Bone loss in CF:A fragmented picture

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INTRODUCTION

Bone loss is highly prevalent in our CF population (70%) and is associated with risk factors: age, low vitamin D levels, low BMI and CFRDM.[1] The evidence that low BMD is a predictor of fracture risk is inconclusive and whilst there are some data to suggest that oral bisphosphonates [2,3] increase BMD, what remains unclear is the clinical significance of bone loss we have documented is in our centre. We hypothesise that bone loss occurs in patients with risk factors described above, and that treatment using the CF Trust Bone Mineralisation Guidelines will result in improvement in BMD. However, we present 4 case studies which show that not all patients "fit this picture" and that treatment does not always result in the predicted outcome.

Case study 1

- o Male aged 37: DF 508 Homozygote, PI, FEV_1 17%, BMI 20.4, CFRDM (on insulin), Vitamin D*23nmol/L, frequent exacerbations, referred for lung transplant.

- o 1st DEXA: Z Scores -3.9 (LS), -2.4 (H) Treated with Alendronate 70mg orally weekly.

- o Rpt DEXA @ 2yrs Z Scores -2.7 (LS) -2.2 (H), (FEV1 21% BMI 25.9, Vitamin D 53nmol/L)

- o Rpt DEXA @ 4yrs Z Scores -2.6 (LS), -2.0 (H) (FEV1 16%, BMI 24.9, Vitamin D 98nmol/L)

- o Rpt DEXA @ 5.5yrs Z Scores -3.9 (LS), -1.9 (H), (FEV15%, BMI 24.8 Vitamin D 41nmol/L)

Summary *Despite treatment for 5 yrs, no improvement in bone loss, however overall decline in health*

Case study 2

- o Female aged 43: DF508/unknown, PI, FEV_1 25%, BMI 17, CFRDM (no insulin) Vitamin D 68nmol/L, frequent exacerbations, NG feeding

- o 1st DEXA: Z Scores -0.4 (LS), -0.5 (H)

- o No treatment (died 6 months post scan)

Summary *No bone loss in patient with severe CF despite multiple risk factors.*

Case study 3

- o Male aged 39: DF508 Homozygote, PI, FEV_1 31%, BMI 21.9, CRDM (on insulin), Vitamin D 82nmols/L

- o 1st DEXA: Z Scores -0.3(LS), -0.7 (H)- No treatment

- o Rpt DEXA @3yrs Z Scores -0.3 (LS), -0.6(H) FEV1 31%, BM1 22.2, Vitamin D 53nmols/L

- o now frequent exacerbations, referred for transplant.

Summary *DEXA scan continues to be in normal range despite worsening health and falling Vitamin D levels.*

Case study 4

- o Male aged 48:DF508/5T-9T, PS, FEV_1 100%, BMI 31.5, Vitamin D 25nmols/L,

- o 1st DEXA: Z Scores -2.5(LS), -1.3(H) Treated with Alendronate 70mg orally weekly

- o Rpt DEXA @ 2.5yrs Z Scores -2.7 (LS), -1.0 (H), (FEV1 101%, BM1 32.4,Vit D 30nmols/L)

Summary *Deterioration in DEXA scan at 2.5yrs post Alendronate treatent, despite stable health in well patient with mild CF.*

*Vitamin D measured as 25-OH Vitamin D (nmols/L range;25-170) LS=Lumbar Spine H=hip

CONCLUSION

These 4 case studies illustrate the diversity of presentation and variability in outcomes with or without treatment for bone loss. Adherence to therapy and individual differences in response to treatment are hard to predict. We conclude that detection and management of bone loss in CF continues to be challenging. Individual treatment decisions should be based on individual measurements, as protocol-based decisions do not suit the circumstances of all individuals.

References
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