328* Multicentre randomised double blind placebo controlled trial assessing the effect of weekly risedronate on bone mineral density in adults with cystic fibrosis

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Low bone mineral density (BMD) is prevalent in adults with cystic fibrosis (CF). The aim of this study was to assess the efficacy, tolerability and safety of risedronate in adults with CF and low BMD.

Patients with a lumbar spine (LS) or total hip (TH) BMD Z-score of −1 or less were randomised to receive risedronate 35 mg weekly or identical placebo, and calcium (1 g) + vitamin D_3 (800 iu), in addition to their standard multivitamin supplements. The primary endpoint was the treatment difference in LS BMD at 24 months.

At baseline in the risedronate (n = 17) vs placebo (n = 19) groups, the mean(SD) age was 30.2(12.0) vs 27.8(8.0) years (NS), FEV_1 56.0(22.2) vs 53.5(28.5) % predicted (NS), BMI 22.5(3.2) vs 21.2(3.1) kg/m^2 (NS), serum 25-OHD 15.3(6.8) vs 15.7(9.9) ng/ml (NS), serum PTH 32.2(19.6) vs 31.1(20.7) pg/ml (NS), LS BMD Z-score −1.42(0.53) vs −1.60(0.48) (NS), and TH BMD Z-score −1.08(0.58) vs −0.78(0.65) (NS), respectively. By 24 months 9/17 risedronate patients remained on study medication (7 patients stopped study medication due to bone pain and 1 died) and 12/19 patients in the placebo group (3 patients withdrew consent and 4 died). After 24 months treatment, the mean(SD) change in LS and TH BMD in the risedronate vs placebo groups (intention to treat analysis) was +3.4(5.0) vs +0.6(3.5)% [p = 0.05] and +1.8(5.3) vs −1.8(3.4)% [p = 0.08].

In conclusion, after two years treatment there was a significant increase in lumbar spine BMD with weekly risedronate compared to placebo. Strategies to reduce the incidence of bone pain following bisphosphonate treatment need to be evaluated in adults with CF.

329 Capillary blood gas sampling at annual assessment − a service review

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Introduction: Following acquisition of appropriate equipment, and relevant training, capillary blood gas sampling (arterialized ear lobe sampling) was performed on clinically stable patients with cystic fibrosis (CF) at the time annual assessment, The aim of this study was to assess the usefulness of this test, and to determine whether we should continue to perform them routinely.

Method: Over 36 months, 173 capillary blood gas samples were undertaken in 148 adult patients with CF. Results were retrospectively collated onto a database and categorized by a respiratory physician and physiotherapist according to normal reference ranges. The diagnosis of acid base disorder was then confirmed using the Siggaard-Anderson acid base chart.

Results: See the table.

	Number	%
Normal	23	13
Hypoxia alone	72	42
all	123	71
Respiratory alkalosis	22	13
compensated	4	2
Metabolic acidosis	11	6
compensated	3	2
Metabolic alkalosis	2	1
compensated	12	7

Discussion: Capillary blood gas sampling has previously been shown to be comparable to arterial sampling as long as a good technique is used. We were surprised by the high prevalence of abnormal results in clinically stable patients. Further work needs to be undertaken to better understand the causes, and implications of abnormal metabolic results in patients with CF.

330 Nutritional status of infants diagnosed with cystic fibrosis by newborn screening

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Background: NBS for CF is relatively new in the UK compared to Australia. Few studies have determined the nutritional status at diagnosis by NBS. The aim was to assess nutritional status at diagnosis by NBS and again at 3 and 12 months of age.

Methods: All infants diagnosed with CF by NBS or with MI, between 2002–2007, at the Royal Children's Hospital, Melbourne, were included. Anthropometric indices at birth, diagnosis, 3, and 12 months were evaluated and levels of fat soluble vitamins A, D and E, at diagnosis and 12 months. Medications were determined by chart review.

Results: 56 infants were included: NBS 44 (79%) or MI 12 (21%). Mean age at diagnosis by NBS was 5.2 weeks. 47 (84%) infants were PI. At diagnosis, 9 (23%) infants diagnosed by NBS and 2 (17%) MI infants had maintained their birth weight percentile. At 3 months 19 (43%) in the NBS group and 2 (17%) in the MI group had regained their birth weight percentile. By 12 months 27 (61%) NBS infants and 5 (42%) MI infants regained their birth weight percentile. 44 (79%) infants were deficient in one of the fat soluble vitamins at diagnosis, vitamin A 23 (44%), vitamin D 32 (67%), vitamin E 6 (12%). At 12 months, 9 (16%) were deficient, vitamin A 1 (2%), vitamin D 9 (20%) and vitamin E 4 (9%). All PI infants, and 2 PS infants had pancreatic enzyme replacement; 18 (32%) had acid suppressants; 51 (91%) used salt replacement; 29 (52%) commenced vitamin supplementation at diagnosis and continued for 12 months.

Conclusion: Nutritional deficiencies are common at the time of diagnosis by NBS. Nutritional parameters are significantly improved around 12 months of age for most babies diagnosed by NBS, but less often for babies with MI.

331 Birth weight, feeding practices and pancreatic enzyme replacement therapy (PERT) in infants diagnosed with cystic fibrosis by newborn screening − 10 years experience

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Good nutrition is an essential part of CF care and is associated with improved outcome. Newborn screening (NS) allows the opportunity to prevent nutritional problems. National NS was introduced in the UK during 2007/8. Infants born at our hospital have been screened for 30 years, 15 years using an IRT/DNA/IRT protocol.

Methods: Feeding and PERT practices of 56 infants diagnosed by NS, between January 2000 and December 2009 were prospectively recorded. Pancreatic insufficiency (PI) was confirmed by faecal pancreatic elastase (FPE-1). Birth weight was taken from newborn records. The incidence of symptomatic gastro-oesophageal reflux (GOR) was documented.

Results: Of 56 infants (36 M), 14 (25%) presented with meconium ileus (MI), 11 required surgery, 6 (11%) are pancreatic sufficient (PS). 38 (68%) are homozygous and 12 (21%) heterozygous (HZ) DF508, 4 are G551D HZ, 3 are R117H 7T HZ (all PS) and 8 have other or no identifiable mutations. Median (range) age at diagnosis (non MI infants) was 3.7 wks (0.9–7.3). Mean birth wt (range) of term infants was M 3.1 kg (1.9–4.1), F 3.3 kg (2.8–4.4). 52 infants started PERT at diagnosis, 2 stopped following increasing FPE-1 values. At 3 mths the median (range) intake of lipase/kg/day was 6431 IU (1096–19,500). 24 (43%) infants had symptomatic GOR and were treated with thickened feeds and/or medical management. 19 were initially breast fed, 5 stopped at diagnosis, 7 continued to 1 yr. 8 MI infants were given a hydrolysed formula and 14 infants received a high energy formula, concentrated formula or feed supplementation.

Discussion: Feeding and PERT requirements of NS infants are varied. Individualised advice is an essential component of care.