Neurology

G110 NATIONAL SENTINEL AUDIT INTO EPILEPSY DEATHS: PAEDIATRIC CASES

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Aims: To benefit children with epilepsy, their families and health professionals involved in their care by understanding better the circumstances that have led to the deaths of children with epilepsy, leading to improved management of care through the more consistent application of guidelines and the development of potential preventative strategies.

Methods: A Department of Health-funded (through 'NICE') and Epilepsy Bereaved-managed audit was undertaken from April 2000 to July 2001. Primary care, secondary care and post-mortem audit tools based on available standards were applied to each child with epilepsy dying before their 18th birthday. The audit tools were completed by trained personnel using each patient's medical notes. Each paediatric death was independently assessed by Consultants in Paediatric Neurology (2), General Paediatrics (l) and Adult Neurology (2) using the three audit tools. Subsequent discussion yielded a single, consensus opinion on each death in areas of the standard of medical care prior to death and the circumstances of the death.

Results: During the study period there were 81 children dying with epilepsy reported to the Office of National Statistics and General Register Office. Complete audit data were available for 22 children. l5 of the children had learning difficulties and 11 had physical difficulties. Judged against available standards, the medical care of these patients was considered to have met the standards in 3, fallen below the standards in 17 and have been unclear in 2. Of the 17 patients with "substandard" care the death was considered to have been probably unavoidable in 3, probably avoidable in l, possibly avoidable in 11 and unclear in 2. Areas of substandard care included inadequate access to specialist epilepsy advice and investigations and the inappropriate use of antiepileptic medication.

Conclusion: There are potentially major implications for the standards of medical care of epilepsy in children, if the results of this audit are representative of current epilepsy management in the UK.

G111 THE CURRENT PRACTICE OF PAEDIATRIC EEG REQUESTS

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Aims: To determine the completion quality of Paediatric EEG request forms, the appropriateness of EEG requests and the usefulness of the EEG report in management.

Methods: A retrospective analysis of 100 consecutive EEG request forms was performed in a Regional Paediatric Neurology department from May–July 2000. Comparisons were made between the clinical history from the referral form and the Technician. A Consultant Paediatric Neurologist determined the appropriateness of each EEG. Prospective questionnaires were sent with 100 consecutive EEG reports, asking whether each EEG had helped management, and if so, how?

Results: Paediatricians requested 62% of EEGs and Paediatric Neurologists requested 38%. Requests were for suspected epilepsy in 49%, suspected encephalopathy in 13% and monitoring progress in 38%. Seizure description was good in 52%, basic in 30% and poor in 18%. The commonest seizure definitions were tonic-clonic (24%), absence (21%) and partial (19%). The EEG Technician's history yielded extra relevant information in 67% and highlighted missing relevant medical details in 6%. The EEG results were abnormal in 55%, normal in 37%, inconclusive in 7% and 1% was not performed. The investigation was deemed to be appropriate in 79% cases and inappropriate in 21% cases. There were 91/100 replies from the prospective questionnaire. Management was helped in 89% (confirming diagnosis of epilepsy in 35% and indicating change of treatment in 12%) and in 11% the EEG was not felt to be useful. Reassurance that a normal EEG excluded a neurological problem was stated in 14%.

Conclusions: Descriptions of seizures by clinicians are suboptimal, and some EEG referrals are inappropriate, but most EEGs help management. Referrals would be improved by enhancing the clinical history and re-educating paediatricians about adherence to EEG guidelines in paediatric practice.

G112 AN EVIDENCE BASED GUIDE TO INVESTIGATION IN NONFEBRILE CONVULSIONS

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Introduction: Nonfebrile seizures occur in 1% of children under 20 years of age, of whom 65% will not have any further seizures. Of those who do, the majority have idiopathic epilepsy. Despite this, considerable resources are spent on excluding underlying pathology in children who present with a seizure.

Methods: 113 patients were investigated for afebrile convulsions during April 2000–2001. A random sample of 31 case notes (27%) have been reviewed so far for investigations performed and final diagnosis. Costings for investigations were obtained. Current best evidence was reviewed.

Results: EEG:30 patients: 21 normal, 9 abnormal:4 petit mal, 2 generalised,3 focal CT :11 (35%): 1 abnormal (NAI) MRI: 2 Metabolic screens: 10 patients—all normal

Final diagnoses: Epilepsy 12 (38%): 4 Absence, 1 Simple partial, 7 generalised

Not Epilepsy 19 (61%): 10 Normal, 2 Vasovagal,2 Breathholding/Reflex anoxic, 2 Autism. 3 Atypical febrile convulsions

Cost: For investigating 15 "normal" patients £3943.

Total cost: For investigating all 31 patients £12209 (est £45200pa)

Discussion: There is evidence that routine investigations following an uncomplicated seizure are unnecessary. A metaanalysis of 1374 patients by the AAP found abnormalities in only 0.4% of patients. Blood glucose should be measured but further investigations should only be done where there is suspicion on history or examination or if the patient is in status. Likewise, CT/MRI should be limited to certain groups and are not recommended for absence, benign rolandic seizures or idiopathic generalised epilepsy. Where an urgent scan is not indicated, an EEG should be done prior to imaging if the seizure classification is undefined. Further investigations e.g. metabolic screens should only be done where there is a high suspicion of underlying disease.

Conclusions: Best evidence suggests that we are doing too many investigations for first nonfebrile seizures. Patients and budget holders bear the costs of our desire to find the obscure diagnosis amongst the common.

