cording to the surgical concept involved, the patients were divided into three subgroups-complete en bloc concept, partial en bloc concept and piecemeal concept. The matching-comparison(en bloc group consisting of the first two subgroups vs. piecemeal group) was conducted for the en bloc concept on the short-term outcomes. Then the patient data after January 2018, when the en bloc concept was routinely integrated into brain tumor surgery in our medical center, were reviewed and analyzed to find out on brain tumor, in childhood. Developmental gene expression data supported by neurolological studies suggest that Wingless (WNT)-MB originate from the lower rhombic lip (LRL), Sonic-Hedgehog (SHH)-MB from the cerebellar hemispheres, and Group 3 and Group 4 MB from the cerebellar vermis. However, there is still insufficient evidence from a neurosurgical perspective supporting this proposed concept. METHODS: Clinical and molecular data from patients aged under 18 years at time of diagnosis were evaluated for a distant intraventricular recurrence. A total endoscopic resection was performed with complete removal of the tumor, no complications, and no perioperative MRI. RESULTS: In the en bloc group, the perioperative outcomes, including hospital stay (p=0.001), PICU stay (p=0.003), total blood loss (p=0.015), transfusion rate (p=0.005) and complication rate (p=0.039), were all significantly improved. The univariate and multivariate regression analysis showed that tumor volume, bottom vessel, and imaging features, like encasing nerve or pass-by vessel, finger-like attachment, ratio of “limited line” and ratio of “clear line” remained independent predictors for the application of en bloc concept in our medical center. CONCLUSION: The application of en bloc concept based on the imaging features can improve the short-term outcomes.

SURG-02. THE SITE OF ORIGIN OF MEDULLOBLASTOMA: DOES THE NEUROSURGICAL PERSPECTIVE SUPPORT THE CURRENT CONCEPT FROM MOLECULAR DATA? Olga Giobanu-Carasu\(^1\), Anna Cho\(^1\), Gregor Kasprian\(^2\), Andreas Peryl\(^1\), Christine Haberl\(^3\), Irene Slavc\(^1\), Josef Haberl\(^1\), Karl Ciobanu-Carausu\(^1\), Department of Neurological Surgery, Weill Cornell Medicine, New York, NY, USA. 2Department of Pathology and Laboratory Medicine, Weill Cornell Medicine, New York, NY, USA. 3Department of Neurology, Division of Neuropathology and Neurochemistry, Medical University of Vienna, Vienna, Austria.

BACKGROUND: Medulloblastoma (MB) are the most common malignant brain tumor in childhood. Developmental gene expression data supported by neurolological studies suggest that Wingless (WNT)-MB originate from the lower rhombic lip (LRL), Sonic-Hedgehog (SHH)-MB from the cerebellar hemispheres, and Group 3 and Group 4 MB from the cerebellar vermis. However, there is still insufficient evidence from a neurosurgical perspective supporting this proposed concept. METHODS: Clinical and molecular data from patients aged under 18 years at time of diagnosis were evaluated for a distant intraventricular recurrence. A total endoscopic resection was performed with complete removal of the tumor, no complications, and no perioperative MRI. RESULTS: In the en bloc group, the perioperative outcomes, including hospital stay (p=0.001), PICU stay (p=0.003), total blood loss (p=0.015), transfusion rate (p=0.005) and complication rate (p=0.039), were all significantly improved. The univariate and multivariate regression analysis showed that tumor volume, bottom vessel, and imaging features, like encasing nerve or pass-by vessel, finger-like attachment, ratio of “limited line” and ratio of “clear line” remained independent predictors for the application of en bloc concept in our medical center. CONCLUSION: The application of en bloc concept based on the imaging features can improve the short-term outcomes.

