

# Development of a Diet Management Scale for Pediatric Patients with Citrin Deficiency

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**Abstract:** Objective: This study aimed to develop a scale to assess the caregivers' ability to manage the dietary needs of pediatric patients with citrin deficiency (CD). Methods: 24 experts from 11 hospitals in mainland China participated in the Delphi research. Item analysis was performed on 61 caregivers of CD patients who were enrolled in the testing of the draft scale, and 153 caregivers attended the validity and reliability testing. The initial scale was made according to the suggestions by the 24 experts. Retest reliability method and Cronbach's  $\alpha$  coefficient method were used to evaluate the external and internal reliability of the scale, respectively; the scale validity was evaluated by content validity and construct validity. Results: A consensus was reached by 24 experts after two rounds of investigation, and a preliminary draft of the scale was formed consisting of three dimensions: Special dietary knowledge, Healthy dietary attitude and Dietary behavior. The expert authority coefficient was 0.887, and the Kendall coefficient was 0.201. The final scale Cronbach's alpha coefficient ( $\alpha = 0.713$ ) showed good internal consistency and balance stability. The retest reliability coefficient of the scale was 0.975, indicating good external stability. The total content validity index (S-CVI) was 0.981, and the content validity index (I-CVI) of each item varied between 0.913 and 1.0. On confirmatory factor analysis, the indicators CMIN/DF, RESAE, PNFI, NFI, TLI, IFI and CFI for the construct validity of the scale were 1.191, 0.053, 0.628, 0.796, 0.940, 0.954, and 0.951, respectively. Conclusion: The scale developed in this paper exhibited promising reliability and validity for the clinicians to evaluate the dietary management ability of the caregivers of pediatric CD patients.

**Keywords:** Scale, Citrin Deficiency, Diet Manage, Caregiver

## 1. Introduction

Citrin deficiency (CD) is a genetic metabolic disease caused by biallelic mutations of SLC25A13 gene [1]. This disease has three clinical phenotypes: Neonatal Intrahepatic Cholestasis caused by Citrin Deficiency (NICCD) [1], Failure to Thrive and Dyslipidemia caused by Citrin Deficiency (FTTDCD) [2], and adult-onset citrullinemia type II (CTLN2) [1]. Previous studies have shown that excessive carbohydrate intake is harmful to CD patients [3-5]. Management of CD requires adherence to a low-carbohydrate while high-protein and high-lipid dietary therapy [6-8]. Growth impairment may occur in children if the diet is not properly managed [9].

Dietary management is the major therapeutic measure for CD, an inborn error of metabolism arising from biallelic mutations of the gene SLC25A13. The effect of dietary management in pediatric patients with CD depends on the management ability of their caregivers; however, to the best of our knowledge, there is currently no specific scale to evaluate such a ability. To address this issue, this study aimed to develop a diet management scale for the caregivers of pediatric patients with CD. We hope that through this scale, caregivers will be able to manage the diet of patients, improve the management abilities of caregivers, and improve the quality of life of children.

## 2. Methods

### 2.1. Study Design

After conducting a thorough literature review to form a pool of potential questions for the scale, experts screened the pool of entries using the Delphi method. Then exploratory factor analysis was done, and questions were divided into 4 dimensions. Finally analysis on the reliability and validity of the scale was completed.

### 2.2. Construction of the Scale Items Pool

Through the literature review, related field expert consultation and group discussion, 31 items were extracted. Among them, 8 items involved the dietary knowledge dimension, 6 items the dietary belief dimension, and 17 items in the eating behavior dimension.

### 2.3. Delphi Expert Consultation

#### 2.3.1. Investigate Experts Into the Standard

Qualifications for the Delphi experts included: 1) Working in a tertiary hospital or college; 2) Acquired intermediate titles; 3) Genetics research in the field of hepatology; 4) Researcher in the field of nutrition; 5) Specialist nurse; 6) Agreement to participate in the research study.

