Bone

G147 RANDOMISED TRIAL OF PHYSICAL ACTIVITY INTERVENTION TO IMPROVE BONE HEALTH OF PRETERM INFANTS IN THE NEONATAL UNIT: RESULTS FROM THE GLASGOW WOMEN & INFANTS' SKELETAL HEALTH (WISH) STUDY

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Introduction: Immobility in preterm infants may partly contribute to osteopenia of prematurity. The role of passive physical exercise in improving bone health in these infants is unclear and requires further study.
Aim: To assess the effect of a physical activity intervention in preterm infants on bone health as assessed by quantitative ultrasound.
Methods: 31 infants born at <33 weeks gestation were randomised to receive a range of motion flexion and extension exercises once daily for 5 days each week starting "early" (n = 15) or "late" (when on 100 kcal/kg per day enteral feeds, n = 16) and continuing until term-corrected gestational age (CGA) or discharge from hospital. All outcomes were compared between groups and with a group of matched historical controls. Tibial quantitative ultrasound measurement of speed of sound (SOS) was performed using the Sunlight Omnisense 7000P scanner.
Results: Tibial SOS showed a significant decline from birth to end of physical activity programme in both "early" and "late" groups with a median change in SOS SDS –1.1 (10th and 90th centiles, −2.9, −0.4) $p<0.005$ and −0.8 (10th and 90th centiles, −3.8, −0.25) respectively, $p<0.005$. Mean SOS SDS decrease in the historical controls was −1.58 (10th and 90th centiles, −2.2, −0.85) and this was not significantly different from the intervention groups. Weight gain and head growth until end of physical activity programme did not show a significant difference between the two study groups or between study infants and controls. No infant was reported to have sustained a fracture. Length of hospital stay was not significantly different between study groups or between study infants and controls. There was no significant increase in sepsis rate, retinopathy of prematurity, or chronic lung disease in study infants. At age 5 months CGA (n = 25), no study infant had a skeletal deformity or fracture, and the rate of adverse neurodevelopmental outcomes was not increased.
Conclusions: The physical activity intervention employed in this study did not confer any benefits on the bone health outcome measure studied and was not associated with any adverse effects.

G148 REDUCED SPINAL BONE MINERAL DENSITY IN PREPUBERTAL CHILDREN WITH NEUROFIBROMATOSIS TYPE 1

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Background: Scoliosis is a common skeletal problem affecting 10–30% of patients with neurofibromatosis type 1 (NF1). It consists of a smooth curve, as in idiopathic scoliosis, or a sharp angled rapidly progressive curve involving ⩽5 vertebrae (dystrophic scoliosis). Recently, patients with NF1 have been shown to have reduced bone mineral density (BMD), which may play a role in the pathogenesis or progression of scoliosis. Our centre is one of four international centres currently evaluating the efficacy of various spinal imaging techniques and BMD as predictors for scoliosis in NF1. In our cohort we hypothesised that lumbar spine (LS) BMD would be reduced in prepubertal children with NF1 with a greater reduction in those with scoliosis or vertebral abnormalities.

Abstract G148 Lumbar spine BMAD and TBMD in neurofibromatosis type 1 children with and without scoliosis or vertebral abnormalities

	No	LS BMAD (g/cm³)	LS TBMD (mg/cm³)
Scoliosis	4	0.16 ± 0.01	± 10
No scoliosis	19	0.17 ± 0.02	148.7 ± 18
		p = 0.41	p = 0.04
Vertebral abnormalities	12	0.16 ± 0.01	135.8 ± 19
No vertebral abnormalities	11	0.19 ± 0.02	153.9 ± 15
		p = 0.01	p = 0.02

BMAD, bone mineral apparent density; LS, lumbar spine; TBMD, trabecular bone mineral density.

