

Method Computerised article search conducted using the electronic databases Medline, Science Direct and the Cochrane Library for the literature review. Interviews were conducted with the child's mother and community paediatrician.

Results After sleep hygiene optimisation and behavioural interventions, sleep onset latency had decreased by one hour. After melatonin, sleep-onset latency; frequency of night terrors and other nocturnal awakenings and daytime behaviour had improved. Total sleep time had increased by 4 h 30 min.

Conclusion Further research should be done to set up more sleep clinics nationwide. Official guidelines or practice pathways should be made to guide professionals in how to manage sleep problems in ASD.

G203(P) STATUS DYSTONICUS PRESENTING IN AN ACUTE SETTING IN ASSOCIATION WITH VIRAL ILLNESSES

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Aims We report 2 cases of status dystonicus seen in our hospital in 2014. Status dystonicus is a rare condition with a potentially life-threatening outcome. The patients' background, clinical features, management, likely triggers, differential diagnosis and outcome is discussed.

Methods Review of charts.

- Review of existing literature.

Description *Case1:* A 9-year-old female with dystonic quadriplegia GMFCS class IV, intellectual disability and gastrostomy feeds was admitted with a febrile illness (pneumonia) and treated with intravenous antibiotics, oxygen and fluid support. Her dystonic movements had not required treatment before this.

During the admission, she developed severe abnormal movements and unusual posturing with sustained hyperpyrexia, sweating and rising creatinine kinase (CK) > 23,000. Blood and urine cultures were negative. She was intubated and ventilated, transferred to PICU, and received chloral hydrate, clonidine, and oseltamivir. She was subsequently confirmed as having Influenza A, H3N2 strain. During recovery, sedation was gradually weaned; however, dystonic movements recurred, requiring institution of a slower weaning regimen. She went home on trihexyphenidyl and remains well.

Case2: A 5-year-old male with microcephaly, spastic quadriplegia GMFCS class V (intra-theal baclofen pump), visual impairment, profound intellectual disability, recurrent urinary tract infections (UTI), nephrocalcinosis, gastrostomy feeds was admitted with *Pseudomonas* UTI and treated with ciprofloxacin.

His recovery was complicated by norovirus gastro-enteritis with dehydration, pre-renal failure and increasingly severe dystonic posturing (tongue protrusion, sustained muscle contractions, opisthotonus) with fever, sweating and rising CK (> 23000). He was treated with: transfer to HDU, close management of fluid and electrolyte balance, sedation with chloral hydrate, clonidine and midazolam.

Resolution of the movement disorder and fever and normalisation of CK followed. He re-presented a month later with similar symptomatology; however, early treatment with hydration, clonidine and chloral hydrate appeared to halt progression to status dystonicus. He went home on low-dose clonidine and remains well.

Conclusion Status dystonicus is a rare condition with a high morbidity and mortality. A rising CK, severe dystonic movements and metabolic derangements suggest the diagnosis. Maintaining a high index of suspicion can identify such cases early and halt further progression. CK is simple test to monitor response to treatment.

G204(P) OSTEOSARCOMA CELL CULTURE ON COLLAGEN SURFACES AND IN HYPOXIA ALTERS MMP EXPRESSION

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Osteosarcoma is the most common primary malignant bone tumour in children. The survival rate has not improved much over the last 25 years, and therefore there is a lot to learn about the pathogenesis of this cancer. The interactions of tumour cells with their environment and hypoxia have been identified as key drivers of tumour growth and metastasis. Matrix-metallo proteinases (MMPs) are involved in this process. MMPs are zinc-endopeptidases that are able to degrade the extra-cellular matrix and are over-expressed in many tumours. Membrane-type (MT1)-MMP and MMP-2 expression is positively associated with tumour progression in a range of tumours, but their role is not well characterised in osteosarcoma.

Two osteosarcoma cell lines were cultured on culture plastic or collagen surfaces in either normoxia or hypoxia. Proliferation was assessed using the SRB assay which showed osteosarcoma cells proliferate slightly slower in hypoxia. Immunofluorescence microscopy was employed to visualise MT1-MMP – this revealed MT1-MMP packaging and localisation was altered in hypoxia and there was formation of invadopodia on collagen. Gelatinase expression, assessed using zymography of supernatants, demonstrated increased proMMP-2 activation by cells cultured on collagen, particularly by U2OS cells. Cell lysates were probed for MT1-MMP using western blotting. ELISA of the culture supernatant was used to measure TIMP-2 expression. Less active MT1-MMP was detected in the lysates of the U2OS cells which coincided with a decreased amount of TIMP-2 detected in the supernatant.

This study contributes to our understanding of the activation of MMPs and the possible role of MT1-MMP in this regard.

G205(P) FIBRODYSPLASIA OSSIFICANS PROGRESSIVA (FOP) AN UNFAMILIAR DISEASE THAT IS NOW IMPORTANT TO DIAGNOSE

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Background FOP is a rare but disabling condition characterised by congenital malformation of the great toes and progressive heterotopic endochondral ossification (HEO). FOP is the most catastrophic disorder of HEO in humans.

