biomedical parameters (7). In cases where there is aversion or difficulty in consuming raw cornstarch, and if the enteral route is to be maintained for over 6-8 weeks, the implementation of a nasogastric tube or percutaneous endoscopic gastrostomy (PEG) is necessary (5). Previous studies have examined the effects of uncooked cornstarch treatment on hypoglycemia and various biochemical parameters; however, the combined impact of uncooked cornstarch as a dietary intervention on growth, development and metabolic control in children has not been explored. Moreover, existing studies have focused on specific subtypes of hepatic glycogen storage disease rather than addressing the condition more broadly (3,4). In our country, literature on the subject is also limited. This study aims to evaluate the nutritional status of patients with different types of hepatic GSD and examine the effects of uncooked cornstarch on metabolic regulation.

MATERIALS AND METHODS

Study Design and Patients

This study is a prospective, single-center study involving patients under 18 years of age diagnosed with hepatic glycogen storage diseases, who were consistently monitored at the Pediatric Metabolism and Nutrition Outpatient Clinic of Şişli Hamidiye Etfal Training and Research Hospital from October 2018 to April 2019. The study aimed to include the entire population of eligible patients. Eleven individuals diagnosed with hepatic GSD were asked to participate in the study; however, one family declined to participate, and two patients were excluded due to not meeting the inclusion criteria. As a result, the study was conducted with 8 patients who met the inclusion criteria out of these. After the completion of the study, power analysis was conducted using the fasting glucose parameter in the G*Power 3.1.9.4 software. It was determined that the sample size (n = 8) achieved a statistical power of 82%. Patients who were fed by nasogastric tube, had an additional chronic disease accompanying GSD, and did not regularly attend follow-up visits were excluded in the study in order not to affect the results of the oral nutrition treatment.

Data Collection

The demographic profiles, clinical histories, and physical exams of the patients were documented. The nutritionist assessed and documented the body weight, height, body mass index (BMI), and waist circumference of the patients.

Body weights were measured using a TESS BT digital scale for patients aged 0-5 years, and a DİKOMSAN BW-200 digital scale with 0.1 kg precision for patients older than 5 years. Heights were measured using an SECA 207 measuring rod for patients aged 0 to 5 years, and an SECA 213 stadiometer for patients older than 5 years. During the height measurement, it was noted that the feet were stationary; the bodies of patients were in an upright position, while those of newborn patients were vertical. The weight-for-age and height-for-age z-scores of the patients were referenced against the standards established by the National Center for Health Statistics-Center for Disease Control (NCHS/CDC) as recommended by the World Health Organization (8).

The waist circumferences of the patients were measured in a standing position with arms extended, at the level of the umbilicus and subcostal area on the sides, utilizing an inelastic measuring tape (9). Plasma fasting glucose, aspartate aminotransferase (AST), alanine aminotransferase (ALT), alkaline phosphatase (ALP), lactate, uric acid, and triglyceride (TG) levels necessary for therapy were documented in the patient files by a pediatric metabolic physician.

The pediatric metabolic physician and the dietitian devised the patients' dietary treatment (10). Dietary interventions were implemented for the patients based on their clinical condition and biochemical parameters. The prescribed dietary composition included 40–50% carbohydrates, 10–15% protein, 25–35% fat, and 0.3–3 g/kg of uncooked cornstarch (10). The patients were questioned biannually for a total of two instances following the initial interview.

The 24-hour dietary recall method was used for dietary assessment of patients. During the face-to-face interview, patients were asked about the foods and beverages they have consumed in the previous 24 hours (on a weekday). The calorie, carbohydrate, protein, fat, and uncooked corn starch content of the meal intake were computed using the Nutrition Information System (NIS) (ver. 7.2) software, with all measurements and assessments conducted by the dietitian.

Ethical Approval

This study has obtained ethical approval from the University of Health Sciences- Şişli Hamidiye Etfal Training and Research Hospital Research and Application Center, Clinical Research Ethics Committee (Date: 25.09.2018, Decision No:107). The subjects were minors, and informed consent was obtained from their parents, with the study conforming to the Declaration of Helsinki requirements.

Statistical Analysis

The data collected from the study were analyzed utilizing the Statistical Package for Social Sciences (SPSS) Version 15.0 software (Chicago: SPSS Inc.). Numerical variables were presented as number (n), percentage (%), median, first quartile (Q1), third quartile (Q3), mean, standard deviation, minimum, and maximum; categorical variables were presented as counts and percentages. The numerical variables in the dependent groups were examined using Repeated Measures Analysis of Variance for normally distributed differences. Paired t test was used for the differences between two normally distributed groups, and Friedman test was used for non-normally distributed differences in groups exceeding two. Subgroup analyses in multiple dependent groups were conducted using the Wilcoxon test in nonparametric testing and interpreted with Bonferroni adjustment. P-value <0.05 was considered statistically significant.

