240 Effect of exercise capacity on bone mineral density in cystic fibrosis

E. Hatziagorou[1], K. Kontouli[1], V. Avramidou[1], F. Kirvasillis[1], E. Kazantzidou[2], J.N. Tsanakas[1]. [1]*Hippokration Hospital, Aristotle University of Thessaloniki, Paediatric Pulmonology Unit, 3rd Paediatric Dept, Thessaloniki, Greece;* [2]*Hippokration Hospital, Radiology Dept, Thessaloniki, Greece*

Aims:

1. To assess bone status in children and adolescents with CF.
2. To correlate Bone Mineral density (BMD) with exercise capacity, FEV_1, physical activity level, disease severity and BMI among CF childhood population.

Methods: Thirty-six patients with CF (mean age: 14.9, mean FEV_1: 81%), participated in the study. All patients had DEXA scan at lumbar spine vertebrae (L2–L4). Results were expressed as Z-scores compared with sex- and age-matched population. Questionnaires were filled in, regarding physical activity levels. All patients performed spirometry and maximal incremental cardiopulmonary exercise testing using a cycle ergometer (10 watt ramp protocol) (Ergoline 200P, Sensor Medics). Differences in disease severity (Shwachman–Kulczycki score, exercise capacity, physical activity level, body mass index [BMI]) were correlated with BMD Z-scores.

Results: The mean BMD values, serum calcium, phosphorus and parathyroid hormone levels were within normal range. BMD Z-scores were 0.34±0.9 (mean±SD). Based on a lumbar spine z-score by DEXA, 8 (22%) of the patients had osteopenia, ($-2.5 <$ z-score < -1) and 2 (5%) had osteoporosis (z-score $\leqslant -2.5$). There was evidence of mild exercise limitation during the cardiopulmonary exercise test, with mean Peak Aerobic Capacity (V′Opeak) 68.77±21.6% predicted. Multiple linear regression identified exercise capacity [VOpeak, VD/VT (ratio of physiologic dead space to tidal volume)], Shwachman score and BMI as significant predictors of BMD ($p = 0.004$, $p = 0.03$, $p = 0.03$, $p = 0.04$ respectively).

Conclusion: Limited exercise capacity, reduced BMI and disease severity are predictors of low BMD among children and adolescents with CF.

241 Iatrogenic suppression of the hypothalamic–pituitary–adrenal axis in cystic fibrosis; a cause for concern

F.J. Gilchrist[1], K.J. Cox[1], R. Rowe[1], A.K. Webb[1], A. Jones[1], R.J. Bright-Thomas[1]. [1]*Manchester Adult Cystic Fibrosis Unit, Manchester, United Kingdom*

Introduction: There are reports of hypothalamic–pituitary–adrenal (HPA) axis suppression caused by inhaled corticosteroids (ICS) when combined with itraconazole. It is caused by itraconazole inhibiting the cytochrome P450 dependent CYP3A4 pathway involved in metabolising some synthetic glucocorticoids. This increases steroid levels which negatively feedback on the HPA axis.

Methods: We performed synacthen tests in 12 patients on itraconazole and ICS and 8 patients on ICS but not itraconazole. We excluded patients on any exogenous source of steroids other than ICS. Itraconazole levels were also taken.

Results: 7/8 in the ICS group and 10/12 in the ICS/itraconazole group were on fluticasone. The median dose of fluticasone equivalent was 500 mcg/day in both groups. All patients on ICS alone had a normal synacthen test. In the ICS/itraconazole group only one patient reached a peak cortisol of 570 nmol/L and 2 had a rise of >200 nmol/L, no patient met both criteria. Five patients had severe HPA axis suppression (basal cortisol <75 nmol/L and peak cortisol of <250 nmol/L). The degree of suppression was not proportional to the itraconazole level.

