inary clinical activity in an adult phase 1 study. Methods: Five children with progressive/refractory CNS tumors harboring an *FGFR* gene alteration following prior therapy were treated with Debio1347 at Memorial Sloan Kettering Cancer Center on single patient use protocols. Patients were treated using the 20 mg tablet formulation at the adult recommended phase 2 dose (80 mg/1.73 m2 * BSA once daily). Toxicities were graded using CTCAEv5.0 and imaging response assessments were performed every 8–12 weeks. RESULTS All AEs were grade 1–2. Most common treatment-related adverse events were hyperphosphatemia, ALT increased and hypoalbuminemia (4 patients). Two patients met criteria for partial response and two patients had stable disease. A 13 month-old patient with a spinal cord high-grade glioma harboring two *FGFR1* mutations (*V592M*, *K687E*) had tumor reduction of 91.7% maintained for 12 months. A 26-month-old patient with a pilomyxoid astrocytoma harboring an *FGFR1-TACC1* fusion had a tumor reduction of 74.5% maintained for 9 months. Molecular characterization of recurrent tumor from this patient demonstrated an *NF1* deletion as a novel molecular mechanism of acquired resistance to FGFR inhibition. Prolonged disease stabilization was noted in an eight year-old patient with metastatic suprasellar pilomyxoid astrocytoma harboring an *FGFR1* mutation (9 months) and in a 14 year-old patient with posterior fossa glioneuronal tumor harboring an *FGFR3-TACC3* fusion (24 months and ongoing). Conclusions: Debio1347 demonstrated tolerable toxicity and promising antitumor efficacy in pediatric patients with refractory FGFR altered gliomas. Specific attention to growth velocity and clinical symptoms with incorporation of imaging assessment of bone growth is warranted. Candidate biomarkers (*FGFR1 V592M* and *K687E* SNVs, *FGFR-TACC* fusions) may guide patient selection. Further studies in this population are warranted.

EPCT-11. RURALITY INDEX SCORE AND PEDIATRIC NEURO-ONCOLOGICAL OUTCOME IN ONTARIO

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Introduction: Rapid access to neurosurgical decisions and definitive management are vital for the outcome of neurocritical patients. There are increased challenges of providing services and to maintain critical infrastructure for rural citizens. The relationship between rurality, marginalization and health outcomes has been identified as associated with higher mortality rates and higher rates of many diseases[G1]. Methods: Employi ng linked administrative databases, we retrospectively analyzed a population based cohort of patients diagnosed with a pediatric brain tumour between 1996 to 2017 in Ontario, Canada. The Ontario Marginalization Index was employed as a surrogate for rurality providing an overall Rurality Index for Ontario (RIO) in addition to the 2016 Ontario Marginalization Index (ON-MARG). Results: Of 1457 patients included, 54.0% were male, 277 of whom were diagnosed in infancy (i.e., < 3 years of age). Income quintile was evenly distributed with 11.5% classified as living in a rural area of Ontario. The median[G2] distance to the nearest pediatric neurosurgical hospital was 59.6km. The rurality index score (RIO) was 0 in 38.8% of children with the majority of patients with a RIO score of <39. The ON-MARG identified 51.9% of patients living in communities with low concentration of individuals without income from employment. A higher RIO score was not a significant factor (Continuous p=0.092/Ordinal p=0.20) associated with length[G3] of follow up, indicating rurality was not a significant factor for determining compliance to[G4] clinical follow-up. However, a trend towards reduced follow-up compliance in the higher RIO score cohort was identified. Conclusion: Rurality and social determinants of health of the region pediatric neuro-oncological patients reside were not associated with patient outcome but a trend towards lower follow-up compliance was identified when children were from regions with RIO>39. Implementation of telehealth follow-up for these patients may overcome barrier to clinical follow-up.[G5]

EPCT-12. NATIONAL MULTICENTERED RETROSPECTIVE REVIEW OF DEMOGRAPHIC, TUMOUR AND INTRAOPERATIVE FEATURES ASSOCIATED WITH THE DEVELOPMENT OF CEREBELLAR MUTISM AFTER PEDIATRIC POSTERIOR FOSSA TUMOUR RESECTION

