

188 Elongation of the small intestine (without histological changes) in young pigs with experimentally induced exocrine pancreatic insufficiency (EPI) – used as a model for children with EPI

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Objectives: Increase of tissue mass of the gastro intestinal tract (GIT) and length of the small intestine (smi) was found in pigs with experimentally induced EPI before. This study aimed to look more detailed at the assumed mechanisms behind these findings.

Methods: In 8 pigs aged 8 weeks the pancreatic duct was ligated (PL), another 4 pigs were sham operated and served as controls (C). 3 weeks post OP the PL-pigs were split into 2 groups (each n=4): PL+E: supplied with Creon® (~6300 Ph.Eur.E lipase/g fat) and PL+0: no enzyme substitution. Animals were pair fed until week 8 post OP, then fed ad libitum. In week 11 post OP pigs were euthanized.

Results: EPI resulted in a significantly reduced prececal (prc.) digestibility of starch (C: 88.4±1.59^a, PL+0: 33.4±15.7^c), while enzyme substitution caused a distinct increase (PL+E: 79.7±3.25^b). When excluding the data of one PL+0 pig (distinctly smallest one) the smi length (m) was significantly higher (p < 0.026) in PL+0 (21.1±1.04^a) compared to those of C (17.2±1.76^b) and PL+E (18.2±1.33^b); [PL+0 n=3; PL+E n=4, C n=4]. Correlation between body length and smi length was bad (R²=0.001) as well as the correlation between smi length and empty bw [bw without GIT; R²=0.214]. Negative correlation between smi length/kg empty bw and prc. digestibility of starch (R²=0.88) and between the mass of cecum per kg empty bw and prc. starch digestibility (R²=0.933) were quite good.

Conclusion: The elongation of the smi in case of EPI has not been described before to our knowledge and seems to be a kind of adaptation. Whether reduced enzymatic digestion or increased bacterial fermentation are the predominant factors is not clear up to date.

189 The variability of carbohydrate antigen 19-9 (CA 19-9) levels in cystic fibrosis patients

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Objectives: CA19-9 is a marker of gastrointestinal malignancies and known to be increased in CF-patients. However, only few data exist on the inter- and intraindividual variability of CA19-9 levels in relation to the clinical status. We therefore investigated plasma CA19-9 levels in 66 CF-patients at various time points.

Methods: Blood was taken in the scope of routine clinical investigations. Serum CA19-9 was determined by ElectroChemiLuminescence Immuno Assay (ECLIA) using a Roche Immunoassayautomat Cobas 6000. Clinical data were collected according to standard protocols, as used in German CF centres.

Results: 37/66 (56.1%) CF-patients had normal CA19-9 levels below 32 IU/ml and 14/66 (21.2%) showed levels above 100 IU/ml (overall mean 57.8 IU/ml; 95% CI 38.1–77.5 IU/ml; median 29.8). There was no relation between CA19-9 and gender, age or PSA status. The intraindividual variability at various time points was remarkably low. One patient with initially high CA19-9 (799.8 IU/ml) showed a sharp increase (1,810 IU/ml) within one month, without any clinical or laboratory sign of pancreatitis, cholestasis or other gastrointestinal pathology. MRI and endosonography revealed multiple cystic formations with dilated branches in the tail of the pancreas, as is seen in intraductal papillary mucinous neoplasms (IPMN).

Conclusion: Despite overall increased levels of CA19-9 in CF-patients only few reveal values above 100 IU/ml. It seems reasonable to follow up these patients more closely, with monitoring of CA19-9 and extended radiological diagnostics were required.

190 Pancreatitis in patients with CF

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Patients with cystic fibrosis may develop either single or recurrent bouts of acute pancreatitis. This complication has been mostly reported in pancreatic-sufficient teenagers and adults with mild pulmonary disease.

Objective: We describe the progress of a previously reported group of patients with CF and pancreatitis.

Methods: Multi-centre retrospective study in 5 CF-Centres of all patients with a diagnosis of CF and pancreatitis.

Results: 36 out of 579 (4.5%) patients with CF had acute pancreatitis at a median age of 14.2 y. 3 had undiagnosed CF when they presented with pancreatitis. 16 (61.5%) had a single episode and 10 (38.5%) two or more. 6 had triggering factors. All presented with abdominal pain and most with nausea, vomiting and malaise. Both lipase and amylase serum concentrations were increased. Abdominal ultrasounds showed various changes. Pulmonary disease was mild in 69.2% (median FEV1 84% Pred.). 46% had pancreatic insufficiency (PI), 34.6% CF-related liver disease, 19.2% nasal polyps and 38.5% had complications related salt losses by the sweat. Most (87.5%) had a satisfactory nutritional condition. 77% carried the F508del mutation (homozygotes or compound-heterozygotes). No major complications were reported.

Conclusions: The frequency of pancreatitis was higher (4.5%) than the 2.9% found in another study and that generally quoted. While the percentage of those with PI (46%) was lower than we found in an earlier report (57%) it still is higher than that generally reported. The condition was more frequent in adolescents with mild pulmonary disease. No any association between clinical, demographic and CF genotype variables and an increased risk of pancreatitis was found.

191 Prospective multinational DIOS survey: concomitant risk factors and treatment alternatives

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Distal intestinal obstruction syndrome (DIOS), a specific complication of CF has an accurate definition based on ESPGHAN CF Working Group (JPGN 2010). This is the first prospective survey on concomitant risk factors and treatment alternatives.

Methods: New cases of DIOS from 2009–2012 were reported through an ECFS website database and analysed by the coordinating centre located in Paris (data under process). Ethical approval was obtained in each country.

Results: 102 cases from 10 countries (28 CF centres) were reported; 60% were males, age was 14.4[6.5–23.5] years and 45% had a complete DIOS. 58% had either a previous DIOS or a recurrent episode during the study period. Frequency of meconium ileus, pancreatic insufficiency, CF liver disease, proton pump inhibitor prescription was respectively 40, 92, 22, 54%; diabetes and chronic *Pseudomonas aeruginosa* in adults, 49 and 68%. Considering the 112 DIOS episodes, outdoor temperature was >20° in 54%, beverage intake was insufficient in 50%, poor compliance with pancreatic enzymes was uncommon (12%), patients were on a high fiber diet or fat diet in only 2 and 9%. All complete DIOS required IV hydration, non opioid (85%), or opioid (17%) pain relief, Gastrografin enema (G) or polyethylene glycol (PEG) lavage in 87%, 4% a colonoscopy and 9% surgery; around half of the incomplete DIOS received IV hydration, non opioid pain relief and G enema or PEG. Overall 84% of the patients were prescribed maintenance therapy.

Conclusion: DIOS is multifactorial but climate and poor compliance do not appear to be major contributing factors. Despite variation in medical management, surgery was required in 2 centres only (9% of complete DIOS).