338* Mineral bone density in children and bone status of adults with cystic fibrosis

E. Colin[1], A.C. Wagner[1], E. Chaillou[1], E. Darviot[1], T. Urban[1], M. Audran[2], J.L. Giniès[1]. [1]*Centre de Ressources et de Compétences pour la Mucoviscidose, Centre Hospitalier et Universitaire, Angers, France;* [2]*Service de Rhumatologie, Centre Hospitalier et Universitaire, Angers, France*

Objective: The aim of this study was to determine whether the Z-scores for bone mineral density measured in childhood and early adolescence and adjusted for chronological age, stature, and bone age, are able to predict adult bone status in patients with cystic fibrosis.
Material and Methods: This study was retrospective. The medical records of 20 patients with cystic fibrosis were examined. All patients had had several bone density test, with at least one since age 18. The adult bone mineral density (BMD) Z-scores were compared with those measured during the first densitometry in childhood and another from prepuberty.
Results: Differences between the chronological age, stature, and bone age were noted at the first and the prepubertal densitometries. When discordance was noted between BMD adjusted for chronological age and BMD adjusted for bone age or stature, the adjustment for chronological age was significantly more predictive of the results seen in adulthood ($p < 0.01$).
Conclusion: This study shows in a small number of patients that the childhood BMD Z-score adjusted for chronological age best predicts adult bone status. Adjustment for stature or bone age may overestimate bone mineralization, thereby delaying the implementation of appropriate therapy.

339 Assessment of bone mass density by quantitative ultrasonography in children with cystic fibrosis

O. Turcu[1], S. Sciuca[1], E. Chioroglo[1]. [1]*Pediatric pulmonology, State Medical and Pharmaceutical University, Chisinau, Moldova*

Background: Reduced body mass density (BMD) has been reported in earlier studies of cystic fibrosis (CF) patients. Quantitative ultrasonography (QUS) method recently has been introduced for the assessment of BMD. This technique is safe, noninvasive, easy to use, and radiation-free, making it ideal for use in children. QUS offers information on skeletal status, matrix microstructure and is important when assessing fracture risk.
Aim: In this our study the frequency of bone and mineral disorders in patients with CF using QUS method was assessed.
Material and methods: The study group included 33 patients with CF (18 males) aged 2.5–21 years. The diagnosis of CF had been made using a clinical features, sweat chloride test and identification of CFTR mutation. CF patients were assessed by QUS at the Sunlight Omnisense 7000S bone sonometer (Israel), witch measures the speed of sound (SOS) propagating axially along the distal 1/3 radius. Z-score was used for bone status evaluation, as standard deviation (DS) from the mean BMD of an age and gender matched control population (Z-score >−1 DS, osteopenia – Z-score <−1 DS and >−2.5 DS, osteoporosis – Z-score <−2.5 DS).
Results: All examined CF patients were divided on two groups according to age of everyone (I group 17 children aged 2.5–12 years, II group 15 children ⩾12 years). Z-score in all children were significantly lower (−2.86±0.3 DS) than age normal values. Children under 12 years, had the mean of Z-score −2.52±0.3 DS. In children older than 12 years, Z-score were −3.22±0.5 DS. The values of Z-score were normal in 3 children.
Conclusion: The monitoring of BMD in patients with CF is an important tool for detection of bone complications (osteopenia, osteoporosis) and fractures prevention with an adequate treatment.

340 Bone loss in CF: a fragmented picture

J. Robertson[1], K. MacDonald[2], J.A. Innes[1]. [1]*Scottish Adult CF Centre, Edinburgh, United Kingdom;* [2]*Queen Margaret University, Edinburgh, United Kingdom*

Bone loss is highly prevalent in our CF population (70%) and is associated with risk factors;age, low vit D levels, low BMI and CFRDM [1]. The evidence that low BMD is a predictor of fracture risk is inconclusive and whilst there is some data to suggest that bisphosphonates increase BMD [2], the significance of bone loss in our centre is unclear. We hypothesised that bone loss occurs in patients with known risk factors and treatment using CF Trust Bone Mineralisation Guidelines results in improved BMD. However, we present 4 case studies which show that not all patients "fit this picture" and that treatment may not result in improved BMD. Of 140 DEXA scans over 6 years we present several individual patients whose results conflict with received wisdom and challenge the new guidelines. Case 1: 37 yr old male: high risk for bone loss(Fev1 17%, BMI 20.4, CFRDM, low VIT D*. Treated with Alendronate for 5 yrs. DEXA scan currently lower than at initial presentation. Case 2: Female aged 43, high risk for bone loss (Fev1 25%, BMI 17 CFRDM, low VIT D) but DEXA scan normal. Case 3: 39 yr old male, CFRDM, VIT D 82. BMI 21.9, Fev1 31% DEXA scan improved despite no treatment and worsening health. Case 4: 48 yr old Male, PS, very healthy (BMI 31.5, FEV1 100% but low Vit D and DEXA −2.8 T score). Deterioration in scan at 2.5 yrs post Alendronate despite stable health. Based on available evidence, we have adopted the guidelines for detection and management of bone loss. However, given the diversity of presentation and differing results after treatment as illustrated, we conclude that detection and management of bone loss continues to be challenging.
*Vitamin D (25-OH nmols/L range;25–170)

Reference(s)

[1] CFT (2007) Bone Mineralisation Guidelines.
[2] (2005). Oral bisphosphonates improve BMD in adults with CF IMJ 98: 270–273.

341 Evaluation of a new nutritional score in patients with cystic fibrosis

G. Alicandro[1], S. Loi[1], C. Speziali[1], L. Valmarana[1], R. Valmarana[1], L. Claut[1], N. Faelli[1], C. Colombo[1]. [1]*Department of Pediatrics, CF Center, Fondazione IRCCS Ospedale Policlinico, Mangiagalli e Regina Elena, University of Milan, Milan, Italy*

Normal growth and nutritional status are associated with better pulmonary function in CF patients. Recently a nutritional score (NS) was proposed for identification of patients at risk of malnutrition (McDonald CM, JPGN 2008), that is based on weight gain, height velocity and BMI percentile.
We have compared this NS with other nutritional indices (Height and BMI percentiles) and investigated the association of nutritional risk classification by each index with pulmonary function. We evaluated 302 patients in the age range between 2–20 years. The agreement in the nutritional categories ranking (high, moderate or low risk) of BMI percentile, height percentile and the NS was analyzed. The relative risk (RR) of having a FEV1 < 80% was determined in the group of patients defined at risk of malnutrition by each parameter.
The NS identified a higher proportion of patients with or at risk of malnutrition than height and BMI percentiles (31.5% vs 26.5% and 15.2% respectively) and showed a moderate agreement with the BMI percentile classification of risk ($k = 0.491$, $p < 0.001$). The RR of having a FEV1 < 80% was increased in patients with BMI percentile <25th (2.2 CI 1.4–3.7) and also <50th (2.3 CI 1.5–3.9). Patients with height percentile <25th and patients identified at risk by the NS did not show a significant RR of having a FEV1 < 80%.
The examined parameters used for nutritional screening differ in nutritional risk classification. The NS and height percentile did not show a significant association with FEV1. In patients with BMI percentile <25th an increased RR of pulmonary deterioration was detected, that remains elevated in those not achieving the CFF nutritional goal (BMI percentile >50th).