Low dose Poly-ICLC (Polyinosinoc-Polyriylic acid stabilized with polylysine and carboxymethylcellulose) has a direct immune enhancing action independent of interferon, including increased antibody response to antigen, and NK cell, T-cell, macrophage and cytokine activation. METHODS: We conducted an immunotherapy trial to evaluate the effect of Poly-ICLC in the children with recurrent or progressive low grade gliomas. Criteria for enrollment: progression by MRI and or visual deterioration. Patients received Poly-ICLC 20mcg/kg/dose IM injection once a week for up to 24 months. RESULTS: 23 patients enrolled, 22 evaluable. Ages 2-20 years. Ten females and 13 males. Location: suprasellar (11), spine (1), disseminated (4), others (7). Six patients with NF-1. Pathology: JPA (11), pilomyxoid (5), no pathology (3), Diffuse Astrocytoma, gangliogioma, Glioneuronal tumor, LGG NOS (1 each). Follow-up range 3 to 58 months. Responses: 43% stable disease, 17% (4/23) partial responses- 3 with NF-1. Visual fields and acuity improved in 33% of patients with NF-1. Two JPA patients that have completed 24 months of therapy: one non-NF with SD and PFS 58 months, one NF-1, PR, PFS for 28 months. Most common toxicities included: fevers, discomfort at the injection site, Grade 1 neurotoxicity, hypopontnea, hypophosphatemia, ALT elevation. One case of grade 4 intratumoral hemorrhage, required medical management alone. No patient required transfusion or admission for fever and neutropenia. No reports of neuropathy. CONCLUSION: Poly-ICLC is well tolerated, minimal toxicity, radiographic and clinical responses were demonstrated in patients with LGG. Larger clinical trials with Poly-ICLC are being designed.

LGG-59. REMARKABLE OBJECTIVE RESPONSE AND FAVORABLE SURVIVAL FOR BRAF-V600E CHILDHOOD LOW-GRADE GLIOMAS TO BRAF INHIBITORS COMPARED TO CONVENTIONAL CHEMOTHERAPY
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Molecular characterization of pediatric low grade glioma (pLGG) over the last decade has identified recurrent alterations, most commonly involving BRAF, and less frequently other pathways including MYB and MYBL1. Many of these molecular markers have been exploited clinically to aid in diagnosis and treatment decisions. However, their frequency and their prognostic significance remain unknown. Further, a significant portion of cases do not have any of these alterations and what underlies these cases is also unknown. To address this we compiled a cohort of 562 patients diagnosed at SickKids from 1990-2017. We identified molecular alterations in 45% (81%) of the cohort. The most frequent events were those involving BRAF (either fusions (most common with KIAA1549 (30%)), or V600E mutations (17%) and NF-1 (22%). Less frequently, we identified recurrent FGFR1 fusions and mutations (3%), MYB/MYBL alterations (2%), H3F3A_K27M mutations (2%) and IDH1_R132H (0.5%), as well as other novel rare events. Survival analysis revealed significantly better progression-free survival (PFS) and overall survival (OS) of BRAF-KIAA1549 patients compared to BRAF-V600E with 10-year OS 97.7% (95%CI 95.5-100) and 83.9% (95%CI 72.5-95.6) respectively. In addition to survival, the molecular alterations predict differences in response to conventional therapeutics; BRAF fused patients show a 46% response-rate, versus only 14% in V600E patients. pLGG harboring H3F3A_K27M progressed early with median PFS of 11 months. In patients with MYB/MYBL1, FGFR1/FGFR2 alterations, we observed only one death (FGFR1 N546K case). The work here represents the largest cohort of pLGGs with molecular profiling and their impact on the clinical behaviour of the disease.

LGG-60. THE GENETIC LANDSCAPE OF PEDIATRIC LOW-GRADE GLIOMAS: INCIDENCE, PROGNOSIS AND RESPONSE TO THERAPY
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INTRODUCTION: The risk of seizures varies between 60% and 100% among low-grade gliomas. The frequency of seizures differs widely according to tumor type. OBJECTIVE: To evaluate the clinical and histopathological features of patients with low grade gliomas. METHODS: We conducted a retrospective review of patients who underwent initial surgery for low grade glioma in a pediatric institution, between 2010 and 2017. Some variables are analyzed such as: age, sex, tumor location, histopathological findings, clinical characteristics of the seizures, surgical treatment, anti epileptic drugs. RESULTS: The main diagnoses were: desembrioplastic astrocytoma (4%), oligodendroglialoma (0.5%), as well as other novel rare events. Survival analysis revealed significantly better progression-free survival (PFS) and overall survival (OS) of BRAF-KIAA1549 patients compared to BRAF-V600E with 10-year OS 97.7% (95%CI 95.5-100) and 83.9% (95%CI 72.5-95.6) respectively. In addition to survival, the molecular alterations predict differences in response to conventional therapeutics; BRAF fused patients show a 46% response-rate, versus only 14% in V600E patients. pLGG harboring H3F3A_K27M progressed early with median PFS of 11 months. In patients with MYB/MYBL1, FGFR1/FGFR2 alterations, we observed only one death (FGFR1 N546K case). The work here represents the largest cohort of pLGGs with molecular profiling and their impact on the clinical behaviour of the disease.

LGG-62. SEIZURES IN LOW-GRADE GLIOMAS: CLINICAL AND HISTOPATHOLOGICAL CHARACTERISTICS
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INTRODUCTION: The risk of seizures varies between 60% and 100% among low-grade gliomas. The frequency of seizures differs widely according to tumor type. OBJECTIVE: To evaluate the clinical and histopathological features of patients with low grade gliomas. METHODS: We conducted a retrospective review of patients who underwent initial surgery for low grade glioma in a pediatric institution, between 2010 and 2017. Some variables are analyzed such as: age, sex, tumor location, histopathological findings, clinical characteristics of the seizures, surgical treatment, anti epileptic drugs. RESULTS: The main diagnoses were: desembrioplastic tumor, gangliogioma, astrocytoma, all as WHO grade I, most of them with seizures as a main symptom, tumors involving the frontal, temporal, and parietal lobes with partial resection or biopsies. And epilepsy that merits treatment with up to two anticonvulsants during follow-up. DISCUSSION: Tumor location and histology influence the risk of epilepsy. Glioneuronal tumors are typically associated with chronic drug-resistant epilepsy in up to 90% -100% of patients. Total macroscopic resection is the strongest predictor of the absence of attacks in addition to clinical factors, such as duration of preoperative attacks, type and control with antiepileptic drugs. CONCLUSION: The management of seizures represents an important aspect of the low grade gliomas, since tumor-related epilepsy affects patients’ quality of life, causes cognitive deterioration, and may result in significant morbidity.