336 Is there any difference in nutritional status between CF patients with and without liver disease

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Patients with cystic fibrosis (CF) are predisposed to develop liver disease (LD). The aim of the study was to compare the growth status of CF patients with and without chronic liver disease.

Material and Methods: The study included seventy-eight CF patients above 3 years who regularly attend our CF Center. We examined the results from the last antropometric measurements. Group one consisted of 21 CF patients with chronic hepatic disease (mean age 15.3±4.7, 13 boys) and a control group consisted of 28 CF patients without hepatic disease (mean age 10.4±6.4, 37 boys). LD was defined by the finding the hepatomegaly, significant or persistent increased serum hepatic enzyme levels, ultrasonographic abnormalities (score >4) and morphologic or functional scintigraphic abnormalities. Nutritional status was evaluated based on anthropometric assessment: height for age z-score, weight for age z-score, z-score weight for height and body mass index. Statistical comparisons between the groups were performed using Student t-test. The difference was considered significant at $p < 0.05$.

Results: Mean values and SD for CF patients with LD were ZH = −0.75±0.97, ZW = 0.57±0.65, ZW/H = 0.13±0.52, BMI = 18.8±1.85. Mean values and SD for CF patients without LD were ZH = −0.51±1.21, ZW = 0.44±1.33, ZW/H = 0.12±1.09, BMI = 17.3±2.10. There wasn't any significant difference between the groups ($p > 0.05$).

There weren't any differences in the anthropometric measurements between our CF patients with chronic liver disease and without liver disease. We can conclude that chronic hepatic disease hasn't any influence on the nutritional status in CF patients.

337 Overweight/obese CF patients, cause for concern?

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Aim: To look at all our adult CF patients with a BMI > 25 to establish if there was any clinical implications to their future management and well-being.

Method: A review of the BMI of all patients attending the all Wales Adult CF Centre assessing age, diabetic status, lipid profile, pancreatic status, FEV_1 and genotype in those with a BMI > 25 over a 3 yr period (2007–2009).

Results: 35 (18%) of 193 patients had a BMI > 25, mean age of these patients was 31 (range 18–51), 63% Male, 34% ΔF508 homozygous. 29% had an elevated Cholesterol (>5.0) and 23% had elevated Triglycerides (>2.0). 31% were pancreatic sufficient. 6/35 had CFRD and 2 had IGT. 4/35 (11%) had an $FEV_1 < 50\%$, 2/35 <30%. All 35 were given basic individual weight reducing advice; those with abnormal lipids were treated appropriately and given dietary advice. 1 patient (BMI > 34) was given a target weight loss and exercise programme prior to referral for transplant assessment. 7/35 patients had a reduction in BMI since receiving weight reducing advice.

Conclusion: Overweight patients tended to be older with better lung function than our general patient population. However, an aging CF population with an increased incidence of CFRD and dyslipidaemia with a high BMI will ultimately lead to associated health complications. A proactive approach advising weight loss/healthy eating and increased exercise has been introduced in these CF patients. However, there is a fine line between quality of life and increased treatment needs; ultimately, should we add to the burden of treatment, especially in those >75% lung function (51%) with no other significant health complications.

338 Evaluation of growth in peri-pubertal cystic fibrosis (CF) subjects with exocrine pancreatic insufficiency (EPI) treated with the novel non-porcine pancreatic enzyme replacement therapy liprotamase

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Background and Aims: A 12-month, open-label, safety and clinical activity study of liprotamase, a non-porcine pancreatic enzyme replacement therapy was conducted in patients with CF and EPI. The objective of this analysis was to evaluate trends in weight, height, and body mass index (BMI) in peri-pubertal subjects.

Methods: Clinically stable CF subjects ⩾7 years of age were enrolled, with fecal elastase <100 μg/g of stool. Subjects took one or more capsules of liprotamase (containing 32,500 units lipase, 25,000 units protease and 3,750 units amylase) in the middle of 3 meals and 2 snacks each day.

Results: A total of 214 subjects were treated with liprotamase and 55 (25.7%) of the 214 subjects were 7 to <12 years old and 57 (26.6%) were 12 to <17 years old. For the overall study population, the mean number of capsules per meal was 5.5. For subjects 7 to <12 years of age, the mean(SD) height z-scores were –0.464(0.742), –0.491(0.718) and –0.594(0.732) at Months 3, 6 and 12, respectively with mean(SD) increases in height from baseline of 1.4(0.92), 2.5(1.32), and 4.9(1.54) cm, respectively. Similar analysis for weight and BMI will be presented.

Conclusions: After an initial period of adjustment, weight showed a steady increase over the 12-month observational period in peri-pubertal subjects with CF-related EPI treated with liprotamase. Importantly, height growth, which is dependent on normal nutritional status, increased over the study period. These data support the use of liprotamase, a non-porcine pancreatic enzyme replacement therapy in growing children with CF-related EPI.

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339* Body composition and lung function in young children with cystic fibrosis

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Objective: Patients with cystic fibrosis (CF) are at risk of malnutrition and studies have found a relationship between fat-free mass (FFM) and lung function (LF). We used the 'gold standard' four-component model (4CM) to assess body composition (BC) and related that to LF.

Methods: BC was measured in 85 CF subjects aged 6–12 yrs and results compared with UK reference data and healthy matched controls. LF (FEV_1 %predicted) was measured and relationships with fat mass (FM) and FFM adjusted for height [FM and FFM indices (FMI and FFMI)] investigated.

Results: Compared to UK ref data CF boys (n = 37) were shorter [mean(sd) height sds; −0.5(1.0), $p < 0.05$] but had higher BMI sds; [0.4(1.0), $p < 0.01$]; CF girls were shorter [−0.6(1.0), $p < 0.001$] with lower BMI sds [−0.3(1.0), $p < 0.05$]. Compared to matched controls CF boys had greater FFMI sds [0.6(1.0), $p < 0.01$] whilst CF girls had lower FMI sds [−0.7(0.9), $p < 0.001$] and mineral mass index sds [−0.8(1.1), $p < 0.001$]. Prepubertal girls (n = 24) had no deficit in mineral. Six children (5 boys) had BMI sds >95th centile; this was due to increased FM in 4 boys. CF girls had lower FEV_1% than boys [mean(sd) 77.6(18.4) v 91.3(20.9), $p < 0.01$]. FM was significantly related to FEV_1% in girls only ($p < 0.01$) but there was no association between FFM and LF in either sex.

Conclusion: CF boys had normal BC, whilst girls had lower FM than controls. Six children had a BMIsds in the obese range but 2 cases were due to high FFM, not FM. We found no association between FFM and LF. The significant positive association between FM and FEV_1% in girls may reflect their poorer nutritional status and this sex difference merits more attention.