332 The effect of long-term oral steroids on growth in children with cystic fibrosis

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Background: Previous randomised controlled trials of high dose oral corticosteroids in cystic fibrosis (CF) have reported serious adverse effects including decreased growth. The long term effects of regular low dose oral corticosteroids on growth are less clear.
Aims: To describe the long term effects on growth of low dose oral steroids on children with CF.
Methods: Case control comparison of growth of subjects receiving regular oral steroids for greater than 6 months, and age and sex matched controls. Height, weight and body mass index (BMI) were collected, and standard deviation scores (SDS) calculated. For each subject the slope of change in height, weight and BMI SDS was calculated by linear regression for 24 months before commencing steroids, and for the period after starting steroids.
Results: 28 cases and 28 controls were identified, with no significant differences between the two groups at baseline. Mean duration of follow up data was 4 years for both groups. Mean daily dose of prednisolone was 0.15 mg/kg. There were no significant differences in slope of weight or BMI SDS between groups. There was a significant difference in slope of height SDS between the cases and controls after commencing steroids (–0.12 v 0.02 SDS/yr; 95% CI diff –0.24, –0.01; $p = 0.032$). Growth suppression was greatest in the first year (–0.17 v 0.10 SDS/yr, 95% CI diff –0.47, –0.07; $p = 0.008$), and there was no significant difference in change in height SDS in the subsequent 2 (–0.06 v –0.10 SDS/yr; $p = 0.6$) or 4 years (–0.03 v –0.04 SDS/yr; $p = 0.9$).
Conclusions: Use of regular low dose oral steroids in CF has a short term effect on growth, but thereafter growth appears normal.

333 Long term effects of inhaled corticosteroids on adult height: data from the Belgian CF Registry (BMR-RBM)

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Background: Inhaled corticosteroids (ICS) are frequently prescribed to patients with cystic fibrosis (CF) without clear evidence of efficacy in the treatment of lung disease. In the Belgian CF Registry, nearly 50% of the patients are reported to use ICS. A previous mixed model analysis of the Belgian CF patients has shown that ICS use is associated with a reduction in the rate of FEV1 decline but this was accompanied by a lower yearly growth (3 mm per year).
Objectives: The objective of the study is to analyse the long-term effects on growth of ICS taken by CF patients during childhood and adolescence by reporting their adult height.
Methods: The adult height of 31 patients never taking ICS (no-ICS group) was compared to that of 40 patients using ICS >50% of time (ICS group). ICS use was documented at least 3 years before attaining adult height. Patients with a transplant and those using oral steroids were excluded. Adult height was expressed in SDS according to CDC references. 55% were F508del homozygous.
Results: Mean adult height SDS was similar in the 2 groups and was respectively –0.28±0.82 SDS in the no-ICS group (at 18.4±2.1 years) and –0.49±1.05. SDS in the ICS group (at 18.6±1.7 years) after a mean follow-up period until adult height of 6.2±1.8 years. Mean FEV1 % predicted obtained at adult height was lower (67.0±20.6%) in the ICS group than in the no-ICS group (83.4±21.3%) $p = 0.002$.
Conclusions: This analysis of long term ICS use shows no difference in adult height between CF patients who never used ICS and those who used ICS during more than 50% of the study period. As previously reported ICS appear preferentially used in patients with worse lung function.

334 10 years on – has the nutritional status in adults attending a large cystic fibrosis centre changed?

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The importance of nutritional care in cystic fibrosis (CF) is well recognised. Malnutrition in CF is associated with poorer pulmonary function and is an independent risk factor for poor survival.
In order to compare the nutritional status of adults attending the Prince Charles Adult CF Centre in 2009 versus 1999 a retrospective chart audit of 212 patients was completed. Data including nutritional status, pancreatic enzyme replacement therapy (PERT) usage, glucose tolerance and dietetic review was collected.
This audit found a significant improvement in mean BMI (21.1±3.0 versus 22.5±3.8; $p = 0.0017$), reflecting an improved nutritional status over the 10 year period. Prevalence of abnormal glucose tolerance has increased (12% versus 26%; $p = 0.004$) most likely due to a vigilant screening program commenced in 2000. Use of commercial oral supplements is higher, notably the percentage of patients using commercial oral supplements in 2009 (26% versus 45%; $p = 0.00001$), is higher than identified in the recent DAA survey of nutrition management practices of Australian Dietitians at 18%. Annual dietetic review is also a statistically significant change (73% versus 90% $p = 0.00009$), this increased review rate despite a two fold increase in the overall CF population may be attributable to a stable and experienced workforce as the dietetic staffing and FTE has remained stable over the 10 year period.

335 Improvement of nutritional status in a cohort of adults with CF

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Introduction: Optimal nutritional management is essential in optimizing quality of life and survival in CF. Since 2004, a dietician has been dedicated to our CF patients.
Objective: To evaluate changes in the nutritional status of the CF patients cared at our adult CF centre.
Method: We compared the nutritional status of 301 patients seen in 2007 with that of 163 patients seen in 1997. Malnutrition was defined as BMI < 18.5 and severe malnutrition as BMI < 16 kg/m^2.
Results: In 2007, mean age was 29.3 yrs (vs 28.8 yrs in 1997). Mean BMI was 20.8±2.3 and 20.1±3.4 kg/m^2 in men and women, respectively (vs 19.2±3.0 and 19.1±2.6 kg/m^2 in 1997). Malnutrition was seen in 77 patients (25.6% as compared to 49.7% in 1997). Severe malnutrition was found in 13 patients (4.3% vs 22.1% in 1997). Malnutrition was more frequent in females (48 patients, 32.6%) than in males (29 patients, 18.8%). This was even more pronounced for severe malnutrition, found in 10 females (6.8%) and 3 males (2.0%). The frequency of exocrine pancreatic insufficiency was unchanged (80.4% vs 83.4% in 1997). Diabetes mellitus was more frequent in 2007 (23.9%) than in 1997 (18.4%), probably because it was detected more actively. More females (33%) than males (16%) had diabetes. Hepatic cirrhosis was present in 6.6% of the patients (vs 9.8 % in 1997). Enteral nutrition had been used in 19 patients, 6.3% (vs 6.7% in 1997). The respiratory function had improved, with mean FEV1 56.3±24.5% pred., as compared to 47.1±25.5% pred. in 1997.
Conclusion: Nutritional status and respiratory function have improved in our CF patients over those last 10 years, probably thanks to a more active dietary management.