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BACKGROUND: Plexiform neurofibromas (PNs) can cause significant morbidity. In this phase 2 study, we assessed imaging and functional outcomes to the MEK-inhibitor Binimetinib in pediatric patients with PNs. METHODS: Children (age 1–17 years) with PN that were progressive or causing significant morbidity were eligible. Binimetinib is dosed twice-daily (starting dose of 32mg/m^2) for maximum of 24 four-week courses. Participants with partial response (PR; >20% decrease in PN volume on central MRI review) at cycle 12 may stay on therapy. Participants undergo MRI and functional assessments at baseline and after courses 4, 8, 12, 18 and 24. Functional assessments are based on PN location. RESULTS: Here we present 1-year response data. Twenty participants (55% male) with median age 12 years (range 2–16 years) enrolled; 19 are evaluable for response. Median baseline tumor volume was 326 ml (range, 8-6661 ml). Fourteen participants (74%) met criteria for PR, with 11 achieving PR by course 5. Median maximal PN volume reduction was 25.5% (range, 9–54%). As of August 2020, 14 participants received at least 12 cycles of Binimetinib; 10 remain on therapy. Off study reasons include treatment associated toxicities (n=2), subject withdrawal (n=2), non-compliance (n=2), prolonged treatment delay (n=1), and lack of response (n=3). Thirteen participants underwent dose reduction. Institution-reported related grade 3 toxicities included dry skin, weight gain, muscle weakness, rash, paronychia, cellulitis, diarrhea, gastric hemorrhage and CPK increase. CONCLUSIONS: Binimetinib appears reasonably well-tolerated and shows promising activity in children with NF1-associated PNs. Outcomes on functional improvement will be reported at the meeting.

NFB-18. IMMUNE FUNCTION IN CHILDREN TREATED WITH TRAMETINIB

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BACKGROUND: Trametinib (Tr) has been applied in the treatment of children with various tumor types, often for prolonged periods. Little is known regarding immune function (IF) following prolonged Tr in this age group. OBJECTIVE: Describe laboratory measures of IF in children on Tr. METHOD: Patients receiving Tr had low grade glioma with BRAF anomalies (6), or neurofibromatosis-1 (16) with glioma or plexiform neurofibroma. IF was evaluated using leukocyte/lymphocyte counts, immunoglobulin levels, and antibody titres. RESULTS: 22 patients received Tr. 2 also received Dabrafenib. Median age at Tr initiation of Tr was 7.75 years. As of June 2020, 7 patients have had IFT; results are pending on 15. Median duration of Tr therapy at time of IF was 3.5 years (0.8 – 4). In these 7 patients, median white cell count was 6.9 x 10^9/L (4.1 – 12.6), neutrophils 4.2 x 10^9/L (1.8 – 6.8) and lymphocytes 3.2 x 10^9/L (1.4 – 7). IgG levels, B cells and CD8 cytotoxic T cells were normal across 7/7 patients:medians 9.47 g/L (8.62 – 17), 0.51 x 10^9/L (0.2 – 1.26) and 0.58 x 10^9/L (0.25 – 2.03) respectively. CD3 and CD4 T cells: median 2.08 x 10^9/L (0.67 – 4.62) and 1.34 x 10^9/L (0.35 – 2.31), borderline low in 1 heavily pre-treated patient. An adequate immune response was present in all 4 vaccine antigens tested in 5/5 patients. CONCLUSION: IF appears relatively intact, relevant for immunisation and infection precautions in children on Tr. Data on the complete cohort will be presented.

NURSING/PATIENT CARE

NURS-01. INTRACEREBROVENTRICULAR DRUG ADMINISTRATION FOR TREATMENT OF PEDIATRIC BRAIN TUMORS

Caroline Fitzgerald, and Kathryn Matson; Boston Children's Hospital, Boston, MA, USA

Intrathecal (IT) chemotherapy, given via lumbar puncture (LP) or an intracerebroventricular (ICV) device has become a safe and effective way to deliver chemotherapy into the cerebrospinal fluid (CSF) space. The blood brain barrier makes treating tumors with CSF dissemination difficult with systemic chemotherapy alone. IT chemotherapy is often necessary for tumors which disseminate into the CSF space including embryonal tumors and choroid plexus carcinomas. It is also used for relapsed or recurrent tumors. Giving IT chemotherapy via an ICV device instead of via an LP can be preferable as it requires no deep sedation and allows for more uniform drug distribution. Drugs given IT include methotrexate, cytarabine, hydrocortisone, etoposide, and topotecan. ICV devices can be placed in patients with adequate CSF flow and a flow study can be done if needed to confirm. Accessing the ICV device for administration of chemotherapy is typically done by a physician or nurse practitioner using sterile technique. Our institution has had success using music therapy and child life specialists for assistance with coping during the procedure as patients are awake. The procedure has few complications the most common being infection usually with skin flora. It can also cause nausea and headache. There are few long term risks.

