

outcome for this teenager. Longterm surveillance for tumour detection is recommended in autoimmune encephalitis.

### G66(P) INTERESTING CASES OF PYRIDOXINE DEPENDENT EPILEPSY IN LATE CHILDHOOD

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We report cases of two children with intractable epilepsy who responded to treatment with pyridoxine in late childhood.

**Case details** Case 1: A 14 year old girl born following an uneventful pregnancy to non consanguineous parents. She has global developmental delay. She developed epilepsy at around 2 years of age. Her extensive investigations including an MRI of brain were normal. She had various types of seizures and these were refractory to antiepileptic medications. A trial of pyridoxine at around 13 years of age surprisingly had a very good effect on her seizure control. We are currently awaiting the result of urinary L- $\alpha$ -amino adipic semialdehyde (AASA).

Case 2: A 4 year old girl born following normal pregnancy developed infantile spasms at 4 months of age. She was treated with numerous courses of steroids with suboptimal response. Extended investigations including the MRI brain were normal. She eventually developed refractory epilepsy with cognitive decline. A trial of pyridoxine at 3 years of age achieved good control of epilepsy and improved her cognition as well. The urinary AASA was negative and the result of PNPO gene mutation is awaited.

**Discussion** Pyridoxine dependent epilepsy is a syndrome that usually presents with neonatal intractable seizures. It may present later in infancy or early childhood. A variety of seizure types are reported, the most common being generalised tonic clonic seizures which progress to status epilepticus. Urinary AASA is usually elevated and the genetic testing is available for this condition now. Our cases responded to the empirical trial of Pyridoxine at a much later age.

**Conclusion** We recommend testing for pyridoxine deficiency and therapeutic trial with pyridoxine in children with refractory epilepsy in all the age groups. Early detection and treatment of this condition not only achieves good seizure control but can possibly improve developmental outcome as well.

### G67(P) CHILDHOOD PARKINSONISM: A RARE COMPLICATION OF HYPOXIC BRAIN INJURY

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**Case report** Child A is a 13-year old boy with a history of poor adherence to his asthma treatment.

He was admitted to hospital with an acute severe exacerbation of asthma, which needed escalation of treatment to intravenous bronchodilators. Despite intensive treatment, he went on to have a cardio-respiratory arrest, necessitating five minutes of cardiopulmonary resuscitation and adrenaline prior to return of his circulation.

Child A spent a week on PICU, after which he underwent neurological rehabilitation on the ward, involving a multidisciplinary team. MRI of his brain confirmed severe hypoxic brain



Abstract G67(P) Figure 1

injury with infarction within the basal ganglia and occipital lobe (Figure 1). Similar to other children with acquired brain injury, he demonstrated 4-limb spasticity, as well as dystonic posturing.

In addition to this, he also displayed Parkinsonian features, which is a rare complication of brain injury. He had a resting tremor, bradykinesia, rigidity and a shuffling gait, with difficulty in turning around. He displayed hypomimic facies, along with a monotonous speech. The above symptoms responded well to co-careldopa.

**Discussion** Poor compliance with asthma medication in children remains a significant problem with major health implications. Our case sustained a severe brain injury as a result of this. Unlike in adults where Parkinsonism is common, this condition is rare in children and easy to miss. The commonest cause of Parkinsonism is the loss of dopaminergic neurones in the substantia nigra of the basal ganglia. Child A's symptoms resolved with co-careldopa which increases dopamine levels in the brain.

We have not come across a similar paediatric case during our literature review. Paediatricians need to be vigilant to identify Parkinsonian features in children with brain injury. These typical signs become apparent to the clinician who is clearly looking out for them. Making this diagnosis correctly is important so that we can prescribe a specific anti-Parkinsonian medication, rather than make the child go through repeated failed trials with incorrect drugs. There are different classes of medication for Parkinsonism and treatment choices are largely based on data from adults.

Recognising this movement disorder is hence vital to support the rehabilitation process and optimise recovery.

### G68(P) A RETROSPECTIVE REVIEW OF EPILEPSY RELATED ADMISSIONS TO THE PAEDIATRIC DEPARTMENT AT A DISTRICT GENERAL NHS TRUST, ENGLAND

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**Aims** Child and Maternal Health (ChiMat) Observatory Data indicate our institution has higher Paediatric Emergency Epilepsy admissions and length of stay, compared to other PCTs in

England. This report has been referred to by Commissioning Groups when reviewing and commissioning services.

The information produced by ChiMat is based on data from the hospital's coding department, who determine the admission reason from discharge letters completed by junior doctors.

We carried out a retrospective audit to review:

- The accuracy of coding
- Medical management of Paediatric Emergency Epilepsy admissions.

**Methods** We reviewed the medical notes of all patients coded as having a Paediatric Emergency Epilepsy admission during 2011. There were 78 patients during this period, and information was collected using a standardised proforma. Inappropriately coded patients were excluded from analysis in the second part of the audit.