Methods: Clinical examination, spinal *x*-ray and magnetic resonance imaging of the spine was undertaken in 23 children (mean age 7.9 ± 1.1 years; 11 boys) with NF1 to detect scoliosis and vertebral abnormalities. BMD of L1–L4 was measured by dual-energy *x*-ray absorptiometry; data were expressed as bone mineral apparent density (BMAD; g/cm³) and values transformed to Z scores using previously published normative data.[1] Volumetric trabecular BMD (TBMD; mg/cm³) of L1–L3 was measured using quantitative computed tomography; values transformed to Z scores using Mindways software (Austin, Texas, USA).
Results: Four children had scoliosis (defined as curve of >10°) and 12 had vertebral abnormalities (loss of vertebral height, scalloping and abnormalities of pedicles or transverse spinous process). Mean Z score of LS BMAD (−0.62; $p<0.05$) and TBMD (−0.90; $p<0.001$) were lower than the zero.
Conclusions: Children with NF1 had reduced LS BMD, which was more marked in the trabecular compartment. This reduction was significantly lower in those with scoliosis or vertebral abnormalities. The ongoing longitudinal study will determine if this reduction is related to progression of scoliosis.

1. *ADC* 2007;**92**:53–9.

G149 SKIN AND BONES

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Introduction: An association between linear sebaceous naevus and hypophosphataemic rickets has been recognised for many years. We present a child with a large melanocytic naevus who developed hypophosphataemic rickets.
Case report: An 11-month-old Asian male infant was referred to the metabolic bone clinic with rickets that had failed to respond to vitamin D treatment. He was born with a giant congenital melanocytic naevus (covering 70% of his body) for which he had received extensive plastic surgery in the neonatal period. At the age of 2 months he was noted to have a low plasma phosphate (1.03 mmol/l) and a low tubular reabsorption of phosphate of 41.7% (N range >85%) for which he was started on phosphate supplements. At age 5 months, in another hospital, he was noted to have rickets for which he was treated with ergocalciferol for presumed vitamin D deficiency. He had bowed legs, swollen wrists and florid rickets on *x*-ray. His biochemistry was consistent with hypophosphataemic rickets, plasma phosphate 0.56 mmol/l, plasma calcium 2.46 mmol/l, alkaline phosphatase 4897 IU/l, parathyroid hormone 96 ng/l, $TmPO_4$/glomerular filtration rate 0.52 mmol/l (n = 1.15–2.44), normal 25OH VitD 35.3 μg/l and a low $1,25(OH)_2D_3$ of 21 pmol/l (n = 43–144). He has responded to treatment with phosphate supplements and α-calcidol. Fibroblast growth factor 23 (FGF 23) was measured and found to be elevated at 232 RU/l

(n<100). A subsequent dermatology review has identified a linear sebaceous naevus in his scalp in addition to his melanocytic naevus.
Discussion: Previous reports suggest that linear sebaceous naevi can lead to excess secretion of the peptide FGF23, which acts as a phosphaturic hormone reducing renal tubular phosphate reabsorption. It also suppresses the activity of one α-hydroxylase leading to low levels of 1,25-dihydroxyvitamin D resulting in hypophosphataemic rickets. Although this phenomenon has not previously been reported with giant congenital melanocytic naevus, our case is otherwise consistent with previous reports, and highlights this rare but important association between skin and bones.

G150 OBJECTIVE MEASUREMENT OF MUSCULOSKELETAL PAIN IN CHILDREN: DEVELOPMENT AND VALIDATION OF A TOOL

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Aims: Management of musculoskeletal disorders necessitates objective assessment of pain. Although there are many tools available in the evaluation of pain internationally, no validated musculoskeletal pain assessment tool is found. Introduction of a model methodology is needed to develop both context- and language-specific musculoskeletal pain tools. This study aims to develop and validate a musculoskeletal pain assessment tool for school-going adolescents.
Methods: The development process of the Adolescent Musculoskeletal Pain Assessment Tool (AMPAT) involved participation of an expert panel (n = 10). Musculoskeletal pain was defined. Word descriptors of musculoskeletal pain were pooled and categorised under each construct of pain experience. Specific body locations were identified. A visual analogue scale quantified the intensity of pain. A composite pain score was formulated. Face, content and consensual validities of the AMPAT were established based on the opinion of the expert panel as the gold standard. A validation study was conducted among two groups, study (with pain, n = 88) and control (no pain, n = 87), of school-going adolescents attending clinics at a larger hospital. Construct validity was appraised by comparing pain scores of the study group before and after intervention to reduce pain and comparing pain scores of the control group. Convergent validity was established by comparing pain scores of study group with AMPAT and Adolescent Paediatric Pain Tool. Discriminant validity was appraised comparing pain scores and scores of Child Medical Fear Scale of the study group.
Results: Construct validity was supported by the significant reduction in composite scores before and after the intervention. A positive linear correlation between composite scores of the AMPAT and Adolescent Paediatric Pain Tool with a Pearson correlation coefficient of 0.68 ($p<0.001$) was reported to establish convergent validity. No significant linear correlation was found between composite scores of AMPAT and Child Medical Fear Scale to support discriminant validity. Reliability analysis showed a Cronbach's α of 0.773.
Conclusions: AMPAT showed high validity and reliability to evaluate musculoskeletal pain among adolescents.

