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BACKGROUND/OBJECTIVE: The introduction of German regimens, supplementing "standard" chemotherapy with both intravenous high-dose (HD-MTX) and intraventricular (IVENT-MTX) methotrexate, and North American regimens incorporating marrow-ablative chemotherapy with autologous hematopoietic cell rescue (HDCx+AuHCR), report encouraging outcomes for young children with medulloblastoma. We performed a comparative outcomes analysis of treatment strategies for young children with ClMB or A/LCMB. DESIGN/METHODS: Data from 12 prospective multi-center trials published between 2005 and 2019 for children <six-years-old with ClMB or A/LCMB were reviewed; survivals were compared. RESULTS: COG-9921, UKCCSG-CNS9204, COG-P9934 and SJYCO7 employing standard chemotherapy with either no or risk-based irradiation, reported 3-5-year event-free survival (EFS) of 17+/-5%, 33+/-28% (ClMB), 14+/-7% and 13.8+/-9% (ClMB) respectively, with reported EFS of 0% for A/LCMB in UKCCSG-CNS9204 and SJYCO7. HIT-SKK'87, HIT-SKK'92 and HIT-SKK'00 incorporating HD-MTX and IVENT-MTX reported 2-10-year EFS of 30–34+/-10–11% for ClMB and 33+/-27% (HIT-SSK'00) for A/LCMB. Head Start HS-I-II combined, CCG-99703 and HS-III employing induction chemotherapy, with or without HD-MTX, followed by single or tandem HDCx+AuHCR reported 3-5-year EFS of 42+/-14%, 50+/-11% and 27+/-6% for ClMB, with EFS for A/LCMB of 38+/-13% (HS-III). Finally, 5-year overall survivals for ACNS0334, without or with induction HD-MTX, are 39% and 69% respectively for ClMB and A/LCMB combined. CONCLUSIONS: A trend towards better outcomes for young children with ClMB and A/LCMB is observed in trials including *either* HD-MTX and IVENT-MTX *or* including HD-MTX-containing induction chemotherapy and HDCx+AuHCR. Trials excluding HD-MTX, IVENT-MTX and HDCx+AuHCR have poorer outcomes.

MBCL-38. UNUSUAL EXTRANEURAL METASTASIS OF PEDIATRIC EMBRYONAL TUMORS: TWO CASE REPORTS

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We report two cases of unusual extraneural metastasis in patients with embryonal tumors without central nervous system disease progression and prolonged survival. The first patient presented at 16 years of age with atypical teratoid rhabdoid tumor of the cervical spine. The tumor was confirmed to have loss of INI1, SMARCB1 deletion of exons 1–3, and heterozygous deletion of 22q11.2. The patient received treatment initially per ACNS0333 with high dose chemotherapy and tandem autologous transplants. The patient developed a biopsy-confirmed liver metastasis six months from diagnosis and, subsequently, had disease progression including liver metastases, bony lesions, muscle involvement, and lung nodules. Two and a half years from diagnosis the patient has still not had a relapse in the CNS. The second patient presented with medulloblastoma isolated to the posterior fossa at 11 years of age and was treated on SJMB03 protocol with craniospinal irradiation and high dose chemotherapy. He had his first recurrence in the temporal lobe three years post treatment. He had multiple recurrences in the brain over the next five years treated with re-resections, adjuvant chemotherapy, and gamma knife radiotherapy. He then developed cervical lymphadenopathy, bony lesions, liver lesions, and lung nodules. Cervical lymph node biopsy confirmed medulloblastoma. Next generation sequencing from recurrent tumor showed somatic mutations in *p53*, *KDM6A*, and *PPP2R1A*. Fourteen years from treatment, he has now developed a temporal lobe lesion. These cases are notable for prolonged survival despite widely metastatic disease and genomics predicting poor prognosis as well as metastatic disease disproportionate to CNS disease.

