360* Nutritional status, body composition and exercise capacity in children with cystic fibrosis

K. Rylance[1], C.J. Taylor[1], A.S. Rigby[2]. [1]*Academic Unit Child Health, University of Sheffield, Sheffield, S.Yorkshire, United Kingdom;* [2]*Dept. Respiratory Medicine, University of Hull, Hull, United Kingdom*

Nutrition is seen as an important facet of CF management. Nutrition is broadly equated with growth and weight gain and assessed by measurement of body mass index (BMI) and weight and height centiles. With adequate pancreatic enzyme replacement therapy most CF affected patients achieve satisfactory growth throughout childhood.
Recently, we have observed excessive weight gain in a proportion of our CF clinic population; this is often associated with poor exercise capacity. This has led us to question the value of weight gain alone as a measure of nutritional sufficiency and to focus on maintaining linear growth and exercise capacity rather than achieving pure weight gain. Our CF population now exhibits normal linear growth and excellent lung function but with a lower fat mass and fat-free mass.
The aim of this study was to measure nutritional status, body composition and exercise capacity in a cross section of CF patients with a view to comparing the relative effects of nutrition and fitness on lung function. Fat-free mass (FFM) and fat body mass (FBM) were derived from dual-energy X-ray absorptiometry (DEXA); exercise capacity was measured using the shuttle walk test. 243 data sets from 115 subjects were analysed.
Overall, the study population was in good health (mean FEV1 90.1%), there were, however, significant gender differences in BMI Z-score, mean –0.25, males –0.4, females –0.12, and %FBM, mean 19.61%, males 16.72%, females 21.20%, and %FFM, mean 76.47%, males 79.57%, females 75.14%. FEV1% correlated weakly with nutritional markers and shuttle distance. Excess nutrition appears to be laid down as fat not lean mass. Lean body mass was important in predicting shuttle distance.

361 Do Danish children with and without CF share nutritional habits?

J. Sundstrup[1], S. Fagt[2], C. Mølgård[1], L. Onsgaard[1], T. Pressler[1]. [1]*Copenhagen CF-Center, Rigshospitalet, Copenhagen, Denmark;* [2]*Danish Institute for Food and Veterinary Research, Copenhagen, Denmark*

Background: CF patients are encouraged a diet providing 120–150% of recommended daily energy intake for age and gender. Fat (F) should provide 35–40% of dietary energy, protein (P) 15–20% and carbohydrates (CHO) 40–50%.
Aims: To compare food recalling data from healthy Danish children to similar data from children with CF.
Methods: Data on energy and macronutrient intake from 47 Danish CF patients aged 4–17 y were calculated, analysed and compared to data from The Danish National Survey of Dietary Habits and Physical Activity 2000–02 (DNSDHPA).
Patients were divided into two groups: 4–9 year-olds (n = 19; f/m 9/10; median age 6.4 y) and 10–17 year-olds (n = 28; f/m 15/13; median age 13.5 y).
Results: Among 4–9 year-old patients, daily energy intake was 19% higher than among their healthy contemporaries. Composition of macronutrients in the diet was 38E% from F, 48E% from CHO and 13E% from P. Corresponding data from DNSDHPA showed a diet composition of 34E% from F, 51E% from CHO and 14E% from P in a similar age group.
Compared to results from DNSDHPA, the group of 10–17 year-old patients had a daily energy intake 32% above their contemporaries. Data on macronutrient intake in the group showed a diet composition of 37E% from F, 48E% from CHO and 14E% from P. Similar figures from DNSDHPA were 32E% from F, 52E% from CHO and 14E% from P.
Conclusions: Energy intake in Danish CF patients was 119–132% of average energy intake in healthy Danish children. Also dietary energy from fat was higher, up to 38E% in CF patients vs. 34E% in the Danish population. In conclusion, Danish CF children follow nutritional recommendations for daily energy intake and FatE% according to recommendations for cystic fibrosis.

