



Original Article

The Effect of Nutritional Counseling via Printed Recipe Cards Versus Verbal on Growth of a Cohort of Egyptian Children with Cystic Fibrosis

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Abstract:

Background: Nutritional management in cystic fibrosis children has a dramatic effect on growth and survival.

Aim of the work: to assess the effect of structured recipe cards nutritional protocol versus verbal nutritional counselling on growth of children cystic fibrosis (CF).

Methods: We prospectively studied a cohort study of 40 children recruited from the CF Clinic and following at Nutrition Clinic, Children's hospital, Cairo University whose caretakers were counselled and were provided with ready-made multiple calculated recipe cards that allowed preparation of specific meals to ensure adequate salt supplementation, high calorie, high fat diet and pancreatic enzyme dose of (1000IU lipase per 1g fat). Anthropometric parameters were recorded at the beginning, after 3 months and 6 months and compared to those of a historical cohort of 13 children with CF who received verbal nutritional counselling only.

Results: Malnutrition was highly prevalent with 64% of patients underweight and 70% stunted. Weight z-score, height z-scores, body mass index z score and middle upper arm circumference improved from -2.64, -2.77, -1.41 to -1.9, -2.22, -1 (p<0.001, p= 0.008, p<0.001) respectively after the intervention. Statistically significant elevation of hemoglobin, albumin and sodium from 11.49gm/dl, 3.85g/dl, 136.6mEq/l to 12.3, 4.44, 139 (p<0.001, p<0.001, p=0.04) respectively and statistically significant reduction of total leucocytic count (TLC), C-reactive protein and CRP/albumin ratio were detected after the intervention. There was no difference between achieved weight, height and BMI z-scores between both groups (p=0.485, p=0.499 and p=0.227 respectively). Mean care taker nutritional counseling session time was 17.9 ±3.8 min for those who received the recipe cards, which was significantly shorter than the mean (48.2±7.3 min) of the other group (p<0.001).

Conclusion: Optimization of nutritional status in children with CF patients resulted in significant improvement of anthropometric, laboratory and inflammatory parameters. Providing care takers with recipe cards allowed shorter counselling session time, and achieved same positive effect on growth as those verbally instructed without recipe cards.

Level of Evidence of Study: IIB (1).

Keywords: Nutrition; cystic fibrosis; anthropometric parameters; salt supplementation; high fat diet; recipe cards.

Abbreviations: BMI: body mass index; CBC: complete blood count; CF: cystic fibrosis; CRP: C reactive protein; FAO/WHO/UNU: food and Agriculture Organization of the United Nations, World Health Organization & United Nations University; MUAC: mid-upper arm circumference TLC: total leucocytic count.

Introduction

Cystic fibrosis (CF) is an autosomal recessive genetic disorder that compromises life expectancy. Early diagnosis is critical for appropriate and prompt treatment to improve patients' quality of life (2). Lung disease is the main cause of morbidity and mortality in patients with



CF, but also many patients present with gastrointestinal, pancreatic and/or hepatic manifestations throughout their lives, which negatively affect the quality of life and survival rate (3). Nutritional status has a close relation to lung function and survival in CF patients. Normal body weight has been associated with good lung function while longer life span has been associated with high energy, high-fat diet. Poor nutrition has been shown to have a bad impact on lung development in pre-adolescent children. Good lung function showed a linear correlation with good nutrition pattern as CF patients grow older (4).

For optimization of nutritional status, every CF patient should undergo thorough nutritional assessment. Also, pancreatic insufficiency is to be assessed and pancreatic enzyme replacement therapy dose is initiated if indicated with proper vitamins, mineral and trace elements supplementation (5). As nutrition in cystic fibrosis is considered a therapeutic intervention, care takers of children with CF receive nutritional counselling sessions. We have previously achieved improvement among CF children who suffered underweight in 78% and stunting in 60% of which decreased to 40 and 45% after the high calorie, high fat, salt supplemented diet (6). We aimed to evaluate effectiveness of supplying the care taker of child with CF by ready-made multiple calculated recipe cards that allowed structured meal planning to ensure adequate salt supplementation, high calorie, high fat diet and pancreatic enzyme dose of (1000IU lipase per 1g fat) on growth of children with CF.