G151 SPINAL BONE MINERAL DENSITY IN CHILDREN AND ADOLESCENTS TREATED WITH CYCLICAL INTRAVENOUS PAMIDRONATE

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Background: Cyclical intravenous pamidronate is increasingly used to treat children and adolescents with fragility fractures due to primary bone disorders, such as osteogenesis imperfecta (OI) and idiopathic juvenile osteoporosis (IJO), and in those with steroid-induced osteoporosis (SIO). In children with moderate to severe forms of OI, this treatment results in a 20–40% annualised increase in lumbar spine (LS) areal bone mineral density (aBMD; g/cm^2) or bone mineral apparent density (BMAD; g/cm^3), measured by dual-energy *x* ray absorptiometry. However, there are scanty data on the effect of this treatment on LS volumetric trabecular bone mineral density (TBMD; mg/cm^3) assessed by quantitative computed tomography.
Aim and methods: The aim of this retrospective study was to compare the increment in LS BMAD and LS TBMD in eight children and adolescents with bone disorders (four with IJO, two with OI and two with SIO) treated with intravenous pamidronate (1 mg/kg, administered on 3 consecutive days, four times (cycles) a year).
Results: After 8.3±2.7 cycles of intravenous pamidronate treatment, there was a significant increase in the LS BMAD Z score (−2.7±1.0 to −0.8±1.3; p = 0.004) but not in the LS TBMD Z score (−3.8±0.7 to −4.0±0.9; p = 0. 49).
Conclusions: It is likely that the increase in LS BMAD in children and adolescents treated with cyclical intravenous pamidronate reflects retention of calcified cartilage in end plates of vertebral bodies, rather than due to an increase in the trabecular bone volume.

G152 TO TREAT OR NOT TO TREAT: MUSCULOSKELETAL PAIN AND PSYCHOLOGICAL FACTORS IN SCHOOL-GOING EARLY ADOLESCENTS

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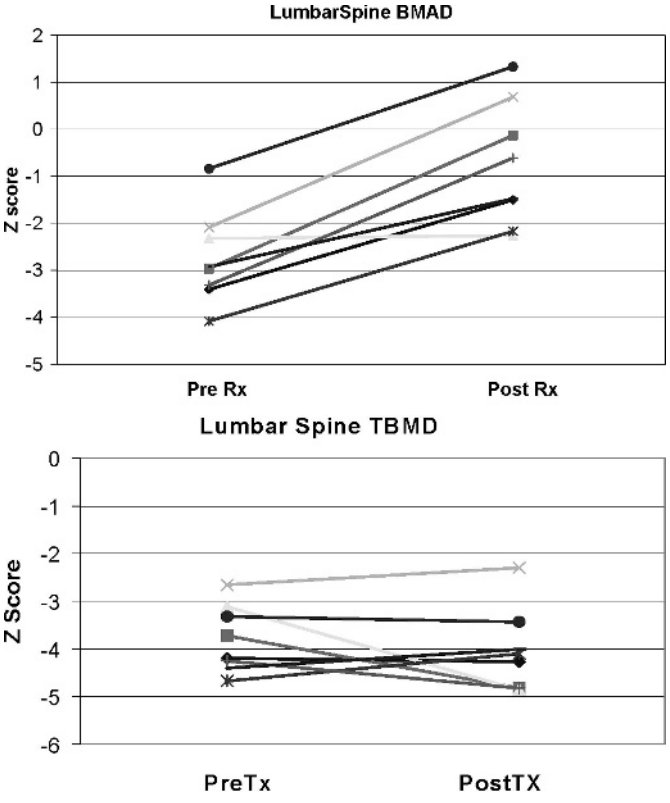
Aims: To describe the relationship between the reporting of musculoskeletal pain and psychological factors in school-going early adolescents.
Method: A sample of 1607 government school children 11–13 years of age in 55 clusters were selected using stratified multistage cluster sampling method. Musculoskeletal pain was assessed with a validated



Abstract G151 Lumbar spine BMAD and TBMD Z-scores before and after treatment with cyclical IV pamidronate. BMAD; bone mineral apparent density; TBMD, trabecular bone mineral density.

