The effects of lung transplantation on the nutritional status of patients with cystic fibrosis

M. Cararà1, S.J. Doe1, A. Anderson1, K. Harispal1, S. Johnston1, K. Stavers1, S.J. Bourke1, 1Adult Cystic Fibrosis Centre, Royal Victoria Infirmary, Newcastle upon Tyne, United Kingdom

Introduction: Many patients with advanced CF are malnourished due to the increased energy demands of lung infection and reduced nutritional intake associated with anorexia and malabsorption. Lung transplantation removes the catabolic effect of infected lungs but patients still have pancreatic insufficiency. We studied the weight change occurring in our patients pre and post transplant.

Results: Twelve patients in our CF Centre had their nutritional status assessed for one year before and after transplantation. Their mean age at transplant was 29.3 (range 19–42) years and mean waiting time to transplant was 16.6 (range 8–63) months. Their mean body mass index (BMI) was generally maintained by intensive nutritional support pre transplant. With the dramatic improvement in lung function most patients gained weight in the year post transplant, but only 4 (33%) had achieved a normal BMI (22–23 kg/m²).

Conclusion: With improved lung function and removal of the catabolic effect of infected lungs, BMI generally improves but patients still have nutritional deficits and require ongoing dietary support.

<table>
<thead>
<tr>
<th></th>
<th>BMI 1 year before Transplant</th>
<th>BMI 1 year post transplant</th>
<th>FEV1 (L) pre Transplant</th>
<th>FEV1 (L) 1 year post Transplant</th>
</tr>
</thead>
<tbody>
<tr>
<td>MA dose (mg)</td>
<td>20 (16–22)</td>
<td>19.6 (16.3–24.3)</td>
<td>22.0 (16.8–29.6)</td>
<td>3.8 (1.61–4.94)</td>
</tr>
</tbody>
</table>

Supplement Snack-Bar: a new approach to improve compliance with oral supplements in children with CF

C. Smith1, J. Lenton3, C. Warde3, P. Seddon1, 1Cystic Fibrosis Clinic, Royal Alexandra Children’s Hospital, Brightton, United Kingdom

Optimal nutrition during childhood in CF is associated with improved survival. However, studies of interventions with oral supplements have shown conflicting results, and it has been suggested that this may be partly because of poor compliance. Contributing factors to poor compliance are prescribing of untried supplements and poor understanding of varieties available: children’s tastes are individual and unpredictable.

To address these issues and improve compliance, we introduced a novel “Supplement snack-bar” into our paediatric CF clinic for an experimental 9-month period, and assessed parents’ and children’s responses by anonymised questionnaires. At each clinic we invited manufacturers to present full-range tasting sessions of their products. Any child felt clinically to require oral supplements was able to browse the range and identify a preferred product before prescribing. Each child over 7 years of age was given one questionnaire to complete and one questionnaire for the accompanying parent.

11 child/parent pairs of questionnaires were distributed; 11 child and 9 parent questionnaires were returned. 55% of parents and 36% of children found the sessions “very helpful”, while a further 44% of parents and 55% of children found them “fairly helpful”. When asked if there had been any effect on subsequent compliance with supplements, 11% of parents and 45% of children felt it had made a “big difference”, while a further 89% of parents and 36% of children felt it had made “some difference” – no parents or children rated “no difference”.

Our results suggest this type of intervention is popular, and may have the potential to improve compliance with nutritional supplements, but a further study using more objective measures of compliance is needed.

The experience of using megestrol acetate (MA) in a large UK adult CF centre

J. Barrett1, B. Ahitan1, E.F. Nash1, J. Whitehouse1, D. Honeybourn1, 1West Midlands Adult Cystic Fibrosis Centre, Birmingham, United Kingdom

Background: Cystic Fibrosis (CF) leads to poor nutritional status in many patients. Appetite stimulants such as MA may be used with conventional nutritional treatments to improve appetite and aid weight gain. We report our experience of using MA in CF adults.

Methods: We identified all patients treated with MA from 2003–2008. We recorded gender, age, CF-related diabetes (CFRD), height, weight, body mass index (BMI) and lung function before starting MA. We recorded duration of treatment, dose, reason for stopping MA and side effects. Nutritional interventions while receiving MA were documented. Weight, BMI and lung function were recorded on stopping MA, as well as 3 and 6 months afterwards.

Results: 25 patients were included (12 male). 2 were post-lung transplantation and 2 were post-liver transplantation. Median age at the time of starting MA was 27.4 yrs (range 18.1–50.8 yrs). 4 patients had CFRD. Median starting dose of MA was 320 mg (range 160–480 mg) and median duration of therapy was 83 days (range 10–756 days). Median weight gain during the treatment period was 4.9 kg (range –0.8–14.7 kg). There was a significant correlation between duration of treatment and weight change over the treatment period (R=0.65, p < 0.001). Weight gain was generally maintained 3 months after discontinuing MA (median 3.6 kg weight gain vs. baseline) and at 6 months (median 3.8 kg weight gain vs. baseline). FEV1% predicted was significantly increased comparing pre and post MA treatment (p < 0.001).

Conclusions: Adult CF patients receiving MA in addition to other nutritional supplementation gain weight and improve lung function. This weight gain is sustained up to 6 months after discontinuing MA. MA is generally well tolerated with few patients reporting adverse events.