SURG-04. ROBOTIC ALIGNMENT SYSTEM CIRQ (BRAINLAB) FOR NAVIGATED BRAIN TUMOR BIOPSY IN CHILDREN

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BACKGROUND: With the incorporation of the robotic alignment system Cirq (Brainlab, Munich, Germany) into our neurosurgical armamentarium, we doubted it is important to study application accuracy of this new device. As a basis, we retrospectively investigated the accuracy of brain tumor biopsies using the non-robotic alignment instrument Varioguide (Brainlab, Munich, Germany). Because of relatively high target registration errors in this historical series, we sought to improve our registration with the introduction of Cirq. Childhood. Develop our experience with the new robotic alignment system Cirq for navigated brain tumor biopsy. 2. Compare patient-to-image registration methods. 3. Evaluate the accuracy of Cirq robotic system as compared to the non-robotic instrument Varioguide. METHODS: All patients (10-19 years old) who underwent a brain tumor biopsy at the Princess Máxima Center, Utrecht, were included. Over the period 2018-2020, data were collected retrospectively (cohort “Varioguide” in which patient-to-image registration was based on preoperative MRI with adhesives). From May 2021 onwards, data were collected prospectively (cohort “Cirq” in which patient-to-image registration was done using 1. a preoperative MRI with adhesives and 2. an intraoperative CT head scan with bone screw fiducials). For both cohorts, Euclidian distances were calculated between the intended target and the obtained target using postoperative MRI scans. PRELIMINARY RESULTS: Nineteen patients were biopsied with Varioguide and nine patients with Cirq. The Cirq robotic system was convenient and safe in use with no intraoperative complications. Patient-to-image registration was more accurate when based on bone screw fiducials as compared to scalp fiducials. The Cirq robotic system, combined with bone fiducials, cerebellar base, and preoperative MRI resulted in a high target accuracy with a maximum target error of 2 millimeters. CONCLUSION: The Cirq robotic alignment system, combined with bone screw fiducial registration, provides a safe and highly accurate method for brain tumor biopsies in children.

SURG-05. SURVIVAL AND FUNCTIONAL OUTCOMES IN PEDIATRIC THALAMIC AND THALAMOPEDUNCULAR LOW GRADE GLIOMAS

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BACKGROUND: Childhood thalamopeduncular gliomas arise at the interface of thalamus and cerebral peduncle. The optimal treatment is total resection but not at the cost of neurological function. We present long-term clinical and oncological outcomes of maximal safe resection. METHODS: Evaluated for a distant intraventricular recurrence. A total endoscopic resection was performed given the local, non-disseminated recurrence pattern. WES results included NFI, PER1, and GLI3 mutations as well as FGFR3 gain, and was negative for TP53 alterations. The WES results included NF1, PER1, and GLI3 mutations as well as FGFR3 gain, and was negative for TP53 alterations. Repeat sequencing on recurrence 4 years later showed persistent NF1 and FGFR3 alterations. Methylation profiling was consistent with “plexus tumor, subclass pediatric B”. Overall, short-term surveillance neuroimaging detected four isolated recurrences on follow-up. During the follow-up period of 5 years, 9 years, and 10 years after initial CPG diagnosis. Mean hospital stay for all recurrences was 1 day with no complications from treatment. CONCLUSION: We describe a patient with 4 isolated recurrences of CPG over a decade, each treated with complete endoscopic removal, and identify an unusual set of molecular features that persisted over time. These outcomes support a strategy encompassing frequent neuroimaging surveillance to facilitate a minimally invasive endoscopic surgical approach at early diagnosis and relatively low tumor mutagenicity.

SURG-03. DURABILITY OF AN EARLY MANAGEMENT STRATEGY FACILITATING ENDOSCOPIC REMOVAL OF RECURRENT CHOROID PLEXUS CARCINOMA

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BACKGROUND: Choroid plexus carcinoma (CPC) is a rare, primarily intraventricular neoplasm with a dismal prognosis. Extent of surgical resection is correlated with improved outcomes but is frequently limited due to tumor vascularity and size. Information related to surgical management and molecular drivers of tumor recurrence is currently limited. Here we characterize a case of multiple recurrent CPC treated solely with sequential endoscopic tumor removals over a 10-year period and highlight its genomic properties. METHODS: We performed a retrospective review of a 16-year-old patient with CPC with local and distal recurrences using repeat excision with minimally invasive neuro-endoscopy. We describe the clinical nuances associated with neuro-endoscopic intervention, mean hospital stay, complications, and perioperative MRL. Whole exome sequencing (WES), targeted sequencing, and methylation profiling results over time are reviewed. RESULTS: Five years after standard treatment, the patient was
SURG-07. THE IMPACT OF EARLY TARGETED THERAPY ON THE NEUROSURGICAL APPROACH TO PEDIATRIC LOW-GRADE NEURO-ONCOLOGY

