286 Dyslipidemia in the Montreal cystic fibrosis cohort

L. Belson[1,2,3], S. Ziai[1,2,3], L. Coderre[1,4], Y. Berthiaume[4,5], R. Rabasa-Lhoret[1,3,4]. [1]*Institut de Recherches Cliniques de Montreal, Montreal, Canada;* [2]*University of Montreal, Nutrition, Montreal, Canada;* [3]*Montreal Diabetes Research Center, Montreal, Canada;* [4]*University of Montreal, Medicine, Montreal, Canada;* [5]*Cystic Fibrosis Clinic, Montreal, Canada*

Introduction: Contrary to the general population, people with cystic fibrosis (CF) have, for primary nutritional recommendation, high-fat and high-calories diets. This is mainly due to the fact that despite pancreatic enzymes supplementation (<80%), intestinal malabsorption persists.
Objectives: Characterize lipid profiles, analyze the causes and potential complications of these abnormalities in CF patients and highlight a possible association with abnormal glucose tolerance.
Subjects: 204 CF subjects and 15 control subjects were included in this study. All subjects underwent an Oral Glucose Tolerance Test (OGTT) to determine their glucose tolerance. Lipid profiles including total cholesterol, HDL, LDL and triglycerides were measured at fasting state. An electronic scale was used to determine body fat, lean mass and body mass index (BMI).
Results: Hypertriglyceridemia (HPTG) was found in 20 CF patients subsequently matched with 20 non-HPTG CF patients. The matching criteria were gender, age, pancreatic insufficiency, genotype and status of glucose tolerance. Comparing the three groups (CF-HPTG, CF non-HPTG and non-CF) showed differences on the secretion and insulin resistance. For a comparable glycemic status, CF-HPTG patients had lower insulin sensitivity resulting in a higher secretion of insulin. BMI, blood pressure, inflammation and lung capacity does not appear to differ between groups. Women tend to be more frequently affected than men to HPTG.
Conclusion: HPTG appears to be an isolated phenomenon and do not seem to have any complications encountered in the general population.

287 Plasma phospholipid arachidonic acid is inversely correlated to the linoleic acid concentration also in patients with cystic fibrosis

B. Strandvik[1,2], B. Holmberg[2]. [1]*Karolinska Institutet, Dept of Biosciences and Nutrition, Stockholm, Sweden;* [2]*University of Gothenburg, Dept of Pediatrics, Lipid Research Laboratory, Gothenburg, Sweden*

Most patients with a severe clinical phenotype have low serum concentrations of linoleic acid (LA) and its metabolite, arachidonic acid (AA) have been reported high–low in different investigations. Warning has been raised that increase of LA in patients with cystic fibrosis (CF) would increase the AA concentration and thereby increase the burden of inflammation. A general increase in eicosanoid metabolism may support such hypothesis. However, in many studies an increase of LA has been shown to down regulate the transformation to AA, giving a negative correlation between LA and AA.
Objective: to investigate if similar or opposite relation was found between LA and AA in CF compared to healthy individuals.
Methods: Plasma phospholipid fatty acid concentration was determined with capillary gas-chromatography in 168 patients with CF aged one to 54 years and compared to 97 healthy children.
There were inverse correlations between LA and AA in both healthy individuals and in patients with CF, being $r = -0.490$, $p < 0.0001$ and $r = -0.278$, $p = 0.0003$, respectively. The difference was mainly due to a wider variation in CF, with generally lower AA (mean (SD) 8.18 (1.46) %, range 5.26–13.04) than in healthy controls (mean (SD) 8.68 (1.24) %, range 5.93–11.57) ($p = 0.002$).
Conclusion: There is no indication that high LA would increase AA in patients with CF. However, intake of meat or other products with high AA, might increase AA in CF patients as well as in healthy individuals.

288 Nutritional education improves energy intake and weight gain in children with cystic fibrosis

E. Hatziagorou[1], M. Chourdakis[1], E. Chrisochoou[1], V. Avramidou[1], J. Tsanakas[1]. [1]*Hippokration Hospital, Aristotle University of Thessaloniki, 3rd Paediatric Dept, Thessaloniki, Greece*

Aim: To compare behavioral and nutritional intervention (BEH) with a usual care control condition (CTL) for cystic fibrosis (CF) children with pancreatic insufficiency. This trial was designed to: (a) compare BEH with CTL over 8 weeks, (b) evaluate the maintenance of BEH at 3- and 6-month follow-up.
Methods: Twenty-four children with CF (aged 8 months to 18 years) that were treated according to the CF Consensus Guidelines for nutritional care (CTL), were reviewed by a dietician and were assigned to BEH. BEH included nutrition counseling to increase energy intake and child behavioral management training. Children were reviewed 2, 3 and 6 months after initial assessment.
Results: BEH led to greater increases in energy intake post-treatment than CTL as measured by calories per day (842 kcal/day vs _131 kcal/day change). At 3-month follow-up, energy intake increased significantly (765 kcal/day vs 475 kcal/day). Weight gain and BMI z score increased significantly (mean, 1.39 vs. 0.85 kg and 0.39 vs. 0.14, respectively.
Conclusions: These preliminary results show that a behavioral – nutritional education intervention was more effective than a usual control education among children with CF.

289 Do children with cystic fibrosis really need more energy?

K. Jones[1], R. Iles[1], D. McShane[1], R. Ross-Russell[1]. [1]*Addenbrookes Hospital, Cambridge, United Kingdom*

Background: Patients with cystic fibrosis (CF) are traditionally thought to need 120–150% of the energy intake of their equivocal counterparts without CF. In more recent years clinicians have begun to question whether an increased need for energy really exists particularly when a child with CF is well and asymptomatic.
Objectives:
1. To establish whether children with CF who are asymptomatic consume a higher energy intake than children without CF by comparing their actual energy intakes with the Estimated Average Energy Requirement (EAR).
2. To assess PERT adequacy.
3. To assess nutritional status in terms of growth and adequacy of weight gain.

Patients and Methods: 32 children with pancreatic-insufficient (PI) CF: 21 boys (age range 3–16.6 yrs) and 11 girls (age range 1–16.9 yrs) completed a 3 day food diary which was computer analysed. Weight, height and BMI were plotted and % weight for height calculated. PERT dosage was assessed by lipase dosage and steathorrea.
Results: The average lipase dose was 10,900 iu/kg/day for the boys and 9,400 iu/kg/day for girls: none of the children reported symptoms associated with steathorrea. All except one child (5%) had an acceptable nutritional status. All children were growing along appropriate centiles. The average intake for energy for each age group did not appear to be significantly different to the EAR with the exception of girls aged 11–14 years whose average energy intake was 20% greater than the EAR
Conclusions: It would seem that when well, asymptomatic children with CF with a good nutritional status, appropriately dosed PERT and regular input from an experienced dietitian need no more energy than their age and sex matched counterparts without CF.