NEUROLOGY

G218 CAUDAVERIC AND HIPPOCAMPAL VOLUMES, AND BEHAVIOURAL AND COGNITIVE PERFORMANCE OF VERY PRETERM INFANTS IN EARLY ADOLESCENCE

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The caudate nuclei and the hippocampus are both involved in the functions of short-term memory and learning. The latter is particularly susceptible to damage from stress, and has been shown to be altered in size in epilepsy, Alzheimer's disease, schizophrenia and post-traumatic stress disorder. Preterm infants (PT) often show learning, motor and behavioural difficulties at school. This study aimed to determine whether structural differences shown on magnetic resonance imaging (MRI) in these areas of the brain were associated with these deficits in PT in adolescence.

94 adolescents (86 PT, 8 term) who had been assessed at school at 12-13 years for motor impairment (Movement ABC), attention deficit and hyperactivity disorder (ADHD, Child and Adolescent Psychiatric Assessment), and intelligence (IQ, Wechsler Intelligence Scale for Children III), underwent MRI of the brain at 15-17 years. Volume measurements of the caudate nuclei and hippocampi were made bilaterally for each child, and the ratio of the left to right structure calculated as a percentage. The neuroradiologist was blind to the results of earlier testing.

Caudate and hippocampal volumes and the hippocampal ratio were significantly higher in the term controls. No significant relationship of volumes or ratio to motors in therapy was observed. PT with ADHD had a significantly smaller right and left hippocampus, PT with a low IQ (<85), had a significantly smaller right caudate nucleus and left hippocampus, and a lower hippocampal ratio when compared to controls. PT with a high IQ standardising for sex and handedness using logistic regression did not alter these findings.

These volumetric differences suggest that specific structural changes during development of the brain in PT may be responsible for behavioural and learning deficits in later life.

G219 SEVERE HYPERKINETIC MOVEMENT DISORDER IN A CHILD TREATED WITH DEEP BRAIN STIMULATION: A CASE REPORT

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Introduction: Deep stimulation of brain structures has been in use for adult movement disorders since 1987 (Pollack, 1997). Its use in childhood movement disorder has not previously been reported in the literature. We would like to report its use in one such case.

Clinical history: This eleven year old male is the first child of unrelated parents. Foetal movements were reduced in the third trimester. Born at term by normal delivery. He was noted to be hypotonic in the neonatal period. Athetoid movements developed in the first year of life. Epilepsy with tonic/ clonic and partial motor seizures occurred at 18 mths. He crawled at 3 yrs, and walked with aids at 5 yrs. He developed a multi-focal, stimulant sensitive and action myoclonus with an exaggerated startle reflex which was non-progressive at 5 yrs. Detailed investigations have all been normal. Intellectual function is generally below that of his age. Management of his movement disorder has included carbamazepine, sodium valproate, haloperidol, tetrazenem, benzhexole, acetazolamide and phenytoin. A trial of clobazam precipitated status choreaticus with severe rhabdomyolysis complicated by hyperpyrexia, renal impairment and disseminated intravascular coagulation.

The failure of medical treatment led to him being referred for assessment for neurosurgical intervention. Following detailed evaluation, he underwent insertion of deep brain stimulators (Medtronic tief II IPG) into the posteroverentral globus pallidus bilaterally.

Results: The patient’s movement disorder has shown sustained improvement at 9 months follow-up. The family is now able to socialise and take trips in a way that was previously impossible. Orthopaedic intervention is now being planned as consistent, supported weight bearing by the patient is now possible. He is no longer injuring himself frequently and is starting to gain weight. Gross Motor Score, ‘posting’, and other evaluations have been performed with the stimulators in the ‘on’ and ‘off’ state. These results will be available within the next 6 weeks.

G220 CHANGING TRENDS IN THE INVESTIGATION AND MANAGEMENT OF FEBRILE CONVULSIONS IN THE FIRST YEAR OF LIFE: LOCAL EXPERIENCE AND A REVIEW OF THE LITERATURE

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The UK guidelines state that lumbar puncture should ‘almost certainly’ be performed following a first febrile convulsion in the first year of life. A retrospective audit was carried out to determine if practice has changed over the last ten years at our centre.