#### 2.3.2. Delphi Expert Questionnaire Standard

The importance of each item on the scale is represented by the mean of the expert score ( $\geq 4$  is important) [10]. The degree of expert authority is represented by the expert authority coefficient (Cr) ( $\geq 0.7$  is considered good) and the positive degree of an expert is represented by questionnaire recovery rate and expert opinion. Coefficient of variation (CV)  $> 0.25$  indicates poor consistency of expert scores. Expert opinion dispersion degree is represented by Kendall's W (0-1). A larger value indicates better coordination.

#### 2.3.3. Delphi Questionnaire Items Deletion Criteria

Items that normally meet any of the following requirements are deleted, depending on the actual situation [11]. This study used: 1) Coefficient of variation  $> 0.25$ ; 2) Mean of expert ratings  $< 4$ ; 3) Outer number of items  $< 20\%$ .

### 2.4. Formation of the Citrin Defective Diet Management Scale

Literature shows that the sample size can be 3 times the number of items [12]. In this research, caregivers of children diagnosed with CD were the test subjects. The investigator explained to each subject the purpose and method of filling out the questionnaire on the "questionnaire star" APP. Items were scored according to the following criteria: *Dimensions 1 scoring criteria*: option plus one point, error option minus 0.5 points; *Dimension 2 scoring criteria*: items were highly dissatisfied, disapproved, uncertain, agree, and strongly endorsed according to the Likert 5 rating; *Dimension 3* items were assigned 1-5 points for never, occasionally, average, often

and always according to the Likert 5 rating scale. The scores of the scales were ranked from high to low, and then the independent sample t test was used to compare the scores of the items in the top 27% (high group) and the last 27% (low group) items, with the retention decision being (CR) value  $> 3$  ( $P < 0.05$ ) [13]. Exploratory factor analysis was then completed to determine the dimension of the scale based on the results of exploratory factor analysis, to optimize the scale structure. The Kaiser-Meyer Olkin (KMO value) and Bartlett sphere test (KMO value  $> 0.7$ ) and Bartlett's spherical test were statistically significant ( $P < 0.05$ ), indicating that items were suitable for the factor analysis [13]. Principal component analysis, skew rotation axis, maximum variation method are used to automatically extract the factor root ( $> 1$ ) and factor load ( $> 0.4$ ) for the factor matrix. A steep slope map helped determine the number of factors, delete multiple loads and the load values that were close. A factor of less than 3 items, after each factor analysis, necessitated re-assessment of the item d to determine if it should be deleted. Each time one item was deleted, the factor analysis was performed again, and the dimension of the item was finally adjusted. When the total variation (the extracted factors combined to explain all variables) reached 50%, the results were acceptable, and if it reached 60%, the factors retained after extraction were ideal [13].

### 2.5. Instrument

This data was complied with Excel 21.0 and SPSS21.0 for data analysis. Communication with subjects was completed using E-mail. The caregiver questionnaire was distributed through questionnaire software.

### 2.6. Sampling

#### 2.6.1. Expert Source

The 24 experts who accepted participation in this study the inquiry were from 10 provinces, autonomous regions and municipalities in China. All experts were from tertiary hospitals or higher education institutions. In addition, one expert as a specialist nurse and the other 23 experts were either senior nursing staff or senior faculty.

#### 2.6.2. Source of Caregivers for Children with Citrin Deficiency

The caregivers who participated in the study were caregivers of children diagnosed with CD. They were recruited from the pediatric outpatient department of the First Affiliated Hospital of Jinan University in China between January 2015 to September 2019.

### 2.7. Ethical Considerations

This study was approved by the Ethics Association of the First Affiliated Hospital of Jinan University. All questionnaires were anonymous and all the caregivers in the survey had signed informed consent forms.

