Poster Presentations

Child Neurology (Epilepsy and EEG)

P.001

Early plasma exchange in FIRES resulting in favorable outcome

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doi: 10.1017/cjn.2015.112

Background: Febrile infection-related epilepsy syndrome (FIRES) is a devastating entity characterized by acute onset of refractory epileptic status preceded by a febrile infection, for which no aetiology has been identified so far. *Methods:* We report the cases of two males presenting with typical FIRES, for whom extensive investigations revealed no specific aetiology. Failure of controlling seizures with multiple anticonvulsants as well as barbiturate coma lead to the decision to try immunotherapy. The first patient received plasma exchange after a negative trial of IVIGs, while the second received plasma exchange in the beginning. *Results:* Significant improvement in the seizure frequency and intensity was obtained following plasma exchange, and weaning of barbiturate coma was successful in the days following treatment. Both patients remain with significant temporal lobe epilepsy, requiring treatment with 3 anti epileptics. However, cognitive outcome is surprisingly good for both, both exhibiting normal IQs and normal everyday function, with the second patient showing even better recovery, possibly due to earlier treatment with plasma exchange. *Conclusion:* Our findings of favourable outcome with plasma exchange favours the auto-immune hypothesis often discussed in FIRES. While awaiting further insight onto the aetiology of this syndrome, we suggest a trial of plasma exchange in patients affected.

P.002

Surgical and seizure outcome in children with DNETs who underwent epilepsy surgery

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doi: 10.1017/cjn.2015.113

Background: Dysembryoblastic neuroepithelial tumors (DNETs) are benign tumors of the cerebral cortex that most commonly occur in children or young adults. Seizures are a frequent presenting feature, with an incidence of 80-100%, and are often an indication for surgical resection. *Methods:* We performed a retrospective chart review of children with DNETs who underwent epilepsy surgery between 1998 and 2014. *Results:* A total of 12 subjects were identified (6 males, 6 females), all of whom had seizures prior to surgical resection. Of these patients, 1 had infantile spasms, 2 had simple partial seizures and 10 had complex partial seizures. Tumors were located in the temporal (n=7), frontal (n=3) or parietal (n=2) cortex. These patients went on to have surgery on average 15 months after seizure onset, 3 had incomplete resections. At an average follow up of 6 years 4 months, all patients were class 1 on Engel's Classification. All but one subject with rare non-disabling seizures were seizure free, with only 6 on medication. Follow up MR imaging revealed tumor recurrence in 1 subject. *Conclusions:* Despite differing seizure seminology and tumor location, surgical resection of these low-grade tumors resulted in excellent seizure outcome even in the setting of incomplete tumor resection.

P.003

A multi-modality approach to identifying primary generalized epilepsy that can mimic focal epilepsy

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doi: 10.1017/cjn.2015.114

Introduction: Evaluating the suitability for surgery in patients with epilepsy requires determining if the epilepsy is focal or generalized. Presurgical workups can indicate focal epilepsy in certain cases of generalized epilepsy (GE). The purpose of this study was to identify distinctive features which characterize patients with primary GE that mimics focal epilepsy. *Method:* We retrospectively identified 19 children with generalized interictal discharges during scalp video-EEG (SVEEG) and underwent invasive monitoring and/or epilepsy surgery. Two children did not undergo resective surgery due to final diagnosis of primary GE (Group A). Seventeen children underwent a resective surgery (Group B). Scalp video-EEG, MEG, MRI, and intracranial video EEG (IVEEG) were reviewed. *Results:* On (SVEEG), the frequency of generalized spike-and-waves (GSW) was 3Hz in Group A and 1.5-2.5Hz in Group B. Group A had only absence seizures , whereas 80% in Group B had multiple types of seizures. Both groups had lateralized MEG dipoles. One patient in Group A had a focal MRI abnormality. In Group A, IVEEG showed GSW of 3 Hz frequency with inconsistent leading. In Group B, IVEEG showed consistent localization of ictal and interictal high frequency oscillations. *Conclusion:* Children with generalized 3 Hz spike-and-waves and only absence seizures may be a contraindication of resective surgery even though some presurgical workup shows focality.

P.004

Decreased nasal nitric oxide in children with isolated midline neuroanatomical defects: a possible indicator of ciliary dysfunction?

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doi: 10.1017/cjn.2015.115

Background: Ciliary mutations cause multi-system disorders, often involving the CNS. We set to evaluate the prevalence of ciliary dysfunction in children with isolated neuroanatomical defects,