

281 Home parenteral nutrition (HPN) in malnourished cystic fibrosis (CF) patients: A safe and effective nutritional therapy

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Objectives: Evaluation of efficacy and complication's rate of HPN in severely malnourished CF patients who failed to improve their nutritional status with dietetic counselling and oral integration.

Methods: We retrospectively analyzed a case series of 7 CF patients reporting clinical and nutritional data, HPN characteristics and complication's rate. 6/7 patients were referred to lung transplantation.

Results: Patients were all females with pancreatic insufficiency, mean age 16.3 yrs (11.5–21.8), FEV1 <35% predicted (6/7), mean BMI Z-score –3.8 prior the HPN. The decision to start the HPN was taken on the basis of clinical status, the reduction of BMI Z-score despite the intensive dietetic treatment during the previous year and the high infusional needs due to reacidizations. The central venous catheters (CVC) were Port-a-Cath (2/7) and PICC (5/7). Mean HPN duration was 12.4 months (4–27), with night-time infusions, 5–6 days/week. Weaning from PN (6/7) was due to lung transplantation (4/7) and to achieved adequacy of oral intakes (2/7). HPN provided 125% of Basal Energy Expenditure with 35% (29–40) of total energy provided by lipids. BMI z-score increased 2.4 folds (–3.8 to –1.6), there were no short-term complications and any modification of daily activities on HPN. CVC related infection was 0.38/1000 infusion days (a single case in a 7 years old Port-a-Cath). Two mechanical complications didn't require hospitalization or total anaesthesia. All patients showed adequate vitamin and bone status without metabolic complications of HPN.

Conclusion: HPN may be valid and flexible nutritional therapy in malnourished CF patients shaping on the whole therapeutic process.

282 Evaluation of nutrition education in a group of children with moderate to severe cystic fibrosis (CF)

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Objective: To evaluate patients' nutritional knowledge before and after a short educational programme.

Methods: 15 pancreatic insufficient children (5–15 years, median age 11), who were enrolled on a joint physiotherapy and dietetic project, completed a 12-item multiple choice nutrition quiz. Questions were asked on diet, pancreatic enzymes and vitamins, to assess baseline knowledge and identify areas to educate. Age-appropriate activities on these topics were designed with accompanying written information. Individualised interactive teaching sessions were delivered as needed by the dietitian to the children 1–2 monthly over 12 months (ward, outpatient or home visits). The quiz was repeated at the end (n = 13). A patient satisfaction survey was completed (n = 12) at the end.

Results: Baseline scores indicated gaps in knowledge (mean 8.7). Scores at the end improved (mean 9.8). Older children (≥11 years) showed a greater improvement, however the mean score for younger children (5–10 years) remained similar pre and post education. Results of the satisfaction survey indicated that sessions helped the children understand their diet better and doing practical activities helped them learn about CF. There was a clear indication that home was not the preferred location for dietetic intervention.

Conclusions: Nutrition education was positively received by patients. Staged one-to-one sessions could be incorporated into the wider clinic on a scheduled outpatient basis. Refinement of the quiz could allow it to be used as a screening tool to assess those who would most benefit from education and specific topics. Younger children need more regular teaching sessions to retain knowledge.

283 Evaluation of a novel behavioural intervention programme (BIP) to modify mealtime behaviour for young children with cystic fibrosis

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Background: Parents of children with CF report more problematic mealtime behaviours and high levels of parental anxiety around mealtimes.

Objective: To evaluate a psychologist/dietitian led BIP aimed at modifying mealtime behaviour in children with CF.

Methods: Parents of 6 pancreatic insufficient children with CF aged 2–6 years attended a BIP consisting of 4×2 hour weekly sessions +1 follow up session 5 weeks later. Content included parenting pyramid framework, use of praise to maximise effect at mealtimes, boundary-setting, increasing food variety, and managing enzyme replacement therapy.

Parents completed questionnaires at the end of the BIP and 6 months later.

Body Mass Index z-score (BMIZ) was calculated at baseline, 3 and 6 months after BIP completion.

Results: All parents completed the BIP and evaluation. Session attendance was 93%.

Evaluation 6 months post-BIP

100% parents (6/6) felt more confident about managing mealtime behaviour and 67% (4/6) felt a lot more confident

83% (5/6) found mealtimes less stressful

83% (5/6) felt their child enjoyed mealtimes more

Parents unanimously reported (open question, no prompt boxes) that the most helpful aspect of the BIP was the opportunity to share experiences.

BMIZ at 3 months compared with baseline was increased (p=0.002) although this was not sustained at 6 months (p=0.38).

Conclusions: A pilot BIP for parents of children with CF shows promising results in modifying mealtime behaviour and reducing stress. BMIZ is significantly increased at 3 months follow up but not 6 months. A key finding from the study is the value that parents place on peer support and the opportunity to share experiences within the framework of an educational programme.

284 The development of guidelines for the use of megestrol acetate (Megace®) as an appetite stimulant in adults with cystic fibrosis

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Objectives: Poor nutritional status remains a challenge in some CF patients, contributing to decline in lung function and premature death. In the USA megestrol acetate (MA) is licensed for use as an appetite stimulant in HIV, AIDS and cancer. Small studies with CF patients have shown weight gain but there is insufficient evidence to routinely recommend use. Following trials of MA with patients failing to respond to, or declining conventional therapies we developed guidelines for the use of this unlicensed drug, considering safety and efficacy.

Methods: We evaluated use of MA in 5 patients. 4 patients had CF-related diabetes, 3 patients were using overnight feeding. Initial dose was 320 mg/day; duration of therapy ranged from 4 weeks to 10 months except 1 patient taking 4 week courses alternate months long term. 4 patients gained weight and reported significant improvement in quality of life; 1 patient reported increased appetite but gained no weight. Side effects were: adrenal insufficiency (permanent in patient taking MA longterm), impotence, hyperphagia, abdominal bloating. Dose adjustment suggested efficacy at lower dose in most patients. Following literature review, and consultation with other users of MA, guidelines were produced regarding identification of patients, dosage, duration of treatment and monitoring of effects, including adrenal suppression.

Conclusion: Use of MA was associated with improvement in nutritional status and quality of life in patients not responding to conventional therapy but is not without side-effects. We recommend an initial dose of 160 mg/d. The development of guidelines ensures appropriate use of MA and monitoring of side-effects.