278 A distinct mechanism for reflux in patients with cystic fibrosis: a study using high resolution manometry-impedance

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Up to 80% of patients with cystic fibrosis (CF) have increased gastro-esophageal reflux (GER) which could be due to low basal lower esophageal sphincter (LES) pressure and a high number of transient LES relaxations (TLESRs).
Aim: To reassess mechanisms of GER and gastro-esophageal pressure gradients (GEPG) in CF using high resolution manometry-impedance (HRM-imp).
Methods: We studied 10 CF patients [age 29 (19–58), 5m/5f] and 11 healthy volunteers [age 27 (20–36), 4m/7f]. HRM-imp was performed in semi-recumbent position for 30 min during fasting and 2 hrs after a standard meal. We measured total reflux and proximal extent of reflux with impedance; LES pressure, TLESRs and GEPG with HRM.
Results: Compared to controls, CF patients had higher number of reflux episodes [19 (13–26) vs. 7 (4–17), $p = 0.03$]; mostly liquid/mixed [16 (7–24) vs. 3 (1–7), $p = 0.007$]. CF patients had more often reflux with a high proximal extent [6 (3–9) vs. 0 (0–1), $p = 0.003$]. The postprandial basal LES pressure was lower in CF [10 (7–12) vs. 16 (9–32), $p = 0.03$] and TLESR was the main mechanism for GER both in CF and controls. The number of TLESRs was similar in CF and controls. However, reflux during TLESRs was more frequent [75 (60–86) vs. 24 (6–50)%, $p = 0.003$] in CF. The GEPG during TLESRs was higher in CF [2.3 (1.8–7.4) vs. 0 (−2–2.5) mmHg, $p = 0.01$]. Such difference was due to lower esophageal pressure in CF patients [−1.5 (−4.3–1.1) vs. 5 (−1–6) mm Hg, $p = 0.02$].
Conclusion: CF patients have increased GER due to higher proportion of TLESRs associated with reflux. Unlike in GERD patients, reflux during TLESRs in CF is probably due to an increased GEPG mainly generated by a reduction in thoraco-esophageal pressure.

279* Growth and pulmonary outcomes during the first two years of life of breastfed and formula-fed infants diagnosed with cystic fibrosis through the Wisconsin Routine Newborn Screening Program

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Background: Optimal feeding (breast milk and/or formula) for infants with cystic fibrosis (CF) is unknown. Recommendations from the CF Foundation are based on limited data.
Objective: We compared growth and pulmonary outcomes between breastfed and formula-fed infants through two years of age.
Design: 103 CF infants born during 1994–2006 and diagnosed through newborn screening in Wisconsin were studied. Breastfed infants were classified by duration of exclusive breastfeeding (ExBF). Exclusive formula-feeding (ExFM) was classified by the formula's caloric density, i.e., standard (20 kcal/oz, ExFM20) throughout infancy or high density (≥22 kcal/oz, ExFM22+) for some duration of infancy.
Results: Fifty-three (51%) infants were breastfed and 50 (49%) were exclusively formula-fed. Among breastfed infants, duration of ExBF ranged from <1 mo (53%), 1–1.9 mo (21%), 2–3 mo (17%) and 4–9 mo (9%). Among ExFM infants, 23 (46%) received high caloric density formula; half ($n = 13$) by age 6 months. Proportionately more infants with pancreatic sufficiency ($n = 9$) were ExBF≥1 mo (44%) and none were ExFM22+, compared to infants with meconium ileus ($n = 24$, 13% ExBF≥1 mo, 38% ExFM22+) or pancreatic insufficiency ($n = 70$, 25% ExBF≥1 mo, 20% ExFM22+), $p = 0.02$. Among infants with pancreatic insufficiency, weight z-scores declined from birth to 6 months ($p < 0.0001$) in the ExBF≥2 mo group, and the number of *Pseudomonas aeruginosa* infections through age 2 years was fewer in breastfed compared to ExFM infants ($p = 0.003$) but did not differ by duration of ExBF.
Conclusion: For infants with CF, exclusive breastfeeding less than 2 months does not compromise growth and is associated with respiratory benefit.

280 Improved early life nutritional outcomes in 303 patients diagnosed with cystic fibrosis from the UK over the last 15 years

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Background: Growth and lung function may be prognostic indicators of disease severity in children with cystic fibrosis (CF). Evidence is accumulating that nutritional status at 2 years of age may also be predictive of pulmonary function (measured by FEV1) by mid childhood, emphasising the importance of early intervention strategies to optimise growth by 2 years. Management outcomes in these children have improved over the last 15 years. We therefore reviewed growth parameters in children diagnosed with CF in our region from 1994–2008.
Methods: The South & South West Regional CF database collects clinical information on over 1000 CF patients from 23 hospitals. Using standard auxological techniques, weight, height and body mass index (BMI) was recorded at annual review and converted to standard deviation scores (SDS) using UK reference data. Relationships between year of diagnosis and growth outcomes were evaluated.
Results: From 1994 to 2008 inclusive 303 children were diagnosed with CF in the first 2 years of life. The mean BMI SDS (SD) for patients aged 0–1 years and 1–2 years increased from −0.73 (1.37) to −0.05 (1.13). There was a significant increase in weight SDS over the study period for 0–1 years ($p < 0.001$) and 1–2 years ($p < 0.05$). BMI SDS also increased significantly at both 0–1 years ($p < 0.05$) and 1–2 years ($p < 0.05$) over the same period. No significant changes in height SDS were identified.
Conclusion: These data demonstrate significantly improved early life growth indices for the studied population over the last 15 years. This is likely to reflect increasing emphasis on nutritional aspects of CF care in the first 2 years of life.

281 Comparison of nutritional status of cystic fibrosis (CF) and non CF children in Albania

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Introduction: In a previous approach to evaluate the epidemiologic data on CF in Albania we found that 85. % of our patients was underweight. So we have the idea that probably this high % was related also to social issue.
Aim & Material: The purpose was comparison of the current growth of our CF patients with a group fulfilling the same age, gender criteria, among the non CF (was considered healthy) children. The following growth indexes were calculated to reflect nutritional status: Weight/Length percentile, Length/height for age percentile and BMI percentile.
Results: 61.4% of CF patients are <15 percentile for length/height for age with a significant differences comparing with normal children (21.4%). The same significant difference between two groups was found for the children in 50–85 percentile and >85 percentile. Concerning the weight for length/height percentile, 20% of CF patients and 16.7% of healthy children were <15 percentile, without a significant difference between groups. The same not significant difference was found and in other percentile. BMI was found <15 percentile in 27.1% of CF patients, and also for 12.9% of healthy children BMI was <15 percentile with a significant difference between, but for both group of children found on 50–85 and over 85 BMI percentile, the difference was not significant.
Conclusion: The nutritional status of CF patients in Albania is poor, but we think it is an extra negative factor which is social economic issue, that is reflected in weight for length/height deficit in normal children.