Hospital has the expertise to build a comprehensive neuro-oncology program. The program currently lacks a dedicated nurse coordinator and "specialist" in neuro-oncology. Ongoing discussions with local stakeholders are aimed to galvanize national support to improve awareness for children with brain tumors and to plan a multidisciplinary neuro-oncology symposium in 2021. In the meantime, telemedicine efforts can support nursing education and re-iterate the multidisciplinary needs for children with brain tumors.

#### LINC-33. MULTIMODALITY MANAGEMENT OF PAEDIATRIC PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA- UPDATED EXPERIENCE FROM A REGIONAL CANCER CENTRE IN NORTH INDIA

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Paediatric primary central nervous system lymphoma(PCNSL) constitutes 1% of all PCNSLs. Data pertaining to paediatric PCNSL (2016-19) was abstracted by retrospective chart review. We identified 7 paediatric patients with PCNSL. None had congenital or acquired immunodeficiency. The median age at presentation was 13 years. The male to female ratio was 4:3. The median ECOG performance status was 2. On neuro-imaging, 3 patients had solitary and 4 patients had multiple lesions. CSF cytology showed atypical cells in 1 patient. None had ocular involvement. Systemic lymphoma work-up was negative in all. Biopsy and resection of tumour were done in 4 patients each. Histopathology revealed DLBCL in 6 and B-cell NHL in 1 patient. All patients underwent induction chemotherapy (median-5 cycles)- modified DeAngelis protocol (IV Methotrexate-2.5g/m<sup>2</sup>,IT Methotrexate-12 mg, Vincristine, Procarbazine and Rituximab-375mg/m<sup>2</sup> every 2 weeks) in 6 and single agent Methotrexate -3.5g/m<sup>2</sup> every 3 weeks in 1 patient. Severe haematological toxicities included grade 3 neutropenia, leucopenia and febrile neutropenia in 2,1 and 1 patient respectively. Radiotherapy(RT) was administered in all-whole brain RT(36-45Gy/20-25fractions/4-5weeks) in 6 patients and craniospinal RT(36Gy/18fractions/3.5weeks) followed by whole brain RT(9Gy/5fractions/1week) in 1 patient(with positive CSF cytology). Subsequently consolidation chemotherapy with 2 cycles of Cytarabine(3g/m<sup>2</sup> IV D1-2 every 3 weeks) was administered in 5 patients. After a median follow-up of 14 months(mean-18.2 months), all patients are in complete radiological remission. Paediatric PCNSL is a rare tumour entity and multimodality management with high dose Methotrexate and Rituximab based chemo-immunotherapy and cranial radiotherapy leads to excellent early clinical outcome.

# LINC-34. OPTIC NERVE INFILTRATION: RARE MANIFESTATION OF CHILDREN WITH ACUTE LYMPHOBLASTIC LEUKEMIA IN REMISSION

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BACKGROUND: Optic nerve infiltration in acute lymphoblastic leukemia is a rare manifestation. This infiltration may appear months in advance as an isolated sign of extramedullary relapse and considered as one of the significant clinical findings of central nervous system leukemia. AIM: To describe a case of rapidly progressive optic nerve infiltration in a girl with ALL in remission. CASE: A 13-year-old girl in full remission following treatment for B-cell acute lymphoblastic leukemia presented with decreased vision and proptosis on the left eye. She completed the chemotherapy course two years before. On physical examination, we found the optic disc swelling in her left eyes. There were no signs of relapse from the hematological, cerebrospinal fluid analysis, and bone marrow aspiration. The orbital CT found a mass on the left retrobulbar (size 29x48x32 mm), suspected of optic nerve glioma. The mass has grown rapidly in a month, and she lost her left sight. The involved eye was exenterated (60x55x40 mm). The histopathology and immunohistochemistry showed the B-cell acute lymphoblastic lymphoma. Unfortunately, the patient could not come for further follow up due to the COVID-19 large-scale social distancing. Two months later, she came with pallor and pain in all of her body. The bone marrow aspiration showed leukemic relapse and she is undergoing chemotherapy. CONCLUSION: Optic nerve infiltration by leukemia requires both diagnostic certainty and urgent management. A routine ophthalmic assessment is recommended in patients with a history of acute lymphoblastic leukemia to diagnose optic nerve involvement due to leukemic infiltration.

