international prospective cohort study. We compared workflow times, reperfusion therapy choices, and 90-day modified Rankin scale (mRS) scores. Results: We included 575 patients, mean age 70.2 years (SD: 13.1) and 48.5% female. There were no significant sex differences in onset-to-CT (males: 115 minutes [IQR: 72-171], females: 114 minutes [IQR: 75-196]) or CT-to-thrombolysis time (males: 24 minutes [IQR: 17-32], females: 23 minutes [IQR: 18-36]). However, female participants had a 12-minute faster CT-to-groin-puncture time, p=0.001. Reperfusion therapies did not significantly differ by sex. Reperfusion therapies included thrombolysis alone (males: 46%, females: 49%), EVT alone (males: 34%, females: 34%), thrombolysis plus EVT (males: 8%, females 9%) and conservative management (males: 12%, females: 8%). Median 90-day mRS was 2 (IQR: 1-4) in both males and females, p=0.1. Conclusions: In the INTERRSeCT cohort, rates of reperfusion therapy, workflow times and 90-day outcomes were similar between sexes, suggesting that women are not subject to any poorer performance in key quality indicators for reperfusion treatment for acute stroke.

B.4

Quantitative electroencephalography to predict post-stroke disability: a systematic review and meta-analysis

*I Sood (Ludhiana) R Injety (Ludhiana) A Farheen (Hershey) S Kamali (Los Angeles) A Jacob (Ludhiana) K Mathewson (Edmonton) B Buck (Edmonton) M Kate (Edmonton)**

doi: 10.1017/cjn.2023.86

Background: We aim to assess the role of quantitative electroencephalography (QEEG) derived indices to predict post-stroke disability. Methods: We included observational studies (sample-size≥10) of patients with stroke who underwent EEG and a follow-up outcome assessment was available either in form of a modified Rankin scale (mRS) or National Institute of stroke scale (NIHSS) or Fugl-Meyer scale (FMA). QEEG indices analyzed were delta-alpha ratio (DAR), delta-theta-alpha-beta ratio (DTABR), brain symmetry (BSI) and pairwise derived brain symmetry (pdBSI). Results: Twelve studies (11 had only ischemic stroke, and one had both ischemic and hemorrhagic stroke), including 513 participants were included for meta-analysis. Higher DAR was associated with worse mRS (n=300, Pearson's r 0.26, 95% CI 0.21-0.31). Higher DTABR was associated with worse mRS (n=337, r 0.32, 95% CI 0.26-0.39). Higher DAR was associated with higher NIHSS (n=161, r 0.42, 95% CI0.24-0.6). Higher DTABR was associated with higher NIHSS (n=172, r 0.49, 95% CI 0.31-0.67). pdBSI was inversely associated with FMA (n=20, r-0.50 95% CI -0.86-(-0.14)) and BSI was not associated with FMA (n=21, r -0.3 95% CI -0.81-0.22). Conclusions: QEEG-derived indices have the potential to assess post-stroke disability. Adding QEEG to the clinical and imaging biomarkers may help in better prediction of post-stroke recovery.

PROSPERO 2022 CRD42022292281

Child Neurology (CACN)

C.1

The molecular diagnostic landscape of children with seizure onset in the first three years of life

D McKnight (San Francisco) H McLaughlin (San Francisco) C Grayson (Burnaby) R Sherrington (Burnaby) N Butterfield (Burnaby) S Aradhya (San Francisco) L DeRienzo (Burnaby) A Morales (San Francisco) A Willcock (San Francisco) B Johnson (San Francisco)*

doi: 10.1017/cjn.2023.87

Background: To clarify the landscape of molecular diagnoses (MDs) in early-onset epilepsy individuals, we determined the prevalent MDs stratified by age at seizure onset (SO) and the time to MD in children with SO <36 months of life. Methods: A panel of up to 302 genes associated with epilepsy was utilized and ordering physicians provided the age of SO. Diagnostic yield analyses were performed for SO ages including <1 mo, 1-2 mo, 3-5 mo, 6-11 mo, 12-23 mo, and 24-35 mo. The time to MD (MD age - SO age) was determined for the top 10 genes in each SO category. Results: 15,074 individuals with SO <36 months of life were tested. Predominant MD findings are as follows: KCNQ2 in neonates with SO at <1mo, KCNQ2 and CDKL5 for SO between 1-2 mo, PRRT2 and SCN1A for SO between 3-11 mo, and SCN1A for SO between 12-36 months. The median time to MD varied by gene. For example, there was no delay in the median time to MD for the GLDC, KCNQ2, and SCN2A genes while the median delay for MECP2, SLC2A1, and other genes was ≥ 12 months. Conclusions: These data highlight the importance of comprehensive early testing in children with early-onset epilepsy.