### 3. Results

#### 3.1. Delphi Expert Consultation Questionnaire Results

##### 3.1.1. Expert Enthusiasm

Of the 24 experts interviewed in this study, one was from a college, 23 from hospitals, 2 nursing experts and 22 clinical medical experts. Expert research interests included gastroenterology, nutrition, hepatology, and chronic disease management. For the 2 rounds of Delphi inquiry 24 packets were distributed. In the first round the effective recovery rate of 100%, and the expert opinion rate was 58.3%. In the second round 23 experts received materials and the effective recovery rate was 95.8% and the expert opinion rate was

52.2%.

##### 3.1.2. Expert Authority

Result of the Expert authority are shown in Table 1.

Table 1. Expert authority.

Stage	Judgment coefficient (Ca)	Familiarity (Cs)	Authority coefficient (Cr)
Round 1	0.942	0.858	0.900
Round 2	0.948	0.826	0.887

##### 3.1.3. Degree of Coordination of Expert Opinions

Result of the degree of coordination of expert opinions are shown in Table 2.

Table 2. Degree of coordination of expert opinions.

Round 1			Round 2		
	Kendall's W	P		Kendall's W	P
Dimension	0.187	P<0.05	Dimension	0.201	P<0.05
Items	0.212	P<0.00	Items	0.30	P<0.00

##### 3.1.4. Second Round of Expert Inquiry Results

Result of the expert inquiry in second round are shown in Table 3.

Table 3. Expert inquiry in second round.

Dimension/Items	Average (Mj)	Full ratio (Kj)	Coefficient of variation (Vj)
A. Special dietary knowledge	5.00±0.00	1.00	0.00
A1. Patients should reduce their intake of carbohydrate food. Which of the following foods are rich in carbohydrate?	4.70±0.47	0.609	0.10
A2. Encourage children to eat foods with higher protein. Which of the following foods are rich in high quality protein?	4.61±0.50	0.609	0.11
A3. Encourage children to eat higher fat foods. Which of the following foods are rich in fat?	4.48±0.51	0.478	0.11
A4. High-sugar fruits are rich in carbohydrates, which of the following should be reduced for children?	4.22±0.60	0.304	0.14
A5. Starchy foods are rich in carbohydrate. Which of the following root vegetables with higher starch content should be reduced for children?	4.39±0.50	0.391	0.11
A6. Which of the following snacks don't require special restrictions?	3.91±0.67	0.174	0.17
A7. Nuts are rich in protein and fatty acids. Which of the following nuts can be given priority in older children?	4.04±0.71	0.217	0.17
Healthy dietary attitude	4.70±0.47	0.696	0.10
B1. Follow the principle of high protein, high fat and low carbohydrate diet to help children with NICCD recover.	4.83±0.39	0.826	0.08
B2. Forcing adolescent children to eat carbohydrate-rich foods may induce adult onset citrullinemia type II.	4.09±0.95	0.435	0.23
B3. Stopping breastfeeding after diagnosis is beneficial to liver function recovery in children with NICCD.	4.74±0.45	0.739	0.09
B4. Stopping breastfeeding after diagnosis is beneficial to stabilize blood lipids in children with NICCD.	2.83±1.64	0.174	0.58
B5. Add food rich in protein and fat to help children grow and develop.	4.26±0.45	0.261	0.11
B6. Follow the principle of low-carbohydrate, high-protein and high fat at 4-6 months, which is beneficial to the growth and development of children with NICCD.	4.61±0.58	0.652	0.13
Dietary behavior	4.83±0.39	0.826	0.08
C1. After the baby is diagnosed, I will follow the doctor's advice to replace lactose-free, medium-chain triglyceride milk powder instead of breastfeeding.	5.00±0.00	1.0	0.00
C2. I will choose the complementary food/staple C3. Food for the children according to the priority order of meat, eggs, fish, beans and dairy products.	4.48±0.59	0.522	0.13
C3. I regularly record the diet and nutrition of each stage of the child.	3.83±0.89	0.304	0.23
C4. I will work with other caregivers in the family to adhere to the child's low-carb, high-fat diet.	4.26±0.75	0.391	0.18
C5. I will not correct the child's eating behavior of eating meat and eggs, milk and soy products.	4.35±0.94	0.522	0.21
C6. Under the guidance of medical staff, I adjusted the type of diet according to the recovery of liver function in children.	4.43±0.66	0.174	0.15
C7. I will follow the doctor's instructions to calculate the total amount of staple food	3.74±1.01	0.174	0.27

