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# Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011

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#### Introduction

Brain tumors are a significant source of cancer-related morbidity and mortality in infants and children. This age group is diagnosed with unique groups of cancers and requires separate reporting in order to accurately portray the state of brain tumors in these populations.

The Central Brain Tumor Registry of the United States (CBTRUS) is the largest population-based registry of primary brain and central nervous system (CNS) tumors in the United States (US), and covers 99.8% of the US population for the period between 2007 and 2011 (for 2011 only, data was available for 50 out of 51 registries). The objective of the CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011 is to provide a comprehensive summary of the current descriptive epidemiology of primary brain and CNS tumors of childhood (0-14 years) in the US population. CBTRUS obtained the latest available data on all newly diagnosed primary brain and CNS tumors from the Centers for Disease Control and Prevention (CDC) National Program of Cancer Registries (NPCR), and the National Cancer Institute (NCI) Surveillance, Epidemiology and End Results (SEER) program for diagnosis years 2007-2011. Incidence counts and rates of primary malignant and nonmalignant brain and CNS tumors are documented by histology, gender, age, race, and Hispanic ethnicity. Mortality and relative survival rates for selected malignant histologies calculated using SEER data for the period 1995-2011 are also presented.

#### **Background**

CBTRUS is currently the only population-based site-specific registry in the US that works in partnership with a public cancer surveillance organization, the CDC's NPCR, and from which data

are directly received under a special agreement. This agreement permits transfer of data through the National Program of Central Registries Cancer Surveillance System (NPCR-CSS) Submission Specifications mechanism. CBTRUS researchers combine the NPCR data with data from the SEER program<sup>1</sup> of the NCI, which was established for national cancer surveillance in the early 1970s. All data from NPCR and SEER originate from tumor registrars who adhere to the Uniform Data Standards (UDS) for malignant and non-malignant brain and CNS tumors as directed by the North American Association of Cancer Registries (NAACCR) (http://www.naaccr.org). Along with the UDS, there are quality control checks and a system for rating each central registry to further insure that these data are reported as accurately and completely as possible. As a surveillance partner, CBTRUS can, therefore, report high quality data on brain and CNS tumors with histological specificity useful to the communities it serves. Its database represents the largest aggregation of populationbased data on the incidence of primary brain and CNS tumors in the US.

#### **Technical Notes**

#### Data Collection

CBTRUS contains incidence data from 51 independent central cancer registries (46 NPCR and 5 SEER registries) representing ~99.8% of the US population for the time period examined in this report (for 1 of 51 registries, data were available only from 2007–2010). Please see *The CBTRUS Statistical Report: Primary and Central Nervous System Tumors Diagnosed in the United States in 2007–2011* for additional information about the way that these data are obtained and processed. <sup>2</sup>

Age-adjusted incidence rates per 100,000 for the entire US for selected other cancers were obtained from the United States Cancer Statistics (USCS),<sup>3</sup> produced by the CDC and the NCI, via

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CDC Wide-ranging Online Data for Epidemiologic Research (WON-DER), for the purpose of comparison with brain and CNS tumor incidence rates. This database includes both NPCR and SEER data and represents nearly 100% of the US population.

Survival data for malignant brain and CNS tumors were obtained from 18 SEER registries for the years 1995 to 2011. This dataset spanning 16 years provides population-based information for approximately 26% of the US population,<sup>4</sup> and is a subset of the data used for the incidence calculations presented in this report. Survival information derived from active patient follow-up is not available in the data that CBTRUS receives from NPCR registries, so the SEER data are used for the generation of these Tables.

Mortality data used in this report are from the National Center for Health Statistics (NCHS) and include deaths where primary brain or CNS tumor was listed as cause of death on the death certificate for all 50 states and the District of Columbia. Population data for each geographic region were obtained from the SEER program website<sup>5</sup> for the purpose of rate calculation.

#### Data Reporting - Definitions

It should be noted that other surveillance organizations and researchers may report brain tumors differently from CBTRUS. The definition of brain and CNS tumors used by SEER, NPCR, and NAACCR in their published incidence and mortality statistics includes tumors located in the following sites with their ICD-O-3 site codes in parentheses: brain, meninges, and other central nervous system tumors (C70.0-9, C71.0-9, and C72.0-9), but excludes lymphoma and leukemia histologies (9590–9989) from all brain and CNS sites. CBTRUS reports data on all tumor morphologies located within the Consensus Conference site definition including lymphoma and other hematopoietic histologies (9590-9989), and olfactory tumors of the nasal cavity [C30.0 (9522-9523)].<sup>2,6</sup> Additionally, CBTRUS reports data on all brain and CNS tumors irrespective of behavior, whereas many reporting organizations may only publish rates for malignant brain and CNS tumors.

The CBTRUS Histology Grouping Scheme used in the CBTRUS Statistical Report: Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011<sup>2</sup> provides the basis for the definition for Gliomas and Embryonal Tumors used throughout this Report. These histologies were re-organized to be more reflective of the clinical organization of brain tumors that are specific to infancy and childhood. The gliomas are further categorized as low grade and high grade gliomas to further enhance their clinical relevance. Specific histologies and corresponding ICD-O-3 codes according to these refined categories can be found in Tables 2a and 2b.

Many other organizations and researchers that report childhood brain tumor statistics do so using the International Classification for Childhood Cancer (ICCC) grouping system<sup>7</sup> for pediatric cancers (Please see the CBTRUS website for additional information on this classification scheme: http://www.cbtrus.org). Frequencies and incidence of childhood brain tumors in the United States using the ICCC are presented in the CBTRUS Statistical Report: Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011.<sup>2</sup>

#### Methods

Counts, means, rates, ratios, proportions, and other relevant statistics were calculated using R 3.1.1 statistical software<sup>8</sup> and/or SEER\*Stat 8.1.5.<sup>9</sup> Statistics are suppressed when counts are fewer than 16 within a cell. However, the data in the suppressed cells are included in the counts and rates for the totals. Note that reported percentages may not add up to 100% due to rounding.

Age-adjusted incidence rates and 95% confidence intervals<sup>10</sup> for malignant and non-malignant tumors and for selected histology groupings by gender, race, Hispanic ethnicity, infant and pediatric age groups were estimated. Age-adjustment was based on one-year age groupings and standardized to the 2000 US standard population. Combined populations for the regions included in this report are shown in Appendix A and Appendix B.

CBTRUS presents statistics on specific brain and CNS tumor patterns in age groups <1, 1-4, 5-9, and 10-14 years. Race categories in this report are all races, white, black, American Indian/ Alaskan Native (AIAN), and Asian/Pacific Islander (API). Other race, unspecified, and unknown race are included in statistics that are not race-specific. Hispanic ethnicity was defined using the NAACCR Hispanic Identification Algorithm, version 2, data element, which utilizes a combination of cancer registry data fields (Spanish/ Hispanic Origin data element, birthplace, race, and surnames) to

**Table 1.** Central Brain Tumor Registry of the United States (CBTRUS), Brain and Central Nervous System Tumor Site Groupings, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011.

Site	ICD-O-3ª Site Code
Frontal lobe of brain	C71.1
Temporal lobe of brain	C71.2
Parietal lobe of brain	C71.3
Occipital lobe of brain	C71.4
Cerebrum	C71.0
Ventricle	C71.5
Cerebellum	C71.6
Brain stem	C71.7
Other brain <sup>b</sup>	C71.8-C71.9
Spinal cord and cauda equina	C72.0-C72.1
Cranial nerves	C72.2-C72.5
Other nervous system <sup>c</sup>	C72.8-C72.9
Meninges (cerebral & spinal)	C70.0-C70.9
Pituitary and craniopharyngeal duct	C75.1-C75.2
Pineal	C75.3
Olfactory tumors of the nasal cavity <sup>b</sup>	C30.0

<sup>&</sup>lt;sup>a</sup>International Classification of Diseases for Oncology, 3rd Edition, 2000. World Health Organization, Geneva, Switzerland.

<sup>&</sup>lt;sup>b</sup>Includes C71.8, Overlapping lesion of brain (Corpus callosum & Tapetum)) and C71.9 Brain, NOS (Intracranial site, Cranial fossa, NOS, Anterior cranial fossa, Middle cranial fossa, Posterior cranial fossa and Suprasellar).

<sup>&</sup>lt;sup>c</sup>Includes ICD-O-3 site code C72.8 Overlapping lesion of brain and CNS when point of origin cannot be assigned and C72.9 Nervous system, NOS (CNS, Epidural, Extradural, Parasellar).

dICD-O-3 histology codes 9522-9523 only.

**Table 2a.** CBTRUS Brain and Central Nervous System Tumor Histology Groupings, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011.

CBTRUS Specific Histology Grouping <sup>a</sup>	Infant and Childhood Report Major Histology Groupings	ICD-O-3 <sup>b</sup> Histology Code
Pilocytic astrocytoma	Pilocytic astrocytoma*	9421
Diffuse astrocytoma	Other low grade glioma*	9400 (excluding site C71.7), 9410, 9411, 9420
	High grade glioma*	9400 (site C71.7 only)
Anaplastic astrocytoma	High grade glioma*	9401
Unique astrocytoma variants	Other low grade glioma*	9383, 9384, 9424
Glioblastoma	High grade glioma*	9440, 9441, 9442/3 <sup>c</sup>
Oligodendroglioma	Other low grade glioma*	9450
Anaplastic oligodendroglioma	High grade glioma*	9451, 9460
Oligoastrocytic tumors	Other low grade glioma*	9382
Ependymal tumors	Ependymal tumors*	9391, 9392, 9393, 9394
Glioma malignant, NOS	Other low grade glioma*	9380 (site C72.3 only)
	High grade glioma*	9380 (site C71.7 only)
	Other glioma*	9380 (excluding sites C71.7 and C72.3)
Choroid plexus tumors	Choroid plexus tumors	9390
Other neuroepithelial tumors	Other glioma*	9363, 9423, 9430, 9444
Neuronal and mixed neuronal-glial tumors	Other low grade glioma*	9412, 9413
	Other glioma*	9442/1 <sup>d</sup>
	Neuronal and mixed neuronal-glial tumors	8680, 8681, 8690, 8693, 9492 (excluding site C75.1), 9493, 9505, 9506, 9522, 9523
Tumors of the pineal region	Tumors of the pineal region	9360, 9361, 9362
Embryonal tumors	Medulloblastoma	9470, 9471, 9472, 9474
	Primitive neuroectodermal tumor	9473
	Atypical teratoid/rhabdoid tumor	9508
	Other embryonal tumors	8963, 9364, 9490, 9500, 9501, 9502, 9504
Nerve sheath tumors	Tumors of cranial and spinal nerves	9540, 9541, 9550, 9560, 9561, 9570, 9571
Other Tumors of cranial and spinal nerves	Tumors of cranial and spinal nerves	9562
Meningioma	Tumors of meninges	9530, 9531, 9532, 9533, 9534, 9537, 9538, 9539
Mesenchymal tumors	Tumors of meninges	8324, 8800, 8801, 8802, 8803, 8804, 8805, 8806, 8810, 8815, 8824, 8830, 8831, 8835, 8836 8850, 8851, 8852, 8853, 8854, 8857, 8861, 8870, 8880, 8890, 8897, 8900, 8901, 8902, 8910, 8912, 8920, 8921, 8935, 8990, 9040, 9136, 9150, 9170, 9180, 9210, 9241, 9260, 9373, 9480
Primary melanocytic lesions	Tumors of meninges	8720, 8728, 8770, 8771
Other neoplasms related to the meninges	Tumors of meninges	9161, 9220, 9231, 9240, 9243, 9370, 9371, 9372, 9535
Lymphoma	Lymphomas and hematopoietic neoplasms	9590, 9591, 9596, 9650, 9651, 9652, 9653, 9654, 9655, 9659, 9661, 9662, 9663, 9664, 9665 9667, 9670, 9671, 9673, 9675, 9680, 9684, 9687, 9690, 9691, 9695, 9698, 9699, 9701, 9702, 9705, 9714, 9719, 9728, 9729
Other hematopoietic neoplasms	Lymphomas and hematopoietic neoplasms	9727, 9731, 9733, 9734, 9740, 9741, 9750, 9751, 9752, 9753, 9754, 9755, 9756, 9757, 9758, 9760, 9766, 9823, 9826, 9827, 9832, 9837, 9860, 9861, 9866, 9930, 9970
Germ cell tumors, cysts and heterotopias	Germ cell tumors	8020, 8440, 9060, 9061, 9064, 9065, 9070, 9071, 9072, 9080, 9081, 9082, 9083, 9084, 9085, 9100, 9101
Tumors of the pituitary	Tumors of the pituitary	8040, 8140, 8146, 8246, 8260, 8270, 8271, 8272, 8280, 8281, 8290, 8300, 8310, 8323, 9492 (Site C75.1 only), 9582
Craniopharyngioma	Craniopharyngioma	9350, 9351, 9352
Hemangioma	Other/unclassified tumors	9120, 9121, 9122, 9123, 9125, 9130, 9131, 9133, 9140
Neoplasm, unspecified	Other/unclassified tumors	8000, 8001, 8002, 8003, 8004, 8005, 8010, 8021
All other	Other/unclassified tumors	8320, 8452, 8710, 8711, 8713, 8811, 8840, 8896, 8980, 9173, 9503, 9580

<sup>&</sup>lt;sup>a</sup>See the CBTRUS 2014 Statistical Report and the CBTRUS website for additional information about the specific histology codes included in each group: http://www.cbtrus.org.

<sup>&</sup>lt;sup>b</sup>International Classification of Diseases for Oncology, 3rd Edition, 2000. World Health Organization, Geneva, Switzerland.

<sup>&</sup>lt;sup>c</sup>Morphology 9442/3 only.

dMorphology 9442/1 only.

<sup>\*</sup>All or some of this histology is included in the CBTRUS definition of gliomas, including ICD-O-3 histology codes 9380-9384, 9391-9460, 9480. See Appendix C for more information on glioma histologies.

Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; NOS, not otherwise specified.

**Table 2b.** ICD-O-3 Morphology Codes for all Histologies Included in Glioma and Embryonal Tumors Infant and Childhood Report Major Histology Groupings, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011.

Infant and Childhood Report Major Histology Groupings	ICD-O-3 <sup>a</sup> Morphology Code	Histology Name	Sub-histologies
Pilocytic astrocytoma	9421/1	Pilocytic astrocytoma	Piloid astrocytoma; Juvenile astrocytoma; Spongioblastoma, NOS
Other low grade glioma	9380/3	Glioma, malignant	Glioma, NOS
	9382/3	Mixed glioma	Oligoastrocytomal; Anaplastic oligoastrocytoma
	9383/1	Subependymoma	Subependymal glioma; Subependymal astrocytoma, NOS; Mixed subependymoma-ependymoma
	9384/1	Subependymal giant cell astrocytoma	
	9400/3	Astrocytoma, NOS	Astrocytic glioma; Astroglioma; Diffuse astrocytoma; Astrocytoma; low grade; Diffuse astrocytoma, low grade; Cystic astrocytoma
	9410/3	Protoplasmic astrocytoma	
	9411/3	Gemistocytic astrocytoma	Gemistocytoma
	9412/1	Desmoplastic infantile astrocytoma	Desmoplastic infantile ganglioglioma
	9413/0	Dysembryoplastic neuroepithelial tumor	
	9420/3	Fibrillary astrocytoma	Fibrous astrocytoma
	9424/3	Pleomorphic xanthoastrocytoma	<b></b>
	9450/3	Oligodendroglioma, NOS	
High grade glioma	9400/3	Astrocytoma, NOS	Astrocytic glioma; Astroglioma; Diffuse astrocytoma; Astrocytoma; low grade; Diffuse astrocytoma, low grade;Cystic astrocytoma
	9401/3	Astrocytoma, anaplastic	. 5 , 11
	9440/3	Glioblastoma, NOS	Glioblastoma multiforme; Spongioblastoma multiforme
	9441/3	Giant cell glioblastoma	Monstrocellular sarcoma
	9442/3	Gliosarcoma	Glioblastoma with sarcomatous component
	9451/3	Oligodendroglioma, anaplastic	
	9460/3	Oligodendroblastoma	
	9380/3	Glioma, malignant	Glioma, NOS
Ependymal tumors	9391/3	Ependymoma, NOS	Epithelial ependymoma; Cellular ependymoma; Clear cell ependymoma; Tanycytic ependymoma
	9392/3	Ependymoma, anaplastic	Ependymoblastoma
	9393/3	Papillary ependymoma	
	9394/1	Myxopapillary ependymoma	
Other glioma	9380/3	Glioma, malignant	Glioma, NOS
	9363/0	Melanotic neuroectodermal tumor	Retinal anlage tumor; Melanoameloblastoma; Melanotic progonoma
	9423/3	Polar spongioblastoma	Spongioblastoma polare; Primitive polar spongioblastoma
	9430/3	Astroblastoma	
	9444/1	Chordoid glioma	Chordoid glioma of third ventricle
	9442/1	Gliofibroma	
Medulloblastoma	9470/3	Medulloblastoma, NOS	Melanotic medulloblastoma
	9471/3	Desmoplastic nodular medulloblastoma	Desmoplastic medulloblastoma; Circumscribed arachnoidal cerebellar sarcoma
	9472/3	Medullomyoblastoma	
	9474/3	Large cell medulloblastoma	
Primitive neuroectodermal tumor (PNET)	9473/3	Primitive neuroectodermal tumor, NOS	PNET, NOS; Central primitive neuroectodermal tumor, NOS; CPNET; Supratentorial PNET

Continued

**Table 2b.** Continued

Infant and Childhood Report Major Histology Groupings	ICD-O-3 <sup>a</sup> Morphology Code	Histology Name	Sub-histologies
Atypical teratoid/ rhabdoid tumor (ATRT)	9508/3	Atypical teratoid/rhabdoid tumor	
Other embryonal tumors	8963/3	Malignant rhabdoid tumor	Rhabdoid sarcoma; Rhabdoid tumor, NOS
	9364/3	Peripheral neuroectodermal tumor	Neuroectodermal tumor, NOS; Peripheral primitive neuroectodermal tumor, NOS; PPNET
	9490/0	Ganglioneuroma	Ganglioneuroblastoma
	9500/3	Neuroblastoma, NOS	Sympathicoblastoma; Central neuroblastoma
	9501/0	Medulloepithelioma, benign	Diktyoma, benign
	9501/3	Medulloepithelioma, NOS	Diktyoma, malignant
	9502/0	Teratoid medulloepithelioma, benign	
	9502/3	Teratoid medulloepithelioma	
	9504/3	Spongioneuroblastoma	

<sup>&</sup>lt;sup>a</sup>International Classification of Diseases for Oncology, 3rd Edition, 2000. World Health Organization, Geneva, Switzerland.

directly and indirectly classify cases as Hispanic or non-Hispanic.<sup>11</sup> The NAACCR regional scheme (http://faststats.naaccr.org/usregions. php) was used for statistics reported by region of the US.

