Reduced bone mineral density (BMD) and altered body composition are well recognised in patients with CF. The causes of reduced BMD are multifactorial.

Aim: To assess the effect of genotype and sex on BMD and body composition in adults with CF.

Methods: BMD was measured at the lumbar spine (LS), femoral necks (FN) and total body (TB) by DXA (GE-Lunar Prodigy) and presented as Z scores. Weight, height and BMI were determined at the time of scanning. Height squared indices were derived for fat free mass (FFMI), total body lean (TBLI), total body bone mineral content (TBBMC) and fat mass (FMI). Patients were divided by sex and categorised as homozygous or heterozygous for the delta F508 mutation.

Results: 246 scans were performed in 125 males, 84 homozygous, mean age 23.8 (SD 5.1); 41 heterozygous, mean age 29.8 (SD 8.5); and 121 females, 76 homozygous, mean age 26.2 (SD 7.1); 45 heterozygous; mean age 26.4 (SD 8.1).

There were no significant differences for any parameter for females. Males homozygous for delta F508 had lower Z LS BMD –0.65 (SD 1.5) p = 0.01, Z TB BMD –0.9 (SD 1.3) compared to 0.02 (SD 1.4) p < 0.0001, FFMI 17.9 (SD 2.0) compared to 18.7 (SD 1.9) p < 0.05, TBBMC 0.86 (SD 0.13) compared to 0.96 (SD 0.14) p < 0.001 and FMI 3.7 (SD 2.2) compared to 4.7 (SD 2.7) p < 0.05.

Conclusion: Reduced bone mineral density in CF is multifactorial. Others have described an association with the delta F508 genotype. Our results support the hypothesis that reduced BMD in CF has a genetic component. Unlike other studies we found a difference between homozygous and heterozygous males. This effect was also exerted on FFMI and FMI.

**Nutritional therapy for catch-up growth in malnourished infants with Cystic Fibrosis – the role of semi-elemental formula with medium chain triglycerides**

S. Mosescu, V. Dan. Clinical Central Children’s Hospital, Bucharest, Romania

Aims: to establish the most efficacious nutritional therapy for catch-up growth to the expected growth parameters in infants with cystic fibrosis (CF).

Methods: have been studied 12 infants with CF diagnosed in our Clinic. All of them were malnourished when the diagnosis of CF was done, with weight for height indices 50%–85%. Their growth was studied comparing their diets: lactose free formula, for 110–120 kcal/kg/day with formula: 6 gained weight and returned to normal growth (66.6%), with formula the same degree of malnutrition. From the 12 infants, 9 were fed with semi-elemental formula with MCT, 3 infants did not show significant correlations. Group B: BMI 18.27±0.36, BMI zscore 0.76±0.16, BFAT% 25.43±2.26, LBM% 74.54±2.27, PEmax 113.72±6.0 and MVV 99.83±11.7. PEmax correlated with LBM% (r 0.704, p 0.016) and BFAT% (r 0.709, p 0.015). PI max and MVV correlated with LBM% (r 0.43, p 0.041), BFAT% (r 0.41, p 0.048) and BMI zscore (r 0.45, p 0.03). The difference in PEmax was statistically significant (t=2.29, p=0.029) but there was no significant difference in PEmax and MVV means between the two groups.

Conclusion: Respiratory muscle strength and endurance are related to nutritional status. Up to 25% reduction of BMI does not affect muscle strength and endurance, provided high LBM% is maintained. A decrease in LBM% will first influence muscle strength and then muscle endurance.