380 The effect of early and late stoma closure on nutritional outcome at 12 months of age in children with meconium ileus secondary to cystic fibrosis

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Introduction: A small but significant number of infants with cystic fibrosis (CF) require neonatal surgery to relieve bowel obstruction from meconium ileus (MI). This often results in stoma formation to encourage feeding, weight gain and recycling of stoma output. We have undertaken a retrospective review to determine the impact of time to stoma closure on nutritional status at 12 months of age.
Methods: A retrospective case note review of patients currently attending the Royal Liverpool Children's Hospital Regional CF Clinic (n, 282). We recorded duration of stoma and weight centile at three stages: stoma formation, closure, and at 12 month review. Complications of stoma and hospital stay were also noted. Stoma duration less than 8 weeks was classed as early closure (E) and more than 8 weeks as late (L). The change in weight centile at 12 month review was used as a measure of nutritional outcome.
Results: 48 children presented with MI (1990–2007); data were available for 26 included in the study. The early and late closure groups consisted of 19 (median time to closure 5 weeks) and 7 patients (13 weeks) respectively. 63% of the early closure group showed an increased weight centile at 12 months compared to 57% of the late group. Stoma duration did not affect post-closure hospital stay. Late stoma closure was associated with increased cases of post closure obstruction (E vs. L; 15.8% vs. 57.1%).
Conclusion: This review suggests that the time to stoma closure does not impact significantly on subsequent nutritional status. More studies are needed to determine the optimal post-operative management in this challenging group of children.

381 Oral protein calorie supplementation effectiveness on nutritional status of children with cystic fibrosis

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A diet therapy and the use of pancreatic enzymes can promote an adequate nutritional status in children with cystic fibrosis – CF.
This study investigated the association between the use of oral protein calorie supplementation and the nutritional status progression of patients with CF treated with pancreatic enzymes, according to the international recommendations.
A total of 149 children aged 1 month to 18 years (mean: 8.9±5.3 years) were evaluated at baseline and after 9 months of intervention. Z-scores for the weight-for-age, height-for-age, weight-for-height and Body Mass Index-for-age were used to characterize the nutritional status. Multiple regression analysis was used to correlate the supplementation and the nutritional status progression, controlled by pancreatic lipase intake, age and sex.
18.1% of the children were wasting at baseline, and 3.2% after 9 months of study. 22.7% of children were underweight before and 15.0% after 9 months of research. 22.6% of the children were stunting before and 15.0% after 9 months. In the multivariate model, the protein supplementation was associated with the weight-for-age improvement ($b = 0.241$; $p = 0.021$) and the calorie supplementation conjugated to the daily dietetic ingestion was associated with weight gain ($b = 0.583$; $p < 0.001$) We observed that the supplementations improved the nutritional status of the group studied. As a long term parameter the stature achieving was associated with the pancreatic lipase intake.

382* Impact of aggressive nutritional intervention on early and late acquisition of CFRD in an adult population?

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Introduction: Pre and post diabetic decline is a feature of cystic fibrosis-related diabetes (CFRD). We examined the nutritional and clinical impact of aggressive nutritional intervention on early and late acquisition of CFRD in a screened adult population.
Method: We report 17 patients with CFRD1 (continued growth at entry) v 31 patients CFRD2 (completed growth), each matched to control populations for age, sex, pseudomonas status.
Mean age at diagnosis (CFRD1) 21.9+3.7 yrs v (CFRD2) 28.3+5.6 yrs. Nutritional intervention was initiated in agreement with patients, according to UK consensus guidelines. Nutritional and clinical indices were recorded annually from 6 yrs pre to 2 yrs post diagnosis and analysed using repeated measures ANOVA.
Results: In the 6 yrs to diagnosis, BMI (CFRD1) remained consistently below CFRD2 [$(F1,44) = 15$, $p < 0.001$]. No differences in FEV1 [$(F6,33) = 21$, $p = 0.3$] or iv antibiotic therapy [$(F1,38) = 0.02$, $p = 0.9$] were evident, but FEV1 deviated increasingly between the younger and older group over time [$(F6,33) = 3$, $p = 0.009$]. A rapid decline in FEV1 (CFRD1) at 1 yr pre-diagnosis (46.9% to 41.5% diff = 5.4, CI 1.6 to 6.5, $p = 0.001$), stabilized on insulin therapy. FEV1 (CFRD2) remained stable as a % of control values throughout the pre-diabetic period. Enteral feeding intensified for both groups and was significantly greater in CFRD1 at diagnosis [65% v 32%, OR 4.5, CI 1.1–19.3, $p = 0.02$].
Conclusion: Aggressive nutritional intervention prevents nutritional decline in older but not younger adults with CFRD. Early acquisition of CFRD results in significant deviation in lung function and greater rate of pulmonary decline in younger adults.

383 Chocolate – that's no way to treat a hypoglycaemic attack!

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Cystic fibrosis-related diabetes (CFRD) is a common complication of Cystic fibrosis (CF). CFRD adversely affects life expectancy, pulmonary function and body weight. Insulin is the recommended treatment choice for the management of CFRD. However glycaemic control is adversely affected by steroid therapy, respiratory infection, dietary habits and patients' concordance with treatment.
Continual glucose monitoring (CGMS) was used to obtain blood glucose profiles in 27 patients who had poor glycaemic control (HbA1c >7%), suspected CFRD but normal fingerstix blood sugar measurements, recurrent hypoglycaemia attacks and those on overnight enteral feeds.
27 patients (17 female) wore the Minimed Gold CGMS for 72 hours and completed a food and activity diary. Following analysis of glucose profiles, these were the recommendations:
– 7 commenced insulin therapy
– 5 were changed to twice daily (BD) background insulin
– 8 were encouraged to increase their bolus insulin dose
– 1 changed to basal bolus insulin from BD insulin
– 3 stopped insulin therapy
– 8 received dietary advice only
As a result of CGMS 24 patients changed their dietary intake and insulin therapy. 14/27 patients had erratic blood sugar profiles explained by overcorrection of hypoglycaemic symptoms and rebound hyperglycaemia. Subsequently resources have been produced to improve patients' knowledge of hypoglycaemia, its symptoms and the most effective treatment. We now have a protocol for the treatment of hypoglycaemia available on patient information prescribing systems, in all in patient rooms and include it in induction training for nursing staff. As patients with CFRD are living longer our aim is to increase dietetic input along with a newly appointed diabetes specialist nurse to provide patients with the knowledge to empower them to achieve optimal glycaemic control.