Estimated numbers of expected malignant and non-malignant brain and CNS tumors were calculated for 2015 and 2016. To project estimates of all primary brain and CNS tumors, age-adjusted brain tumor incidence rates for 2007–2011 were multiplied by the projected population. Projected population estimates for 2015 and 2016 were obtained from the interim projections from 2000–2030 based on the 2000 Census.<sup>5</sup>

Age-adjusted mortality rates for deaths resulting from all malignant brain and CNS tumors were calculated using the mortality data available in the CDC WONDER Online Database provided by NCHS.<sup>12</sup> The SEER cause of death recode<sup>13</sup> was used to categorize all mortality data used in this report. In addition to total age-adjusted rate for the US, age-adjusted rates are presented by gender and state.

SEER\*Stat 8.1.5 statistical software was used to estimate one-, two-, three-, four-, five-, and ten-year relative survival rates for primary malignant brain tumor cases diagnosed between 1995–2011 in eighteen SEER areas. <sup>9,14</sup> This software utilizes life-table (actuarial) methods to compute survival estimates and accounts for current follow-up.

Survival analysis was conducted using multiple-year cohorts, which include all persons diagnosed during the time period specified for the survival calculation. Second or later primary tumors, cases diagnosed at autopsy, cases in which race or sex is coded as other or unknown, and cases known to be alive but for whom follow-up time could not be calculated, were excluded from the SEER survival data analyses (~1% of total cases of malignant primary brain tumor in children under 15 in the SEER database from 1973 – 2011). Survival was not calculated for non-malignant tumors as collection of these cases has only been mandated since 2004, and therefore, not enough time has elapsed to accurately calculate relative survival. Please note that survival statistics are reported for pilocytic astrocytoma,

which has traditionally been included as a malignant tumor for cancer registration purposes although this tumor is clinically considered to be non-malignant. This decision has been influenced by the importance of location in the CNS to the morbidity and mortality caused by brain and CNS tumors.

Total deaths by specific histology group were calculated using data on primary malignant brain tumor cases diagnosed between 1995–2011 in eighteen SEER areas. <sup>9,14</sup> Using only persons that died due to disease, we used month of diagnosis, year of diagnosis, survival months, and age of diagnosis to calculate approximate month and year of death and approximate age at death.

Five-year conditional survival estimates were calculated for brain tumor cases diagnosed between 1995–2011 in eighteen SEER areas using SEER\*Stat 8.1.5 statistical software. <sup>9,14</sup> Conditional survival is an estimate of the probability that a patient will survive for a specific time period given that they have already survived a certain number of years. For example, 5-year conditional survival for a child who has lived two years since their diagnosis with pilocytic astrocytoma is 98.5%, which means that 98.5% of children 0–14 years who have already survived two years will eventually survive five years.

#### Results

Cancer is a significant source of morbidity and mortality for infants and children ages 0-14 years in the US. The overall average annual age-adjusted incidence rate for children 0-14 years between 2007 and 2011 was 5.26 per 100,000 population (16,044 total tumors). Approximately 1 in 2,000 children born from 2009–2011 will be diagnosed with a primary malignant brain or CNS tumor by the time they are 14 years. <sup>16</sup> These tumors continue to be the most common solid tumor in infants and children 0-14 years.

In children ages 1-4 and 5-14 years cancer is the  $4^{th}$  and  $2^{nd}$  most common causes of death, respectively (Figure 1a). Brain and CNS tumors are the most common cause of cancer death in children 0-14 years in the United States (Figure 1b).

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**Table 3.** Average Annual Age-Adjusted Incidence Rates<sup>a</sup> for Childhood Brain and Central Nervous System Tumors by Site, and Age at Diagnosis, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011.

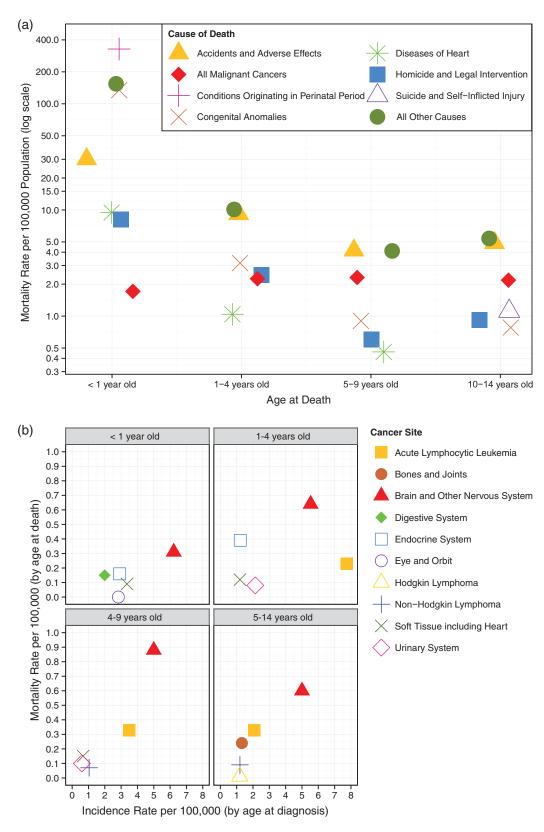
Histology				Age At	Diagn	osis (years)									
	0-14			<1			1-4			5-9			10-14	+	
	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI
Frontal, temporal, parietal, & occipital lobe	2,522	0.83	(0.80-0.86)	163	0.81	(0.69-0.94)	556	0.69	(0.63-0.75)	729	0.73	(0.68-0.78)	1,074	1.04	(0.98-1.10)
Frontal lobe of brain	876	0.29	(0.27 - 0.31)	51	0.25	(0.19 - 0.33)	218	0.27	(0.24-0.31)	238	0.24	(0.21-0.27)	369	0.36	(0.32 - 0.39)
Temporal lobe of brain	1,037	0.34	(0.32 - 0.36)	72	0.36	(0.28 - 0.45)	213	0.26	(0.23 - 0.30)	311	0.31	(0.28 - 0.35)	441	0.43	(0.39 - 0.47)
Parietal lobe of brain	457	0.15	(0.14-0.16)	30	0.15	(0.10-0.21)	101	0.13	(0.10-0.15)	133	0.13	(0.11-0.16)	193	0.19	(0.16-0.21)
Occipital lobe of brain	152	0.05	(0.04-0.06)	_	-	-	24	0.03	(0.02 - 0.04)	47	0.05	(0.03 - 0.06)	71	0.07	(0.05-0.09)
Cerebrum	979	0.32	(0.30 - 0.34)	64	0.32	(0.24 - 0.41)	261	0.32	(0.29 - 0.37)	332	0.33	(0.30-0.37)	322	0.31	(0.28 - 0.35)
Ventricle	1,019	0.33	(0.31-0.35)	202	1.00	(0.87 – 1.15)	312	0.39	(0.34 - 0.43)	233	0.23	(0.20-0.26)	272	0.26	(0.23 - 0.30)
Cerebellum	3,001	0.98	(0.95-1.02)	131	0.65	(0.54 - 0.77)	1,017	1.26	(1.19 - 1.34)	1,072	1.06	(1.00-1.13)	781	0.76	(0.71 - 0.81)
Brain stem	1,997	0.66	(0.63 - 0.69)	89	0.44	(0.35 - 0.54)	663	0.82	(0.76 - 0.89)	784	0.78	(0.72 - 0.83)	461	0.45	(0.41 - 0.49)
Other brain <sup>b</sup>	2,439	0.80	(0.77 - 0.83)	310	1.54	(1.37 – 1.72)	740	0.92	(0.85 - 0.99)	711	0.71	(0.66-0.76)	678	0.66	(0.61-0.71)
Spinal cord and cauda equina	683	0.22	(0.21-0.24)	74	0.37	(0.29 - 0.46)	175	0.22	(0.19 - 0.25)	171	0.17	(0.15-0.20)	263	0.25	(0.22-0.29)
Cranial nerves	1,104	0.36	(0.34-0.38)	80	0.40	(0.31 - 0.49)	480	0.59	(0.54 - 0.65)	324	0.32	(0.29 - 0.36)	220	0.21	(0.19 - 0.24)
Other nervous system <sup>c</sup>	307	0.10	(0.09 - 0.11)	65	0.32	(0.25 - 0.41)	108	0.13	(0.11-0.16)	81	0.08	(0.06-0.10)	53	0.05	(0.04 - 0.07)
Meninges (cerebral & spinal)	316	0.10	(0.09 - 0.12)	36	0.18	(0.13 - 0.25)	57	0.07	(0.05 - 0.09)	69	0.07	(0.05 - 0.09)	154	0.15	(0.13 - 0.17)
Pituitary and craniopharyngeal duct	1,252	0.41	(0.39 - 0.44)	17	0.08	(0.05-0.13)	141	0.18	(0.15-0.21)	401	0.40	(0.36 - 0.44)	693	0.67	(0.62 - 0.72)
Pineal & olfactory	425	0.14	(0.13 - 0.15)	24	0.12	(0.08 - 0.18)	82	0.10	(0.08 - 0.13)	114	0.11	(0.09 - 0.14)	205	0.20	(0.17 - 0.23)
Pineal	413	0.14	(0.12-0.15)	23	0.11	(0.07 - 0.17)	78	0.10	(0.08-0.12)	114	0.11	(0.09 - 0.14)	198	0.19	(0.16-0.22)
Olfactory tumors of the nasal cavity <sup>b</sup>	-	-	_	-	-	_	-	-	-	-	-	_	-	-	_
TOTAL <sup>c</sup>	16,044	5.26	(5.18-5.34)	1,255	6.22	(5.88-6.58)	4,592	5.53	(5.53-5.86)	5,021	5.00	(4.86-5.14)	5,176	5.00	(4.87-5.14)

 $<sup>^{\</sup>mathrm{a}}$ Rates are per 100,000 and are age-adjusted to the 2000 US standard population.

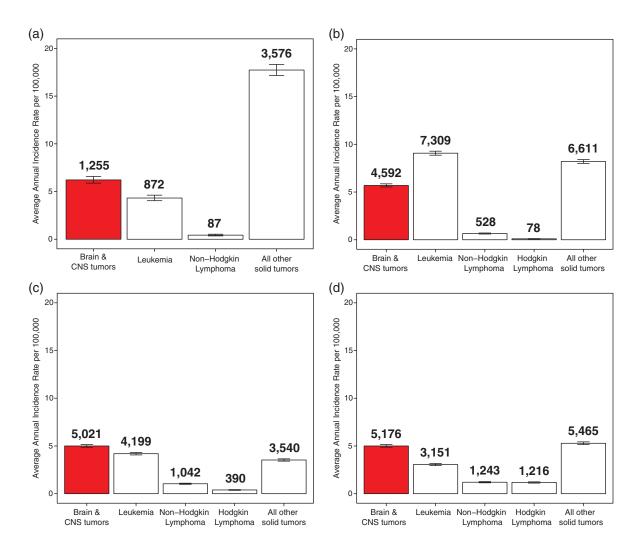
<sup>&</sup>lt;sup>b</sup>Refers to all brain tumors including histologies not presented in this table.

<sup>&</sup>lt;sup>c</sup>ICD-O-3 histology codes 9522 – 9523 only.

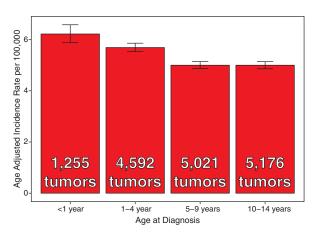
<sup>-</sup> Counts and rates are not presented when fewer than 16 cases were reported for the specific histology category. The suppressed cases are included in the counts and rates for totals. Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; CI, confidence interval.



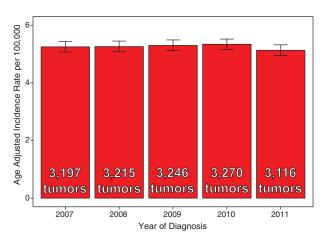
**Fig. 1.** (a) Average Annual Mortality Rates and Total Deaths for Top 5 Causes of Death and Death Due To Malignant Neoplasms for Children 0–14 by Age Groups (NVSS 2007–2011), (b) Average Annual Mortality Rates and Total Deaths for Top 5 Causes of Death Due to Cancer for Children 0–14 by Age Groups, 2007–2011 (NVSS 2007–2011, CBTRUS 2007–2011, USCS 2007–2011)



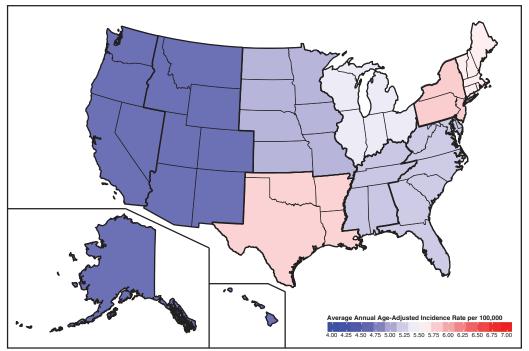
**Fig. 2.** Average Annual Age-Adjusted Incidence Rates of All Primary Brain And CNS Tumors in Comparison to Leukemias And Lymphomas in (a) Infants (<1 Year Old), (b) Children 1–4 Years, (c) Children 5–9 Years, and (d) Children 10–14 Years (CBTRUS 2007–2011, USCS 2007–2011)



**Fig. 3.** Average Annual Age-Adjusted Incidence Rates of Primary Brain and CNS Tumors by Age Group (N = 16,044) (CBTRUS 2007 – 2011)



**Fig. 4.** Annual Age-Adjusted Incidence Rates of Primary Brain and CNS Tumors by Year of Diagnosis (N = 16,044) (CBTRUS 2007–2011)



New England: Connecticut, Maine, Massachusetts, New Hampshire, Rhode Island, Vermont; Middle Atlantic: New Jersey, New York, Pennsylvania; South Atlantic: Delaware, District of Columbia, Florida, Georgia, Maryland, North Carolina, South Carolina, Viginia, West Virginia; East North Central: Illinois, Indiana, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, South Dakota, West South Central: Arkansas, Louisiana, Oklahoma, Texas; Mountain: Arizona, Colorado, Idaho, Montana, New Mexico, Utah, Wyoming; Pacific: Alaska, California, Hawaii, Nevada, Oregon, Washington.

Fig. 5. Average Annual Age-Adjusted Incidence Rates of All Primary Brain and CNS Tumors by Region of the United States (0–14 Years) (N = 16,044) (CBTRUS 2007–2011)

#### Comparison to Other Common Childhood Cancers

Average annual age-adjusted incidence rates for primary brain and CNS tumors, leukemias, and lymphoma in the United States are presented by age in Figures 2a (age < 1 year), 2b (ages 1–4 years), 2c (ages 5–9 years), and 2d (ages 10–14 years). Brain and CNS tumors were the most common cancer in children <1, and 5–14. For those aged 1–4 years, leukemias were the most commonly occurring cancer though brain and CNS tumors were still the most commonly occurring solid tumor across all age groups 0–14 years.

#### Overall Incidence by Age Group and Year of Diagnosis

Incidence of brain and CNS tumors was highest in infants (<1 year old), who had an overall incidence rate of 6.22 per 100,000 (1,255 tumors), followed by children ages 1-4 years who had an incidence rate of 5.53 per 100,000 (4,592 tumors). Children ages 5-14 years had an age-adjusted incidence of 5.00 per 100,000 (5-9: 5,021 tumors; 10-14: 5,176 tumors) (Figure 3). Incidence of brain and CNS tumors was stable over the time period examined (Figure 4).

#### Incidence by Region of the United States, And Age Group

Incidence of brain and CNS tumors varied by region of the United States (Figure 5). Overall age-adjusted incidence was highest in the Middle Atlantic (5.78 per 100,000, 95% CI: 5.53–6.02) and West South Central (5.75 per 100,000, 95% CI: 5.51–5.99) regions, and lowest in the Mountain (4.69 per 100,000, 95%

CI: 4.40–4.99) and Pacific (4.69 per 100,000, 95% CI: 4.51–4.88) regions.

Incidence by region and age groups is presented in Table 7.

- Incidence in infants (<1 year old) was highest in West South Central (7.00 per 100,000, 95% CI: 6.04–8.07), Middle Atlantic (6.85 per 100,000, 95% CI: 5.85–7.97), and East North Central (6.80 per 100,000, 95% CI: 5.88–7.81). It was lowest in West North Central (5.08 per 100,000, 95% CI: 6.04–8.07), and Pacific (5.31 per 100,000, 95% CI: 4.57–6.12).
- Incidence in children 1-4 years was highest in West South Central (6.10 per 100,000, 95% CI: 5.65-6.59) and Middle Atlantic (6.08 per 100,000, 95% CI: 5.60-6.60). It was lowest in East South Central (4.75 per 100,000, 95% CI: 4.16-5.41) and Pacific (4.99 per 100,000, 95% CI: 4.63-5.38).
- Incidence in children 5-9 years was highest in West South Central (5.60 per 100,000, 95% CI: 5.20-6.02), and lowest in Mountain (4.25 per 100,000, 95% CI: 3.78-4.77)
- Incidence in children 10-14 years was highest in Middle Atlantic (5.82 per 100,000, 95% CI: 5.41-6.25), New England (5.41 per 100,000, 95% CI: 4.76-6.12) and West South Central (5.39 per 100,000, 95% CI: 5.00-5.80). It was lowest in West North Central (4.43 per 100,000, 95% CI: 3.94-4.95) and Pacific (4.50 per 100,000, 95% CI: 4.19-4.82).

