Commentary

A System for Classifying Cancers Diagnosed in Adolescents and Young Adults

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INTRODUCTION

The discipline of adolescent and young adult (AYA) oncology is advancing rapidly, making it timely to offer a new system for classifying cancers in AYAs. Neoplasms in AYAs constitute a different array of types from those in children and older adults, and many cancers that are histologically and anatomically similar in AYAs to those in other age groups are different biologically.

Information on topography (anatomic location) and morphology (histology and behavior) is collected and coded by cancer registries according to the International Classification of Diseases for Oncology (ICD-O), which was first published in 1976[1] and now is in its third edition (ICD-O-3).[2] A new ICD-O-3.2[3] was released in April 2019 for implementation in 2020. The topography codes range from C00.0 to C80.9, and the morphology codes from 8000/0 to 9993/3. The first 4 digits of the morphology code indicate the specific histology, and the digit after the slash indicates the behavior (3, malignant; 2, in situ; 1, borderline or uncertain; and 0, benign).

Morphology is more useful than anatomic site for classifying cancers in children and AYA cancer classification benefits likewise, as undertaken by Birch and colleagues for AYAs aged 15 to 24 years in 2002.[4] On the basis of their scheme, a classification was developed in 2006 for AYAs aged 15 to 29 inclusive.[5] By raising the upper age limit from 29 to 39 years, as was recommended at a consensus conference in the United States,[6] the array of cancer types contrasts markedly with that in childhood[7]; notably, the proportion of all cases in AYAs aged 15 to 39 years that are diagnosed as carcinomas (42%) is greater than that in AYAs aged 15 to 29 years (32%)[8] and much greater than that in children (3.5%).[9]

Simultaneously, there has been an evolving characterization of the biology of cancers in AYAs,[10] and further insights have been provided by recent World Health Organization (WHO) Blue Books together with the ICD-O-3.2. In this report, AYA refers to the group aged 15 to 39 years inclusive, unless otherwise specified.

This new classification is the outcome of international conferences, engaging AYA oncologists, other clinicians, epidemiologists, pathologists, and molecular and cellular biologists, combined with expertise in analysis and classification of data from international population-based cancer registries.

The previous classification[5] was expanded on the bases of age, disease distribution, new diagnostic criteria, and biologic considerations. Surveillance, Epidemiology, and End Results (SEER) program registry data from 2004 to 2017 were used to illustrate the proposed classification by 5-year age groups compared to individuals aged <15 years and those aged ≥40 years. The SEER registry then represented 28.5% of the total AYA population in the United States. These analyses include data from 18 SEER registries selected to represent the country's geography and racial/ethnic mix.[11] The SEER data were accessed using SEER*Stat software (version 8.3.6). The total numbers of cases in the groups aged 0 to 14, 15 to 29, 15 to 39, and ≥40 years are 45,860, 123,851, 353,269, and 6,159,548, respectively.

In addition to 11 main categories containing all invasive cases, together with benign and borderline tumors of the central nervous system (CNS), the new classification also includes a supplementary category containing in situ tumors

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TABLE 1. Adolescent and Young Adult Classification by Main Categories and Selected Age Groups: Frequencies and Rates Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017[a]

	Age at Diagnosis, y									
Classification	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
Frequency: Total counts										
1. Leukemias and related disorders	13,365	2986	2768	3116	3824	4869	241,888	272,816	8870	17,563
2. Lymphomas	4989	4346	5942	6568	7332	9028	329,515	367,720	16,856	33,216
3. CNS and other intracranial and intraspinal neoplasms	11,721	4782	5781	7789	9748	11,545	205,337	256,703	18,352	39,645
4. Sarcomas	4331	2546	2222	2381	2813	3426	64,715	82,434	7149	13,388
5. Blood and lymphatic vessel tumors	350	293	527	963	1293	1374	13,068	17,868	1783	4450
6. Nerve sheath tumors	852	463	615	832	1179	1632	22,311	27,884	1910	4721
7. Gonadal and related tumors	1561	2614	5810	7687	7557	6888	79,356	111,473	16,111	30,556
8. Melanoma, malignant	499	1010	3102	5712	8195	10,845	261,119	290,482	9824	28,864
9. Carcinomas[b]	1678	3859	9420	21,312	42,023	73,225	4,256,805	4,408,322	34,591	149,839
10. Miscellaneous specified neoplasms	6330	133	107	159	234	379	24,869	32,211	399	1012
11. Unspecified malignant neoplasms except CNS	58	46	187	278	446	748	165,937	167,700	511	1705
A. In situ	126	655	2437	4403	7520	13,295	494,628	523,064	7495	28,310
Totals										
All sites (malignant/invasive)	41,020	19,466	31,837	50,304	76,127	113,535	5,493,717	5,826,006	101,607	291,269
CNS (benign/borderline)	4714	3612	4644	6493	8517	10,424	171,203	209,607	14,749	33,690
All sites (malignant/invasive) and CNS (benign/borderline)	45,734	23,078	36,481	56,797	84,644	123,959	5,664,920	6,035,613	116,356	324,959
Grand total: All sites (invasive and in situ) and CNS (benign/borderline)	45,860	23,733	38,918	61,200	92,164	137,254	6,159,548	6,558,677	123,851	353,269
Incidence, rate per 100,000[c]										
1. Leukemias and related disorders	5.45	3.51	3.21	3.64	4.63	5.92	45.23	22.14	3.46	4.25
2. Lymphomas	2.05	5.11	6.90	7.68	8.87	10.98	60.92	29.50	6.51	8.00
3. CNS and other intracranial and intraspinal neoplasms	4.80	5.62	6.71	9.11	11.80	14.04	37.76	20.69	7.09	9.61
4. Sarcomas	1.77	2.99	2.58	2.79	3.40	4.17	11.90	6.65	2.79	3.23
5. Blood and lymphatic vessel tumors	0.14	0.34	0.61	1.13	1.56	1.67	2.44	1.47	0.68	1.08
6. Nerve sheath tumors	0.35	0.54	0.71	0.97	1.43	1.98	4.01	2.21	0.74	1.16
7. Gonadal and related tumors	0.64	3.07	6.74	8.99	9.14	8.38	14.56	8.98	6.15	7.26
8. Melanoma, malignant	0.20	1.19	3.60	6.68	9.92	13.19	47.87	23.17	3.72	7.12
9. Carcinomas[b]	0.69	4.54	10.93	24.93	50.85	89.05	775.10	347.15	13.11	37.94
10. Miscellaneous specified neoplasms	2.55	0.16	0.12	0.19	0.28	0.46	4.60	2.62	0.16	0.25
11. Unspecified malignant neoplasms except CNS	0.02	0.05	0.22	0.33	0.54	0.91	31.08	13.53	0.19	0.43
A. In situ	0.05	0.77	2.83	5.15	9.10	16.17	90.18	41.35	2.83	7.12
Totals										
All sites (malignant/invasive)	16.73	22.88	36.95	58.84	92.12	138.08	1,004.03	461.25	38.91	72.12
CNS (benign/borderline)	1.93	4.25	5.39	7.59	10.31	12.68	31.45	16.86	5.68	8.20
All sites (malignant/invasive) and CNS (benign/borderline)	18.66	27.12	42.34	66.43	102.43	150.75	1035.48	478.11	44.59	80.32
Grand total: All sites (invasive and in situ) and CNS (benign/borderline)	18.72	27.89	45.17	71.58	111.53	166.92	1125.66	519.45	47.42	87.44
Unclassified (excluded from analysis), N	0	0	1	0	3	1	15	20	1	5

Abbreviation: CNS, central nervous system.

[a]Values are based on data from the Surveillance, Epidemiology, and End Results Program, National Cancer Institute, November 2019 submission.

[b]This category excludes in situ carcinomas.

[c]Rates are per 100,000 and age-adjusted to the 2000 United States Standard Population[12] (19 age groups – Census P25-1130).

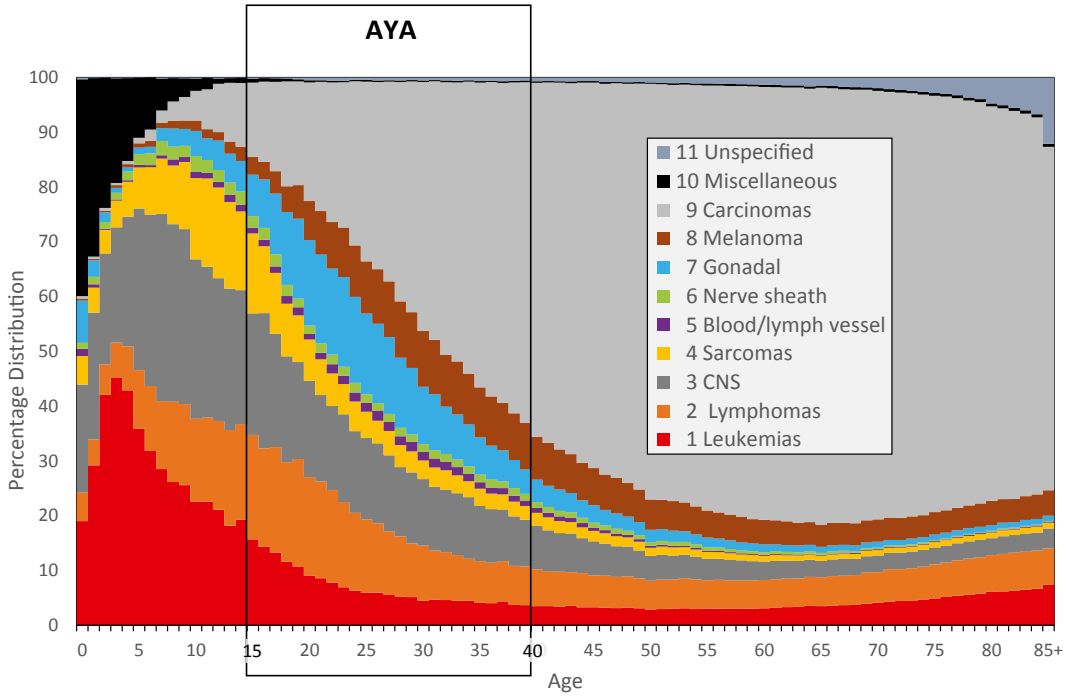

FIGURE 1. The percentage distribution of adolescent and young adult (AYA) classification categories 1 through 11 (excluding in situ) is illustrated by single year of age from infants to age ≥85 years (US Surveillance, Epidemiology, and End Results Program 18 areas, 2004-2017). CNS indicates central nervous system.

(Table 1).[12] Further stratifications are provided in subsequent tables. Gonadal tumors are separated into testicular and ovarian primary site and further stratified into germ cell and non-germ cell tumors. Separate categories have been created for more primary sites of carcinomas than in the previous classification, and there are specific subcategories for neuroendocrine tumors in several locations.

The variation in types of tumors as a function of patient age is striking. A shift among the major categories occurs across the age spectrum (Fig. 1). Within the AYA range, the percentages of cancers accounted for by leukemias, lymphomas, CNS tumors, and sarcomas all decrease with age. The percentage that are carcinomas increases with age throughout the range, and there is a marked increase in the percentage that are melanomas until around age 25 years. Uniquely, the highest percentage of cancers that are gonadal and related tumors occurs in the middle of the AYA age range.

The detailed definitions for each of the main categories and subcategories are presented in Tables 2 through 8. To be applicable to data coded according to the ICD-O-3 and subsequent revisions, all eligible histology codes from ICD-O-3, ICD-O-3.1, and ICD-O-3.2 have been included. When the morphology (histology) code is present and the topography (primary site) code is absent in the tables, the latter includes all primary sites (codes C00.0-C80.9).

Specific categories were created for entities representing a higher proportion of all cancers in AYAs than in children and/or adults in Tables 9 through 19. These include:

- Acute promyelocytic leukemia, other forms of acute myeloid leukemia, chronic myeloid leukemia, and Hodgkin lymphoma (Table 9);
- Noninvasive pituitary tumors (Table 10);
- Kaposi and synovial cell sarcomas, hemangioblastoma, tufted hemangioma, and malignant peripheral nerve sheath tumors outside the CNS (Table 11);
- Seminoma, embryonal carcinoma and mixed germ cell tumors of the testis, and superficial spreading melanoma (Table 12);
- The primary sites of the carcinomas (Table 13);
- Infiltrating duct carcinoma of the breast and squamous carcinoma of the uterine cervix (Table 17); and
- Paraganglioma outside the CNS (Table 19).