Adolescent Musculoskeletal Pain Assessment Tool. A strengths and difficulties questionnaire was administered to detect psychiatric disorder status. Knowledge on a known person with musculoskeletal pain and functional disabilities were enquired. A case–control study was done to show the relationship of recurrent musculoskeletal pain and psychological factors (cases = 463, controls = 463).

Results: 52.1% were men and 47.9% women. Anytime musculoskeletal pain prevalence was 71.2% (CI: 69.0 to 73.3%), which were: recurrent (35.9%; CI: 33.6 to 38.3%), acute (19.0%; CI: 17.2 to 21.0%) and one-time (16.2%; CI: 14.5 to 18.1%) pain. In psychiatric disorder status, 82.5% were normal and 17.5% were abnormal. Of the psychiatric problems identified, many (25.8%) tend to have abnormal peer problems and 17.3% showed conduct problems. 43% knew about a well known person suffering from musculoskeletal pain. 46% had functional disabilities 1–7 days during the last month. Abnormal psychiatric disorder status (OR = 1.72, 95% CI: 1.17 to 2.54), emotional problems (OR = 2.29, 95% CI: 1.51 to 3.50) and hyperactivity problems (OR = 1.85, 95% CI: 1.23 to 2.80) were significantly associated with higher risks of recurrent musculoskeletal pain. Presence of a functional disability (OR = 2.15, 95% CI: 1.62 to 2.85) and knowledge of a known person with a similar complaint (OR = 1.76, 95% CI: 1.33 to 2.33) were found to be posing a similar high risk.

Conclusions: The relative importance of psychosocial factors as risk factors of musculoskeletal pain highlights the need for careful assessment and management of such children. Management should be based on the category of pain and psychological status and aim at promoting healthy life-styles, thus emphasising the need for avoiding over-medicalisation.

G153 CINACALCET IN PRIMARY HYPERPARATHYROIDISM IN CHILDREN

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Introduction: Cinacalcet is a calcimimetic agent that increases the sensitivity of the calcium-sensing receptor (CaSR) to activation by extracellular calcium. This results in a reduction in parathormone (PTH) secretion with a concomitant decrease in plasma calcium. It is currently licensed for adults with secondary hyperparathyroidism due to end-stage renal disease and adults with primary hyperparathyroidism where parathyroid surgery is contraindicated. It is not currently licensed for use in children.

Methods: We report the use of cinacalcet in two children with primary hyperparathyroidism.

Case reports: A 14-year-old girl with primary hyperparathyroidism was experiencing recurrent loin pain due to renal calculi. It was not possible to localise a parathyroid adenoma and parathyroidectomy proved unsuccessful. She was treated with cinacalcet in an initial dose of 30 mg daily and subsequently 30 mg twice daily. Her plasma calcium and serum PTH fell from 3.04 mmol/l and 104 ng/l respectively to 2.56 mmol/l and 29 ng/l (NR 11–32) within 4 months of treatment. An infant who presented with hypercalcaemia (6.97 mmol/l) and elevated PTH (675 ng/l) had neonatal severe hyperparathyroidism due to a homozygous CaSR gene mutation. Despite parathyroid surgery on two occasions she continued to have a persistently elevated PTH (172 ng/l) and required a low calcium diet to prevent hypercalcaemia. Cinacalcet was commenced in an initial dose of 0.5 mg/kg per day. There was an initial response with serum PTH falling to 26 ng/l and plasma calcium falling from 3.26 mmol/l to 2.08 mmol/l within 1 month. However, this was not sustained despite increasing the dose to 1.0 mg/kg per day and cinacalcet had to be discontinued after 4 months of treatment. She required further parathyroid surgery to remove her remaining two parathyroid glands before her hypercalcaemia resolved.

Conclusions: Primary hyperparathyroidism is a rare condition in children. Although parathyroid surgery with an experienced surgeon is the management of choice this may not always be successful. In such situations it is worth considering the use of cinacalcet even if it is a short-term measure before further surgery. Additional studies of the use of cinacalcet in children are indicated.