SUMMARY

Introduction: Chronic disease in childhood can negatively influence the development of skeleton. Bone disease has been known for more than 20 years as a complication of cystic fibrosis. It is probably secondary and has a multifactorial origin. Bone health in CF patients is influenced by nutritional state, disorders of vitamin metabolism, liver disease, inflammation, low physical activity, hypogonadism and delayed puberty. The **goal** of this work was to asses bone mineral density in two groups of CF patients, identificate influencing factors and assess positives and negatives of two densitometric methods. **Patients:** Tübingen (CF-Tübingen) – 33 patients, 15 female, age 4,5-43,7 years, median 10,4 years, 79% <18 years. Hradec Králové (CF-HK) – 42 patients, 26 female, age 4,3 – 45 years, median 10,9 years, 69% <18 years. **Methods:** CFTR mutation, pankreatic sufficiency, microbial colonisation, anthropometric parameters, serum levels of Ca, phosphate, IgG, vitamin D, ALP activity, FEV1, body composition by BIA (CF-Tübingen), body composition and bone mineral density by dual energy X-ray absorptiometry (BMD, BA, BMC) of the whole body (CF-Tübingen) and BMD of lumbar spine L1-L4 (CF-HK). In CF-Tübingen, proximal and distal forearm was measured by peripheral quantitative computed tomography (pQCT). In distal forearm, volumetric trabecular bone density was measured (TBD). In proximal forearm, cross-section area (CSA) of muscle and bone components and volumetric cortical bone mineral density (CBD) was measured. **Results:** Patients CF-Tübingen had normal whole-body BMD ($p = 0.393$), thin cortical bone and small proximal forearm muscle CSA ($p < 0,001$). CSA of cortical bone was dependent on proximal forearm muscle CSA ($R^2=0,75$, $p < 0,001$). Patients had low BMD L1-L4, which persisted also after height- or weight-adjustment in patients < 15 years of age. BMD and BMC were dependent especially on weight and height. Increased vitamin D and calcium supplementation, based on urinary calcium excretion, lead to BMD increase higher than expected in patients < 20 years of age. **Conclusion:** In CF patients, normal whole body BMD and low lumbar spine BMD can be expected, even after height adjustment. In CF patients, body weight can be used for BMD adjustment in DXA. PQCT is a good complementary densitometric method. It enables a complex assesment of the relationship between bone and muscle. Wider use of pQCT should be encouraged.