in a non-invasive and objective manner. *Methods:* SsVEPs were obtained in ten children with OPGs and 42 controls ages 3 to 21. The stimuli consisted of two circular dartboard patterns stimulating fovea and peripheral zones at two flickering frequencies, so that central and peripheral visual fields could be assessed simultaneously. The test consisted of eight stimuli presentations of 10 seconds. *Results:* Results indicate significantly lower ssVEP amplitudes in children with OPGs (M = 2.52, 95% CI [1.13, 3.92]) compared to controls (M = 13.26, 95% *CI* [8.85, 17.67]) in the central visual field (p = .021). However, no between group differences were detected in the peripheral field (p >.05). There were no significant differences between age groups (p > 0.05). *Conclusions:* This objective, affordable, and non-invasive method appears to be effective in detecting central visual field deficits in children with OPGs rapidly and consistently.

A.03

Analyses of surgical and MRI factors associated with cerebellar mutism

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Background: The surgical risk factors and neuro-imaging characteristics associated with cerebellar mutism (CM) remain unclear and require further investigation. We aimed to examine surgical and MRI findings associated with CM in children following posterior fossa tumor resection. *Methods:* Using our data registry, we retrospectively collected data from pediatric patients who acquired CM and were matched based on age and pathology type with patients not acquiring CM after posterior fossa surgery. The strength of association between surgical and MRI variables and CM were examined using odds ratios (ORs) and corresponding 95% confidence intervals (CIs). *Results:* A total of 22 patients were included. Medulloblastoma was the most common pathology among CM patients (91%). Tumor attachment to the floor of the fourth ventricle (OR, 6; 95% CI, 0.7-276), calcification/hemosiderin deposition (OR 7; 95% CI 0.9-315.5), and post-operative peri-ventricular ischemia on MRI (OR, 5; 95% CI, 0.5-236.5) were found to have the highest association with CM. *Conclusions:* Our results may suggest that tumor attachment to the floor of the fourth ventricle, pathological calcification, and post-operative ischemia are relatively more prevalent in patients with CM. Collectively, our work calls for a larger multi-institutional study of CM patients to further investigate the determinants and management of CM to potentially minimize its development and predict onset.

A.04

Functional investigations of CIC and ATXN1L in Oligodendroglioma

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Background: Oligodendroglioma (ODG), a molecularly defined subtype of glioma, is a treatment responsive, slow growing tumour strongly associated with IDH mutation and 1p19q co-deletion. Mutations in *Capicua* (*CIC*), located on chromosome 19q, have been found in up to 70% of IDH mutated, 1p19q co-deleted ODGs; suggesting that loss or altered function of CIC may be crucially associated with ODG's unique biology. CIC and ATXN1L have previously been implicated in neurodegeneration, however, this interaction has not been studied in cancer. *Methods:* Transcriptome profiling of CIC knockout HEK293 cell lines generated using CRISPR was performed using microarray. CIC and ATXN1L interaction was confirmed using immunoprecipitation and immunofluorescence. Transcript and protein changes of CIC targets were tested using RT-qPCR and Western blot following ATXN1L siRNA knockdown. *Results:* Transcriptomic profiling of CIC knockout cell lines resulted in a list of candidate CIC target genes validated against clinical samples. Immunoprecipitation and immunofluorescence confirmed CIC and ATXN1L interaction. Derepression of candidate CIC targets at transcript and protein levels was seen upon siRNA knockdown of ATXN1L. *Conclusions:* The interaction between CIC and ATXN1L is necessary for the repression of CIC target genes, including known oncogenes. Further research into the relationship between CIC and ATXN1L may lead potentially novel avenues of therapeutic approaches for less favorable gliomas.

A.05

An epidemiologic study of SLC52A2-related Riboflavin Transport Deficiency

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Background: Riboflavin transporter deficiency (RTD), formerly known as Brown-Vialetto-van Laere syndrome, is an early-onset neurodegenerative disorder with distinctive phenotypes. RTD is caused by mutations in either the *SLC52A2* or *SLC52A3* genes that encode riboflavin transporters RFVT-2 and RFVT-3, respectively. *Methods:* This was a 3-year retrospective case review from the Cure RTD International Registry. *Results:* 73 individuals (~60% female, 14 deceased) from 56 families had genetically confirmed RTD Type 2, including 30 novel *SLC52A2* mutations (24 missense, 2 nonsense, 4 deletion). The mean ages at symptom onset and at diagnosis were 2.4 years (SD 1.5, range 0.25 – 8, n=63) and 12.0 years (SD 10.2, range 0.75 – 52, n=56) respectively. Most common presenting symptoms were sensory ataxia (n=43), sensorineural hearing loss (n=22), nystagmus/visual loss secondary to optic atrophy (n=14), upper limb weakness (n=11), and respiratory insufficiency (n=9). Treatment included high dose riboflavin, other supplements, and supportive care; 7 individuals required transfusions for anemia pre-riboflavin treatment and 17 (25%) received a cochlear implant. The minimum prevalence of RTD was estimated to be 1 per million, with >100 new

cases each year. *Conclusions:* This is the largest case series of RTD to date. Early recognition and prompt riboflavin treatment is essential for survival and optimal outcome.