Flare-ups are episodic; immobility is cumulative. The discovery of the ACVR1 gene as the cause of FOP has allowed identification of possible therapeutic targets. Palovarotene, a retinoic acid receptor gamma agonist, is currently in Phase 2 clinical trials to reduce HEO during acute flares.

Aim To describe the clinical presentation and current management of FOP.

Methods A 7 year old boy with a history of hallux valgus, recurrent painful episodes of soft tissue swelling and new abnormal bone formation, was assessed clinically and radiologically.

Results Review of the clinical history and radiographs taken in infancy revealed the diagnosis of FOP. This had not been previously recognised, although he had been seen in a specialist Hallux Valgus clinic as a baby.

Conclusions As specific treatments are now becoming available for this life-limiting condition, it is essential that all neonatologists, paediatricians, paediatric oncologists and orthopaedic surgeons consider a diagnosis of FOP if a baby or child presents with bilateral hallux valgus and/or episodes of swelling with evidence of ossification. These children should now be referred to a paediatric metabolic bone clinic to consider genetic testing and for specialist management.

G206(P) RECOGNISING THE RISK FACTORS: MISSED OPPORTUNITIES TO PREVENT RICKETS

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Aim Implementing the NICE guidance published in November 2014 for vitamin D supplementation of at-risk groups, including all children under-five years of age, would help reduce childhood rickets due to Vitamin D deficiency. Whilst awareness of Vitamin D is increasing, we often assume that UK infants have sufficient calcium dietary intake. This case series highlights rickets arising when insufficient dietary calcium compounds the effects of insufficient Vitamin D.

Methods We reviewed case-notes, biochemical and radiological data of four children diagnosed with rickets at our hospital during the past 18 months.

Results One child presented aged seven months with seizures. She had a prolonged corrected QT interval secondary to profound hypocalcaemia (corrected calcium 1.49 mmol/L), and vitamin D Deficiency (vitamin D <6 nmol/L). The remaining three infants (aged 16–20 months) presented with bony deformities characteristic of rickets, and Vitamin D supplementation had recently been commenced in primary care. Total Vitamin D levels were 26.3, 38.9 and 51.1 nmol/L each with a significant proportion of the total Vitamin D as D2 consistent with supplementation. These three cases had normo-calcaemia (2.3–2.54 mmol/L). Radiology showed rachitic changes. All cases had pigmented skin, were predominantly breastfed and had no vitamin D supplementation during the first year of life. The clinical and radiological rachitic changes were more marked than expected for the levels of Vitamin D. All were cow's milk allergic (confirmed on specific IgE testing) and were slow to wean, with prolonged breast feeding. The children had inadvertently been on low calcium diets without supplementation. All children have made good progress with subsequent appropriate calcium and vitamin D supplementation.

Conclusion This series illustrates commonly recognised risk factors for rickets, namely skin pigmentation, exclusive and prolonged breastfeeding, and the lack of Vitamin D supplementation. Undoubtedly the lack of vitamin D was contributory to the development of rickets and appropriate supplementation

could have minimised the adverse skeletal effects. These cases also highlight that a calcium-deplete diet is an additive risk factor for the development of rickets. Infants with cow's milk protein allergy comprise a clinical group in whom it is important to consider the need for calcium supplementation alongside Vitamin D.

G207(P) TO REVIEW THE OUTCOME OF HIP SCREENING ULTRASOUND SCAN (USS) FOR DEVELOPMENTAL DYSPLASIA OF HIP (DDH) IN HIGH RISK BABIES

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Aim The aim of our study was to review the outcome of hip screening ultrasound scan in high risk babies for further evaluation of developmental dysplasia of hip in the district general hospital.

Methods This is a retrospective study. We looked at the outcome of the USS hip performed for the period of 18 months from June 2013 to October 2014 in our trust in high risk patient group with breech presentation, family history of DDH, twin deliveries with one of the twin born with breech presentation and on the clinical suspicion after clinical examination of the hip. Data was collected using hospital notes, PACS system was used for USS results.

Results In total 553 USS hip was performed for further evaluation of DDH out of which only 4 scans were abnormal. Three of this abnormal scans were in babies born with breech presentation and one was in a baby who's mother had history of DDH needed intervention in past. Out of this 4 abnormal scans 3 scans were Graf Type IIa and one was Graf type D.

Conclusion In our experience we have observed a significantly low percentage of clinically relevant USS results for DDH even in selected high risk babies.

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G208 REFLECTIONS AND NEW DIRECTIONS IN CHILDREN AND YOUNG PEOPLE'S PALLIATIVE CARE

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In June 2010, the Department of Health (DoH) in the UK released a call to apply for funding to support projects focused on benefiting the lives of children and young people with palliative and complex health care needs and their families. A programme of work was subsequently developed from 2010–2014 including an innovative e-learning programme and important projects seeking to explore issues around competence of care skills and communication challenges in the field of children and young people with palliative care.

Aims The aim in this workshop is to share reflections from a recent narrative literature review as part of a programme of work which focused on confidence and competence of care skills. Results of the review will be shared but will provide a platform for critical debate around challenges and new directions in children and young people's palliative care.