362 Meconium ileus, nutritional values and lung function

A.M. Olesen[1], J. Sundstrup[2], K. Nilsson[3], I.E. Moen[4]. [1]*Aarhus University Hospital, Aarhus, Denmark;* [2]*Rigshospitalet, Copenhagen, Denmark;* [3]*Karolinska University Hospital, Stockholm, Sweden;* [4]*Ullevaal University Hospital, Oslo, Norway*

Aim: To compare nutritional values and lung function in CF patients with a history of meconium ileus (MI) and CF patients without meconium ileus.
Method: Data from 601 CF patients with pancreatic insufficiency (PI) (100 MI) <30 years from seven Scandinavian CF centres were analysed.
We evaluated the nutritional values: BMI zscore, vitamin A, D and E deficiency, and lipase/kg bodyweight (bw)/day.
Furthermore we evaluated FEV1.0
Results: We found no significant difference in BMI zscore between the two groups. We saw a tendency of lower BMI in the MI group of patients <10 years (mean –0.22 versus 0.06 for the non MI group).
Vitamin A, D and E deficiency (defined as serum vitamin values < the lowest normal value for the age) was present in both groups but with no significant difference between the groups.
Units of lipase/kg bw/day was significantly different between the two groups (median 5470 units of lipase/kg bw/day in the non MI group versus 7890 units of lipase/kg bw/day in the MI group). However we found no significant difference in the group of patients <10 years.
The FEV1.0 % of predicted did not differ between the two groups, neither for the whole group nor in the different age groups (mean FEV1.0 value for both MI and non MI patients <10 years was 93% of predicted; mean FEV1.0 value for MI patients >10 years was 82% and for non MI >10 years 79%).
Conclusion: Our data showed only a significant difference between the MI and non MI group in the intake of lipase/kg bw/day. Other nutritional values and FEV1.0 were not significantly different between the groups.

363 Bone mineral density in cystic fibrosis patients – a 3-years follow-up and intervention

E. Očenášková[1], H. Vaníček[1], O. Pozler[1], A. Jebavá[2]. [1]*Paediatric Department, Faculty Hospital, Hradec Králové, Czech Republic;* [2]*Institute for Clinical Biochemistry and Diagnostics, Faculty Hospital, Hradec Králové, Czech Republic*

The aim of this study was to assess prospectively bone mineral density (BMD) in cystic fibrosis (CF) patients and the effect of vitamin D (vit.D) and calcium (Ca) supplementation adjustment. Subjects: 45 CF patients (aged 4.3–45 years (y), 27 female, 27% >18 y).
Methods: DXA (dual-energy X-ray absorptiometry) BMD of lumbar spine L1-L4, correction of BMD Z-score according to height and weight. Other parameters: anthropometry, FEV1, serum levels of Ca, total IgG, vit.D, alkaline phosphatase, calciuria. All measurements were repeated yearly, conducted 4×. Ergocalciferol and Ca supplementation were adjusted according to calciuria every 3 months.
Results: Weight, height and BMI were lower than in normal population [mean(SD) Z-score = –1.2(1.14), –1.01(1.16) and –0.48(0.90), respectively]. Weight and height Z-score did not change during follow up, BMI Z-score decreased. BMD was lower than in normal population in all yearly measurements [mean(SD) Z-score = –1.7(1.0), –1.2(0.9), –1.3(1.1) and –1.4(1.0), $p < 0.001$), also after correction. Between the 1st and 2nd measurement, BMD Z-score increased in patients aged <18 y ($p = 6.6 \times 10^{-7}$) and then stayed stabilized, in patients >18 y did not change. Ergocalciferol and Ca supplementation was increased between the 1st and 2nd measurement. 25(OH)-vit.D and 1,25(OH)2-vit.D levels were normal and did not change during follow-up, however, 70% values of 25(OH)-vit.D levels were <75 nmol/L. Calciuria was in normal range and decreased during follow-up.
Conclusions: Low BMD may be present already in prepubertal CF patients. Vitamin D and Ca supplementation adjustment according to calciuria may help to improve lumbar spine BMD in CF patients.