Subjects and Methods

This is a prospective interventional observational study. It was conducted in cooperation with Clinical Nutrition Clinic, and Cystic Fibrosis Clinic, Children's Hospital, Cairo University, between March 2019 and December 2019. Informed consent was obtained from parents before enrollment. The study was approved by the Scientific Research Ethics Committee of Pediatrics Department-Faculty of Medicine-Cairo University (approval code: MS-163-2019), in compliance with Helsinki declaration guidelines (7).

Participants

The study included 40 children with confirmed cystic fibrosis, by positive sweat chloride test and/or genetic study. Their parents/care takers received verbal nutritional counselling and recipe cards. The studied parameters were compared to those of another historical cohort of 13 children whose care takers did not receive readymade recipe cards.

Methods

All enrolled children underwent the following at baseline, after 3 and after 6 months of the intervention throughout the study period to ensure good compliance:

- Full history taking including demographic data, motor and mental developmental history, feeding difficulties, onset and quality of complementary feeding, gastrointestinal and chest symptoms, age at diagnosis and other siblings with CF.
- Detailed nutritional history with a 24-h dietary recall for caloric, protein and fat intakes, compliance to a previously prescribed nutritional regimen, difficulties encountered, previous vitamins supplementation and salt replacement.
- A detailed history of pancreatic enzyme replacement therapy.
- Clinical assessment for signs of vitamins and mineral deficiency, and clinical examination.
- Anthropometric measurements: weight, height/length (below 2years old), body mass index (BMI) and mid-upper arm circumference (MUAC) were measured. All measures were processed by WHO Anthro Plus 3.2 software to obtain z-scores (standard deviation scores) for all anthropometric parameters .
- Routine laboratory tests: Complete blood picture (CBC), kidney function tests, electrolytes and minerals (Sodium, Potassium, Calcium, Phosphorus), albumin, liver function tests.
- Total leucocytic count, C-reactive protein (CRP) and CRP/albumin ratio were assessed as markers for inflammation.
- Urinary sodium as indicator of sodium balance was measured in a spot urinary sample.



Nutritional intervention:

Their caretakers received verbal nutritional counselling, and received a written structured nutritional support plan with readymade recipe cards containing the desired proportion of every macronutrient. Recipe cards were designed by the registered dietitians of the Pediatric Clinical Nutrition Clinic at Cairo University. High caloric, high fat dietary regimens were prescribed to contain from 1000 to 2200 kcal/day with fats representing 40-45% of total calories of vegetable oil and butter. Every meal contained in the card is labelled with the exact fat content in grams and corresponding pancreatic enzyme dosage. Calories were prescribed based on WHO/FAO/UNU recommendation for height age multiplied by stress factor 1.2-1.5 according to the severity of malnutrition (8). The ready-made meals allowed proper and easy adjustment of pancreatic enzyme dose and minimized the variability of the nutritional regimens provided. Medium-chain triglycerides containing formula was not included in pancreatic enzyme dosage (6). Pancreatic enzymes (Creon capsules 10000 international unit "IU") on pureed apple were used to achieve pancreatic enzyme replacement of 1000IU lipase for every gram of dietary fat. Salt was supplemented in the form of sachets (520mg sodium chloride, 30mg potassium chloride, 580mg sodium citrate (dihydrate) and 2.7g anhydrous glucose per 4.2g sachet). One daily sachet was prescribed for children younger than 2 years and two daily sachets were prescribed for older children with close monitoring of serum and urinary sodium levels. The historical cohort group received same instructions but not the ready made recipe cards of specific meals.

Statistical Analysis

Data were processed using Statistical Package for Social Science (IBM SPSS) version 23. The quantitative data were presented as mean, standard deviations and ranges when parametric and median with inter-quartile range (IQR) when non parametric. Also, qualitative variables were presented as number and percentages. The comparison between groups regarding qualitative data was done by using Chi-square test and/or Fisher exact test when the expected count in any cell found less than 5. The comparison between two paired groups with quantitative data and parametric distribution was done by using Paired t-test. The comparison between more than two independent groups with quantitative data and parametric distribution was done by using one way ANOVA. Pearson correlation coefficients test was used to assess the correlation between two quantitative parameters in the same group. The confidence interval was set to 95% and the margin of error accepted was set to 5%. So, the p-value was considered significant as the following: P-value > 0.05: Non significant (NS), P-value < 0.05: Significant (S), P-value < 0.001: Highly significant (HS).

