framework for long term conditions in order to improve the service delivery to children with epilepsy.

Method A questionnaire was developed using CG137epilepsy NICE guideline. Patients with epilepsy who were seen over the period of January–May 2014 were selected to receive the questionnaires that included parents feeling once the diagnosis is made. Patients were seen both in hospital and community settings.

Results 40 received the questionnaires, 34 returned (85%). Most (30/34) were younger than 10 years at the time of diagnosis. Co-morbidities were seen in 62% including cerebral palsy, learning difficulty, autism, ADHD and some had combinations. The high number relates to seeing more children in special schools over the period of the audit. 18/40 reported significant emotional problems at diagnosis: sad, terrified, devastated and shocked. Information on lifestyle including water safety was provided to 25/34 (70%). Information on SUDEP was not provided to 23/34 (68%).

Conclusion There is a wide variation in practice within the Department. There is no epilepsy clinic with no epilepsy nurse. The information provided to patients was very patchy, too little or given too late.

Action Proforma was put in place, covered classification as per Multi-axial diagnostic scheme: Description of seizures, Seizure type, Syndrome, Aetiology and Co-morbidity. Referral letter is sent to the community nursing team for early home visit and an early liaison with school to improve outcome. Group training for the parents was commenced.

G200(P) A CASE HIGHLIGHTING THE IMPACT OF UNCORRECTED SCOLIOSIS

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Method This is a case of a previously well 17 year old girl with cerebral palsy secondary to Lissencephaly. She presented with a history of intractable vomiting progressing over the course of a few months secondary to severe worsening scoliosis.

Upon further investigation, it was felt that the extensive degree of her scoliosis was rotating her abdominal anatomy in such a manner to be the main cause of her vomiting. She was reviewed by the orthopaedic team who felt that correction of her scoliosis was indicated however this was not without significant risks to the patient especially given her extensive recent weight loss secondary to the vomiting. However, without the procedure the patient would continue to vomit and losing weight and may eventually die.

Result The parents of the patient were keen to proceed with the operation despite the high risks as they felt their daughter's quality of life had deteriorated significantly since the onset of the scoliosis and recurrent vomiting.

The patient underwent the scoliosis correction and made a good recovery.

Conclusion This case presents a dilemma in which without correction of her scoliosis, the patient would continue to vomit and lose weight (medical therapy had failed). However, undertaking such a major operation in this patient carried a significant mortality risk.

The case also highlights the importance of close follow up of these patients as this child did not develop scoliosis until her teenage years and she missed her follow up appointments due to an address change.

Pictures of the scoliosis (X ray/CT are available to accompany the case)

G201(P) DO BASELINE BLOOD PRESSURE AND HEART RATE IN CHILDREN WITH A SPINAL CORD INJURY VARY DEPENDING ON THEIR LEVEL OF INJURY?

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Children with a spinal cord injury may have blood pressure and heart rate differences from the expected values of that age. Understanding this potential problem is important for optimal management.

Aims To establish any association between level of spinal injury and ASIA (American Spinal Injury Association) impairment scale severity score with heart rate, blood pressure and diurnal variation in blood pressure. The association between age, gender and BMI on heart rate and blood pressure was also explored.

Method Retrospective records were reviewed for 32 children admitted to a National Spinal Injury Centre for more than 4 rehabilitation periods between 2011 and 2013 (107 admissions). Patient and injury data were established. The first recorded morning and evening blood pressures and heart rates were collated.

Data was analysed using MS Excel 2010 and IBM SPSS v.20.

Results The mean age at first admission was 10.2 years, 41% were male, and 59% had an injury at or above T6. Only 12% of the admissions had complete cardiovascular data.

The associations between ASIA score and BP centile group; ASIA score and diurnal variation; level of injury and BP centile group; and level of injury and diurnal variation, were not statistically significant.

Increasing age was significantly associated with a lower heart rate (OR 0.094, p value <0.001) and systolic BP with increasing BMI (OR 2.97, 95% CI(1.439, 6.137)

Conclusion The changes observed with age and BMI can be accepted as normal physiological change. That no statistical association between the injury related factors, ASIA score and level of injury, and cardiovascular measures was observed could be due to the poor data quality, and no conclusion can therefore be reached from this finding.

Improved measurement and recording of height, weight and cardiovascular observations is paramount for optimal cardiovascular management in this patient group.

G202(P) SLEEP MANAGEMENT IN AUTISTIC SPECTRUM DISORDER

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Abnormal sleep affects 44–89% of children with ASD. Currently there are no official UK guidelines for the management of sleep with ASD.

Aim To discuss sleep management using a case study of 9 year old boy with ASD.

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 quadriparesis 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 quadriparesis GMFCS class V (intra-thecal baclofen pump), visual impairment, profound intellectual disability, recurrent urinary tract infections (UTI), nephcalcinosis, 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, ophisthotonus) 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.