#### Distribution by Site and Age Group

The distribution of brain and CNS tumors by site is shown in Figure 6, and the distribution of tumors by site in each age

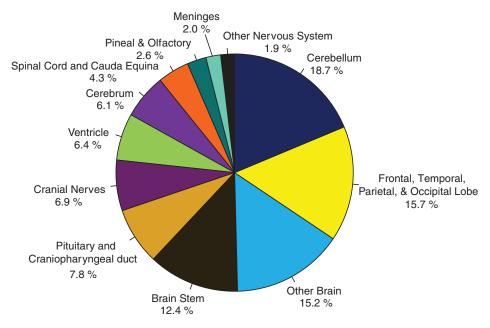


Fig. 6. Distribution of All Primary Brain and CNS Tumors by Site (0-14 Years) (N = 16,044) (CBTRUS 2007-2011)

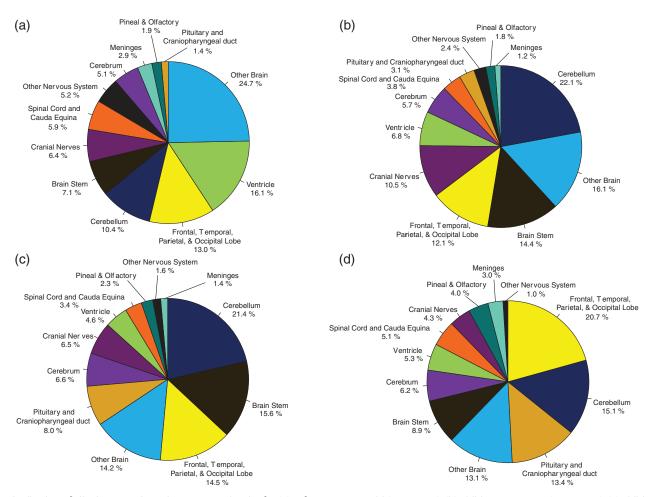


Fig. 7. Distribution of all Primary Brain and CNS Tumors by Site for (a) Infants <1 Year Old (N=1,255), (b) Children 1-4 Years (N=4,592), (c) Children 5-9 Years (N=5,021), and (d) Children 10-14 Years (N=5,176) (CBTRUS 2007-2011)

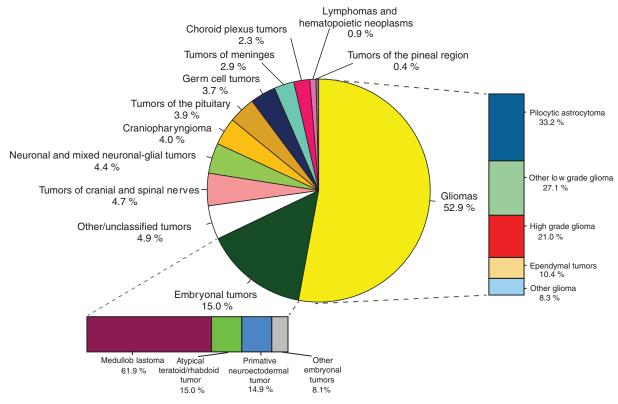


Fig. 8. Distribution of All Primary Brain and CNS Tumors by Histology Groupings (0-14 Years) (N = 16,044) (CBTRUS 2007-2011)

group is shown in Figures 7a-7d. Frequencies for each age group are presented in Table 3.

- The most common site was the cerebellum (18.7%), followed by the frontal, temporal, parietal, and occipital lobes (15.7%).
- The most common site in infants (<1 year old) was other brain (24.7%), followed by ventricle (16.1%). Other brain is a designation used in cancer registry data when the location of a tumor is not identified in a patient's record, or when a tumor involves multiple locations in the brain (Please see Table 1 for more information about the specific sites included in these groups).
- The most common site in children 1–4 years was the cerebellum (22.1%), followed by other brain (16.1%) and brain stem (14.4%).
- In children 5–9, cerebellum was also the most common site (21.4%), followed by brain stem (15.6%), and frontal, temporal, parietal, and occipital lobes (14.5%)
- In children 10-14, the most common site of disease was the frontal, temporal, parietal and occipital lobes (20.7%).

## Distribution and Incidence by Histologic Group and Age Group

The distribution of brain and CNS tumors by histologic group is shown in Figure 8, and the distribution of tumors by histologic group in each age group is shown in Figures 9a-9d. Frequencies for each age group are presented in Tables 4 and 5.

- The most common histologic group in all ages was glioma (52.9%), of which the majority were pilocytic astrocytoma (33.2%) and other low grade gliomas (27.1%).
- In infants (<1 year old), gliomas (37.2%) and embryonal tumors (24.9%) were the most commonly occurring tumor type. Of embryonal tumors, 42.9% were atypical teratoid/rhabdoid tumors.
- In children 1–4 years, gliomas (58.1%) and embryonal tumors (20.2%) were the most common tumor type.
- Gliomas (56.5%) and embryonal tumors (14.8%) were also the most common histologic groups in children 5–9 years. Medulloblastoma represented 80.4% of all embryonal tumors in this age group.
- In children 10–14 years, gliomas (48.6%), tumors of the pituitary (8.9%), and embryonal tumors (8.3%) were the most commonly occurring histologic types.

#### Incidence by Gender

Overall, approximately 52.8% of all tumors occurred in males (8,479 total tumors) and 47.2% occurred in females (7,565 total tumors). Counts and incidence rates by histologic groups and gender are presented in Table 4.

- Most histologies were more common in males, or equivocal between genders.
- Embryonal tumors, especially medulloblastoma, were more common in males. Age-adjusted incidence of embryonal tumors was 0.91 per 100,000 in males, as compared to 0.65 per 100,000 in females.

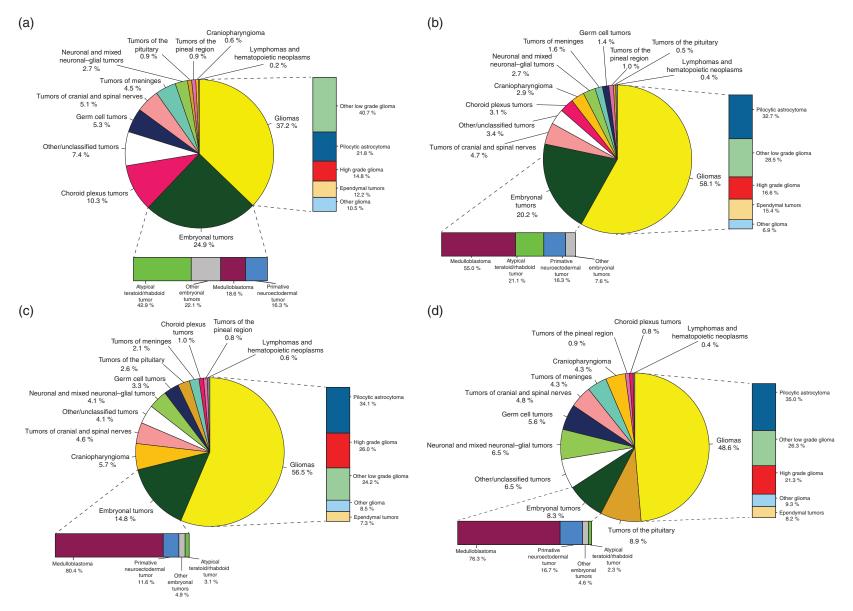


Fig. 9. Distribution of All Primary Brain and CNS Tumors by Histology Groupings for (a) Infants <1 Year Old (N = 1,255), (b) Children 1-4 Years (N = 4,592), (c) Children 5-9 Years (N = 5,021), and (d) Children 10-14 Years (N = 5,176) (CBTRUS 2007-2011)

	Total					Male			Female			IRR (Male:Female)	
Histology	N	% of All Tumors	Median Age	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	IRR	p-value
Gliomas	8,487	52.9%	6.0	2.78	(2.72 – 2.84)	4,386	2.81	(2.73 – 2.90)	4,101	2.75	(2.67-2.83)	0.98	0.29
Pilocytic astrocytoma	2,821	17.6%	7.0	0.93	(0.89 - 0.96)	1,452	0.93	(0.89 - 0.98)	1,369	0.92	(0.87 - 0.97)	0.98	0.67
Other low grade glioma	2,296	14.3%	6.0	0.75	(0.72 - 0.78)	1,188	0.76	(0.72 - 0.80)	1,108	0.74	(0.70 - 0.79)	0.98	0.58
High grade glioma	1,784	11.1%	7.0	0.59	(0.56 - 0.62)	898	0.58	(0.54 - 0.62)	886	0.60	(0.56 - 0.64)	1.03	0.55
Ependymal tumors	879	5.5%	4.0	0.29	(0.27 - 0.30)	510	0.32	(0.30 - 0.35)	369	0.24	(0.22 - 0.27)	0.76	< 0.01
Other glioma	707	4.4%	7.0	0.23	(0.22-0.25)	338	0.22	(0.20 - 0.24)	369	0.25	(0.22 - 0.27)	1.14	0.09
Choroid plexus tumors	362	2.3%	1.0	0.12	(0.10-0.13)	200	0.13	(0.11-0.14)	162	0.11	(0.09 - 0.12)	0.85	0.14
Tumors of the pineal region	701	4.4%	6.5	0.23	(0.21 - 0.25)	384	0.25	(0.22-0.27)	317	0.21	(0.19 - 0.24)	0.86	0.05
Neuronal and mixed neuronal-glial tumors	140	0.9%	9.0	0.05	(0.04 - 0.05)	71	0.05	(0.04 - 0.06)	69	0.05	(0.04 - 0.06)	1.01	1.00
Embryonal tumors	2,413	15.0%	4.0	0.79	(0.76 - 0.82)	1,429	0.91	(0.87 - 0.96)	984	0.65	(0.61-0.70)	0.72	< 0.01
Medulloblastoma	1,494	9.3%	6.0	0.49	(0.47 - 0.52)	929	0.60	(0.56 - 0.64)	565	0.38	(0.35 - 0.41)	0.63	< 0.01
Primitive neuroectodermal tumor	360	2.2%	3.5	0.12	(0.10-0.13)	197	0.13	(0.11-0.14)	163	0.11	(0.09 - 0.13)	0.86	0.18
Atypical teratoid/rhabdoid tumor	363	2.3%	1.0	0.12	(0.10-0.13)	197	0.12	(0.11 - 0.14)	166	0.11	(0.09 - 0.13)	0.88	0.24
Other embryonal tumors	196	1.2%	1.0	0.06	(0.05 - 0.07)	106	0.07	(0.06-0.08)	90	0.06	(0.05 - 0.07)	0.88	0.42
Tumors of cranial and spinal nerves	758	4.7%	7.0	0.25	(0.23 - 0.27)	403	0.26	(0.23 - 0.28)	355	0.24	(0.21-0.26)	0.93	0.30
Tumors of meninges	458	2.9%	9.0	0.15	(0.14-0.16)	226	0.14	(0.13-0.16)	232	0.16	(0.14-0.18)	1.07	0.49
Lymphomas and hematopoietic neoplasms	70	0.4%	6.0	0.02	(0.02 - 0.03)	46	0.03	(0.02 - 0.04)	24	0.02	(0.01-0.02)	0.55	0.02
Germ cell tumors	590	3.7%	9.0	0.19	(0.18 - 0.21)	358	0.23	(0.21-0.26)	232	0.16	(0.14-0.18)	0.68	< 0.01
Tumors of the pituitary	625	3.9%	12.0	0.20	(0.19 - 0.22)	227	0.15	(0.13 - 0.17)	398	0.27	(0.24-0.29)	1.83	< 0.01
Craniopharyngioma	648	4.0%	8.0	0.21	(0.20 - 0.23)	326	0.21	(0.19 - 0.24)	322	0.22	(0.19 - 0.24)	1.03	0.76
Other/unclassified tumors	792	4.9%	9.0	0.26	(0.24-0.28)	423	0.27	(0.25 - 0.30)	369	0.25	(0.22-0.27)	0.91	0.19
TOTAL <sup>b</sup>	16,044	100.0%	7.0	5.26	(5.18-5.34)	8,479	5.44	(5.32-5.56)	7,565	5.07	(4.95-5.18)	0.93	< 0.01

<sup>&</sup>lt;sup>a</sup>Rates are per 100,000 and are age-adjusted to the 2000 US standard population.

<sup>&</sup>lt;sup>b</sup>Refers to all brain tumors including histologies not presented in this table.

<sup>-</sup>Counts are not presented when fewer than 16 cases were reported for the specific histology category. Suppressed cases are included in the total count. Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; CI, confidence interval; IRR: incidence rate ratio.

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Table 5. Average Annual Age-Adjusted Incidence Rates for Childhood Brain and Central Nervous System Tumors by Major Histology Groupings, Histology, and Age at Diagnosis, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011

	Age At	Diagnosis	s (years)									
	<1			1-4			5-9			10-14		
Histology	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI
Gliomas	467	2.32	(2.11-2.54)	2,667	3.31	(3.18-3.44)	2,836	2.82	(2.72-2.92)	2,517	2.44	(2.34-2.53)
Pilocytic astrocytoma	102	0.51	(0.41 - 0.61)	871	1.08	(1.01 - 1.16)	968	0.96	(0.90 - 1.02)	880	0.85	(0.80 - 0.91)
Other low grade glioma	190	0.94	(0.81 - 1.09)	760	0.94	(0.88 - 1.01)	685	0.68	(0.63 - 0.74)	661	0.64	(0.59 - 0.69)
High grade glioma	69	0.34	(0.27 - 0.43)	443	0.55	(0.50 - 0.61)	736	0.73	(0.68 - 0.79)	536	0.52	(0.48 - 0.57)
Ependymal tumors	57	0.28	(0.21 - 0.37)	410	0.51	(0.46 - 0.56)	206	0.20	(0.18 - 0.23)	206	0.20	(0.17 - 0.23)
Other glioma	49	0.24	(0.18 - 0.32)	183	0.23	(0.20 - 0.26)	241	0.24	(0.21 - 0.27)	234	0.23	(0.20 - 0.26)
Choroid plexus tumors	129	0.64	(0.53 - 0.76)	142	0.18	(0.15 - 0.21)	50	0.05	(0.04 - 0.07)	41	0.04	(0.03 - 0.05)
Tumors of the pineal region	34	0.17	(0.12 - 0.24)	124	0.15	(0.13 - 0.18)	205	0.20	(0.18 - 0.23)	338	0.33	(0.29 - 0.36)
Neuronal and mixed neuronal-glial tumors	-	-	-	47	0.06	(0.04 - 0.08)	38	0.04	(0.03 - 0.05)	44	0.04	(0.03 - 0.06)
Embryonal tumors	312	1.55	(1.38 - 1.73)	929	1.15	(1.08 - 1.23)	741	0.74	(0.68 - 0.79)	431	0.42	(0.38 - 0.46)
Medulloblastoma	58	0.29	(0.22 - 0.37)	511	0.63	(0.58 - 0.69)	596	0.59	(0.55 - 0.64)	329	0.32	(0.29 - 0.36)
Primitive neuroectodermal tumor	51	0.25	(0.19 - 0.33)	151	0.19	(0.16 - 0.22)	86	0.09	(0.07 - 0.11)	72	0.07	(0.05 - 0.09)
Atypical teratoid/rhabdoid tumor	134	0.66	(0.56 - 0.79)	196	0.24	(0.21 - 0.28)	23	0.02	(0.01 - 0.03)	-	-	-
Other embryonal tumors	69	0.34	(0.27 - 0.43)	71	0.09	(0.07 - 0.11)	36	0.04	(0.03 - 0.05)	20	0.02	(0.01 - 0.03)
Tumors of cranial and spinal nerves	64	0.32	(0.24 - 0.41)	214	0.27	(0.23 - 0.30)	230	0.23	(0.20 - 0.26)	250	0.24	(0.21 - 0.27)
Tumors of meninges	57	0.28	(0.21-0.37)	73	0.09	(0.07 - 0.11)	105	0.10	(0.09 - 0.13)	223	0.21	(0.19 - 0.24)
Lymphomas and hematopoietic neoplasms	-	-	-	17	0.02	(0.01 - 0.03)	28	0.03	(0.02 - 0.04)	22	0.02	(0.01 - 0.03)
Germ cell tumors	67	0.33	(0.26 - 0.42)	66	0.08	(0.06-0.10)	165	0.17	(0.14-0.19)	292	0.28	(0.25 - 0.32)
Tumors of the pituitary	-	-	-	23	0.03	(0.02 - 0.04)	129	0.13	(0.11-0.15)	462	0.44	(0.40 - 0.48)
Craniopharyngioma	-	-	-	132	0.16	(0.14-0.19)	288	0.29	(0.26 - 0.32)	221	0.21	(0.19 - 0.24)
Other/unclassified tumors	93	0.46	(0.37 - 0.56)	158	0.20	(0.17 - 0.23)	206	0.21	(0.18 - 0.24)	335	0.32	(0.29 - 0.36)
TOTAL <sup>b</sup>	1,255	6.22	(5.88-6.58)	4,592	5.53	(5.53-5.86)	5,021	5.00	(4.86-5.14)	5,176	5.00	(4.87-5.14)

<sup>&</sup>lt;sup>a</sup>Rates are per 100,000 and are age-adjusted to the 2000 US standard population. <sup>b</sup>Refers to all brain tumors including histologies not presented in this table.

<sup>-</sup> Counts and rates are not presented when fewer than 16 cases were reported for the specific histology category. The suppressed cases are included in the counts and rates for totals. Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; CI, confidence interval.

