315 Two-year outcomes of behavior and nutrition treatment for young children with Cystic Fibrosis

S.W. Powers[1], C. Piazza-Waggoner[1], K.A. Driscoll[1], J.S. Jones[2], J. Acton[3], C. Daines[3]. [1]*Division of Behavioral Medicine & Clinical Psychology, Cincinnati Children's Hospital Medical Center,* [2]*Division of Developmental & Behavioral Pediatrics, Greenville Hospital System Children's Hospital;* [3]*Cystic Fibrosis Center, Cincinnati Children's Hospital Medical Center, Cincinnati, USA*

Aims: To evaluate whether changes in energy intake and growth velocity due to behavior and nutrition treatment (BEH) for young children with CF were maintained at 2-yr follow-up (f-up).
Methods: Two-yr f-up data for 9 subjects who completed an 8-week BEH intervention, which included nutrition counseling and parental child behavior management training. For f-up assessments three 24-hour recall diet diaries and height (ht) & weight (wt) were obtained. Average daily energy intake and growth velocity (ht & wt) were outcome variables. Velocity was benchmarked against expected velocities for a same age child without CF at the 50th percentile (US 2000 Center for Disease Control growth charts).
Results: The average daily energy intake was 2,589±519 kcal/day at the 2-yr f-up. This compares favorably to data from post-treatment (2151±301 kcal/day). Pre-treatment intake was 1,381±198 kcal/day. Subjects exceeded the goal of 120% RDA/day for energy intake at 2-yr f-up.
The average wt velocity was 4.94/2-yr (Median: 4.6). Seven of 9 subjects were above benchmark. The average ht velocity was 15.8/2-yr (Median: 15.7). All subjects were above benchmark.
Conclusions: Upon returning to standard care, young children with CF who received BEH continue to maintain clinically significant increases in energy intake, and demonstrate patterns of normal growth at 2-yr post-treatment. Findings suggest this intervention is durable and families are able to continue to implement skills and knowledge acquired during treatment.
Supported by US National Institutes of Health.

316 An audit of the food provision for inpatients with Cystic Fibrosis

J. Roberts, R. Richmond, H. Musson, A.M. Jones, A.K. Webb. *Manchester Adult CF Centre, UK*

Introduction: During an inpatient stay nutrition is provided by four sources: hospital catering, ward snacks, enteral feeds/supplements and patient's own food. Complaints about inpatient food provision occur daily at the centre. To address the problem, quantitative and qualitative information about the food provision was collected.
Aims: To determine:
1. If patients are meeting their energy requirements during an inpatient stay.
2. The percentage of their energy requirements provided by each source.
3. Patient satisfaction with food provision.

Methods: Food eaten by each patient was determined over three consecutive days using two methods:
1. Three-day weighed food intakes (for food provided by catering)
2. Three-day food diaries (for all other sources of food eaten).

Patients also completed a questionnaire that assessed their satisfaction with inpatient food provision. Patients sex, age and body mass index (BMI) were noted. Energy intakes were calculated using Microdiet nutrient analysis software. Patient energy requirements were taken as 120% of the UK Estimated Average Requirement.
Results: Nine patients completed the audit (six male). Mean (range) BMI = 19.2 (15.7–23.4). 5/9 patients met their energy requirements. The mean (range) energy intake provided from each source (expressed as a percentage of energy requirements) is as follows: catering 48.8% (6–84.3); ward snacks 4.4% (0–14.4); feeds/supplements 19.1% (0–86.7); own food 22.6% (3.9–68.2). Satisfaction with food provision: 3/9 felt it was poor, 2/9 satisfactory and 4/9 good.
Conclusion: Whilst the catering service provides the majority of the energy intake it falls short of providing patient's energy requirements. The ward snacks arrangement was designed to supplement the hospital catering provision but this audit shows that it fails in this task.

317 Anemia in adult patients with cystic fibrosis (CF)

S. Hansen[1], L. Houghton[2], A. Freitag[1], R. Labiris[1]. [1]*Hamilton Health Science, McMaster University, Hamilton, ON;* [2]*Acadia University Wolfville, NS, Canada*

Aim: To determine the prevalence of anemia in adult patients with CF.
Method: Sixty pts (mean age 28 y; range 19–53 y) had ferritin, iron, TIBC, transferrin saturation (TS), hemoglobin (Hb), MCV, RBC folate, B12, C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) measurements. All pts are prescribed 2 ADEK tablets/day containing 0.2 mg folic acid (FA) and 12 µg cyanocobalamin per tablet. Results are reported as mean±SD.
Results: Mean Hb was 140±14.1 g/L with normal MCV (86.9±4.4 fL) reported in 55/60 (91.7%) pts. Serum iron (11±5.1 µmol/L) was <12 µmol/L in 36/60 (60%) yet, TIBC remained normal. Serum ferritin concentrations (35.5± 37.1 µg/L) were low with 13/60 (21.7%) pts exhibiting concentrations <23 µg/L. The inflammatory markers CRP and ESR were elevated in 78% (47/60) and 66.7% (40/60) pts, respectively. TS < 16% indicative of Fe deficiency was noted in 25/60 (41.7%) pts. Elevated B12 (740.5±426.3 pmol/L) and RBC folate (1823±682 nmol/L) were reported in 45% (27/60) and 53.3% (32/60) pts, respectively.
Conclusion: Chronic inflammation, as demonstrated by elevated CRP and ESR, is likely the cause of Fe deficiency anemia in adult CF pts. Elevated B12 and RBC folate are likely secondary to the provision of such within the ADEK multivitamin tablet. Canada's mandatory FA fortification of the food supply in 1998, has also contributed to significant blood folate improvements. Concern about exposure to excessive FA intake masking B12 deficiency was not seen in our population. Newer concerns are emerging showing that unmetabolized FA in plasma due to excessive intake is associated with decreased NK cytotoxicity. These data suggest that further increases in supplemental FA should be prescribed with caution in the era of postfortification.

318 Hypovitaminosis A in a child with Cystic Fibrosis

J. Lowdon, I.J.M. Doull. *CF Unit, The Children's Hospital for Wales, Cardiff, UK*

Introduction: Patients with CF may have vitamin A deficiency due to maldigestion and malabsorbtion, as well as poor compliance with vitamin supplementation. As well as clinical manifestations of vitamin deficiencies, low vitamin levels have also been associated with compromised clinical status1 and reduced lung function2.
Patient: An 11 year old male with CF, who has frequent pulmonary infections, resected bowel due to meconium ileus presentation, liver disease and poor compliance with vitamin and pancreatic enzyme treatment, presented with a history of night blindness. Previous vitamin A levels had been low (0.06 µmols/l). Attempts had been made to correct this by increasing the vitamin A supplementation (5000 IU).
Results: Opthamology examination revealed a right relative papillary defect with poor visual fields on the right and colour vision absent on the right. He had bilateral pallor of optic nerves and a faint hypopigmentation of fundus in the peripherary. Discussion: Immediate, initial treatment is intramuscular administration of vitamin A in a dose of 100 000 IU of retinol daily for three days. Half the dose was then given orally in an oil solution daily for 2 weeks. A marked improvement was noted.
Conclusion: Adequate supplementation, with regular monitoring and prompt action on low levels, is essential to prevent co morbidity due to vitamin A deficiency, especially in patients with additional risk factors such as a resected bowel.

References

[1] Rayner et al. Arch Dis Childh 1989. 64: 1151–1156.
[2] Carr et al. Paed Pulmn 1996. 13 (suppl): 317.