A.06

Selective amygdalohippocampectomy in pediatric medically refractory temporal lobe epilepsy yields worse seizure outcomes

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Background: Selective amygdalohippocampectomy (SAH) is a surgical option in well-selected cases of pediatric medically refractory temporal lobe epilepsy (TLE). The objective of this study was to compare the surgical outcome and the rate of reoperation for ongoing or recurrent seizures between SAH and anterior temporal lobectomy (ATL) in pediatric TLE. *Methods:* Retrospective review of 78 pediatric intractable TLE patients referred to the Comprehensive Epilepsy Program at our institution between 1988 and 2015 treated initially with either a trans-middle temporal gyrus SAH (19) or ATL (59). Patients underwent baseline long-term video electroencephalography and 1.5-Tesla MRI. Neuropsychological testing was performed preoperatively and 12-months postoperatively (including reoperations). *Results:* The mean follow-up was 64 months (range, 12-186 months). The average age at initial surgery was 10.6 ± 5 years with an average delay of 5.7 ± 4 years between seizure onset and surgery. Ultimately 78% were seizure-free (61/78) at most recent follow-up. Seizure freedom after initial surgical treatment was achieved in 81% of patients who underwent ATL (48 patients) versus 42% in SAH (8 patients; $p<0.001$). Of patients with ongoing disabling seizures following SAH, reoperation (ATL) was offered in 8 resulting in seizure freedom in 63%, without interval neuropsychological decline. *Conclusions:* SAH amongst well-selected pediatric TLE results in significantly worse seizure control compared with ATL.

A.07

Genomics of atypical dyskinetic cerebral palsy – opportunities for improved diagnosis and management

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Background: Cerebral palsy (CP) is a debilitating disorder (1). Based on neuromotor impairments it is divided to spastic, dyskinetic and ataxic types (2). Inborn Errors of Metabolism (IEMs), monogenic and chromosomal disorders mimic CP (3). We aimed to identify causal genetic variants in patients with atypical dyskinetic CP in whom known IEMs were ruled out. Timely diagnosis is essential for proper management, especially in conditions that mimic CP and are treatable. *Methods:* We enrolled 23 patients with unexplained atypical dyskinetic CP, for whole exome sequencing. Variants were filtered against public and in-house databases to identify variants predicted as damaging (*in silico* tools and ACMG criteria). We applied a virtual gene panel of known and suspected CP and movement disorder genes and investigated each sample. *Results:* The participants presented with symptoms including: spasticity, dystonia, choera-athetosis, ataxia and cognitive delays. We identified 23 diagnoses: 13 dominant,6 recessive and 4 X-linked. 12 patients had movement disorders. In 4, the diagnoses enabled targeted treatment (neurotransmitter supplements in Unverricht Lundborg diseases (CSTB) and PAK3 deficiency, deep brain stimulation in GNAO1 deficiency, medical diet in Glutaric Aciduria (GCDH). *Conclusions:* Whole Exome Sequencing contributes to establishing diagnosis in patients with atypical dyskinetic CP resulting in precision medicine and improved health outcomes.

CNS/CSCN CHAIR'S SELECT ABSTRACTS

B.01

Impact of work-hours and sleep on well-being and burnout for physicians-in-training: the prospective RATE Study

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Background: Wearable activity trackers are an innovative tool for measuring sleep and physical activity. The Resident Activity Tracker Evaluation (RATE) is a prospective observational study evaluating the impact of work-hours, sleep, and physical activity on resident well-being, burnout, and job satisfaction. *Methods:* Residents were recruited from: 1. general surgery and orthopedics (SURG), 2. internal medicine and neurology (MED) and 3. anesthesia and radiology (RCD). Groups 1 and 2 do not enforce on-call duration restrictions and group 3 had 12-hour restricted-call durations (RCD). Participants wore FitBit activity trackers for 14 days and completed four validated surveys assessing self-reported health, sleepiness, burnout, and job satisfaction. *Results:* Fifty-nine residents completed the study. 778 days of activity and 244 on-call periods were tracked. Surgical residents worked 24 more hours per week than non-surgical residents (84.3 vs 60.7). Surgical residents had 7 less hours of sleep per week and reported significantly higher Epworth Sleepiness scores. Nearly two-thirds of participants (61%) scored high burnout on the Maslach depersonalization subscore. Total steps per day and self-reported well-being, burnout, and job satisfaction were comparable between the groups. *Conclusions:* Despite a positive correlation between work-hours and sleepiness, burnout and well-being were similar among residents. Physical activity did not prevent burnout. These findings are relevant to work-hours policies.