

A.04

Anatomic variation of the Circle of Willis in perinatal stroke

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Background: Perinatal stroke is a common disorder in neonates with unknown etiology. Previous studies have linked anatomic variations in the Circle of Willis to adult stroke. This study aimed to understand the potential relationship between circle anatomy and common forms of perinatal stroke: NAIS, APPIS, and PVI. **Methods:** 94 subjects (62 NAIS/APPIS, and 32 PVI) were identified from the Alberta Perinatal Stroke Project. Inclusion criteria were: MRI-confirmed perinatal stroke, 3D-TOF MRA, and absence of other disorders. Images were classified as complete, incomplete posterior circulation, incomplete anterior circulation, and incomplete anterior and posterior circulation. Fisher Exact Test compared completeness against stroke type and segment absence ipsilateral to stroke. Mann-Whitney U compared completeness and lesion volume. **Results:** Completeness was more common in PVI than NAIS/APPIS ($p=0.500$) and in healthy controls than total stroke population ($p=0.251$). Ipsilesional absent segments were more frequent in NAIS/APPIS ($p=0.270$). NAIS/APPIS patients with complete CoW had larger median lesion volume was compared to those with incomplete circles ($p=0.484$), with contralateral absence ($p=0.943$), and with ipsilateral absence ($p=1.00$). The opposite was found in PVI patients for all lesion volume comparisons ($p=0.321, 0.362, 0.739$ respectively). **Conclusions:** Circle anatomy is highly variable in perinatal stroke. Absence of segments is not associated with stroke type, lesion side, and lesion volume.

A.05

Ten years of experience with lamotrigine for the treatment of neonatal and infantile seizures

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Background: The therapeutic value of second-generation anticonvulsants such as lamotrigine has not been clearly established in neonates and infants with seizures. To address this issue, we assessed the efficacy of lamotrigine for treating neonatal and infantile seizures, detailed the dosing regimens used, and described its tolerability and safety profile. **Methods:** This retrospective study included patients (age 0-12 months) diagnosed with seizures and treated with lamotrigine, as monotherapy or adjunctive therapy, by pediatric neurologists at Centre mère-enfant Soleil du CHUQ from 2004 to 2014. The frequency of seizures and EEG patterns were compared before and after introduction of lamotrigine during the first months of life. Data on initial and maintenance doses, rate and magnitude of dosing increments, and adverse effects were collected. **Results:** Treatment with lamotrigine was initiated in 32 neonates and 13 infants. At first follow-up (mean duration 3 months), 76 % ($n = 34$) showed a significant ($\geq 50\%$) reduction of seizures and 64% ($n = 29$) improvement of EEG pattern compared to baseline. The efficacy in monotherapy and adjunctive therapy was similar. A single case of

cutaneous hypersensitivity reaction requiring cessation of treatment was reported. **Conclusions:** This study suggests that lamotrigine is a useful, safe, and well-tolerated anticonvulsant alternative for the treatment of seizures in neonates and infants.

A.06

Ataluren: an overview of clinical trial results in nonsense mutation Duchenne Muscular Dystrophy (nmDMD)

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Background: Ataluren is the first drug to treat the underlying cause of nmDMD. **Methods:** Phase 2 and 3 studies of ataluren in nmDMD were reviewed, with efficacy and safety/tolerability findings summarized. **Results:** Ataluren nmDMD trials include: a Phase 2a proof-of-concept study ($N=38$); a Phase 2b randomized controlled trial (RCT) ($N=174$); an ongoing US-based open-label safety extension study ($N=108$); an ongoing non-US-based open-label safety/efficacy extension study ($N=94$); and a Phase 3 RCT, ACT DMD ($N=228$), whose primary endpoint was change in six-minute walk distance (6MWD) over 48 weeks. The proof-of-concept study demonstrated increased dystrophin production in post-treatment muscle biopsies from ataluren-treated patients with nmDMD. The Phase 2b results demonstrated an ataluren treatment effect in 6MWD, timed function tests, and other measures of physical functioning. The Phase 3 ACT DMD results demonstrated an ataluren treatment effect in patients with nmDMD in both primary and secondary endpoints, particularly in those with a baseline 6MWD of 300-400m. Ataluren was consistently well-tolerated in all three trials, as well as in the ongoing extension studies. Trial findings will be presented in detail. **Conclusions:** The totality of the results demonstrates that ataluren enables nonsense mutation readthrough in the dystrophin mRNA, producing functional dystrophin and slowing disease progression.