Results

The study recruited 40 children with CF, 4 of them failed to attend the follow-up visits. The included 36 had a median age (IQR) of 10.5 (7-21) months, involving 22 boys (61.1%) and 14 girls (38.9%), with only 8 patients (22%) born to consanguineous parents. The historical cohort group comprised 9 (69.2%) boys and 4 (30.7%) girls. They were age and gender matched ($p=0.386$, $p=0.602$ respectively).

Effect on Nutritional Status

Nutritional parameters showed statistically significant differences before and after the intervention, with a significant increase in caloric, protein and fat intakes ($p=0.001$, $p<0.001$, $p=0.007$ respectively) and excellent compliance to the pancreatic enzyme dose of 1000IU lipase for every gram of dietary fat, and administration method ($p<0.001$) (Table 1). Malnutrition was highly prevalent among the studied patients with underweight (weight for age z-score ≤ -2 standard deviation "SD") represented 64% of the study group (23/36) while stunting (length/height for age z-score ≤ -2 SD) represented 70% (25/36) which significantly decreased to 45% and 58% respectively after the intervention (Table 1).

There were significant improvements in z-scores of weight for age ($p<0.001$), height for age (p -value=0.008), BMI for age ($p<0.001$) and MUAC for age ($p<0.001$) over a period of 6 months (Table I, Figure 1). There were statistically significant positive correlations between baseline fat intake and weight for age z-score at baseline ($r=0.410$, $p=0.020$) and follow up ($r=0.389$, $p=0.028$). Similarly, there were statistically significant positive correlations between baseline fat

intake and height for age z score at baseline ($r=0.413$, $p=0.017$) and follow up ($r=0.408$, $p=0.018$). These correlations suggest the role of high-fat proportion of diet in achieving better anthropometric parameters. There was no significant improvement in family-reported chest symptoms, frequency or duration of hospitalization before or after the intervention ($p=0.564$). As regard the gastrointestinal symptoms in the study group, there is a highly statistically significant improvement in steatorrhea ($p<0.001$).

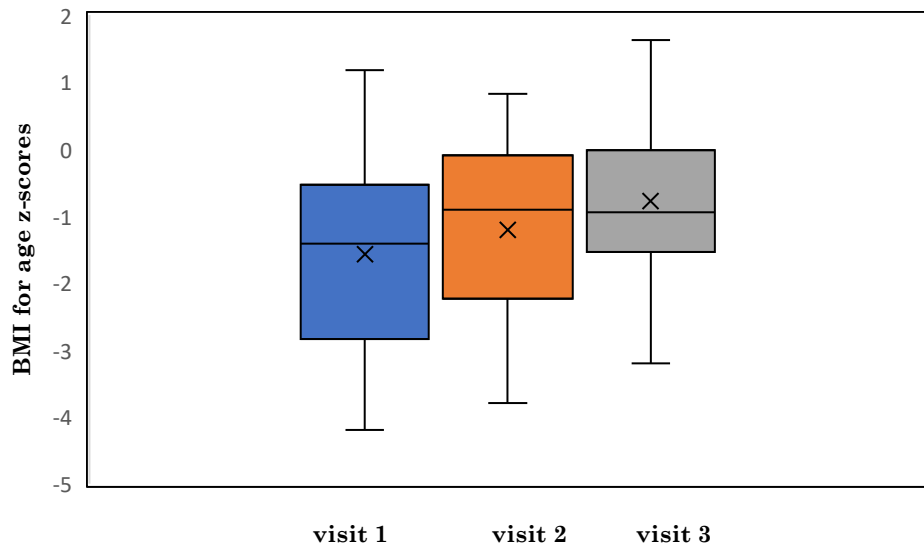


Figure 1: Comparison of the BMI z-score between the follow up visits.

Table 1: Nutritional assessment of the study group before and after using the readymade recipe cards.

