

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

A Sergeant (Hamilton) MM Kameda-Smith (Hamilton)* B Manoranjan (Hamilton) B Kumar (Hamilton) J Duckworth (Hamilton) T Petrelli (Hamilton) K Savage (Hamilton) O Ajani (Hamilton) B Yarascavitch (Hamilton) C Samaan (Hamilton) K Scheinmann (Hamilton) C Alyman (Hamilton) S Almenawer (Hamilton) F Farrokhvar (Hamilton) AJ Fleming (Hamilton) SK Singh (Hamilton) N Stein (Hamilton)*

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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

D Wong (Vancouver) V LeBlanc (Vancouver) S Chittaranjan (Vancouver) S Chan (Vancouver) J Song (Vancouver) M Lee (Vancouver) M Marra (Vancouver) S Yip (Vancouver)*

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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

JK Mah (Calgary) M Menezes (Sydney) K Massey (Calgary)*

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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