Compared with the previous classification[5] used for recent reports on incidence[13] and survival,[14] the system proposed here groups bone and soft-tissue sarcomas

TABLE 2. Categories 1 (Leukemias) and 2 (Lymphomas): Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology	ICD-O-3 Primary Site	Behavior[a]
1. Leukemias and related disorders			
1.1 Acute lymphoblastic leukemia	9826, 9835-9836		3
	9687, 9811-9819, 9827, 9837	C42.0-C42.1, C42.4	3
1.2 Acute myeloid leukemia			3
1.2.1 Acute promyelocytic leukemia	9866		3
1.2.2 Other acute myeloid leukemia	9840, 9861, 9865, 9867, 9869-9874, 9877-9879, 9891, 9895-9898, 9910-9912, 9920, 9931		3
1.3 Chronic myeloid leukemia	9863, 9875-9876, 9945-9946		3
1.4 Chronic lymphocytic leukemia	9823	C42.0-C42.1, C42.4	3
1.5 Polycythemia vera	9950		3
1.6 Essential thrombocythemia	9962		3
1.7 Primary myelofibrosis	9961		3
1.8 Myelodysplastic syndrome (MDS)	9980, 9982-9989, 9991-9993		3
1.9 Other and unspecified leukemias and related disorders			
1.9.1 Hairy cell leukemia	9940		3
1.9.2 Other lymphocytic/lymphoblastic leukemias	9820, 9831-9834, 9948		3
1.9.3 Other myeloid leukemias	9860, 9930, 9963-9964		3
1.9.4 Leukemias of mixed phenotype	9805-9809		3
1.9.5 Mast cell diseases	9740-9742		3
1.9.6 Other	9800-9801, 9960, 9965-9968, 9975		3
2. Lymphomas			
2.1 Non-Hodgkin lymphomas			
2.1.1 Lymphoblastic	9727-9729		3
	9811-9819, 9827, 9837	C00.0-C41.9,C42.2-C42.3,C44.0-C80.9	3
2.1.2 Burkitt	9687	C00.0-C41.9, C42.2-C42.3, C44.0-C80.9	3
2.1.3 Diffuse large B-cell (DLBCL)	9596, 9673, 9680, 9684, 9688-9689, 9712, 9737-9738		3
2.1.4 Primary mediastinal large B-cell excluded from DLBCL	9679		3
2.1.5 Anaplastic T-cell and null-cell excluding NK/T-cell	9700, 9702, 9705, 9708-9709, 9714-9718		3
2.1.6 Follicular	9690-9691, 9695, 9698		3
2.1.7 NK/T-cell (excluded from anaplastic T-cell)	9719		3
2.1.8 MALT (Mucosa-associated lymphoid tissue)	9699		3
2.1.9 Other non-Hodgkin lymphoma NOS	9591, 9670, 9971		3
	9823	C00.0-C41.9,C42.2-C42.3,C44.0-C80.9	3
2.2 Hodgkin lymphoma			
2.2.1 Hodgkin NLP	9659		3
2.2.2 Hodgkin classic, other	9650-9655, 9661-9665, 9667		3
2.3 Myeloma	9731-9734		3
2.4 Cutaneous lymphomas	9597, 9701, 9725, 9726		3
2.5 Other B-cell and T-cell lymphomas	9671, 9675, 9678, 9724, 9735		3
2.6 Other lymphomas, specified and unspecified			
2.6.1 Histiocytic and dendritic cell neoplasms	9749-9759		3
2.6.2 Lymphoma NOS	9590, 9760-9762, 9764-9769, 9970		3

Abbreviations: ICD-O-3, International Classification of Diseases for Oncology, Third Edition; NK, natural killer cell; NLP, nodular lymphocyte-predominant; NOS, not otherwise specified; PMLCL, primary mediastinal large B-cell lymphoma.

[a]A behavior code of 3 indicates malignant.

TABLE 3. Category 3 (Central Nervous System and Other Intracranial and Intraspinal Neoplasms): Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology	ICD-O-3 Primary Site	Behavior[a]
3. CNS and other intracranial and intraspinal neoplasms[b]			
3.1 Astroglial and related neoplasms			
3.1.1 Oligodendriogliomas			
3.1.1.1 Oligodendrioglioma, benign/borderline	9382, 9385, 9450-9451, 9460		0, 1
3.1.1.2 Oligodendrioglioma, invasive	9382, 9385, 9450-9451, 9460		3
3.1.2 Glioblastomas/gliofibromas			
3.1.2.1 Gliofibroma, benign/borderline	9440-9442, 9445		0, 1
3.1.2.2 Glioblastoma, invasive	9440-9442, 9445		3
3.1.3 Ependymomas			
3.1.3.1 Ependymoma, benign/borderline	9383, 9391-9394, 9396		0, 1
3.1.3.2 Ependymoma, invasive	9383, 9391-9394, 9396		3
3.1.4 Other astrocytoma/astroglial neoplasms			
3.1.4.1 Pilocytic astrocytoma	9421		0, 1, 3
3.1.4.2 Other astrocytoma/astroglial, benign/borderline	9380-9381, 9384, 9400-9401, 9410-9412, 9420, 9424-9425, 9431-9432		0, 1
3.1.4.3 Other astrocytoma/astroglial, invasive	9380-9381, 9384, 9400-9401, 9410-9412, 9420, 9424-9425, 9431-9432		3
3.2 Medulloblastoma and other invasive embryonal CNS tumors	9260, 9364-9365, 9470-9478, 9480, 9508	C70.0-C72.9, C75.1-C75.3	3
3.3 Neuroblastomas/ganglioneuromas			
3.3.1 Ganglioneuroma, benign/borderline	9490, 9500, 9523	C70.0-C72.9, C75.1-C75.3	0, 1
	9523	C30.0	0, 1
3.3.2 Neuroblastoma/ganglioneuroblastoma, invasive	9490, 9500, 9523	C70.0-C72.9, C75.1-C75.3	3
	9523	C30.0	3
3.4 Neuronal and mixed neuronal-glial neoplasms			
3.4.1 Neuronal and mixed neuronal-glial, benign/borderline	8680-8693, 9413, 9492-9493, 9505-9506, 9509	C70.0-C72.9, C75.1-C75.3	0, 1
3.4.2 Neuronal and mixed neuronal-glial, invasive	8680-8693, 9413, 9492-9493, 9505-9506, 9509	C70.0-C72.9, C75.1-C75.3	3
3.5 Meningiomas			
3.5.1 Meningioma, benign/borderline	9530-9539		0, 1
3.5.2 Meningioma, invasive	9530-9539		3
3.6 Choroid plexus neoplasms			
3.6.1 Choroid plexus, benign/borderline	9390	C70.0-C72.9, C75.1-C75.3	0, 1
3.6.2 Choroid plexus, invasive	9390	C70.0-C72.9, C75.1-C75.3	3
3.7 Craniopharyngiomas			
3.7.1 Craniopharyngioma, benign/borderline	9350-9352	C70.0-C72.9, C75.1-C75.3	0, 1
3.7.2 Craniopharyngioma, invasive	9350-9352	C70.0-C72.9, C75.1-C75.3	3
3.8 Pituitary neoplasms			
3.8.1 Pituitary, benign/borderline	8040, 8140, 8146, 8246, 8260, 8270-8273, 8280-8281, 8290, 8300, 8310, 8323, 9582	C70.0-C72.9, C75.2	0, 1
	8000-8679, 8694-8709, 8800-8814, 8816-9059, 9110, 9139-9140, 9180-9342, 9363-9379, 9386-9389, 9395, 9421-9422, 9443, 9470-9477, 9479-9480, 9491, 9501-9504, 9507, 9510-9522, 9572-9993	C75.1	0,1
3.8.2 Pituitary, invasive	8040, 8140, 8146, 8246, 8260, 8270-8273, 8280-8281, 8290, 8300, 8310, 8323, 9582	C70.0-C72.9, C75.2	3
	8000-8361, 8380-8589, 8680-8693, 8936, 8940, 8941, 8963, 9050-9055, 9363, 9492, 9501-9506, 9582	C75.1	3

TABLE 3. *Continued*

Category	ICD-O-3 Histology	ICD-O-3 Primary Site	Behavior[a]
3.9 Pineal neoplasms			
3.9.1 Pineal, benign/borderline	8000-8679, 8694-8709, 8800-8814, 8816-9059, 9110, 9139-9140, 9180-9342, 9360-9379, 9386-9389, 9395, 9421-9422, 9443, 9470-9477, 9479-9480, 9491, 9501-9504, 9507, 9510-9522, 9572-9993	C75.3	0, 1
	9360-9362	C70.0-C72.9, C75.1-C75.2	0, 1
3.9.2 Pineal, invasive	8000-8709, 8714-8719, 8827, 8834, 8898, 8905, 8930-8960, 8964-8981, 9000-9039, 9045-9055, 9110-9119, 9192-9200, 9241, 9262, 9350-9363, 9373, 9395, 9490-9492, 9500-9507, 9510-9523, 9582	C75.3	3
	9360-9362	C70.0-C72.9, C75.1-C75.2	3
3.10 Other and unspecified CNS neoplasms			
3.10.1 Other and unspecified CNS, benign/borderline	8000-8039, 8041-8139, 8141-8145, 8147-8245, 8247-8259, 8261-8265, 8311-8322, 8324-8679, 8694-8709, 8800-8814, 8816-9059, 9110, 9139-9140, 9180-9342, 9363-9379, 9386-9389, 9395, 9421-9422, 9443, 9470-9477, 9479-9480, 9491, 9501-9504, 9507, 9510-9522, 9572-9581, 9583-9993	C70.0-C72.9, C75.2	0, 1
	8720-8799, 9423, 9430, 9444	C70.0-C72.9, C75.1-C75.3	0, 1
3.10.2 Other and unspecified CNS, invasive	8000-8005, 8935-8936, 9050-9055, 9363, 9501-9504	C70.0-C72.9, C75.2	3
	9423, 9430, 9444		3

Abbreviations: CNS, central nervous system; ICD-O-3, International Classification of Diseases for Oncology, Third Edition.
[a]For behavior codes, 0 indicates benign, 1 indicates borderline malignancy, 2 indicates in situ, and 3 indicates malignant (note that, except for the CNS, behavior codes 0 and 1 are not collected by many cancer registries).
[b]For CNS germ cell tumors, see category 7.3 under Gonadal and related tumors.

together and adds 2 new groups: *blood and lymphatic vessel tumors* and *nerve sheath tumors*. The new groups occur more frequently in AYAs than in children and/or represent a higher proportion of all cancers than in adults (Tables 9-19). Categories 1, 2, and 3 (leukemias, lymphomas, and CNS tumors, respectively) have been expanded considerably with respect to specified entities, reflecting the influence of the most recent WHO Blue Books (*Tumours of the Haematopoietic and Lymphoid Tissues*[15] and *Tumours of the Central Nervous System*[16]) and updates in the ICD-O-3.2. Detailed frequencies and incidence rates by 5-year age groups are provided in Supporting Tables 1 and 2.

A final group, category 11—*unspecified malignant neoplasms except CNS*—is the smallest main category (<0.5% of all cases). An additional 20 cases (<0.001%), including 5 in the AYA group, were unclassified and excluded from the current analyses (see Table 1).

SPECIFIC CONSIDERATIONS

Subgroupings

To enhance flexibility many subgroups were added, eg, the primary site of the carcinomas (Table 13) and often the types (histologies) by primary site (Tables 14-18). Carcinomas are <10% of cancers in those aged <15 years but, by age 33 years, they are >50% (Fig. 1). In some cases, detailed subgroups that are more clinically or epidemiologically relevant have been created within a broader category, eg, the separation of nodular lymphocyte-predominant Hodgkin lymphoma from classical Hodgkin lymphomas.[17,18] Within neoplasms of the gastrointestinal tract, neuroendocrine tumors of the appendix are especially prevalent in AYAs, with a striking recent increase in incidence (Fig. 2), as also noted by others and caused in part by a change in reportability.[19] Broad ranges of histology codes, especially for carcinomas, are intended to capture all histologies occurring at individual sites and may include histologies that would not be found for that particular site.

Cross References

These are provided for 2 subgroups to maximize ease of identification/location:

- CNS germ cell tumors are classified under gonadal and related tumors (Table 5, category 7.3) and are referenced with a note on category 3 in Table 3 (CNS).
- Kaposi sarcoma is classified under blood and lymphatic vessel tumors (Table 4, category 5.2.1) and is referenced with a note on category 4 (sarcomas) in Table 4.