**Table 6.** Average Annual Age-Adjusted Incidence Rates<sup>a</sup> for Brain and Central Nervous System Tumors by Major Histology Groupings, Histology, and Race<sup>b</sup>, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011

	White			Black			AIAN			API			
Histology	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	
Gliomas	6,786	2.92	(2.85 – 2.99)	1,104	2.21	(2.08-2.34)	68	1.25	(0.97 – 1.59)	470	2.78	(2.54 – 3.05)	
Pilocytic astrocytoma	2,290	0.99	(0.95 - 1.03)	343	0.68	(0.61 - 0.76)	20	0.36	(0.22 - 0.56)	149	0.88	(0.74 - 1.03)	
Other low grade glioma	1,866	0.80	(0.77 - 0.84)	269	0.54	(0.47 - 0.60)	19	0.35	(0.21 - 0.55)	126	0.74	(0.62 - 0.89)	
High grade glioma	1,370	0.59	(0.56 - 0.62)	286	0.58	(0.51 - 0.65)	-	-	_	99	0.59	(0.48 - 0.72)	
Ependymal tumors	699	0.30	(0.28 - 0.32)	118	0.23	(0.19 - 0.28)	-	-	-	45	0.26	(0.19 - 0.35)	
Other glioma	561	0.24	(0.22 - 0.26)	88	0.18	(0.14-0.22)	-	-	_	51	0.31	(0.23 - 0.41)	
Choroid plexus tumors	299	0.13	(0.11 - 0.14)	33	0.06	(0.04 - 0.09)	-	-	_	26	0.15	(0.10-0.22)	
Tumors of the pineal region	562	0.24	(0.22 - 0.26)	86	0.17	(0.14 - 0.21)	-	-	_	44	0.26	(0.19 - 0.35)	
Neuronal and mixed neuronal-glial tumors	87	0.04	(0.03 - 0.05)	43	0.09	(0.06 - 0.12)	-	-	-	-	-	-	
Embryonal tumors	1,964	0.84	(0.81 - 0.88)	282	0.56	(0.49 - 0.62)	16	0.29	(0.17 - 0.47)	123	0.71	(0.59 - 0.85)	
Medulloblastoma	1,237	0.53	(0.50 - 0.56)	151	0.30	(0.26 - 0.36)	-	-	-	76	0.44	(0.35 - 0.56)	
Primitive neuroectodermal tumor	288	0.12	(0.11 - 0.14)	52	0.10	(0.08 - 0.13)	-	-	-	-	-	-	
Atypical teratoid/rhabdoid tumor	289	0.12	(0.11 - 0.14)	43	0.08	(0.06 - 0.11)	-	-	-	26	0.14	(0.09 - 0.21)	
Other embryonal tumors	150	0.06	(0.05 - 0.08)	36	0.07	(0.05 - 0.10)	-	-	-	-	-	-	
Tumors of cranial and spinal nerves	565	0.24	(0.22 - 0.26)	112	0.22	(0.18 - 0.27)	-	-	_	66	0.39	(0.30 - 0.50)	
Tumors of meninges	366	0.16	(0.14 - 0.17)	56	0.11	(0.08 - 0.14)	-	-	_	30	0.18	(0.12 - 0.26)	
Lymphomas and hematopoietic neoplasms	51	0.02	(0.02 - 0.03)	-	-	_	-	-	_	-	-	_	
Germ cell tumors	440	0.19	(0.17 - 0.21)	59	0.12	(0.09 - 0.15)	-	-	_	85	0.52	(0.41 - 0.64)	
Tumors of the pituitary	481	0.21	(0.19 - 0.23)	86	0.17	(0.14 - 0.21)	-	-	_	48	0.30	(0.22 - 0.40)	
Craniopharyngioma	488	0.21	(0.19 - 0.23)	104	0.21	(0.17 - 0.25)	-	-	_	46	0.27	(0.20 - 0.37)	
Other/unclassified tumors	615	0.27	(0.24 - 0.29)	94	0.19	(0.15-0.23)	-	-	_	71	0.42	(0.33-0.53)	
TOTAL <sup>c</sup>	12,704	5.46	(5.37-5.56)	2,069	4.12	(3.94-4.30)	134	2.46	(2.06-2.92)	1,020	6.05	(5.69-6.44)	

 $<sup>^{\</sup>mathrm{a}}$ Rates are per 100,000 and are age-adjusted to the 2000 US standard population.

<sup>&</sup>lt;sup>b</sup>Individuals with unknown race were excluded

<sup>&</sup>lt;sup>c</sup>Refers to all brain tumors including histologies not presented in this table.

<sup>-</sup> Counts are not presented when fewer than 16 cases were reported for the specific histology category. Suppressed cases are included in the total count.

Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; CI, confidence interval; AIAN, American Indian/Alaskan Native; API, Asian/Pacific Islander.

**Table 7.** Average Annual Age-Adjusted Incidence Rates<sup>a</sup> for Brain and Central Nervous System Tumors by Major Histology Groupings, Histology, and Hispanic Ethnicity<sup>b</sup>, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011

	Hispanic			Non-Hispan	ic	
Histology	N	Rate	95% CI	N	Rate	95% CI
Gliomas	1,512	2.13	(2.02-2.24)	6,975	2.98	(2.91-3.05)
Pilocytic astrocytoma	458	0.65	(0.59 - 0.71)	2,363	1.01	(0.97 - 1.05)
Other low grade glioma	372	0.52	(0.47 - 0.58)	1,924	0.82	(0.79 - 0.86)
High grade glioma	372	0.53	(0.48 - 0.59)	1,412	0.60	(0.57 - 0.64)
Ependymal tumors	195	0.27	(0.23 - 0.31)	684	0.29	(0.27 - 0.31)
Other glioma	115	0.16	(0.13 - 0.19)	592	0.25	(0.23 - 0.27)
Choroid plexus tumors	81	0.11	(0.09 - 0.13)	281	0.12	(0.11 - 0.14)
Tumors of the pineal region	119	0.17	(0.14-0.21)	582	0.25	(0.23 - 0.27)
Neuronal and mixed neuronal-glial tumors	30	0.04	(0.03 - 0.06)	110	0.05	(0.04 - 0.06)
Embryonal tumors	505	0.69	(0.63 - 0.75)	1,908	0.82	(0.78 - 0.85)
Medulloblastoma	302	0.42	(0.37 - 0.47)	1,192	0.51	(0.48 - 0.54)
Primitive neuroectodermal tumor	70	0.10	(0.07 - 0.12)	290	0.12	(0.11 - 0.14)
Atypical teratoid/rhabdoid tumor	97	0.12	(0.10-0.15)	266	0.11	(0.10-0.13)
Other embryonal tumors	36	0.05	(0.03 - 0.07)	160	0.07	(0.06 - 0.08)
Tumors of cranial and spinal nerves	128	0.18	(0.15-0.22)	630	0.27	(0.25 - 0.29)
Tumors of meninges	79	0.11	(0.09 - 0.14)	379	0.16	(0.14-0.18)
Lymphomas and hematopoietic neoplasms	19	0.03	(0.02 - 0.04)	51	0.02	(0.02 - 0.03)
Germ cell tumors	119	0.17	(0.14-0.21)	471	0.20	(0.18 - 0.22)
Tumors of the pituitary	158	0.24	(0.20-0.28)	467	0.20	(0.18 - 0.21)
Craniopharyngioma	154	0.22	(0.19 - 0.26)	494	0.21	(0.19 - 0.23)
Other/unclassified tumors	186	0.27	(0.23 - 0.31)	606	0.26	(0.24 - 0.28)
TOTAL <sup>c</sup>	3,090	4.36	(4.21-4.56)	12,954	5.53	(5.43-5.62)

<sup>&</sup>lt;sup>a</sup>Rates are per 100,000 and are age-adjusted to the 2000 US standard population.

#### Incidence by Race and Ethnicity

Counts and incidence rates by histologic groups and race are presented in Table 6.

- Incidence of brain and CNS tumors was highest in Whites and Asian/Pacific islanders (API). Overall age-adjusted incidence in these groups was 5.46 per 100,000, and 6.05 per 100,000, respectively.
- Gliomas and embryonal tumors were most common in white children, with age-adjusted incidence rates of 2.92 per 100,000 and 0.84 per 100,000, respectively.
- Germ cell tumors and tumors of the cranial and spinal nerves were most common in API children, with age-adjusted incidence rates of 0.52 per 100,000 and 0.39 per 100,000, respectively.

Counts and incidence rates by histologic groups and ethnicity are presented in Table 7.

• Incidence of brain and CNS tumors was highest in non-Hispanic children, with an overall age-adjusted incidence of 5.53 per 100,000 as compared to 4.36 per 100,000 in Hispanic children.

 Specific histologies that occurred more frequently in non-Hispanic children included: pilocytic astrocytomas, other low grade gliomas, tumors of the pineal region, medulloblastoma, and tumors of cranial and spinal nerves. Other histologies occurred at similar rates within both groups.

#### Incidence by Age Groups

Overall incidence and incidence of specific histologies varied by age at diagnosis. Counts and incidence rates by histologic groups and age are presented in Table 5 and Figure 10.

- Incidence of embryonal tumors, choroid plexus tumors, and germ cell tumors were highest in infants. Among the embryonal tumors, ATRT occurred notably more frequently in infants.
- Incidence of choroid plexus tumors drops significantly from children <0 to children 0-4 years.
- Gliomas were most common in children ages 1-4, though children ages 5-9 had the highest incidence of high grade gliomas.
- Incidence of high grade glioma peaked in this age group.
- Pilocytic astrocytomas were most common in children 1-4.

<sup>&</sup>lt;sup>b</sup>Hispanic ethnicity is not mutually exclusive of race; Classified using the North American Association of Central Cancer Registries Hispanic Identification Algorithm, version 2 (NHIA v2).

<sup>&</sup>lt;sup>c</sup>Refers to all brain tumors including histologies not presented in this table.

<sup>-</sup> Counts are not presented when fewer than 16 cases were reported for the specific histology category. Suppressed cases are included in the total count. Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; CI, confidence interval.

**Table 8.** Average Annual Age-Adjusted Incidence Rates<sup>a</sup> for Childhood Brain and Central Nervous System Tumors by Region of the United States and Age at Diagnosis, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011

		Age At	Diagno	osis (years)												
		0-14			<1			1-4			5-9			10-14		
Region	Included states	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI	N	Rate	95% CI
New England	Connecticut, Maine, Massachusetts, New Hampshire, Rhode Island, Vermont;	720	5.59	(5.19-6.01)	50	6.38	(4.74-8.41)	192	5.96	(5.14 – 6.86)	228	5.33	(4.66-6.07)	250	5.41	(4.76-6.12)
Middle Atlantic	New Jersey, New York, Pennsylvania;	2,175	5.78	(5.53 – 6.02)	168	6.85	(5.85-7.97)	590	6.08	(5.60-6.60)	654	5.29	(4.89-5.71)	763	5.82	(5.41-6.25)
South Atlantic	Delaware, District of Columbia, Florida, Georgia, Maryland, North Carolina, South Carolina, Virginia, West Virginia	2,948	5.20	(5.01 – 5.39)	230	6.09	(5.33-6.93)	884	5.87	(5.49 – 6.27)	897	4.81	(4.50-5.14)	937	4.90	(4.59 – 5.22)
East North Central	Illinois, Indiana, Michigan, Ohio, Wisconsin	2,482	5.40	(5.19 – 5.61)	198	6.80	(5.88 – 7.81)	701	5.91	(5.48 – 6.36)	777	5.08	(4.73 – 5.45)	806	5.06	(4.72 – 5.42)
East South Central	Alabama, Kentucky, Mississippi, Tennessee	943	5.19	(4.87 – 5.53)	82	6.77	(5.39-8.41)	228	4.75	(4.16-5.41)	318	5.29	(4.72 – 5.90)	315	5.13	(4.58 – 5.73)
West North Central	Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, South Dakota	1,046	5.08	(4.78 – 5.40)	70	5.08	(3.96-6.42)	327	5.94	(5.31-6.62)	345	5.08	(4.56 – 5.65)	304	4.43	(3.94 – 4.95)
West South Central	Arkansas, Louisiana, Oklahoma, Texas	2,285	5.75	(5.51 – 5.99)	189	7.00	(6.04-8.07)	656	6.10	(5.65-6.59)	737	5.60	(5.20-6.02)	703	5.39	(5.00-5.80)
Mountain	Arizona, Colorado, Idaho, Montana, New Mexico, Utah, Wyoming		4.69	(4.40-4.99)	80	5.68	(4.50 – 7.06)	315	5.49	(4.90-6.13)	295	4.25	(3.78-4.77)	292	4.33	(3.85-4.86)
Pacific	Alaska, California, Hawaii, Nevada, Oregon, Washington	2,463	4.69	(4.51 – 4.88)	188	5.31	(4.57 – 6.12)	699	4.99	(4.63 – 5.38)	770	4.53	(4.22 – 4.87)	806	4.50	(4.19-4.82)
TOTAL <sup>b</sup>	<u> </u>	16,044	5.26	(5.18-5.34)	1,255	6.22	(5.88-6.58)	4,592	5.53	(5.53-5.86)	5,021	5.00	(4.86-5.14)	5,176	5.00	(4.87-5.14)

<sup>&</sup>lt;sup>a</sup>Rates are per 100,000 and are age-adjusted to the 2000 US standard population.

<sup>&</sup>lt;sup>b</sup>Refers to all brain tumors including histologies not presented in this table.

<sup>-</sup> Counts and rates are not presented when fewer than 16 cases were reported for the specific histology category. The suppressed cases are included in the counts and rates for totals. Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States; CI, confidence interval.

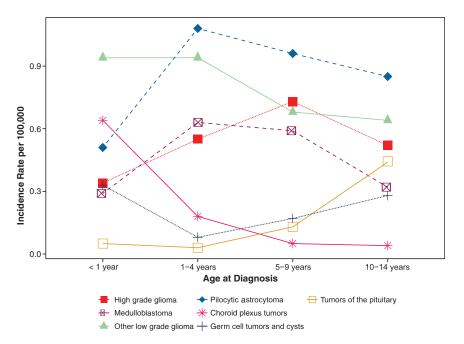


Fig. 10. Age-Adjusted Incidence Rates of Brain and CNS Tumors by Selected Histologies and Age Groups (CBTRUS 2007-2011)

Table 9. Estimated Number of Cases<sup>a,b</sup> of Brain and Central Nervous System Tumors by Age, Major Histology Groupings, and Histology, 2015, 2016

Histology	2015 Es	timated N	lew Cases	5		2016 Es	timated N	New Cases	5	
	0-14	<1	1-4	5-9	10-14	0-14	<1	1-4	5-9	10-14
Gliomas	1,810	100	590	610	510	1,820	110	590	620	520
Pilocytic astrocytoma	600	-	190	210	180	610	-	190	210	180
Other low grade glioma	490	-	170	150	130	490	-	170	150	140
High grade glioma	380	-	100	160	110	390	-	100	160	110
Ependymal tumors	190	-	90	-	-	190	-	90	-	-
Other glioma	150	-	-	50	50	150	-	-	50	50
Choroid plexus tumors	80	-	-	-	-	80	-	-	-	-
Tumors of the pineal region	150	-	-	-	70	150	-	-	-	70
Neuronal and mixed neuronal-glial tumors	-	-	-	-	-	-	-	-	-	-
Embryonal tumors	510	70	210	160	90	520	70	210	160	90
Medulloblastoma	320	-	110	130	70	320	-	110	130	70
Primitive neuroectodermal tumor	80	-	-	-	-	80	-	-	-	-
Atypical teratoid/rhabdoid tumor	80	-	-	-	-	80	-	-	-	-
Other embryonal tumors	-	-	-	-	-	-	-	-	-	-
Tumors of cranial and spinal nerves	160	-	50	50	50	160	-	50	50	50
Tumors of meninges	100	-	-	-	-	100	-	-	-	-
Lymphomas and hematopoietic neoplasms	-	-	-	-	-	-	-	-	-	-
Germ cell tumors	120	-	-	-	60	120	-	-	40	60
Tumors of the pituitary	130	-	-	-	90	130	_	-	-	90
Craniopharyngioma	140	-	-	60	-	140	_	-	60	-
Other/unclassified tumors	170	-	-	50	70	170	-	-	50	70
TOTAL <sup>c</sup>	3,420	280	990	1,080	1,050	3,440	280	990	1,090	1,060

<sup>&</sup>lt;sup>a</sup>Source: Estimation based on CBTRUS (NPCR and SEER 2007 - 2011) data, and US Census population estimates.

<sup>&</sup>lt;sup>b</sup>Rounded to the nearest 10. Numbers may not add up due to rounding.

<sup>&</sup>lt;sup>c</sup>Refers to all brain tumors including histologies not presented in this table.

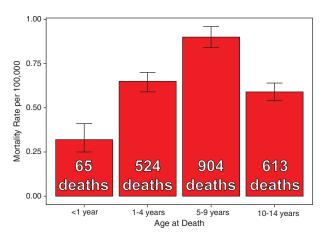
<sup>-</sup> Estimated number is less than 50 and may affect totals.

Abbreviations: CBTRUS, Central Brain Tumor Registry of the United States.

- Medulloblastomas were most common in children ages 1-4 (0.63 per 100,000), though incidence was similar in children 5-9 (0.59 per 100,000).
- Pituitary tumors increase in incidence with age and were most common in children ages 10–14.

#### Number of Estimated New Cases for 2015 And 2016

The estimated number of cases of all primary brain and CNS tumors for 2015 and 2016 by histology and age are shown in Table 9.



**Fig. 11.** Average Annual Age-Adjusted Mortality Rates for Malignant Primary Brain and CNS Tumors by Age Groups (N = 2,106) (NVSS 2007–2011)

- For 2015, the total estimated new cases in children 0-14 years is 3.420.
- For 2016, the total estimated new cases in children 0-14 years is 3,440.

## Mortality Rates by Region of the United States and Age Group

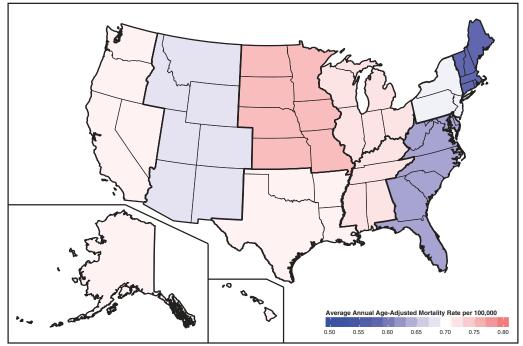
Mortality rates due to malignant brain and CNS tumor varied by age group, with the highest mortality occurring in children 5–9 years (0.90 per 100,000) at time of death (Figure 11), and the lowest mortality rates in infants (<1 year at time of death) (0.32 per 100,000).

