# Platform Presentations

## CACN Chair's Select Abstract Presentations

## A.01

CACN 2015 President's Prize

### Lidocaine for status epilepticus in pediatrics

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*Introduction:* Our goal was to perform a systematic review of the literature on the use of intravenous lidocaine in pediatrics for status epilepticus (SE) and refractory status epilepticus (RSE) to determine its impact on seizure control. M*ethods:* All articles from MEDLINE, BIOSIS, EMBASE, Global Health, HealthStar, Scopus, Cochrane Library, the International Clinical Trials Registry Platform (inception to November 2014), and gray literature were searched. The strength of evidence was adjudicated using both the Oxford and GRADE methodology by two independent reviewers. *Results:* Overall, 20 original studies were identified, with 19 manuscripts and 1 meeting abstracts. Two hundred and thirty-five pediatric patients were treated for 252 episodes of SE/RSE. Patients had varying numbers of anti-epileptic drugs (AEDs), 2 to 8, on board prior to lidocaine therapy. During 20 of the 252 (7.9%) episodes of SE/RSE, phenytoin was on board. The dose regimen of lidocaine varied, with some utilizing bolus dosing alone; others utilizing a combination of bolus and infusion therapy. Overall, 60.0% of seizures responded to lidocaine, with complete cessation and greater than 50% reduction seen in 57.6% and 12.3% respectively. Patient outcomes were sparingly reported. *Conclusions:* There currently exists level 2b, GRADE C evidence to support the consideration of lidocaine for SE and RSE in the pediatric population.

## A.02

### The long-term outcome of children with refractory epilepsy after a vagal nerve stimulator implantation: CHU Sainte-Justine experience

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*Background:* Debate persists in Canada about the cost and benefit of vagal nerve stimulation in patients with refractory epilepsy. The aim of our study was to evaluate the impact of a vagal nerve stimulator on the seizure frequency and the admission rate of children with refractory epilepsies over five years of follow-up. *Methods:* 52 patients were implanted between 2000-2013. Of these, 37 were followed at CHU Sainte-Justine and 21 kept seizure diaries. Seizure frequency was compared to the baseline at 6 months, 12 months, 24 months and 60 months of follow up using a multivariate ANOVA analysis. The hospitalization rate was calculated as the mean difference between the number of hospitalizations prior to and after the implantation. *Results:* Seizure frequency decreased by 58% at 6 months, by 61% at 12 months, by 53% at 24 months and by 63% at 60 months of follow up respectively compared to the baseline ($p< 0.001$). The hospitalization rate decreased by 50.87% after surgery ($p< 0.001$). *Conclusion:* In our population, vagal nerve stimulation has a sustained impact on seizure frequency and hospitalization rates. This supports previous data from our group and others on cost-effectiveness of the technique in children with refractory epilepsy.

## A.03

### Increased focal and diffuse cerebral demand after concussion

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*Aim:* To examine cortical activation during a memory task in children with and without post-concussion symptoms (PCS) following concussion. *Methods:* A case-controlled study within the Play-Game Trial (www.playgametrial.ca). Children (aged 8-18 years) with PCS at 1-month post-injury were eligible. The fMRI task was a working memory task. Pre-processing and single-subject analysis were performed in FSL. Group activation and inter-group difference maps were extracted. *Results:* 11 symptomatic, 12 asymptomatic, and 11 controls without concussion participated. Groups were similar in age (14.9, 14.0, and 13.8yrs; p=0.46), sex (p=0.984) and time post-injury (symptomatic: 37d; asymptomatic 35d; p=0.573). Compared with controls, symptomatic children demonstrated greater activation especially in the bilateral orbito-frontal cortex and cerebellum. A similar, less pronounced pattern was observed in asymptomatic subjects. *Conclusions:* Similar to adult studies, increased network activation may represent decreased "efficiency" and explain the cognitive fatigue in PCS. Further, children who are "asymptomatic" may not yet be fully recovered.

## A.04

### Neonatal hemorrhagic stroke: population-based epidemology

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*Background:* Stroke is a leading cause of perinatal brain injury and cerebral palsy. Term neonatal hemorrhagic stroke (NHS) is a common syndrome with poorly defined epidemiology. We aimed to determine incidence and mechanisms within a large population-based NHS sample. *Methods:* The Alberta Perinatal Stroke Project (APSP), a provincial registry ascertained NHS cases using exhaustive ICD-9/10 code searching (1992-2012, >2400 chart reviews). Prospective cases were captured through the Calgary Pediatric Stroke Program from 2007-2014 (n=387). All NHS cases underwent structured chart review using a data capture form and blinded review

of neuroimaging. Provincial live births were obtained from statistics Canada. Outcomes were extrapolated to the Pediatric Stroke Outcome Measure (PSOM). *Results:* We identified 74 cases: 49 NHS (26 retrospective, 27 prospective), 4 presumed perinatal HS (PPHS), and 21 hemorrhagic transformation (HT) of ischemic injury. Incidence of NHS was 1:8800 live births (1:5820 for all forms). HT was common (28.4%) including global, arterial venous ischemic lesions. Presumed perinatal hemorrhagic stroke presented with epilepsy. No risk factor was identified in 68% of cases. Outcomes were abnormal (PSOM 1 or more) in 30% and better in the HT group. *Conclusion:* NHS occurs in 1:8800 live births. Imaging classification is essential to define mechanisms.

