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BACKGROUND: We have previously documented the presence of diagnostic delays in children with central nervous system (CNS) tumors in the United States. This study serves to expand and validate the previously established baseline from symptom onset to definitive diagnosis in children with newly-diagnosed CNS tumors. **DESIGN:** The medical records of children with newly-diagnosed CNS tumors were retrospectively reviewed from January 2004 to December 2017 at Nationwide Children's Hospital, Akron Children's Hospital and Riley Hospital for Children at IU Health. Records were reviewed for age, gender, tumor type, presenting symptoms, number of healthcare visits prior to diagnosis, time interval (in months) from onset of symptoms to definitive diagnosis and any associated genetic syndromes. **RESULTS:** Of the 768 patients with newly-diagnosed CNS tumors, the median time interval from symptom onset to definitive diagnosis was 40.5 days while the mean symptom interval was 144 days (range < 1 to 5,475 days). The median age of diagnosis was 7 years, with a male predominance (57%). This expanded cohort continues to reveal that pediatric brain tumor patients most often seek care at the primary care level, although many patients were seen in various multiple subspecialty clinics prior to diagnosis. **CONCLUSIONS:** This multi-institutional cohort study updates our previously documented single state time interval and provides a consistent Midwest "benchmark" to improve awareness for children with brain tumors through the adaptation of the UK 'HeadSmart,' now renamed 'BrainFirst.' Additionally, future work could include a prospective registry to better examine potential risk factors for delays in diagnosis.

EPID-12. TEMPORAL AND GLOBAL GEOGRAPHIC VARIATION IN THE INCIDENCE OF PEDIATRIC CNS TUMORS, 1998–2012

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AIMS: To describe the temporal and geographic variation in the incidence of pediatric CNS malignancies worldwide, presenting analyses by sex, period, region, and histological subtype between 1998 and 2012. **METHODS:** Data were extracted from volumes IX to XI of the Cancer Incidence in 5 Continents, covering the periods 1998–2002 (1), 2003–2007 (2), and 2008–2012 (3). We pooled data from 44 countries, classifying them into 6 regions (Africa (AF), Asia (AS), Oceania (O), Europe (E), Central/South America (CSA), North America (NA)). Age-standardized incidence rates (ASIR per million, 0–19 years) were calculated and temporal variation was evaluated using incidence rate ratios (IRR) (95% CI). **RESULTS:** The highest incidence (Period 3) was observed in NA (34.0 and 30.2 for males and females, respectively). Astrocytic tumors were predominant in all regions, with percentages ranging between 24.5% (E, females) and 45.6% (NA, females). Increasing trends (Period 3 x 1) were observed in AS (IRR=1.15, 95% CI 1.06–1.25), CSA (IRR=1.25, 95% CI 1.01–1.55), and NA (IRR=1.05, 95% CI 1.03–1.07), for males and in AS (IRR=1.15, 95% CI 1.05–1.26) and NA (IRR=1.08, 95% CI 1.06–1.11) for females. Geographic discrepancies in time-trends were observed for astrocytomas, ependymomas, medulloblastomas, other embryonal tumors, and other specified tumors. Reductions in the incidence of unspecified tumors from period 1 to 3 were noted in E, AS, and NA, ranging from -20% (E, females) to -66% (AS, females). **CONCLUSIONS:** Heterogeneous trends and improvement in the registration of histological types were noted. Geographic variation can help to raise hypotheses to investigate etiologic factors.