Methods: Patients were identified using the hospital admissions books for 1999 and 1998. The notes of all those between 6 months’ and 1 year with an admitting diagnosis of convulsion, fit or seizure were examined individually. For those who satisfied the diagnostic criteria for a febrile convulsion data were retrieved to determine the patient characteristics, seizure duration, investigations done, subsequent management and final diagnosis.

Results: 30 children between 6 months to a year of age were admitted to our centre following their first febrile convolution (17 in 1998 and 13 in 1989). The demographics of both groups were very similar. There was a significant decrease in the number of children undergoing lumbar puncture (0/17 in 1998 vs. 8/13 in 1989 (p<0.01)) and a trend towards less blood tests, less antibiotic use and shorter hospital stays in 1998.

Discussion: Systematic review of the literature demonstrates that it is extremely rare for bacterial meningitis to present as a febrile convulsion. In the absence of other physical signs (drowsiness, bulging fontanelle or rash) then the minimum incidence of bacterial meningitis is approximately 0.1% (6/5522). Furthermore, the diagnostic yield of lumbar puncture in this group is only 66% (4/6) which raises questions about the value and risks of lumbar puncture in this age group.

LUMBAR PUNCTURE FOLLOWING THE FIRST FEBRILE CONVULSION: A SURVEY OF PRACTICE IN THE UNITED KINGDOM IN THE FIRST YEAR OF LIFE

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Guidelines produced by the British Paediatric Association state that paediatricians should ‘almost certainly’ perform a lumbar puncture in children under one year, even in the absence of signs of meningitis as these may be occult. A survey of 100 paediatric registrars was performed to determine current practice across the UK.

Methods: 120 hospitals were randomly selected from the Directory of Hospital Telephone Numbers. From these, 100 paediatric registrars from hospitals in every region in the UK were contacted by telephone. Firstly they were asked if there were any guidelines concerning the management of febrile convulsions in their department. Then they were asked whether they would perform a lumbar puncture on a ten-month-old child who had had its first fifteen-minute febrile convolution. They were all told that the child had a runny nose and a temperature of 38°C but no abnormal physical signs (bulging fontanelle, drowsiness or residual focal neurological).

Results: 46 (out of 100) registrars said that they would and 54 said that they would not perform a lumbar puncture. The presence or knowledge of departmental guidelines had little effect on the likelihood of performing a lumbar puncture.

Local guidelines Lumbar puncture

| Yes 42 | Yes 21 (50%) |
| No 28 | No 20 (50%) |
| Don’t know 30 | Yes 14 (47%) |
| No 16 (53%) |

Conclusion: There is currently no consensus amongst middle grade paediatric registrars on whether lumbar puncture should be performed following a first febrile convolution in the first year of life.

G221 NORMAL DECUSCATION OF CORTICOSPINAL AXONS AND AXONAL PROJECTION TO LUMBAR SURAL SEGMENTS IN HUMAN SUBJECTS WITH L1CAM MUTATIONS

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Background: L1CAM gene (Kc28) mutations are linked to neurological syndromes characterised by spastic diplegia. The L1CAM knockout mouse shows failure of corticospinal (CST) axonal decussation and projection beyond the cervical spinal cord. Our hypothesis was that similar pathology underlies the spasticity of males hemizygous for L1CAM mutations.

Methods: Studies were performed on 7 adult carrier females and 9 hemizygous males (aged 2–29 years). Transcranial magnetic stimulation (TMS) excited the CST and responses were recorded bilaterally in biceps brachii, abductor digiti minimi and quadriceps femoris. Subthreshold CST conditioning of the stretch reflex of biceps and quadriceps was performed. The CST was examined in post-mortem brain and spinal cord material from a one month old male with L1CAM mutation.

Results: TMS evoked contralateral and ipsilateral responses in all three muscles, which were in the normal range in upper limb muscles but had abnormally high thresholds and delayed onset in quadriceps. Ipsilateral responses were smaller, of higher threshold and delayed relative to contralateral, indicative of CST decussation. The patterns of CST modulation of the stretch reflex in biceps and quadriceps were abnormal, suggesting a reduced...
projection to inhibitory interneurones at both segmental levels. Abnormality of CST function was greater in hemisymmetric males than in carrier female. Post mortem examination of the CST revealed normal decussation and axonal pro-
tections to lumbar spinal segments. 