C.2

SUNFISH parts 1 and 2: 4-year efficacy and safety data of risdiplam in types 2 and 3 spinal muscular atrophy (SMA)

JK Mah (Calgary) M Oskoui (Montreal) JW Day (Palo Alto) N Deconinck (Brussles) E Mazzone (Rome) A Nascimento (Barcelona) K Saito (Tokyo) C Vuillerot (Lyon) G Baranello (London) O Boespflug-Tanguy (Paris) N Goemans (Leuven) J Kirschner (Freiburg) A Kostera-Pruszczyk (Warsaw) L Servais (Oxford) J Braid (Welwyn Garden City) M Gerber (Basel) K Gorni (Basel) C Martin (Welwyn Garden City) W Yeung (Welwyn Garden City) RS Scalco (Basel) E Mercuri (Rome)*

doi: 10.1017/cjn.2023.88

Background: SMA affects individuals with a broad age range and spectrum of disease severity. Risdiplam (EVRYSDI®) is a centrally and peripherally distributed, oral SMN2 pre-mRNA splicing modifier. Methods: SUNFISH is a multicenter, two-part, randomized, placebo-controlled, double-blind study in patients with Types 2/3 SMA. Part 1 assessed the safety, tolerability and

pharmacokinetics/pharmacodynamics of different risdiplam dose levels in patients with Types 2/3 SMA. Part 2 assessed the efficacy and safety of the selected dose of risdiplam versus placebo in Type 2 and non-ambulant Type 3 SMA. In Part 2, participants were treated with risdiplam or placebo for 12 months, then received risdiplam in a blinded manner until month 24. At month 24, patients were offered the opportunity to enter the open-label extension phase. Results: Change from baseline in MFM32 total score (Part 2- primary endpoint) in patients treated with risdiplam versus placebo was met at month 12. These increases in motor function were sustained in the second and third year after risdiplam treatment. Here we present 4-year efficacy and safety data from SUNFISH. Conclusions: SUNFISH is ongoing and will provide further long-term efficacy and safety data of risdiplam in a broad population of individuals with SMA.

C.3

Development and validation of a prediction model for perinatal arterial ischemic stroke in term neonates

R Srivastava (Edmonton) M Dunbar (Calgary) M Shevell (Montreal) M Oskoui (Montreal) A Basu (Newcastle upon Tyne) M Rivkin (Boston) E Shany (Beer-Sheva) L de Vries (Utrecht) D Dewey (Calgary) N Letourneau (Calgary) MD Hill (Calgary) A Kirton (Calgary)*

doi: 10.1017/cjn.2023.89

Background: Perinatal arterial ischemic stroke (PAIS) is a focal brain injury in term neonates, identified postnatally but presumed to occur around birth. Early risk detection and targeted treatments are limited. We developed and validated a diagnostic risk prediction model from common clinical factors to predict a term neonate's probability of PAIS. Methods: A diagnostic prediction model was developed using multivariable logistic regression. Common pregnancy, delivery, and neonatal clinical factors were collected across four registries. Variable selection was based on peer-reviewed literature. Participant inclusion criteria were term birth and no underlying predisposition to stroke. The primary outcome was discriminative accuracy of the model predicting PAIS, measured by the concordance (C-) statistic. Results: 2571 participants (527 cases, 2044 controls) were eligible for analysis. Nine variables were included in the model – maternal age, tobacco exposure, recreational drug exposure, pre-eclampsia, chorioamnionitis, maternal fever, emergency c-section, low 5-minute Apgar score, and sex – to predict the risk of PAIS in a term neonate. This model demonstrated good discrimination between cases and controls (C-statistic 0.73) and model fit (Hosmer-Lemeshow p=0.20). Conclusions: Clinical variables can be used to develop and internally validate a model of PAIS risk prediction. Identifying high-risk neonates for early screening and treatment could reduce lifelong morbidity.

C.4

Understanding the role of deep brain stimulation for Refractory Status Dystonicus in children: case series and systematic review

LM Vogt (Toronto) H Yan (Toronto) B Santyr (Toronto) GM Ibrahim (Toronto) C Gorodetsky (Toronto)*

doi: 10.1017/cjn.2023.90

Background: Status dystonicus (SD) is a life-threatening form of dystonia with limited treatments available. We sought to better understand the processes, outcomes, and complications of deep brain stimulation (DBS) for pediatric SD through a systematic review alongside an institutional case series. Methods: Data regarding treatment, stimulation parameters, dystonia severity and outcomes was collected for the case series (n=7) and systematic review (n=70, conducted in accordance with PRISMA guidelines). This was analysed descriptively (rates, outcome measures). For the case series we created probabilistic voxel-wise maps for improvement in dystonia based on brain region stimulated. Results: All patients in our case series and > 95% of patients in the systematic review had resolution of SD with DBS, typically within 2-4 weeks. Most patients in the review (84%) and all patients in the case series had DBS implanted to the globus pallidus internus. In terms of dystonia severity scores, there was a mean improvement of 25% (case series) or 49% (systematic review). Reported mortality was 4% in the systematic review. Conclusions: DBS for pediatric SD is feasible and safe. It allows for increased survival as well as quality of life - however risks still exist. More work is needed to determine timing, eligibility, and stimulation parameters.

C.5

Highlighting a novel, stepwise pathway for the in-hospital management of children with acutely worsening dystonia

LM Vogt (Toronto) K Yang (Toronto) G Tse (Stanford) C Gorodetsky (Toronto)*

doi: 10.1017/cjn.2023.91

Background: Dystonia is common in children with acquired and inherited neurological disorders. Status dystonicus (SD) is the most severe form of dystonia that can lead to life-threatening complications if not treated promptly. We identified a local provider knowledge gap in the acute management of dystonia, leading to uncertainty and delays in care. To our knowledge, no in-hospital clinical pathway exists for the ward-based management of acute dystonia. We hypothesized that a stepwise clinical pathway would standardize and improve comfort in managing hyperacute dystonia. Methods: We formed a multidisciplinary working group and developed a pathway based on literature review and expert consensus. Aims of the pathway included: