Are the UK Cystic Fibrosis Trust recommendations for bone health achievable?

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Objectives: Audit of current practice at the Bristol Adult CF Centre against the 2007 UK CF Trust recommendations in regard to dual-energy X-ray absorptiometry (DEXA) scans, calcium intake, vitamin D status and bisphosphonate treatment.

Methods: A retrospective case note audit for patients that had an annual review in 2010. Data was compared with the 2007 UK CF Trust recommendations for bone health.

Results: 106 patients were invited for annual review in 2010: 93 (88%) attended. Of these 93 patients 84 (90%) had a DEXA scan within the last 3 years, 7 (8%) did not attend, 1 (1%) refused and 1 (1%) scan was not ordered. Despite measures to optimise bone health, 17 (20%) patients still met clinical criteria for bisphosphonate treatment of whom 13 (15%) were receiving treatment. In 4 patients (5%) treatment was delayed for clinical reasons. 7 patients (8%) were taking bisphosphonates that no longer met the criteria. Serum 25-OH vitamin D was measured in 86 patients (92%): 59 (69%) had a level below the recommended 75 nmol/l. All patients had been prescribed vitamin supplements appropriately. Poor adherence to vitamin therapy was identified in 11 patients (12%).

Calcium intake was assessed in 81 patients (87%): 38 (47%) had a calcium intake below the recommended 1300 mg/day even with ongoing dietician support.

Conclusion: This audit has highlighted the challenges of meeting the UK CF Trust recommendations relating to vitamin D status and calcium intake despite dietician advice and monitoring. It has also raised awareness that there are a small number of patients receiving bisphosphonate treatment that may no longer be clinically indicated.

Lactose intolerance is a risk factor for decreased bone mineral density in pancreatic insufficient cystic fibrosis children

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Several risk factors of decreased bone mineral density (BMD) in cystic fibrosis (CF) children have been identified. The aim of the study was to define other determinants of occurring abnormalities.

The study comprised 78 pancreatic insufficient CF patients, with mild/moderate lung disease, no liver involvement and diabetes mellitus, not taking systemic steroids. Nutritional status (standardized body height and weight), lumbar BMD (DEXA), pulmonary function (FEV1), serum vitamin D3, calcium intake and single nucleotide polymorphism upstream of the lactase gene was assessed in all subjects.

In subjects with −13910C/C genotype (predisposing to adult-type hypolactasia), the presence of lactose malabsorption was assessed (hydrogen-methane breath test). With a use of logistic regression it was proved that BMD was dependent upon two qualitative factors: age (p < 0.03) and body weight (p < 0.04) and one quantitative factor: lactose malabsorption (p < 0.05). Such a relationship was not documented for lactase genotype.

In conclusion, lactose malabsorption seems to be an independent risk factor for decreased BMD in CF children.

Correction of low bone mass density in patients with cystic fibrosis

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Background: Bone disease is a common complication that progresses with age, severity of lung damage and nutritional disorders in patients with cystic fibrosis (CF).

Aim: To study the effect of calcium supplements and vitamin D on low bone mass density (BMD) in patients with CF.

Materials and Methods: A group of 42 patients with CF was examined to measure low BMD status and the effects of therapy. Chest deformation was found in 1/3 of schoolchildren with CF and in two toddlers with a very severe disease evolution. Children with CF were divided into two groups: first group – 26 children younger than 12 years, and the second – 21 children older than 12 years. Calcium supplements with vitamin D were given to correct the low values of BMD in children with CF. This study lasted for 3 months. The control group consisted of 21 children with CF (8 children <12 years and 13 children >12 years) with no calcium supplements for the period of this study. Quantitative ultrasound was performed (Omnisense 7000P) and Z-score of BMD was assessed.

Results: Initial Z-score showed decreased values in both study groups: −2.83 ± 0.44 SD in CF children <12 years and −3.61 ± 0.50 SD in CF patients >12 years. The level of initial Z-score in the control group were −2.73 ± 0.51 SD (CF children <12 years) and −3.82 ± 0.62 SD (CF children >12 years). Three months of treatment with calcium supplement produced an increase of Z-score to −2.44 ± 0.47 SD in children <12 years and to −2.85 ± 0.49 SD in older patients comparative to the control groups: −2.77 ± 0.38 SD (p < 0.05) and −3.21 ± 0.52 SD (p < 0.01).

Conclusion: Calcium supplements with vitamin D in CF patients have a beneficial influence on BMD Z-score.

Bone mineral density measurement in cystic fibrosis patients with CFTR I1234V mutation in a large kindred family in Qatar

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Objective: To determine the spectrum of skeletal and prevalence of CF-related low bone mineral density (BMD) in children and adults with CFTR I1234V mutation in a large Arab kindred family in Qatar.

Methodology: Twenty-six CF patients aged more than 10 years who attended the CF clinic at Hamad Medical Corporation between November 2009 till April 2010. Lumbar spine, total hip, and whole-body bone mineral density were measured by DEXA scan. Serum Ca, phosphorus, alk. phosphatase, 25-OHD vitamin D, and vitamin K were measured. Age, gender, BMI, FEV1 and bacterial colonization were correlated with BMD. The Z score was used for those who were less than 21 years and T score was used in those who were 21 year or older.

Results: Mean age of 17.29±4.95(SE) years, range 10–33 years. All the patients were pancreatic sufficient.

Mean BMD Z scores for patients 20 years or younger were −0.69±1.96 (L1-L4), −0.49±1.92 (total hip), and −0.38±0.86 (total body). Mean BMD T scores for patients 21 years or older were 0.14±1.13 (L1-L4), 0.38±1.01 (total hip) and 0.52±1.03 (total body). Seven CF patients had low BMD less than −1 giving the prevalence of CF-related low BMD of 26.9%. BMI correlated with BMD lumbar and total body Z or T scores but no significant correlation with hip Z or T score. Vitamin D deficiency was found in 20 CF patients (76.9%). BMD was not significantly correlated with gender, Pseudomonas aeruginosa and FEV1.

Conclusion: CF-related low BMD does occur in patients with mild CFTR mutation associated with pancreatic sufficient. A larger study in a cohort of CF patients in the Arabian gulf region is required to determine the risk factors of CF-related bone disease.