Average annual age-adjusted mortality rates by region of the United States are presented in Figure 12. The highest mortality was in the West North Central region (0.75 per 100,000, 95% CI: 0.63 – 0.87), and the lowest mortality rate was in New England (0.58 per 100,000, 95% CI: 0.46 – 0.73).

#### Relative Survival by Site

Relative survival after diagnosis with a brain and CNS tumor varies by site. One-year, two-year, five-year, and ten-year survival rates by site are presented in Table 10.

- Brain stem tumors had lower relative survival than tumors diagnosed at any other location, with one- and ten-year survival of 69.2% and 45.6%, respectively.
- Tumors of the cranial nerve had the highest survival rates, with 99.9% one-year and 97.8% ten-year survival.



New England: Connecticut, Maine, Massachusetts, New Hampshire, Rhode Island, Vermont; Middle Atlantic: New Jersey, New York, Pennsylvania; South Atlantic: Delaware, District of Columbia, Florida, Georgia, Maryland, North Carolina, South Carolina, Siryinia; Mest Virginia; East North Central: Illinois, Indiana, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennesses; West North Central: Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, South Dakota, South Dakota, South Central: Arkansas, Louisiana, Oklahoma, Texas; Mountain: Arizona, Colorado, Idaho, Montana, New Mexico, Utah, Wyoming; Pacific: Alaska, California, Hawaii, Nevada, Oregon, Washington.

**Fig. 12.** Average Annual Age-adjusted Mortality Rates for Malignant Primary Brain and CNS Tumors by Region of the United States (N = 2,106) (NVSS 2007 - 2011)

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**Table 10.** One-, Two-, Five-, and Ten-Year Relative Survival Rates<sup>a</sup> for Malignant Brain and Central Nervous System Tumors by Site<sup>b</sup>, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011<sup>c</sup>

			1-Year		2-Year		5-Year		10-Year	
ICD-O-3 CODE	SITE <sup>b</sup>	N	%	95% CI	%	95% CI	%	95% CI	%	95% CI
C71.1	Frontal lobe of the brain	406	87.5	(83.8-90.4)	80.1	(75.7-83.8)	73.5	(68.5 – 77.9)	69.4	(63.9-74.2)
C71.2	Temporal lobe of the brain	429	91.2	(88.0-93.6)	86.9	(83.1 - 89.9)	81.9	(77.5 – 85.5)	76.3	(70.7 - 81.0)
C71.3	Parietal lobe of the brain	253	88.5	(83.8-92.0)	80.5	(74.8 - 85.0)	73.9	(67.4 - 79.3)	69.4	(62.0 – 75.7)
C71.4	Occipital lobe of the brain	92	95.5	(88.4 - 98.3)	89.6	(81.0 - 94.5)	83.4	(73.4 - 89.9)	78.2	(66.7-86.1)
C71.0	Cerebrum	623	83.2	(79.9 - 85.9)	73.2	(69.4 - 76.7)	68.5	(64.4 - 72.2)	65.7	(61.3 - 69.8)
C71.5	Ventricle	508	85.3	(81.8-88.1)	78.7	(74.7 - 82.2)	71.5	(67.0-75.6)	67.5	(62.4-72.0)
C71.6	Cerebellum	2,047	91.5	(90.2-92.6)	86.0	(84.3 – 87.5)	80.8	(78.9-82.6)	76.8	(74.5 – 78.9)
C71.7	Brain stem	1,610	69.2	(66.9 - 71.5)	54.9	(52.3 - 57.4)	48.7	(46.1-51.3)	45.6	(42.8 - 48.4)
C71.8-C71.9	Other brain	1,279	86.3	(84.2 - 88.1)	82.0	(79.7 - 84.1)	74.9	(72.2 - 77.3)	70.5	(67.3 – 73.3)
C72.0-C72.1	Spinal cord and cauda equina	333	87.4	(83.3-90.6)	81.8	(77.1 - 85.7)	78.7	(73.6 - 82.9)	75.1	(69.0 - 80.1)
C72.2-C72.5	Cranial nerves	529	99.9	(97.0-100.0)	99.7	(98.0 - 99.9)	98.6	(96.7 – 99.4)	97.8	(95.2-99.0)
C72.8-C72.9	Other nervous system	59	84.3	(71.9-91.6)	80.6	(67.5-88.8)	67.3	(52.4 – 78.5)	63.6	(47.6 – 76.0)
C70.0-C70.9	Meninges (cerebral and spinal)	_	_	_	_	_	_	_	_	_
C75.1-C75.2	Pituitary and craniopharyngeal duct	41	100.0	(100.0 - 100.0)	97.2	(81.2-99.6)	93.7	(76.5-98.4)	93.7	(76.5-98.4)
C75.3	Pineal	289	89.8	(85.5-92.9)	83.2	(78.1-87.3)	76.2	(70.1-81.2)	68.2	(60.4 – 74.8)
C30.0 <sup>d</sup>	Olfactory tumors of the nasal cavity	41	92.5	(78.3 – 97.5)	81.3	(64.5 – 90.6)	75.0	(57.2-86.3)	75.0	(57.2-86.3)
All Codes	All Sites	8,564	85.5	(84.7-86.2)	78.2	(77.3-79.1)	72.6	(71.5-73.6)	68.7	(67.6-69.6)

<sup>&</sup>lt;sup>a</sup>The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases.

<sup>&</sup>lt;sup>b</sup>The sites referred to in this table are loosely based on the categories and site codes defined in the SEER Site/Histology Validation List.

cestimated by CBTRUS using Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER\*Stat Database: Incidence – SEER 18 Regs Research Data + Hurricane Katrina Impacted Louisiana Cases, Nov 2013 Sub (1973 – 2011 varying) - Linked To County Attributes - Total U.S., 1969 – 2012 Counties, National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistics Branch, released April 2014, based on the November 2013 submission.

dICD-O-3 histology codes 9522 – 9523 only.

<sup>-</sup> Rates are excluded when calculated based on a population of less than 50, or when less than 16 remain alive in the survival period.

Abbreviation: CBTRUS, Central Brain Tumor Registry of the United States; SEER, Survival, Epidemiology and End Results; CI, confidence interval.

**Table 11.** One-, Five-, and Ten-Year Relative Survival Rates<sup>a</sup> for Malignant Brain and Central Nervous System Tumors by Site<sup>b</sup> and Year of Diagnosis, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011<sup>c</sup>

				1-Year		5-Year		10-Yeo	ar
ICD-O-3 CODE	SITE <sup>b</sup>	Years of Diagnosis	N	%	95% CI	%	95% CI	%	95% CI
C71.0-C71.4	Supratentorial	1973-1976	71	77.4	(65.7-85.5)	56.0	(43.6-66.7)	53.1	(40.8-64.0)
		1977-1981	136	77.3	(69.3-83.4)	59.6	(50.8-67.3)	53.7	(44.9-61.7)
		1982-1986	159	83.0	(76.1-88.0)	63.9	(55.9-70.9)	59.6	(51.4-66.8)
		1987-1991	212	85.0	(79.4-89.1)	70.3	(63.6-76.0)	68.5	(61.7-74.4)
		1992-1996	298	87.0	(82.6-90.3)	74.5	(69.2-79.1)	71.3	(65.7-76.1)
		1997-2001	431	85.6	(431-85.6)	72.1	(67.6-76.1)	68.0	(63.3 – 72.3)
		2002 – 2006	614	87.8	(84.9-90.1)	74.1	(70.3 – 77.4)	69.8	(64.1-74.8)
		2007-2011	640	87.8	(84.9-90.3)	74.4	(68.8-79.3)		_
C71.5	Ventricle, NOS	1973 – 1976	-	-	-	-	-	-	-
		1977-1981	-	-	-	-	-	-	-
		1982-1986	-	-	-	-	-	-	-
		1987-1991	-	-	-	-	-	-	-
		1992-1996	68	79.6	(67.9-87.4)	62.0	(49.3-72.4)	60.6	(47.9-71.1)
		1997-2001	133	83.5	(76.0-88.9)	71.5	(63.0-78.4)	64.4	(55.5-72.0)
		2002 – 2006	158	87.4	(81.1-91.7)	71.4	(63.6-77.8)	_	_
		2007 - 2011	192	84.8	(78.5-89.4)	_	_	_	_
C71.6	Cerebellum, NOS	1973-1976	143	74.1	(66.1-80.5)	52.3	(43.8-60.1)	43.9	(35.6-51.9)
C/ 110	cereseitarry ress	1977-1981	230	82.3	(76.7 – 86.7)	62.4	(55.7-68.3)	57.3	(50.6-63.4)
		1982-1986	200	86.6	(81.0-90.6)	71.5	(64.6-77.3)	68.1	(61.0 – 74.1)
		1987-1991	220	87.7	(82.6-91.4)	74.9	(68.6-80.2)	71.8	(65.3 – 77.3)
		1992-1996	334	89.6	(85.8-92.4)	77.6	(72.7-81.7)	74.2	(69.1 – 78.6)
		1997 – 2001	528	91.1	(88.3-93.3)	82.3	(78.8-85.4)	78.6	(74.8-81.9)
		2002 – 2006	667	92.5	(90.2-94.3)	81.3	(78.0-84.1)	_	_
		2007 - 2011	707	91.9	(89.5-93.8)	78.7	(72.2-83.8)	_	_
C71.7	Brain stem	1973-1976	_	_	_	_	_	-	
		1977-1981	76	51.4	(76.0-51.4)	25.1	(16.0-35.2)	23.8	(14.9-33.8)
		1982-1986	125	57.5	(48.3 – 65.6)	35.7	(27.3-44.1)	32.5	(24.4-40.8)
		1987-1991	154	62.4	(54.2-69.5)	33.6	(26.2-41.1)	33.6	(26.2-41.1)
		1992-1996	236	58.4	(51.8-64.4)	42.7	(36.3-48.9)	37.6	(31.4-43.8)
		1997-2001	371	68.9	(64.0-73.4)	47.7	(42.6-52.7)	45.6	(40.4-50.6)
		2002-2006	558	69.9	(65.9 – 73.6)	51.6	(47.3 – 55.7)	45.7	(39.5-51.7)
		2007-2011	603	69.7	(65.6-73.3)	46.6	(40.6-52.4)		(39.3-31.7)
671.0.671.0	Oth an Duale							-	- (/2 / 50.0)
C71.8-C71.9	Other Brain	1973 - 1976	125	69.7	(60.8-77.0)	54.6	(45.4–62.8)	51.5	(42.4-59.9)
		1977 - 1981 1982 - 1986	125 101	77.5 78.4	(69.1 – 83.9) (69.0 – 85.3)	59.7 59.4	(50.4-67.7) (49.1-68.3)	54.9 57.3	(45.6-63.2) (47.0-66.4)
		1987-1991	125	80.1	(71.9-86.1)	61.0	(51.8–68.9)	57.0	(47.8-65.2)
		1992-1996	227	86.4	(81.2-90.3)	70.5	(64.1 – 76.0)	66.5	(59.9 – 72.3)
		1997-2001	300	83.7	(79.0-87.5)	70.3	(66.8-77.0)	68.0	(62.3 – 73.1)
		2002 - 2006	454	85.9	(82.3-88.8)	74.1	(69.7–77.9)	-	(02.5-75.1)
		2007-2011	438	87.6	(83.9-90.5)	76.7	(69.3-82.6)	_	_
C70.0-C70.9, C72.0-C72.9	Other Nervous System	1973-1976	58	70.9	(57.3 – 80.9)	58.9	(45.1 – 70.3)	52.1	(38.5-64.1)
C75.1-C75.3 C30.0 <sup>d</sup>	Carici Nervous System	1977-1981	86	78.1	(67.7-85.5)	61.9	(50.7–71.3)	58.5	(47.3 – 68.2)
		1982-1986	89	84.3	(74.8-90.4)	63.9	(52.9-73.0)	60.6	(49.6-70.0)
		1987-1991	112	89.3	(81.9-93.8)	78.6	(69.7-85.2)	74.2	(64.8-81.4)
		1992-1996	152	92.2	(86.6-95.5)	82.4	(75.3-87.6)	77.2	(69.6-83.2)
		1997-2001	277	91.7	(87.8-94.5)	81.6	(76.5-85.7)	79.9	(74.5-84.2)
		2002 – 2006	452	92.7	(89.9-94.8)	87.1	(83.6-89.9)	-	-
		2007-2011	478	93.0	(90.0-95.0)	-	-	-	-

Continued

Table 11. Continued

				1-Year		5-Year		10-Year	
ICD-O-3 CODE	SITE <sup>b</sup>	Years of Diagnosis	N	%	95% CI	%	95% CI	%	95% CI
All Codes	All Sites	1973-1976	467	70.5	(66.1-74.4)	51.2	(46.5-55.6)	45.9	(41.3-50.4)
		1977-1981	690	76.0	(72.6-79.0)	56.3	(52.5-59.9)	51.7	(47.9-55.4)
		1982-1986	706	78.8	(75.6-81.6)	59.8	(56.1-63.3)	56.3	(52.5-59.9)
		1987-1991	878	81.1	(78.4-83.6)	64.2	(60.9-67.3)	61.6	(58.2-64.7)
		1992-1996	1,321	82.7	(80.6-84.7)	69.2	(66.7-71.7)	65.4	(62.7-67.9)
		1997-2001	2,048	84.5	(82.8-86.0)	71.7	(69.6-73.6)	68.1	(66.0-70.1)
		2002-2006	2,915	85.9	(84.6-87.1)	73.3	(71.6-74.8)	69.2	(67.0-71.3)
		2007-2011	3,076	85.7	(84.4-87.0)	71.9	(69.3-74.3)	_	_

<sup>&</sup>lt;sup>a</sup>The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases.

Abbreviation: CBTRUS, Central Brain Tumor Registry of the United States; SEER, Survival, Epidemiology and End Results; CI, confidence interval.

For most sites, relative survival after diagnosis has improved over time, and these data are presented in Table 11.

- For tumors that occur supratentorially (in the cerebrum, frontal lobe, occipital lobe, parietal lobe, and temporal lobe), 1-year survival for tumors diagnosed from 1977–1981 was 77.3%, whereas 1-year survival was 87.8% from 2007–2011. 10-year survival for these tumors was 53.7% from 1977–1981, and 69.8% from 2002–2006.
- For tumors that occur in the brain stem, 1-year survival was 51.4% for tumors diagnosed from 1977–1981, as compared to 69.7% for tumors diagnosed from 2007–2011. 10-year survival for these tumors was 23.8% from 1977–1981, and 45.7% from 2002–2006.

#### Relative Survival by Histologic Group

Relative survival after diagnosis with primary brain and CNS tumor varies by histologic type of tumor, and is presented in Table 12.

- ATRT and high grade glioma were the histologic groups with the lowest relative survival after diagnosis. ATRT had one-, five-, and ten-year survival of 48.1%, 27.5%, and 26.0%, respectively. High grade glioma had one-, five-, and ten-year survival of 55.6%, 28.4%, and 25.7%, respectively.
- Pilocytic astrocytoma and other low grade glioma had some of the highest survival rates after diagnosis. Ten-year survival with these tumors was 95.9% and 84.6%, respectively.

For most histologies, relative survival after diagnosis has improved over time, and these data are presented in Table 13.

 For all malignant brain tumors, 1-year survival was 70.5% from 1973 – 1976, as compared to 85.7% from 2007 – 2011. 10-year

- survival was 45.9% in 1973–1976, as compared to 69.2% in 2002–2006.
- Survival after diagnosis with high grade glioma remained relatively stable, with 1-year survival rates of 59.3% from 1982-1986, and 57.1% from 2007-2011. 10-year survival was 30.8% from 1982-1986, and 24.7% from 1997-2001.

#### Relative Survival by Age Group and Histologic Group

Relative survival rates generally improved with increasing age at diagnosis. One-year, two-year, five-year, and ten-year relative survival rates by histologic group and age groups are presented in Table 14.

- Overall, survival was better in older children. Though there
  was not much difference in relative survival between children
  ages 1-4 and 5-9 years, differences could be seen in
  histologies that depend on treatment with radiation such
  as medulloblastoma and PNET.
- Though infants generally had poor survival, the long-term survival of infants with high grade gliomas was higher than children of other ages, with a ten-year survival of 54.1%.
- Five-year survival in children 10-14 years was 79.2% for all brain tumors, as compared to infants where five-year survival was 55.3%.
- For embryonal tumors, infants (<1 year old) had one-year survival of 52.8%, whereas those 10-14 years had relative survival of 92.2%.</li>

### Distribution of Deaths due to Selected Histologic Groups and Site

Between 2007 and 2011, there were 734 total deaths in children ages 0–14 due to primary malignant brain tumors between 1995

<sup>&</sup>lt;sup>b</sup>The sites referred to in this table are loosely based on the categories and site codes defined in the SEER Site/Histology Validation List.

Estimated by CBTRUS using Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER\*Stat Database: Incidence – SEER 18 Regs Research Data + Hurricane Katrina Impacted Louisiana Cases, Nov 2013 Sub (1973 – 2011 varying) – Linked To County Attributes – Total U.S., 1969 – 2012 Counties, National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistics Branch, released April 2014, based on the November 2013 submission.

<sup>&</sup>lt;sup>d</sup>ICD-O-3 histology codes 9522-9523 only.

<sup>-</sup> Rates are excluded when calculated based on a population of less than 50, when less than 16 remain alive in the survival period, or when not enough follow up time has passed to calculate survival for the listed period.

