LAVENDER. Here, we report the safety and efficacy results of LILAC and LILAC-2, open-label extension studies of LAVENDER. Methods: Females with RTT, aged 5–21 years, received twice-daily, oral trofinetide in LILAC for 40 weeks. Participants who completed LAVENDER and LILAC continued trofinetide in LILAC-2, a 32-month extension study. Safety assessments included the incidence of adverse events (AEs). Efficacy endpoints included the Rett Syndrome Behaviour Questionnaire (RSBQ) and the Clinical Global Impression–Improvement (CGI-I) scale. Results: Overall, 154 patients were enrolled in LILAC. The most common AEs were diarrhea (74.7%) and vomiting (28.6%). The mean (standard error [SE]) change from the LAVENDER baseline to Week 40 in the LILAC study in RSBQ was -7.3 (1.62) and -7.0 (1.61) for participants treated with trofinetide and placebo in LAVENDER, respectively. Mean (SE) CGI-I scores compared with the LILAC baseline at Week 40 were 3.1 (0.11) and 3.2 (0.14) for patients treated with trofinetide and placebo in LAVENDER, respectively. Similar safety and efficacy trends were observed in LILAC-2. Conclusions: Trofinetide continued to improve symptoms of RTT in LILAC and LILAC-2 with a safety profile consistent with LAVENDER.

P.046

Review of the management of Wernicke encephalopathy in pediatrics

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doi: 10.1017/cjn.2024.153

Background: Wernicke encephalopathy (WE) is a neurological emergency defined by acute encephalopathy, oculomotor dysfunction, and ataxia. Pediatric cases of WE are underdiagnosed despite having a similar incidence to adults. There are no available treatment guidelines for pediatric WE. Prompt treatment with thiamine can prevent devastating consequences. Methods: A rapid review of the literature of the past 20 years with selected relevant older articles was conducted for the research question "How does child and adolescent thiamine therapy management for Wernicke Encephalopathy compare to adult guidelines?" All articles reporting the investigation, management and treatment of Wernicke encephalopathy – both non alcohol related and alcohol-related pediatric cases – were included. Articles not reporting clinical outcomes were excluded. Results: Eleven case studies including one available review article, met the inclusion and exclusion criteria. An algorithm was created for the organization of published reports of the management of WE for children and adolescents. Key considerations were included for the prevention, identification, acute and ongoing management of patients with WE. Conclusions: The recognition of risk factors for thiamine deficiency and symptoms of acute WE should prompt immediate treatment with thiamine – as a routine and safe therapy in the pediatric population.

P.047

Survey of caregivers of individuals with NBIAs to identify relevant quality of life outcomes

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doi: 10.1017/cjn.2024.154

Background: Neurodegeneration with Brain Iron Accumulation (NBIA) is a heterogenous group of disorders with the common theme of iron accumulation in the basal ganglia. These disorders typically present in childhood with progressive neurodegeneration and neuropsychiatric symptoms. Caring for an individual with NBIAs is intensive, however it is unknown what factors impact caregiver well-being and quality of life. Methods: Common themes were obtained via literature review of quality of life surveys in children with neurological and chronic illnesses. Five domains were addressed: Diagnosis, Communication, Symptom Management, Clinical Experience and Resources/Support. The survey was approved by the Family Advisory Committee at the CHEO Research Institute and the CHEO REB. The survey was distributed via the Rare Connect Platform to Canadian caregivers. Results: Survey responses are being analyzed and will be presented at the CNSF. Within each domain, Likert scales will be analyzed. Domains will be ranked according to the caregiver responses. Conclusions: Results of this survey will assist in developing care management guidelines, resources for families and help with future advocacy for patients and families affected by NBIAs. The results will also help guide future NBIA Canada Family Conferences.

P.048

Prevalence, type and risk factors of intracranial hemorrhage in term neonates: a systematic review and meta-analysis

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doi: 10.1017/cjn.2024.155

Background: Intracranial hemorrhage (ICH) in newborns poses a significant challenge to wellbeing and development. In preterm neonates, germinal matrix hemorrhage is most common. In term neonates, prevalence and type of ICH has not been well elucidated. This systematic review aims to assess prevalence, type, and risk factors of ICH in term neonates. Methods: A systematic review was conducted. Inclusion criteria was ICH in neonates born at 37+ weeks gestation. Exclusion criteria was one type of ICH, one risk factor, sample size <20, text not in English, full text not accessible. Eligible studies were evaluated by two authors, data was extracted and analyzed using a predesigned template and MetaXL. Results: A total of 1226 records were initially identified and 20 studies were included in the final analysis. The overall prevalence of ICH was 9.3%. This was subdivided into an asymptomatic subgroup (5.8%) and symptomatic subgroup (29.3%). Analysis showed CT detected ICH most commonly. Extra-axial hemorrhage was most commonly

detected (~30%), with subdural more common than extradural hemorrhages. The odds of having an ICH was significantly higher with instrumental delivery (3.75%). Conclusions: This shows that prevalence of ICH is relatively high in symptomatic children. Measured prevalence varies according to the type of modality used for screening.