MBCL-41. LYMPHOHEMATOPOIETIC TOXICITY IDENTIFIED IN PATIENTS WITH MEDULLOBLASTOMA RECEIVING CRANIOSPINAL IRRADIATION

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BACKGROUND: Medulloblastoma (MB) is the most common malignant brain tumor of childhood. MB easily disseminates through the spinal fluid. Surgery followed by radiotherapy, applied to the entire craniospinal axis (CSI), and adjuvant chemotherapy, represent the treatment of choice for patients aged ≥3 years. Since the bone marrow of the skull and vertebral column are the major hematopoietic organs, we investigated the myelosuppressive effect of irradiation treatment in patients with MB retrospectively. METHODS: Medical records of newly diagnosed MB patients treated at our hospital from 2007–2019 were analyzed. Children <3 years old were excluded because they did not receive CSI to avoid potential neurotoxicity. RESULTS: Medical records of 18 patients (11 males and 7 females, aged 6–26, median 11 years) were reviewed. Eight patients were stratified as high-risk disease and 10 patients with standard risk. All patients received CSI (dosage range 23.4–39.6 Gy based on disease risk) and posterior fossa boost. All patients developed lymphocytopenia (<0.5×109/L) during irradiation, and for 11 of 18 patients, lymphocytopenia (<0.2×109/L) was severe. Although 13 patients recovered from the lymphocytopenia before the initiation of chemotherapy, five patients underwent chemotherapy without recovery. Conversely, only six patients developed neutropenia (<1.0×109/L), and five of the six patients were <10 years old. CONCLUSION: Although infectious episode associated with lymphocytopenia was not observed in this study, CSI treatment in children and adolescents may induce immunodeficient condition particularly in the lymphocytic system. Pediatric oncologists should pay attention to the impaired immunity of patients with MB who receive CSI.

MBCL-43. RECURRENT MEDULLOBLASTOMA – LONG-TERM SURVIVAL WITH A "MEMMAT" BASED ANTIANGIOGENIC APPROACH

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INTRODUCTION: Patients with recurrent medulloblastoma have a poor prognosis with only around 8% of patients surviving at 5 years irrespective of salvage therapy used. We report on 29 patients from four institutions treated with a "MEMMAT" based antiangiogenic combination therapy. PATIENTS AND METHODS: From 11/2006 to 06/2016, 29 patients were diagnosed with a recurrent medulloblastoma (19 first, 10 multiple recurrences). Median age at start of antiangiogenic therapy was 10 years (range 1–27). Subgroup of medulloblastoma was available in 18 patients and was group 3 or 4 in all except two (one WNT, one SHH-infant). For their current relapse patients received an antiangiogenic combination therapy consisting of bevacizumab, thalidomide, celecoxib, fenofibrate, and etoposide, alternating with cyclophosphamide and augmented with intraventricular therapy (etoposide and liposomal cytarabine). RESULTS: As of 01/2020, 8/29 patients are alive at a median of 44 months after recurrence. 6/8 surviving patients are currently in CCR between 66 and 134 months after recurrence that prompted MEMMAT therapy. Two patients are again in remission after intercurrent relapses 105 and 102 months after first starting MEMMAT therapy. Five patients died of another cause (accident, leukemia, septicemia). OS (median 44 months) was 44±10% at 5 years and 39±10% at 10 years, PFS was 33±10% at 5 years and 28 ±9% at 10 years. Therapy was well tolerated and toxicities were manageable. CONCLUSION: Our results suggest that antiangiogenic metronomic chemotherapy has clinical activity in recurrent medulloblastoma. Further investigation with an international phase II study is ongoing (MEMMAT; ClinicalTrials.gov Identifier: NCT01356290).