TABLE 4. Categories 4 (Sarcomas), 5 (Blood and Lymphatic Vessel Tumors), and 6 (Nerve Sheath Tumors): Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology	ICD-O-3 Primary Site	Behavior[a]
4. Sarcomas[b]			
4.1 Osteosarcoma	9180-9187, 9192-9210		3
4.2 Chondrosarcoma	9220-9221, 9230-9231, 9240, 9242-9243		3
4.3 Ewing family of tumors			
4.3.1 Bone	9260, 9364-9365, 9473	C40.0-C41.9	3
4.3.2 Soft tissue	9260, 9364-9365, 9473	C00.0-C39.9, C42.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
4.4 Fibromatous neoplasms			
4.4.1 Myxofibrosarcoma	8811		3
4.4.2 Malignant fibrous histiocytoma[c]	8830-8831, 8835-8836		3
4.4.3 Other fibromatous neoplasms	8810, 8813-8814, 8820-8826, 8832-8833, 9252		3
4.5 Liposarcoma	8850-8881		3
4.6 Synovial sarcoma	9040-9043		3
4.7 Leiomyosarcoma	8890-8893		3
4.8 Rhabdomyosarcoma	8900-8904, 8910, 8912, 8920-8921		3
4.9 Gastrointestinal stromal tumor, malignant	8936	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
4.10 Spindle cell sarcoma	8801	C00.0-C39.9, C42.0-C80.9	3
4.11 Epithelioid sarcoma	8804		3
4.12 Desmoplastic small round cell tumor	8806	C00.0-C39.9, C42.0-C80.9	3
4.13 Chordoma	9370-9379		3
4.14 Giant cell sarcoma	8802	C00.0-C39.9, C42.0-C80.9	3
4.15 Other soft tissue sarcomas	8840-8849, 8894-8897, 8982-8983, 8990-8991, 9044-9045, 9251, 9580-9581		3
	8935	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
	8800, 8803, 8805	C00.0-C39.9, C42.0-C80.9	3
4.16 Other bone tumors	8812, 9250, 9261, 9270-9342		3
	8000-8005, 8800-8803, 8805-8806	C40.0-C41.9	3
5. Blood and lymphatic vessel tumors			
5.1 Benign blood and lymphatic vessel tumors, CNS			
5.1.1 Hemangioblastoma and tufted hemangioma	9161	C70.0-C72.9, C75.1-C75.3	0, 1
5.1.2 Cavernous hemangioma	9121	C70.0-C72.9, C75.1-C75.3	0, 1
5.1.3 Other	8710-8714, 8815, 9120, 9122-9160, 9170-9175	C70.0-C72.9, C75.1-C75.3	0, 1
5.2 Malignant blood and lymphatic vessel tumors, all sites			
5.2.1 Kaposi sarcoma	9140		3
5.2.2 Other	8710-8714, 8815, 9120-9138, 9141-9175		3
6. Nerve sheath tumors			
6.1 Benign, CNS			
6.1.1 Neurilemmoma	9560	C70.0-C72.9, C75.1-C75.3	0, 1
6.1.2 Other	9540-9550, 9561-9571	C70.0-C72.9, C75.1-C75.3	0, 1
6.2 Malignant			
6.2.1 MPNST (Malignant peripheral nerve sheath tumor)			
6.2.1.1 CNS	9540-9542	C70.0-C72.9, C75.1-C75.3	3
6.2.1.2 Peripheral	9540-9542	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
6.2.2 Other	9543-9571		3

Abbreviations: CNS, central nervous system; ICD-O-3, International Classification of Diseases for Oncology, Third Edition.
[a]For behavior codes, 0 indicates benign, 1 indicates borderline malignancy, 2 indicates in situ, and 3 indicates malignant.
[b]This category does not include Kaposi sarcoma (see category 5.2.1).
[c]This category includes undifferentiated pleomorphic sarcoma.

TABLE 5. Categories 7 (Gonadal and Related Tumors) and 8 (Melanoma): Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology	ICD-O-3 Primary Site	Behavior[a]
7. Gonadal and related tumors			
7.1 Testis			
7.1.1 Germ cell and trophoblastic			
7.1.1.1 Seminoma	9060-9064	C62.0-C62.9	3
7.1.1.2 Embryonal carcinoma	9070	C62.0-C62.9	3
7.1.1.3 Endodermal sinus (yolk sac tumor)	9071	C62.0-C62.9	3
7.1.1.4 Teratoma	9080-9084	C62.0-C62.9	3
7.1.1.5 Mixed germ cell	9085, 9086	C62.0-C62.9	3
7.1.1.6 Choriocarcinoma and other trophoblastic	9100-9105	C62.0-C62.9	3
7.1.1.7 Other	9065, 9072	C62.0-C62.9	3
7.1.2 Non-germ cell			
7.1.2.1 Carcinoma	8010-8589	C62.0-C62.9	3
7.1.2.2 Sex cord	8590-8671	C62.0-C62.9	3
7.2 Ovary			
7.2.1 Germ cell and trophoblastic			
7.2.1.1 Teratoma	9070, 9072-9084	C56.9	3
7.2.1.2 Dysgerminoma	9060	C56.9	3
7.2.1.3 Yolk sac	9071	C56.9	3
7.2.1.4 Mixed germ cell	9085, 9086	C56.9	3
7.2.1.5 Other germ cell and trophoblastic	9061-9065, 9090-9091, 9100-9105	C56.9	3
7.2.2 Non-germ cell			
7.2.2.1 Carcinoma			
7.2.2.1.1 Adenocarcinoma			
7.2.2.1.1.1 Clear cell adenocarcinoma	8310	C56.9	3
7.2.2.1.1.2 Cystadenocarcinoma	8440-8474	C56.9	3
7.2.2.1.1.3 Mixed cell adenocarcinoma	8323	C56.9	3
7.2.2.1.1.4 Mucinous adenocarcinoma	8480-8482	C56.9	3
7.2.2.1.1.5 Endometrioid	8380-8383	C56.9	3
7.2.2.1.1.6 Other adenocarcinoma	8140-8231, 8250-8309, 8311-8322, 8324-8379, 8384, 9110	C56.9	3
7.2.2.1.2 Other carcinoma	8010-8139, 8240-8249, 8390-8439, 8483-8589	C56.9	3
7.2.2.2 Sex cord and other specialized gonadal	8590-8671	C56.9	3
7.3 Germ cell and trophoblastic, CNS	9060-9065, 9070-9073, 9080-9085, 9090-9091, 9100-9105	C70.0-C72.9, C75.1-C75.3	0, 1, 3
7.4 Germ cell and trophoblastic excluding CNS, ovary, testis	9060-9065, 9070-9073, 9080-9085, 9090-9091, 9100-9105	C00.0-C56.8, C57.0-C61.9, C63.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
7.5 Non-germ cell specified tumors excluding CNS, ovary, testis	8590-8671	C00.0-C55.9, C57.0-C61.9, C63.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
7.6 Fibroepithelial including Brenner, excluding breast phyllodes	9000-9019, 9021-9030		3
8. Melanoma, malignant			
8.1 Superficial spreading/low cumulative sun damage melanoma	8743		3
8.2 Nodular melanoma	8721		3
8.3 Other malignant	8720, 8722-8742, 8744-8790		3

Abbreviations: CNS, central nervous system; ICD-O-3, International Classification of Diseases for Oncology, Third Edition.
[a]For behavior codes, 0 indicates benign, 1 indicates borderline malignancy, 2 indicates in situ, and 3 indicates malignant.

TABLE 6. Carcinomas: Categories 9.1 Through 9.5: Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology[a]	ICD-O-3 Primary Site	Behavior[b]
9. Carcinomas[c]			
9.1 Thyroid carcinoma			
9.1.1 Medullary	8345, 8510-8513	C73.9	3
9.1.2 Hurthle cell carcinoma	8290	C73.9	3
9.1.3 Papillary	8050-8053, 8260, 8341-8344, 8503-8507	C73.9	3
9.1.4 Follicular	8330-8335, 8339	C73.9	3
9.1.5 Papillary with follicular variant	8340	C73.9	3
9.1.6 Other	8010-8046, 8054-8257, 8261-8289, 8291-8325, 8336-8337, 8346-8502, 8508-8509, 8514-8589	C73.9	3
9.2 Other carcinoma of head and neck			
9.2.1 Nasopharyngeal carcinoma			
9.2.1.1 Nasopharyngeal carcinoma, squamous	8050-8089	C11.0-C11.9	3
9.2.1.2 Nasopharyngeal carcinoma, other	8010-8049, 8090-8589	C11.0-C11.9	3
9.2.2 Oral cavity, lip, and pharynx			
9.2.2.1 Oral cavity, lip, and pharynx, squamous	8050-8089	C00.0-C06.9, C09.0-C10.9, C12.0-C14.8	3
9.2.2.2 Oral cavity, lip, and pharynx, mucoepidermoid	8430-8439	C00.0-C06.9, C09.0-C10.9, C12.0-C14.8	3
9.2.2.3 Oral cavity, lip, and pharynx, other	8010-8049, 8090-8429, 8440-8589	C00.0-C06.9, C09.0-C10.9, C12.0-C14.8	3
9.2.3 Salivary gland			
9.2.3.1 Salivary gland, acinar	8550-8552	C07.9-C08.1, C08.8-C08.9	3
9.2.3.2 Salivary gland, other malignant	8010-8543, 8560-8589	C07.9-C08.1, C08.8-C08.9	3
9.2.4 Other carcinoma of head and neck	8010-8589	C30.0-C32.9, C76.0	3
9.3 Carcinoma of gastrointestinal tract			
9.3.1 Carcinoma of esophagus	8010-8589	C15.0-C15.9	3
9.3.2 Carcinoma of stomach			
9.3.2.1 Stomach, neuroendocrine			
9.3.2.1.1 Neuroendocrine tumor (NET)	8240-8245, 8248-8249	C16.0-C16.9	3
9.3.2.1.2 Neuroendocrine carcinoma (NEC)	8013, 8041-8045, 8246	C16.0-C16.9	3
9.3.2.2 Stomach, signet ring	8490	C16.0-C16.9	3
9.3.2.3 Stomach, other adenocarcinoma	8140-8239, 8250-8489, 8491-8579	C16.0-C16.9	3
9.3.2.4 Stomach, other invasive	8010-8012, 8014-8040, 8046-8139, 8247, 8580-8589	C16.0-C16.9	3
9.3.3 Carcinoma of small intestine			
9.3.3.1 Small intestine, neuroendocrine			
9.3.3.1.1 NET	8240-8245, 8248-8249	C17.0-C17.9	3
9.3.3.1.2 NEC	8013, 8041-8045, 8246	C17.0-C17.9	3
9.3.3.2 Small intestine, other	8010-8012, 8014-8040, 8046-8239, 8247, 8250-8589	C17.0-C17.9	3
9.3.4 Carcinoma of colon			
9.3.4.1 Appendix			
9.3.4.1.1 NET	8240-8245, 8248-8249	C18.1	3
9.3.4.1.2 NEC	8013, 8041-8045, 8246	C18.1	3
9.3.4.1.3 other	8010-8012, 8014-8040, 8046-8239, 8247, 8250-8589	C18.1	3
9.3.4.2 Colon excluding appendix			
9.3.4.2.1 Colon excluding appendix, neuroendocrine			
9.3.4.2.1.1 NET	8240-8245, 8248-8249	C18.0, C18.2-C18.9	3
9.3.4.2.1.2 NEC	8013, 8041-8045, 8246	C18.0, C18.2-C18.9	3
9.3.4.2.2 Colon excluding appendix, adenocarcinoma	8140-8239, 8250-8579	C18.0, C18.2-C18.9	3
9.3.4.2.3 Colon excluding appendix, other	8010-8012, 8014-8040, 8046-8139, 8247, 8580-8589	C18.0, C18.2-C18.9	3

