Pancreatitis in cystic fibrosis patients

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Introduction: Patients with Cystic Fibrosis (CF) may develop single or sometimes recurrent bouts of acute pancreatitis and this complication has been mostly reported in adolescents or adults with a pancreatic sufficient (PS) phenotype. In addition patients with idiopathic chronic pancreatitis are often found to carry mild CFTR mutations.

Goal: To describe the features of a group of CF patients who had pancreatitis and to investigate its prevalence and possible risk-factors.

Patients and Methods: The clinical records of patients with CF under care in 5 CF Units who had a diagnosis of pancreatitis were reviewed and clinical and genetic details are presented.

Results: Twenty out of 485 (3.1%) patients had pancreatitis, 66.7% males, average age 15.04 years. In 3 (20%) pancreatitis preceded the diagnosis of CF. Eight (53.3%) had a single episode, 4 (26.7%) had 2 and 3 (20%) had multiple bouts. Clinical features: Nutrition (Weight >P10 80%); Height >P10 73.3%); No & Mild lung disease 73.3%, Pancreatic insufficiency (PI) 53.3%, Nasal polyps & Sinusitis 46.6%, Liver disease 33.3%, Hypochloremic alkalosis 33.3%, Meconium ileus/DIOS 20%, Glucose intolerance 13.3%.


Conclusions: The prevalence of pancreatitis in this study was 3.1% and over half of the patients had pancreatic insufficiency underlying that this complication is not restricted to patients carrying mild mutations.

Pancreatitis versus cystic fibrosis patients

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898 (430 F) Scandinavian CF patients took part in a comprehensive study of their nutritional status, 7/8 CF centers participating. Median age was 18.5 (0.3–65.9) y. As part of the study the pancreatic enzyme replacement therapy was investigated.

Results: 771 patients were pancreatic insufficient. Enzyme preparations used: low dose (5,500 lipase) 28%, medium dose (10,000 lipase) 51% and high dose (25,000 lipase) 17%. Use of high lipase preparations increased with age and differed marked, Spain 3 centres (from): H: Hospjalg 9%/5 to Oslo 48%/9. Units of lipase differed between countries and between children and adults. Median units of lipase/kg BW per principal meal and frequency of DIOS. Preliminary data showed reported amount lipase intake to be higher than that registered in the 7-day pre-coded dietary food record.

Conclusions: Danish CF patients had a higher intake of lipase than CF patients in Norway and Sweden but used no high lipase preparations. A subgroup of patients having a large intake of lipase needs to be investigated further.

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Liver transplantation in children with cystic fibrosis: single centre experience

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Introduction: Advances in medical management have improved life expectancy in patients with cystic fibrosis (CF). With prolonged survival CF associated liver disease (CFLD) is now recognised as a major manifestation of CF with significant morbidity. Liver transplantation (LTx) is effective therapy in patients with CFLD and hepatic dysfunction prior to deterioration of lung function.

Aim: To assess the long term outcome of LTx in children with CFLD in our unit.

Methods: Retrospective review of all children with CFLD transplanted between 1989–2006. Data reviewed: demographic details, indications for LTx, diabetic status, survival, renal function [calculated glomerular filtration rate (cGFR) ml/min/1.73m2], lung function and nutritional status.

Results: 19 children (12M, 7F) had LTx. Median age at transplant was 11.5 years (2.1–16.5). Indications were progressive liver failure with deteriorating pulmonary function (n=18)and acute liver failure (n=1).14/19 are alive. Median follow up is 7 years (0.6–15.5). 2/19 had retransplant (1 primary graft failure, 1 chronic rejection after 9 years). 6 were diabetic pre LTx and 6 developed IDDM post LTx.

Summary: One and 5 year survival was 95% and 80% respectively, with an overall long term survival rate of 72%. LTx for CFLD does not improve nutritional status, but leads to improvement in lung function during first two years.

Conclusion: LTx is effective therapy for CFLD.