Conclusions: These data show abnormalities of CST function in hemi-
symmetric males who “carrier” females which is greater in the projection to the lower limb. However, anatomical and neurophysiological evidence reveal CST axo-
nal decussation and projection to the lumbar spinal segments. While these results support the hypothesis that LICAM has an important role in CST devel-

opment, they do not indicate significant abnormality of CST axonal guidance as suggested by the LICAM knockout mouse.

G225 INHERITED EARLY ONSET SEVERE AXONAL POLYNEUROPATHY WITH RESPIRATORY FAILURE AND AUTONOMIC INVOLVEMENT

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We report dizygotic twin infants with early onset of severe axonal polyneu-
ropathy presenting with respiratory failure due to diaphragm weakness. A few

siblings who had had been similarly affected died from respiratory failure at nine months of age. Both twins had persistent tachycardia, increased sweating and hypertension secondary to a autonomic involvement. This is an unreported pre-

sentation of presumed autosomal recessively inherited hereditary motor sen-
sory neuropathy.

Twin male black infants born at 36 weeks gestation presented at six months of age with acute respiratory failure requiring mechanical ventilation for over six weeks following which they underwent tracheostomies to receive continuous ventilatory support. There was no maternal polyhydramnios or parental consan-
guinity. Neurological examination revealed generalised hypotonia, wasting and flexion contractures of distal limb muscles with absent deep tendon reflexes. Cognitive function was normal. Bilateral diaphragmatic paralysis was confirmed by ultrasound. Persistent hypertension refractory to betablocker therapy responded to phenoxycbenzamine. Extensive investigations on serum and urine, imaging studies of brain, kidney and heart were normal. EMG showed profuse fibrillations at rest. Mixed median nerve action potentials and sensory action potentials were absent. Nerve biopsy showed reduced proportion of myelinated axons with preservation of unmyelinated axon popu-

lation and axonal degeneration. Muscle biopsy revealed typical features of

denervation. Duplication of chromosome 17p11.2 was not detected in either infant. The twins are now 10 months of age and continue to receive ventilatory support.

Congenital forms of axonal polyneuropathy causing early respiratory failure are extremely rare. To conclude, this description would seem to be a unique disorder, the gene defect for which is unknown.

G226 MRI APPEARANCE AND CLINICAL SIGNIFICANCE OF PUNCTATE LESIONS IN THE NEONATAL BRAIN

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Aims: Although considered as having a haemorrhagic origin, the clinical sig-
pcificance of punctate lesions in neonatal brain MRI is largely unknown. Our aims were to study the incidence of these lesions in a large cohort of newborns and to relate anatomical characteristics to clinical outcome.

Methods: Punctate lesions were characterised for number [-3, 3-10]; appearance [clump, linear]; distribution [anterior, middle, posterior, uni- or bilat-

eral]; signal intensity (SI) [SI on T1, T2]; association with minor [cortical highlight-

ing, discrete oedema] and major [malformation, PVL, infarction, intraventricul-

ar bleeding, cortical atrophy] abnormalities.

Results: We analysed 110 MRI scans (92 neonates, born in 1998). Punctate lesions were observed in 15/50 preterm neonates. The number of lesions were either <3 (n=3, all clumps), 3-10 (n=5, 3 linear) or >10 (n=7, 6 linear). SI was uniformly increased on T1 and reduced on T2. The lesions were seen peri-


tricularly and in the centrum semiovale (14/15), posterior (8/15) and anterior (3/ 15). All other brain abnormalities were related to asphyxia and were either minor (11/15) or major (4/15). Lesions preterms without punctate lesions (35/50) brain abnormalities were either absent (18/35), minor (4/35) or major (13/35). Punctate lesions were seen in 24/24 term neonates, organised in clumps, with only minor additional brain abnormalities. Clinical outcome of infants with iso-

lated punctate lesions (i.e. involving only minor additional brain abnormalities) is highly favourable (n=13, mean age=14.5 months, delayed development of language in 1 patient).