Dimension/Items	Average (Mj)	Full ratio (Kj)	Coefficient of variation (Vj)
that the child needs to add.			
C8. I will adjust the child's diet according to the nutritional status of the child.	4.00±0.95	0.304	0.24
C9. Children with this disease have special dietary preferences. I will not force children to eat foods rich in carbohydrates, noodles, etc.	4.70±0.56	0.739	0.12
C10. I will not test the tolerance of children to rice noodles.	3.65±0.83	0.174	0.23
C11. I will ask the medical staff about my doubts about diet management.	4.52±0.79	0.652	0.17
C12. If the child enters school, I will communicate with the teacher and tell the principle of eating.	4.57±0.73	0.696	0.16
C13. If the child enters school, I will prepare a high-protein, high-fat, low-carbohydrate meal for the child to take to school.	3.48±1.04	0.217	0.30
C14. If the child enters school, I will train the child's special diet knowledge and self-catering management skills.	4.70±0.64	0.783	0.14
C15. I will actively learn about Citrin deficiency disease nutrition management.	4.61±0.50	0.609	0.11
C16. If the child enters school, I will tell the child about the toxicity of alcohol to the liver.	3.70±1.11	0.348	0.30
C17. If the child's disease is improved, I will try to add lactose-containing dairy products to the child.	3.13±1.10	0.130	0.35
C18. I will pay attention to whether the food ingredients are harmful to the health of the children.	4.26±0.81	0.478	0.19

### 3.2. The Results of Scale Entry Screening

#### 3.2.1. Item Analysis Result

A total of 80 questionnaires were distributed to caregivers of children with CD, and the 61 questionnaires were effectively recovered. The effective recovery rate was 76.3% in this study.

The frequency analysis of the answers to the 26 entries of the initial scale shows that the selection rate of the other items is less than 80% except that the selection rate of the original item C14 is 94% (>80%). Entry C14 was deleted and the remaining entries were retained for further analysis.

#### 3.2.2. Cronbach's $\alpha$ Coefficient Result

The Cronbach's  $\alpha$  coefficient results for each dimension on the first draft of the scale are shown in Table 4. The Cronbach's  $\alpha$  coefficient of the corresponding dimension and totals after deleting the entry is shown in Table 5. The result shows that after deleting the item A3, the Cronbach's  $\alpha$  coefficient of dimension one rises to 0.767, which is higher than the original 0.740, so A3 was deleted. After deleting the item A8, the dimension Cronbach's  $\alpha$  coefficient of one rises to 0.745, which is higher than the original 0.733. After

deleting the item B13, the Cronbach's  $\alpha$  coefficient of dimension two rises to 0.903, which is higher than the original 0.725. After deleting the item C14, the Cronbach's  $\alpha$  coefficient rose to 0.767, higher than the original 0.733, and was deleted. After deleting the item C18, the Cronbach's  $\alpha$  coefficient of dimension three rose to 0.768, which was higher than the original 0.731. After deleting the item C19, in dimension three, the Cronbach's  $\alpha$  coefficient rises to 0.770, which is higher than the original 0.732. After deleting the item C21, the Cronbach's  $\alpha$  coefficient of dimension three rises to 0.777, which is higher than the original 0.736. The specific values are shown in Table 5.