RESULTS

The demographic characteristics, type of GSD, and diagnostic results of the patients are presented in Table 1. The study comprised 6 boys (75%) and 2 girls (25%). The patients' ages varied from 1 to 18 years, with a mean age of 8.0±5.9 years. Their age at diagnosis varied from a minimum of 2 months to a maximum of 132 months.

Table 1: Demographic characteristics, GSD subtype, and diagnostic findings of patients

Age Mean±SD (Min-Max)		8.0±5.9 (1-18)
Sex n (%)	Male	6 (75.0)
	Female	2 (25.0)
Age at diagnosis (Min-Max)		43.6± 49.2 (2-132)
Type of GSD n (%)	Fanconi- Bickel Send.	1 (12.5)
	GSD Type 0	1 (12.5)
	GSD Type 1	1 (12.5)
	GSD Type 3	4 (50.0)
	GSD Type 9	1 (12.5)
Initial Diagnosis Findings n (%)	Hepatomegaly, hypoglycemia	4 (50.0)
	Hypoglycemia	2 (25.0)
	Hepatomegaly, Hyperglycemia	1 (12.5)
	Hepotamegaly, Splenomegaly	1 (12.5)

SD: Standart Deviation; Min: Minimum; Max: Maksimum; GSD:Glycogen Storage Diseases.

Table 2: Comparison of patients' anthropometric measurements and biochemical parameters

			Subgroup Analyses		
			0^{th} month vs. 3^{rd} month	0^{th} month vs. 6^{th} month	3^{rd} month vs.6^{th} month
		Median (Q_1-Q_3)	p	p	p
Weight (kg)≠	0^{th} month	17.0(14.2-36.7)	0.105	**0.039**	0.265
	3^{rd} month	19.0(14.7-37.4)			
	6^{th} month	20.1(15.6-37.2)			
	p	**0.038**			
Height (cm) ≠	0^{th} month	0.97(0.87-1.34)	**0.008**	**0.001**	**0.001**
	3^{rd} month	1.01(0.87-1.37)			
	6^{th} month	1.02(0.91-1.4)			
	p	**0.001**			
BMI (kg/m^2) ≠	0^{th} month	18.8(17.6-21.2)	-	-	-
	3^{rd} month	19.0(18.5-20.6)			
	6^{th} month	19.4(17.6-20.6)			
	p	0.920			
Waist circumference ≠	0^{th} month	57.0(52-63)	**0.023**	**0.013**	0.833
	3^{rd} month	61.4(53.5-66.2)			
	6^{th} month	61.0(55.5-64.5)			
	p	**0.008**			
Fasting Glucose (mg/dl) ≠	0^{th} month	72(61.5-80)	0.147	**0.008**	0.240
	3^{rd} month	97(74.5-100)			
	6^{th} month	93(82.5-104)			
	p	**0.024**			
ALT*	0^{th} month	191 (72.1-238.5)	0.107	0.208	**0.017**
	3^{rd} month	174(52.5-189.5)			
	6^{th} month	198(75.5-309)			
	p	**0.044**			
AST*	0^{th} month	198(81.5-217)	-	-	-
	3^{rd} month	149(76-351.5)			
	6^{th} month	215(115-385.5)			
	p	0.303			
ALP*	0^{th} month	178(169.6-218)	-	-	-
	3^{rd} month	184(173-205.5)			
	6^{th} month	197(165-213)			
	p	0.607			
Uric acid*	0^{th} month	4.55(4.16-4.74)	-	-	-
	3^{rd} month	4.10(3.77-5.45)			
	6^{th} month	4.40(3.95-4.75)			
	p	0.508			

*Wilcoxon test; *Friedman test; BMI: Body Mass Index; ALT: Alanine aminotransferase; AST: Aspartate aminotransferase; ALP: Alkaline phosphatase; TG: Triglyceride*

Table 2: Comparison of patients' anthropometric measurements and biochemical parameters-continued

			Subgroup Analyses		
			0th month vs. 3rd month	0th month vs. 6th month	3rd month vs.6th month
		Median (Q_1-Q_3)	p	p	p
TG≠	0th month	281(133.5-357.7)	-	-	-
	3rd month	234(137-309.5)			
	6th month	215(124-322)			
	p	0.343			
Lactate≠	0th month	2.7(1.9-3.5)	-	-	-
	3rd month	2.5(1.95-3.4)			
	6th month	2.6(1.5-4.1)			
	p	0.670			

