

CACN PLATFORM PRESENTATIONS

D.01

Video diversion improves success rate of fundoscopic exam in children: a randomized controlled trial

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Background: Fundoscopy is an important component of the neurological examination, but can be challenging in uncooperative children. This study explores whether playing a video during eye examination, improves the success, duration and ease of pediatric funduscopy. **Methods:** We completed a prospective, multi-clinic, block-randomized trial. Patients 1-4 years were recruited in the emergency department, neurology, spinal cord and general pediatric clinic. Patients were randomized (by eye examined) to video/non-video assisted funduscopy. Successful exams were defined as visualizing the fundus within 60 seconds. Time to visualize optic disc was recorded and difficulty of exam was examined using a 10-point Likert scale. **Results:** 101 subjects were recruited, with a mean age of 2.8 years. Overall, there was a 20% absolute improvement in the success rate of visualizing the optic disc in the video versus non-video group ($p < 0.01$). Time to visualize optic disc was also improved ($\Delta 5.3s$, $p < 0.01$). Improvement in ease of examination with video were noted by caregivers and practitioners ($p < 0.01$). **Conclusion:** Playing a video improved the ease, duration and most importantly the success of funduscopy in younger children. This simple, inexpensive adjunct has great potential to improve the ease and efficacy of this aspect of the neurological examination.

D.02

2015 update on triple therapy for pyridoxine-dependent epilepsy

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Background & Objectives: Of patients with pyridoxine-dependent epilepsy (PDE) due to ATQ deficiency, 75% suffer intellectual disability despite adequate seizure control with high dose pyridoxine. We aimed to assess the safety and efficacy of two novel therapeutic strategies to reduce accumulation of toxic intermediates in this cerebral lysine degradation defect. **Methods:** In two open-label observational studies, seven children with confirmed ATQ deficiency were started on dietary lysine restriction with regular nutritional monitoring, and outcome evaluation pipercolic acid, AASA levels in body fluids; development/cognition via age-appropriate tests and parental observations; epilepsy). Subsequently additional arginine supplementation was initiated to reduce cerebral lysine flux (cation transporter competitive inhibition). **Results:** Lysine-restriction was well tolerated and diet is safe, resulted in reduction of lysine intermediates in all body fluids in all patients (up to 80% reduction AASA

in cerebrospinal fluid), with beneficial effects on seizure control and psychomotor development. Additional arginine fortification resulted in normalization of biomarkers and dramatic improvement of psychomotor development. **Discussion:** Triple therapy is effective, especially if implemented early; studies for PDE newborn screening have been initiated. For dissemination and evidence generation, our PDE Consortium published Recommendations, developed a Digital Diet App and established a RedCap study database (www.pdeonline.org).

D.03

Canadian Paediatric Neurology Workforce survey and consensus statement

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Background: Little knowledge exists on the availability of paediatric neurology positions. Our study aimed to: 1) obtain information regarding the availability of positions for paediatric neurologists in academic centres; 2) survey paediatric neurology trainees regarding their perceptions of employment issues and career plans; 3) survey practicing community paediatric neurologists 4) convene a group of paediatric neurologists to develop a consensus regarding how to address these workforce issues. **Methods:** Surveys addressing workforce issues regarding paediatric neurology in Canada were sent to: 1) all paediatric neurology program directors in Canada who then solicited information from division heads and from paediatric neurologists in surrounding areas; 2) paediatric neurology trainees in Canada and; 3) community paediatric neurologists. A meeting was held with relevant stakeholders to develop a consensus on how to approach employment issues. **Results:** We found that the number of projected positions in academic paediatric neurology is fewer than the number of paediatric neurologists that are being trained. Paediatric neurology residents are concerned about job availability and desire more career counselling. **Conclusions:** There is a current and projected clinical demand for paediatric neurologists despite a lack of academic positions. Training programs should focus on community neurology as a viable career option.

D.04

Interplay between impaired brain structure and function in term newborns with congenital heart disease

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Background: Term neonates with congenital heart disease (CHD) demonstrate a high incidence of white matter injury (WMI), together with increased average diffusivity and decreased fractional anisotropy (FA) on MR diffusion tensor imaging. EEG background activity is a robust measure of functional brain maturation that becomes less discontinuous, contains more fast activity and shows higher complexity of EEG patterns with increasing age. We sought to determine the association between structural brain abnormalities and functional brain maturation in term neonates with CHD. **Methods:**

Thirteen term newborns with CHD underwent pre-operative MR imaging and continuous EEG recordings (cEEG). The proportion of cEEG with discontinuous vs. continuous background activity was quantified by visual analysis. During continuous epochs, we differentiated between two states: wakefulness/active sleep vs. quiet/transitional sleep, and applied algorithms to measure spectral power and EEG complexity. *Results:* Three patients had multifocal WMI which was associated with greater EEG background discontinuity ($P<0.05$). Moreover, lower white matter diffusivity was associated with higher power of fast activity ($P<0.05$ for wakefulness/active sleep EEG pattern), while higher white matter FA showed a trend toward being associated with increased EEG complexity ($P<0.1$ for quiet/transitional sleep pattern). *Conclusions:* In this series of term neonates with CHD, structural and microstructural white matter abnormalities are associated with impaired maturation of brain function.