#### LINC-35. THE ST. JUDE GLOBAL ACADEMY NEURO-ONCOLOGY TRAINING SEMINAR: A MULTIDISCIPLINARY, INTERNATIONAL EDUCATION PROGRAM

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The success of the treatment of children with central nervous system (CNS) tumors relies on an effective multidisciplinary team, with up-to-date

and broad knowledge and skills. The St. Jude Global Academy Neuro-Oncology Training Seminar was launched as course in globally applicable content in pediatric neuro-oncology with a focus on multidisciplinary teams in low- and middle-income countries (LMICs). To identify the content that is most relevant for the learners, a needs assessment survey that included evaluation of team dynamics, treatment capacity, existing knowledge, and educational goals was designed. Survey questions in 11 domains were answered by 24 sites in LMICs across the world. This information was used to create the course that consists of two components: a 9-week online course and a 10-day workshop at the St. Jude campus. 72 participants from 11 institutions enrolled in the online portion and 20 participants were selected based on grades to attend the workshop. A retrospective post-test evaluation established that learners improved their understanding of the barriers to care, possible solutions to improve care, understanding of diagnosis and treatment, and methodology to implement projects (p<0.01). All participating teams developed projects that are locally implemented. Those present at the workshop formed a multidisciplinary, international collaborative group (Global Alliance in Pediatric Neuro-Oncology). This experience establishes that educational programs with systematically created curricula can not only improved knowledge but be a mechanism to share experiences and create collaborative networks. Ultimately, patient outcomes will be tracked to monitor the true impact of the course.

# LINC-36. TRILATERAL RETINOBLASTOMA: A REPORT OF FOUR CASES

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Retinoblastoma is the most common primary malignant intraocular cancer that usually develops in early childhood. About 5% of those patients are at risk of developing trilateral retinoblastoma (TRB). In developing countries, most of them came in the late stage; therefore, ocular and patient survival rates are lower than in developed countries. From 2015–2019, we found four cases of trilateral retinoblastoma. Two of them had bilateral retinoblastoma, and two had unilateral retinoblastoma. They all presented with leukocoria and had no family history of retinoblastoma. The mean age was 13.8 months (range 9–24 months of age). The diagnosis of trilateral retinoblastoma was made from initial head CT/MRI. They were treated during treatment. Trilateral retinoblastoma is usually fatal and needs multidisciplinary treatment care. In developing countries, it is important to evaluate distant metastasis. Head CT or MRI from the initial diagnosis to exclude the trilateral retinoblastoma.

#### LINC-38. 500 CONSECUTIVE SURGICAL CASES FROM THE PEDIATRIC ONCOLOGY NEUROSURGERY GROUP: UNDERSTANDING THE PERSPECTIVE OF A TERTIARY CENTER IN BRAZIL

<u>Felipe Hada Sanders.</u> Hamilton Matushita, and Manoel Jacobsen Teixeira; USP, Sao Paulo, SP, Brazil

With this presentation we aim to present cases submitted to surgery by the same group of surgeons since 2010, presenting the physical structure, medical assistance, scientific production and the challenges that we need to overcome in the second decade of the twenty-first century, in a developing country.

#### LINC-39. PERFORMANCE STATUS OF PEDIATRIC PATIENTS WITH CENTRAL NERVOUS SYSTEM TUMORS TREATED IN MEXICO, A SINGLE-CENTER EXPERIENCE