*Wilcoxon test; *Friedman test; BMI: Body Mass Index; ALT: Alanine aminotransferase; AST: Aspartate aminotransferase; ALP: Alkaline phosphatase; TG: Triglyceride*

The anthropometric measurements and biochemical parameters of the patients during the follow-up period are summarized in Table 2. A significant change in body weight, height, and waist circumference were recorded during the 6-month follow-up period (p=0.038; p<0.001; p=0.008, respectively). Significant variations were observed in the biochemical parameters of individuals consuming uncooked cornstarch, specifically in fasting glucose and ALT levels (p=0.024; p=0.044, respectively). Additionally,the median initial nighttime fasting duration was 6 hours (IQR: 4.5–10), which increased to 7 hours (IQR: 5.0–10.5) at six months. The median initial maximum daytime fasting time was 3.5 hours (IQR: 3.0–3.5), which was recorded as 3.5 hours (IQR: 3.5–4.0) after six months.

The patients' anthropometric measurements were assessed based on the initial z-scores. Changes in median z-scores for body weight, height, and BMI at baseline, 3-month, and 6-month follow-up are shown in Figure 1. There was no significant difference in the median z-scores of patients' body weight, height, and BMI (p>0.05). Based on weight-for-age z-scores, 4 patients (50%) were classified as extremely underweight, 1 patient (12.5%) as underweight, 2 patients (25%) as normal, and 1 patient (12.5%) as overweight. Based on height-for-age z-scores, 6 patients (75%) exhibited severe stunting, whereas 2 patients (25%) were classified as normal. In the assessment of BMI-for-age z-scores, it was noted that 2 patients (25%) were classified as wasted, 2 patients (25%) were classified as normal, and 4 patients (50%) were categorized as overweight.

Table 3 presents a comparison of dietary treatment and 24-hour dietary intake of the patients throughout the trial. A statistical difference was observed between the average calories provided in the nutritional treatment in the 3rd month and the average calories consumed by the patients (p=0.001). A difference was also found between the average protein percentage given to the patients in 0th and 3rd months and the average protein percentage in the patients' food consumption (p=0.012; p=0.029, respectively). It was found that the patients' initial mean uncooked cornstarch consumption was significantly different from the amonunt given in the diet treatment (r=0.043).

DISCUSSION

Dietary intervention plays a crucial role in the management of all hepatic glycogen storage diseases (GSDs). The objective of nutritional treatment is to achieve metabolic regulation, therefore mitigating potential complications and promoting the growth and development of patients. Energy and macro- and micro-nutrient requirements in nutritional treatment vary depending on the type of illness, the patient’s age, and associated comorbidities. Uncooked cornstarch and/or nutritional supplementation are advised to maintain blood glucose homeostasis (5,11,12).

Table 3: Comparison of patients' recommended nutritional requirements with the recorded dietary intake

Variables		Mean±SD	Difference Mean± SD	%95 CI Min	%95 CI Max	P#
Energy-0th month (kcal)	Diet	1681.9±325.9	21.2±209.1	-53.6	196.0	0.783
	Food Consumption	1660.7±344.7				
Energy-3rd month (kcal)	Diet	1693.8±321.2	196.3±82.5	127.3	265.3	**0.001**
	Food Consumption	1497.5±300.6				
Energy-6th month (kcal)	Diet	1731.3±352.5	173.0±210.4	-2.9	348.9	0.053
	Food Consumption	1558.3±260.7				
Carbohydrate-0th month (%)	Diet	39.8±8.04	-7.9±12.7	-18.3	3.00	0.134
	Food Consumption	47.4±9.30				
Carbohydrate-3rd month (%)	Diet	41.0±7.8	-4.2±9.9	-12.5	4.1	0.272
	Food Consumption	45.2±7.9				
Carbohydrate-6th month (%)	Diet	41.6±6.1	-3.4±7.8	-9.9	3.2	0.264
	Food Consumption	44.9±7.5				
Protein-0th month (%)	Diet	20.3±4.3	4.5±4.9	0.4	8.6	**0.012***
	Food Consumption	15.8±4.5				
Protein-3rd month (%)	Diet	19.9±3.9	2.3±2.4	0.3	4.4	**0.029**
	Food Consumption	17.6±5.5				
Protein-6th month (%)	Diet	18.9±3.7	0.9±1.7	-0.5	2.3	0.186
	Food Consumption	18.1±3.9				
Fat-0th month (%)	Diet	39.9±4.2	3.2±9.5	-4.7	11.1	0.373
	Food Consumption	36.8±6.9				
Fat-3rd month (%)	Diet	39.1±4.2	1.8±11.3	-7.6	11.3	0.660
	Food Consumption	37.2±9.2				
Fat-6th month (%)	Diet	39.5±2.9	2.5±8.5	-4.6	9.5	0.437
	Food Consumption	37.0±7.9				
Uncooked Cornstarch-0th month (g/kg)	Diet	0.96±0.87	0.45±0.74	-0.17	1.07	**0.043***
	Food Consumption	0.51±0.17				
Uncooked Cornstarch-3rd month (g/kg)	Diet	0.89±0.89	0.43±0.67	-0.14	0.99	0.116
	Food Consumption	0.46±0.37				
Uncooked Cornstarch -6th month (g/kg)	Diet	0.75±0.74	0.28±0.48	-0.12	0.68	0.145
	Food Consumption	0.47±0.38				

