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OTHR-06. KNOWLEDGE GAP AMONG HEALTH CARE PROVIDERS IN CHILDHOOD CENTRAL NERVOUS SYSTEM TUMORS: RESULT FROM AN INTERNATIONAL MULTICENTER SURVEY
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BACKGROUND: Central nervous system (CNS) tumors in children are associated with a longer delay in diagnosis. One of the contributing factors is the lack of awareness regarding childhood CNS tumors and their presentation among health care providers. To evaluate the knowledge gap among health care providers, we conducted a cross-sectional survey that was distributed globally. METHODS: The survey was disseminated to health care practitioners via electronic mail in November 2018 and it was closed in March 2020. The participants were asked to complete a pre-test survey, re-quested to view a CNS tumor education seminar, and subsequently complete a post-test survey. The survey had nine questions focusing on CNS tumor symptoms, pre-diagnosis symptom interval (PSI), and imaging indication. The knowledge gap was evaluated with pre-test and post-test scores. RESULTS: 889 pre-test and 392 post-test responses were received. The majority of the respondents were from Asia, with a percentage of 73.1% and 87.5% in pre-test and post-test respectively. For the pre-test, the median score for accurate answers was 40.0% (range:13-1-92%). Interestingly, a high rate of correctness was achieved in the post-test with a median score of 77.1% (14.9-98.2%). In the pre-test, only 18.7% of the participants responded precisely that Cushings' tris is a less common symptom and just 15.0% recognized that older children >10 years old are at risk for endocrine sequelae. Surprisingly, 21.9% falsely reported that patients with malignant tumors experience the longest PSI, and 54.5% of the respondents wrongly selected medulloblastoma as the commonest CNS tumor. Overall, the pre-test scores among pediatricians and professionals with >10 years of experience due not demonstrate improved knowledge when compared to other specialties. CONCLUSIONS: The survey analysis showed a significant knowledge gap regarding childhood CNS tumors among health care providers. Therefore, raising professional awareness is very important and can be achieved through targeted educational strategies.

OTHR-07. A NEW FRAMEWORK FOR MISSED VALUE TOLERANT DATA INTEGRATION
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Dataset integration is common practice to overcome limitations, e.g., in statistically underpowered omics datasets. This is of particular importance when analyzing rare tumor entities. However, combining datasets leads to the introduction of biases, so called ‘batch effects’, which are due to differences in quantification techniques, laboratory equipment or used treatment protocols. A common problem is the old batch quantification for features like gene transcripts or proteins within a dataset. These missing values can appear at random in a given dataset and also get introduced by combination of multiple datasets. Currently, strategies beyond common normalization for such datasets exist, but are either missing entirely or are unable to handle the presence of data points and therefore rely on error-prone data imputation. We introduce a framework that enables batch effect adjustments for combined datasets while avoiding data loss by appropriately missing values without imputation. The underlying idea is based on a matrix dissection approach, adjusting common information from the integrated dataset under guarantee of sufficient data presence. The strategy is implemented within the R environment and linked with popular software stacks that are built on top of R. Successful data adjustment is exemplarily shown for proteomic data generated by different quantification approaches and LC-MS/MS instrumentation setups.

OTHR-08. PEDIATRIC NEUROLOGIC ASSESSMENT IN NEURO-Oncology (pNANO) SCALE: A TOOL TO ASSESS NEUROLOGIC FUNCTION FOR RESPONSE ASSESSMENT IN NEURO-Oncology
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CONCLUSIONS: The Neurologic Assessment in Neuro-Oncology (pNANO) Scale was developed to provide a standardized metric to objectively measure neurologic function in adult brain tumor patients, complements radiographic assessment in evaluating outcomes of neuro-oncology patients in clinical trials and clinical practice. Currently, there is no standardized measure for neurologic function in pediatric neuro-oncology patients despite their distinct clinical presentations and tumor locations. Therefore, we developed a dedicated pediatric pNANO Scale. METHODS: An expert panel of pediatric neuro-oncologists convened bi-weekly over 5 months to draft the pNANO Scale as an objective and quantifiable measure of neurologic function in children that can be administered during routine examination by pediatric-trained providers of any subspecialty and be utilized together with other indicators to assess response in clinical trials. RESULTS: Ten relevant domains of neurologic function were identified based on common pediatric brain tumor locations: gait, strength, sensation, cranial nerve function, visual fields, facial strength, coordination, scintillations, extraocular movements, dysarthria and dysphasia. For each domain, developmental age appropriate levels of function were defined and categorized. Each domain, based on direct observation and testing during any routine neurological examination, can be used for longitudinal monitoring. CONCLUSIONS: The pNANO Scale has been developed and aims to provide an objective metric of neurologic function for pediatric brain tumor patients. This scale will be tested for reliability, feasibility and inter-observer variability. Consistent evaluation of neurologic function using pNANO along with radiographic assessment will enable more comprehensive and standardized response assessment in pediatric neuro-oncology patients enrolled in clinical trials.

