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Original Article

Dietary intake and lipid profile in children and adolescents with cystic fibrosis

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Abstract

Background: Cystic fibrosis (CF) patients are advised to derive 35% of their daily energy intake from dietary fat. Whether this high fat intake is associated with dyslipidaemia is unknown. We described the lipid profile and dietary intake in paediatric patients with CF.
Methods: 110 fasting lipid concentrations of 110 Dutch patients with CF were studied, along with 86 measurements of dietary intake. For the total group and for boys and girls separately, the lipid profile and the dietary intake were investigated. The cross-sectional relationship between the lipid concentrations and dietary intake was determined.
Results: The mean dietary fat intake was ≥35% of the total energy intake, along with a considerable consumption of saturated fat. We found lower concentrations of cholesterol, high-density lipoprotein cholesterol and low-density lipoprotein cholesterol, and increased concentrations of triglyceride and triglyceride to high-density lipoprotein cholesterol ratios. Lipid concentrations were not associated with dietary fat intake.
Conclusion: This study lacks variation in dietary fat intake to exclude an effect on lipid concentrations as the distribution of dietary fat intake remained constant at a high level. Elevated triglyceride concentrations and triglyceride to high-density lipoprotein cholesterol ratios suggest an increased risk of cardiovascular disease. Any negative consequences of a high dietary fat intake on the overall lipid profile later in life cannot be excluded.

Keywords: Cystic fibrosis; Dietary intake; Lipid profile; Fat intake; Cholesterol; Triglyceride

1. Introduction

Cystic fibrosis (CF) is a life-threatening genetic disorder, characterized by chronic pulmonary inflammation. Most patients with CF also have pancreatic insufficiency and consequently intestinal fat malabsorption [1]. This might compromise the nutritional status (NS), which is, along with the pulmonary function, a strong predictor of morbidity and mortality [2], and as a result, monitoring the NS has become a key objective in CF-care. For this purpose, international CF-guidelines advocated a high-caloric intake [3,4] in which at least 35% of the energy should be derived from dietary fat [5]. An adverse effect of the latter is a disproportional saturated fat consumption [6,7] which was found well above the advised limit of 10% of the daily energy intake [8]. In non-CF paediatric patients, excessive saturated fat consumption causes lipid abnormalities [9,10] and, consequently, a deviated triglyceride (TG) to high-density lipoprotein cholesterol (HDL-C)

Abbreviations: HDL-C, high-density lipoprotein cholesterol; LDL-C, low-density lipoprotein cholesterol; TG, triglyceride; TG/HDL-ratio, triglyceride to high-density lipoprotein cholesterol ratio; NS, nutritional status; CFA, coefficient of fat absorption; FPG, fasting plasma glucose; OGTT, oral glucose tolerance test; 2-h-PG, 2-h-post-glucose; CFRD, CF related diabetes mellitus; FEV_1, pulmonary function assessed by forced expiratory volume per 1 s; CFLD, CF related liver disease.

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ratio. Both are associated with an increased risk of cardiovascular disease [11,12]. In patients with CF, it is unknown whether there is a relationship between the lipid profile and dietary fat intake. This knowledge has become highly relevant as the outlook for patients with CF has dramatically improved over the last few decades [13]. We therefore set out to characterize the overall lipid profile and the association between dietary total fat intake, dietary saturated fat intake and serum lipid concentrations. We also studied a possible association between serum lipid concentrations and coefficient of fat absorption, nutritional status, glucose tolerance, liver disease, and genotype in a large study sample of paediatric patients with CF.

2. Methods

A dynamic cohort of Dutch children (born between 1995 and 2004) with proven CF and proven pancreatic insufficiency, were studied retrospectively from March 2013 to December 2015. All children received medical care at the CF-centre of the University Medical Centre Utrecht. Each child was confirmed as having CF by a positive sweat test and/or the presence of two CF-mutations, as well as clinical signs of CF and/or a positive family history. Pancreatic insufficiency was confirmed when having a documented history of fat malabsorption with a coefficient of fat absorption (CFA) outcome <85%, and/or a faecal elastase concentration < 15 μg/g stool. After an overnight fast serum lipid profile, glucose tolerance, weight and height were measured during routine annual review. Dietary data were collected through 3-day dietary food records in clinically stable patients and simultaneously, a stool collection was done to measure faecal fat excretion. These results, along with demographic information were stored in a longitudinal database. This study included children, aged 10 to 18 years who had at least an overnight fast measurement of lipid concentrations along with a weight and height measurement. All patients, or parents/guardians of the young patients, provided written informed consent for the storage and analysis of their data. The study was performed in accordance with the guidelines of the medical ethics board of the University Medical Centre Utrecht.