		1-Yr		2-Yr		3-Yr		4-Yr		5-Yr		10-Yr	
Histology	N	%	95% CI	%	95% CI								
Gliomas	5,814	87.2	(86.3-88.1)	80.6	(79.5-81.6)	78.1	(77.0-79.2)	76.8	(75.6-77.9)	76.0	(74.8-77.2)	73.3	(72.0 – 74.6)
Pilocytic astrocytoma	2,131	98.8	(98.2-99.2)	98.5	(97.9-99.0)	98.1	(97.3-98.6)	97.4	(96.5-98.1)	97.1	(96.1-97.8)	95.9	(94.6-96.9)
Other low grade glioma	936	95.4	(93.8-96.6)	91.7	(89.7-93.4)	89.6	(87.4-91.5)	88.3	(85.9-90.4)	87.3	(84.7-89.4)	84.6	(81.6-87.1)
High grade glioma	1,347	55.6	(52.9-58.3)	35.0	(32.3 - 37.6)	30.0	(27.4-32.6)	28.9	(26.3-31.5)	28.4	(25.9-31.0)	25.7	(23.1-28.4)
Ependymal tumors	655	93.3	(91.0-95.0)	86.3	(83.2-88.9)	80.4	(76.9-83.5)	75.8	(71.9-79.2)	72.7	(68.6-76.4)	63.8	(58.9-68.3)
Other glioma	745	96.9	(95.3-98.0)	94.4	(92.3-95.9)	93.1	(90.9-94.9)	92.6	(90.2-94.4)	92.4	(89.9-94.2)	91.2	(88.3-93.4)
Choroid plexus tumors	78	81.3	(70.3 - 88.5)	76.7	(65.0-84.9)	68.2	(55.5 – 77.9)	64.5	(51.5-74.9)	60.5	(47.2 – 71.5)	58.0	(44.2-69.4)
Tumors of the pineal region	41	97.5	(82.7-99.7)	91.7	(76.1 - 97.3)	88.5	(71.8-95.6)	88.5	(71.8-95.6)	88.5	(71.8-95.6)	88.5	(71.8-95.6)
Neuronal and mixed neuronal-glial tumors	83	84.8	(74.7-91.1)	74.8	(63.2-83.2)	66.7	(54.4 – 76.5)	59.3	(46.4-70.2)	54.9	(41.5-66.4)	43.4	(29.0-57.0)
Embryonal tumors	1,908	0.08	(78.1-81.8)	70.3	(68.1-72.4)	66.6	(64.3 - 68.8)	64.3	(62.0-66.5)	62.1	(59.7-64.4)	55.9	(53.3-58.5)
Medulloblastoma	1,124	86.8	(84.7-88.7)	79.4	(76.8-81.7)	75.2	(72.5 - 77.8)	72.8	(69.8-75.4)	70.1	(67.0-72.9)	63.0	(59.5-66.4)
Primitive neuroectodermal tumor	442	76.9	(72.7-80.6)	64.1	(59.3-68.4)	60.2	(55.4-64.8)	58.1	(53.2-62.7)	56.0	(51.1-60.7)	49.2	(43.9 – 54.4)
Atypical teratoid/rhabdoid tumor	197	48.1	(40.8-55.0)	32.5	(25.7 - 39.5)	29.5	(22.8-36.5)	28.6	(21.9 - 35.7)	27.5	(20.8 - 34.7)	26.0	(19.1 - 33.4)
Other embryonal tumors	145	80.2	(72.6-86.0)	70.2	(61.7-77.1)	68.5	(59.9-75.7)	65.7	(56.8-73.2)	64.7	(55.7-72.3)	63.4	(54.3 – 71.3)
Tumors of cranial and spinal nerves	-	-	-	-	-	_	-	-	-	-	-	-	-
Tumors of meninges	92	76.5	(66.2-84.1)	64.2	(53.0 - 73.4)	58.9	(47.6-68.6)	57.6	(46.1-67.4)	56.0	(44.5-66.0)	48.3	(36.2-59.4)
Lymphomas and hematopoietic neoplasms	55	87.3	(75.2-93.7)	85.4	(72.8 - 92.4)	83.2	(70.0 - 90.9)	80.7	(66.8-89.2)	80.7	(66.8-89.2)	76.2	(59.3-86.9)
Germ cell tumors	383	93.3	(90.2-95.4)	91.2	(87.7-93.7)	89.9	(86.2-92.7)	88.1	(84.2-91.2)	86.6	(82.3-89.9)	80.1	(74.1-84.8)
Other/unclassified tumors	97	59.9	(49.3-69.0)	52.9	(42.2-62.5)	50.3	(39.6-60.1)	50.3	(39.6-60.1)	50.3	(39.6-60.1)	43.4	(31.6-54.6)
TOTAL: All Brain and Other Nervous System <sup>d</sup>	8,564	85.5	(84.7-86.2)	78.2	(77.3–79.1)	75.4	(74.4–76.4)	73.8	(72.8–74.8)	72.6	(71.5-73.6)	68.7	(67.6–69.9)

<sup>&</sup>lt;sup>a</sup>The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases.

Abbreviation: CBTRUS, Central Brain Tumor Registry of the United States; SEER, Survival, Epidemiology and End Results; CI, confidence interval; NOS, not otherwise specified.

<sup>&</sup>lt;sup>b</sup>Rates are an estimate of the percentage of patients alive at one, two, three, four, five, and ten year, respectively.

<sup>&</sup>lt;sup>c</sup>Estimated by CBTRUS using Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER\*Stat Database: Incidence - SEER 18 Regs Research Data + Hurricane Katrina Impacted Louisiana Cases, Nov 2013 Sub (1973 – 2011 varying) - Linked To County Attributes - Total U.S., 1969 – 2012 Counties, National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistics Branch, released April 2014, based on the November 2013 submission dIncludes histologies not listed in this table.

<sup>-</sup>Rates are excluded when calculated based on a population of less than 50, or when less than 16 remain alive in the survival period.

**Table 13.** One-, Five-, and Ten-Year Relative Survival Rates<sup>a,b</sup> for Selected Malignant Brain and Central Nervous System Tumors by Histology and Year of Diagnosis, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007 – 2011<sup>c</sup>

			1-Yr		5-Yr		10-Yr	
Histology	Years of Diagnosis	N	%	95% CI	%	95% CI	%	95% CI
Gliomas	1973-1976	310	74.2	(68.9 – 78.7)	56.1	(50.3-61.4)	52.0	(46.2-57.4)
	1977-1981	476	77.0	(72.9 - 80.5)	59.7	(55.2-64.0)	55.4	(50.8-59.8)
	1982-1986	486	80.9	(77.0 - 84.1)	63.6	(59.1-67.7)	60.5	(56.0-64.8)
	1987-1991	636	82.9	(79.7-85.6)	66.9	(63.1 – 70.5)	65.0	(61.1-68.6)
	1992-1996	894	85.2	(82.7 - 87.3)	74.3	(71.3 - 77.1)	71.2	(68.1-74.1)
	1997-2001	1,334	86.3	(84.4-88.1)	75.8	(73.3 – 78.0)	73.2	(70.6-75.5)
	2002 - 2006	2,029	87.8	(86.2 - 89.1)	76.5	(74.6 - 78.3)	74.1	(71.7 – 76.3)
	2007-2011	2,096	87.2	(85.6-88.6)	74.5	(71.5-77.2)	_	-
Pilocytic astrocytoma	1973-1976	-	-	-	-	_	-	_
	1977-1981	_	_	_	_	_	_	-
	1982-1986	56	94.7	(84.1-98.3)	92.9	(81.9-97.3)	91.2	(79.7-96.3)
	1987-1991	109	95.5	(89.3-98.1)	88.2	(80.5-93.1)	88.2	(80.5-93.1)
	1992-1996	270	98.9	(96.6-99.7)	96.0	(92.8-97.8)	94.3	(90.6-96.6)
	1997-2001	500	98.8	(97.4-99.5)	97.5	(95.6-98.6)	96.1	(93.9–97.6)
	2002 – 2006	741	98.7	(97.5-99.3)	97.2	(95.7-98.2)	-	-
	2007-2011	771	98.9	(97.7–99.4)	-	-	-	-
Other low grade glioma	1973 – 1976	117	85.5	(77.6-90.7)	77.0	(68.2 – 83.7)	72.8	(63.7-80.1)
	1977 – 1981	243	86.5	(81.5-90.2)	76.7	(70.8-81.6)	71.1	(64.8-76.4)
	1982-1986	209	89.0	(83.9-92.6)	78.0	(72.1-83.4)	74.6	(68.0-80.0
	1987-1991	233	93.2	(89.0-95.8)	85.4	(80.1-89.4)	82.5	(76.9-86.9
	1992 – 1996	236	95.4	(91.8-97.4)	88.6	(83.7-92.1)	85.7	(80.4-89.7
	1997-2001	242	93.8	(89.8-96.2)	89.2	(84.4-92.5)	86.2	(80.9-90.0)
	2002 – 2006	326	95.7	(92.8-97.5)	85.2	(80.8-88.7)	-	_
	2007 – 2011	283	96.5	(93.4-98.2)	-	-	_	-
High grade glioma	1973 – 1976	67	50.8	(38.3-62.0)	26.9	(17.0-37.9)	-	-
	1977-1981	89	46.1	(35.5-56.0)	20.3	(12.7 - 29.2)	19.2	(11.8-28.0
	1982-1986	128	59.3	(50.2-67.2)	33.2	(25.1-41.4)	30.8	(23.0-39.0)
	1987-1991	169	55.7	(47.8-62.8)	33.0	(26.0-40.2)	32.5	(25.5-39.6)
	1992-1996	207	51.6	(44.6-58.2)	32.1	(25.8–38.6)	29.7	(23.5-36.0)
	1997 - 2001	321	54.2	(48.5-59.4)	27.9	(23.1 – 32.9)	24.7	(20.1 – 29.6)
	2002-2006	450	54.3	(49.5–58.8)	28.1	(24.0-32.4)	-	(20.1 25.0)
								_
	2007-2011	502	57.1	(52.4-61.4)	25.3	(19.7 – 31.2)	-	-
Ependymal tumors	1973 - 1976	-	-	- (60.2 0 ( 5)	-	- (477 (22)	-	-
	1977-1981	51	74.7	(60.3-84.5)	29.5	(17.7-42.3)	27.6	(16.2 - 40.2)
	1982 – 1986	56	80.5	(67.4-88.8)	46.2	(32.7 – 58.6)	42.5	(29.3 – 55.0)
	1987 – 1991	80	88.9	(79.6-94.1)	50.1	(38.7 – 60.5)	46.4	(35.2 – 56.9)
	1992 – 1996	107	87.0	(79.0-92.1)	63.7	(53.8 – 72.0)	54.4	(44.5-63.3)
	1997 – 2001	134	94.8	(89.3-97.5)	71.1	(62.4 – 78.1)	64.0	(55.1 – 71.7)
	2002 – 2006	241	94.2	(90.4–96.6)	72.4	(66.2 – 77.6)	58.3	(45.9-68.8)
	2007 – 2011	231	92.5	(87.8-95.4)	75.4	(60.4-85.3)	-	_
Other glioma	1973 – 1976	76	79.0	(68.0-86.6)	60.7	(48.8 – 70.7)	56.8	(44.9-67.1)
	1977-1981	52	74.9	(60.6-84.7)	51.4	(36.9-64.1)	47.5	(33.3-60.5)
	1982-1986	-	-	-	-	-	-	-
	1987-1991	-	-	-	-	-	-	-
	1992-1996	74	93.3	(84.5-97.2)	82.6	(71.8-89.6)	81.3	(70.3-88.5)
	1997-2001	137	95.0	(89.6-97.6)	89.9	(83.5-93.9)	89.2	(82.6-93.4)
	2002-2006	271	98.2	(95.6-99.3)	93.7	(90.0-96.1)	-	-
	2002-2000	309	97.1	(94.2-98.5)	-	(50.0 50.1)		
	2007-2011	309	57.1	(34.2-30.3)	_	_	-	-

Continued

Table 13. Continued

			1-Yr		5-Yr		10-Yr	
Histology	Years of Diagnosis	N	%	95% CI	%	95% CI	%	95% CI
Embryonal tumors	1973-1976	121	67.1	(57.9-74.7)	41.5	(32.6-50.1)	31.5	(23.4-39.9)
	1977-1981	158	75.4	(67.9-81.4)	47.6	(39.6-55.2)	42.0	(34.2-49.6)
	1982-1986	155	79.5	(72.2-85.0)	55.0	(46.8-62.4)	49.9	(41.8-57.5)
	1987-1991	175	77.2	(70.2 - 82.7)	55.9	(48.2-62.9)	51.4	(43.6-58.5)
	1992-1996	296	75.8	(70.4-80.3)	55.5	(49.7-61.0)	50.8	(44.9-56.3)
	1997-2001	508	80.9	(77.2 - 84.1)	63.4	(59.1-67.5)	57.7	(53.2-61.9)
	2002 – 2006	606	79.5	(76.1-82.5)	61.5	(57.5-65.3)	53.7	(48.0-59.1)
	2007-2011	677	80.3	(76.9-83.2)	60.9	(54.4-66.7)	-	-
Medulloblastoma	1973-1976	100	67.0	(56.9-75.3)	40.1	(30.5 - 49.5)	28.1	(19.7-37.2)
	1977-1981	138	77.6	(69.7-83.7)	47.2	(38.7-55.3)	40.8	(32.5-48.9)
	1982-1986	120	82.0	(73.6-87.6)	57.6	(48.3 – 65.9)	51.9	(42.6-60.4)
	1987-1991	109	82.6	(74.0-88.5)	63.2	(53.3 – 71.5)	56.8	(46.9-65.5)
	1992-1996	158	78.6	(71.3-84.2)	62.1	(54.1-69.2)	57.0	(48.9-64.4)
	1997-2001	273	86.8	(82.2-90.3)	72.1	(66.3 – 77.0)	65.7	(59.7-71.1)
	2002-2006	353	86.7	(82.6-89.8)	61.5	(57.5-65.3)	_	_
	2007-2011	419	88.9	(85.3-91.7)	60.9	(54.4-66.7)	_	_
Primitive neuroectodermal tumor	1973-1976	-	-	-	-	-	_	_
Timilare nearoccioacimal camor	1977-1981	_	_	_	_	_	_	_
	1982-1986	_	_	_	_	_	_	_
	1987-1991	_	_	_	_	_	_	_
	1992-1996	123	72.4	(63.6-79.5)	45.6	(36.6 – 54.2)	41.5	(32.7-50.1)
	1997-2001	162	75.8	(68.4-81.7)	55.2	(47.1 – 62.5)	47.9	(39.9-55.5)
	2002 – 2006	146	79.4	(71.9-85.2)	59.1	(50.6-66.7)	49.3	(36.1-61.3)
	2007-2011	103	74.8	(64.8-82.3)	_	_	_	_
Atypical teratoid/rhabdoid tumor <sup>d</sup>	2002-2006	69	39.8	(28.2-51.2)	19.9	(11.3 – 30.2)	_	_
	2007-2011	111	52.8	(42.7-61.9)	_	_	_	_
TOTAL: All Brain and Other Nervous System <sup>de</sup>	1973-1976	467	70.5	(66.1-74.4)	51.2	(46.5-55.6)	45.9	(41.3-50.4)
	1977-1981	690	76.0	(72.6–79.0)	56.3	(52.5-59.9)	51.7	(47.9-55.4)
	1982-1986	706	78.8	(75.6-81.6)	59.8	(56.1-63.3)	56.3	(52.5-59.9)
	1987-1991	878	81.1	(78.4–83.6)	64.2	(60.9-67.3)	61.6	(58.2-64.7)
	1992-1996	1,321	82.7	(80.6-84.7)	69.2	(66.7–71.7)	65.4	(62.7–67.9)
	1997-2001	2,048	84.5	(82.8-86.0)	71.7	(69.6–73.6)	68.1	(66.0-70.1)
	2002-2006	2,915	85.9	(84.6-87.1)	73.3	(71.6-74.8)	69.2	(67.0-71.3)
	2007-2011	3,076	85.7	(84.4-87.0)	71.9	(69.3-74.3)	_	•

<sup>&</sup>lt;sup>a</sup>The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases.

<sup>&</sup>lt;sup>b</sup>Rates are an estimate of the percentage of patients alive at one, two, three, four, five, and ten year, respectively.

Estimated by CBTRUS using Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER\*Stat Database: Incidence - SEER 18 Regs Research Data + Hurricane Katrina Impacted Louisiana Cases, Nov 2013 Sub (1973 – 2011 varying) - Linked To County Attributes - Total U.S., 1969 – 2012 Counties, National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistics Branch, released April 2014, based on the November 2013 submission

<sup>&</sup>lt;sup>d</sup>Atypical teratoid/rhabdoid tumors were first included in the WHO classification of tumors of the central nervous system in the 2000 revision. <sup>e</sup>Includes histologies not listed in this table.

<sup>-</sup> Rates are excluded when calculated based on a population of less than 50, when less than 16 remain alive in the survival period, or when not enough follow up time has passed to calculate survival for the listed period.

Abbreviation: CBTRUS, Central Brain Tumor Registry of the United States; SEER, Survival, Epidemiology and End Results; CI, confidence interval; NOS, not otherwise specified.

