Plasma amino acid levels in cystic fibrosis patients

H.P. Ioannou1, M. Fotoulaki1, K. Vasilaki1, P. Augoustides-Savvopoulou2, 14th Pediatric Department, Aristotle University, Thessaloniki, Greece; 2Metabolic Laboratory of the 1st Pediatric Department, Aristotle University, Thessaloniki, Greece

Aim: To determine plasma amino acid levels in cystic fibrosis patients and to correlate these profiles with their nutritional status and liver disease.

Methods: Plasma amino acid profiles were measured by ion-exchange high performance liquid chromatography in 36 stable cystic fibrosis (CF) patients and were compared to those of 30 age-matched healthy controls. Possible relationships between amino acid levels of CF patients, their nutritional status and liver disease were also studied.

Results: In CF patients (age 20±9.9 yr), levels of histidine, arginine and cysteine were reduced, whereas levels of glycine were increased as opposed to controls (p < 0.05). Branched-chain amino acids were lower in malnourished patients, but differences reached statistical significance only for valine and isoleucine (p < 0.05). The ratio of valine to glycine correlated significantly with body mass index z-score (r = 0.58, p < 0.05). The ratio of branched-chain to aromatic amino acids (leucine + isoleucine + valine)/(phenylalanine + tyrosine), known as Fischer’s ratio, was significantly lower in patients with biliary cirrhosis (2.19±0.97) than in CF patients without liver disease (3.03±0.4, p < 0.01) and control subjects (3.2±0.6, p < 0.01).

Conclusions: The results of the present study suggest that in cystic fibrosis patients some changes occur in their plasma amino acid profiles, changes which are not consistent with a “disease specific” pattern. As expected, Fischer’s ratio, a well known amino-index in advanced liver disease, was reduced in CF liver cirrhosis. Of interest, low levels of arginine were observed, contributing perhaps to the reduced nitric oxide formation reported in the airways of CF patients.

Will my CF diet make me ill?

E. Lake, R. Barnes, A. Morton, S. Wolfe. 1Cystic Fibrosis Trust, Bromley, United Kingdom; 2Regional CF Unit, St James’ hospital, Leeds, United Kingdom

Background: Through Peer Reviews carried out by the CF Trust it became apparent that there is disparity in dietetic provision in the UK.

Aim: We wished to establish how adults with CF and parents of children with CF felt about their CF diet.

Methods: The CF Trust initiated a postal survey amongst patients and parents of children with CF in the UK.

Results: 5300 questionnaires were sent, 21% responded. A significant number had concerns over the perceived unhealthy nature of the diet they were advised to eat, and worried that it would lead to health problems in the future. They had heard repeatedly that the high fat, high sugar diet could lead to heart trouble, cancer and other health problems for the population at large. With increased longevity, this was a cause of concern to them. Some of them felt that a high sugar diet was likely to directly contribute to the development of CF related diabetes. Parents of children with CF reported difficulty in ensuring their child had an appropriate diet when at school. This was thought to be due to the emphasis put on healthy eating in society and at school in particular.

Discussion: These concerns and beliefs indicate the need for and importance of better education in relation to the nutritional needs and problems in CF. In this health and food conscious age, those with CF and those who care for them, including schools, need to understand the importance of an appropriate diet to maximise the length and quality of their life. Unless this is achieved, many children, adolescents and adults may make food choices which are detrimental to their long-term health.

Is dietetics the cinderella of the CF multidisciplinary team?

E. Lake, R. Barnes, A. Morton, S. Wolfe. 1Cystic Fibrosis Trust, Bromley, United Kingdom; 2Regional CF Unit, St James’ hospital, Leeds, United Kingdom

Background: Through Peer Reviews carried out by the CF Trust it has become apparent that there is disparity in dietetic provision in the UK.

Aim: We wished to establish the number of people with CF suffering with perceived weight and digestive problems and whether there is appropriate specialist CF dietetic support, given by dietitians with experience, commitment and time.

Methods: The CF Trust initiated a postal survey amongst adults with CF and the parents of children with CF in the UK.

Results: 5300 questionnaires were sent, 21% responded. It was clear that many responders felt strongly about this aspect of their health. 31% of parents and 41% of adults reported putting on or maintaining weight as being an area of concern. While just over three quarters of parents said their child was growing well, many said their child was small for their age or smaller than their peers. 54% reported problems with their digestion, including ongoing stomach pains, frequent stools, blockages and difficulty in adequately managing enzymes. Again, a wide range of problems were reported, and some saw having to take enzymes as a problem in itself.

Discussion: Whilst approximately half of patients felt that nutritional aspects of their CF were well managed and well controlled, half had significant problems, which were of real concern to them and created a lot of anxiety. Parents saw the matter of gaining weight as involving a lot of hard work and effort. The question as to whether these problems were inevitable and inherent in the condition or whether better specialist dietetic support would help alleviate many of these issues is an important one. The survey also assessed the variable level of specialist dietetic input, which could well be a significant contributory factor to these health problems.