351 Plasma amino acid levels in cystic fibrosis patients

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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 acids levels of CF patients, their nutritional status and liver disease were also studied.
Results: In CF patients (age 20±9 y), 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)/(phenynalanine + 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.

352 Will my CF diet make me ill?

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Background: Through Peer Reviews carried out by the CF Trust it became apparent 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.

353 Is enough attention paid to diet, weight and digestion?

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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.

354 Is dietetics the cinderella of the CF multidisciplinary team?

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Background: Do patients and parents of children with CF understand the level of dietetic support they should be getting? We wished to see if there was appropriate specialist dietetic support for those with CF attending CF Centres/shared care clinics.
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. Over 80% reported good support, with 17% having concerns. 16% percent reported not seeing a dietitian at all or only having contact with a dietitian only once in the last year, often at annual review. Some reported feeling let down, describing a derisory level of support or no support at all. Some patients felt the problem was lack of cover for sick or maternity leave or lack of adequate time. There was often no dietitian in shared care clinics and access to specialist dietitians was determined by arrangements with the specialist centre. Some patients were concerned whether their dietitian was a specialist. Some believed their dietitian knew very little about CF, consequently they often avoided contact, especially if they were being offered limited advice. 25% did not know the name of their specialist dietitian.
Discussion: This survey has highlighted a lack of clarity in how a specialist dietitian is recognised, particularly lack of a qualification recognising knowledge and experience. Responders observed that dietetics was seen as less important – they saw doctors, nurses and physios more often. Overall, the findings from this survey signify a current serious lack of appropriate dietetic support for many of those with CF.