		Baseline	Follow up (6 months)	P value
Calorie intake (kcal/kg/day)	Mean ± SD	100.28 ± 32.44	111.33 ± 33.69	0.001
Protein intake (gm/kg)	Mean ± SD	1.69 ± 0.79	2.6 ± 1.28	0.000
Fat intake (% of calories)	Mean ± SD	38.75 ± 3.66	40.83 ± 3.48	0.007
Pancreatic enzyme dose (IU/ 1g fat)	Mean ± SD	764.06±218.47	968.48±101.98	0.000
Weight Z score	Median (IQR)	-2.64 (-3.8 – -1.56)	-1.9 (-2.47 – 0.63)	<0.001
Height Z score	Median (IQR)	-2.77 (-3.81 – -1.47)	-2.22 (-2.9 – -1.39)	0.008
BMI Z score	Median (IQR)	-1.41 (-2.72 – -0.64)	-1 (-2 – 0)	<0.001
MUAC, (cm)	Mean ± SD	12.21 ± 2.14	14.17 ± 1.47	<0.001
Weight z-score				
• More than -2 SD	n (%)	13 (36%)	20 (55%)	<0.001
• -2 to -3 SD		9 (25%)	10 (28%)	
• Less than -3 SD		14 (39%)	6 (17%)	
Height z-score				
• More than -2 SD	n (%)	11 (30%)	15 (42%)	<0.001
• -2 to -3 SD		10 (28%)	13 (36%)	
• Less than -3 SD		15 (42%)	8 (22%)	

BMI: body mass index; IQR: interquartile range; MUAC: mid upper arm circumference; SD: standard deviation; IU: international unit; n: number.

Effect on Laboratory Parameters

At baseline evaluation 11 patients (31%) were anemic (Hb <11g/dl). They sustained significant improvement in hemoglobin level and mean corpuscular volume after the intervention (only 3 patients were anemic, with $p= 0.001$). Seven patients (19%) suffered initially from hypoalbuminemia (serum albumin ≤ 3.5 g/dl) with a statistically significant difference after the intervention ($p<0.001$). All patients were initially normocalcemic ($Ca \geq 8.5$ mg/dl) with a

statistically significant improvement of total and ionized calcium levels after the intervention. Phosphorus and alkaline phosphatase levels showed statistically significant reductions (Table 2). Upon initial evaluation of sodium balance, ten patients (28%) were hyponatremic (serum sodium less than 135mg/dl) while 26 patients (72%) were normonatremic (serum sodium 135-145mg/dl). All patients became normonatremic after the intervention with statistically significant improvement of serum sodium levels. Urinary sodium showed a statistically significant improvement before and after the intervention (Table 2). Baseline serum sodium showed statistically significant positive correlations with weight for age, height for age and mid-upper arm circumference for age z-scores at baseline and follow up (Table 3) (Figure 2). These correlations confirm the importance of proper salt supplementation at the beginning of any nutritional regimen to achieve normal sodium balance and subsequently normal progression of growth parameters. There was highly significant improvement in total leucocytic counts (Figure 3), CRP and CRP/albumin ratio (Figure 4) ($p < 0.001$ for all markers). C-reactive protein values showed a statistically significant negative correlation with the fat intake ($r=0.453$, $p=0.010$), pointing to the possible immunomodulatory role of high dietary fat in CF patients.

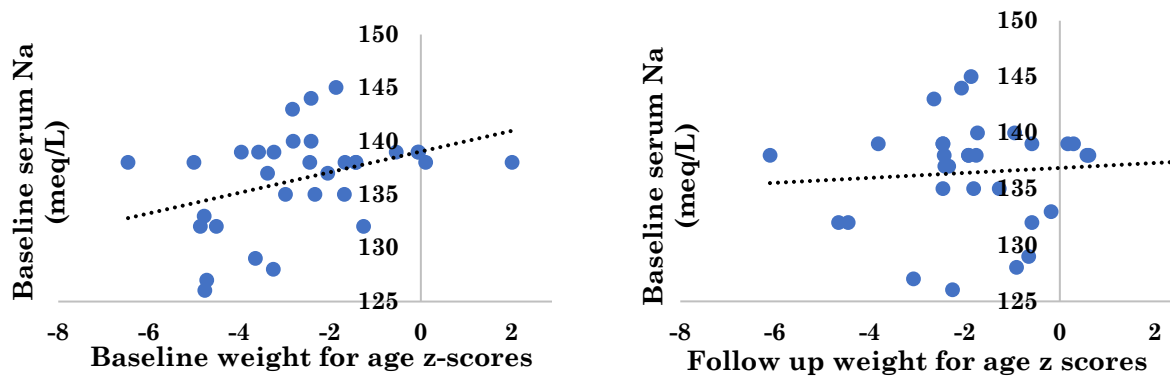


Figure 2: Correlation of serum sodium to weight for age z-scores at baseline and follow-up.