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BACKGROUND: Central nervous system (CNS) tumors are the most common solid neoplasms in the pediatric age, they comprise about a quarter of all cancers at this age. Little is known about the specific epidemiology of this group in Mexico and there are no reports of results focused on the Performance Status of patients who are treated in a multidisciplinary setting. OBJECTIVE: To describe the Performance Status of CNS pediatric patients after being treated with a multidisciplinary approach in a tertiary center. METHODS: We report a retrospective chart review of all pediatric patients who presented to the Neuro-Oncology Clinic at Teleton Pediatric Oncology Hospital in Queretaro, Mexico, from December 2014 to January 2020. We analyzed age, gender, the extent of surgical resection and histopathology. Performance Status was assessed using ECOG and Karnofsky/ Lansky scores during every patient's last follow-up visit. RESULTS: A total of 56 patients were treated, epidemiology and histopathology variants are similar to those described in the international literature. With a median follow-up of 33 months, 35 patients are alive (62.5%), 28 of them (74.2%)

have an excellent Performance Status (ECOG score 0 or Lansky/Karnofsky  $\geq$  90), 5 (14.2%) scored ECOG 1–2 and only 4 (11.4%) scored ECOG 3–4. CONCLUSIONS: A multidisciplinary approach with a focus on Performance Status and the potential for neurological recovery is essential in the management of pediatric patients with CNS tumors. Efforts should be aimed at reducing post-surgical morbidity and early rehabilitation to reintegrate patients into society in the long term.

#### LINC-40. VERY YOUNG PATIENTS AND CENTRAL NERVOUS SYSTEM TUMORS: A SINGLE-CENTER EXPERIENCE IN AN UPPER-MIDDLE-INCOME COUNTRY

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Tumors of the central nervous system comprise nearly a quarter of all childhood cancers and are the most frequent solid tumor in the pediatric population. Primary central nervous system tumors (PCNST) are a rare and heterogeneous group of tumors responsible for high mortality and morbidity. Around 10% of primary CNS tumors occur during the first year of life with almost half of them during the first six months. About 18% of these tumors appear before the age of two years. Very young children differ from older children and adolescents regarding the incidence and location of different histological entities of CNS tumors. We aimed at providing descriptive epidemiological data and report the outcome in a tertiary center from December 2013 to January 2020 for all histological subtypes of primary central nervous system tumors in very young patients, defined as patients younger than three years. We collect data from 19 patients treated in an oncology exclusive tertiary center in Mexico between 2013 and 2020. This study aims to relate factors such as age, radiotherapy, surgery, chemotherapy with Lansky Performance Scale and determine the impact, not only in the overall survival but also in the quality of life.

## LINC-41. TREATMENT OF RECURRENT MEDULLOBLASTOMA IN CHILDREN IN LOW INCOME SETTINGS

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INTRODUCTION: Children with recurrent medulloblastoma after initial therapy have very poor prognosis due to limited second line treatment options and significant treatment-related morbidity. METHODS: A retrospective chart review of 18 children with recurrent or progressive medulloblastoma, treated initially with risk-adapted therapy in Western Ukrainian Specialized Pediatric Medical Centre from 2012 to 2019, was performed. RESULTS: All patients received first line multimodal treatment: surgery, distant beam radiotherapy and chemotherapy. Recurrent disease in 11 patients presented with metastatic dissemination and in 7 patients as local relapse. The median time to recurrence was 10 months. The median follow-up after diagnosis of recurrent disease diagnosed was 2 years and 2 months. Second line therapy included re-surgery (5 cases), radiation therapy (10 cases) and various cytostatic agents as monotherapy or combination - carboplatin, cisplatin, cyclophosphamide, etoposide, methotrexate, temozolomide, lomustine. Patients treated with radiotherapy for salvage had prolonged local control compared to those that received chemotherapy only. On follow-up 8 children are currently alive. CONCLUSION: Recurrent and progressive medulloblastoma had a poor prognosis with a 2-year overall survival (OS) of 28% on different salvage therapy. The variety in the treatment of all patients experiencing recurrence was observed due to low income country settings. The factors that influenced higher survival after recurrence of medulloblastoma were longer time to relapse, and local pattern of relapse/progression.

