

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.