**Results** 10 of the 78 patients (12.8%) were exclusively under the care of the adult physicians, with age range 16–19 years. Of the other 68 patients, 15 were incorrectly coded (22%).

Review of the medical management in the remaining 53 admissions, showed areas for improvement in medication adherence, patient education and awareness, and community management plans.

**Conclusions** This study has shown the importance of accurate data coding, as this is used to review the service we provide, highlighting exceptional practice as well as areas which require improvement. Variation in practice and value in healthcare are the current quality indicators which are used, to compare hospitals and clinicians, and to continue the quality improvement cycle. It is therefore in the interest of all to engage with clinical coding to ensure accurate, robust data is being used.

Our audit has led to more streamlined management of patients with epilepsy, including the consideration of more community-based management plans and proposal for a Paediatric Epilepsy Specialist Nurse. There has also been quality improvement effects, including introduction of a weekly epilepsy-related admissions report which is reviewed for accuracy, monthly epilepsy peer-review meeting to review all admissions and challenging cases, importance of accurate coding on discharge letters being emphasised to junior doctors at induction, and introduction of a checklist for management of patients with epilepsy.

### G69(P) NARCOLEPSY – AN IMPORTANT BUT RARE PAEDIATRIC DIAGNOSIS

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**Aim** To present patients investigated for narcolepsy in order to increase awareness of its clinical features.

**Methods** 5 patients referred with possible narcolepsy to the sleep clinic.

**Results**

1. 8 year old boy with excessive daytime sleepiness and cataplexy. HLA typing has shown HLA- DQB\*06:02. MSLT showed short sleep latency. Awaiting lumbar puncture for CSF analysis of orexin level. Commenced on methylphenidate with good response.
2. 6 year old boy with 4 year history of falling asleep quickly including very short distances in the car, at school, and before finishing meals. HLA typing is positive for DQB1\*06–02. Commenced on methylphenidate.

3. 13 year old boy with 5 year history of falling asleep several times a day often missing his bus stop and has recurrent naps in school. Commenced on methylphenidate.
4. 13 year old girl who falls asleep in lessons however related to boredom rather than irresistible desire to fall asleep. Found to be anaemic. Commenced on iron supplementation.
5. 8 year old girl initially presenting with excessive daytime sleepiness with hallucinations on sleeping and waking. She was HLA DQB\*06–02 positive suggesting narcolepsy with hypnagogic and hypnopompic hallucinations. MSLT not diagnostic of narcolepsy but had epileptiform changes on EEG. Commenced on clonazepam with improvement in hallucinations.

**Conclusion** Narcolepsy is an autoimmune neurological disorder characterised by excessive daytime sleepiness associated with cataplexy, hypnagogic/hypnopompic hallucinations and sleep paralysis. This frightening onset of symptoms often occurs in childhood or adolescence with a 10–14 year diagnostic delay. Often multiple misdiagnoses such as hypothyroidism, depression and epilepsy are made along the way. By presenting these patients, the features of narcolepsy are highlighted enabling Paediatricians to consider this as a diagnosis. Better recognition and earlier diagnosis can lead to earlier onset of therapeutic intervention thus leading to a lower impact on academic performance and social development.

### G70(P) RADIOLOGICAL INVESTIGATIONS OF MIDLINE INFANTILE HAEMANGIOMAS OVERLYING THE SCALP OR SPINE: 5 YEAR SINGLE CENTRE EXPERIENCE

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**Aim** To review the outcomes of investigations for midline infantile haemangiomas (IH) overlying the head or spine in a tertiary paediatric dermatology department.

**Methods** Case notes and radiological results were reviewed retrospectively for all patients with a clinical diagnosis of IH overlying the midline of the scalp, neck or spine seen over the 5 year period October 2009–14.

**Results** 43 patients were identified (31 girls). The median age at the first specialist clinic appointment was 5 months (range 2–23 months). In 26 patients the IH were located on the scalp, in five on the back of the neck, in four overlying the thoracic spine and in eight overlying the lumbar spine and or sacrum. 24 lesions were more than 5 cm and 19 less than 5 cm in size at the time of the first appointment, 10 of which were plaque-type IH. 18/43 patients had MRI of either the brain or spine following the initial assessment. MRI was normal in 15 patients, including in all lumbosacral lesions. In two cases of plaque-type IH a diagnosis of PHACES syndrome was confirmed. In one child where the IH was a palm-sized lesion over the thoracic spine, MRI showed extension of the IH into the spinal canal, with complete compression of the cord between T4 and T9. The child was neurologically asymptomatic at the first assessment, but the result prompted treatment with propranolol.

**Conclusion** This retrospective series of midline IH seen over a five year period in a tertiary centre has identified a significant underlying lesion in a child with a thoracic spinal IH. The current literature from experts in the field recommends MRI for lumbosacral lesions. Our internal guidelines will now dictate an