TABLE 6. *Continued*

Category	ICD-O-3 Histology[a]	ICD-O-3 Primary Site	Behavior[b]
9.3.5 Carcinoma of rectum			
9.3.5.1 Rectum, neuroendocrine			
9.3.5.1.1 NET	8240-8245, 8248-8249	C19.9-C20.9	3
9.3.5.1.2 NEC	8013, 8041-8045, 8246	C19.9-C20.9	3
9.3.5.2 Rectum, adenocarcinoma	8140-8239, 8250-8579	C19.9-C20.9	3
9.3.5.3 Rectum, other	8010-8012, 8014-8040, 8046-8139, 8247, 8580-8589	C19.9-C20.9	3
9.3.6 Carcinoma of anus			
9.3.6.1 Anus, squamous	8050-8089	C21.0-C21.8	3
9.3.6.2 Anus, other	8010-8046, 8090-8589	C21.0-C21.8	3
9.3.7 Carcinoma of liver and intrahepatic bile ducts (IBD)			
9.3.7.1 Liver and IBD, cholangiocarcinoma	8160-8169	C22.0-C22.1	3
9.3.7.2 Liver and IBD, hepatocellular carcinoma	8170-8189	C22.0-C22.1	3
9.3.7.3 Liver and IBD, other	8010-8159, 8190-8589	C22.0-C22.1	3
9.3.8 Carcinoma of gallbladder and other extrahepatic biliary	8010-8589	C23.9-C24.9	3
9.3.9 Carcinoma of pancreas			
9.3.9.1 Pancreas, neuroendocrine			
9.3.9.1.1 NET	8150, 8240, 8249	C25.0-C25.9	3
9.3.9.1.2 NEC	8013, 8041-8045, 8246	C25.0-C25.9	3
9.3.9.1.3 Neuroendocrine, other	8151-8153, 8155-8156, 8241-8245, 8248	C25.0-C25.9	3
9.3.9.2 Pancreas, adenocarcinoma	8140-8149, 8160-8239, 8250-8579	C25.0-C25.9	3
9.3.9.3 Pancreas, other	8010-8012, 8014-8040, 8046, 8050-8139, 8154, 8157, 8247, 8580-8589	C25.0-C25.9	3
9.3.10 Other carcinoma of gastrointestinal tract	8010-8589	C26.0-C26.9	3
9.4 Carcinoma of lung, bronchus, and trachea			
9.4.1 Small cell carcinoma, NEC	8041-8045	C33.0-C34.9	3
9.4.2 Non-small cell carcinoma			
9.4.2.1 Non-small cell, adenocarcinoma	8140-8239, 8250-8579	C33.0-C34.9	3
9.4.2.2 Non-small cell, neuroendocrine			
9.4.2.2.1 Non-small cell NET	8240-8245, 8248-8249	C33.0-C34.9	3
9.4.2.2.2 Non-small cell NEC	8013, 8246	C33.0-C34.9	3
9.4.2.3 Non-small cell, other	8010-8012, 8014-8040, 8046-8139, 8247, 8580-8589, 9050-9059	C33.0-C34.9	3
9.5 Carcinoma of skin (if collected)	8010-8589	C44.0-C44.9	3

Abbreviation: ICD-O-3, International Classification of Diseases for Oncology, Third Edition.
[a]These include carcinoma histology codes that may not occur for a particular site.
[b]Excludes carcinoma in situ; a behavior code of 3 indicates malignant.
[c]This category excludes in situ carcinoma.

TABLE 7. Carcinomas: Categories 9.6 Through 9.9: Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology[a]	ICD-O-3 Primary Site	Behavior[b]
9.6 Carcinoma of breast			
9.6.1 Breast, infiltrating duct	8500, 8522-8523	C50.0-C50.9	3
9.6.2 Breast, adenocarcinoma	8140-8230, 8250-8313, 8321-8323, 8360-8384, 8401-8402, 8410-8420, 8440-8450, 8460-8490, 8503-8504, 8509, 8550-8552, 8570-8574	C50.0-C50.9	3
9.6.3 Breast, lobular	8520, 8524	C50.0-C50.9	3
9.6.4 Breast, phyllodes	9020	C50.0-C50.9	3
9.6.5 Breast, medullary	8510-8513	C50.0-C50.9	3
9.6.6 Breast, Paget	8540-8543	C50.0-C50.9	3
9.6.7 Breast, ductal	8507, 8514, 8521	C50.0-C50.9	3
9.6.8 Breast, metaplastic	8575	C50.0-C50.9	3
9.6.9 Breast, inflammatory	8530	C50.0-C50.9	3
9.6.10 Breast, other	8010-8131, 8231-8249, 8314-8320, 8324-8350, 8390-8400, 8403-8409, 8430, 8452-8454, 8501-8502, 8505-8506, 8508, 8525, 8560-8562, 8580-8589	C50.0-C50.9	3
9.7 Carcinoma of genital sites excluding ovary and testis			
9.7.1 Carcinoma of uterine cervix			
9.7.1.1 Cervix, squamous	8050-8089	C53.0-C53.9	3
9.7.1.2 Cervix, adenosquamous	8560-8570	C53.0-C53.9	3
9.7.1.3 Cervix, adenocarcinoma	8140-8559, 8571-8579	C53.0-C53.9	3
9.7.1.4 Cervix, other	8010-8049, 8090-8139, 8580-8589	C53.0-C53.9	3
9.7.2 Corpus uteri			
9.7.2.1 Corpus uteri, adenocarcinoma			
9.7.2.1.1 Corpus uteri, endometrioid	8380-8383	C54.0-C55.9	3
9.7.2.1.2 Corpus uteri, other adenocarcinoma	8140-8379, 8384-8579	C54.0-C55.9	3
9.7.2.2 Corpus uteri, other	8010-8139, 8580-8589, 8930-8931, 8950	C54.0-C55.9	3
9.7.3 Carcinoma of vulva and vagina	8010-8589	C51.0-C51.9, C52.9	3
9.7.4 Carcinoma of penis	8010-8589	C60.0-C60.9	3
9.7.5 Carcinoma of prostate	8010-8589	C61.9	3
9.7.6 Other genital	8010-8589	C57.0-C57.9, C58.9, C63.0-C63.9	3
9.8 Carcinoma of urinary tract			
9.8.1 Carcinoma of kidney			
9.8.1.1 Kidney, adenocarcinoma			
9.8.1.1.1 Kidney, renal cell	8310-8319	C64.9	3
9.8.1.1.2 Kidney, other adenocarcinoma	8140-8309, 8320-8579	C64.9	3
9.8.1.2 Kidney, other	8010-8139, 8580-8589	C64.9	3
9.8.2 Carcinoma of bladder			
9.8.2.1 Urinary bladder, transitional cell carcinoma	8120-8139	C67.0-C67.9	3
9.8.2.2 Urinary bladder, other carcinoma	8010-8119, 8140-8589	C67.0-C67.9	3
9.8.3 Other urinary	8010-8589	C65.9, C66.9, C68.0-C68.9	3
9.9 Other invasive carcinomas			
9.9.1 Adrenocortical carcinoma	8010-8589	C74.0-C74.9	3
9.9.2 Unknown primary	8010-8589, 9010	C80.9	3
9.9.3 Thymic carcinoma	8010-8589	C37.9	3
9.9.4 Carcinoma of other and ill-defined sites	8010-8589	C38.0-C42.4, C47.0-C49.9, C69.0-69.9, C75.0, C75.4-C75.9, C76.1-C80.8	3

Abbreviation: ICD-O-3, International Classification of Diseases for Oncology, Third Edition.
[a]These include carcinoma histology codes that may not occur for a particular site.
[b]Excludes carcinoma in situ; a behavior code of 3 indicates malignant.

TABLE 8. Categories 10 (Miscellaneous Specified Neoplasms), 11 (Unspecified Malignant Neoplasms Except Central Nervous System, and A (In Situ): Histology, Primary Site, and Behavior

Category	ICD-O-3 Histology	ICD-O-3 Primary Site	Behavior[a]
10. Miscellaneous specified neoplasms			
10.1 Other pediatric and embryonal tumors			
10.1.1 Wilms tumor	8959-8960	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
10.1.2 Olfactory and non-CNS neuroblastoma	9490, 9500	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
	9522	C00.0-C26.9, C30.0, C30.1-C69.9, C70.0-C72.9, C73.0-C75.0, C75.1-C75.2, C75.4-C80.9	3
10.1.3 Other embryonal non-CNS tumors	8963-8964, 8970-8973, 8981, 9501-9521, 9523	C00.0-C26.9, C30.1-C69.9, C73.0-C75.0, C75.4-C80.9	3
	8963-8964, 8970-8973, 8981, 9501-9521	C30.0	3
	8963	C70.0-C72.9, C75.1-C75.3	3
10.2 Other specified tumors			
10.2.1 Paraganglioma, non-CNS	8680-8693, 8700	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
10.2.2 Other specified neoplasms	8932-8934, 8937-8939, 8941-8949, 8951, 8980, 9110	C00.0-C56.8, C57.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
	8932-8934, 8937-8939, 8941-8949, 8951, 8980	C56.9	3
	9050-9055	C00.0-C32.9, C35.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
	8930-8931, 8950	C00.0-C53.9, C56.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
	8370-8375	C75.1	3
11. Unspecified malignant neoplasms except CNS	8000-8005	C00.0-C39.9, C42.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
	8940	C00.0-C69.9, C73.0-C75.0, C75.4-C80.9	3
A. In situ			
A.1 Melanoma in situ (if collected)			
A.1.1 Superficial spreading melanoma in situ	8743		2
A.1.2 Lentigo maligna	8742		2
A.1.3 Other in situ melanoma	8720-8741, 8744-8790		2
A.2 Colon including appendix, in situ	8000-8719, 8800-9589	C18.0-C18.9	2
A.3 Rectum, in situ	8000-8719, 8800-9589	C19.9-C20.9	2
A.4 Anus, in situ	8000-8719, 8800-9589	C21.0-C21.8	2
A.5 Breast, in situ	8000-8719, 8800-9589	C50.0-C50.9	2
A.6 Cervix uteri, in situ (not collected by some registries)	8000-8719, 8800-9589	C53.0-C53.9	2
A.7 Ovary, in situ	8000-8719, 8800-9589	C56.9	2
A.8 Vulva and vagina, in situ	8000-8719, 8800-9589	C51.0-C51.9, C52.9	2
A.9 Penis, in situ	8000-8719, 8800-9589	C60.0-C60.9	2
A.10 Prostate in situ including PIN III[b]	8000-8719, 8800-9589	C61.9	2
A.11 Urinary bladder, in situ	8000-8719, 8800-9589	C67.0-C67.9	2
A.12 Other in situ	8000-8719, 8800-9589	C00.0-C17.9, C22.0-C49.9, C54.0-C55.9, C57.0-C58.9, C62.0-C66.9, C68.0-C80.9	2

Abbreviations: CNS, central nervous system; ICD-O-3, International Classification of Diseases for Oncology, Third Edition; PIN III, prostatic intraepithelial neoplasia grade 3.
[a]For behavior codes, 2 indicates in situ, and 3 indicates malignant.
[b]PIN III is not collected by all registries.

TABLE 9. Categories 1 (Leukemias and Related Disorders) and 2 (Lymphomas): Total Cases and Percentage Distribution Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017