Conclusions: Punctate lesions are predominantly seen in preterm neo-

nates, most commonly linearly organised and bordering the lateral ventricles, their origin most likely being haemorrhagic. Caution is needed in interpreting isolated punctate lesions as being pathologic, since so far most of these patients have a normal developmental outcome.

G227 AN ATYPICAL CASE OF ALPERS’ SYNDROME

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Case Report: We present a case of a boy aged 1 year 7 months with mild developmental delay and poor growth. Both this boy and his elder sister were on the “at risk register”.

Investigations showed elevated serum and CSF lactates with generalised aminoaciduria and abnormal liver function tests. Neurophysiology, neuroimaging,

and skeletal survey were normal. Skin fibroblast cultures showed normal fatty acid oxidation studies, pyruvate dehydrogenase and pyruvate carbox-

ylase, and muscle biopsy was normal on histochemistry and EM. Cytochrome

oxidase IV was low normal.

He was admitted at 1 year 10 months to ITU with left sided convulsive movements. He was floppy with a reduced level of consciousness. EEG


285.
showed focal electrical status epilepticus over the right hemisphere. There was worsening of liver function tests and clotting results. A CT scan was normal, however, 2 days later a follow-up MRI showed diffuse cortical thickening, hyperintensity and bilateral thalamic lesions on T2 weighted images becoming more pronounced on a follow up MRI scan. CT scanning by this stage demonstrated global cerebral swelling and hypodensities to be a "cerebral reverse sign". Despite ventilation and treatment with anticonvulsants he continued to have myoclonus and died of a respiratory tract infection. Postmortem findings were obliteration of occipital lobe medially, and sponginess involving frontal and parietal lobes. The liver showed fatty change, bile duct proliferation, inflammation and nodular regeneration.

Conclusion: This was an atypical case of Alper's disease as intractable seizures occurred very late after his initial presentation. His later MRI scans were consistent with severe hypoxic ischaemic encephalopathy.

GUILLAIN-BARRE SYNDROME AND JAPANESE ENCEPHALITIS PRESENTING AS ACUTE FLACCID PARALYSIS IN SOUTHERN VIETNAM

G228

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As part of the surveillance programme for poliomyelitis, all children with an acute flaccid paralysis (AFP) admitted to a infectious diseases hospital in southern Vietnam from 1996–1999 were prospectively followed between 1996–1999. 15 and of 43 children were diagnosed with Guillain-Barre syndrome (GBS) clinically and 9 patients had evidence of acute Japanese encephalitis (JE) infection. Compared with the JE cases, the GBS cases were more likely to have had a preceding illness, arm involvement, asymmetrical and distal weakness, whereas the JE cases were more likely to have fever at onset of weakness, neck stiffness, urinary retention and the later development of encephalopathic features. 3 GBS and 2 JE patients developed respiratory failure and one patient died in each group. All surviving GBS patients made a functional recovery by 12 months but 7 with JE had severe residual flaccid weakness resembling the sequelae of paralytic poliomyelitis.

Electrodiagnostic criteria divided the GBS cases into 3 with acute inflammatory demyelinating polyneuropathy (AIDP), 11 with acute motor axonal neuropathy (AMAN) and 1 unclassifiable. The AMAN group only differed clinically from the AIDP group in having no sensory deficits and less bladder involvement. However 7 patients in the AMAN group had raised levels of anti-ganglioside antibodies compared to none of the AIDP cases, and 4 of these had anti-GD1a antibodies, a feature also highly significant in cases of AMAN reported in other studies.

As the incidence of polio declines in Vietnam, knowledge of the predominant local causes of AFP needs to be revised. Distinguishing GBS and JE myelitis, may be difficult based on admission features. However making a specific diagnosis is important because of the different prognosis and for planning future preventative measures.

MOYAMOYA DISEASE IN A NORTH-EASTERN POPULATION

G229

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Moyamoya disease is a rare cerebrovascularopathy of unknown origin and has primary and secondary types. The primary form is most commonly seen in Japan where it affects nearly 4000 people.