MBCL-46. TREATMENT OF RECURRENT WINGLESS-ACTIVATED MEDULLOBLASTOMA (WNT-MB) INCORPORATING MARROW-ABLATIVE THIOTEPA AND CARBOPLATIN CHEMOTHERAPY (HDCX) AND AUTOLOGOUS HEMATOPOIETIC PROGENITOR CELL RESCUE (AUHPCR): A DUAL REPORT

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BACKGROUND: Wnt-MB infers an excellent prognosis, and metastatic disease is rare. However, specific treatment strategies and patterns of failure for patients with recurrent Wnt-MB are unknown. We report two cases of

recurrent beta-catenin nucleopositive Wnt-MBs treated with an irradiation-sparing strategy, incorporating HDCx/AuHPCR. PATIENT 1: A nine-year-old female experienced local recurrence of a non-metastatic Wnt-MB nine months after gross total resection (GTR) followed by 18Gy craniospinal irradiation (CSI) with primary site boost to 54Gy, accompanied by weekly vincristine, followed by a maintenance regimen of nine cycles of cisplatin/lomustine/vincristine alternating with cyclophosphamide/vincristine every third cycle. GTR of the relapsed tumor was followed by three cycles of HDCx/AuHPCR. She is disease-free for over three years following relapse treatment. PATIENT 2: A 17-year-old male initially underwent GTR, followed by 23.4Gy CSI with 54Gy posterior fossa boost with concomitant weekly vincristine, followed by a maintenance regimen that included nine alternating cycles of vincristine/lomustine/cisplatin and cyclophosphamide/vincristine. Isolated right frontal horn metastatic recurrence developed 19 months post-treatment; three cycles of irinotecan/temozolomide/bevacizumab and gamma-knife radiosurgery produced complete response. A second isolated metastatic recurrence within the left frontal horn occurred 13 months post-treatment, which was treated with two cycles of cyclophosphamide/etoposide followed by two cycles of HDCx/AuHPCR. MRI of the brain showed no residual tumor one month post-treatment. He currently awaits follow-up stereotactic radiosurgery. CONCLUSION: Patients with recurrent Wnt-MB may be treated with curative intent using a multidisciplinary approach that includes HDCx/AuHPCR, and minimization or avoidance of re-irradiation.

MBCL-48. OUTCOMES OF TREATMENT BASED ON THE ST. JUDE MEDULLOBLASTOMA-96 REGIMEN FOR JAPANESE CHILDREN WITH MEDULLOBLASTOMA

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Medulloblastoma is a type of malignant embryonal tumor in childhood that is considered to require multiagent chemotherapy followed by radical resection and craniospinal irradiation (CSI). However, the outcomes of chemotherapy for this tumor in Japan are unclear. Here, we performed a multicenter retrospective study to determine the prognosis of pediatric medulloblastoma patients in Japan treated with the St. Jude medulloblastoma-96 (SJMB96) regimen. Thirty patients with newly diagnosed medulloblastoma received treatment with the SJMB96 regimen at Juntendo University Hospital in Tokyo (n=10), Saitama Medical University International Medical Center in Saitama (n=10), and Tohoku University Hospital in Miyagi (n=10) from 2011 to 2018. All patients underwent tumor resection and CSI, with radiation doses of 23.4Gy for standard-risk patients (n=11) and 39.6Gy for high-risk patients (n=19). Six weeks after radiation therapy, patients received four cycles of high-dose chemotherapy with autologous peripheral blood stem cell transplantation according to the SJMB96 regimen. We found that 5-year overall survival was 80.8% among standard-risk patients and 74.2% among high-risk patients. No treatment-related deaths occurred. Eight patients who experienced recurrence died within 80 months of diagnosis. As these treatment outcomes are comparable to those previously reported outside of Japan, our findings indicate that this regimen is a therapeutic option for medulloblastoma patients in Japan.