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**Table 14.** One-, Two-, Five-, and Ten-Year Relative Survival Rates<sup>a,b</sup> for Selected Malignant Brain and Central Nervous System Tumors by Age Groups, CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011<sup>c</sup>

			1-Yr		2-Yr		5-Yr		10-Yr	
Histology	Age Group (years)	N	%	95% CI	%	95% CI	%	95% CI	%	95% CI
Gliomas	<1	314	81.7	(76.8-85.7)	78.5	(73.3-82.8)	71.0	(65.1 – 76.1)	68.2	(61.4-74.0
	1-4	1,833	90.9	(89.5-92.2)	84.5	(82.7-86.2)	80.0	(77.9 - 81.9)	77.0	(74.6-79.2
	5-9	1,974	81.5	(79.7-83.2)	73.9	(71.8 - 75.8)	70.4	(68.2 - 72.5)	68.0	(65.6 – 70.3
	10-14	1,693	91.0	(89.5-92.3)	84.5	(82.6-86.2)	79.2	(77.0-81.2)	76.5	(74.1 – 78.8
Pilocytic astrocytoma	<1	74	93.4	(83.7-97.4)	91.8	(81.7-96.5)	80.1	(66.7-88.5)	80.1	(66.7-88.5
	1-4	685	99.1	(98.0-99.6)	98.5	(97.1 - 99.2)	97.5	(95.8 - 98.5)	95.9	(93.1-97.6
	5-9	717	98.8	(97.7 - 99.4)	98.8	(97.7 - 99.4)	98.0	(96.5 - 98.9)	97.2	(94.9-98.4
	10-14	655	98.9	(97.7-99.5)	98.9	(97.7 - 99.5)	97.4	(95.5-98.5)	96.3	(93.7-97.8
Other low grade glioma	<1	59	90.5	(79.0-95.8)	90.5	(79.0-95.8)	77.1	(62.1-86.7)	70.7	(50.8 – 83.7
3 3	1-4	254	94.7	(91.0-96.9)	92.4	(88.2-95.2)	88.9	(83.8-92.4)	88.9	(83.8-92.4
	5-9	260	94.8	(91.3-97.0)	91.4	(87.1-94.3)	88.8	(83.9-92.3)	85.8	(79.9 – 90.1
	10-14	363	97.2	(94.8-98.5)	91.6	(88.1-94.2)	86.7	(82.3-90.0)	82.7	(77.4-86.9
High grade glioma	<1	69	57.4	(44.7 – 68.3)	54.1	(41.4-65.3)	54.1	(41.4-65.3)	54.1	(41.4-65.3
J J 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1 1	1-4	308	61.1	(55.4-66.4)	34.7	(29.3 – 40.3)	27.3	(22.1-32.7)	24.4	(19.1–30.
	5-9	591	45.6	(41.5-49.6)	25.9	(22.3 – 29.7)	20.8	(17.4-24.4)	18.4	(15.0-22.
	10-14	379	66.5	(61.4-71.1)	45.9	(40.5 – 51.0)	36.9	(31.7-42.1)	33.3	(27.9 – 38.
Ependymal tumors	<1	51	77.6	(62.8-87.1)	70.6	(55.0-81.6)	60.1	(43.8-73.0)	51.4	(33.4 – 66.
-μ - · · - · · · · · · · · · · · · · · ·	1-4	309	92.8	(89.2-95.3)	85.8	(81.0-89.4)	71.4	(65.1 – 76.7)	59.4	(51.6-66.
	5-9	167	95.6	(91.0-97.9)	86.7	(80.1-91.2)	73.9	(65.5-80.5)	67.0	(57.1 – 75.
	10-14	128	97.6	(92.6-99.2)	93.1	(86.7-96.5)	79.2	(70.0-85.9)	73.6	(63.2-81.
Embryonal tumors	<1	219	52.8	(45.8–59.3)	41.7	(34.8-48.4)	36.5	(29.7 – 43.4)	34.0	(27.2 – 41.
	1-4	768	74.3	(71.0 – 77.3)	63.4	(59.7-66.8)	55.8	(51.9 – 59.5)	51.9	(47.7 – 55.
	5-9	602	90.9	(88.3-93.0)	81.7	(78.2 – 84.6)	73.1	(69.0 – 76.7)	63.1	(58.1-67.
	10-14	319	92.2	(88.5-94.7)	85.4	(80.7-88.9)	73.9	(68.0 – 78.9)	67.4	(60.6 - 73.
Medulloblastoma	<1	65	55.9	(42.8-67.2)	48.6	(35.5-60.5)	41.6	(28.5 – 54.2)	38.2	(24.7 – 51.
ricadioblastoria	1-4	388	81.0	(76.6-84.6)	71.4	(66.4 – 75.7)	62.3	(56.8-67.3)	58.0	(52.0-63.
	5-9	445	91.9	(88.8-94.1)	84.6	(80.7 – 87.7)	75.7	(71.0-79.7)	65.3	(52.6 - 05.
	10-14	226	95.8	(92.1-97.8)	91.8	(87.0-94.8)	80.7	(73.8-85.9)	74.6	(66.6-81.
Primitive neuroectodermal tumor	<1	-	-	(92.1-97.0)	-	(67.0-34.6)	-	(73.8-83.9)	-	(00.0-81.
Thirlitive neuroectodennat tamor	1-4	205	75.6	(69.0-81.0)	62.5	(55.3-68.9)	54.4	(47.1-61.2)	49.4	(41.6 – 56.
	5-9	123	89.1	(82.0-93.5)	76.0	(67.1-82.8)	67.4	(57.8 – 75.2)	57.3	(45.9 – 67.
	3-9 10-14	71	81.6	(70.4-88.9)	66.9	(54.5 – 76.6)	55.4	(42.6-66.5)	57.5 46.6	(33.3 – 58.9
Atypical teratoid/rhabdoid tumor	10-14 <1	71 67	40.2	(28.3 – 51.9)	19.7	(10.8 – 30.5)	55.4 -	(42.0-00.5)	40.0	(33.3-36.
Αιγρικάι τειαιοία/παράσια ταπιοί	<1 1-4	111	40.2 48.6		36.7		- 33.5	(2/, 1 / 2 2)	- 33.5	(2), 1 (2.1
	1-4 5-9			(38.9-57.7)		(27.4-46.1)		(24.1-43.2)		(24.1-43.2
	5-9 10-14	_	_	-	_	-	-	_	_	_

Germ cell tumors	\ \	ı	ı	I	ı	ı	ı	ı	I	ı
	,									
	1-4	I	I	ı	I	ı	I	ı	I	ı
	5-9	92	8.76	(91.3 - 99.4)	96.5	(89.3 - 98.9)	90.5	(79.3 - 95.8)	87.7	(74.7 - 94.3)
	10 - 14	248	97.1	(94.0 - 98.6)	94.4	(80.6 - 96.8)	89.5	(84.4 - 92.9)	80.8	(72.9 - 86.6)
TOTAL: All Brain and Other Nervous System <sup>d</sup>	7	989	2.79	(63.8-71.3)	61.0	(57.3-65.2)	55.3	(51.1 - 59.3)	53.2	(48.7–57.4)
	1-4	2,758	85.4	(84.0 - 86.7)	78.0	(75.9-79.1)	71.9	(70.0-73.6)	9.89	(9.2–70.6)
	5–9	2,775	84.5	(83.0 - 85.8)	77.0	(75.0-78.2)	71.5	(69.6–73.3)	67.0	(0.69-6.49)
	10-14	2,395	91.5	(90.3–92.6)	85.0	(83.8–86.8)	79.2	(77.4–80.9)	75.1	(72.9–77.1)

The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases

Rates are an estimate of the percentage of patients alive at one, two, five, and ten year, respectively. Rates were not presented for categories with 50 or less cases and were suppressed Estimated by CBTRUS using Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER\*Stat Database: Incidence - SEER 18 Regs Research Data + Hurrifor rates where less than 16 cases were surviving within a category.

cane Katrina Impacted Louisiana Cases, Nov 2013 Sub (1973–2011 varying) - Linked To County Attributes - Total U.S., 1969–2012 Counties, National Cancer Institute, DCCPS, Surveilance Research Program, Cancer Statistics Branch, released April 2014, based on the November 2013 submission. less than 16 remain alive in the survival period. or when Rates are excluded when calculated based on a population of less than 50, <sup>4</sup>Includes histologies not listed in this table.

Epidemiology and End Results; CI, confidence interval; NOS, not otherwise specified.

Survival,

Abbreviation: CBTRUS, Central Brain Tumor Registry of the United States; SEER,

and 2011 in the 18 SEER registries. The distribution of these deaths by histology grouping is presented in Figure 13a, and the distribution of these by site is presented in Figure 13b.

- High grade gliomas were the cause of the greatest proportion of deaths (43.8%), followed by medulloblastoma (14.3%) and ATRT (9.8%).
- Brain stem tumors were the cause of the greatest proportion of deaths (37.9%), followed by cerebellar tumors (16.6%).

#### Five-Year Conditional Survival after Diagnosis by Selected Histologic Groups

Relative survival provides data on cancer prognosis that is useful at a population level, but these numbers may not be informative for individual patients. In the case of individuals that have already survived a year, or several years after diagnosis with their brain tumor, conditional survival estimates provide information about the likelihood that they will survive into the next period of time. Five-vear conditional survival estimates for selected alioma and embryonal sub-types are presented in Figure 14. Five-year conditional survival estimates by age groups for selected brain and CNS tumor subtypes are also presented in Table 15.

- At the time of diagnosis with a high grade glioma, there is a 28.1% probability that a child will live five additional years. For children that have already survived two years after diagnosis, there is an 80.0% chance that they will reach five years of survival.
- At the time of diagnosis with an ATRT, there is a 27.4% probability that a child will live five additional years. For children that have already survived one year after diagnosis, there is a 54.3% chance that they will reach five years of survival.

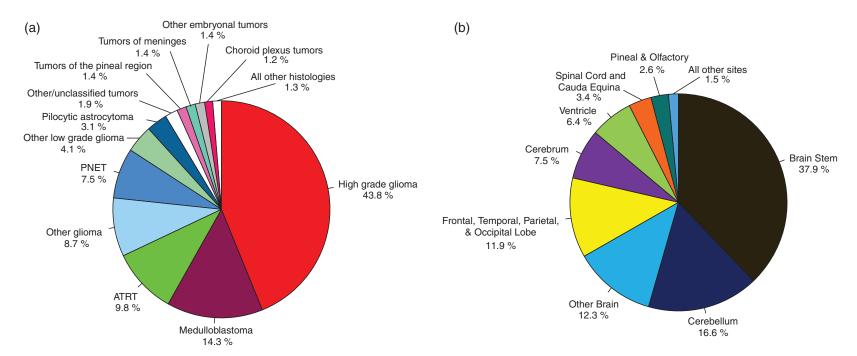
#### Descriptive Summary of Gliomas, and Embryonal Tumors

The data in the CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007-2011 are synthesized to describe the two most common histology groups in infants and children: gliomas and embryonal tumors.

#### Gliomas

- Gliomas are the most common histology group of primary brain and CNS tumor in children 0-14 years (Table 3).
- Gliomas account for 52.9% of all primary brain and CNS tumors in children 0-14 years (Figure 9).
- Pilocytic astrocytoma represent 33.2% of gliomas, followed by other low grade gliomas (27.1%), high grade gliomas (21.0%), and ependymal tumors (10.4%) (Figure 9).
- Incidence of gliomas is highest in New England (3.19 per 100,000), and Middle Atlantic (3.06 per 100,000) (Figure 15).
- Site of tumor varies by age and specific glioma histology. For infants, other low grade gliomas occur most frequently supratentorially (cerebrum, frontal, occipital, temporal, parietal and occipital lobes) and in other nervous system (including the optic nerve). In children 1-4 years, high grade gliomas occur more often in the brain stem (Figure 16).

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**Fig. 13.** Distribution of Total Deaths Due to Primary Malignant Brain and CNS Tumor in Children 0–14 at Time of Death from 2007–2011 by (a) Histology Groupings (N = 734), and (b) Site (N = 734) (SEER 1995–2011)

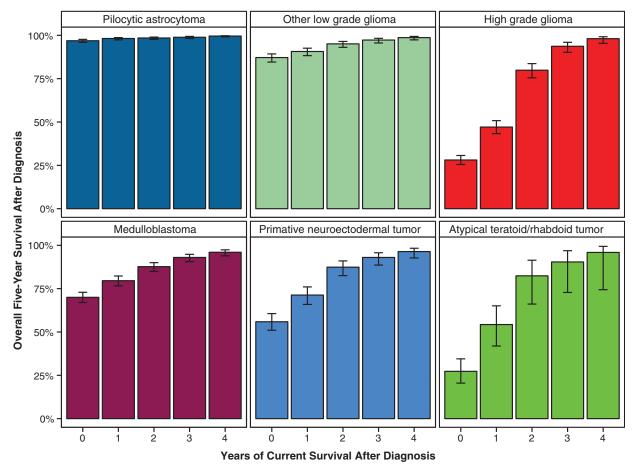


Fig. 14. Five-Year Conditional Survival by Age Groups and Selected Glioma and Embryonal Histology Groupings (SEER 1995 - 2011)

#### **Embryonal Tumors**

- Embryonal tumors are the 2<sup>nd</sup> most common type of primary brain and CNS tumor in children 0-14 years (Table 3).
- Embryonal tumors account for 15.0% of all primary brain and CNS tumors in children 0–14 years (Figure 9).
- Medulloblastomas represent 61.9% of all embryonal tumors, followed by atypical teratoid/rhabdoid tumors (ATRT) (15.0%), primitive neuroectodermal tumors (PNET) (14.9%) (Figure 9).
- Incidence of embryonal tumors is highest in New England (0.85 per 100,000), South Atlantic (0.84 per 100,000), and East North Central (0.81 pre 100,000) (Figure 17).

#### Strengths and Limitations

CBTRUS is the largest population-based registry of primary brain and CNS tumors in the US and covers 99.8% of the U.S. population (for 2011 only, data was available for 50 out of 51 registries). The CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011 contains the most up-to-date population-based data on primary childhood brain tumor and CNS tumors available through the surveillance system in the United States.

The histologic grouping scheme used in this report represents a re-organization from that used in the CBTRUS Statistical Report: Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007-2011<sup>2</sup> in order to better reflect the histologies that are particularly relevant to infant and childhood brain tumors. Comparison of the statistics presented in this report to those included in the overall CBTRUS Report has been affected and may be difficult. Furthermore, the grouping scheme and definition of primary brain tumors used in this report differs from those used by other cancer surveillance organizations. This report includes both malignant and non-malignant tumors, and hematopoietic tumors of the CNS. Other reporting agencies may chose not to include hematopoietic neoplasms, and non-malignant brain and CNS tumors. Additionally many other organizations use the ICCC grouping scheme to report on childhood brain tumor incidence and mortality. In light of these differences, caution should be used if attempting to compare the statistics included in this report to those presented by other organizations. All analyses were undertaken with the overall intention to present meaningful and relevant statistical information to the communities working with infant and childhood brain and CNS tumors.

Registration of individual cases is conducted by cancer registrars at the institution where diagnosis occurs and is then transmitted to the central registry, which further transmits this information to NPCR or SEER. Central cancer registries (both

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**Table 15.** Five-Year Conditional Survival Rates<sup>a,b</sup> for Selected Malignant Brain and Central Nervous System Tumors by Age Groups<sup>c</sup>

				Years	of current survi	val after (	diagnosis				
		At Dio	gnosis	1 year	rs	2 years	5	3 years	i	4 years	;
Histology	Age Group (years)	%	95% CI	%	95% CI	%	95% CI	%	95% CI	%	95% CI
Gliomas	<1	71.2	(65.3 – 76.3)	86.6	(80.9-90.7)	90.4	(85.0-94.0)	93.3	(88.2-96.3)	98.6	(94.4-99.6)
	1-4	80.0	(78.0 - 81.9)	87.2	(85.3-88.9)	94.4	(92.9-95.6)	97.9	(96.8-98.7)	99.0	(98.2-99.5)
	5-9	70.2	(68.0 - 72.3)	84.2	(82.2-86.0)	94.9	(93.4-96.1)	97.2	(96.0-98.1)	98.9	(97.9-99.4)
	10-14	79.1	(76.9 - 81.1)	86.0	(84.0-87.8)	93.3	(91.7-94.7)	96.7	(95.3-97.6)	98.7	(97.7-99.3)
	0-14	76.0	(74.7-77.1)	85.9	(84.8-86.9)	94.0	(93.2-94.8)	97.1	(96.4-97.6)	98.9	(98.4-99.2)
Pilocytic astrocytoma	<1	80.2	(66.8 - 88.6)	85.7	(72.0-93.0)	87.1	(73.4-94.0)	88.7	(74.9-95.2)	97.4	(82.9-99.6)
	1-4	97.5	(95.8-98.5)	98.4	(96.8-99.2)	99.0	(97.6-99.6)	99.4	(98.0-99.8)	100.0	#
	5-9	98.0	(96.4-98.9)	99.2	(97.7-99.7)	99.2	(97.7-99.7)	99.3	(97.9-99.8)	99.3	(97.9-99.8)
	10-14	97.4	(95.5-98.5)	98.4	(96.7-99.3)	98.4	(96.7-99.2)	99.2	(97.6-99.7)	99.8	(98.0-100.0
	0-14	97.1	(96.1-97.8)	98.3	(97.4-98.8)	98.5	(97.7-99.0)	99.0	(98.3-99.4)	99.6	(99.1-99.9)
Other low grade glioma	<1	76.7	(61.6 - 86.5)	84.9	(68.9-93.0)	84.8	(68.9-92.9)	93.5	(76.2-98.3)	96.4	(77.2-99.5)
	1-4	88.8	(83.8-92.4)	93.0	(88.3-95.8)	96.1	(91.8 - 98.1)	98.1	(94.1-99.4)	98.7	(94.7-99.7)
	5-9	88.9	(84.1-92.4)	93.0	(88.5-95.7)	97.2	(93.3-98.8)	97.7	(93.8 - 99.1)	99.4	(95.5-99.9)
	10-14	86.7	(82.3 - 90.0)	88.4	(84.2-91.6)	94.6	(90.9-96.8)	97.1	(93.8-98.6)	98.7	(95.8-99.6)
	0-14	87.3	(84.8-89.4)	90.8	(88.4-92.7)	95.2	(93.2-96.6)	97.3	(95.7-98.4)	98.8	(97.4-99.4)
High grade glioma	<1	54.5	(41.8-65.6)	91.9	(76.7-97.3)	100.0	#	100.0	#	100.0	#
	1-4	27.1	(22.0 - 32.6)	42.3	(34.7-49.6)	78.8	(68.4-86.1)	96.8	(87.5-99.2)	98.2	(88.2-99.8)
	5-9	20.3	(16.9 - 23.9)	39.7	(33.7-45.7)	78.3	(69.7-84.7)	92.4	(84.6-96.3)	98.7	(91.0-99.8)
	10-14	36.5	(31.3 - 41.7)	52.7	(45.9-59.0)	78.3	(70.5 - 84.3)	92.0	(85.0-95.8)	97.1	(91.0 - 99.1)
	0-14	28.1	(25.6-30.7)	47.1	(43.3-50.8)	80.0	(75.5-83.7)	93.8	(90.3-96.1)	98.1	(95.4-99.2)
Ependymal tumors	<1	59.9	(43.5 - 72.9)	77.1	(57.6-88.4)	84.8	(64.4-94.0)	84.8	(64.4-94.0)	100.0	#
	1-4	71.3	(65.0 - 76.7)	76.1	(69.7-81.3)	81.5	(75.1-86.4)	91.6	(85.9-95.0)	95.7	(90.7 - 98.1)
	5-9	73.7	(65.3 - 80.4)	76.5	(68.0-83.0)	85.1	(76.7-90.6)	88.1	(79.9-93.1)	94.3	(86.7-97.6)
	10-14	79.2	(69.9-85.9)	80.5	(71.3-87.1)	85.1	(75.9-90.9)	88.6	(79.6 - 93.7)	94.8	(86.5-98.0)
	0-14	72.7	(68.6-76.3)	77.3	(73.2-80.9)	83.5	(79.5-86.8)	89.5	(85.9-92.2)	95.4	(92.4-97.2)
Embryonal tumors	<1	36.6	(29.8 - 43.4)	68.3	(57.8-76.7)	86.5	(75.6 - 92.8)	90.3	(79.5-95.5)	98.1	(87.1-99.7)
	1-4	55.8	(51.9 - 59.4)	72.4	(68.2 – 76.2)	87.8	(84.0-90.8)	93.7	(90.3-95.9)	96.2	(93.2-97.9)
	5-9	73.2	(69.2 – 76.8)	80.2	(76.2-83.6)	89.0	(85.4-91.7)	94.9	(91.9-96.8)	97.4	(94.9-98.7)
	10-14	73.8	(67.8 - 78.8)	79.5	(73.5-84.3)	86.1	(80.2 – 90.3)	89.3	(83.7-93.1)	93.2	(88.0-96.2)
	0-14	62.1	(59.7-64.4)	76.2	(73.8-78.5)	87.8	(85.6-89.7)	93.0	(91.1-94.5)	96.2	(94.6-97.3)
Medulloblastoma	<1	42.0	(29.0-54.6)	73.1	(52.7-85.8)	86.1	(62.6-95.3)	90.1	(65.9 – 97.4)	_	_
	1-4	62.1	(56.5 – 67.1)	74.0	(68.2 – 78.9)	86.8	(81.2 – 90.8)	93.1	(88.0-96.0)	96.1	(91.6-98.3)
	5-9	75.8	(71.2 – 79.9)	82.3	(77.7-86.0)	88.8	(84.5-91.9)	94.9	(91.3-97.0)	97.4	(94.4-98.8)
	10-14	80.6	(73.6-85.9)	84.1	(77.2-89.1)	87.3	(80.5-91.9)	90.0	(83.3 – 94.1)	92.6	(86.2-96.1)
	0-14	70.1	(67.1-72.9)	79.6	(76.6-82.3)	87.8	(85.0-90.1)	93.0	(90.6-94.9)	96.0	(93.9-97.4)