Aims: to document the case presentation, features and natural history of Moyamoya within a Caucasian population.

Methods: retrospective case notes review of all children with Moyamoya under our care

Results: 6 children were identified with primary Moyamoya disease. 3 presented with stroke, 1 with learning disabilities and migraine, 1 with transient ischaemic attacks and 1 with migraine and choreoathetosis. Age at presentation ranged between 9 months (stroke) and 8 years (choreoathetosis). Apart from the 2 children who presented directly to our unit with stroke where a diagnosis of Moyamoya was confirmed in the acute period, this took between 3 and 11 years in the remaining group. All 4 children presenting under the age of 4 years had mild or no associated disabilities. 3 of the children had received 14 MRI/CT procedures. 1 child had surgery within 1 month of presentation and has a normal cognitive profile; the other 2 children had their surgery more than 5 years from presentation and have learning disabilities. Of the children presenting under the age of 4 years, 2 are siblings with an affected mother, the third has an affected mother and the fourth child’s mother has not yet been imaged.

Conclusion: Familial Moyamoya occurs in 10% of the affected Japanese population and has recently been localised to Chromosome 3p. Although Moyamoya occurrence is low in Caucasians, the presence of “cerebral reverse sign” in our very small series we found that half the children had an affected parent. Young children at presentation had a poor prognosis. With the advent of MRA, screening of first degree relatives should be considered.

THE DEVELOPMENT OF A CONSENSUS BASED GUIDELINE FOR THE MANAGEMENT OF THE CHILD WHO HAS HAD A SEIZURE

G230


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Introduction: Seizures affect 3% of children and account for 5% of all medical attendances to paediatric accident and emergency (A&E).

Aims: To develop an evidence based practice guideline for the child who presents to A&E having had a seizure, using the Delphi method.

Method: A systematic search of electronic databases 'Medline', 'Cochrane' and 'Embase' was performed. Relevant quality publications were graded according to their strength of evidence. Statements were made with an evidence level of recommendation. Publications used, the literature review and derived statements were sent to a national panel (selected to be broadly representa- tive) of 30 medical and nursing staff. They were asked to rate their level of agreement with each statement on a 1-5 Likert scale and to comment. Consensus agreement was pre-defined as 83% of panelists rating 7-9. The results were fed back in two rounds to the panel with amendments for further rating and comments.

Results: 42 statements were made, 2 were based on level III evidence, 7 implied by level III, 23 on level V and 10 were derived from panelists comments. 10 reached consensus in round 1, 14 in round 2, 10 in round 3 and 8 did not achieve consensus. These 8 were alternatives to those that did, or concerned issues that could be decided at a local level.

Conclusions: This process resulted in a comprehensive evidence and consensus based guideline on the management of the child presenting to A&E having had a seizure. An analysis of the impact of this guideline will be presented separately.

WORSTER-DUFFY SYNDROME (W.D.S.) A MILD TETRAPLEGIC PERISYLVIAN CEREBRAL PALSY: A REVIEW OF 47 CASES

G231

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Aims: To clarify the diagnosis of Worster-Dufty Syndrome and to record associated features and complications.

Methods: Retrospective case-note analysis of 47 children thought to have W.D.S. on clinical grounds i.e. a congenital upper motor neurone bulbary palsy.

Results: The study children had significant bulbary problems (with 80% still needing a modified diet and a similar number with speech impairments requir- ing augmentative communication methods at last review). There were also high rates of predictable bulbary complications (86% had dribbling; 60% had glue ear; gastro-oesophageal reflux in 40%; history of poor nutrition in 40%; aspiration in 40%). Most of the children had additional complex impairments (91% had mild pyramidal tetraplegia; 81% learning impairments; 60% malfor- mations/deformations; 41% neuropsychiatric problems and 28% epilepsy). Over half of the children had significant medical problems in the first year but the mean age at diagnosis was 6 years.

There were no obvious causes in pregnancy or birth. 6 children had a family history of W.D.S. 32% (12/37) had abnormal neuroimaging including 5 with bilateral perisylvian polymicrogyria.