	Age at Diagnosis, y									
Category	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
1. Leukemias and related disorders, N	13,365	2986	2768	3116	3824	4869	241,888	272,816	8870	17,563
Percentage of category 1										
1.1 Acute lymphoblastic leukemia	76.5	51.5	32.0	21.5	15.9	13.3	2.6	7.7	34.9	24.8
1.2 Acute myeloid leukemia	15.1	27.8	33.9	32.0	31.6	27.3	18.5	19.1	31.2	30.2
1.2.1 Acute promyelocytic leukemia	1.1	5.9	7.9	7.3	7.3	6.6	1.1	1.5	7.0	6.9
1.2.2 Other acute myeloid leukemia	14.0	22.0	26.0	24.7	24.4	20.7	17.4	17.6	24.2	23.3
1.3 Chronic myeloid leukemia	2.3	7.2	14.5	18.2	19.7	18.3	7.9	8.2	13.3	16.1
1.4 Chronic lymphocytic leukemia	0.1	0.5	0.5	1.3	2.4	5.4	25.2	22.5	0.8	2.4
1.5 Polycythemia vera	0.3	2.1	3.1	5.3	6.5	7.9	5.9	5.6	3.6	5.4
1.6 Essential thrombocythemia	0.7	2.4	6.0	8.4	9.8	10.1	5.8	5.7	5.6	7.8
1.7 Primary myelofibrosis	—[a]	—[a]	—[a]	0.4	0.6	1.2	1.7	1.5	0.2	0.6
1.8 Myelodysplastic syndrome	1.8	3.9	3.8	4.1	4.9	5.3	23.8	21.5	3.9	4.5
1.9 Other and unspecified leukemias and related disorders	3.0	4.5	6.1	8.7	8.7	11.2	8.6	8.3	6.5	8.3
1.9.1 Hairy cell leukemia	—[a]	—[a]	—[a]	0.7	1.7	3.9	1.4	1.4	0.3	1.6
1.9.2 Other lymphocytic/lymphoblastic leukemias	0.2	0.8	0.4	0.9	1.0	1.0	1.3	1.2	0.7	0.8
1.9.3 Other myeloid leukemias	0.6	0.9	1.7	2.0	1.6	1.7	0.8	0.9	1.5	1.6
1.9.4 Leukemias of mixed phenotype	0.8	1.2	0.6	0.7	0.5	0.4	0.1	0.2	0.8	0.6
1.9.5 Mast cell diseases	0.1	0.2	0.5	0.9	0.8	0.8	0.2	0.2	0.6	0.7
1.9.6 Other	1.4	1.4	2.8	3.6	3.1	3.3	4.7	4.4	2.6	2.9
2. Lymphomas, N	4989	4346	5942	6568	7332	9028	329,515	367,720	16,856	33,216
Percentage of category 2										
2.1 Non-Hodgkin lymphomas	49.8	36.4	34.3	42.6	51.8	60.8	64.4	62.7	38.1	47.3
2.1.1 Lymphoblastic	15.1	5.4	2.9	2.7	1.8	1.3	0.3	0.7	3.5	2.5
2.1.2 Burkitt	12.1	4.9	3.3	2.8	3.2	3.1	0.7	1.1	3.5	3.3
2.1.3 Diffuse large B-cell (DLBCL)	7.4	12.0	13.9	18.0	21.1	24.6	28.3	27.1	15.0	18.9
2.1.4 Primary mediastinal large B-cell excluded from DLBCL	0.3	1.0	1.6	1.7	1.8	1.5	0.1	0.2	1.5	1.6
2.1.5 Anaplastic T-cell and null-cell excluding NK/T-cell	9.1	7.3	6.9	7.6	8.9	9.4	5.4	5.7	7.3	8.2
2.1.6 Follicular	1.3	1.2	1.5	3.2	6.6	11.4	12.9	12.1	2.1	5.6
2.1.7 NK/T-cell (excluded from anaplastic T-cell)	0.3	0.2	0.5	0.7	0.8	0.8	0.2	0.2	0.5	0.7
2.1.8 MALT (Mucosa-associated lymphoid tissue)	0.9	2.0	1.5	2.9	3.9	4.5	6.6	6.2	2.2	3.2
2.1.9 Other non-Hodgkin lymphoma NOS	3.3	2.3	2.2	3.1	3.6	4.3	10.0	9.3	2.6	3.3
2.2 Hodgkin lymphoma	28.5	59.8	62.3	52.5	40.4	26.7	4.8	8.8	57.8	45.5
2.2.1 Hodgkin NLP	3.2	3.1	2.3	2.1	2.2	1.6	0.4	0.6	2.4	2.1
2.2.2 Hodgkin classic, other	25.3	56.6	60.0	50.4	38.3	25.1	4.5	8.3	55.4	43.4
2.3 Myeloma	0.2	0.3	0.7	1.4	4.0	7.6	24.7	22.4	0.8	3.4
2.4 Cutaneous lymphomas	—[a]	—[a]	0.1	0.2	0.1	0.4	0.2	0.2	0.1	0.2
2.5 Other B-cell and T-cell lymphomas	0.5	0.3	0.3	0.7	0.8	0.7	1.3	1.3	0.4	0.6
2.6 Other lymphomas, specified and unspecified	20.9	3.2	2.4	2.6	2.8	3.8	4.5	4.6	2.7	3.0
2.6.1 Histiocytic and dendritic cell neoplasms	19.9	2.1	0.9	1.0	0.9	0.9	0.2	0.6	1.2	1.1
2.6.2 Lymphoma NOS	1.0	1.2	1.5	1.6	1.9	3.0	4.3	4.0	1.5	2.0

Abbreviations: NK, natural killer cell; NLP, nodular lymphocyte-predominant; NOS, not otherwise specified; PMLCL, primary mediastinal large B-cell lymphoma.
[a]The statistics are not displayed for this group because there were less than 6 cases.

TABLE 10. Category 3 (Central Nervous System and Other Cranial and Intraspinal Neoplasms): Total Cases and Percentage Distribution Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017

Category	Age at Diagnosis, y									
	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
3. CNS and other intracranial and intraspinal neoplasms, N[a]	11,721	4782	5781	7789	9748	11,545	205,337	256,703	18,352	39,645
Percentage of category 3										
3.1 Astroglial and related neoplasms	55.1	35.0	31.3	29.8	27.8	26.3	27.1	28.7	31.6	29.1
3.1.1 Oligodendriogliomas	1.4	2.9	4.8	6.7	7.1	6.1	2.0	2.5	5.1	5.9
3.1.1.1 Oligodendrioglioma, benign/borderline	—[b]	—[b]	—[b]	—[b]	—[b]	—[b]	—[b]	0.0	—[b]	—[b]
3.1.1.2 Oligodendrioglioma, invasive	1.4	2.9	4.8	6.7	7.0	6.1	2.0	2.5	5.1	5.9
3.1.2 Glioblastomas/gliofibromas	3.1	3.7	4.0	4.5	4.9	6.4	18.6	15.8	4.1	5.0
3.1.2.1 Gliofibroma, benign/borderline	—[b]	—[b]	—[b]	—[b]	—[b]	—[b]	0.0	0.0	—[b]	—[b]
3.1.2.2 Glioblastoma, invasive	—[b]	—[b]	4.0	4.5	4.9	6.4	18.6	15.8	4.1	5.0
3.1.3 Ependymomas	6.5	4.4	4.4	4.0	3.5	3.3	1.4	2.0	4.3	3.8
3.1.3.1 Ependymoma, benign/borderline	0.8	1.6	1.7	1.7	1.7	1.3	0.6	0.8	1.7	1.6
3.1.3.2 Ependymoma, invasive	5.7	2.9	2.7	2.3	1.8	2.0	0.8	1.2	2.6	2.2
3.1.4 Other astrocytoma/astroglial neoplasms	44.2	24.0	18.1	14.6	12.4	10.4	5.1	8.3	18.2	14.5
3.1.4.1 Pilocytic astrocytoma	19.3	10.0	4.2	2.2	1.5	1.0	0.2	1.5	4.8	2.9
3.1.4.2 Other astrocytoma/astroglial, benign/borderline	3.3	1.3	0.7	0.4	0.3	0.3	0.1	0.3	0.8	0.5
3.1.4.3 Other astrocytoma/astroglial, invasive	21.7	12.6	13.2	12.0	10.6	9.2	4.8	6.5	12.5	11.1
3.2 Medulloblastoma and other embryonal tumors, invasive	14.9	4.0	2.8	1.8	1.1	0.8	0.1	1.0	2.7	1.7
3.3 Neuroblastomas/ganglioneuromas	0.9	0.3	0.2	0.1	0.1	0.1	0.0	0.1	0.2	0.1
3.3.1 Ganglioneuroma, benign/borderline	0.1	0.2	—[b]	0.1	—[b]	0.1	0.0	0.0	0.1	0.1
3.3.2 Neuroblastoma/ganglioneuroblastoma, invasive	0.8	—[b]	0.1	—[b]	—[b]	—[b]	0.0	0.1	0.1	0.1
3.4 Neuronal and mixed neuronal-glial neoplasms	7.5	8.6	5.0	3.2	2.3	1.6	0.4	1.2	5.2	3.4
3.4.1 Neuronal and mixed neuronal-glial, benign/borderline	7.2	8.3	4.8	3.1	2.2	1.5	0.3	1.1	5.0	3.3
3.4.2 Neuronal and mixed neuronal-glial, invasive	0.2	0.3	0.2	0.2	0.1	0.1	0.0	0.1	0.2	0.1
3.5 Meningiomas	2.0	5.8	9.0	13.4	20.7	28.4	50.4	43.2	10.0	18.0
3.5.1 Meningioma, benign/borderline	1.8	5.6	8.8	13.2	20.4	28.1	49.8	42.6	9.8	17.7
3.5.2 Meningioma, invasive	0.1	0.2	0.3	0.2	0.3	0.4	0.6	0.5	0.2	0.3
3.6 Choroid plexus neoplasms	2.6	1.0	0.4	0.5	0.3	0.4	0.1	0.3	0.6	0.5
3.6.1 Choroid plexus, benign/borderline	1.8	0.9	0.4	0.5	0.3	0.4	0.1	0.2	0.6	0.4
3.6.2 Choroid plexus, invasive	0.8	—[b]	—[b]	—[b]	—[b]	—[b]	0.0	0.0	0.0	0.0
3.7 Craniopharyngiomas	4.7	2.6	1.6	1.2	1.1	1.0	0.6	0.9	1.7	1.3
3.7.1 Craniopharyngioma, benign/borderline	4.7	2.6	1.6	1.2	1.1	1.0	0.6	0.9	1.7	1.3
3.7.2 Craniopharyngioma, invasive	—[b]	—[b]	—[b]	—[b]	—[b]	—[b]	—[b]	0.0	—[b]	—[b]
3.8 Pituitary neoplasms	5.7	38.2	45.6	46.6	43.4	38.9	17.5	20.8	44.1	42.4
3.8.1 Pituitary, benign/borderline	5.7	38.2	45.5	46.5	43.4	38.9	17.5	20.8	44.0	42.4
3.8.2 Pituitary, invasive	—[b]	—[b]	—[b]	0.1	—[b]	0.1	0.1	0.1	0.1	0.1
3.9 Pineal neoplasms	1.1	1.3	1.0	0.9	0.7	0.4	0.2	0.3	1.1	0.8
3.9.1 Pineal, benign/borderline	0.3	0.6	0.6	0.6	0.4	0.2	0.1	0.2	0.6	0.4
3.9.2 Pineal, invasive	0.8	0.7	0.5	0.3	0.2	0.2	0.1	0.1	0.5	0.3
3.10 Other and unspecified CNS neoplasms	5.4	3.3	3.0	2.5	2.5	2.1	3.6	3.5	2.9	2.6
3.10.1 Other and unspecified CNS, benign/borderline	4.4	2.8	2.5	1.9	1.9	1.6	1.9	2.0	2.3	2.0
3.10.2 Other and unspecified CNS, invasive	1.0	0.5	0.6	0.6	0.6	0.5	1.7	1.5	0.6	0.5

Abbreviation: CNS, central nervous system.

[a]For CNS germ cell tumors, see category 7.3 under Gonadal and related tumors.

[b]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 11. Categories 4 (Sarcomas), 5 (Blood and Lymphatic Vessel Tumors), and 6 (Nerve Sheath Tumors): Total Cases and Percentage Distribution Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017