# A.05

## Health-related quality of life in children with Duchenne muscular dystrophy: a follow-up study

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*Background:* Improvement of health-related quality of life (HRQOL) is a major goal in chronic disease management and HRQOL has become an important outcome in clinical trials. Longitudinal data on HRQOL are needed to elucidate change over time and to assess effectiveness of interventions; such research is lacking in the paediatric Duchenne Muscular Dystrophy (DMD) population. *Methods:* We followed up participants from our initial HRQOL study in 2013 a year and a half later. Multidimensional generic and disease-specific measures from the Pediatric Quality of Life Inventory were used to assess HRQOL from child and parent perspectives. Mean changes in HRQOL were calculated. *Results:* Data collection is ongoing and currently, data from 16 families (out of the initial 98) are available. Preliminary results indicated that by both child and parent reports, there were declines in all domains of HRQOL except for social function, in which there was a slight improvement. Mean decline in HRQOL scores ranged from 1.6 to 8.6 for child reports; and 3.3 to 7.7 for parent reports. *Conclusion:* HRQOL of boys with DMD deteriorates over time. Our results may be helpful in interpreting patient reported outcomes in forthcoming clinical trials and determining minimally clinically important changes in this population.

# A.06

## KCNQ2 mutations: genotype-phenotype association beyond epilepsy

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*Background:* KCNQ2 abnormalities were described in infants with benign familial neonatal seizures (BFNS) and epileptic encephalopathy (EE). Associated features possibly include abnormal neuroimaging findings such as hypomyelination and/or T2 high signal of basal ganglia. *Methods:* This report describes 4 infants carrying different heterozygous KCNQ2 variants and 2 infants with 20q13.33 deletions encompassing KCNQ2 gene. *Results:* The different KCNQ2 mutations led to EE in 3 patients and included a novel de novo missense variant, p.Arg201Cys/c.601C>T, in an infant with severe EE and global developmental delay, hyperkinetic movement disorder, autonomic dysfunction with chronic hypoventilation, apnea, low GABA levels in CSF, and hypomyelination. She died at age 3 years of respiratory failure. One patient with BFNS and normal MRI has a previously reported c.508delG frame shift mutation in KCNQ2. Of the two de novo 22q13.33 deletions (1.2Mb versus 254.1 Kb) the larger caused a more severe phenotype, including focal epilepsy from infancy until 4 years, moderate developmental delay and diffuse brain volume loss. *Conclusions:* Along with varied epilepsy phenotypes and neuroimaging findings KCNQ abnormalities were associated with severe autonomic dysfunction and reduced CSF GABA levels. This might have further treatment implications, besides that the altered potassium channel function itself presents a therapeutic target.

# A.07

## Brain stimulation and constraint for hemiparesis after perinatal stroke: The PLASTIC CHAMPS trial

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*Background:* Perinatal stroke causes hemiparetic cerebral palsy. Constraint therapy (CIMT) improves function in congenital hemiparesis and adult stroke. Repetitive transcranial magnetic stimulation (rTMS) may improve function in adult stroke. The two have not been tested in perinatal stroke. *Methods:* PLASTIC CHAMPS (www.clinicaltrials.gov/NCT01189058) was a controlled factorial trial of rTMS and CIMT in perinatal-stroke hemiparesis. Children 6-18 years participated in a 2 week peer-supported motor learning camp, randomized to daily inhibitory rTMS (1200 stimulations, contralesional M1), CIMT, both or neither. Primary outcomes were Assisting Hand Assessment (AHA) and Canadian Occupational Performance Measure (COPM) at 1, 8, and 24 weeks. Quality-of-life, safety and tolerability were evaluated. Change was assessed across treatment groups over time (linear mixed effects model). *Results:* All forty-five subjects completed the trial (median 11.4yrs). COPM scores increased >100% with maximal gains at 6 months ($p<0.002$). Addition of rTMS and/or CIMT doubled the chances of clinically significant gains. Combined rTMS+CIMT resulted in larger AHA gains at all time points (6 months $p=0.006$). CIMT or rTMS alone had more modest effects. Neither treatment decreased function in either hand. Procedures were well tolerated. *Conclusions:* Children with hemiparesis participating in intensive, psychosocial rehabilitation programs perceive marked increases in function. Non-invasive brain stimulation may enhance motor learning therapy in perinatal stroke hemiparesis.