**319** Immuno-Reactive Trypsine and external pancreatic status in cystic fibrosis children

E. Chaillou1, A.C. Wagner1, M. Beaumesnil1, C. Bonnemains1, E. Darviet1, J.L. Giniès1,1 Centre de Ressources et de Compétences pour la Mucoviscidose, Centre Hospitalier et Universitaire, Angers, France

**Background:** The aim of our study was to evaluate the significance of the Immuno-Reactive Trypsine (IRT) as a marker for exocrine pancreatic insufficiency, assessed on fecal elastase, in pancreatic sufficient and insufficient cystic fibrosis (CF) children.

**Patients and Methods:** From August 2006 to January 2008, fifty-six children followed-up in CF Centre, Department of Pediatrics, Angers (France), were studied. From their fecal elastase concentration they were divided into 2 groups: pancreatic sufficient or insufficient (>200 mg/g of stool or ×100 mg/g of stool). For every patient, IRT concentration was assessed on a dried blood sample.

**Results:** The fifty pancreatic sufficient patients all had IRT concentrations upper than 9.5 ng/mL whatever their age. Thirty-four of the 41 pancreatic insufficient patients had IRT concentrations lower than 9.5 ng/mL whatever their age, and 33 of the 35 pancreatic insufficient patients over than 4 years-old had IRT concentrations lower than 9.5 ng/mL. IRT concentration lower than 9.5 ng/mL measured in a more than 4 years-old CF child permit to affirm that he has pancreatic insufficiency, with sensibility of 94%, specificity of 100%, positive predictive value of 100% and negative predictive value of 83%.

**Conclusion:** From the age of 4 in cystic fibrosis children, IRT concentration is correlated with external pancreatic status. This could be used as a marker for exocrine pancreatic insufficiency when difficulties to obtain stools in consultation.

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**320** Pancreatic status in cystic fibrosis patients depending upon phenotype-genotype correlations

L. Bober1, O. Munduk1, O. Hnateyk12, H. Makkukh1. 1Lviv Regional Specialized Children’s Hospital and Cystic Fibrosis regional centre, Lviv, Ukraine; 2Institute of Hereditary Pathology of Academy of Medical Science of Ukraine, Lviv, Ukraine

The influence of genetic and epigenetic factors on functional status of pancreas in patients with cystic fibrosis and on course of the disease in general has been analysed.

96 cases of CF were verified at Lviv regional CF centre and 89 CF patients were under observation in terms from 2 month to 12 years. Among them 13 (13.4%) died – 9 (69.2%) girls and 4 (30.8%) boys. The studied group of patients was divided on the genotype differences and disease manifestation severity and the obtained results of glucose-toleration test, sonographical investigation data and faecal elastase-1 level were compared.

It was determined that CFTR genotype influences on exocrine pancreatic insufficiency manifestation. “Major” CFTR mutation associates with severe exocrine pancreatic insufficiency phenotype. “Minor” changes of alleles dominate on “severe” and could result sufficient exocrine function of pancreas. Both negative (for the F508del mutation) and positive (for the 3849+10kbC → T, R334W, 3272–11A>G mutations) correlations between genotype and the degree of exocrine pancreatic insufficiency have been revealed.

CF patient genotype and the degree of exocrine pancreatic insufficiency don’t seem to determine his endocrine status and dynamics of progression of fibrotic and atrophic changes in pancreas. It is established that older age of cystic fibrosis patients, progression of fibrotic and atrophic changes in pancreas as well as severe pulmonary insufficiency, are all predictors of deterioration in endocrine function of pancreas.

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**321** Clinical outcome in cystic fibrosis patients with or without meconium ileus

K. Venkatesh1, C.J. Taylor1. 1Academic Unit of Child Health, Sheffield Children’s Hospital, Sheffield, United Kingdom

**Aims:** Meconium ileus (MI) is the presenting symptom in 20% of patients with cystic fibrosis (CF). The aims of the present study were to assess the clinical outcomes in cohorts of CF patients with MI at 5, 10 and 15 years in comparison to patients without MI.

**Methods:** Clinical records of CF patients with and without MI admitted to a tertiary referral centre were examined. In patients with MI, data on birth weight, genotype, type and extent of surgery, duration of parental nutrition were recorded. In both groups clinical status at 5, 10, and 15 years were accessed from annual review records.

**Results:** Eighteen patients (7 F) (with MI) and 16 controls (5 F) (without MI) were reviewed. Of the patients, 8 were homozygous for ∆F508, 4 were simple MI and 11 patients received parenteral nutrition for a median of 24 days (10–120 days). Compared with controls, the median weight, height, FEV1, FVC, Shwachman score, at 5 years, were 16.9 kg (13.6–23) [20 kg (14.5–27.8)*], 105 cm (102–120) [109.8 cm (100.1–121.0)*], 82% (36–121) [86% (83–94)*], 82% (36–121) [92% (85–107)*], and 93 (83–113) [96 (75–92)]; at 10 years were 30.4 kg (25–39) [32.4 kg (22.9–46.8)], 134 cm (127–140) [138 cm (126–149)], 76% (66–72) [83% (64–102)*], 76% (42–98) [93% (83–113)*] and 85 (65–87) [84 (59–94)]; and at 15 years, 52 kg (28–65) [42.6 kg (34.7–59.8)*], 161 cm (135–170) [156.7 cm (140–185)], 77% (63–104) [86% (49–119)*], 80% (77–99) [90% (63–118)*], and 71 (57–79) [71.5 (62–81)].

Abnormal findings on ultrasound scan (USS) of the liver were seen 36% (45), 70% (35)*, and 85% (60)* of patients at 5, 10 and 15 years. *p < 0.05.

**Conclusion:** The study has shown that patients without MI have better growth and lung function parameters throughout childhood. Abnormal USS of liver is significantly higher in patients with both simple and complicated MI.

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**322** Intestinal inflammation is present in young infants with cystic fibrosis

A. Lisowska1, J. Walkowiak1. 1Department of Gastroenterology & Metabolism, Poznan University of Medical Sciences, Poznan, Poland; 2Department of Dietetics, University of Life Sciences, Poznan, Poland

**Background:** It has been documented that intestinal inflammation is one of the major features of cystic fibrosis (CF). However, the age of its onset is not well known. Therefore, we have aimed to assess whether intestinal inflammation is present in young CF infants at diagnosis.

**Material and Methods:** The study comprised 10 infants detected in CF neonatal screening programme. The age of infants at the assessment was in range from 6 to 10 weeks. Intestinal inflammation was assessed by the measurement of fecal calprotectin concentrations (Immunoagnostik AG, Bensheim, Deutschland). Three children were pancreatic sufficient at the assessment (fecal elastase-1 concentrations: 200, 350 and 680 mcg/g of stool), the remaining seven were pancreatic insufficient (fecal elastase-1 concentrations from 6 to 33 mcg/g of stool).

**Results:** The range of fecal calprotectin concentrations was from 7.5 to 400 mg/L (median 81.2; mean ± SEM: 42.5 ± 37.3). In 8 (80%) infants abnormal calprotectin results were obtained. The highest value, significantly different than in the other subjects, was observed in an infant who was operated due to meconium ileus. However, the second child who experienced meconium ileus and was treated in a conservative mode presented with normal calprotectin concentration (7.5 mg/L).

**Conclusions:** Intestinal inflammation is already present in young CF infants. Supported by: Polish Ministry of Science and Higher Education (grant No. N40605431/2126.)