Quinn T. Ostrom: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain

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Primitive neuroectodermal tumor	<1	33.2	(19.5-47.6)	-	-	-	-	-	-	-	-
	1-4	54.3	(46.9-61.1)	69.7	(61.4-76.5)	87.1	(79.1-92.2)	93.7	(86.4-97.1)	95.6	(88.6 - 98.3)
	5-9	67.2	(57.6-75.1)	75.5	(65.6 - 82.9)	88.5	(79.0 - 93.9)	94.2	(85.2 - 97.8)	96.9	(88.1 - 99.2)
	10-14	55.5	(42.7 - 66.5)	65.7	(51.5 – 76.7)	83.0	(67.5 - 91.6)	87.0	(71.5-94.4)	96.9	(80.0 - 99.6)
	0-14	56.0	(51.0-60.7)	71.4	(66.0-76.1)	87.5	(82.5-91.1)	93.0	(88.7-95.8)	96.5	(92.7-98.3)
Atypical teratoid/rhabdoid tumor	<1	12.2	(5.0-22.7)	30.1	(12.2 - 50.3)	-	-	_	-	-	-
	1-4	33.9	(24.3 - 43.7)	66.2	(49.3 – 78.6)	90.9	(66.9 - 97.8)	94.1	(64.9-99.2)	94.1	(65.0 - 99.2)
	5-9	_	_	_	_	_	_	_	-	_	_
	10-14	_	_	_	_	_	_	_	_	_	_
	0-14	27.4	(20.6-34.6)	54.3	(42.0-65.1)	82.5	(66.1-91.4)	90.5	(72.8-96.9)	95.9	(74.3-99.4)
Germ cell tumors	<1	-	-	_	-	-	_	_	-	-	_
	4 /										
	1-4	-	-	-	-	-	-	_	-	-	-
	1-4 5-9	- 90.4	- (79.1-95.8)	- 92.4	- (80.4-97.1)	93.7	- (81.3-97.9)	93.6	- (81.3-97.9)	- 97.4	- (83.1-99.6)
			- (79.1 - 95.8) (84.6 - 93.0)	- 92.4 92.3	- (80.4-97.1) (87.5-95.2)	93.7 94.8	- (81.3 - 97.9) (90.4 - 97.2)	93.6 96.7	- (81.3 – 97.9) (92.6 – 98.5)	- 97.4 98.2	- (83.1 – 99.6) (94.4 – 99.5)
	5-9	90.4	,		,		,		•		,
TOTAL: All Brain and Other Nervous System <sup>d</sup>	5-9 10-14	90.4 89.6	(84.6-93.0)	92.3	(87.5-95.2)	94.8	(90.4-97.2)	96.7	(92.6-98.5)	98.2	(94.4-99.5)
TOTAL: All Brain and Other Nervous System <sup>d</sup>	5-9 10-14 <b>0-14</b>	90.4 89.6 <b>86.8</b>	(84.6-93.0) <b>(82.6-90.1)</b>	92.3 <b>92.9</b>	(87.5-95.2) <b>(89.1-95.4)</b>	94.8 <b>95.0</b>	(90.4-97.2) <b>(91.4-97.1)</b>	96.7 <b>96.3</b>	(92.6-98.5) <b>(92.9-98.1)</b>	98.2 <b>98.2</b>	(94.4-99.5) <b>(95.2-99.3)</b>
TOTAL: All Brain and Other Nervous System <sup>d</sup>	5-9 10-14 <b>0-14</b> <1	90.4 89.6 <b>86.8</b> 55.5	(84.6-93.0) ( <b>82.6-90.1</b> ) (51.2-59.5)	92.3 <b>92.9</b> 81.1	(87.5-95.2) ( <b>89.1-95.4</b> ) (76.5-84.9)	94.8 <b>95.0</b> 89.5	(90.4-97.2) (91.4-97.1) (85.2-92.6)	96.7 <b>96.3</b> 93.1	(92.6-98.5) (92.9-98.1) (89.2-95.6)	98.2 <b>98.2</b> 98.2	(94.4-99.5) ( <b>95.2-99.3</b> ) (95.2-99.3)
TOTAL: All Brain and Other Nervous System <sup>d</sup>	5-9 10-14 <b>0-14</b> <1 1-4 5-9	90.4 89.6 <b>86.8</b> 55.5 71.9 71.5	(84.6-93.0) (82.6-90.1) (51.2-59.5) (70.1-73.7) (69.6-73.2)	92.3 <b>92.9</b> 81.1 82.9 83.1	(87.5-95.2) (89.1-95.4) (76.5-84.9) (81.1-84.5) (81.4-84.7)	94.8 <b>95.0</b> 89.5 92.4 92.9	(90.4-97.2) <b>(91.4-97.1)</b> (85.2-92.6) (91.0-93.6) (91.5-94.1)	96.7 <b>96.3</b> 93.1 96.8 96.2	(92.6-98.5) (92.9-98.1) (89.2-95.6) (95.7-97.6) (95.1-97.1)	98.2 98.2 98.2 98.4 98.3	(94.4-99.5) (95.2-99.3) (95.2-99.3) (97.5-98.9) (97.4-98.9)
TOTAL: All Brain and Other Nervous System <sup>d</sup>	5-9 10-14 <b>0-14</b> <1 1-4	90.4 89.6 <b>86.8</b> 55.5 71.9	(84.6-93.0) ( <b>82.6-90.1</b> ) (51.2-59.5) (70.1-73.7)	92.3 <b>92.9</b> 81.1 82.9	(87.5-95.2) ( <b>89.1-95.4</b> ) (76.5-84.9) (81.1-84.5)	94.8 <b>95.0</b> 89.5 92.4	(90.4-97.2) (91.4-97.1) (85.2-92.6) (91.0-93.6)	96.7 <b>96.3</b> 93.1 96.8	(92.6-98.5) (92.9-98.1) (89.2-95.6) (95.7-97.6)	98.2 <b>98.2</b> 98.2 98.4	(94.4-99.5) ( <b>95.2-99.3</b> ) (95.2-99.3) (97.5-98.9)

<sup>&</sup>lt;sup>a</sup>The cohort analysis of survival rates was utilized for calculating the survival estimates presented in this table. Long-term cohort-based survival estimates reflect the survival experience of individuals diagnosed over the time period, and they may not necessarily reflect the long-term survival outlook of newly diagnosed cases.

Abbreviation: CBTRUS, Central Brain Tumor Registry of the United States; SEER, Survival, Epidemiology and End Results; CI, confidence interval; NOS, not otherwise specified.

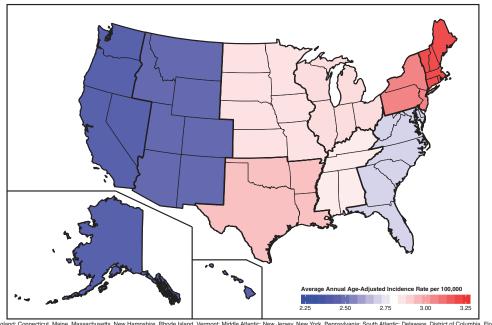
<sup>&</sup>lt;sup>b</sup>Rates are an estimate of the percentage of patients alive at one, two, five, and ten year, respectively. Rates were not presented for categories with 50 or less cases and were suppressed for rates where less than 16 cases were surviving within a category.

<sup>&</sup>lt;sup>c</sup>Estimated by CBTRUS using Surveillance, Epidemiology, and End Results (SEER) Program (www.seer.cancer.gov) SEER\*Stat Database: Incidence - SEER 18 Regs Research Data + Hurricane Katrina Impacted Louisiana Cases, Nov 2013 Sub (1973–2011 varying) - Linked To County Attributes - Total U.S., 1969–2012 Counties, National Cancer Institute, DCCPS, Surveillance Research Program, Cancer Statistics Branch, released April 2014, based on the November 2013 submission.

<sup>d</sup>Includes histologies not listed in this table.

<sup>-</sup> Rates are excluded when calculated based on a population of less than 50, or when less than 16 remain alive in the survival period.

<sup>#</sup> Statistic could not be calcualted



Georgia, Maryland, North Central: Iowa, Kansas, Minnesota, West Virginia; East North Central: Illinois, Indiana, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, Suth Alabama, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, Suth Alabama, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, Suth Alabama, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Mississippi, Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Mississippi, Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Mississippi, Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Mississippi, Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Mississippi, Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Alabama, Mississippi, Tennessee; West North Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Alabama, Mississippi, Tennessee; West North Central: Alabama, Mississippi, Tennessee; West North Central: Alabama, Mississippi, Tennessee; West North Central: Alabama,

Fig. 15. Average Annual Age-Adjusted Incidence Rates of All Gliomas by Region of the United States (0-14 Years) (N = 8,487) (CBTRUS 2007-2011)

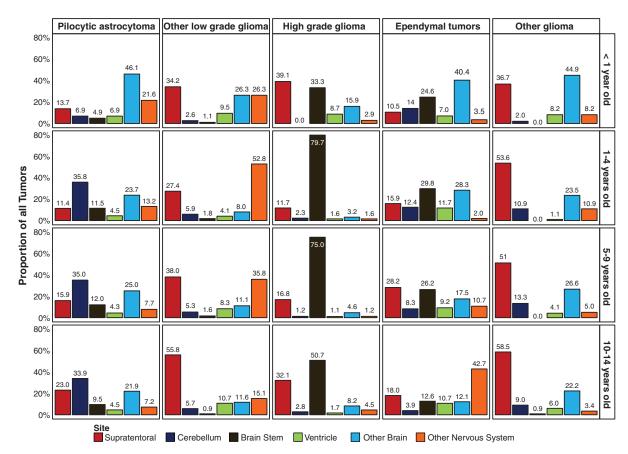
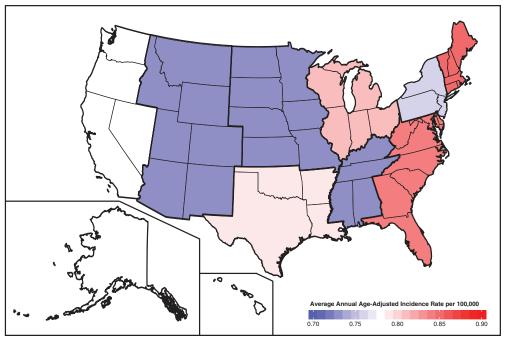


Fig. 16. Number and Distribution of Sites by Specific Glioma Histology Groupings and Age Groups (N = 8,487) (CBTRUS 2007 - 2011)



New England: Connecticut, Maine, Massachusetts, New Hampshire, Rhode Island, Vermont; Middle Atlantic: New Jersey, New York, Pennsylvania; South Atlantic: Delaware, District of Columbia, Florida, Georgia, Maryland, North Carolina, South Carolina, Virginia, West Virginia; East North Central: Illinois, Indiana, Michigan, Ohio, Wisconsin; East South Central: Alabama, Kentucky, Mississippi, Tennessee; West North Central: Iowa, Kansas, Minnesota, Missouri, Nebraska, North Dakota, South Dakota; West South Central: Arkansas, Louisiana, Oklahoma, Texas; Mountaim: Arizona, Colorado, Idaho, Montana, New Mexico, Utah, Wyomino: Pacific: Alabaca, California, Hawaii, Newada, Oregon, Washindton.

Fig. 17. Average Annual Age-Adjusted Incidence Rates of All Embryonal Tumors by Region of the United States (0-14 Years) (N = 2,413) (CBTRUS 2007-2011)

NPCR and SEER) only report cases to the CDC and NCI for persons that are residents of that particular state, so duplicate records should not occur for persons that may have traveled across state lines for treatment. No mechanism exists for central pathology review of cases, and registration is based on histology information contained in the patient's medical record.

The SEER 18 population dataset used for the survival analyses is a subset of the larger CBTRUS dataset and only covers approximately 26% of the US population as compared to the 99.8% population coverage of the larger dataset. Survival estimates obtained from this dataset may be less reliable as representations of 'real' relative survival rates for the US than if they were based on data from a larger portion of the population.

#### **Concluding Comment**

The CBTRUS Statistical Report: Alex's Lemonade Stand Foundation Infant and Childhood Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2007–2011 comprehensively describes the current population-based incidence of primary malignant and non-malignant brain and CNS tumors in children ages 0–14 years, collected and reported by central cancer registries covering approximately 99.8% of the United States population (for 2011 only, data were available for 50 out of 51 registries). Overall, brain and CNS tumors are the most common solid tumor, the most common cancer, and the most common cause of cancer death in infants and children 0–14 years. This report aims to serve as a useful resource for researchers, clinicians, patients, and families.

#### **Abbreviations**

AIAN – American Indian/Alaskan Native

API – Asian/Pacific Islander

AYA – Adolescents and Young Adults ATRT – Atypical Teratoid/Rhabdoid Tumor

CBTRUS - Central Brain Tumor Registry of the United States

CDC – Centers for Disease Control and Prevention

CSS – Cancer Surveillance System

CI – Confidence interval CNS – Central nervous system

ICD-O-3 - International Classification of Diseases for Oncology,

Third Edition

ICCC – International Classification of Childhood Cancer

NAACCR - North American Association of Central Cancer

Registries

NCDB – National Cancer Data Base

NCHS - National Center for Health Statistics

NCI – National Cancer Institute NOS – Not otherwise specified

NPCR – National Program of Cancer Registries PNET – Primitive Neuroectodermal Tumor

SEER - Surveillance, Epidemiology and End Results

USCS – United States Cancer Statistics WHO – World Health Organization

#### **Acknowledgments**

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Appendix A. Average Annual Populations<sup>a</sup> for 2007–2011, By Age, Gender and Race

Age Group	White	Black	AIAN	API	Total
Male					
<1 year	1,554,737	348,286	38,661	119,032	2,060,715
1-4 years	6,256,217	1,358,815	150,358	470,874	8,236,265
5-9 years	7,849,153	1,655,085	180,670	562,182	10,247,090
10-14 years	8,107,511	1,744,870	183,262	547,649	10,583,292
Total	23,767,618	5,107,057	552,951	1,699,737	31,127,362
Female					
<1 year	1,484,983	336,915	37,926	112,585	1,972,409
1-4 years	5,970,115	1,313,813	145,785	455,644	7,885,358
5-9 years	7,478,677	1,601,608	175,695	559,237	9,815,218
10-14 years	7,701,865	1,684,402	179,050	534,779	10,100,096
Total	22,635,640	4,936,739	538,457	1,662,246	29,773,081

<sup>&</sup>lt;sup>a</sup>Population data source for 51 population-based geographic regions: Estimates from the United States. Bureau of the Census http://seer.cancer.gov/popdata/index.html.

**Appendix B.** Average Annual Populations<sup>a</sup> for 2007–2011, by Age, Gender, and Hispanic Ethnicity

Age Group	Non-Hispanic	Hispanic	Total
Male			
<1 year	1,531,632	529,084	2,060,715
1-4 years	6,190,889	2,045,376	8,236,265
5-9 years	7,902,989	2,344,101	10,247,090
10-14 years	8,331,023	2,252,269	10,583,292
Total	23,956,533	7,170,829	31,127,362
Female			
<1 year	1,462,286	510,124	1,972,409
1-4 years	5,923,047	1,962,311	7,885,358
5-9 years	7,565,538	2,249,680	9,815,218
10-14 years	7,942,964	2,157,132	10,100,096
Total	22,893,835	6,879,246	29,773,081
-			

<sup>&</sup>lt;sup>a</sup>Population data source for 51 population-based geographic regions: Estimates from the U.S. Census Bureau http://seer.cancer.gov/popdata/index. html.

Abbreviations: AIAN, American Indian Alaskan Native; API, Asian Pacific Islander.