340* Longitudinal changes in bone mass in children with cystic fibrosis: effect of size adjustment using bone mineral apparent density

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Objective: Patients with cystic fibrosis (CF) are at risk of poor growth, sub-optimal bone mineralization and osteoporosis. Bone monitoring using dual-energy X-Ray absorptiometry (DXA) is recommended from age 10 yrs, however, DXA derived bone mineral density (BMD) standard deviation scores (sds) do not fully adjust for body size and may give deceptive results for small children. We assessed (1) trends in BMD over time, (2) effect of size-adjustment using Bone Mineral Apparent Density (BMAD).
Methods: 56 children (32 girls) with CF had DXA measurements (GE Lunar Prodigy) of the lumbar spine (L2-L4) at baseline (7–12 yrs) and 2 yrs, providing BMDsds for age and sex; 34 children were measured at 4 yrs. BMAD was calculated as $BMC/BA^{1.5}$ and BMADsds derived for age and sex using UK reference data.
Results: See the table.

Characteristics and Bone Mineral Density SDS

Year	Boys				Girls			
	N	Height	BMD	BMAD	N	Height	BMD	BMAD
0	24	-0.7(1.1) $p<0.01$	-0.2(0.7)	0(1.1)	32	-0.7(1.2) $p<0.01$	-0.7(1.1) $p<0.001$	-0.6(1.0) $p<0.001$
2	24	-0.6(1.1) $p<0.05$	-0.4(0.8) $p<0.05$	0.1(1.1)	32	-0.5(1.2) $p<0.05$	-1.1(0.9) $p<0.001$	-0.7(0.9) $p<0.001$
4	15	-0.6(1.4)	-0.8(1.1) $p<0.05$	-0.9(1.0)	19	-0.5(1.3)	-1.1(1.3) $p<0.001$	-0.6(1.0) $p<0.05$

Seven children had BMDsds <–2 with normal BMADsds; 8 children had both BMDsds and BMADsds <–2. Twenty children had BMADsds at least 0.5 sds lower than BMDsds. Two children <10 yrs had BMD and BMADsds <–2.
Conclusion: Mean BMDsds was low and fell progressively with age especially in girls. Although use of BMADsds reduced the apparent bone deficit and may avoid mis-diagnosis in some children small for age, in others BMADsds was lower than BMDsds. Two children <10 yrs had low BMADsds but would not have been scanned under current UK guidelines.

341 Dietary intake and DXA total body measurements in children and adolescents with cystic fibrosis

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Careful management of dietary intake and growth is extremely important in cystic fibrosis (CF). The aim of our study was to evaluate dietary intake and body composition in a group of paediatric CF patients.
We evaluated 27 subjects (M16/F11) aged 5–17 years (mean = 12.3 yrs). Dietary intake was assessed with a 24 h recall diary. Body composition was evaluated with DXA total-body measurements (whole body bone mineral content BMC, lean body mass LBM and fat mass FM) using the Hologic QDR Discovery A fan-beam scanner (Hologic, Inc., Bedford, Massachusetts), running software version 12.4 (fast-array mode). According to international consensus, DXA measurements without head were used.
Dietary intake in our patients was appropriated in total Kcal intake (mean = 2022.3 kcal/die) and in nutrients composition (lipids 35%, proteins 19%, carbohydrates 46%, calcium 1126 mg/die, vitamin D 296UI/die). We found no difference between pre-pubertal and pubertal patients and between *Pseudomonas aeruginosa* positive and negative patients.
In our population BMC, LBM and FM were lower than normal (mean Z-score for BMC = –1.87, LBM = –1.06 and FM = –0.54).
We found a significant correlation between total Kcal intake and: BMC ($p=0.001$), LBM ($p<0.001$), FM ($p=0.02$). We also found that protein intake correlates with BMC ($p=0.003$), LBM ($p<0.001$), FM ($p=0.05$) and lipids intake correlates with BMC ($p=0.028$) and LBM ($p=0.013$).
In our patients there is also a significant correlation ($p=0.012$) between lipids and vitamin D intake, probably due to the high consumption of nuts and peanuts, rich in monounsaturated acids, and olive oil.
Conclusion: Body composition can be reduced despite adequate caloric intake.

342 Defining and classifying height in children with cystic fibrosis: comparison of z-score height for age and z-score target height

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Background: Nutritional status has a significant effect on survival in patients with Cystic Fibrosis (CF). A part of the assessment of nutritional status is height measurement. In clinical practice, measured height is often compared with reference values of healthy children by converting it to z-score height-for-age (HFA). A limitation of this method is that the genetic potential is not taken into consideration. Therefore height expressed as z-score target height (TH) might be more accurate.
Objective: The purpose of this study was to compare classification of height expressed as z-score HFA with z-score TH in Dutch children with CF.
Design: In a cohort of 169 Dutch children (90 boys) with CF (age 11.1±3.7 years), height was measured with a stadiometer to the nearest 0.5 cm and expressed as z-score HFA. TH is derived from the average of the two parents' heights, with an adjustment added for boys and subtracted for girls. To express current height related to TH, z-score for TH was subtracted from HFA. Z-score <–2 was classified as malnutrition.
Results: Mean height expressed as HFA and TH was respectively –0.5±1.0 (2.2 – –2.8) and –1.0±1.0 (1.7 – –3.4). Assessment of the nutritional status according to HFA and TH showed that respectively 52 (31%) and 25 (15%) children were in a good nutritional status (z-score ⩾0). According to HFA and TH respectively, 14 (8%) and 26 (15%) children were malnourished.
Conclusion: Defining height according to HFA and TH results in a widespread outcome. Therefore, the use of z-score TH might be helpful to optimize the assessment of the nutritional status in individual patients with CF.

343 Does fat free mass index correlate better with pulmonary function than body mass index in adult CF patients before and after lung transplantation?

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Background: Malnutrition in cystic fibrosis (CF) patients is correlated with pulmonary function. Body mass index (BMI) might not be the best marker because it does not distinguish between fat mass (FM) and fat-free mass (FFM).
Aim of the study: To determine if fat free mass index (FFMI) correlates better with pulmonary function than BMI.
Methods: The study population consisted of 43 (25 men) CF patients who were screened for lung transplantation (mean age 29 y). FFMI, BMI and FEV1 were measured during out clinic visits. 22 subjects underwent lung transplantation. Measurements performed just before and after lung transplantation were included.
Results: At screening mean FEV1 was 23% for men (range 13%-42%) and 28% for women (range 22%-38%). Mean BMI was 19.6 kg/m^2 for men (range 16.1–28.4) and 19.9 kg/m^2 for women (range 16.5–23.4). Mean FFMI was 16.1 kg/m^2 for men (range 12.5–22.4) and 14.5 kg/m^2 for women (range 12.6–16.7) Both BMI and FFMI were correlated with FEV1 in men (both $r=0.75$, $p<0.001$), but not in women ($r=0.10$, $p=0.70$ and $r=0.26$, $p=0.29$ respectively). In patients just before lung transplantation ($n=22$) mean FEV1 was 21% (range 13%-41%) for men ($n=14$) and 24% (range 21%-30%) for women ($n=8$). In this sub group both BMI and FFMI were correlated with FEV1 ($r=0.56$, $p=0.008$ and $r=0.51$, $p=0.02$ adjusted for sex). After lung transplantation (>1 year, $n=15$) no correlations were observed for either BMI or FFMI to FEV1 (all $p>0.05$). Lung function increased to a mean FEV1 of 80% (range 39–109) in men and 79% (range 71–98) in women.
Conclusion: BMI and FFMI correlate similar to pulmonary function tests before and after lung transplantation.