OTHR-09. THE PREVALENCE AND COMPLEX MANAGEMENT OF MEK INHIBITOR INDUCED CUTANEOUS SIDE EFFECTS
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BACKGROUND: Cutaneous side effects commonly occur with MEK inhibitor (MEKi) therapy and can be challenging to manage. METHODS: A retrospective chart review was performed on sixteen pediatric patients treated with MEKi therapy for at least three months. These patients were diagnosed with either a brain tumor, plexiform neurofibroma, Langerhans Cell Histiocytosis, or Parkes Weber Syndrome (PWS). OBJECTIVES: To describe cutaneous side effects from MEKi therapy, compare the side effect profiles of patients treated for different diagnoses, and compare the side effect profiles between different MEKi agents. RESULTS: The most prevalent cutaneous toxicities for all eligible patients were acneiform rash (81%, most common), paronychia (50%, second most common), hair changes and xerosis. Mean number of cutaneous skin toxicities were 3.2 (trametinib group: 3.4, selumetinib group: 2.8). Only one patient, treated with trametinib for PWS, developed paronychia within the first cycle of therapy; average time to development was 2.6 months into therapy. Most exhibited improvement overtime following intervention. Those patients who experienced an acneiform rash, the average number of recommended interventions were 3.1. Sixty two percent of these patients developed acne within the first cycle; average time to development was 2.6 months into therapy. Most exhibited improvement overtime following intervention. Those patients who experienced paronychia were treated with a mean of 3.1 interventions – 37.5% developed paronychia within the first cycle of therapy; average time to development was 3.9 months into therapy. CONCLUSIONS: Cutaneous side effects are common, occur early in therapy, and require multiple interventions. The number and complexity of these interventions may further
complicate the overall management of cutaneous side effects given that the responsibility of the administration falls on the patient. This differs from traditional chemotherapy regimens.

**OTHR-10. PILOCYTIC ASTROCYTOMA WITH RESPECT TO TREATMENT**

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AIM: To describe the sizes of pilocytic astrocytoma with respect to treatment.

Methodology Pilocytic pilocytic astrocytomas cases from 2001 to 2021 were retrospectively reviewed in this Institutional Review Board approved study. Imaging reports, location of tumour, maximum dimension of tumour at diagnosis, treatment given (operation/chemotherapy/radiotherapy), degree of tumour excision were captured. RESULTS: Imaging was available in 33 with 23 centered in the posterior fossa (1 extending into thalamus), 4 in suprasellar region, 2 in cerebral hemisphere, 2 in thalamus, 1 in pineal thalamic region and 1 in cervices tumour spine. Tumor dimension at presentation varied from 5.40 cm x 2.34 cm. Tumor size at presentation did not show significant correlation with age. 30 patients underwent operation with tumours completely excised in 15 and partially excised in 14 and no postoperative information for 1. Three patients where tumour involved the thalamus, did not have operation and were given radiotherapy, average size of tumour being 4.57 ± 1.13 cm, compared to the 5.59 ± 2.34 cm of tumours that underwent operation (p=0.06). Completely excised tumours measured 6.29 ± 2.04 cm at presentation while incompletely excised ones measured 4.76 ± 2.53 cm, not significantly different (p=0.09). Unoperated tumours were statistically smaller than those completely excised (p=0.02). One of the completely excised tumours was located in the parietal cerebral hemisphere with the rest of the 15 in the posterior fossa. Seven of the completely excised tumours were located in the posterior fossa with 4 in suprasellar region, 1 in thalamus, 1 in spine and 1 in cerebral hemisphere. 3 patients with uncompletely excised tumours (1 cerebral, 1 post fossa, 1 spine) had post-operative radiation while 2 suprasellar tumours were given post-operative chemotherapy.

CONCLUSION: Completely excised tumours are mainly located in posterior fossa. Tumours that were not operated on are located in thalamus and significantly smaller than tumours which are completely excised.

**OTHR-11. THE EFFECTS OF THE COVID-19 PANDEMIC ON THE TIME TO DIAGNOSIS IN PEDIATRIC PATIENTS WITH PRIMARY CENTRAL NERVOUS SYSTEM TUMORS**

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Primary central nervous system (CNS) tumors are a leading cause of death and disability amongst pediatric cancer patients. The early identification of symptoms of tumour biology in preventing diagnostic delays. In 2018, Akron Children’s Hospital published data on our response time to brain tumor diagnosis and launched educational programs in an effort to decrease diagnostic delays due to the unique pandemic constraints. A retrospective chart review was performed on patients at Akron Children’s Hospital to evaluate both for pre- (diagnosed Jan 1, 2018-February 29, 2020) and post-diagnosis (diagnosed Mar 1, 2020-June 8, 2021) groups. Both subsets were evaluated statistically and were similar in all respects including demographics, symptomatology, tumor location, tumor type, and survival. The pre-COVID-19 group demonstrated a median TDI of 43.5 days, while the post-COVID-19 group demonstrated a median TDI of 47.5 days. Low vs. high-grade lesions increased from 32 to 59 days and for high-grade lesions decreased from 60 to 27.5 days in the post-pandemic cohort. Overall, this demonstrates a maintained average time to diagnosis for patients despite the pandemic restrictions in place. In addition, the median TDI of 47.5 days. Low vs. high-grade lesions suggest that tumors with a more subtle onset of symptoms were disproportionately affected, and highlight a population for intervention during the continued pandemic.

