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Research Article

Nutritional Status of Children with Cystic Fibrosis Followed in Consultation

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Abstract

Cystic fibrosis is the most common genetic disease in children of transmission, autosomal recessive. The nutritional status is a major clinical element of monitoring, prognosis and management, with this in mind, we carried out this study in order to evaluate the dietary behavior of children with cystic fibrosis , to describe their quantitative and qualitative calorie intake in macro and micronutrients and to compare their nutritional status with normal children of the same age through a case-control study involving 30 children aged 2 to 16 years (10 with cystic fibrosis and 20 control children). The study took place between February 1 and May 31, 2019.

Results: Of the 10 patients interviewed, there are 6 boys and 4 girls. The average age was 4 years with extremes of 3 months to 12 years. 80% of children with cystic fibrosis were underweight, unsatisfactory observance of the diet was objectified in relation to the recommended nutritional intake. Daily energy intake of cystic fibrosis children was lower than that of normal children. All patients have a low plasma level of HDL (0.25 ± 0.05 g / l) and cholesterol (0.98 ± 0.26 g / l) , while the level of Triglycerides (0.45 ± 0.23g / l) was within the limit lower. All children with cystic fibrosis regardless of their age have a deficiency in fat-soluble vitamins compared to normal children. All children with cystic fibrosis presented a deficiency in Zinc, Selenium and Magnesium and in Calcium and Sodium. Water intake was insufficient, with averages below standards.

onclusion: It is imperative to advise patients to take 3 balanced meals per day and a minimum of 2 snacks per day, with high calorie foods. In addition, it is necessary to set up dietary education sessions for better nutritional intake to compensate for dietary errors that can lead to sometimes dreadful consequences.

INTRODUCTION

Cystic fibrosis is a potentially serious genetic disease from the pediatric age. Its incidence is variable, it is more common in the Caucasian population of northern and central Europe. It affects an average of 1 child in 2,500 births, of which 1 in 25 is heterozygous. The pathology is much rarer in Asian or African populations. Its exact incidence in Algeria is unknown due to a lack of studies [1].

Cystic fibrosis is linked to mutations in the CFTR gene (Chromosome 7), responsible for alterations of the CFTR protein whose function is to regulate the transmembrane transport of chlorine. Autosomal recessive monogenic pathology, more than 2000 mutations have been identified including *F508del* responsible for 80% of cases of cystic fibrosis. In addition to allelic heterogeneity and the occurrence of multiple mutations in the same gene, a wide variety of other factors can influence the phenotype. The absence of functional CFTR protein in the membrane of epithelial cells leads to abnormally viscous mucous secretions responsible for stasis, obstruction responsible for pathological changes in the various organs expressing CFTR protein such as sweat glands, lungs, pancreas , liver, intestines and genital tract [2,3].

Nutritional status is a major clinical element in the monitoring, prognosis and management of cystic fibrosis. Nutritional management is essentially based on supplementation with pancreatic and vitamin extracts, an appropriate diet and in some cases by more aggressive treatment with enteral nutrition [3].

METHOD

Case-control, prospective, cross-sectional, descriptive and analytical study involving 30 children aged 2 to 16 followed in pediatric pneumology consultation in the department. These children were divided into two groups: cystic fibrosis children (n=10) and normal children (n=20). The study took place between February 1 and May 31, 2019. The objectives of the study were to assess the eating behavior of children with cystic fibrosis, to describe their quantitative and qualitative caloric intake in maro and micronutrients and to compare their nutritional status with normal children of the same age.

The data was collected via a questionnaire comprising several items after informed consent from the parents. During the interview; the following information was collected: age, sex, history of illness. The dietary survey was conducted using the

3-day recall technique. From the food records, the frequency of food consumption of the different groups was determined and the conversion of food into nutrient was carried out. The frequency of taking snacks, number of meals were noted. Basal metabolic rate was calculated; for each of the patients, from the equations of Harris and Benedict. Blood samples were taken for the evaluation of: total cholesterol, HDL, LDL, triglyceride, total protein. Data analysis was performed using SPSS version 25 software. A value of p <0.05 is considered significant.

RESULTS

Of the 10 patients questioned, there are 6 boys and 4 girls with a sex ratio of 1.5. The average age was 4 years with extremes of 3 months to 12 years. The notion of consanguinity is found in 9 out of 10 patients.

Of the 20 control children, there are 10 boys and 10 girls with a sex ratio of 1. The average age was 5 years with extremes of 2 months. 80% of children with cystic fibrosis are underweight and only 20% are normal weight, on the other hand, 85% of children in the control group are of normal build and 5% are thin. 60% of patients present with digestive manifestations associated with respiratory manifestations, 40% suffer from pure digestive manifestations.

