planned protocol therapy but relapsed 6 months following the completion of therapy. In both cases, relapse was local and disseminated. Further accrual was halted. Both subjects were salvaged with CSI/XRT followed by adjuvant chemotherapy. Of the remaining 4 subjects, two had recently completed planned protocol therapy at the time of study closure and received CSI/XRT while in remission and remain in remission approximately one year from the completion of treatment. One subject aborted protocol therapy and transitioned to a Head Start regimen and remains in remission 10 months from completion of therapy. The final subject had just completed protocol therapy and had new areas of restricted diffusion concerning for early relapse. Went on to receive CSI/XRT but subsequently relapsed and is now receiving salvage chemotherapy. CONCLUSIONS: Chemotherapy following ACNS0331, omitting CSI/XRT, appears to be insufficient for the treatment of non-metastatic WPM.

MBCL-26. FACTORS ASSOCIATED WITH LONGER SURVIVAL AFTER FIRST RECURRENCE IN MEDULLOBLASTOMA BY MOLECULAR SUBGROUP AFTER RISK-BASED INITIAL THERAPY

Murali Chintagumpala[1], Colton Terhune[2], Lin Tong[3], Eric Bouffet[4], Ute Bartels[4], Michael Fisher[5], Tim Hassall[6], Shridharan Gururangan[7], Kristin Schroeder[8], Jordan Hansford[9], Dong Anh Khuong Quang[9], Richard Cohn[10], Stewart Kellie[11], Geoffrey McCowage[12], Kyle Smith[3], Paul Northcott[3], Giles Robinson[3], and Amar Gajjar[3]; [1]Texas Children's Hospital, Houston, TX, USA, [2]University of South Hampton, South Hampton, United Kingdom, [3]St. Jude Children's Research Hospital, Memphis, TN, USA, [4]Hospital for Sick Children, Toronto, Ontario, Canada, [5]Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, USA, [6]Children's Health Queensland, Brisbane, Queensland, Australia, [7]University of Florida, Gainsville, FL, USA, [8]Duke University, Durham, NC, USA, [9]Royal Children's Hospital, Melbourne, Victoria, Australia, [10]Sydney Childre, Sydney, New South Wales, Australia, [11]Westmead Children's, Sydney, New South Wales, Australia, [12]Westmead Children's, Sydney, NSW, Australia

OBJECTIVE: To evaluate differences in time to recurrence among molecular subgroups of medulloblastoma treated on a single protocol and to identify factors associated with survival after first recurrence. METHODS: Time to recurrence following SJMB03 treatment was compared across methylation subgroups among relapsed patients. Therapies received subsequent to relapse were noted. Kaplan-Meier methods and log-rank tests were used for statistical analyses. RESULTS: 74 of 330 medulloblastoma patients developed recurrence after initial therapy. (38 Standard-Risk; 36 High-Risk). The 2- and 5-year survival after first recurrence was 30.4% and 14.6% respectively. DNA methylation-based subgroups from initial diagnosis were SHH (n=14), Group 3 (n=24), Group 4 (n=26), and unclassified (n=8). None of the pts with WNT MB had recurrent disease. Median time to first recurrence was 1.23, 0.91, and 3.09 years in SHH, Group3, and Group 4 respectively. Group 4 patients had longer post-recurrence survival than others (p-value=0.0169). Clinical risk at diagnosis (p-value=0.337), anaplasia (p-value=0.4032), *TP53* (p-value=0.1969), *MYC* (p-value=0.8967), and *MYCN* (p value = 0.9404) abnormalities were not associated with post progression survival. Patients who received any therapeutic modality (chemotherapy, re-radiation and second surgery) had longer survival and those who had all three (n=10) had the best outcome (p-value<0.0001). CONCLUSION: Outcome after recurrence in medulloblastoma is dismal, however, association with subgroups is still present. Group 4 patients had a longer time to recurrence and post progression survival. No other prognostic factor at initial diagnosis was associated with outcome after recurrence. Patients who received all 3 types of conventional therapy had better survival.

MBCL-27. ASSOCIATION OF MEDULLOBLASTOMA WITH CHARCOT-MARIE-TOOTH DISEASE

Kenichiro Watanabe[1], Kazuyuki Komatsu[1], Koji Kawaguchi[1], Risa Makino[1], Takayuki Takachi[1], Taemi Ogura[1], Yasuo Horikoshi[1], Ryuji Ishizaki[2], Hideto Iwafuchi[3], and Yuzuru Tashiro[2]; [1]Department of Hematology and Oncology, Shizuoka Children's Hospital, Shizuoka, Japan, [2]Department of Neurosurgery, Shizuoka Children's Hospital, Shizuoka, Japan, [3]Department of Pathology, Shizuoka Children's Hospital, Shizuoka, Japan

Charcot-Marie-Tooth disease (CMT) is one of the most common hereditary neurological disorders and damages peripheral nerves that results in motor and sensory disturbance. Association of medulloblastoma (MBL) with CMT has been rarely reported. A one-year-old male was referred to our hospital because of cerebellar mass. He had partial resection of the tumor, and was pathologically diagnosed as having desmoplastic nodular medulloblastoma. He received chemotherapy according to the HIT protocol, however, developed severe peripheral neurotoxicity in the initial stage of the treatment. Reinvestigation of family history revealed his mother, grandmother, and aunt had muscle weakness. We suspected he had an inherited neurological disease including CMT, and discontinued administration of vincristine. Fluorescence in situ hybridization analysis detected duplication of PMP22 gene located on 17p11.2, confirming the diagnosis of CMT1A. He completed the rest of chemotherapy without vincristine, and remained in complete remission for four years from the end of treatment. In the literature, there are reports of patients with CMT who developed MBL and were complicated with severe peripheral neurotoxicity due to the use of vincristine. The present case, along with previous reports, suggests that medulloblastoma can develop in patients with CMT and reminds the importance of recalling the possibility of CMT when patients develop severe chemotherapy-induced peripheral neurotoxicity upon use of vincristine. Desmoplastic nodular medulloblastoma may be successfully treated by chemotherapy without vincristine.