Conclusion: In our experience W.D.S. is not uncommon, is relatively easily diagnosed and is crucial not to miss as the management of these child's multiple impairments is complex and requires a careful team approach.

W.D.S. is a clearly a form of cerebral palsy, as a syndrome that includes motor impairment arising from static damage to the brain in early life. Its core ele- ments are a supravalvar paresis, a mild spastic tetraplegia and a significant excess of cognitive and behavioural impairments and epilepsy.

MAGNETIC RESONANCE IMAGING OF THE NEONATAL CENTRAL NERVOUS SYSTEM ON THE NEONATAL INTENSIVE CARE UNIT

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Aims: To assess the role of a dedicated magnetic resonance scanner on a neonatal intensive care unit.

Methods: A 0.2 Tesla permanent magnet system (Nichi, Innsversion) was installed in a room adjacent to the NICU. Imaging protocols were optimised on 60 normal controls. Subsequently MR imaging was performed on 40 babies with suspected intracranial haematoma (32 had patent channels). All patients had spin echo T1 weighted images, 25 had spin echo T2 weighted images and 3 had diffusion weighted imaging. All images were reported independently by two experienced radiologists. The results were compared with the clinical, cranial ultrasonography and conscious sedation in 25 of the MR scan.

Results: One hundred babies have been successfully scanned to date, 68 normal and 32 with pathology. Of the 32 cases with pathology the ultrasound and MR results were in agreement in 13cases. In 16 the MR images provided...
additional information to the ultrasound and this had management implications in 7 patients. Two patients had normal ultrasounds and the MR showed middle cerebral artery stenosis. One patient had a large posterior fossa mass and the other had schizencephaly not seen on ultrasound. One had severe hydroce- phalus on ultrasound, which was proven as hydrancephaly with MR im- aging. Hydropncephaly was seen in 2 patients several days earlier on MR than Ultrasound. In 3 cases ultrasound demonstrated grade one intra- ventricular haemorrhages not seen on the MR images.

**Conclusion:** Low field strength MR imaging provides a valuable method of imaging and provides additional information to ultrasound.

**G233**

**COLLIDING BABIES WITH HYDROCEPHALUS AND LARGE HEAD, A NEW SYNDROME?**

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Hydrencephaly has been noted in several syndromes, ie Jøgensen Larsson syn- drome, (global delay, epilepsy, diplegia), Retts syndrome (ataxia, neuropa- thy, etc), Rudi syndrome (epilepsy and mental impairment) and Tay syndrome. However we are unaware about its association with hydrocephalus. We present two brothers with hydrencephaly born to non consanguineous white par- ents.

Case one, now a three year old, was noted to have partachment like skin, ectropion, cracking of skin at knee and groin and contractures of knee, ankle and elbow. His weight and head circumference were between 10th and 25th Centile.

His skin seems to be improving with treatment. Unfortunately at 6 months of age he presented with vomiting and had clinical signs of hydrocephalus. Cranial CT scan confirmed that hydrocephalus was due to acquired stenosis. He has required revision of shunts since then. He is displaying delay in all areas of development and is being treated for epilepsy. We would show clinical photograph, growth chart and cranial CT scan.

Case two, now a one year old, had partachment like skin and low set deformed ears. His skin is much better. His developmental milestones are normal. Now he has a rather large head (weight between 30th-75th Centile and head circum- ference 75th-90th Centile) but so far has not developed hydrocephalus.

We have excluded X-linked ichthyosis due to steroid sulfatase deficiency and feel that ichthyosis is of autosomal recessive origin. We would highlight the significance of measuring head circumference and monitoring development in children with ichthyosis. If our second case develops neurological problems then we would have to expand our battery of investigations.

**G234**

**CLINICAL AND ELECTROENCEPHALOGRAPHIC SEIZURES IN COMA: RELATIONSHIP TO OUTCOME**

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**Aims:** Prolonged and multiple seizures are associated with brain damage in humans. Status epilepticus may occur in acute encephalopathy but prognosis has been assumed to be related to etiology and the severity of underlying process rather than to the seizures. This study examines the hypothesis that the prognosis for coma in childhood is determined, at least in part, by the presence of seizures and/or status epilepticus.