NURS-02. CLINICAL MANAGEMENT OF PATIENTS RECEIVING CAR T CELL THERAPY FOR CNS TUMORS

Susan Holtzclaw, and Corrine Hoeppner; Seattle Children's Hospital, Seattle, WA, USA

Chimeric antigen receptor (CAR) T cells are an innovative new therapy with proven efficacy in some pediatric cancers such as leukemia and lymphoma, but much less experience in solid tumors, especially tumors of the central nervous system (CNS). Seattle Children's has three open Phase 1 CAR T cell studies (BrainChild-01, -02, and -03 targeting HER2, EGFR, and B7-H3, respectively) for recurrent/refractory CNS tumors and DIPG (BrainChild-03 only). As of December 2019, four patients have been treated at Seattle Children's Hospital with CAR T cells infused on a weekly schedule through indwelling catheters into the tumor resection cavity or ventricular system. Given the scrutiny of clinical care needed for Phase 1 studies, we are now able to report detailed clinical information that we have learned during the treatment of these patients. Clinical care includes the judicious use of steroids, the clinical support of patient's symptoms pre- and post-infusion, and the management of peritumoral edema. We will also discuss the psychosocial support needed for families who travel long distances to receive this therapy compounded by the many emotional components of being enrolled on any Phase 1 trial. Case studies and experience from a Nurse Practitioner role will be provided and discussed.

NURS-03. DEVELOPMENT OF A PATIENT-HELD TREATMENT SUMMARY FOR PAEDIATRIC CNS TUMOUR PATIENTS

Rachel McAndrew, Bernadine Wilkie, Mark Brougham, and Jo Phillips; NHS Lothian, Edinburgh, Scotland, United Kingdom

BACKGROUND: Following the Scottish Government Cancer Plan 2012–15*(1)* 'End of Treatment' summaries for paediatric oncology patients treated in SE Scotland have been successfully implemented. However, it became evident that the particular needs of patients with CNS tumours were not adequately captured on the standardised documentation. METHODS: In view of these difficulties an alternative document was prepared specifically for this patient cohort by the multi-disciplinary team, including Nurse Specialists, Paediatric Neuro-oncology and Neuro-psychology. This was designed to be a flexible, fluid summary to be used for all such patients regardless of tumour grade or treatment modality and included those undergoing surveillance only. OUTCOMES: The document is primarily completed by the Neuro-Oncology Nurse Specialist alongside the patient and family, usually following initial treatment and is used alongside their holistic needs assessment. The document is circulated to all involved professionals, including Primary Care, and a copy is retained by the patient. This then provides a concise source of information detailing diagnosis and treatment, any specific ongoing sequelae and details of red flag symptoms to alert patients and health professionals to the potential of relapse or other associated significant health problems. These treatment summaries are currently being piloted and have been well received thus far. They will be formally audited in due course with the aim to use nationally throughout Scotland in future.

NURS-04. COMBINATION OF NEURO-ONCOLOGY AND DERMATOLOGY CLINICS IMPROVE THE MANAGEMENT AND KNOWLEDGE OF SKIN-RELATED TOXICITIES WITH MEK AND BRAF TARGETED THERAPY

Tara McKeown, Irene Lara-Corrales, and Andrea Cote; Hospital for Sick Children, Toronto, ON, Canada

BACKGROUND: The recent advancement in treating pediatric low grade glioma has led to upfront use of MEK and BRAF (MAPK) inhibitor therapy. At the Hospital for Sick Children we are the National leaders in treating pediatric oncology diagnosis with MAPK therapies. DESIGN: After treating several patients on MAPK inhibitors with various degrees of skin toxicity, we found we had poor and inconsistent access to dermatology services and as oncology practitioners had limited front-line knowledge about skin management. It was determined that a more formalized expertise and time with dermatology was needed. In 2018, in combination with the derma-