D.05

[18F]-sodium fluoride PET/CT is an imaging-derived biomarker of hydroxyapatite expression in carotid plaque: sub-study of the Canadian Atherosclerosis Imaging Network (CAIN-2)

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Background: It has been hypothesized that [18F]-sodium fluoride (NaF) uptake imaged with positron emission tomography (PET) binds to hydroxyapatite molecules expressed in regions with active calcification. Therefore, we aimed to validate NaF as a marker of hydroxyapatite expression in high-risk carotid plaque. *Methods:* Eleven patients (69 ± 5 years, 3 female) scheduled for carotid endarterectomy were prospectively recruited for NaF PET/CT. One patient received a second contralateral endarterectomy; two patients were excluded (intolerance to contrast media and PET/CT misalignment). The bifurcation of the common carotid was used as the reference point; NaF uptake (tissue to blood ratio - TBR) was measured at every PET slice extending 2 cm above and below the bifurcation. Excised plaque was immunostained with Goldner's Trichrome and whole-slide digitized images were used to quantify hydroxyapatite expression. Pathology was co-registered with PET. *Results:* NaF uptake was related to the extent of hydroxyapatite expression ($r=0.45$, $p<0.001$). Upon classifying bilateral plaque for symptomatology, symptomatic plaque was associated with cerebrovascular events (3.75 ± 1.1 TBR, $n=9$) and had greater NaF uptake than clinically silent asymptomatic plaque (2.79 ± 0.6 TBR, $n=11$) ($p=0.04$). *Conclusion:* NaF uptake is related to hydroxyapatite expression and is increased in plaque associated with cerebrovascular events. NaF may serve as a novel biomarker of active calcification and plaque vulnerability.

D.06

The diverse manifestations of tuberous sclerosis complex: the experience of a provincial TSC clinic

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Background: Recent consensus recommendations for Tuberous Sclerosis Complex (TSC) stress the importance of multidisciplinary follow-up for these patients. The objective of our study was to review the manifestations of TSC seen in our hospital to determine the care needs of this population. *Methods:* This was a systematic, retrospective chart review of children with TSC treated at our institution. Patients were identified through epilepsy and clinical neurophysiology databases. *Results:* The study population comprised 81 patients, born between 1987-2014, who were a median 10 (Range 0.2-23.2) years of age at last follow-up. 88% of patients had epilepsy, including 30% with a history of infantile spasms. Developmental delay was reported in 65%, while 40% had intellectual disability. Psychiatric co-morbidities occurred in 49%. The most common psychiatric diagnoses were autism (25%), ADHD (19%), and anxiety (16%). Cardiac rhabdomyomas occurred in 35% of patients and renal angiomyolipomas in 42%, while only 4% had polycystic kidneys. Subependymal giant cell astrocytomas were observed in 14% of patients. 86% had skin manifestations. *Conclusions:* This study reaffirms the multi-system manifestations of TSC and the need to provide comprehensive, multidisciplinary care. As many children are still very young, the prevalence of autism and intellectual disability is likely underestimated.

D.07

Epilepsy surgery in tuberous sclerosis complex: the BC Children's Hospital experience

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Background: Epilepsy occurs in up to 90% of patients with Tuberous Sclerosis Complex (TSC) and is often refractory to medications. Our objective was to assess the safety and outcome of epilepsy surgery in children with TSC at our institution. *Methods:* We performed a systematic, retrospective chart review of children with TSC who underwent epilepsy surgery at our institution. Patients were identified through epilepsy and clinical neurophysiology databases. *Results:* 19 patients (out of 81 with TSC) underwent surgery between 1995-2014. Median age at surgery was 4.2 (Range 1.1-15.6) years, with patients having failed a median 4 (Range 0-10) anti-seizure medications. Surgery comprised corpus callosotomy in 2 and resection of one or more tubers in 17. 2 patients had a subsequent second resection. Minor neurologic deficits occurred after 14% of surgeries. Median follow-up was 2.4 years (Range 0.3 -13.8 years) following surgery. At last follow-up, 47% were seizure free, including 2 patients off anti-seizure medication. *Conclusions:* Epilepsy surgery is safe and effective in carefully selected TSC patients, with the majority having a good seizure outcome. Children with epilepsy secondary to TSC should be referred for epilepsy surgery assessment.