EPID-13. A POPULATION-BASED ANALYSIS OF CNS TUMOR DIAGNOSES, TREATMENT, AND SURVIVAL IN CONGENITAL AND INFANT AGE GROUPS

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BACKGROUND: Congenital (<3 months) and infant (3 to 11 months) brain tumors are biologically different from tumors in older children, but epidemiology of these tumors has not been studied comprehensively. Insight into epidemiological differences could help tailor treatment recommendations by age and increase overall survival (OS). **METHODS:** Population-based data from the SEER 18 registries was obtained for 14,493 0-19-year-olds diag-

nosed with CNS tumors between 1990 and 2015. Incidence, treatment, and survival were analyzed using Chi-square and Kaplan-Meier analyses. **RESULTS:** Between the <3 month, 3–5 month, 6–11 month, and 1–19 year age groups, tumor type distribution differed significantly ($p < 0.001$); high-grade glioma (HGG) was most common in the <3-month-olds, while low-grade glioma (LGG) was most common in the other groups. 5-year OS for all tumors was 36.7% (<3 months), 56.0% (<3–5 months), 63.8% (6–11 months), and 74.7% (1–19 years) (log rank $p < 0.001$). OS by tumor type was worst for <3-month-olds with LGG, medulloblastoma, and other embryonal tumors; OS was worst for 3–5-month-olds with ependymoma, <1-year-olds collectively with atypical teratoid-rhabdoid tumor, and 1–19-year-olds with HGG (log rank $p < 0.02$ for all tumor types). <3-month-olds were least likely to receive any treatment for each tumor type and least likely to undergo surgery for all except HGG. <1-year-olds were far less likely than 1–19-year-olds to undergo radiation for embryonal tumors, as expected, but were also less likely to undergo chemotherapy. **CONCLUSIONS:** Congenital/infant CNS tumors differ pathologically, therapeutically, and prognostically from those in older children. Treatment changes could help address poorer outcomes for these young patients.

EPID-14. GABRIELLA MILLER KIDS FIRST DATA RESOURCE CENTER: COLLABORATIVE PLATFORMS FOR ACCELERATING RESEARCH IN PEDIATRIC CANCERS & STRUCTURAL BIRTH DEFECTS

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Since launching to the public in September 2018, the Gabriella Miller Kids First Data Resource Center (DRC) has made an increasing number of pediatric genomic studies available to the research community. Currently, 1.3 PBs of genomic and clinical data drawn from 12,000 participants are available across a variety of pediatric cancer and structural birth defect studies. The DRC has architected a secure, cloud-based platform with over 1,300 users that supports the ability of researchers to not only find, access, and reuse data, but also integrate, collaborate, and analyze data quickly at scale. Users can use integrations with platforms such as Cavatica for bioinformatics workflows and PedcBioPortal for cancer genomic visualizations. Additionally, a set of framework services, powered by Gen3, provide a foundation for interoperability with other large-scale data sources, platforms, and a growing ecosystem of analysis and visualization applications. These integrations allow users to search across both TARGET and Kids First clinical data in one location while allowing data governance to be maintained by the original approvers. The new "explore data" feature allows users to search across all studies in order to identify virtual cohorts. Within the portal, these cohorts can be saved and shared with collaborators for iterative refinement and analysis. With appropriate approvals, the associated genomic data can be accessed and analyzed seamlessly in Cavatica or other platforms with interoperable framework services. Additionally, gene searching capabilities will be available in 2020. Data is free to download and cloud credits are available for analysis support.

EPID-15. THE INTERNATIONAL DIFFUSE INTRINSIC PONTINE GLIOMA (DIPG)/DIFFUSE MIDLINE GLIOMA (DMG) REGISTRY AND REPOSITORY (IDIPGR) EXPANSION

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Established in April 2012, the mission of the IDIPGR is to provide secure integrated data sets including clinical, pathologic, radiologic and molecular genomics to the research community to promote hypothesis driven research. Over 600 data points per patient are securely stored on a CCHMC constructed web resource and domain using the open-source data mart