Table 2. Laboratory parameters of the study group before and after the recipe cards use.

	Visit 1 (Mean ± SD)	Visit 2 (Mean ± SD)	Visit 3 (Mean ± SD)	P value
Hb (gm/dl)	11.49 ± 1.1	11.83 ± 0.93	12.32 ± 0.97	0.001
MCV (fl)	76.45 ± 7.39	75.31 ± 6.33	72.5 ± 5.57	0.022
MCH (pg)	25.86 ± 5.19	26.03 ± 4.79	25.73 ± 3.04	0.947
HCT (%)	34.67 ± 4.21	36.15 ± 2.65	36.16 ± 3.37	0.246
PLT (cells/mm ³)	360.8 ± 133.44	337.56 ± 98.1	348.25 ± 98.4	0.479
Albumin (g/dl)	3.85 ± 0.43	4.19 ± 0.23	4.44 ± 0.21	<0.001
UREA (15-45mg/dl)	15.12 ± 7.42	14.25 ± 3.19	15.31 ± 3.68	0.503
Creatinine (0.3-0.7mg/dl)	0.45 ± 0.1	0.47 ± 0.11	0.47 ± 0.1	0.473
AST (10-38 IU/l)	32.42 ± 12.45	30.78 ± 8.65	28.66 ± 4.3	0.223
ALT (10-40 IU/L)	30.94 ± 13.78	29.88 ± 12.04	29.84 ± 5.74	0.790
Total Calcium (≥ 8.5 mg/dl)	9.68 ± 0.53	10.18 ± 0.32	10.38 ± 0.22	<0.001
Ionized Calcium (mg/dl)	4.65 ± 0.45	4.98 ± 0.32	5.01 ± 0.29	<0.001
Phosphorus (3.5-5.5mg /dl)	6.08 ± 1.05	6.14 ± 0.86	5.64 ± 1.11	0.040
Alkaline phosphatase (80-644U/L)	323.27 ± 134.6	279.25 ± 63.6	267.41 ± 41.3	0.034
Sodium (135-150mEq/l)	136.58 ± 4.56	138.06 ± 3.05	139 ± 1.85	0.012
Potassium (3.5-5.2mEq/l)	4.15 ± 0.55	4.36 ± 0.4	4.41 ± 0.34	0.040
Urinary sodium (40-220 mmol)	61.85 ± 14.23	74.53 ± 13.31	75.78 ± 13.9	<0.001

Normal accepted values are expressed between brackets for studied parameters.

CRP: C-reactive protein; HCT: hematocrit; HB: hemoglobin; MCH: mean corpuscular hemoglobin; MCV: mean corpuscular volume; PLT: platelet count; TLC: total leucocytic count.

The achieved weight, height or BMI z-scores of the studied cohort was not statistically different from that achieved by the nutritional support without recipe cards ($p=0.485$, $p=0.499$ and $p=0.227$ respectively). Mean care taker nutritional counseling session time was 17.9 ± 3.8 min for those who received the recipe cards, which was significantly shorter than the mean (48.2 ± 7.3 min) of the other group ($p < 0.001$). Both groups were compliant to diet.

Table 3. Correlations of serum sodium with anthropometric parameters at baseline and follow up.

	Baseline serum Sodium		Follow up serum Sodium	
	r	p-value	r	p-value
Weight for age (z-score)				
• Baseline	0.379	0.032	0.136	0.466
• Follow up	0.361	0.042	0.237	0.199
Height for age (z-score)				
• Baseline	0.349	0.046	0.098	0.594
• Follow up	0.348	0.047	0.224	0.218
MUAC for age (z-score)				
• Baseline	0.499	0.003	0.288	0.110
• Follow up	0.416	0.016	0.291	0.106

MUAC: mid-upper arm circumference.