Our results indicate an unsatisfactory observance of the diet in relation to the recommended and recommended nutritional intakes (per day and per week). 60% of patients take 3 meals with 1 snack. While 30% take only 2 meals a day. A significant difference (p=0.03), was found between the 2 groups of children regarding the frequency of cereal consumption. Half of children with cystic fibrosis do not consume cereals daily and only (20%), consume cereals more than twice a day. The consumption of fruits and vegetables was significantly different between the 2 groups of sick and healthy children. However, the majority of children with cystic fibrosis respect the daily consumption of fruits and vegetables either once or several times a day, i.e. 40% and 30% respectively, while only 30% do not respect these recommendations. A significant difference (p=0, 04) was found between the 2 groups of children regarding the frequency of consumption of dairy products. 46.6% of children with cystic fibrosis consume dairy products more than twice a day and 23.3% do not consume these foods daily.

The consumption of meat, eggs, fish was significantly different (p=0.006) between the two groups of children. It is noted that the majority of children with cystic fibrosis consume meat, eggs, fish twice a week. These children prefer the consumption of eggs over meat and organ meats, while fish are rarely eaten.

A significant difference (p=0.04) was found between the 2 groups of children regarding the frequency of fat consumption. 20% of patients do not consume fat daily.

Regarding the frequency of consumption of sweet products, a very significant difference (p=0.008), was found between the two groups of children with cystic fibrosis and normal children. We note that the majority of children with cystic fibrosis consume sweet foods daily either once a day or several times a day (respectively 40% and 30%). 30% do not consume these foods daily.

Table 1 below shows that the daily energy intake of CF children is lower than that of normal children. It is also noted that there is a significant difference between the energy intake of sick and healthy patients.

Table 2 below shows that the contribution of the different macronutrients in the energy intake of the two groups of subjects is respected. In addition, the carbohydrate intake of the two groups is lower for children with cystic fibrosis and normal for healthy children. However, the percentages of the lipids of the two groups are within the norms.

All patients have a low plasma level of HDL (0.25 ±0.05 g / l) and cholesterol (0.98 ±0.26 g / l) , while the level of Triglycerides (0.45 ± 0.23g / l) is in the lower limit . This means that there is a fat malabsorption caused by a pancreatic deficit. We find that all children with cystic fibrosis, regardless of their age, have a deficiency in fat-soluble vitamins compared to normal children and also to recommended intakes. Below are the data summarized in Table 3.

We found that all children with cystic fibrosis are deficient in Zinc, Selenium and Magnesium and in Calcium and Sodium (Table 4).

Water intake is insufficient, with averages below standards in the two groups studied (Table 5).

DISCUSSION

80% of children with cystic fibrosis were underweight, our results are similar to literature data which show that 90% of cystic fibrosis children were stunted in a study carried out in 2014 by Le Bourgois [2].

The consumption of fruits and vegetables was significantly different between the 2 groups of sick and healthy children. Strandvik [4], shows that the consumption of fruits and vegetables is essential in cystic fibrosis and this to correct the antioxidant deficit. A study by Munck [5], indicates that the intake of fruits rich in vitamin C is important. Indeed, vitamin C is involved in the stimulation of the immune system, and thus helps the body to better cope with microbial infections which are favored during cystic fibrosis.

A significant difference (p=0.04), was found between the 2 groups of children concerning the frequency of consumption of dairy products . The relationship between cystic fibrosis and the consumption of dairy products has been reported by McBean(6) who shows that the ingestion of milk and dairy products promotes the production of mucus and that the latter leads to an aggravation of the symptoms of cystic fibrosis. However,

Table 1: Comparison between the two groups of children according to caloric intake.

	Children	
	cystic fibrosis	**Normals**
Daily Energy Intake (Kcal/d)	795.24±216.05	1257.06±457.82
Daily Energy Expenditure (Kcal/d)	1062±299.225	1324.21±538.30
P	0.0012	0.0048

Table 2: Daily intakes of macronutrients.

Groups Bring	Normal children (Mean ± standard deviation	Cystic fibrosis children (Mean ± standard deviation	Recommended intake (%)
Carbohydrate intake (%)	49.55 ±5.43	41.87 ±10.77	45-50
Fat intake (%)	36.27 ±6.28	35.39 ±2.48	35-40
Protein intake (%)	17.11 ±7.66	16.25 ±4.59	12-15

Table 3: Daily intake of fat-soluble vitamins.