development framework Harvest (PMID:24303304) ('Links'). Genomic data is stored in the cloud-enabled VIVA platform and accessed through cross-platform integration and standardization algorithms for comparison across datasets. Features include source identification, data wrangling, and standardization of molecular and phenotypic data (2017), a web-enabled data mart that provides phenotype-genotype query/exploration, along with raw and processed data file downloads to authorized investigators (Harvest, 2017), additional tools for filtering and analysis of genomic datasets at the level of a phenotype, sample, gene, and variant (VIVA, 2017–2018), and uploaded digitized slides (Aperio, 2019). The IDIPGR Repository stores abstracted datasets for >1020 patients with DIPG/DMG, of whom 366 have tumor tissue available through biopsy and/or autopsy, and centrally reviewed and digitized specimens from 124 patients. The Repository contains >5000 radiology studies from >700 patients, with >550 patients centrally reviewed, and genomics data from 80 patients. Currently 27 IDIPGR approved projects utilize these datasets. The DIPG/DMG Registry constructed a robust database platform and integration system that provides the infrastructure to promote highly collaborative, international, hypothesis-driven research. Broadening collaboration among investigators for hypothesis-driven research studies will lead to better classification and more effective treatment of patients with DIPG and DMG.

EPID-16. INTEGRATION OF EHR AND CANCER REGISTRY DATA TO CONSTRUCT A PEDIATRIC NEURO-ONCOLOGY SURVIVORSHIP COHORT AND IMPROVE LONG-TERM FOLLOW-UP CARE

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BACKGROUND: Pediatric neuro-oncology (PNO) survivors suffer long-term physical and neurocognitive morbidity. Comprehensive care addressing late effects of brain tumors and treatment in these patients is important. Clinical guidelines offer a framework for evaluating late effects, yet lack of extended follow-up is a significant barrier. The electronic health record (EHR) allows novel and impactful opportunities to construct, maintain, and leverage survivorship cohorts for health care delivery and as a platform for research. **METHODS:** This survivorship cohort includes all PNO cases ≤18-years-old reported to the state-mandated cancer registry by our institution. Data mining of the EHR for exposures, demographic, and clinical data identified patients with lack of extended follow-up (>1000 days since last visit). Explanatory variables included age, race/ethnicity, and language. Primary outcome included date of last clinic visit. **RESULTS:** Between January 1, 2013 and December 31, 2018, there were 324 PNO patients reported to our institutional registry with ongoing analysis to identify the specific survivorship cohort. Thirty patients died with an overall mortality of 9.3%. Two-hundred-and-sixteen patients were seen in PNO clinic, of which 18.5% (n=40) did not receive extended follow-up. Patients without extended follow-up were an average of 3.5 years older up (p<0.01); however, there was no significant difference in preferred language (p=0.97) or race/ethnicity (p=0.57). **CONCLUSION:** Integration of EHR and cancer registry data represents a feasible, timely, and novel approach to construct a PNO survivorship cohort to identify and re-engage patients without extended follow-up. Future applications include analysis of exposures and complications during therapy on late effects outcomes.

EPID-17. A SINGLE INSTITUTE EXPERIENCE IN THE REGISTRATION STUDY OF PEDIATRIC SOLID TUMOR IN JAPAN CHILDREN'S CANCER GROUP

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A larger scale prospective registration study has been conducted nationwide in Japan since 2011, called as "registration study in pediatric solid tumor", in Japan Children's Cancer Group (JCCG). In this study, clinical data and surgical specimen are collected into the National Center for Child Health. Kyoto University Hospital has participated in this study since IRB approval in 2011. We reviewed our registered patients to the registration study and assessed the current status. 40 patients with pediatric brain tumors participated in this study from 2011 to 2020. There were 13 intracranial germ cell tumors, 9 medulloblastomas, 6 gliomas in 4 diffuse midline gliomas, 4 pilocytic astrocytoma, and 4 other types of tumor. The informed consent was obtained from 36 patients by pediatricians and 3 patients by neurosurgeons. Twenty-five surgical specimens were collected for central review. The reasons of no surgical specimens were nonsurgical management in 6 patients and no enough FFPE sample in 3 patients. There was no discrepancy between central review and institutional diagnosis. The status of clinical data entry was complete in 13 patients and uncomplete in 9 patients.

