324 Small intestine bacterial overgrowth (SIBO) in CF patients

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The use of azythromycin in CF patients has been suggested as a predisposing factor for SIBO. Hydrogen breath test (BT) is commonly used as a diagnostic tool for its detection. The aim of the present study was to assess the significance of: (1) clinical status for the occurrence of SIBO and (2) the measurement of methane expiration for the diagnosis of SIBO.

Material and Methods: The study comprised 31 CF patients without previous use of azythromycin. In all subjects glucose hydrogen/methane BT was performed. The nutritional status (Z-scores for body weight), exocrine pancreatic secretion (fecal elastase-1, ELISA) and lung function (FEV1) were also determined.

Results: In 12 (38.7%) CF patients abnormal BT results were found. In addition, 4 (12.9%) subjects had borderline values. In 4 (12.9%) patients methane production was documented. Methane results created basis for the diagnosis of SIBO in 33.3% (4 out of 12) of cases. Nutritional and clinical status was not statistically different in patients with/without SIBO (Table).

Similarly, *P. aeruginosa* colonization was not different in both subgroups (66.7% vs. 60.0%).

Conclusions: For the detection of SIBO in CF patients combined measurement of hydrogen and methane should be applied. Excessive bacterial colonization of small intestine does not seem to be extensively related to clinical expression of the disease.

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Parameter	Subgroup, median (mean±SEM)	
	With SIBO (n = 12)	Without SIBO (n = 15)
Z-score for body weight	−1.22 (−1.06±0.06)	−0.87 (−0.99±0.19)
FEV1 (%)	73 (77±6)	88 (84±4)
Fecal elastase-1 (μg/g of stool)	7 (10±2)	10 (57±19)

325* The effect of tegaserod in patients with distal intestinal obstruction syndrome

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Background: Distal intestinal obstruction syndrome (DIOS) is defined as partial or complete bowel obstruction due to abnormally viscid mucofaecal material in the terminal ileum and caecum in individuals with cystic fibrosis (CF). The syndrome encompasses a spectrum from acute obstruction to chronic pain, bloating and bowel dysfunction. The long-term management of the most severe cases of DIOS remains unsatisfactory. The prokinetic cisapride was shown to be effective but has subsequently been withdrawn. Tegaserod is a 5-HT4 partial agonist which has been shown to accelerate transit in the gastrointestinal tract and might therefore be useful in this setting.

Methods: The Royal Brompton Hospital provides specialist CF care. A series of patients, including cases with both acute obstruction and chronic symptoms, was given a therapeutic trial of tegaserod. After three months of treatment, patients were reviewed, using a visual-analogue scale to assess the subjective global relief.

Results: 10 patients received tegaserod 2 mg twice daily. The age of onset of DIOS could be established in 7 with a mean of 22.0 years (range 14–33). The mean age at tegaserod treatment was 34.2 years (range 24–52). 8 cases had required admission at some point for acute obstructive episodes. Subjective global relief of symptoms at 3 months was scored at a mean of 49% (0–80%). 9 patients reported some improvement, 1 no change and none reported deterioration. Patients consistently reported improvements in pain, bloating and an increase in stool frequency.

Discussion: This uncontrolled case series presents the first evidence that tegaserod could be a useful therapy for DIOS. We feel that a randomised controlled trial of tegaserod in DIOS is justified.

326 Is intestinal inflammation present in CF patients?

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Despite of pancreatic substitution some CF patients fail to achieve a good nutritional status. In the stool of CF cases older than 4 years of age we proved increased concentration of A1AT compared to the control group, indicating increased permeability of the bowel wall. In order to learn if this feature is of inflammatory origin, we examined lactoferrin concentration in stool by ELISA (IBD-SCAN; TechLab). Stool concentration of lactoferrin was examined in 50 CF patients at the age 0.01–26.7 years (median 8.7). Assessment of lactoferrin was supplemented by the examination of A1AT concentration and polymorphonuclear (PMN) Elastase activity (using spectrophotometry with substrate Glt.Ala.Ala.Pro.Val.pNA). In all cases concentration of Elastase-1 (ELISA Elastase-1; Bioserv Diagnostic, Germany), clinical status, nutrition, lung function, CRP and therapy were assessed. Median of lactoferrin concentration in the stool of CF patients was 5.7 (2.8–130μg/ml). In 22/50 patients it was above the population norm (>7.24 μg/ml), comparable to that of patients with ulcerative colitis in remission. As expected the concentration of lactoferrin significantly correlates with PMN Elastase activity ($r = 0.44$; $p < 0.01$). Increased lactoferrin is more frequently associated with lower concentration of Elastase-1 ($p = 0.05$) and the presence of Class I–III CFTR gene genotypes. However, no correlation was found with CRP, A1AT, nutritional status, pancreatic substitution, abdominal pain, lung function, bacterial infection and/or therapy. In nearly half of examined cases there was lactoferrin-based evidence of intestinal inflammation, but surprisingly no correlation with nutritional status was observed. This issue needs further study.

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327 Fatty acid status in patients with CF-related liver disease

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In cystic fibrosis (CF), the genotype is known to influence the fatty acid (FA) composition. Diabetes mellitus and liver disease can influence FA status. Until now, there are no data on the influence of CF related diabetes mellitus and CF related liver disease (CFRLD) on the FA status of CF patients. The aim of this study was to evaluate whether the presence of CFRLD influences the FA status.

Method: A fasting blood sample for determination of serum vitamin E and phospholipid FA composition was withdrawn in 79 CF patients, with stable pulmonary disease, under regular control in our CF centre. Patients with CFRLD (n = 13) were compared to CF patients with the same severe genotype (n = 66).

Results: The CF patients with CFRLD, have a more pronounced decrease in DHA (22:6 n-3) and an increase of docosatetraenoic acid (22:4 n-6) compared to the CF patients of the same genotype. There were no significant differences in the precursors of these FAs.

Conclusion: DHA concentration in patients with CFRLD is more profoundly decreased than in their genotype controls. The presence of CFRLD should be taken into account in future FA studies in CF patients.

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