**Table 4.** Cronbach's  $\alpha$  coefficient results on the first draft of the scale.

Dimension	Number of items	Cronbach's $\alpha$ coefficient
Dimension 1	8	0.733
Dimension 2	5	0.871
Dimension 3	13	0.779

Dimension 1 represents Special dietary knowledge; Dimension 2 represents Healthy dietary attitude; Dimension 3 represents Dietary behavior.

**Table 5.** Result of the Item filtering.

Items	Overall Scale (0.786)	Dimension 1 (0.733)	Dimension 2 (0.871)	Dimension 3 (0.779)	Retention
A1	.770	.702			Reserved
A2	.767	.683			Reserved
A3	.798	.750			Delete
A4	.764	.665			Reserved
A5	.776	.695			Reserved
A6	.788	.736			Delete
A7	.772	.661			Reserved
A8	.788	.742			Delete
B9	.778		.834		Reserved
B10	.779		.840		Reserved
B11	.776		.823		Reserved
B12	.775		.807		Reserved
B13	.774		.921		Delete
C14	.787			.785	Delete
C15	.780			.759	Reserved

Items	Overall Scale (0.786)	Dimension 1 (0.733)	Dimension 2 (0.871)	Dimension 3 (0.779)	Retention
C16	.773			.779	Reserved
C17	.782			.759	Reserved
C18	.784			.778	Reserved
C19	.785			.777	Reserved
C20	.779			.743	Reserved
C21	.791			.789	Delete
C22	.783			.746	Reserved
C23	.779			.744	Reserved
C24	.776			.747	Reserved
C25	.776			.747	Reserved
C26	.778			.745	Reserved

(Consider deleting the underlined items in the Table5).

### 3.3. Final Scale Reliability and Validity Test

#### 3.3.1. General Situation of the Research Object

A total of 135 questionnaire were distributed to caregivers of children with CD, and the 133 questionnaires were effectively recovered. The effective recovery rate was 98.52% in this study.

#### 3.3.2. Exploratory Factor Analysis Results

The KMO value is 0.727, and the Bartlett spherical test  $\chi^2 = 636.181$ ,  $P < 0.00$ , suitable for line factor analysis. Principal component analysis and skewed axis automatically propose 4 common factors (feature value  $> 1$ ). In Figure 1, the gravel map slows down after the fifth factor.

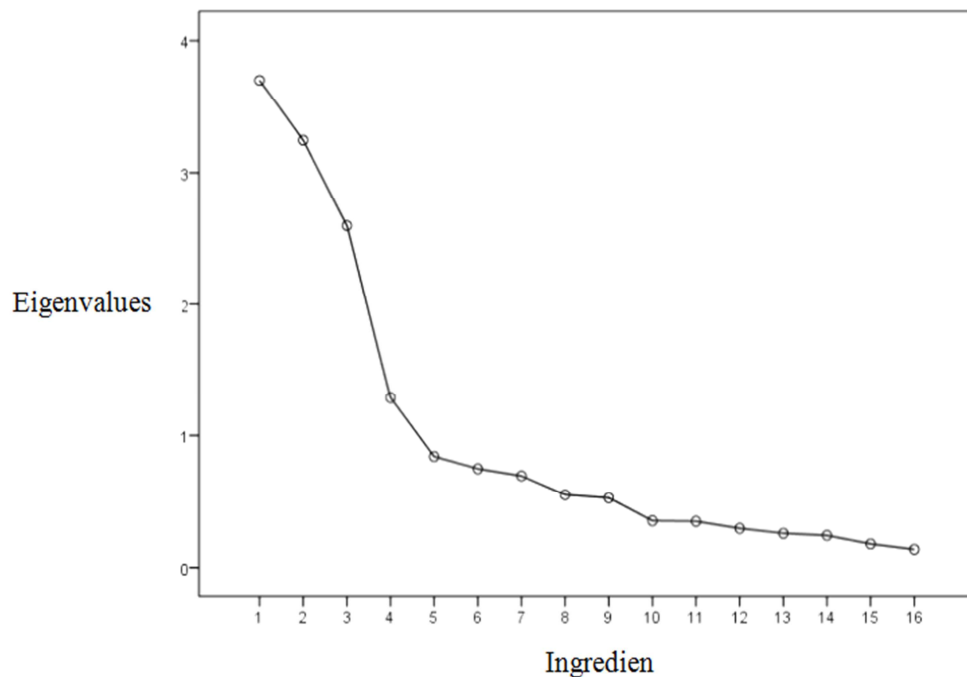


Figure 1. Gravel map.