These registration data including pathological diagnosis, molecular diagnosis, treatment, clinical information in patients with pediatric brain tumor are very important to realize current status. To conduct this study certainly, the collaboration among pediatrician, neurosurgeon, and supporting staff should be needed in collecting specimens and clinical data.

EPID-18. TRENDS IN INCIDENCE AND SURVIVAL OF MALIGNANT PEDIATRIC CENTRAL NERVOUS SYSTEM TUMORS IN THE NETHERLANDS

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BACKGROUND: Variation in survival of pediatric central nervous system (CNS) tumors is large between countries. Within Europe, the Netherlands had one of the worst reported survival rates of malignant CNS (mCNS) tumors during 2000–2007. **METHODS:** Using the Netherlands Cancer Registry, we evaluated trends in incidence and survival of pediatric mCNS tumors (behavior /3, 5th digit in the morphology code) diagnosed between 1990–2017. **RESULTS:** 839 newly-diagnosed mCNS tumor patients <18 years were registered between 1990–2017. Incidence of mCNS tumors remained stable (average incidence rate, 21.6 per million person-years). However, an increased incidence of malignant gliomas, NOS was found (Estimated Annual Percentage Change (EAPC) 11.6% p<0.001). This appears to be related to a registration shift between 1990–1999 and 2000–2009 as brainstem tumors increased (+25%, n=79) for astrocytomas and other gliomas but decreased (-31%, n=32) for unspecified intracranial and intraspinal neoplasms. Overall, 5-year observed survival (5Y-OS) of mCNS tumors increased from 51% in 1990–1999 to 61% in 2010–2017 (P-for-trend<0.001). This increase was not constant over time, as 5Y-OS for the period 2000–2009 was 47%. The only significant decrease in survival was found for malignant astrocytomas and other gliomas with a 5Y-OS of 56% in 1990–1999 decreasing to 48% in 2010–2017 (P-for-trend<0.001). **CONCLUSION:** Between 1990–2017 incidence of mCNS tumors in the Netherlands remained stable and survival increased. However, a decrease in survival was seen for malignant astrocytomas and other gliomas, which is partially explained by the registration shift of brainstem tumors. The impact of this shift on survival for all mCNS tumors is subject to further research.

ETMR AND OTHER EMBRYONAL TUMORS

ETMR-01. TREATMENT OUTCOME OF TWO CASES WITH HIGH-GRADE NEUROEPITHELIAL TUMOR WITH BCOR ALTERATION

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INTRODUCTION: High grade neuroepithelial tumor with BCOR exon 15 internal tandem duplication (HGNET BCOR) is a recently described tumor entity of the central nervous system (CNS) with a distinct methylation profile and characteristic genetic alteration. We report the outcome of two cases after 1st line multimodality therapy. **MATERIAL AND METHOD:** A 7 year old girl with a ventricular tumour and a 6 year old boy with a tumour in the occipital region with infiltration of the transverse and sigmoid sinus were both diagnosed based on histology and methylation with HGNET-BCOR. No spinal or liquor dissemination were found at diagnosis in both cases. Treatment consisted of radical resection of the tumour. In the case of the lesion with sinus infiltration residual tumour in the vessel could not be removed. Both children were postoperatively treated with radiotherapy (craniospinal 36 Gy and boost to 54 Gy), concomitant Vincristin and adjuvant Cisplatin, Lomustine and Vincristine. **RESULTS:** The girl developed a local recurrence at the primary tumour site 18 months after diagnosis. Reoperation showed the same histology. Start of 2nd line chemotherapy with Temozolomid and Irinotecan is being discussed. The boy with sinus infiltration developed seven months after diagnosis multiple liver, lung and bone metastasis. Biopsy of a liver lesion showed HGNET-BCOR. He was treated with Temozolomid, Irinotecan and died nine months after diagnosis. **CONCLUSION:** We report two cases with failure after 1st line treatment for HGNET-BCOR. To our knowledge HGNET-BCOR with development of hematological disease dissemination is a rare finding.