	Age at Diagnosis, y									
Category	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
4. Sarcomas, N[a]	4331	2546	2222	2381	2813	3426	64,715	82,434	7149	13,388
Percentage of category 4										
4.1 Osteosarcoma	24.0	28.0	15.5	8.3	5.1	4.2	2.1	4.8	17.5	11.5
4.2 Chondrosarcoma	1.8	3.2	6.2	7.4	7.1	6.9	4.7	4.8	5.5	6.2
4.3 Ewing family of tumors	19.5	20.7	14.1	9.2	4.6	3.4	0.9	3.3	14.8	9.7
4.3.1 Bone	14.4	13.5	8.5	4.2	1.9	1.2	0.3	1.9	8.9	5.4
4.3.2 Soft tissue	5.1	7.2	5.6	4.9	2.7	2.3	0.6	1.4	6.0	4.3
4.4 Fibromatous neoplasms	8.7	10.2	18.2	26.3	28.9	25.3	16.9	17.3	18.1	22.2
4.4.1 Myxofibrosarcoma	0.9	1.6	2.5	2.4	2.5	2.5	3.0	2.8	2.1	2.3
4.4.2 Malignant fibrous histiocytoma[b]	0.9	1.0	1.3	1.6	2.0	2.0	7.8	6.4	1.3	1.6
4.4.3 Other fibromatous neoplasms	6.9	7.6	14.5	22.3	24.4	20.8	6.1	8.1	14.6	18.3
4.5 Liposarcoma	1.0	3.1	5.0	6.9	10.6	12.4	14.5	12.7	4.9	8.0
4.6 Synovial sarcoma	3.3	6.4	8.0	7.9	7.0	5.1	1.7	2.6	7.4	6.8
4.7 Leiomyosarcoma	0.7	1.1	3.1	4.6	8.8	13.3	17.7	15.1	2.9	6.8
4.8 Rhabdomyosarcoma	28.0	10.9	5.6	4.0	2.7	2.3	1.6	3.5	7.0	4.9
4.9 Gastrointestinal stromal tumor, malignant	0.3	0.9	1.6	3.2	5.0	6.6	15.0	12.4	1.9	3.8
4.10 Spindle cell sarcoma	0.8	1.2	2.0	1.6	2.3	2.6	3.2	2.9	1.6	2.0
4.11 Epithelioid sarcoma	0.9	1.8	2.1	1.9	1.5	1.9	0.7	0.9	1.9	1.8
4.12 Desmoplastic small round cell tumor	1.1	1.8	3.1	2.8	1.1	0.7	0.1	0.4	2.6	1.8
4.13 Chordoma	1.1	1.1	1.6	1.9	2.1	2.5	1.6	1.6	1.5	1.9
4.14 Giant cell sarcoma	0.3	0.5	0.6	1.5	2.2	2.1	5.5	4.6	0.9	1.5
4.15 Other soft tissue sarcomas	7.3	6.0	10.7	9.9	9.2	8.8	12.3	11.5	8.8	8.9
4.16 Other bone tumors	1.1	3.0	2.7	2.6	1.9	1.9	1.6	1.7	2.8	2.4
5. Blood and lymphatic vessel tumors, N	350	293	527	963	1293	1374	13,068	17,868	1783	4450
Percentage of category 5										
5.1 Benign blood and lymphatic vessel tumors, CNS	86.9	84.3	53.5	38.7	36.0	33.5	31.8	35.2	50.6	41.1
5.1.1 Hemangioblastoma and tufted hemangioma	14.0	35.8	22.0	15.9	14.6	12.9	10.9	12.4	21.0	16.6
5.1.2 Cavernous hemangioma	48.6	34.8	22.6	15.5	13.8	13.1	11.4	13.4	20.8	16.4
5.1.3 Other	24.3	13.7	8.9	7.4	7.5	7.5	9.5	9.5	8.9	8.0
5.2 Malignant blood and lymphatic vessel tumors, all sites	13.1	15.7	46.5	61.3	64.0	66.5	68.2	64.8	49.4	58.9
5.2.1 Kaposi sarcoma	—[c]	3.1	35.5	50.8	52.7	54.9	32.4	35.6	38.4	47.7
5.2.2 Other	11.7	12.6	11.0	10.5	11.4	11.6	35.8	29.2	11.0	11.3
6. Nerve sheath tumors, N	852	463	615	832	1179	1632	22,311	27,884	1910	4721
Percentage of category 6										
6.1 Benign, CNS	88.4	79.7	79.7	84.4	88.4	90.4	94.4	92.8	81.7	86.4
6.1.1 Neurilemmoma	29.3	65.2	69.8	78.4	83.5	87.0	91.8	87.9	72.4	80.2
6.1.2 Other	59.0	14.5	9.9	6.0	4.9	3.4	2.6	4.9	9.3	6.2
6.2 Malignant	11.6	20.3	20.3	15.6	11.6	9.6	5.6	7.2	18.3	13.6
6.2.1 MPNST (Malignant peripheral nerve sheath tumor)	9.9	18.8	17.4	13.6	10.3	8.5	4.7	6.1	16.1	12.0
6.2.1.1 CNS	1.1	—[c]	1.1	0.7	—[c]	0.4	0.3	0.4	0.9	0.6
6.2.1.2 Peripheral	8.8	17.9	16.3	12.9	10.1	8.0	4.4	5.7	15.2	11.4
6.2.2 Other	1.8	1.5	2.9	2.0	1.3	1.2	0.9	1.0	2.2	1.6

Abbreviation: CNS, central nervous system.

[a]For Kaposi sarcoma, see category 5.2.1.

[b]This category includes undifferentiated pleomorphic sarcoma.

[c]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 12. Categories 7 (Gonadal and Related Tumors) and 8 (Melanoma): Total Cases and Percentage Distribution Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017

	Age at Diagnosis, y									
Category	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
7. Gonadal and related tumors, N	1561	2614	5810	7687	7557	6888	79,356	111,473	16,111	30,556
Percentage of category 7										
7.1 Testis	15.4	62.1	80.6	82.1	77.6	67.8	12.9	30.2	78.3	75.8
7.1.1 Germ cell and trophoblastic	15.2	61.8	79.8	81.7	77.1	67.2	12.6	29.8	77.8	75.2
7.1.1.1 Seminoma	0.6	7.3	22.1	36.4	45.7	46.7	9.7	16.7	26.5	35.8
7.1.1.2 Embryonal carcinoma	0.8	8.8	10.2	8.8	6.0	3.8	0.5	2.4	9.3	7.3
7.1.1.3 Endodermal sinus (yolk sac tumor)	7.8	1.2	1.2	1.1	0.6	0.5	0.1	0.4	1.1	0.9
7.1.1.4 Teratoma	2.7	5.0	4.9	2.8	1.9	1.2	0.2	0.9	3.9	2.8
7.1.1.5 Mixed germ cell	2.7	34.0	34.6	27.9	19.8	13.1	1.8	8.0	31.3	24.4
7.1.1.6 Choriocarcinoma and other trophoblastic	0.4	3.3	3.1	2.3	1.6	0.9	0.1	0.7	2.8	2.0
7.1.1.7 Other	—[a]	2.3	3.7	2.4	1.4	0.9	0.2	0.7	2.9	2.1
7.1.2 Non-germ cell	—[a]	0.3	0.7	0.4	0.5	0.6	0.3	0.4	0.5	0.5
7.1.2.1 Carcinoma	—[a]	0.2	0.4	0.2	0.2	0.3	0.1	0.2	0.3	0.3
7.1.2.2 Sex cord	—[a]	—[a]	0.3	0.2	0.3	0.3	0.2	0.2	0.2	0.2
7.2 Ovary	29.8	22.1	12.0	12.3	17.5	28.2	85.8	66.4	13.8	18.0
7.2.1 Germ cell and trophoblastic	26.6	15.3	6.3	4.2	3.1	1.9	0.3	1.9	6.7	4.7
7.2.1.1 Teratoma	11.3	5.5	2.3	1.5	1.5	0.9	0.1	0.8	2.5	1.9
7.2.1.2 Dysgerminoma	5.6	4.9	1.8	1.4	0.7	0.4	0.0	0.5	2.1	1.4
7.2.1.3 Yolk sac	3.7	2.3	0.9	0.5	0.3	0.3	0.0	0.3	0.9	0.6
7.2.1.4 Mixed germ cell	4.8	2.3	1.0	0.5	0.3	0.2	0.0	0.3	1.0	0.6
7.2.1.5 Other germ cell and trophoblastic	1.3	0.3	0.2	0.2	0.2	0.2	0.1	0.1	0.2	0.2
7.2.2 Non-germ cell	3.2	6.9	5.7	8.2	14.4	26.2	85.5	64.5	7.1	13.2
7.2.2.1 Carcinoma	1.5	4.9	5.2	7.3	13.2	24.4	84.1	63.2	6.2	12.0
7.2.2.1.1 Adenocarcinoma	0.8	3.9	4.4	6.1	11.4	21.6	75.6	56.7	5.1	10.4
7.2.2.1.1.1 Clear cell adenocarcinoma	—[a]	—[a]	—[a]	0.1	1.0	1.9	4.8	3.6	0.1	0.7
7.2.2.1.1.2 Cystadenocarcinoma	0.4	1.7	2.3	2.8	4.7	9.7	43.3	32.1	2.4	4.6
7.2.2.1.1.3 Mixed cell adenocarcinoma	—[a]	—[a]	0.2	0.3	0.6	1.0	3.6	2.7	0.2	0.5
7.2.2.1.1.4 Mucinous adenocarcinoma	—[a]	1.7	1.2	1.7	1.9	2.3	3.2	2.8	1.5	1.8
7.2.2.1.1.5 Endometrioid	—[a]	—[a]	0.4	0.9	2.6	5.2	8.4	6.6	0.6	2.1
7.2.2.1.1.6 Other adenocarcinoma	—[a]	—[a]	0.3	0.3	0.6	1.5	12.3	8.9	0.3	0.6
7.2.2.1.2 Other carcinoma	0.7	1.1	0.7	1.2	1.8	2.8	8.5	6.5	1.0	1.6
7.2.2.2 Sex cord and other specialized gonadal	1.7	2.0	0.6	0.8	1.2	1.8	1.3	1.3	0.9	1.2
7.3 Germ cell and trophoblastic, CNS	32.4	10.4	2.4	1.2	0.7	0.6	0.2	1.1	3.1	1.9
7.4 Germ cell and trophoblastic excluding CNS, ovary, testis	22.4	5.4	5.0	4.3	4.2	3.4	0.8	2.1	4.7	4.3
7.5 Non-germ cell specified tumors excluding CNS, ovary, testis	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	0.0	0.0	—[a]	—[a]
7.6 Fibroepithelial including Brenner excluding breast phyllodes	—[a]	—[a]	—[a]	—[a]	0.1	—[a]	0.3	0.2	0.0	0.0
8. Melanoma, malignant, N	499	1010	3102	5712	8195	10,845	261,119	290,482	9824	28,864
Percentage of category 8										
8.1 Superficial spreading/low cumulative sun damage melanoma	13.8	34.5	40.1	40.2	40.9	40.5	28.6	29.7	39.6	40.3
8.2 Nodular melanoma	7.2	5.3	5.4	4.4	4.6	4.7	6.9	6.6	4.8	4.7
8.3 Other malignant	79.0	60.2	54.6	55.4	54.4	54.9	64.6	63.7	55.6	55.0

Abbreviation: CNS, central nervous system.
[a]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 13. Total Cases and Distribution of Category 9 (Carcinomas) by Primary Site Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017[a]

	Age at Diagnosis, y									
Category	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9. Carcinomas, N[a]	1678	3859	9420	21,312	42,023	73,225	4,256,805	4,408,322	34,591	149,839
Percentage of category 9										
9.1 Thyroid carcinoma	45.5	57.5	52.3	39.1	28.2	20.0	2.8	3.6	44.8	28.0
9.2 Other carcinoma of head and neck	15.6	9.9	6.5	4.4	3.8	3.9	4.3	4.3	5.6	4.3
9.2.1 Nasopharyngeal carcinoma	4.5	2.6	1.3	0.7	0.5	0.6	0.2	0.2	1.0	0.7
9.2.2 Oral cavity, lip, and pharynx	2.7	2.3	2.0	1.9	1.9	2.1	2.7	2.7	2.0	2.0
9.2.3 Salivary gland	7.9	4.3	2.7	1.4	1.0	0.6	0.3	0.3	2.1	1.1
9.2.4 Other carcinoma of head and neck	0.5	0.7	0.6	0.5	0.5	0.6	1.1	1.1	0.5	0.6
9.3 Carcinoma of gastrointestinal tract	21.9	19.7	17.1	15.3	15.6	16.8	22.6	22.4	16.3	16.3
9.3.1 Carcinoma of esophagus	—[b]	—[b]	0.2	0.3	0.2	0.4	1.2	1.2	0.2	0.3
9.3.2 Carcinoma of stomach	0.7	1.1	1.8	1.7	2.0	2.0	1.9	1.9	1.6	1.9
9.3.3 Carcinoma of small intestine	—[b]	0.4	0.4	0.5	0.5	0.6	0.6	0.6	0.4	0.5
9.3.4 Carcinoma of colon	12.3	11.5	8.1	6.7	6.3	6.3	8.3	8.3	7.6	6.6
9.3.4.1 Appendix	11.2	8.6	4.1	2.1	1.2	0.8	0.2	0.3	3.4	1.5
9.3.4.2 Colon excluding appendix	1.1	3.0	4.0	4.6	5.1	5.5	8.1	8.0	4.2	5.1
9.3.5 Carcinoma of rectum	—[b]	1.7	3.4	3.5	3.8	4.2	3.5	3.5	3.3	3.9
9.3.6 Carcinoma of anus	—[b]	—[b]	0.2	0.3	0.4	0.6	0.5	0.5	0.2	0.4
9.3.7 Carcinoma of liver and intrahepatic bile ducts	6.1	2.8	1.6	1.1	0.9	1.0	2.4	2.3	1.4	1.1
9.3.8 Carcinoma of gallbladder and other extrahepatic biliary	—[b]	0.2	0.2	0.3	0.3	0.4	0.8	0.8	0.2	0.3
9.3.9 Carcinoma of pancreas	2.1	1.7	1.1	0.9	1.1	1.3	3.2	3.1	1.1	1.2
9.3.10 Other carcinoma of gastrointestinal tract	—[b]	—[b]	0.1	0.1	0.1	0.1	0.2	0.2	0.1	0.1
9.4 Carcinoma of lung, bronchus, and trachea	3.4	2.3	2.6	2.3	2.3	3.0	15.6	15.1	2.4	2.7
9.5 Carcinoma of skin (if collected)	1.0	0.6	0.4	0.3	0.2	0.2	0.4	0.4	0.4	0.2
9.6 Carcinoma of breast	0.5	1.3	6.8	17.6	26.9	34.3	18.9	19.2	12.8	27.3
9.7 Carcinoma of genital sites excluding ovary and testis	0.5	1.6	6.9	13.5	15.1	13.4	23.2	22.9	10.4	13.2
9.7.1 Carcinoma of uterine cervix	0.4	0.9	4.9	9.8	10.0	7.6	0.8	1.1	7.5	8.2
9.7.2 Corpus uteri	—[b]	0.5	1.7	3.2	4.3	4.5	3.7	3.7	2.5	4.0
9.7.3 Carcinoma of vulva and vagina	—[b]	—[b]	0.2	0.3	0.5	0.6	0.5	0.5	0.3	0.5
9.7.4 Carcinoma of penis	—[b]	—[b]	0.1	0.1	0.1	0.1	0.1	0.1	0.1	0.1
9.7.5 Carcinoma of prostate	—[b]	—[b]	—[b]	0.0	0.1	0.5	18.0	17.4	0.0	0.3
9.7.6 Other genital	—[b]	—[b]	0.1	0.1	0.1	0.1	0.2	0.2	0.1	0.1
9.8 Carcinoma of urinary tract	6.2	5.2	5.5	6.0	6.6	7.2	10.3	10.2	5.8	6.7
9.8.1 Carcinoma of kidney	5.1	3.4	3.7	4.4	5.1	5.5	4.1	4.1	4.1	5.1
9.8.2 Carcinoma of bladder[c]	1.1	1.8	1.8	1.5	1.4	1.5	5.7	5.6	1.6	1.5
9.8.3 Other urinary	—[b]	—[b]	—[b]	0.1	0.1	0.1	0.5	0.5	0.0	0.1
9.9 Other invasive carcinomas	5.3	1.9	1.9	1.5	1.3	1.3	2.0	2.0	1.6	1.4
9.9.1 Adrenocortical carcinoma	3.4	0.5	0.3	0.2	0.1	0.1	0.0	0.0	0.2	0.1
9.9.2 Unknown primary	0.8	0.5	0.9	0.7	0.7	0.7	1.6	1.6	0.7	0.7
9.9.3 Thymic carcinoma	—[b]	0.5	0.4	0.3	0.2	0.2	0.1	0.1	0.4	0.3
9.9.4 Carcinoma of other and ill-defined sites	1.0	0.3	0.3	0.3	0.2	0.2	0.2	0.2	0.3	0.2