MBCL-50. DISMAL OUTCOME OF HIGH RISK MEDULLOBLASTOMA TREATED WITH CHEMOTHERAPY FIRST APPROACH IN MALAYSIA

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INTRODUCTION: Patients with high risk medulloblastoma are treated either with high dose chemotherapy or hyperfractionated radiotherapy. Both approaches are not feasible in resource-limited countries. POG9031 trial has reported favourable outcome for high risk medulloblastoma using standard chemotherapy and radiotherapy only. Hence, we have adopted the protocol using chemotherapy first approach due to logistical reasons. OBJECTIVE: To review the outcome of children diagnosed with high risk medulloblastoma in Hospital Kuala Lumpur. METHODS: Patients diagnosed with high risk medulloblastoma between January 2015 and June 2018 treated using the chemotherapy first approach as per POG9031 protocol were identified. Data was then extracted and analysed. RESULTS: Nine patients were identified, 3 boys and 9 girls. Median age was 9.3 years (range 2.6 – 15.9 years). Median follow up for survivors are 3.6 years. Five patients (55.6%) had macroscopic metastatic disease at diagnosis. All patients had significant residual disease post-op. Only 3 patients are disease free till last follow up, giving a 3 years event free survival of 16%. Of the 6 patients who had relapsed, 4 have died, giving a 3 years overall survival of 46%. Patients with no metastasis at diagnosis (M0) fared better with 3 years event free survival of 38%, but 3 years event free survival for patients with macroscopic metastatic disease (M+) was 0%. CONCLUSION: Outcome of children with high risk medulloblastoma treated with chemotherapy first approach was dismal.

MBCL-51. POST-AUTOLOGOUS HEMATOPOIETIC CELL TRANSPLANTATION (AUHCT) PRACTICES FOR YOUNG CHILDREN WITH MALIGNANT BRAIN TUMORS

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BACKGROUND: "Head Start" protocols have used autologous hematopoietic stem cell transplant (AuHSCT) for infants and young children with malignant brain tumors in order to avoid cranial irradiation. The post-AuHSCT practice for children with a brain tumor diagnosis varies greatly. The goal of this research study is to explore practices and attitudes about post-AuHSCT care for children with brain tumors. DESIGN: An anonymous REDCap survey link was provided to all site primary investigators and additional support personnel at "Head Start" institutions. The survey questions defined the role of the medical provider completing the form and explored the various practices relating to transition, management, communication and overall satisfaction. RESULTS: Twenty-one individual replies have been received so far. The majority report that prophylactic medicines were discontinued upon WBC recovery; however, management of discontinuation was split evenly between the neuro-oncology and stem-cell transplant teams. Nearly half of responders follow T-cell recovery following transplant without immunology guidance. Post-AuHCT vaccination practices are highly variable, with no clear consensus. Lastly, most responders reported adequate ease of transition and communication between the neuro-oncology and transplant teams. CONCLUSIONS: This work underscores the need for both multidisciplinary communication for children with brain tumors in the post-AuHCT period and for the development of standardized vaccination and other prophylaxis practices.

MBCL-52. ENDOCRINE PROFILE AFTER MEDULLOBLASTOMA TREATMENT

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BACKGROUND: Treatment of medulloblastoma has evolved substantially with more chemotherapy, risk-adapted dosing of radiotherapy (RT) and new RT techniques. We present the endocrine profile for our patients treated over a 20-year period. METHODS: The charts of patients treated for medulloblastoma between 1/1/00 and 31/12/19 were reviewed. 105 were available. Group 1 received chemotherapy alone, Group 2 received 23.4 Gy whole CNS RT with a posterior fossa (PF) boost to 54 Gy, Group 3 received > 35 Gy whole CNS RT with PF boost to 54–59 Gy, Group 4 received PF RT to 54 Gy. All received chemotherapy according to national guidelines or clinical trials relevant at the time. RESULTS: Group 1 (M:F 11:6, 7 survivors mean age 2 years range 1–7) had no endocrinopathies. At 5 years from diagnosis Group 2 (M:F 15:13) and Group 3 (M:F 35:14) had the following % RESULTS: Survival 77:61; Growth Hormone deficiency 92:100; Thyroid deficiency 75:81; ACTH deficiency 42:33. Girls were more likely to need sex hormone replacement than boys. Group 4 (M:F 7:5 mean age 2) were all treated in the first decade. 3 survivors, one GH deficiency, one thyroxine deficiency, one both. CONCLUSIONS: There is a trend to earlier endocrinopathies in the group 3 vs group 2 patients, but it does not reach statistical significance. Girls are more likely to need sex hormone replacement than boys. This investigation provides a contemporary profile of