Poster Presentations

Child Neurology (Epilepsy and EEG)

P.001

Early plasma exchange in FIRES resulting in favorable outcome

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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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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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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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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,

by measuring nasal nitric oxide (nNO), a screening test for Primary Ciliary Dyskinesia (PCD). *Study design:* We measured nNO levels of 26 children with congenital midline CNS defects. We evaluated the effect of age, gender, and anomaly (brain, spinal cord, or combined) on measurements. We compared our results to the previously established normal range (153.6-509.9 nL/min), and to the cutoff for PCD (77 nL/min). *Results:* The range for nNO in our cohort was 56.5-334.7 nL/min, with age, gender, and anomaly not having a significant effect. The overall mean, 217.7 nL/min, was significantly lower than that of normal children, 314.51 nL/min ($p<0.01$). Four subjects (15.4%) had nNO levels below the lower end of normal, with two (7.7%) having values fitting the cutoff for PCD. *Conclusions:* We report an association between ciliary dysfunction and isolated midline neuroanatomical defects, not in context of any known syndrome. This suggests that genes causing isolated CNS defects, may be implied in the function of cilia. Longitudinal studies are required to investigate whether children with abnormal measurements suffer from any respiratory sequelae.

P.005

Utilization of transition care management plans to facilitate transition of adolescents with epilepsy into the adult healthcare system

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Background: For adolescents with epilepsy, there is often a poor system in place to meet their individualized transition needs. Our objectives were 1) to develop epilepsy-specific transition care management plans (TCMPs) to ensure access, and attachment to adult healthcare providers, and 2) to identify strategies for providing support during the transition period, including through the development of physician and patient (or caregiver) navigated web-based tools, resources and recommendations for health system improvements. *Methods:* Physicians and nurses with expertise in areas including adult and pediatric epilepsy, family medicine, psychiatry, and varied allied health professionals were engaged to generate epilepsy-related TCMPs. *Results:* Through an iterative process spanning the course of over a year, TCMPs were developed to cover areas including: treatment responsive and resistant epilepsy, ketogenic diet, epilepsy surgery, women's issues, mental health, and psychosocial aspects of epilepsy. The TCMPs referenced established guidelines and best practices in the literature wherever possible. Caregiver roles and responsibilities were outlined, remaining cognoscent of available provincial resources. *Conclusions:* Epilepsy specific TCMPs can be developed through a collaborative approach between pediatric and adult healthcare providers, easing the patient experience, creating educated accountability, and providing a forum to identify and address gaps of care in adolescents with epilepsy.

Child Neurology (General Pediatric Neurology)

P.006

Increased healthcare services utilization in the tuberous sclerosis complex population in Quebec

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Background: Tuberous sclerosis complex (TSC) is a neurocutaneous syndrome that can present with many disabling neurological symptoms, the most common being seizures. Although it is a chronic systemic syndrome, healthcare utilization and long-term outcome of subjects with TSC are not well defined. The goal of this study was to evaluate the direct cost and long-term outcome of TSC compared to other forms of epilepsy and healthy controls. *Methods:* Our provincial health care database was interrogated to determine use of medical services by patients with TSC, epilepsy and healthy controls from 1996-2011. Data on demographics, outcomes and health care utilization were analyzed. *Results:* 1004 TSC, 41,934 with epilepsy and 41,934 controls were identified. The prevalence of TSC was 1/7,872 compared to 1/189 for epilepsy. TSC experienced more hospitalizations, medical visits and prescription drug use, resulting in higher total health care costs. Their most common admission diagnosis was seizures and age at death was significantly lower: 61,3 years old for TSC vs 69,6 and 76,6 years old for epilepsy and controls, ($p<0,001$). *Conclusions:* TSC subjects have a significantly higher burden of disease than other subjects with epilepsy. These results stress the need for specialized services in this population through the lifespan.

P.007

Topographical orientation as a model of plasticity in children with perinatal stroke

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Background: Children with perinatal stroke go on to develop most cognitive skills (e.g. language) due to brain plasticity; however, their performance is usually poor when compared to age-matched controls, indicating a reduced potential compared to uninjured children. To date, how plasticity after early injury affects the development of complex cognitive skills remains uncertain. Here, we use topographical orientation, which relies on integration of several cognitive processes underlain by widespread neural networks, as a model to test plasticity in complex behaviour. *Methods:* Children with perinatal stroke and age-matched controls were tested with a neuropsychological battery and a novel navigation task. In addition, for each patient, we obtained the most recent MRI scan to assess the effects of lesion characteristics on performance at the navigational task. *Results:* Children with history of injury performed worse than controls, and their scores were not different based on lesion's laterality, location or functional region affected. In particular, involvement of regions known to contribute to spatial orientation did not result in significantly decreased performance. *Conclusions:* As seen in other