[a]This category excludes in situ carcinomas.
[b]Statistics are not displayed for this group because there were less than 6 cases.
[c]For the bladder, the classification separates in situ and invasive carcinomas, but the Surveillance, Epidemiology, and End Results data combine them as invasive.

TABLE 14. Carcinomas of Thyroid and Head and Neck: Distribution of Histologies Within Selected Primary Sites Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017[a]

	Age at Diagnosis, y									
Category 9: Carcinomas of Thyroid and Head and Neck	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9.1 Thyroid carcinoma, N	764	2220	4922	8340	11845	14647	118,006	160,744	15,482	41,974
Percentage of category 9.1										
9.1.1 Medullary	9.6	1.8	0.9	1.1	1.1	1.1	2.0	1.8	1.1	1.1
9.1.2 Hurthle cell carcinoma	0.8	1.0	0.9	1.1	1.1	1.2	2.6	2.2	1.0	1.1
9.1.3 Papillary	57.1	68.8	70.1	69.7	67.5	67.1	59.8	61.9	69.7	68.2
9.1.4 Follicular	8.6	6.5	5.9	5.5	5.0	4.6	5.2	5.2	5.8	5.2
9.1.5 Papillary with follicular variant	19.9	20.3	20.9	21.7	24.6	25.2	27.4	26.4	21.3	23.6
9.1.6 Other	4.1	1.6	1.3	0.9	0.8	0.8	3.0	2.5	1.1	0.9
9.2.1 Nasopharyngeal carcinoma, N	75	101	119	140	210	412	6644	7701	360	982
Percentage of category 9.2.1										
9.2.1.1 Nasopharyngeal carcinoma, squamous	48.0	39.6	61.3	54.3	46.7	56.1	66.6	64.7	52.5	52.7
9.2.1.2 Nasopharyngeal carcinoma, other	52.0	60.4	38.7	45.7	53.3	43.9	33.4	35.3	47.5	47.3
9.2.2 Oral cavity, lip, and pharynx, N	45	89	192	404	778	1523	114,880	117,911	685	2986
Percentage of category 9.2.2										
9.2.2.1 Oral cavity, lip, and pharynx, squamous	37.8	36.0	64.6	71.3	77.5	84.8	95.1	94.6	64.8	78.3
9.2.2.2 Oral cavity, lip, and pharynx, mucoepidermoid	51.1	51.7	24.5	20.0	12.2	8.6	1.1	1.4	25.4	13.4
9.2.2.3 Oral cavity, lip, and pharynx, other	—[b]	12.4	10.9	8.7	10.3	6.6	3.8	3.9	9.8	8.3
9.2.3 Salivary gland, N	133	165	252	295	408	473	12,941	14,667	712	1593
Percentage of category 9.2.3										
9.2.3.1 Salivary gland, acinar	29.3	35.8	33.3	24.1	20.8	26.0	9.5	11.6	30.1	26.5
9.2.3.2 Salivary gland, other malignant	70.7	64.2	66.7	75.9	79.2	74.0	90.5	88.4	69.9	73.5

[a]This category excludes in situ carcinomas.
[b]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 15. Carcinomas of Digestive System: Distribution of Histologies Within Selected Primary Sites Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017[a]

	Age at Diagnosis, y									
Category 9: Carcinomas of Digestive System	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9.3.2 Carcinoma of stomach, N	11	43	171	352	832	1458	81,116	83,983	566	2856
Percentage of category 9.3.2										
9.3.2.1 Stomach, neuroendocrine	—[b]	14.0	8.8	6.8	11.4	13.2	7.1	7.3	8.0	11.6
9.3.2.1.1 Neuroendocrine tumor (NET)	—[b]	—[b]	6.4	5.4	9.7	10.7	5.4	5.5	6.2	9.5
9.3.2.1.2 Neuroendocrine carcinoma (NEC)	—[b]	—[b]	—[b]	—[b]	1.7	2.5	1.7	1.8	1.8	2.1
9.3.2.2 Stomach, signet ring	—[b]	41.9	39.8	38.6	41.1	34.2	17.2	17.9	39.2	37.2
9.3.2.3 Stomach, other adenocarcinoma	—[b]	39.5	47.4	52.6	44.8	49.5	71.4	70.6	50.0	48.2
9.3.2.4 Stomach, other invasive	—[b]	—[b]	4.1	2.0	2.6	3.2	4.3	4.2	2.8	2.9
9.3.3 Carcinoma of small intestine, N	—[b]	15	41	97	212	425	23,929	24,724	153	790
Percentage of category 9.3.3										
9.3.3.1 Small intestine, neuroendocrine	—[b]	93.3	43.9	66.0	62.7	66.4	62.5	62.6	62.7	64.7
9.3.3.1.1 NET	—[b]	66.7	39.0	51.5	54.7	53.6	49.8	49.9	49.7	53.2
9.3.3.1.2 NEC	—[b]	—[b]	—[b]	14.4	8.0	12.7	12.7	12.6	13.1	11.5
9.3.3.2 Small intestine, other	—[b]	—[b]	56.1	34.0	37.3	33.6	37.5	37.4	37.3	35.3
9.3.4.1 Appendix, N	188	330	387	445	507	552	9,971	12,380	1162	2,221
Percentage of category 9.3.4.1										
9.3.4.1.1 NET	88.3	88.5	83.7	71.0	62.3	52.9	34.7	41.7	80.2	69.3
9.3.4.1.2 NEC	11.2	9.7	10.6	13.9	8.3	6.9	3.1	4.4	11.6	9.7
9.3.4.1.3 other	—[b]	1.8	5.7	15.1	29.4	40.2	62.2	53.9	8.2	21.0
9.3.4.2 Colon excluding appendix, N	18	114	373	983	2124	4061	344,062	351,735	1470	7655
Percentage of category 9.3.4.2										
9.3.4.2.1 Colon excluding appendix, neuroendocrine	—[b]	5.3	3.8	3.1	3.3	2.0	1.4	1.4	3.4	2.6
9.3.4.2.1.1 NET	—[b]	5.3	2.9	2.0	2.2	1.5	0.8	0.9	2.5	1.9
9.3.4.2.1.2 NEC	—[b]	—[b]	—[b]	1.0	1.1	0.5	0.6	0.6	0.9	0.8
9.3.4.2.2 Colon excluding appendix, adenocarcinoma	94.4	94.7	95.4	94.7	95.8	97.0	96.6	96.6	94.9	96.3
9.3.4.2.3 Colon excluding appendix, other	—[b]	—[b]	—[b]	2.2	0.9	1.0	1.9	1.9	1.7	1.1
9.3.5 Carcinoma of rectum, N	—[b]	65	318	743	1611	3054	148,518	154,313	1126	5791
Percentage of category 9.3.5										
9.3.5.1 Rectum, neuroendocrine	—[b]	38.5	31.1	25.2	18.7	16.4	9.7	10.0	27.6	19.2
9.3.5.1.1 NET	—[b]	32.3	26.7	22.9	16.8	14.4	8.4	8.7	24.5	17.1
9.3.5.1.2 NEC	—[b]	—[b]	4.4	2.3	1.9	2.0	1.3	1.3	3.1	2.2
9.3.5.2 Rectum, adenocarcinoma	—[b]	61.5	68.2	74.2	79.6	81.8	87.0	86.7	71.8	79.3
9.3.5.3 Rectum, other	—[b]	—[b]	—[b]	—[b]	1.7	1.8	3.3	3.3	0.6	1.5
9.3.6 Carcinoma of anus, N	—[b]	—[b]	20	55	169	421	22,008	22,676	78	668
Percentage of category 9.3.6										
9.3.6.1 Anus, squamous	—[b]	—[b]	60.0	69.1	85.8	86.2	81.5	81.6	67.9	84.0
9.3.6.2 Anus, other	—[b]	—[b]	40.0	30.9	14.2	13.8	18.5	18.4	32.1	16.0
9.3.7 Carcinoma of liver and intrahepatic bile ducts (IBD), N	102	107	147	244	378	707	101,024	102,709	498	1583
Percentage of category 9.3.7										
9.3.7.1 Liver and IBD, cholangiocarcinoma	—[b]	5.6	8.2	13.1	22.0	21.4	11.9	12.0	10.0	17.9
9.3.7.2 Liver and IBD, hepatocellular carcinoma	94.1	91.6	88.4	79.5	68.8	70.4	82.0	81.9	84.7	74.5
9.3.7.3 Liver and IBD, other	—[b]	—[b]	—[b]	7.4	9.3	8.2	6.0	6.1	5.2	7.5

TABLE 15. *Continued*

Category 9: Carcinomas of Digestive System	Age at Diagnosis, y									
	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9.3.9 Carcinoma of pancreas, N	36	66	108	202	457	957	136,346	138,172	376	1790
Percentage of category 9.3.9										
9.3.9.1 Pancreas, neuroendocrine	30.6	45.5	46.3	49.5	42.5	33.4	6.7	7.1	47.9	38.8
9.3.9.1.1 NET	16.7	24.2	14.8	26.7	19.0	15.5	2.9	3.1	22.9	17.9
9.3.9.1.2 NEC	—[b]	19.7	28.7	21.8	22.8	16.5	3.7	3.9	23.4	19.6
9.3.9.1.3 Neuroendocrine, other	—[b]	—[b]	—[b]	—[b]	—[b]	1.5	0.1	0.1	1.6	1.3
9.3.9.2 Pancreas, adenocarcinoma	63.9	53.0	49.1	46.5	51.9	60.1	81.8	81.4	48.4	55.5
9.3.9.3 Pancreas, other	—[b]	—[b]	—[b]	4.0	5.7	6.5	11.5	11.4	3.7	5.7

[a]This category excludes in situ carcinomas.
[b]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 16. Carcinomas of Lung, Bronchus, and Trachea: Distribution of Histologies Within Selected Primary Sites Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017[a]