**Methods:** The patients were observed by members of the nursing and medical teams and any clinical signs suggestive of seizures were noted. A cerebral function analysing monitor (Medaid) was used to monitor either 1 or 2 channels of EEG. The presence, number and duration of any EEG seizure discharges in an individual patient were noted and were compared with survival and outcome (normal or moderate handicap=good; severe handicap, vegetative state or death=poor) using the Mann-Whitney U-test.

**Results:** 188 patients (median age 2, range 0-15 y; 52% male) with a wide range of aetologies (113 hypoxic-ischaemic encephalopathy, 45 cerebral malaria, 11 encephalitis, 2 meningitis, 7 head injury, 3 Reye’s syndrome and 7 miscellaneous) were studied. Survival was related to aetiology (p=0.0001), Glasgow coma score on admission (p=0.0001) and mean arterial pressure on admission (p=0.004), but not to number (p=0.3), duration (p=0.5) and duration of the longest seizure (p=0.7). Outcome was related to aetiology (p=0.0001), Glasgow coma score on admission (p=0.0001), EEG score on admission (p=0.0001), EEG score on admission (p=0.007), and to number (p=0.006), duration (p=0.004) and duration of the longest seizure (p=0.003).

**Conclusions:** Although survival and outcome for coma in childhood may be predicted at least in part on admission, seizures after admission appear to be a potentially modifiable cause of neurological morbidity and warrant interventions designed for prevention or treatment.

**G235**

**MUTATIONS IN THE TRKA GENE IN KUWAITI PATIENTS WITH CONGENITAL INSENSITIVITY TO PAIN WITH ANHYDROSIS (CIPA)**

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We describe mutations in the TRKA gene in 7 children from 5 consanguineous Kuwaiti families with CIPA. CIPA is a rare autosomal recessive disorder characterised by unexplained fever, anhydrosis, mental retardation and absence of pain resulting in self-mutilation of fingers, tongue, lips and joints. The TRKA gene encodes a tyrosine kinase receptor for nerve growth factor, which ensures survival of embryonic sensory & sympathetic neurones. The following mutations were found: a single base deletion in exon 1 (c.284 del A) of the gene in 2 sibs and a G to A substitution in the 5’ splice site of intron 7 (IVS7 + IG-A) in another 2 sibs (These have been reported in Am.J.Hum.Genet.64: 1570, 1999).

Two unrelated children had a novel C to T transition at nucleotide 610 (c.610 C>T) in exon 5, which changes Gin a termination codon at amino acid 176 (Gin 176X). The parents of all these 6 patients were heterozygous. The last patient showed an 8 base-pair insertion in exon 13, but the mother did not show the same mutation. Mutations of the TRKA gene are associated with CIPA.

**G236**

**THE IMPACT OF IDIOPATHIC GAIT DISORDERS**

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Many children are in hospital with gait disorders requiring treatment, some physical, some idiopathic. The prevalence of idiopathic gait disorders in chil- dren is unknown. The impact of school absence, investigations and therapy may be substantial. The intent of this study was to estimate the incidence and impact of idiopathic gait disorders in children admitted with gait disorders in a tertiary children’s hospital.

We evaluated prospectively all the children admitted with a gait disorder requiring treatment in the form of physiotherapy at Birmingham Children’s hospital, using a standardized pro forma, during a three-month period (March and June 99).

103 children (ages 2 to 16) were admitted with gait disorders (57 female), 45 children were admitted under orthopaedics, 32 neurosurgery, 26 other. Eight (ages 10 to 15) had an idiopathic gait disorder (5 female). All 8 children exhib- ited function impairment, pain and school absence. Some had lethargy (6) and sensory symptoms (4). Most reported an antecedent illness (5) and a similar illness history in a acquaintance (4). All children were above average at school and most had many friends (7). Results were normal of physical and laboratory and imaging examinations.

The symptoms resolved after therapeutic admission (1 week to 6 months).

Idiopathic acquired gait disorders are 7.7% of gait disorders in admitted children. The economic and social impact is substantial with regard to diag- nosis, investigations, treatment and school absence.