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Background: Cerebellar mutism (CM) is a condition characterized by a significant lack or loss of speech in children following posterior fossa (PF) surgery. The biological origin of CM remains largely unclear and remains the subject of ongoing debate. Despite multidisciplinary rehabilitative interventions, the outcome is less favorable than initially described. Given the treatment refractory nature of CM, central to its management is prevention. Methods: A national multi-centered retrospective review of all the children undergoing posterior fossa resection at 4 Canadian academic pediatric institutions was undertaken. Patient, tumour, surgical features suggested to be associated with the post-operative development of CM were reviewed to identify pre-operative and intra-operative factors that may predict post-operative CM occurrence. Results: 258 pediatric patients were identified after posterior fossa lesion resection. Mean age at surgery was 6.74 years (SD 4.60) and 42.2% were female. Frozen section was available in 90.3% of cases. The majority of final tumour histology was medulloblastoma (35.7%), pilocytic astrocytoma (32.6%), ependymoma (17.1%) and exophytic glioma (1.2%). Intra-operative impression of adherence to the floor of the 4th ventricle was negative in 47.7%, positive in 36.8% of cases. The extent of resection assessed intraoperatively as gross total resection was 69.8% of cases. Intra-operative abrupt changes in blood pressure and/or heart rate was identified in 19.4% and 17.8% of cases. CM was experienced in 19.5% of patients (N=50), with the majority of cases identified by post-operative day 7. The clinical resolution of CM as mainly assessed by a neurosurgeon (86%) and was complete, significantly resolved, slight improvement, no improvement or deterioration in 56.0%, 8.0%, 20.0%, 14.0%, 2.0% respectively. Conclusion: As a devastating surgical complication, identifying and understanding the biological origin of CM is the first step to complication avoidance. Maximal safe resection irrespective of intra-operative pathology remains the goal to avoid the devastating complication of CM.

EPCT-13. SINGLE INSTITUTION RETROSPECTIVE ANALYSIS OF TUMOR MUTATIONAL BURDEN AND SURVIVAL IN PEDIATRIC BRAIN TUMORS

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Tumor mutational burden (TMB) has been studied across numerous cancer types as a means of risk stratification. To examine the prognostic relevance of TMB to pediatric central nervous system (CNS) tumors, we conducted a retrospective analysis of patients at Albany Medical Center diagnosed from 2012 to present. Patients were <21 at diagnosis, had a primary CNS tumor and available genomic data. Forty-seven patients were included – 22 low-grade gliomas, 10 high-grade gliomas, 5 medulloblastomas, 3 ependymomas, 2 choroid plexus carcinomas, and 5 other CNS tumors, with a median follow up of 36 months, median age at diagnosis 10 (1–19), and 47% female. Median TMB was 1 mutation per megabase (mut/mb); range 0–6. Nine patients did not have available TMB data. Twenty-seven patients had driver mutations and other alterations implicated in cancer development including, including *BRAF-KIAA1549 fusion* (n=6), *NF1 loss* (n=5), *FGFR1 amplification* (n=4), *TP53 inactivation* (n=4), *BRAF V600E mutation* (n=3), and *H3F3A K28M mutation* (n=3). Patients with low TMB (<3 muts/mb; n=24) versus high TMB (≥3 muts/mb; n=14) had a survival of 87% versus 71%, respectively, at last follow-up. Of note, all but one patient in the low TMB cohort had localized disease at diagnosis versus three in the high TMB cohort. High TMB was more prevalent in high- (45%, 9/20) versus low-grade histologies (22%, 4/18). Patients with *BRAF* alterations had LGGs and low TMB (0–1 muts/mb) with all patients surviving at last follow up. Of the eight deaths observed (median 18 months from diagnosis) TMB was high in 4, low in 3, and unknown in 1; all had high-grade histology. Although limited, our data suggests higher TMB may be associated with worse outcome. This analysis will be expanded via a multi-institutional review of TMB and genomic alterations in pediatric CNS patients to better identify high-risk patients requiring alternative treatment strategies.

EPCT-14. GD2 CAR T-CELLS MEDIATE CLINICAL ACTIVITY AND MANAGEABLE TOXICITY IN CHILDREN AND YOUNG ADULTS WITH H3K27M-MUTATED DIPG AND SPINAL CORD DMG

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Background: We previously discovered high expression of the disialoganglioside GD2 on H3K27M+ gliomas and demonstrated preclinical efficacy of intravenous (IV) GD2-targeted chimeric antigen receptor (CAR) T-cells in preclinical models of H3K27M-mutated diffuse intrinsic pontine glioma (DIPG) and diffuse midline gliomas (DMGs). We are now conducting a Phase I clinical trial (NCT04196413) of autologous GD2-targeting CAR T-cells for H3K27M+ DIPG and spinal cord DMG. Here we present the