Topics	Vit A (µg/d)	VitD (mg/d)	VitE (µg/d)	Vit K(µg/d)
Normals	328.9±160.87	6.27±9.91	3.165±1.55	18.07±8.08
cystic fibrosis	313.06±97.99	2.71±1.29	3.46±3.06	15±9.2

Table 4: Daily intake of minerals and trace elements.

Topics	Zn(mg/d)	Mg (mg/d)	Se (µg/d)	Sodium (mg/d)	Ca (mg/d)
Normals	7.525±3.3	186.825±88.6	25±5	1292.7±726.05	689.1±277.86
cystic fibrosis	4.64±1.58	111.36±34.76	19±2	998.2± 313.72	678.2±459.87

Table 5: Daily fluid intake.

Boys			**Girls**		
Normals	**cystic fibrosis**	Water (L/d) (Standards)	**Normals**	**cystic fibrosis**	Water (L/d) (Standards)
Mean (L/d) deviation			Mean (L/d) deviation		
0.78 ±0.31	0.64 ± 0.06	1.6	0.71 ±0.41	0.65 ±0.49	1.6

milk and dairy products are important sources of calcium which should not be missing in a healthy and balanced diet.

The consumption of meat, fish and eggs was significantly different (p=0.006), between the two groups of children. The relationship between the consumption of these foods and the severity of cystic fibrosis was reported by Freedman [7], who found that the fatty fish containing omega 3 have the power to reduce the inflammatory profile, they provide essential amino acids which have a role in maintaining muscle mass including that of the respiratory muscles and thus causes a reduction in respiratory function disorders.

20% of children with cystic fibrosis do not consume fat on a daily basis. The relationship between the consumption of these foods and the severity of the disease has been reported by Smyth and Panchaud [8,9], these authors advise in the case of cystic fibrosis fats of vegetable origin than those of animal origin. The consumption of healthy fats that contain Omega 3 leads to an improvement in the infectious state but also to the reduction of inflammatory flare-ups.

It is noted that the majority of children with cystic fibrosis consume sugary foods daily. The onset of diabetes is a frequent complication in patients with cystic fibrosis, so it is necessary to reduce foods with a high glycemic index as much as possible [10].

Panchaud [9], reported that in children with cystic fibrosis, energy requirements are higher than those of the general population estimated at 110 to 120% of the recommended daily intake. Corey [11], had observed that a high energy intake leads to weight gain, better lung function and longer survival, it is necessary to limit carbohydrate intake because very high carbohydrate intake risks increasing the occurrence of diabetes with the progression of the disease. Fat-soluble vitamin intake (A, D, E, K) has a more important role in cystic fibrosis patients because of their malabsorption, which is why Munch [12], objectified that supplementation with these vitamins is necessary for all CF patients . Knowing that vitamin A and E are major antioxidants and have a role in the fight against chronic lung inflammation and periods of superinfection. While vitamin D and K supplementation is also important as vitamin K deficiency is linked to altered gut flora and vitamin D deficiency induces osteopenia which can lead to osteoporosis.

All children with cystic fibrosis in different age groups have a deficiency in Zinc, Selenium and Magnesium and in Calcium and Sodium. Munck [13], demonstrated that there is a deficiency in trace elements such as Zinc, Selenium and Magnesium in children with cystic fibrosis and this is due to fat malabsorption. Furthermore, Vic [14], reported that these deficiencies can increase susceptibility to infections. Sodium requirements are higher in children with cystic fibrosis than in other children, following the loss of sodium chloride through sweat. Water intake is insufficient in children with cystic fibrosis with averages below the standards. Good hydration makes it possible to obtain bronchial secretions that are thinner, less sticky, easier to drain and expectorate and reduces the risk of constipation [15].

CONCLUSION

Children with cystic fibrosis showing growth retardation secondary to a lipid deficiency linked mainly to fat malabsorption

associated with a deficiency in fat-soluble vitamins, trace elements and antioxidants in the majority of cases.

The diet of children with cystic fibrosis must contain fruits and vegetables rich in antioxidant vitamins as well as foods high in trace elements (selenium and zinc) and dairy products, it is imperative to advise patients to take 3 balanced meals, a minimum of 2 snacks per day, with high-calorie foods such as oilseeds and dried fruits. It is currently recommended to carry out a complete biological assessment twice a year in order to correct any nutritional, electrolyte, trace element deficiencies and vitamins. In addition, it is necessary to set up dietary education sessions for better nutritional intake to compensate for dietary errors with sometimes dreadful consequences.

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