2.1. Serum lipid profile

In children starting at the age of ten years, blood samples were taken yearly after an overnight fast for estimation of total cholesterol, HDL-C and TG by using enzymatic methods. LDL-C was calculated with the Friedewald equation [14]. Total cholesterol, HDL-C, LDL-C and TG concentrations were expressed as mmol/L, and as a percentage of the reference values for age-equivalent healthy controls [15]. The triglyceride to high-density lipoprotein cholesterol (TG/HDL-C) ratio was calculated by subdividing TG by HDL-C.

2.2. Nutritional assessment

Weight was measured in patients in their underwear, to the nearest 0.1 kg using a digital weight balance. Height was measured with a stadiometer (Holtain, Crymich, UK) to the nearest 0.5 cm. Age- and gender-specific z-scores for weight and height were computed by using the reference values for their nationality (Growth analyser 4 RCT, 2010, Dutch Growth Foundation). Further, height measurements were adjusted for genetic potential by calculating z-scores height-for-age-adjusted-for-target height [16].

Yearly, patients received written instructions on completing a 3-day dietary food record including a request to maintain the child's usual dietary intake. All food and beverages consumed were recorded in portion size or weight during two weekdays and one weekend-day whenever possible. A registered dietician (JWW) calculated the food records at the time of the individual completing the record. All food records were coded and analysed according to a standardized approach, using the Dutch Food Composition Table (2010) of the Dutch Nutrition Centre. The energy intake was expressed as kcal, and the total fat, saturated fat, protein and carbohydrate intake were expressed both as an absolute intake and as a percentage of energy intake (En%).

2.3. Clinical measurements

Starting at the age of ten years, patients were annually screened for glucose intolerance with an oral glucose tolerance test (OGTT) (1.75 g/kg glucose, maximum dosage 75 g glucose). Fasting plasma glucose (FPG) level and the 2-h-post-glucose load (2-h-PG) level were monitored. A FPG < 7.0 and a 2-h-PG level < 7.8 were considered as normal, and a FPG level <7.0 mmol/L and a 2-h-PG level between 7.8–11.1 mmol/L as impaired. A FPG level < 7.0 mmol/L and a 2-h-PG level ≥ 11.1 determined the diagnosis of CF related diabetes mellitus (CFRD) without fasting hyperglycaemia. A FPG level > 7 mmol/L or a 2-h-PG level > 11.1 mmol/L determined the diagnosis of CFRD [17].

A fat balance study was performed to measure the fat excretion in faeces, and to calculate the CFA. In conjunction with the 3-day dietary intake assessment, a home-based 72-hour stool collection was obtained to determine the mean faecal fat content of this 3-day collection. The stool collection started on day two of the dietary intake assessment and ending one day after dietary recording, The CFA was then calculated from the mean dietary fat intake of the 3-day dietary record and the mean daily faecal fat output and expressed as a percentage.

Pulmonary function assessed by forced expiratory volume per 1 s (FEV_1), was obtained from maximal expiratory flow curves (Masterscreen, Viasys Healthcare, Höchberg, Germany) and expressed as the percentage of the predicted value for a given height and gender (FEV_1% pred.) [18]. For each child, the highest FEV_1% pred. measured in the preceding calendar year was given.

CF related liver disease (CFLD) was diagnosed if at least two of the following variables were present: hepatomegaly confirmed by ultrasound, other abnormalities of the liver parenchyma on ultrasound, and/or persistently increased liver enzymes [19]. Additionally, patients were categorized based on their CF-transmembrane-conductance-regulator-mutation. Patients who were either homozygous or compound heterozygous for class I, II and III mutations were classified as severe, and