 # Paired t Test ; Wilcoxon Test; SD: Standart Deviation; Min: Minimum; Max: Maksimum*

Hepatic GSDs are typically diagnosed during infancy or childhood. During this period of rapid growth, it is essential to monitor growth, development, and treatment effectiveness. Santos et al. (13) found that, according to age, 76.1% of individuals diagnosed with GSD Type 1 who had uncooked cornstarch therapy were overweight, whereas 19% exhibited stunting. In another study, 40% of patients were classified as

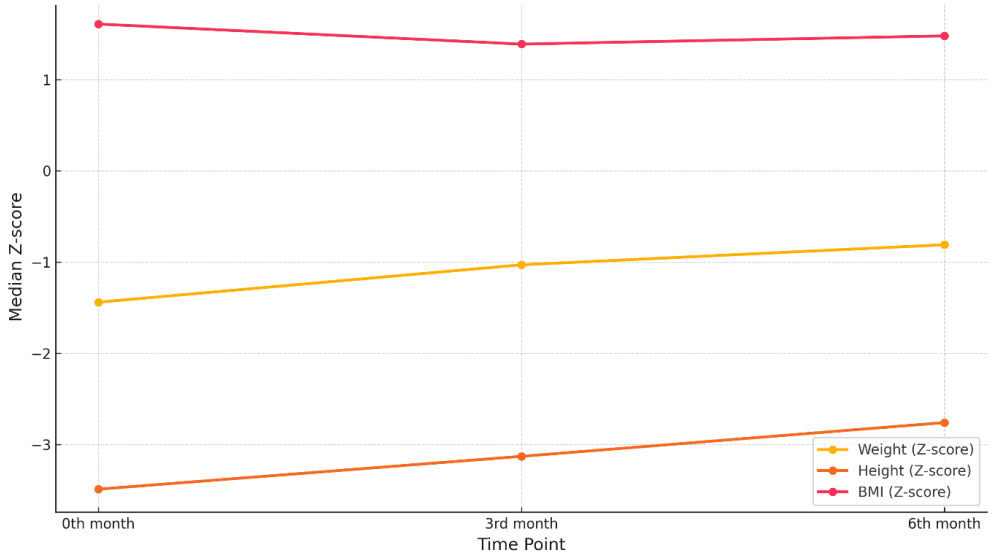


Figure 1. Median Z-scores of body weight, height, and BMI at baseline (0th month), 3rd month, and 6th month follow-up.

obese, 32% as normal weight, and 28% as overweight, according to their age. Moreover, the patients' weight-for-age z-scores were considerably elevated compared to the control group (14). An analysis of the height-for-age z-scores in GSD Type 1 patients revealed that 35.7% were normal height, 28.5% were severely stunted, 28.5% were stunted, and 7.1% were extremely tall (15). Another study in GSD Type 1 patients who had uncooked cornstarch therapy for over five years experienced notable height gains (16). Similarly, patients with GSD Type 3 also experienced height increases after one year of cornstarch treatment (17). In constrast to these studies, we found that half of the patients in our study were severely underweight according to weight-for-age z-score before starting regular nutritional therapy. Consistent with earlier findings, significant stunting was observed according to height-for-age z-score. However, a non-significant increase in body weight and height of z-skore was noted during follow-up, suggesting that the dietary intervention effectively promoted growth.