G113 CHILDHOOD CEREBROVASCULAR DISEASE AND STROKE LIKE ILLNESS IN THE UNITED KINGDOM AND EIRE, A DESCRIPTIVE EPIDEMIOLOGICAL STUDY

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The incidence of childhood stroke has been estimated to be 2.5–13.5/100 000 per annum in North America and Europe. A regional study in Birmingham suggested a figure of at least 2/1 000 000 per annum for the United Kingdom and Eire, with an anticipated annual number of cases of at least 200.

This 14 month descriptive epidemiological study, under the BPSU Orange Card Scheme and funded by the Stroke Association, aimed to clarify the incidence and aetiology of stroke, stroke-like illness and cerebrovascular disease in children between birth (>36 weeks gestation) and 16 years. Current investigation and management protocols have been examined. Parallel surveillance involved cardiologists, haematologists, neurosurgeons and radiologists.

The study commenced in 1 January 2001 and we are reporting the first 9 months of this ongoing project. So far there have been 273 notifications. Taking into account incorrect referral and 20 duplications, there were 223 confirmed cases (which excluded 5 cases of Vein of Galen and 4 of Sturge-Weber syndrome). This provides an estimated incidence of 2.475 cases per 100 000 per annum and a calculated yearly estimate of 297 cases, which is in line with the Birmingham pilot regional study. The BPSU identified 200 (86%) of the cases. The remainder were from neurosurgeons (9 cases, 3.9%), radiologists (10 cases, 4.3%), haematologists (2 cases, 0.86%) and cardiologists (2 cases, 0.86%). From 103 for whom the data are available, 53% were male and median age was 3.5 years (range 1 day–16 years; 13 neonates). For the first 5 months, there have been 7 deaths and 4 recurrences by 6 months, for approximate mortality and recurrence rates of 6% and 3% respectively over this follow-up period.

Childhood stroke is an important cause of childhood morbidity and mortality. These data will allow the appropriate development of services and of intervention trials aimed at secondary (and eventually primary) prevention.

G114 THE CANADIAN AND BRITISH PERSPECTIVES OF PAEDIATRIC INTELLECTUAL AND NEUROLOGICAL DETERIORATION(PIND): ARE THE RESULTS COMPARABLE?

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Presented on behalf of the Canadian Paediatric Surveillance Program participants and principle investigator with co-operation from the British Paediatric Surveillance Unit.

Objectives: To identify, via active surveillance, children with PIND in order to determine its incidence; to evaluate and classify cases in order to investigate possible cases of variant Creutzfeldt-Jakob disease (vCJD).

Abstract G114

PIND Diagnosis	CPSP	BPSU
Neuronal ceroid lipofuscinosis	8	55
Gangliosidosis	0	50
Mitochondrial encephalopathies	8	38
Mucopolysaccharidoses	6	33
Krabbe's disease, Rett syndrome	4,3	0
Cri encephalophy	2	0
Adrenoleukodystrophy	2	28
Nieman-Pick-type C	1	22

Methods: Paediatricians reported cases on a monthly check-off form to the British paediatric surveillance unit and the Canadian paediatric surveillance program. Each unit used the same case definition and protocol and had an expert panel review challenging cases. Reporting periods were 2 years for CAN and nearly 4 years for UK.

Results: CAN reported one 13 year old iatrogenic CJD but no vCJD; the UK did not have any CJD but detected 4 cases of vCJD (4 confirmed, 1 probable). Both countries identified a variety of neurological disorders:

Conclusion: Results are comparable. One exception was the incidence of gangliosidosis where CAN reported zero cases, most likely due to an effective screening program of the at risk population. Other exceptions and similarities will be discussed. Overall, the study increased awareness of the complexity of PIND differential diagnoses.

G115 CEREBROSPINAL FLUID (CSF) PRESSURES IN CHILDREN ON INTENSIVE CARE. DOES APPROPRIATE BEDSIDE NEUROLOGY PREDICT NORMAL CSF PRESSURES?

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Background: Children with suspected meningoencephalitis often present with clinical features of raised intracranial pressure (ICP) requiring admission for a period of neuro-intensive care. Lumbar punctures (LP) are postponed in these children until there is no evidence of raised ICP on clinical examination, thus representing normal bedside neurology.

Aim: To determine the CSF manometer pressure in children receiving an LP after a period of neuro-intensive care.

Methods: A prospective study was undertaken from March to November 2001 to include all patients admitted to a Paediatric Intensive Care Unit (PICU) who required an LP. This was performed when clinical assessment revealed no signs of raised ICP after a period of neuro-intensive care. CSF opening pressure was measured in the lateral decubitus position using a manometer.

Results: 34 patients were included in the study. The median age was 18 months (range 4 days–16yrs). CSF microscopy was abnormal in 7 (20.5%) of these patients. The duration of neuro-intensive care prior to LP ranged from 0.5-15 days, with 11 (32%) LPs performed after more than 1 day. The CSF manometry pressure was >20cmH2O in 22 (65%) cases. The median CSF manometry pressure was 24cmH2O (range 0–40; interquartile range 14–31).

Conclusions: The CSF manometry pressure was high in the majority of patients who required an LP on PICU. This was found in patients who had no clinical evidence of raised ICP at the time of LP following a period of neuro-intensive care. Our study suggests that had LP been performed earlier in the course of the illness CSF pressure might have been higher. Further studies are needed to determine the role of manometer CSF pressure measurement in this group of patients.