**OTHR-12. INCIDENTALOMAS; SHOULD BENIGN, INDETERMINATE MRI FINDINGS BE DISCUSSED AT THE PAEDIATRIC NEURO-ONCOLOGY MDT**

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BACKGROUND: In 2020, 503 patients were discussed at the weekly neuro-oncology MDT meeting, at a UK tertiary paediatric hospital. This was a 50% increase from 2016. Anecdotally, an increased number of non-tumours were being discussed. OBJECTIVES: To determine whether Paediatric Neuro-oncology MDT is consistent with follow up of benign, incidental findings on MRI scans and whether these lesions should be discussed. METHODS: MDT discussion notes were reviewed for all incidental and benign MRI head findings between 05/09/18 and 02/09/20.RESULTS: 59 MRI head lesions, that were deemed to be both incidental and benign were discussed at meetings from the local hospital; this may be due to availability of the MDT or complexity of the patient cohort. Findings discussed included pineal cysts (24), arachnoid cysts (3), epidermoid cysts (1), pituitary lesions (4) and other indeterminate lesions or signal change (27). The majority of children received serial MRI imaging after MDT. There were large inconsistencies in the follow-up interval recommended, even within similar pathologies. For example, pineal cysts follow up ranged from no repeat imaging to multiple repeat scans with 6 and 12 month intervals. ‘Complex’ cysts were more likely to receive follow up but there is little consensus between radiologists about criteria for a ‘complex cyst’. None of the lesions reviewed changed significantly over time. Perhaps most strikingly, only 2/27 of the ‘indeterminate lesions’ have been discharged from the MDT. CONCLUSIONS: As clinical reasoning is seldom recorded in reports or MDT discussion notes the reasons for different surveillance recommendations is difficult to ascertain retrospectively. However, as MRI scans become more readily available incidental and being findings will increase, potentially putting great pressure on an already stretched service. Serial MRI imaging also creates unnecessary anxiety for both the child and their family.

**OTHR-13. EFFECTIVITY AND TOXICITY OF OFF-LABEL TREATMENT WITH TRAMETINIB MONOTHERAPY OR IN COMBINATION WITH DABRAFENIB IN CHILDREN WITH RELAPSED OR REFRACTORY BRAIN TUMOR**

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INTRODUCTION: Most pediatric low grade gliomas, and a substantial part of PXAs, have a BRAF alteration/activated MAPK pathway. This group often requires several lines of therapy, which coincides with significant morbidity. There is growing evidence that molecular targeted treatment may be beneficial for this population. Here we report the toxicity and efficacy of patients treated off-trial with either trametinib monotherapy or in combination with dabrafenib.

METHODS: We performed a single-center retrospective chart review of all neuro-oncological patients who received trametinib monotherapy or in combination with dabrafenib (comparative). The study was approved by the Princes Maxima Center between April 1st, 2018 and December 31st, 2021.

RESULTS: 22 patients (ages 1-21 years) were included of whom 14 received trametinib monotherapy (BRAF-KIAA fusion, n=10) and eight received dabrafenib/dabrafenib combination therapy (BRAFV600 mutation, n=8). All patients on trametinib monotherapy (n=14) developed skin problems such as rash (100%), dry skin (86%), paronychia (71%) and eczema (64%). Eight patients (57%) had at least one adverse event (AE) grade 3. Patients on trametinib and dabrafenib showed similar toxicity, although with lower prevalence and no paronychia. One patient (7%) on trametinib developed an AE grade 3. Median treatment time of trametinib was 514 days (IQR 455) and for trametinib and dabrafenib 360 days (IQR 512). Five patients (36%) on trametinib are still on treatment and nine patients (64%) stopped treatment due to e.g. tumor progression or toxicity. All patients on trametinib and dabrafenib are still on treatment. Best overall response of trametinib monotherapy (n=14 evaluated) was observed as partial response (50%), stable disease (33%) and progressive disease (17%). Combination therapy (n=7 evaluated) brought 100% partial response. CONCLUSION: Dermatologic toxicities are mostly seen in trametinib monotherapy or in combination with dabrafenib. Despite moderate toxicity patients seem to benefit from treatment. Results suggest that combination therapy has a more favorable toxicity profile than monotherapy.

**OTHR-14. RESPONDING TO THE COVID CHALLENGE: EVALUATING A NEW METHOD OF ADVERSE EVENT RECORDING IN RESPONSE TO REMOTE WORKING**

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BACKGROUND: The SARS-COV2 pandemic had huge impact on how clinical research is conducted when clinical research coordinators (CRC) traditional working to working remotely. An urgent transition of paper documentation into electronic formats had to occur without compromising participant safety or data integrity. Adverse event (AE) reporting had previously been captured in various paper formats with wet signature. AEs, attribution, severity, and clinical significance had to be changed into being electronically captured and incorporated into the clinical record that captures the events in

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