Clinical Neurophysiology (CSCN)

Epilepsy and EEG

P.049

Acetazolamide use for myoclonus: case report of 2 patients with progressive myoclonic epilepsy and literature review

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doi: 10.1017/cjn.2024.156

Background: Cortical myoclonus originates at cerebral cortex, predominantly occurring on voluntary movements. Few case reports described usage of Acetazolamide (ACZ) for myoclonus. Methods: Chart review of 2 patients was performed. Literature review was conducted on myoclonus and ACZ using Pubmed. Results: 22-year-old female was diagnosed with Progressive Myoclonic Epilepsy (PME) secondary to a KCNC1 mutation. Her symptoms started at 10 years old with bilateral tonic clonic seizures (BTCS), later developing progressive ataxia and myoclonus, involving face and limbs, which worsened with stimulus and menses. Medications included Perampanel, Clonazepam and Levetiracetam, however myoclonus was still limiting. At the age of 19, ACZ 250 mg BID was started for 2 weeks around her menses. Follow up revealed significant improvement of myoclonus, resulting in better ambulation, balance and speech, sustained 2.5 years after. 67-year-old male presented BTCS at the age of 53 along with cortical myoclonus, dementia and ataxia, leading to diagnosis of PME with a mutation on IRF2BPL. Improvement of myoclonus occurred with ACZ 250 mg BID biweekly, although balance and cognition still deteriorated. Conclusions: Previous literature outlines 4 cases of action myoclonus that responded to ACZ. We believe that ACZ should be considered to treat myoclonus, especially in cases with cortical involvement and hormonal fluctuations.

P.050

Spike source localizations between the three non-REM sleep stages: resemblances to wakefulness and distinctions from REM sleep

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doi: 10.1017/cjn.2024.157

Background: Sleep-wake states (SWS) affect the expression of interictal epileptiform discharges ("spikes"), which affects resultant source localization calculations used in epilepsy evaluation. We hypothesize that spike localizations from non-REM sleep 1-3 are most concordant with one another. Methods: We used Standardized low-resolution brain electromagnetic tomography (sLORETA) in Curry 8 software to calculate source localization voxels of spikes in N1-3, REM, or wakefulness (W). We assessed voxel concordance between N1-N2-N3/N1-N2-W/N1-N3-W/N2-N3-W/REM-N1-N2/REM-N1-N3/REM-N2-N3/REM-N1-W/REM-N2-W/REM-N3-W. We classified concordances into those containing and not containing a SWS (e.g. N1 vs. not-N1 = N1-N2-N3/N1-N2-W/N1-N3-W/REM-N1-N2/REM-N1-N3/REM-N1-W vs. REM-N2-W/REM-N3-W/REM-N2-N3/N2-N3-W) for comparison. Results: Concordances did not differ for N1-3 or W. However, concordances with REM were lower than those without REM as a fraction of source localization space (median 32.1% vs. 56.1%, $p<0.001$) and cortical grey matter (median 20.4% vs. 27.3%, $p=0.003$). Conclusions: As expected, source localizations from spikes in N1, N2, and N3 did not significantly differ from one another because these three states are constituent members of non-REM sleep. Surprisingly, however, source localizations derived from awake spikes – not a constituent of non-REM sleep – also did not differ. In contrast, REM was most different by reproducibly exhibiting the least three-way concordance. These findings reinforce the unique localizing ability of REM sleep.

Movement Disorders

P.051

Parkinson's disease tremor can show entrainment and distractibility with tapping test

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doi: 10.1017/cjn.2024.158

Background: Electrophysiological tests such as the tapping test are used to distinguish functional and organic tremors, in which patients with functional tremor commonly show entrainment and amplitude reduction (>50% decrease relative to baseline) of contralateral tremor during tapping. While these features are suggested to be specific to functional tremor, the tapping test in Parkinson's disease (PD) tremor has not been tested. Methods: We evaluated 18 PD patients (2F, age 64.17±7.30 [mean±SD] years) with rest and postural tremors using surface electromyography and triaxial accelerometry. Patients were recorded while tapping at 1, 3 and 5 Hz with the contralateral arm at rest or outstretched. Tremor amplitude and frequency were calculated using power spectrum analysis from accelerometer recordings. Results: Reduction of rest tremor amplitude was observed in 3/18 patients during 1 and 3 Hz tapping. Reduction was seen in 3/16 and 1/16 patients with postural tremors at 1 and 3 Hz tapping, respectively. Frequency shifts (>1.5 Hz) were observed in 3/18 rest tremors and 6/16 postural tremors. Seven patients exhibited rest and/or postural tremor entrainment during 3 or 5 Hz tapping. Conclusions: Distractibility and entrainment can be found in PD