The most prominent clinical manifestation of hepatic GSD is hypoglycemia, particularly nocturnal hypoglycemia. If left untreated, hypoglycemia can lead to severe complications. Therefore, blood glucose levels are routinely monitored during treatment. The application of raw cornstarch has long been a favored therapeutic approach, particularly for nocturnal hypoglycemia (5,18,19). Uncooked cornstarch has been found to be more effective than dextrose in reducing nocturnal hypoglycemia in patients with GSD Type 1 (18), and to sustain elevated blood glucose levels for a longer duration compared to an isocaloric diet (20). Furthermore, its long-term use has been shown to help maintain stable blood glucose levels and prevent chronic complications (21). In addition, Ross et al. (22) reported that modified cornstarch maintained nocturnal blood glucose levels for a longer period than conventional cornstarch in individuals with GSD Type 1. Similarly, Hanaa et al. (17) demonstrated that uncooked cornstarch significantly reduced hypoglycemic seizure episodes in patients with GSD Type 3. Herbert et al. (23) found that a cornstarch-based diet given by the metabolism dietician stabilized blood glucose levels throughout the day and night. In our study, patients' daytime and nocturnal fasting periods were at the desired level and increased slightly over six months, though not significantly.

In the hepatic glycogen storage patients, the glycogen build up due to dysfunction in glycogenesis and glycogenolysis processes leads to the functional degeneration of the liver. To identify these deteriorations, levels of ALT, AST, triglyceride (TG), cholesterol, uric acid, and serum lactate are assessed, therefore preventing problems arising from nutritional therapy (24,25). It was reported that the uncooked cornstarch therapy did not reduce cholesterol and TG levels in GSD Type 1 patients receiving nasogastric tube feeding (15). Another

study showed no significant differences in AST, ALT, TG, cholesterol, or serum uric acid levels between patients consuming regular versus modified cornstarch, with values remaining stable (22). A comparative study revealed that the administration of uncooked cornstarch resulted in a notable reduction in AST levels exclusively in Type 0, 6, and 9 (25). Karwowski et al. (26) reported in their case report that marked transaminase elevations in a patient with GSD Type III were due to cell membrane damage due to glycogen accumulation in hepatocytes. In our study, ALT and AST levels increased over six months; however, only the ALT elevation reached statistical significance in the final three months. We think that this increase is due to glycogen damage accumulated in liver cells, as in the study in the literature. The reduction in ALP, TG, cholesterol, uric acid, and lactate levels, however there was not statistically significant, might have mitigated the risk of hepatic consequences.

Dietary therapy remains the primary intervention for managing this condition, and adherence to the prescribed nutritional regimen is essential for a favorable prognosis. Chehida et al. (27) found that inadequate intake of energy, protein, and cornstarch consumption was associated with hepatic, endocrine, and cardiac problems in 50 GSD Type 3 patients. In our study, we found that patients' energy and protein intake was below the recommended levels, similar to what is reported in the literature. The reasons for this may be the children's metabolic problems, as well as the reluctance to consume high-protein foods, which is common in children, and the low socioeconomic status of families (28). This is attributed to children's aversion to high-protein meals such as meat, the low socioeconomic level of families, and the elevated cost of animal-derived foods. Conversely, their intake of carbohydrates and fats adhered to the prescribed levels. We also discovered that patients' intake of uncooked cornstarch at the initial assessment was markedly below the required quantity. This might stem from families feeling uneasy about consuming items outside their normal routine and lacking a comprehensive awareness of the significance of ingesting uncooked cornstarch.

Our study has several limitations. GSD is a rare metabolic disorder and nasogastric tube feeding is a common treatment approach for this condition. Therefore, the sample size was limited by the exclusion of patients who were fed by nasogastric tube. Furthermore, due to socioeconomic inadequacies and transportation difficulties experienced by patients coming from outside the city, the follow-up period was limited to only six months, which was insufficient to evaluate the long-term effects.

CONCLUSION

Our findings demonstrate that a structured and closely monitored dietary intervention involving uncooked cornstarch improves in body weight, fasting glucose levels, linear growth, and metabolic regulation. Uncooked cornstarch appears to be an effective nutritional strategy in the dietary management of hepatic glycogen storage diseases, contributing to favorable changes in disease-related metabolic processes. To strengthen the level of evidence in this area, randomized controlled trials with larger patient cohorts and rigorous follow-up protocols are warranted. Future research should focus on the impact of appropriate dietary interventions on the nutritional status, growth, and development of children.

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Conflict of interest: The authors have no conflict of interest to declare.

Ethical approval: Ethical approval for the study was granted by the University of Health Sciences- Şişli Hamidiye Etfal Training and Research Hospital Research and Application Center, Clinical Research Ethics Committee (Date: 25.09.2018, Decision No:107), and the study adhered to the Declaration of Helsinki standards.

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