#### 3.3.3. Reliability Analysis

Result of the Retest reliability are shown in Table 6. Result of the Cronbach's  $\alpha$  coefficient are shown in Table 7.

Table 6. Result of the Retest reliability.

Dimension	First Score	Second Score	Pearson Correlation Coefficient	P
Special dietary knowledge	24.23±7.78	24.37±7.17	0.990	<0.000
Healthy dietary attitude	18.50±1.82	18.37±1.56	0.893	<0.000
Dietary behavior	18.37±3.80	18.33±2.91	0.916	<0.000
Risk prevention	8.23±2.19	8.27±2.10	0.997	<0.000
Total Scale	71.92±9.31	69.33±9.28	0.975	<0.000

Table 7. Result of the Cronbach's  $\alpha$  coefficient.

Dimension	Number of items	Cronbach's $\alpha$ coefficient
Special dietary knowledge	5	0.756
Healthy dietary attitude	4	0.884

Dimension	Number of items	Cronbach's $\alpha$ coefficient
Dimension 3	2	0.746
Dietary behavior	5	0.822
Total Scale	16	0.713

### 3.3.4. Validity Analysis

The total content validity index (S-CVI) is 0.981, and the content validity index (I-CVI) of each item is between 0.913 and 1.0. The results of confirmatory factor analysis showed that the model was slightly modified by the correlation of the residual terms. The corrected structural equation model is

shown in Figure 2. Except for  $NFI=0.769<0.9$ ,  $CMIN/DF=1.191$ ,  $RMSEA=0.053$ ,  $PNFI=0.628$ ,  $TLI=0.940$ . The fitting indexes such as  $IFI=0.954$  and  $CFI=0.951$  are all within the reference range, and the model has a good matching degree with the scale (Figure 2).