Table: Synacthen test results

	Cortisol level [nmol/L], median (range)		
	Basal	30 min	Rise
ICS & itraconazole (n = 12)	171 (22–414)	373 (59–574)	179 (37–236)
ICS alone (n = 8)	352 (41–518)	674 (571–904)	383 (210–449)

Conclusions: All patients on ICS and itraconazole had HPA axis suppression and in some this was severe. In contrast, all those on ICS alone were adrenal sufficient. Therefore, in patients with CF, treatment with a combination of ICS and itraconazole should be avoided. If this is not possible, the dose of ICS should be reduced and the degree of suppression monitored with a synacthen test.

242 Are we missing salt depletion in cystic fibrosis?

S. Sharp[1], C. Marks[1], K. Chetty[2], L. Jason[2], P. Seddon[2]. [1]*Brighton and Sussex Medical School, Child Health, Brighton, United Kingdom;* [2]*Royal Alexandra Children's Hospital, Brighton, United Kingdom*

Objectives: Salt depletion was the clinical feature which first differentiated cystic fibrosis from coeliac disease, but in temperate climates it has not been considered a common problem. Most guidelines do not suggest routine sodium supplementation, few clinics screen for sodium depletion outside infancy, and there is no agreement on the best screening measure. Public health initiatives in the general population are leading to lower salt intakes. We planned to determine the prevalence of sodium depletion in our clinic, as measured by three screening methods, and to compare the sensitivity of these methods.

Methods: Urine samples were taken at annual review from 28 children (mean age 9 years). Urinary sodium (UNa), urinary sodium:creatinine ratio (UNaCr) and urinary sodium:potassium ratio (UNaK) were measured. If blood electrolytes were available, fractional sodium excretion (FeNa) was also calculated. Sodium depletion was defined by UNa <20 mmol/L, UNaCr <17, UNaK <2 and FeNa <0.5%.

Results: Using these criteria, sodium depletion was detected by UNa in only 4% (1 child), but by UNaCr in 88%, UNaK in 82%, and FeNa in 77%. FeNa correlated strongly with UNaCr and UNaK, but not with UNa. Sodium depletion was not limited to infants and preschool children. None of the children had clinical symptoms suggestive of sodium depletion.

Conclusions: Our results suggest that subclinical sodium depletion may be common across the age range in children with CF, and that simple urinary sodium concentration is an insensitive screening test. Further work is needed to test the clinical implication of our findings, and the accepted thresholds for diagnosing sodium depletion.

243 "My child tastes salty": children's and parents' understanding of salt losses in cystic fibrosis

C. Marks[1], S. Sharp[1], K. Chetty[2], C. Warde[2], P. Seddon[2]. [1]*Brighton and Sussex Medical School, Child Health, Brighton, United Kingdom;* [2]*Royal Alexandra Children's Hospital, Brighton, United Kingdom*

Background: Sweat salt losses are an intrinsic feature of cystic fibrosis (CF), especially with high environmental temperature, exertion or intercurrent illness. Despite this, guidance regarding when and how to supplement salt is often patchy and inconsistent. We explored children's and parents understanding of salt losses in CF and the actions they take to prevent sodium depletion.

Method: Questionnaires were given to 23 parents of children attending a paediatric CF clinic, and to their children if aged ⩾11 years (n = 10) to assess knowledge of salt losses, scenarios in which salt intake should be increased and preferred methods of salt supplementation.

Results: Responses were received from 100% of parents and 90% of children. 53% of respondents were aware that children with CF lose more salt than those without. 74% of parents and 89% of children would increase salt intake if on holiday in a hot climate, however only 36% of parents and 13% of children would increase salt intake if sweating with exercise. Given choices for salt supplementation, among parents 22% chose table salt, 52% salt tablets and 26% salt solution; among children 22% chose table salt, 56% tablets and 0% salt solution. 91% of respondents would like more information about salt in CF.

Conclusions: Our findings show a lack of understanding of the importance of adequate salt intake in children with CF and their parents, especially when exercising. Education on salt intake is needed, particularly in the light of general health messages promoting low salt intake.