Nutritional status of Russian children with cystic fibrosis diagnosed before and after neonatal screening

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Objectives: The mandatory cystic fibrosis (CF) neonatal screening was introduced in Russia everywhere since 2006. The aim of the study was to elucidate whether early diagnosis of CF by neonatal screening had a positive influence on the nutritional status of CF children.

Methods: Weight and height of 112 CF children (216 measurements in all) under 6 years of age were recorded according to standard methods and expressed as standard deviation scores (z-score) and percentiles adjusted for age and gender using WHOAnthro- and WHOAnthroPlus programs. 45 children (93 measurements) diagnosed by symptoms formed group I and 67 children (123 measurements) born between 2006-2012 and diagnosed by neonatal screening formed group II.

Results: The mean body mass index (BMI) was higher in II group (14.89±1.86 [I] and 15.56±1.64 [II] p=0.001). The mean weight for height (37.2±4.98, p=0.016), weight for age (29.9±38.9, p=0.017) and BMI (34.6±45.8, p=0.017) percentiles were also slightly higher in group II. Height for age percentile (31.6±37.9) did not differ between groups. However, the part of stunted (height for age z-score <-2; 6.5/7.7%) and wasted (weight for age <-2; 9.7/22.8%) was higher in group II, the same as low (<-2) BMI z-score (8.6/13.8%).

Conclusion: Certain improvement of nutritional status mean indexes can be observed in children diagnosed by neonatal screening. We suppose that the higher portion of patients with low z-scores in after-screening group II is a result of early diagnosis but inadequate treatment in some distant Russian regions.

Conventional and novel assessment tools for the evaluation of nutritional status of children with cystic fibrosis

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Objectives: To assess the nutritional status among children with CF using conventional, as well as novel nutrition assessment tools. To compare conventional and novel nutrition assessment tools.

Methods: Nutrition status of 64 children with CF (56.25% males, mean age 10.2 years, range 0.1–18 years) was evaluated. Anthropometry was expressed as z-scores for age and sex using WHO Anthro software and classified following the WHO criteria. The proportion of patients below the “BMI p goal” (BMI p=50%) was also evaluated. Phase angles (PHA) and body cell mass (BCM) were assessed by BIA. Three-day food intake was recorded and analyzed using “Food Processor”. Overall diet quality was assessed using the Diet Quality Index International (DQI-I).

Results: Seven out of the 64 patients (10.9%) were malnourished (Weight for Age z-score <-2). Thirty-one out of 64 patients (48.4%) did not fulfill the BMIp-50th goal. Intake/Requirements Ratio (median) was 77.9% for energy and 63.5% for protein. The mean PHA values were 4.84±1.05. PHA values were significantly correlated with body mass index (BMI) (P=0.027), BMC (p=0.047), energy intake (p=0.049) and age of diagnosis (p=0.002). Statistically significant correlation was found between Smart Score and energy intake (p=0.015), protein intake (p=0.034), as well as weight for age z-score (p=0.001) and height for age z-score (p=0.003).

Conclusions: Novel nutrition assessment tools (PHA and smart score values) correlate with traditional nutrition assessment tools, indicating the nutritional status of children and adolescents with CF.

Composition of fatty acids in erythrocytes and its correlation with the nutritional status, genotype, pancreatic and pulmonary function in cystic fibrosis patients

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Objectives: To evaluate the composition of fatty acids (FA) in erythrocytes and its correlation with the nutritional status, genotype, pancreatic and pulmonary function in cystic fibrosis (CF) patients.

Methods: Observational cross-sectional study in CF children and adolescents clinically stable, attending in Brazil reference center. We analyzed the FA in erythrocytes phospholipids by gas chromatography. The energy and FA intake were obtained by three 24-hour recalls. The anthropometric indicators (BMI and height-for-age) were also assessed in CF Consensus recommends. Pancreatic insufficiency was classified by fecal elastase-1 and fecal fat excretion. Lung function impairment was based on the %FEV1 predicted. The genotyping was done by polymerase chain reaction. The Pearson correlation, ANOVA and t-test were used by SPSS 17.0 with 5% significance.

Results: We observed in erythrocytes of 27 patients (138.8±42 months) low concentrations of palmitoleic acid, oleic acid, linoleic acid (LA) and docosahexaenoic acid (DHA) and high concentrations of arachidonic acid (AA), dipalmitoyl-Linolenic acid (DDL). Correlations were found between the intake of DHA and LA: DHA ratio (r=0.738 p=0.000) and AA: DHA ratio (r=0.629 p=0.001) in erythrocytes, and EPA intake with ALA concentration (r=0.576 p=0.003). The height-for-age was related with AA:DHA ratio in cells (p=0.035). In DF508 homozygous patients AA concentrations in erythrocytes were greater compared to heterozygous (p=0.038).

Conclusion: Imbalance and deficiency of FA were observed in erythrocytes of CF pediatric patients and related to ingestion of FA, height-for-age and DF508 homozygous mutation.

Impact of n-3 fatty acids supplementation on inflammatory markers in cystic fibrosis patients

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Objectives: To investigate the effects of n-3 fatty acids supplementation on inflammatory markers in cystic fibrosis (CF) children and adolescents.

Methods: A prospective clinical trial, before-after study, held in Brazil CF reference center. Patients with clinically stable conditions, between 5 and 20 years, were supplemented with fish oil capsules (180mg of EPA – Eicosapentaenoic acid and 120mg of DHA – Docosahexaenoic acid/1g), during 3 months (20 to 30 mg/kg/day). Serum tumor necrosis factor (TNF-α) and interleukins (IL-1, IL-6 and IL-8) were analyzed by ELISA and C-reactive protein (CRP) by Nephelometry. The genotyping was done by polymerase chain reaction. Patients were divided into smaller (children group-CG) and larger than 10 years (adolescents group-AG). Comparisons were performed using Wilcoxon and McNemar test, with 5% significance, by SPSS software version 17.0.

Results: 57 patients were studied (CG=26, AG=31). In CG 19.2% were DF508 homozygous and 53.8% heterozygous and in AG 97.6% and 45.2% respectively. Reduction of TNF-α serum concentrations were observed in both groups after supplementation (p<0.05). The concentrations of seric IL-1 β had a tendency to increase and IL-6 were significantly increased in CG. Seric IL-8 concentration was not modified. CRP levels (>5mg/ml) that indicates patient inflammation were reduced in 57.8% (CG) and 15.5% (AG) after supplementation (p<0.05).

Conclusion: n-3 fatty acids supplementation reduced seric concentration of TNF-α and the percentage of patients undergoing inflammation, should be considered during treatment of CF pediatric patients, especially with DF508 mutation.