

277 Bone mineral density in patients with Cystic Fibrosis of Yaroslavl regional center

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Aims: to assess the frequency of osteopenia in cystic fibrosis (CF) patients and the clinical, functional and biochemical variables associated with reduced bone mineral density (BMD).

Patients and methods: BMD of the lumbar spine (L2-L4) was examined in 26 patients with CF aged 2–22 years using dual energy X-ray absorptiometry. Anthropometric variables, disease severity, pulmonary function, exercise capacity, serum calcium, phosphate, 25(OH)vitamin D, C-end telopeptides, osteocalcin levels, gonadal hormones concentration were assessed and related to BMD.

Results: In 34.6% patients using the tables of conjugated values of anthropometric and densitometric indices BMD was significantly reduced. Z-score was -1.8 ± 0.93 SD of normal values and correlated with the body mass index, and the Shwachman score. Forced expiratory volume in 1 second, vital capacity, exercise capacity were lower, residual volume higher in children with osteopenia. The serum levels of calcium and 25(OH)vitamin D were lower than normal in 39% and 48% cases respectively, but there was no significant difference in children with normal and reduced BMD. Urine calcium excretion was decreased in 57% cases and related with bone mass and BMD. Serum C-end telopeptides levels were elevated in 72.7% patients. Osteocalcin were decreased in 63.6% cases. Testosterone and oestradiol deficiency was determined in 61.1% of males and 42.8% females respectively, but decreased BMD did not correlate with lowered gonadal hormones concentration.

Conclusion: Osteopenia is frequently observed in CF patients. Nutrition status, disease severity are factors most highly correlated with decreased BMD. The dual X-ray absorptiometry should be an integral part of investigation in CF subjects in order to prevent and treat osteoporosis.

279 The retrospective assessment of bone mass density in patients with Cystic Fibrosis

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Regarding the lifestyle quality handicap associated with CF, limited spontaneous mobility and application of steroidtherapy, there is a potential disorder of bone tissue mineralization.

Aims: the aim was the retrospective (annual), quantitative assessment of bone mass density change in patients with cystic fibrosis and assessment of prophylaxis in cases with osteopenia and osteoporosis.

Methods: The study was performed in group of 21 CF patients (8F, 13M) aged 3–24 yrs. Total bone mineral density and lumbar spine bone mineral density of L2-L4 region was evaluated. Obtained results were expressed as Z-scores. The results were also presented using the authentic method of percentiles. Osteopenia was assumed when the BMD was $<1SD$ and osteoporosis when $<2SD$. Values of total BMD below 5th perc. were equivalent to osteopenia. In all patients daily supplementation of vit.D3 (800 units) and calcium (800–1200 mg) was provided.

Results: In 4 females (50%) Z-score BMD SPINE was $<2SD$ which equals values $<5\%$. In 5 males (38.5%) Z-score BMD SPINE was below 2SD which was at 5th perc. and below, and in 5 males (38.5%) BMD SPINE was below 1SD indicating osteopenia. In annual evaluation only in one female and in one male the BMD SPINE values had an increasing trend and were within normal percentile values.

Conclusions: The calcium supplementation in CF patients is necessary but not always sufficient. In the advanced cases of CF related osteoporosis other the use of other confections should be considered and assessed. The annual assessment of total and lumbar bone mass density is essential for therapy modification.

278 Bone mineral density (BMD) in Cystic Fibrosis patients (CFP)

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Despite the advances in clinical and nutritional treatments of CFP, complications such reduction of the BMD are described.

The objective of this study was to evaluate BMD in CFP. A transversal descriptive study at the University Hospital was done. CFP older than 10 years were selected. Nutritional status was assessed by anthropometrics indices weight/age, height/age and body mass index (kg/m²). Lumbar spine BMD (segment L2-L4), femur and whole body were evaluated by a dual-energy x-ray absorptiometry (DEXA). This exam also allows the measurement of both lean and fat mass (kg). The dietary food intake and pulmonary function test was determined. Mean age (14.4 ± 3.85 years) male (58.6%). The weight/age and height/age Z-scores demonstrated malnutrition: 60.9% and 65.2%, respectively. The evaluation of bone densitometry determined that 58.6% of the patients had reduced bone mineral density at the lumbar spine and 42.3% of them had it on the entire body. There was a positive correlation between BMD of lumbar spine, femur and whole body and age, body mass index and lean mass ($p < 0.05$). The pulmonary disease severity and pancreatic insufficiency present no association with BMD. The dietary food intake revealed adequate values for nutrients that were beyond a 100% of the European Consensus on Cystic Fibrosis recommendation.

Conclusion: The results on this study demonstrated a significant change in BMD and this finds are in concordance to the scientific literature. The maintenance of an adequate nutritional status may guarantee effective protection for bone mass loss at the lumbar spine and at the whole body. A prevention bone mineralization is important in CF patients because it might reduce fracture risk.

280* Serum phospholipid fatty acid pattern is associated with bone growth in children but not adults with Cystic Fibrosis

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Essential fatty acids (EFA) have proved to be important for normal bone mineral density (BMD) and bone growth in animal studies. Patients with cystic fibrosis (CF) often have low serum EFA levels. The aim of the study was to analyse if BMD was related to FA status in patients with CF.

Methods: Bone mass was studied prospectively in patients with CF, aged 6–33 yrs (35 children and 19 adults), during a two-year period (Gronowitz et al. *Pediatrics* 2004; 114: 435). BMD was measured with dual x-ray absorptiometry, and FA concentrations in serum phospholipids were determined with capillary gas-liquid chromatography (*J Lipid Res* 2001; 42: 359)

Results: The CF patients showed low linoleic acid concentration and high ratio of arachidonic acid (AA) to docosahexaenoic acid (DHA) in serum in both children and adults compared to healthy controls. The ratio of eicosatrienoic acid (ETA) and AA, an index of EFA deficiency, was high and increased further during two years in the children, as did the total concentration of saturated FA. Bone mineral content in lumbar spine (LS) and femoral neck (FN) were positively correlated to the most common saturated FA, palmitic acid only in the children with CF, $r = 0.6$, $p = 0.0001$ and $r = 0.5$, $p = 0.001$, respectively. The LS BMD z-score and change in z-score correlated negatively to the AA/DHA ratio in children (both $r = -0.4$, $p = 0.004$ and 0.02 , respectively), but not in adults. The EFA deficiency index was positively correlated to the bone area of LS ($p < 0.01$) and FN ($p < 0.001$) in both children and adults.

Conclusion: Our results imply that FA status influenced bone growth in CF children, but not in adults, suggesting that FA status would be important for bone modelling but not for bone remodelling.