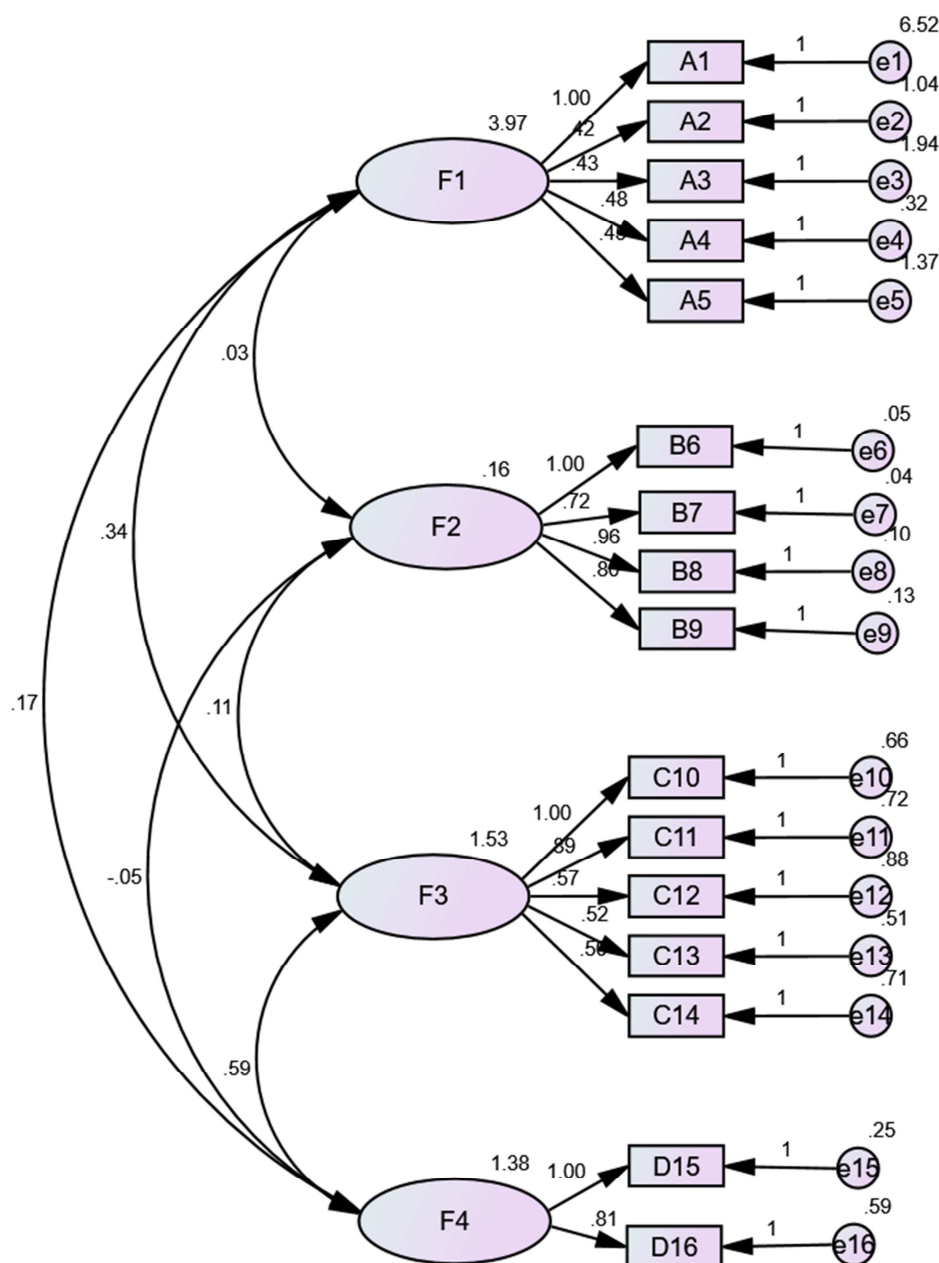


Figure 2. Verification factor analysis structural equation model diagram.

## 4. Discussion

With the continuous advancement of molecular diagnostics,

more and more patients with Citrin deficiency have been diagnosed [14-16]. At present, the clinical research on this disease mostly focuses on molecular diagnosis, clinical manifestations and treatment. Children as a special group lack

self-management ability, and their management ability depends on the caregiver, but there is currently no clinical scale to evaluate the caregiver's ability to manage the diet of children with citrin deficiency.

#### **4.1. Rationality Analysis of Scale Dimensions and Items**

There are six steps in the construction of the scale, which are to clarify the purpose of measurement, assign dimensions and weights, establish the original pool of entries, select expression of the entries, pre-experiment and analysis, revision and improvement [13]. This study is guided by the above steps to ensure the rigor of the scale development. In this study, an expert correspondence form consisting of 3 dimensions and 31 items was formed through literature review, expert consultation in related fields, and group discussions. The dimensions and items of the scale were modified through two rounds of Delphi expert correspondence. The caregivers of children with Citron deficiency disease tested the expression of the items, and finally formed an initial scale of 3 dimensions and 31 items.

#### **4.2. Reliability Analysis of Expert Consultation**

The experts invited in this study are from 10 provinces, autonomous regions, and municipalities directly under the Central Government in China. The institutions represented are all tertiary hospitals or colleges and universities. In addition to one professional nursing specialist, the rest of the experts held titles of deputy senior and above. They all had profound knowledge and rich experience in clinical nursing, clinical nutrition and hereditary liver disease ensuring the representativeness and authority of the Delphi expert consultants.