#### LINC-42. EPIDEMIOLOGICAL OVERVIEW OF CHILDHOOD CNS TUMORS IN THE NEUROSURGICAL UNIT IN YEREVAN, ARMENIA

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BACKGROUND: Central nervous system (CNS) tumors are the second most common malignant neoplasms among children worldwide. The cur-

rent paper aims to analyze the situation in pediatric neuro-oncology in Armenia from the neurosurgical perspective. METHODS: We have collected data of pediatric patients with CNS tumors treated in the Neurosurgery department of "Surb Astvasamayr" Medical Center from 01.01.2010 till 01.12.2019. Incidence by gender, age at diagnosis, and histopathology results were calculated. Survival rates were calculated based on the follow-up results performed until 30.12.2019. RESULTS: Hospital-based data showed that during the previous 10 years 47 patients with CNS tumors received neurosurgical treatment in the unit, among them 66% were females. 38.3%, 31.9% and 29.8% of diagnosed patients were aged 0-4, 5-9, and 10-18 respectively. In 41 cases, the disease was not disseminated at diagnosis. The most common observed malignancies were low-grade gliomas (21.3%) and embryonal tumors (19.1%), followed by high-grade gliomas (14.9%) and ependymal tumors (8.5%). Follow-up information only for 33 patients is available. From them, 14 are dead and 19 alive. Survival rates in most common groups were 62.5%, 80%, 50%, and 50% respectively. The median follow-up time was 18 months (range 1-113 months). CONCLU-SION: Similar to the data reported in the literature, low-grade gliomas, and embryonal tumors are the most frequent pediatric CNS tumors in Armenia. On the other hand, the pediatric CNS tumor survival rates are lower compared to those reported in developed countries.

### LINC-43. FACTORS LEADING TO DIAGNOSTIC DELAY FOR CHILDREN WITH PRIMARY TUMORS OF CENTRAL NERVOUS SYSTEM (CNS) IN QATAR

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INTRODUCTION: Median time to diagnosis for primary CNS tumors for children in Qatar has been reported to be 28 days. However, a wide variation in diagnostic times is seen. This study was undertaken to analyze the factors leading to delay in diagnosis. METHODS: Data were retrospectively analyzed for children who had diagnostic delay (more than 28 days) from September 2006 to February 2020. Presenting symptoms, number and type of healthcare contacts and presenting symptom interval (PSI) were reviewed. Parental delay (PSI-1) was defined as the date of onset of first symptom to the date of first healthcare contact. Healthcare delay (PSI-2) was defined as date of first healthcare contact to the date of diagnostic scan. RE-SULTS: Twenty-four patients were identified with diagnostic delay. Median age at diagnosis was 48.2 (range 5.4-171.6) months with an equal sex distribution. Fifteen (62.5%) patients were older than 3 years, 13(54%) patients had low grade glioma, 16 (66.7%) had supratentorial tumors and 12 (50%) presented with raised intracranial pressure. Diagnosis was made after a median 3 (range 1-8) healthcare contacts. Nineteen (79%) patients presented to primary care. Median PSI was 132 (31-783) days. Parental delay (PSI-1) was 35 (0-496) days, while healthcare delay (PSI-2) was 41 (0-562) days. Endocrine (241 days) and oculo-visual (184 days) symptoms were associated with the longest PSI. CONCLUSIONS: There was no significant difference between parental and healthcare delay. Endocrine and oculo-visual symptoms were associated with longest PSI. Increased awareness is required for early recognition of signs suggestive of CNS tumors.

### MEDULLOBLASTOMA (CLINICAL)

#### MBCL-01. METHYLATION PROFILING OF PEDIATRIC MEDULLOBLASTOMA IN SAUDI ARABIA IN A CLINICAL SETTING PERMITS SUB-CLASSIFICATION AND REVEALS NEW OUTCOME PREDICTIONS

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Medulloblastoma (MB) is the most common childhood malignant brain tumor. DNA methylation profiling has rapidly advanced our understanding of MB pathogenesis at the molecular level, MBs can be sub-grouped according to methylation patterns from FPPE samples into Wingless (WNT-MB), Sonic Hedgehog (SHH-MB), Group 3 (G3) and Group 4 (G4) WNT-MB and SHH-MB subgroups are characterized by gain-of function mutations that activate oncogenic cell signalling whilst G3/G4 tumors show recurrent chromosomal alterations. each subgroup has distinct clin-