MBCL-28. LONG-TERM FOLLOW-UP RESULTS OF REDUCED DOSE CRANIOSPINAL RADIOTHERAPY AND TANDEM HIGH-DOSE CHEMOTHERAPY IN PATIENTS WITH HIGH-RISK MEDULLOBLASTOMA

Ji Won Lee[1], Do Hoon Lim[2], Meong Hi Son[1], Ki Woong Sung[1], Hee Won Cho[1], Hee Young Ju[1], Ju Kyung Hyun[1], Keon Hee Yoo[1], Hye Lim Jung[3], Hong Hoe Koo[1], Yeon-Lim Suh[4], Yoo Sook Joung[5], and Hyung Jin Shin[6]; [1]Department of Pediatrics, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Republic of Korea, [2]Department of Radiation Oncology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul,Republic of Korea, [3]Department of Pediatrics, Kangbuk Samsung Hospital, Sungkyunkwan University School of Medicine, Seoul, Republic of Korea, [4]Department of Pathology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Republic of Korea, [5]Department of Psychiatry, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Republic of Korea, [6]Department of Neurosurgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Republic of Korea

BACKGROUND: In this study, we report the follow-up results of reduced-dose of craniospinal radiotherapy (CSRT) followed by tandem high-dose chemotherapy (HDCT) in patients with high-risk medulloblastoma (MB). METHODS: Newly diagnosed high-risk MB patients (metastatic disease, postoperative residual tumor > 1.5 cm^2 or large cell/anaplastic histology) over 3 years of age were enrolled in this study. Two cycles of pre-RT chemotherapy, RT including reduced-dose CSRT (23.4 or 30.6 Gy), 4 cycles of post-RT chemotherapy and tandem HDCT were given. NanoString and DNA sequencing were done with archival tissues. RESULTS: Forty patients were enrolled, and molecular subgrouping was possible in 21 patients (2 WNT, 3 SHH, 8 Group 3 and 8 group 4). All patients including two patients who experienced progression during the induction chemotherapy underwent HDCT. Relapse/progression occurred only in four patients (10-year cumulative incidence 10.4 ± 0.3%). However, six patients died from treatment-related mortality (TRM) (4 acute TRMs and 2 late TRMs) resulting in 18.5 ± 0.5% of 10-year cumulative incidence. Taken together, the 10-year event-free survival and overall survival were 71.1 ± 8.0% and 68.9 ± 8.5%, respectively. Late effects were evaluated in 25 patients and high-tone hearing loss, endocrine dysfunction, dyslipidemia, and growth retardation were common. CONCLUSIONS: Strategy using tandem HDCT following reduced-dose CSRT showed promising results in terms of low relapse/progression rate, however, the high TRM rate indicates that modification of HDCT regimen and careful selection of patients who can have benefit from HDCT will be needed in the future study.

MBCL-29. PHASE I/II STUDY OF SEQUENTIAL HIGH-DOSE CHEMOTHERAPY WITH STEM CELL SUPPORT IN CHILDREN YOUNGER THAN 5 YEARS OF AGE WITH HIGH-RISK MEDULLOBLASTOMA

Christelle Dufour[1], Julien Masliah-Planchon[2], Marie-Bernadette Delisle[3], Anne Geoffray[4], Rachid Abbas[1], Franck Bourdeaut[2], Anne-Isabelle Bertozzi[3], Cecile Faure-Conter[5], Celine Chappe[6], Emilie De Carli[7], Natacha Entz-Werle[8], Fanny Fouyssac[9], Nicolas Andre[10], Christine Soler[11], Claire Pluchart[12], Gilles Palenzuela[13], Pierre Leblond[14], and Jacques Grill[1]; [1]Gustave Roussy, Villejuif, France, [2]Curie Institute, Paris, France, [3]Toulouse University Hospital, Toulouse, France, [4]Fondation Lenval Children's Hospital, Nice, France, [5]Institut d'Hématologie et d'Oncologie pédiatrique, Lyon, France, [6]Rennes University Hospital, Rennes, France, [7]University Hospital, Angers, France, [8]CHU of Strasbourg, Strasbourg, France, [9]Children's Hospital, Nancy, France, [10]CHU Timone, Marseille, France, [11]CHU of Nice, Nice, France, [12]CHU of Reims, Reims, France, [13]CHU of Montpellier, Montpellier, France, [14]Oscar Lambret, Lille, France

PURPOSE: To assess the 3-year EFS rate of children younger than 5 years of age with high-risk medulloblastoma (MB) treated according to the prospective multicenter trial HR MB-5. PATIENTS AND METHODS: After surgery, all children received 2 cycles of Etoposide- Carboplatine. If par-