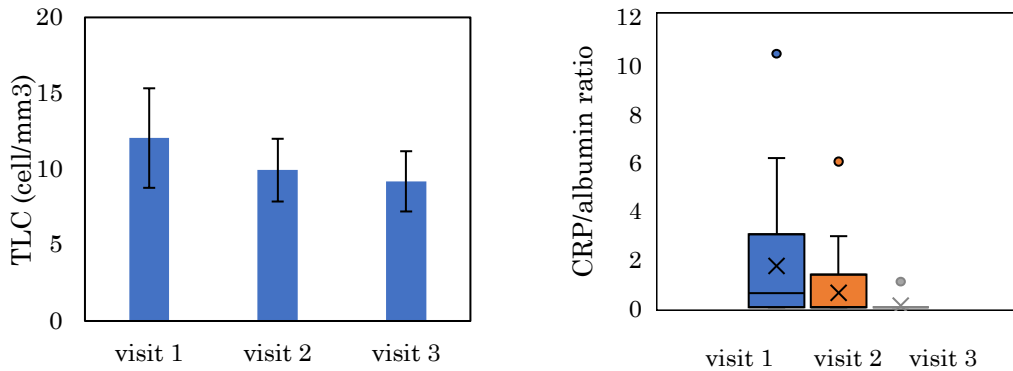


Figure 3: Total leucocytic count C-reactive protein/ albumin ratio of the study group.

Table 4. Comparison of anthropometric parameters initial visit and follow up according to use of readymade recipe cards.

	Group that used readymade Recipe Cards		Group that did not use readymade Recipe Cards		P value
	Mean	SD	Mean	SD	
<i>Initial measurements at visit 1</i>					
Weight z-score	-3.49	1.8	-2.13	1.24	0.288
Height z-score	-2.49	2.2	-3.25	1.5	0.146
BMI z-score	-1.57	0.909	-0.5	0.909	0.0055
<i>Initial measurements after 6 months</i>					
Weight z-score	-3	1.68	-1.76	1.46	0.485
Height z-score	-1.81	2.23	-1.81	1.77	0.499
BMI z-score	-0.77	1.07	-1.03	0.860	0.227

Discussion

High fat diets are established as a an essential nutritional treatment in CF (9). Many diet plans have been adopted by children with CF that included high fat content, but were associated with complications as increased risk of cardiovascular disease, or difficulties in child compliance (10). Our devised nutritional protocol was accepted by the children and their care givers.



initially, 64% of the study group were underweight and 70% had stunted growth. The high prevalence of malnutrition in our patients might be attributed to later diagnosis being a rare disease in Egypt (2) as cystic fibrosis is not included in the Egyptian neonatal screening program. Hence, malnutrition might be attributed to later age at presentation upon referral to our tertiary center, or the lack of a structured meal plan, as the malnutrition frequency was reported to be far less by others (11, 12).

It is interesting that the recipe cards of meals did not improve the growth parameters. All anthropometric parameters improved after an average of six months of structured and calculated high fat, high calorie, salt supplementation and optimum pancreatic enzyme dosage especially the weight and height and middle-upper arm circumference, irrespective of mode of delivery of instructions and nutritional counseling. Underweight and stunting decreased from 64% and 70% to 45% and 58% respectively which was reported in earlier study (6). Yet, the cards allowed delivery of information in shorter nutritional counselling sessions. It seems that the printed ready-made recipe cards allowed easier and more convenient demonstration to care taker. Visual aids for nonformal education has been long recognized as a definite tool (13, 14). The time of physician-patient caretaker discussions to achieve growth decreased by almost 63% by using the readymade recipe cards. The visual aids not only reduces need for verbal communication, but crosses barriers of language and illiteracy (14).

It is interesting that there was adherence and compliance to high fat, high calorie diet, salt supplementation and pancreatic enzyme dose. Anthropometric assessment is essential to follow up the adherence and compliance to therapy and diet, we adopt regular every 3 months visits. We have not evaluated if shorter or longer term visits would achieve better effect on the growth parameters. Yet, the achieved improvement in our studied group remains humble compared to others. Our findings support that the earlier institution of suitable diet is an important determining factor of growth and development in children with CF (15). Hence, the need for inclusion of CF in the Egyptian neonatal screening program (16).

We did not assess impact of the diet on lipid profile, or blood pressure assessments, or on mental wellbeing, mental development as they were beyond the scope of this study. It is not clear if the diet needs to be tailored to achieve better improvement among children with CF.

Author Contributions:

All authors shared in conceptualization, supervising, data curation, data analysis, writing original draft, data interpretation, writing original draft, supervising and revising. All authors reviewed the final manuscript. All authors have read and agreed to the published version of the manuscript.

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CONFLICT OF INTEREST

The authors declare no conflict of interest in connection with the study.

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