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A.07

Head circumference in preterm neonates: size at birth and postnatal growth predict neurodevelopment at 18 months

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Background: We determined the association between head circumference (HC) at birth and through neonatal intensive care with neurodevelopmental outcome in preterm neonates, accounting for brain injury on MRI. **Methods:** 169 neonates born 24-32 weeks gestation were studied prospectively with serial MRI. HC was measured at birth and discharge from neonatal intensive care. Outcome was assessed at 18 months corrected age using Bayley Scales of Infant & Toddler Development III motor and cognitive scores. Using multivariate linear regressions we evaluated the association between HC

and outcomes, accounting for severity of brain injury and postnatal infection. *Results*: 46 neonates had HC <10th percentile at birth (SHC) which predicted poorer motor (~4 points; $p=0.001$) and cognitive (~4 points; $p=0.005$) outcomes, relative to those with normal HC at birth. In 9 of these neonates, SHC persisted to discharge; they had dramatically lower motor scores (15 points; $p=0.004$) and cognitive scores (12 points; $p<0.001$), even after adjusting for known risk factors. Those born with SHC whose HC normalized by discharge did not show significantly poorer outcomes than those born with normal HC. *Conclusions*: The relationship between small HC at birth and adverse neurodevelopmental outcomes can be attenuated with normalization of head growth through the period of neonatal intensive care.

CNS / CSCN CHAIR'S SELECT ABSTRACTS

B.01

CNS Francis McNaughton Memorial Prize

Predictors of dysphagia screening after acute ischemic stroke: Who gets tested?

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Background: Dysphagia is a devastating complication of stroke and can lead to malnutrition, immobility, aspiration pneumonia, and death. Guidelines advocate screening all patients with acute stroke for swallowing impairment. However, previous research suggests only 60% are screened, and it is unclear what factors contribute to receiving dysphagia screening. *Methods*: We used the Ontario Stroke Registry to identify patients who were admitted to Regional Stroke Centres from 2010-2013. We used multivariable regression to identify predictors of receiving a dysphagia screen within 72 hours. *Results*: Among 7172 patients with acute ischemic stroke, 1705 patients (23.8%) did not undergo screening. Factors increasing the odds of being tested were: Stroke unit admission (adjusted odds ratio aOR 6.5), presenting with speech deficits (aOR 1.9) or weakness (aOR 1.5), or receiving thrombolysis (aOR 1.9). Seizure (aOR 0.49) and mild stroke (aOR 0.59 vs moderate stroke) decreased the odds of being tested. Among those with mild strokes who received a swallowing screen, 33% failed. *All $p<0.0001$. *Conclusions*: Patients with mild stroke are at risk of not being screened for dysphagia, despite a significant fail rate among those tested. This may expose untested patients to a higher risk of complications from dysphagia, and suggests a gap in process of care that should be addressed.

B.02

CSCN Herbert Jasper Prize

Burst-suppression EEG is reactive to photic stimulation in comatose children with acquired brain injury

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Background: Burst-suppression is an electroencephalographic pattern observed during coma and reflects severe encephalopathy. We investigated the reactivity of burst-suppression to photic stimulation in children with acquired brain injury. *Methods*: Intensive care unit electroencephalographic monitoring recordings containing burst-suppression were obtained from 5 comatose children with acquired brain injury of various etiologies. Intermittent photic stimulation was performed at 1 Hz for 1 minute to assess reactivity. We quantified reactivity by measuring the change in the burst ratio (fraction of time in burst) following photic stimulation. *Results*: Photic stimulation evoked bursts in all patients, resulting in a transient increase in the burst ratio, while the mean heart rate remained unchanged. The regression slope of the change in burst ratio, referred to as the standardized burst ratio reactivity, correlated with subjects' Glasgow Coma Scale scores. *Conclusions*: Reactivity of the burst-suppression pattern to photic stimulation occurs across diverse coma etiologies. Standardized burst ratio reactivity appears to reflect coma severity. Measurement of burst ratio reactivity may represent a simple bedside tool to monitor coma severity in critically ill children.

B.03

The Canadian neurology graduate survey

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Background: Planning for neurology training necessitated a reflection on the experience of graduates. We explored practice characteristics, and training experience of recent graduates. *Methods*: Graduates from 2010-2014 completed a survey. *Results*: Response rate was 37% of 211. 56% were female. 91% were adult neurologists. 65% practiced in an outpatient setting. 63% worked in academics. 85% completed subspecialty training (median 1 year). 36% work 3 days a week or less. 82% took general call (median 1 night weekly). Role preparation was considered very good or excellent for most; however poor or fair ratings were 17% in advocacy and 8% in leadership. Training feedback was at least "good" for 87%. Burnout a few times a week or more was noted by 5% (6% during residency, particularly PGY1 and 5). 64% felt overly burdened by paperwork. Although most felt training was adequate, it was poor or fair at preparing for practice management (85%) and personal balance