

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 and piecemeal group) was conducted to investigate the effect of 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 the predictors associated with the application of en bloc concept. 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 multinomial logistic 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?

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BACKGROUND: Medulloblastoma (MB) are the most common malignant brain tumor in childhood. Developmental gene expression data supported by neuroradiological 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 who were operated on a histologically confirmed MB at the Department of Neurosurgery of the Medical University of Vienna between 1990 and 2020 were retrospectively analyzed. The location of the tumor origin was defined based on operative reports, surgical videos and preoperative imaging data by an experienced neurosurgeon blinded to the subgroup information. RESULTS: Sufficient data were available in 53 patients. In 28.6% (2 / 7) WNT-MB, 66.7% (6 / 9) SHH-MB and 70.3% (26 / 37) Group 3 and Group 4 MB, the intraoperatively defined site of origin corresponded well with the cellular origin suspected from the molecular subgroup. Within the WNT-subgroup, 57.1% (4 / 7) originated from the vermis, 28.6% (2 / 7) from the LRL and 14.3% (1 / 7) from the cerebellar hemisphere. The origin of SHH-MB was predominantly located in cerebellar hemispheres (66.7% (6 / 9)), while 33.3% (3 / 9) originated from the vermis. Of Group 3 and Group 4 MB, 70.3% (26 / 37) had their origin in the vermis and 29.7% (11 / 37) in the LRL. CONCLUSION: Our results indicate that there is a considerable level of inconsistency between the intraoperatively observed site of origin and the expected cellular origin based on the molecular subgroup, especially in WNT-MB. This discrepancy needs to be discussed when it comes to surgical decision-making accounting for risk stratification.

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

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BACKGROUND: Choroid plexus carcinoma (CPC) is a rare, primarily intraventricular neoplasm with a dismal prognosis. Extent of surgical removal 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 multiply 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 female treated for CPC with local and distal recurrences undergoing repeat excision with minimally invasive neuro-endoscopy. We describe technical nuances associated with neuro-endoscopic intervention, mean hospital stay, complications, and perioperative MRI. Whole exome sequencing (WES), targeted sequencing, and methylation profiling results over time are reviewed. RESULTS: Five years after standard treatment, the patient was

evaluated for a distant intraventricular recurrence. A total endoscopic resection was performed given the local, non-disseminated recurrence pattern. WES results included NF1, PER1, and GLI3 mutations as well as FGFR3 gain, and was negative for TP53 alterations. Repeat sequencing on a 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, all treated with complete endoscopic resections at 5 years, 6.5 years, 9 years, and 10 years after initial CPC 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 CPC over a decade, each treated with complete endoscopic removal, and identify an unusual set of molecular alterations that persisted over time. These outcomes support a strategy encompassing frequent neuroimaging surveillance to facilitate a minimally invasive endoscopic surgical approach at early detection and relatively low tumor mutagenicity.

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

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BACKGROUND: With the incorporation of the robotic alignment system Cirq (Brainlab, Munich, Germany) into our neurosurgical armamentarium, we deemed it important to study application accuracy of this new device. As a baseline, we retrospectively reviewed our historical data on 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. AIMS: 1. Share our experience with the new robotic alignment system Cirq for navigated brain tumor biopsies. 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 (0-19 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 adhesive scalp fiducials). From 2021 onwards, data were collected prospectively (cohort "Cirq" in which patient-to-image registration was done using 1. A preoperative MRI with adhesive scalp fiducials 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 fiducial registration, 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:

Retrospective review of prospectively collected data: demography, symptomatology, imaging, extent of resection, surgical complications, histology, functional and oncological outcome. **RESULTS:** During 16-year period (2005-20) 21 patients were treated at our institution. These were 13 girls and 8 boys (mean age 7.6 years). Presentation included progressive hemiparesis in 9 patients, raised intracranial pressure in 9 patients and cerebellar symptomatology in 3 patients. The tumor was confined to the thalamus in 6 cases. Extent of resection was judged on postoperative imaging as total (6), near-total (6) and less extensive (9). Surgical complications included progression of baseline neurological status in 6 patients, 5 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 6.1 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 thalamopeduncular 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-06. MANAGEMENT OUTCOME (5YEARS OR MORE) OF PAEDIATRIC VERTEBRAL HAEMANGIOMAS 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 METHOD:** 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 records. 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 from 60 month to 144 months. **RESULTS:** There were 6 male and 8 female patients with sex ratio of 3:4. All were located in dorsal spine with single level involvement. Upper dorsal spine involvement was more common (10 cases 71.43%) than lower dorsal spine involvement (4 case 28.57%). All patients had weakness of lower limbs with features of myelopathy, urinary bladder symptoms were present in 6 patients local site pain in 1. D1 involvement was present in 1, D3 in 2, D4 in 2, D5 in 2, D6 in 3, D8 in 1, D10 in 1, D11 in 1 and D12 in 1. All patients have improvement in power of both lower limbs after surgery. Bladder symptom and pain resolved in all patients. **CONCLUSION:** Good postoperative results can be achieved in pediatric symptomatic vertebral hemangioma with minimal complications. Individualization of treatment for each patient should be done with avoidance of selecting more blood loss procedures like corpectomies.

#### **SURG-07. THE IMPACT OF EARLY TARGETED THERAPY ON THE NEUROSURGICAL APPROACH TO PEDIATRIC LOW-GRADE GLIOMA**

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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 and basal ganglia. Both received a biopsy and molecular analysis was performed in the girl's 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 impairment occurred. Molecular testing showing BRAF V600E mutation on the initial tumor biopsy was obtained; 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% reduction of the mass after one week, increased up to 65% after six months. Therefore, no ulterior 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 decisions, hopefully 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-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 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 EMBASE and MEDLINE/PubMed databases revealed 20 eligible publications encompassing 186 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 glioblastoma (GBM, 21/27, 78%), anaplastic ependymoma WHO grade III (10/14, 71%), and anaplastic astrocytoma (4/6, 67%). In contrast, cases of pilocytic astrocytomas (PAs) and medulloblastomas 5-ALA-guided surgery did not show consistent fluorescent signals and 5-ALA was considered helpful only 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 fluorescence signal and concentration of protoporphyrin IX (PPIX) concentration. A threshold of 4µg/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 resection of malignant, contrast enhancing, supratentorial 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

#### **SURG-09. HYPOTHALAMIC INVOLVEMENT IN PEDIATRIC CRANIOPHARYNGIOMAS; HOW TO DEFINE AND TO REALIZE OPTIMAL NEUROSURGICAL RESECTIONS**

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The optimal neurosurgical strategy for pediatric craniopharyngiomas is on debate with shifts from gross total resection to limited resections in combination with local radiotherapy. The most relevant question underlying this ongoing discussion is how to prevent additional surgical morbidity to the hypothalamus. Limited neurosurgical resection in combination with local radiotherapy has been proved a safe alternative to gross total resection concerning tumor control. Pre-operative anatomical grading of hypothalamic involvement has been proposed by Puget et al using MRI discriminating between no hypothalamic involvement (grade 0), hypothalamic displacement (grade 1), and hypothalamic disruption (grade 2). Subsequently, post-operative hypothalamic damage can also be assessed on post-op MRI by the grading of the Vile et al in no discernable damage (grade 0), abnormality of floor of third ventricle (grade 1), or damaged or deficient floor of third ventricle (grade 2). Three questions remain to be settled: 1. How to define pre-operatively the extent of resection 2. How to realize pre-operatively the intended limited resection, and three. How to determine post-operatively the (additional) surgical damage to the hypothalamus. We retrospectively evaluated a series of 25 children with craniopharyngioma being resected in our center from June 2018 until January 2022, with pre- and post-operative grading of hypothalamic involvement. In total 11 patients were downgraded on the post-operative compared to the pre-operative scale; either by 1 grade