Prevalence and treatment of reduced BMD in a cystic fibrosis cohort undergoing lung transplantation

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Background: As life expectancy in cystic fibrosis (CF) increases, long term sequelae and their treatment assume greater importance. Osteoporosis is a prime example.

Objectives: To examine the prevalence and treatment of reduced bone mineral density (BMD) in CF patients referred for lung transplantation as a basis for implementing improved clinical practice.

Methods: Single centre retrospective analysis of bone health and its management in CF patients who underwent lung transplantation 01/2008–07/2012. We measured BMD via dual-energy X-ray absorptiometry (DEXA) scan and examined multiple risk factors for reduced BMD.

Results: 41 patients with CF underwent primary lung transplantation, comprising 63% females. Mean age at transplant was 27 years (95% CI 24–30), mean BMI 18.35 (95% CI 17.56–19.13). 5/41 patients (12%) had a documented history of fracture. DEXA was undertaken in 39/41 (95%) of patients prior to transplantation, with t-scores reported in 27/41 patients. The mean t-score at L2-L4 was −1.4 (95% CI −1.0 to −1.8); and at the femoral neck −1.1 (95% CI −0.7 to −1.5). 13/27 (48%) patients met WHO criteria for osteoporosis and 6/27 (22%) for osteoporosis. There was significant variability in the measurement and treatment of risk factors.

Conclusions: Clinically significant reduced BMD is frequent in CF patients referred for consideration of lung transplantation. Treatment of reduced BMD and its risk factors was variable, prompting implementation of a number of clinical practice improvement measures including pre-emptive management prior to referral.

Cystic fibrosis related osteopenia: What is the etiology?

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Objectives: The increase in life expectancy of cystic fibrosis (CF) patients has raised new clinical problems like osteopenia and related fractures. The etiology of osteopenia in CF is multi-factorial. Here, we aim to search osteopenia and vitamin D deficiency frequency in children with CF.

Methods: Bone mineralization was assessed using a dual energy X-ray absorptiometry (DEXA) scan. Areal bone mineral density (BMD) values were determined as g/cm² and Z-score. Vitamin D level was assessed by plasma 25(OH)D3. 25(OH)D3 level <10 ng/ml and 10–25 ng/ml was defined as severe and mild to moderate deficiency respectively.

Results: 24 children (50% female) were included. Mean age was 9.1±4.5 years. 20 (83%) patients were receiving vitamin D at 400–800 IU doses/day. The mean BMD and Z-score were 0.87 and −0.81±1.1 respectively. Eleven patients (46%) had decreased BMD values (Z-score <−1). The mean vitamin D level was 24.6±2.2 ng/ml and it was low in 58% (severe in 5%, mild to moderate in 53%) of patients. BMD Z-score values were lower in patients with bacterial colonisation with pseudomonas (p=0.019) and 25(OH)D3 levels were lower in patients with pancreas insufficiency (p=0.03). No correlation was detected between vitamin D levels and BMD Z-scores.

Conclusion: We conclude that osteopenia is a common problem in children with CF and the infection and inflammation are the major etiologic factors, other than the vitamin D levels in CF related osteopenia. However, the vitamin D deficiency is very common in CF, even they are supplemented with vitamin D. Optimization of vitamin D supplementation doses is required.

Bone turnover in patients with cystic fibrosis

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Osteoporosis is frequent problem for adult patients with cystic fibrosis (CF). But more attention should be given to the effects of the disease on bone turnover and bone mineral status especially in young patients.

Aim: To determine bone mineral status and markers of bone turnover in CF patients.

Methods: The study recruited 70 CF patients (range 5–32 y), 30 female, divided in 3 groups: prepubertal, pubertal and adults. Osteocalcin, [c]crosclips, PTH, 25(OH)D, calcium, phosphorus and alkaline phosphatase in serum were investigated.

Results: Prepubertal group included 37 patients (range 5–11.99 y). Mean value for osteocalcin was 81.06±21.1 ng/ml, for [c]crosclips 1.33±0.3 ng/ml, 25(OH)D was 25.56±12.1 ng/ml and PTH was 31.7±12.3 pg/ml. Calcium was 2.4±1.2 mmol/l and alkaline phosphatase 240±12.3 IU. Pubertal group included 17 CF patients (range 12–18 y). Mean value for osteocalcin in this group was 77.03±11.2 ng/ml, for [c]crosclips 1.42±0.5 ng/ml, for 25(OH)D 22.07±10 ng/ml and for PTH 40.6±11.3 pg/ml. Calcium was 2.3±1.3 mmol/l and alkaline phosphatase 230±13.7 IU. The group with adult CF patients (range 19–32 y) has osteocalcin 27.66±12.1 ng/ml, [c]crosclips 58.0±3.9 ng/ml, 25(OH)D 20.4±13 ng/ml and PTH 65.9±19.3 pg/ml. Calcium was 2.4±1.1 mmol/l and alkaline phosphatase 240±11.6 IU. There was no difference in bone mineral status or for levels of vitamin D in all three groups. Levels of markers for bone turnover in bone turnover in serum were elevated in prepubertal and pubertal children with CF.

Conclusion: There is a possibility of a very early onset of defective bone mineralization in CF independent of severe inflammation and nutritional status.

Renal function impairment in cystic fibrosis

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In Cystic Fibrosis (CF) renal function impairment is of major concern.

Objectives: To ascertain the prevalence of impaired renal function (GFR <90 ml/min) and associated risk factors in patients (pts) followed in our specialized CF centre.

Methods: Renal function, demographics, genotype, FEV1, pancreatic status, BMI, diabetes (CFRD), airways bacterial colonization, inhaled and iv aminoglycoside use were registered. Patients were grouped according to their GFR: group A (<90 ml/min) and group B (≥90 ml/min).

Results: 33 patients were studied, group A (13; 45%) and group B (20; 55%).

Proteinuria: 65.2±47.7 vs 28.8±28.3 mg/dL. Serum creatinine: 0.7±0.2 vs 0.8±0.2 mg/dL. Cystatin C 0.61±0.06 vs 0.62±0.86 mg/L. Age: 30.6±6.7 vs 24±5.4 yrs. Time since diagnosis: 15.1±12.8 vs 16.6±6.4 yrs. DelF508 mutation identified 11 vs 17 pts. FEV1 pred was 47.6±19.5 vs 56.4±27.0%. Pseudomonas aeruginosa chronic airways colonization: 10 vs 13 pts. Pancreatic insufficiency: 9 vs 13. BMI was 20.7±3.0 vs 19.6±3.2 kg/m². Cumulative iv aminoglycoside therapy: 26.3±9 vs 76.4±148 days. Inhaled aminoglycosides 8 vs 14 pts.

Conclusion: Patients with GFR impairment were older (p=0.02, r2=−0.41), had more time elapsed since diagnosis and worst disease severity. Creatinine clearance was negatively correlated (p=0.002, r2=−0.55) with proteinuria. GFR impairment seems to be irrespective of genotype, airways colonization, CFRD, BMI, antibiotic use, serial biomarkers.

Serial measurements of GFR and proteinuria are mandatory because GFR impairment may ensue although serum creatinine remains normal.