Prevalence of bone demineralisation in Russian adult CF patients and associated risk factors

S. Krasovsky1, I. Baranova2, E.L. Amelina1, 1Cystic Fibrosis, Pulmonology Institute, Moscow, Russia; 2Russian State Medical University, Moscow, Russia

Decreased bone mineral density (BMD) is a serious problem for clinicians who care for adult CF patients. The aim of our study was to determine the prevalence of decreased BMD in adults with CF, and to assess risk factors for reduced bone mass in this group of patients. Patients and methods. 56 patients (23 males, mean age 22.2±4.3 yrs) were included in the study. BMD (expressed as a Z score) of L2–L4 spine and proximal femur were measured with Dual-Energy X-ray Absorbtionmetry (DEXA) scans on Hologic-4500A. Patients were divided into 3 groups: Z < -1 normal BMD, -1 > Z > -2 osteopenia, Z < -2 osteoporosis. Correlations between BMD and age, sex, anthropometric data, the age of pulmonary and intestinal manifestations setup, glucocorticoid use, physical activity, lung function, delF508 mutation, SatO2, dyspnea severity (MRC scale).

Results: BMD was reduced in a total of 75% (n=42) patients. Osteopenia was diagnosed in 35.7% (n=20), osteoporosis – in 39% (n=22). Reduced bone density significantly correlated with FEV1 (p=0.02), FVC (p=0.03), SatO2 (p=0.01), physical activity (p=0.01), dyspnea severity (p=0.02).

Conclusion: Bone decomposition affects the majority of adult CF patients and is strongly related to pulmonary disease severity. The basic CF care program should include bone metabolism therapy.

Proteinuria in patients with cystic fibrosis: is there an association with genotype?

E.L. Gamboa1, J.M. Cemlyn-Jones1, 1Pulmonology Department, Coimbra University Hospital, Coimbra, Portugal

Introduction: CFTR is expressed in all nephron segments but in CF patients there is no well-established renal phenotype and no major primary renal dysfunction. Objective: To evaluate proteinuria in our CF population and its correlation with genotype. Secondary objectives were to assess correlation of proteinuria with CF related diabetes and disease severity. Methods: Prospective analysis of 22 CF patients that were evaluated for 24 hour protein excretion within a 6-month period. Patients were divided into two groups: A (proteinuria <150 mg/day) and B (proteinuria >150 mg/day). Genotype data were obtained from patient files and the two main mutations, AF508 and R334W, were considered. Due to the existence of genotype AF508 /R334W two categories were created to enable statistical analysis so that in category 1 this genotype was filed as “AF508” and in category 2 as “R334W”. CF related diabetes data were obtained from patient files and disease severity was assessed by acute exacerbations in the last semester and FEV1, measured during the study period.

Results: In group A all patients (100%) were AF508 when considering category 1 and 86.7% for category 2. In group B 60% of patients were R334W when considering category 1 and 80% for category 2. Therefore, mutation AF508 tended to be associated to normal values of proteinuria and mutation R334W tended to be associated with higher proteinuria values (p=0.009 for category 1 group analysis and p=0.014 for category 2 group analysis).

No significant relationship was found between the different groups and the other variables.

Conclusions: Results suggest a possible mutation association eventually according to the mechanism by which they disrupt CFTR function and renal phenotype.