Deficiencies of essential fatty acids and vitamin E in cystic fibrosis

H. Böhles, H. Heid, K. Stehr, and W. Fekl

With 5 figures

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Introduction

Problems of lipid assimilation are well known in cystic fibrosis of the pancreas (7, 10, 19, 21). Deficient activity of pancreatic lipase has been fully described (19). In consequence, dietary substitution with pancreatic enzymes has been established as standard therapeutic approach in cystic fibrosis (7, 10). The effect of a disturbed assimilation on serum and tissue levels of lipids continue to be the subject of further studies (3, 8). In particular, polyunsaturated fatty acids and vitamin E have become a center of interest during the last years (1, 2, 3, 5, 8, 18, 20, 22, 23, 24, 25).

Materials and methods

Study population

The study was done on 25 patients (13 female; 12 male) with cystic fibrosis. The diagnosis was confirmed by sodium sweat analysis. The patients are under regular control of the outpatient department of the University Children's Hospital of Erlangen. The age of the patients ranged from 6 months up to 16 years.

24 control children were matched for age and sex. These were patients of the hospital with mild upper respiratory tract infections or orthopedic problems. Their history showed no intestinal problems. The parents of all our patients were informed about the purpose of our study and gave their consent.

Growth velocity

The growth velocity for height of the cystic fibrosis patients was calculated for the year before the beginning of the study. Data were taken from the charts. Data were available for 12 patients with cystic fibrosis and 11 control children.

Analysis

About 3 cc of blood were taken from patients and controls in the fasting state during the morning hours. Patients were selected randomly as they presented at the outpatient department for their routine check up. Total serum vitamin E was determined within half an hour after the withdrawal of blood. The remaining serum was kept frozen under nitrogen for the later fatty acid analysis. The fatty acid analysis was done within a month after the sampling of blood.
Vitamin E

Total serum vitamin E was determined using the method of Quaife et al. (17).

Fatty acids

Lipids were extracted from serum according to Folch et al. (4). The total lipid was chromatographed on silica gel G in hexane/diethyl ether/acetic acid (80:20:1; v:v:v). Fatty acid methyl esters of the separated serum triglyceride, of the cholesterol ester and of the phospholipid fractions were prepared (5% methanolic H₂SO₄; 3 hr; 70 °C) and separated by gas liquid chromatography (Hewlett-Packard 5830 A equipped with hydrogen flame ionization) using glass columns packed with 10% DEGS on 100/120 mesh chromosorb W-HP. Analysis started at 160 °C followed by temperature programming at 2°/min to 200 °C, from then, isothermally. Commercially obtained standards were used for reference.

Results

Serum tocopherol

Serum tocopherol was decreased considerably in the patients with cystic fibrosis (0.30 ± 0.26 mg/dl) compared to the control group (1.02 ± 0.24 mg/dl) (p = 0.01) Fig. 1). The results in the cystic fibrosis group show a wide scatter, ranging from 1.07 mg/dl to immeasurable values (≤ 0.00 mg/dl).

Fatty acids

The fatty acid distribution in the cystic fibrosis group showed no significant difference in any of the studied ester fractions, compared to the control group (fig. 2, 3, 4). However, there is a wide scatter in all three ester fractions and trends towards differences are demonstrable. The concentration differences observed can be seen best in the cholesterol ester fraction.

![Fig. 1. Total serum tocopherol levels in patients with cystic fibrosis and in controls (mean ± 1 SD).](image-url)