The recovery rates of the two questionnaires were 100% and 95.8%, and the expert opinion submission rates were 58.3% and 52.2% in this study. The results show that the experts participating in the survey were highly motivated and the consultation results are reliable. With the expert authority coefficient ( $Cr$ )  $\geq 0.70$ , the consultation result is authoritative [10]. The  $Cr$  values of the two rounds of consultation in this study were 0.900 and 0.887 meaning there was a high degree of expert authority.

The coefficient of variation ( $v_j$ ) and Kendall's  $W$  are used to indicate the degree of coordination of expert opinions,  $v_j < 0.25$ , and Kendall's  $W$  value is 0-11 [17]. The dimensions of Kendall's  $W$  are 0.187 and 0.201, the Kendall's  $W$  of the items are 0.212 and 0.30 ( $P < 0.05$ ) in the two surveys of this study. The level of expert coordination maintained an upward trend in this study.

#### **4.3. Analysis of the Screening Process for Items**

Screening was performed again by item analysis, Cronbach's  $\alpha$  coefficient method, and exploratory factor analysis in this study.

The item analysis method is to test the difference between the scores of the high- and low-level survey respondents on each item. In order to evaluate whether the items have good

discrimination and 27% is usually used as the cutoff value [13]. All items meet the screening criteria in this survey ( $CR > 3.0$ ,  $P < 0.001$ ) meaning that all items had a high degree of discrimination for survey subjects at different levels, and no items were deleted by this method.

Cronbach's  $\alpha$  method performs item screening from the perspective of internal consistency. The principle of this method is that if an entry is deleted, Cronbach's  $\alpha$  is rises, which means that the item will reduce the internal consistency of the scale, and this entry should be deleted. The researchers deleted the items A3, A8, B13, C14, C18, C19, and C21, to assure the internal consistency of the scale.

Exploratory factor analysis screens items from a representative perspective. If an item has a large load on a common factor, that item can reflect the characteristics of the dimension represented by the common factor, but if an item has a large load on two or more common factors, the attribute of the entry is not strong and the representation is poor. Exploratory factor analysis was repeated 7 times, and 4 common factors were extracted. Finally, the dimensions of the scale were re-divided.

#### **4.4. Evaluation**

The feasibility of the scale is expressed by the recovery rate and efficiency. Usually, the effective recovery rate should be  $\geq 85\%$ , and the effective recovery rate is 86.6% in this survey, and means the scale has high acceptability. Cronbach's  $\alpha$  is an index used to evaluate the degree of internal consistency of the scale, and it is required to be  $> 0.7$ . Cronbach's  $\alpha$  in each dimension is 0.786, 0.733, 0.871, 0.779 in this study, and indicates that the internal consistency of this scale is high. The retest reliability coefficient of the scale is 0.975, which shows good external stability. The total content validity index (S-CVI) is 0.981, and the content validity index (I-CVI) of each item is between 0.913 and 1, which showed the high content validity in this study. Confirmatory factor analysis was used for structural inspection of the scale. The researchers slightly modified the model, and the fitting indicators such as IFI and CFI were within the reference range, indicating that the model matches the scale well and the scale results are good.

## **5. Conclusions**

The first draft of the citrin deficiency disease diet management scale was formed through two rounds of the Delphi method, and consisted of 3 dimensions and 31 items. The final scale was formed by re-screening all items through classical measurement theory, which ultimately consisted of 4 dimensions and 16 items. Screening methods included item analysis, Cronbach's  $\alpha$  coefficient method, and factor analysis. All analyses indicated the scale had high reliability and validity for determining the caregivers, of children with CD, attitudes, knowledge and behaviors concerning dietary practices. The scale can be confidently used to evaluate the caregiver's ability to manage a child's dietary needs when dealing with CD.

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