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We report two cases of pediatric low-grade glioma (pLGG) treated with vemurafenib, an oral BRAF-inhibitor: a 12-year-old girl with involvement of basal ganglia, hypothalamus, and diencephalic junction; a 3-year-old boy, with an optic pathway/hypothalamic glioma extending along the left optic tract to involve the right basal ganglia. Both received a biopsy and molecular analysis was performed in the tumor, showing BRAF V600E mutation. Therefore, instead of surgical removal planned by neurosurgeons, first-line treatment with vemurafenib was started: after one month 45% reduction of the mass according to RANO criteria was found, as well as better balance control and strong reduction of the right arm paresis; five months later, a 70% shrinkage was detected, stabilized to 76% after a year. The young boy first started chemotherapy with vincristine and carboplatin, but at the end of the induction phase the tumor had increased and ascites, hydrocephalus, and visual and motor impairment occurred. These were improved with further reduction. Since then, vemurafenib treatment was started; therefore, the surgical option was postponed and therapy with vemurafenib started. After only three days, visual acuity and muscle tone improved; brain MRI showed a 34% tumor shrinkage, increased up to 63% after one month. Therefore, no ulceration surgery was necessary. In pLGG, the neurosurgical biopsy is essential to let an early and rapid molecular diagnosis of BRAF mutations and guide subsequent targeted therapies. Our cases demonstrate how a prompt radiological response to vemurafenib and the related clinical improvement can influence both therapeutic and surgical decision-making, hope- fully reducing the occurrence of second neurosurgery with associated risks of neurological sequelae. To our knowledge, this is the first report assessing such a quick shrinkage in pLGG treated with vemurafenib, highlighting the importance of an early investigation of BRAF status in all cases of LGG in children.

SURG-06. MANAGEMENT OUTCOME (5 YEARS OR MORE) OF PEDIATRIC VERTEBRAL HEMANGIOMAS PRESENTING WITH MYELOPATHY

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INTRODUCTION: Vertebral hemangiomas are very rare in pediatric age group. In this study we analysed the outcomes of patient treated at department of neurosurgery , All india institute of medical sciences New Delhi since may 2010 to January 2022 with at least 5 year follow up. MATERIALS AND METHODS: All patients up to 18 years age of vertebral haemangioma treated at our hospital from may 2010 to January 2022 with at least 5 year follow up. All had features of myelopathy;Patients demography, clinical details and follow up and complications were retrieved from hospital record. Functional clinical outcomes were measured using ASIA score. Mean age was 14.21 years with range of 12 years to 18 years. All patients had magnetic resonance imaging and computed tomography of spine in preoperative and postoperative period. Mean follow up was 78 months with range of 59 to 122 months.

RESULTS: During 16-year period 25 children with craniopharyngioma being resected in our institution. In total 11 patients were downgraded to grade I by pinhole test and cranial nerves examination; in total 6 and less extensive (9). Surgical complications included progression of baseline neurological status in 6 patients, 3 of these gradually improved to preoperative status. All tumors were classified as low-grade gliomas. Disease progression was observed in 9 patients (median progression free survival 7.3 years). At last follow-up (median 6.6 years) all patients were alive; median Lansky score of 90. Seven patients were without evidence of disease, 6 had stable disease, 7 stable following progression and 1 had progressive disease managed expectantly. CONCLUSION: Pediatric patients with low grade thalamospeduncular gliomas have excellent long-term functional and oncological outcomes even when gross total resection is not achievable. Surgery should aim at total resection; however neurological function should not be endangered due to excellent chance for long-term survival.

SURG-08. 5-AMINOLEVULINIC ACID (5-ALA)-GUIDED RESECTION OF PEDIATRIC BRAIN TUMORS

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BACKGROUND: 5-Aminolevulinic acid (5-ALA)-guided resection of gliomas in adults enables better differentiation between tumor and normal brain, allowing a higher degree of resection, and improved patient outcomes. In recent years, several reports have emerged regarding the use of 5-ALA-guided resection in other brain tumor entities, including pediatric brain tumors. Since gross total resection (GTR) of many brain tumors in children is crucial, the role of 5-ALA-guided resection requires elucidation. METHODS: A systematic literature review of MEDLINE/PubMed and other databases revealed 186 eligible publications encompassing 5-ALA-guided operations on pediatric brain tumors. To reduce bias, publications were revised independently by two authors. RESULTS: 5-ALA-guided resection enabled the surgeons to identify the tumor more easily and was considered helpful mainly in cases of low malignant gliomas, like gangliogliomas, well differentiated astrocytoma and pleomorphic xanthoastrocytoma. 5-ALA was considered helpful in 12% and 22% of cases, respectively. Accumulation of fluorescent porphyrins seems to depend on WHO tumor grading. In case fluorescence signal was considered helpful, it was associated to a greater degree of resection. One study showed an association between visible fluorescent signal and concentration of protoporphyrin IX (PPIX) concentration. A threshold of 4 μg/ml was required in order to visualize the fluorescence signal. The rate of adverse events related to 5-ALA was negligible, especially new postoperative sequelae. CONCLUSION: 5-ALA could play a role in resecting malignant, contrast enhancing, pediatric brain tumors. At present, we are conducting a prospective phase I-II multicenter clinical trial to evaluate side effects and feasibility of 5-ALA guided surgery.