238 Relation between fatty acid (FA) composition and clinical status/genotype in CF?

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Aim: evaluate the relation of clinical parameters & genotype with the serum phospholipids FA composition in CF patients.
Methods: CF patients, with stable pulmonary disease, followed for min 6 m were taken a blood sample for phospholipids FA composition & vitamin E concentration. The genotype, age, pancreatic function, nutritional status, caloric intake, pulmonary function & presence of pseudomonas colonisation, liver disease or diabetes mellitus were recorded. Patients were divided in 2 groups according to genotype (group A: mutation class I, II, or III, group B: mutation class IV, V).
Results: CF patients (group A and B together) have significant lower docosahexaenoic acid (DHA) ($P < 0.007$) and linoleic acid (LA) ($P < 0.0001$) and higher dihomogammalinolenic acid (DHGLA) ($P < 0.0001$), oleic acid (OA) ($P < 0.0001$) and mead acid (MA) ($P < 0.0001$), resulting in an increased ratio of arachidonic acid (AA)/DHA ($P < 0.004$), MA/AA ($p < 0.0001$) and OA/LA ($P < 0.0001$).
Group A had compared to group B a lower LA ($p < 0.002$) and a higher DHGLA ($p < 0.002$), 22:4 ω6 ($p < 0.03$), 22:5 ω6 ($p < 0.03$) and 20:3 ω9 ($p < 0.04$). There was however no significant difference between the groups for pulmonary function, nutritional status, age, vitamin E concentration. There was no relation of FA with gender, pulmonary function, pseudomonas colonisation, diabetes mellitus, pancreatic function, nutritional status or caloric intake. Patients with CF liver disease had an even lower DHA than the patients of the same genotype.
Conclusion: Fatty acid disturbances are related to genotype and the presence of CF related liver disease. Future studies should take these parameters into account.

239 Pilot study for evaluation of essential fatty acids deficiency in Slovak CF patients

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Background: Essential fatty acids deficiency (EFAD) has been reported for more than 40 years. Its most frequent symptoms are: skin lesions, increased susceptibility to infections, impaired growth and hepatic steatosis. Although etiology of EFAD is still unknown, it is clear that correction of this defect via: nutrition, oral supplementation or intravenous administration can improve health status of CF patients.
Aim: Assessment of serum levels of linoleic acid (LA), α-linolenic acid (αLnA) and arachidonic acid (AA) in Slovak CF patients and correlation of the estimated levels with pancreatic insufficiency (PI), BMI and FEV1.
Methods: 79 patients with CF and 40 healthy persons as controls were included into the study. Fatty acids concentrations were estimated in serum samples by capillary gas chromatography and quantified in μg/ml.
Results: Patients with CF had significantly lower concentrations of LA (16.4±2.9) than healthy controls (21.8±4.7). Comparison of LA levels between the PI subgroup (16.5±3.0) and the non-PI patients (15.7±1.8) showed that decreased concentration of LA is not related to pancreatic functions. Levels of αLnA in the serum samples of the CF patients were small (1.0±1.1) but not significantly different from the controls (1.8±0.4). Concentration of AA in the CF patients (3.7±1.4) was slightly higher in comparison with the controls (2.1±0.6) what is in consonance with previously published data.
Conclusions: The study can support the hypothesis that EFAD is a result of an increased demand for linoleic acid due to increased production of its metabolite – arachidonic acid.

240 Influence of pancreatic status on plasma and erythrocyte membrane fatty acid composition in CF

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Abnormal fatty acid (FA) composition in plasma (P) and erythrocyte membrane (RBC) has been increasingly reported in CF. Discrepancies have been described and could be related, at least partly, to the pancreatic status (Coste 2007). P and RBC FA compositions were analysed by GC-MS method using a 30 m × 0.25 μm ID column after transesterification with acetyl chloride in 68 CF patients (median age = 14.5 y) and 29 age-matched controls. CF patients were categorized according to the pancreatic status as sufficient (PS) or insufficient (PI).
PI patients displayed significantly lower linoleic acid (LA) and docosahexaenoic acid (DHA). A decreasing trend ($p = 0.08$) was noted for DHA in PS patients. The product LAxDHA appeared reduced in both CF groups and reached statistical significance ($p < 0.0001$) in PI patients as compared to controls. These results indicate that pancreatic status plays a major role in circulating and cellular fatty acid composition in CF.

Plasma and erythrocyte LA and DHA levels in CF patients according to the pancreatic status

FA (g/100 g)	Controls (n = 29)	CF PS (n = 12)	CF PI (n = 56)
LA P	25.6±5.2	27.0±2.7	20.3±5.1[a+b]
LA RBC	8.3±1.0	8.5±0.7	7.1±1.3[a+b]
DHA P	1.9±0.7	1.6±0.5	1.4±0.6[a]
DHA RBC	4.5±1.0	4.0±1.1	3.5±1.1[a]

Data are expressed as means ± SD. Statistical differences at p-value level <0.0003 are indicated for comparison between PI and [a]control or [b]PS group.

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241 Comparison of inpatient blood glucose monitoring with UK Cystic Fibrosis Trust Guidelines

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Glucose tolerance varies in people with Cystic Fibrosis (CF) over time. Hyperglycaemia may develop in individuals without CF Related Diabetes (CFRD) on steroids or during exacerbations. The UKCF Trust recommend measuring blood glucose on admission, pre and post meals, pre, 2 hours into and post enteral tube feeding, and considering insulin if levels are persistently raised (>7 mmols/L) at any time.
Aim: To compare blood glucose monitoring with UKCF Trust Guidelines.
Methods: From November-December 2006, all CF patients admitted with an exacerbation underwent blood glucose monitoring (n = 10) aged 18–33 years, mean BMI 19.7 (15.7–21.5). Based on the most recent oral glucose tolerance test, 30% had normal glucose tolerance (NGT), 30% had impaired glucose tolerance (IGT) and 40% had CFRD (75% on insulin). Monitoring was compared with UKCF Trust Guidelines.
Results: 20% of patients had blood glucose measured on admission. In the NGT group, the mean frequency of pre and post meal monitoring was 4% compared with 32% and 5% in the IGT group, and 57% and 26% in the CFRD group. The mean BMI of the IGT group was 17.9 and all were on enteral feeding. The mean frequency of pre, 2 hours into and post feed monitoring in this group was 60%, 70% and 30% respectively. Of which, a mean 34% of pre feed, 96% of 2 hours into and 53% of post feed values were >7 mmols/L. However, insulin was not commenced. Within the CFRD group, mean pre and post meal values >7 mmols/L were 85% and 83% respectively.
Conclusion: In our group of undernourished patients, blood glucose monitoring was inconsistent and sub-optimal compared with UKCF Trust Guidelines. Management of glycaemic status during an exacerbation was therefore difficult to assess. Results suggest that some patients should have been considered for insulin treatment.