Category 9: Carcinomas of Lung, Bronchus, and Trachea	Age at Diagnosis, y									
	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9.4 Carcinoma of lung, bronchus, and trachea; N	57	88	243	486	949	2223	663,109	667,155	817	3989
Percentage of category 9.4										
9.4.1 Small cell carcinoma, neuroendocrine carcinoma (NEC)	—[b]	—[b]	2.9	2.3	5.1	7.3	13.0	12.9	2.4	5.8
9.4.2 Non-small cell carcinoma	100.0	97.7	97.1	97.7	94.9	92.7	87.0	87.1	97.6	94.2
9.4.2.1 Non-small cell, adenocarcinoma	59.6	17.0	35.0	39.3	50.2	52.5	43.4	43.4	35.6	48.5
9.4.2.2 Non-small cell, neuroendocrine	33.3	65.9	46.1	40.9	26.1	17.5	3.5	3.6	45.2	25.2
9.4.2.2.1 Non-small cell neuroendocrine tumor	29.8	54.5	37.9	35.6	21.7	13.0	1.4	1.5	38.3	20.2
9.4.2.2.2 Non-small cell NEC	—[b]	11.4	8.2	5.3	4.4	4.5	2.1	2.1	6.9	5.0
9.4.2.3 Non-small cell, other	—[b]	14.8	16.0	17.5	18.7	22.8	40.1	40.0	16.8	20.6

[a]This category excludes in situ carcinomas.
[b]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 17. Carcinomas of Breast and Female Genital Sites: Distribution of Histologies Within Selected Primary Sites Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017 [a]

Category 9: Carcinomas of Breast and Female Genital Sites	Age at Diagnosis, y									
	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9.6 Carcinoma of breast, N	9	50	636	3752	11,314	25,117	803,728	844,606	4438	40,869
Percentage of category 9.6										
9.6.1 Breast, infiltrating duct	—[b]	48.0	81.9	88.0	89.6	88.5	81.4	81.7	86.6	88.6
9.6.2 Breast, adenocarcinoma	—[b]	—[b]	4.7	2.7	2.8	3.1	4.8	4.8	3.0	3.0
9.6.3 Breast, lobular	—[b]	—[b]	—[b]	1.2	1.8	3.5	9.8	9.4	1.1	2.8
9.6.4 Breast, phyllodes	—[b]	44.0	6.1	1.8	0.6	0.4	0.2	0.2	2.9	0.7
9.6.5 Breast, medullary	—[b]	—[b]	—[b]	1.1	1.0	0.8	0.3	0.3	0.9	0.8
9.6.6 Breast, Paget	—[b]	—[b]	—[b]	0.5	0.5	0.6	0.4	0.4	0.5	0.6
9.6.7 Breast, ductal	—[b]	—[b]	—[b]	0.5	0.5	0.5	0.4	0.4	0.5	0.5
9.6.8 Breast, metaplastic	—[b]	—[b]	—[b]	0.8	0.6	0.4	0.4	0.4	0.7	0.5
9.6.9 Breast, inflammatory	—[b]	—[b]	1.1	0.7	0.8	0.6	0.4	0.4	0.7	0.7
9.6.10 Breast, other	—[b]	—[b]	3.8	2.9	1.8	1.6	1.9	1.9	3.0	1.8
9.7.1 Carcinoma of uterine cervix, N	6	34	458	2099	4196	5574	34,586	46,953	2591	12,361
Percentage of category 9.7.1	1									
9.7.1.1 Cervix, squamous	—[b]	32.4	71.2	67.4	65.4	63.9	67.3	66.7	67.6	65.2
9.7.1.2 Cervix, adenosquamous	—[b]	—[b]	3.1	2.9	4.1	4.2	3.5	3.6	2.9	3.9
9.7.1.3 Cervix, adenocarcinoma	100.0	32.4	14.8	23.3	26.5	28.4	24.3	24.9	21.9	26.4
9.7.1.4 Cervix, other	—[b]	29.4	10.9	6.4	4.1	3.5	4.9	4.8	7.5	4.5
9.7.2 Corpus uteri, N	—[b]	20	156	675	1795	3324	156,706	162,677	851	5970
Percentage of category 9.7.2										
9.7.2.1 Corpus uteri, adenocarcinoma	—[b]	70.0	85.3	93.6	95.1	94.6	95.0	95.0	91.5	94.3
9.7.2.1.1 Corpus uteri, endometrioid	—[b]	45.0	65.4	73.6	77.2	76.6	69.6	69.8	71.4	76.0
9.7.2.1.2 Corpus uteri, other adenocarcinoma	—[b]	—[b]	19.9	20.0	17.9	18.0	25.4	25.2	20.1	18.2
9.7.2.2 Corpus uteri, other	—[b]	30.0	14.7	6.4	4.9	5.4	5.0	5.0	8.5	5.7

[a]This category excludes in situ carcinomas.
[b]Statistics are not displayed for this group because there were less than 6 cases.

TABLE 18. Carcinomas of Kidney and Bladder: Distribution of Histologies Carcinomas of Kidney and Bladder: Distribution of Histologies Within Selected Primary Sites Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017[a]

	Age at Diagnosis, y								
Category 9: Carcinomas of Kidney and Bladder	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
9.8.1 Carcinoma of kidney, N	85	133	349	936	2159	4047	173,623	181,332	1418	7624
Percentage of category 9.8.1										
9.8.1.1 Kidney, adenocarcinoma	100.0	97.7	97.7	99.3	99.4	99.0	97.5	97.6	98.7	99.0
9.8.1.1.1 Kidney, renal cell	68.2	76.7	79.4	85.7	87.8	86.6	83.1	83.2	83.3	86.3
9.8.1.1.2 Kidney, other adenocarcinoma	31.8	21.1	18.3	13.6	11.6	12.4	14.4	14.4	15.4	12.7
9.8.1.2 Kidney, other	—[b]	—[b]	2.3	0.7	0.6	1.0	2.5	2.4	1.3	1.0
9.8.2 Carcinoma of bladder, N[c]	19	68	168	326	599	1134	243,277	245,591	562	2295
Percentage of category 9.8.2										
9.8.2.1 Urinary bladder, transitional cell carcinoma	84.2	92.6	91.7	93.6	92.5	92.2	94.9	94.9	92.9	92.4
9.8.2.2 Urinary bladder, other carcinoma	—[b]	—[b]	8.3	6.4	7.5	7.8	5.1	5.1	7.1	7.6

[a]This category excludes in situ carcinomas.
[b]Statistics are not displayed for this group because there were less than 6 cases.
[c]For the bladder, the classification separates in situ and invasive carcinomas, but the Surveillance, Epidemiology, and End Results data combine them as invasive.

Evolution of Histology Codes

Changes in the descriptors of morphology over time have resulted in greater specificity, but these make comparisons over time and across countries/registries difficult. For example, in the ICD-O-3.2, precursor B-cell acute lymphoblastic leukemia, which was simply defined by histology code 9836/3 in the ICD-O-3, consists of all histology codes from 9811/3 through 9819/3, but only when combined with primary site codes C42.0, C42.1 or C42.4; the same histology codes with any other primary site define precursor B-cell lymphoblastic lymphoma. Likewise, changes in behavior codes have occurred in pilocytic astrocytoma, borderline tumors of the ovary, and carcinoids, often affecting reportability. The very use of the term carcinoid may be discontinued.[20] As terminology evolves and specific histology codes are added, the *not otherwise specified* (NOS) group may decrease, eg, subgroups of specific adenocarcinomas may increase while adenocarcinoma NOS may decrease over time.

DISCUSSION

Reporting Requirements

A particularly important consideration is comparisons across populations or time periods. Many registries use the behavior code to determine reporting eligibility. The proposed classification can be used as a data-validation tool. Although unclassified cases could result from the adoption of new codes or represent the first occurrence of a hitherto unrecognized primary site, they may also highlight coding errors. Registration practice may vary even within a behavior code with respect to specific histology/site combinations, such as basal and squamous carcinomas of the skin and uterine cervix neoplasia in situ.

When comparing data from different sources using this system, it will be essential to ensure that the analyses of in situ tumors are based on primary sites common to the data sets. There is a specific consideration for the reporting of in situ tumors of the bladder, shown separately from invasive tumors in the classification; however, in the SEER registry data, these frequently are combined.

Impact of WHO Blue Books and Future Influence of the ICD-O-3.2

The new classification incorporates updated histologies from the ICD-O-3.2 based on the Blue Books fourth revision. The ICD-O-3.1 and ICD-O-3.2 have several changes to the behavior code from nonmalignant of /0

TABLE 19. Miscellaneous Specified Neoplasms, Unspecified Malignant Neoplasms Except in the CNS and in situ Tumors: Total Cases and Percentage Distribution Based on Surveillance, Epidemiology, and End Results 18 Areas, 2004-2017

	Age at Diagnosis, y									
Category	<15	15-19	20-24	25-29	30-34	35-39	≥40	All Ages	15-29	15-39
10. Miscellaneous specified neoplasms, N	6330	133	107	159	234	379	24,869	32,211	399	1012
Percentage of category 10										
10.1 Other pediatric and embryonal tumors	99.7	69.9	46.7	23.9	24.8	16.4	3.7	23.4	45.4	29.7
10.1.1 Wilms tumor	29.2	21.8	15.0	5.0	4.7	4.0	0.5	6.3	13.3	7.8
10.1.2 Olfactory and non-CNS neuroblastoma	38.8	37.6	24.3	13.2	16.7	8.7	2.2	9.9	24.3	16.7
10.1.3 Other embryonal non-CNS tumors	31.7	10.5	7.5	5.7	3.4	3.7	1.0	7.1	7.8	5.2
10.2 Other specified tumors	0.3	30.1	53.3	76.1	75.2	83.6	96.3	76.6	54.6	70.3
10.2.1 Paraganglioma, non-CNS	0.3	18.8	24.3	22.0	17.1	11.9	2.3	2.3	21.6	16.9
10.2.2 Other specified neoplasms	—[a]	11.3	29.0	54.1	58.1	71.8	94.1	74.3	33.1	53.4
11. Unspecified malignant neoplasms except CNS, N	58	46	187	278	446	748	165,937	167,700	511	1705
A. In situ, N	126	655	2437	4403	7520	13,295	494,628	523,064	7495	28,310
Percentage of category A										
A.1 Melanoma in situ (if collected)	75.4	53.4	46.0	52.2	50.4	40.3	42.0	42.2	50.3	45.6
A.1.1 Superficial spreading melanoma in situ	—[a]	5.8	6.3	6.5	5.9	4.9	2.4	2.6	6.4	5.5
A.1.2 Lentigo maligna	—[a]	—[a]	1.1	1.2	1.9	2.5	11.0	10.5	1.1	2.0
A.1.3 Other in situ melanoma	69.8	47.3	38.6	44.4	42.6	33.0	28.5	29.0	42.8	38.1
A.2 Colon including appendix, in situ	—[a]	1.1	0.5	0.6	0.9	1.2	3.3	3.2	0.6	1.0
A.3 Rectum, in situ	—[a]	—[a]	0.5	0.7	0.5	0.7	1.3	1.3	0.6	0.6
A.4 Anus, in situ	—[a]	4.1	9.5	10.4	9.0	7.2	1.8	2.1	9.6	8.3
A.5 Breast, in situ	—[a]	2.0	3.4	7.0	15.4	31.5	40.8	39.7	5.4	20.3
A.6 Cervix uteri, in situ (not collected by some registries)	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]
A.7 Ovary, in situ	—[a]	1.5	1.4	1.0	0.6	0.4	0.1	0.1	1.2	0.7
A.8 Vulva and vagina, in situ	7.1	35.0	35.2	23.5	19.5	15.4	5.1	5.9	28.3	19.9
A.9 Penis, in situ	—[a]	—[a]	1.8	2.5	1.3	0.9	0.6	0.6	2.1	1.3
A.10 Prostate in situ including PIN III[b]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	0.0	0.0	—[a]	—[a]
A.11 Urinary bladder, in situ[c]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]	—[a]
A.12 Other in situ	14.3	2.1	1.7	2.1	2.5	2.4	5.1	4.9	2.0	2.3

Abbreviations: CNS, central nervous system; PIN III, prostatic intraepithelial neoplasia grade 3.

[a]Statistics are not displayed for this group because there were less than 6 cases.

[b]PIN III is not collected by all registries.

[c]For the bladder, the classification separates in situ and invasive carcinomas, but the Surveillance, Epidemiology, and End Results data combine them as invasive.

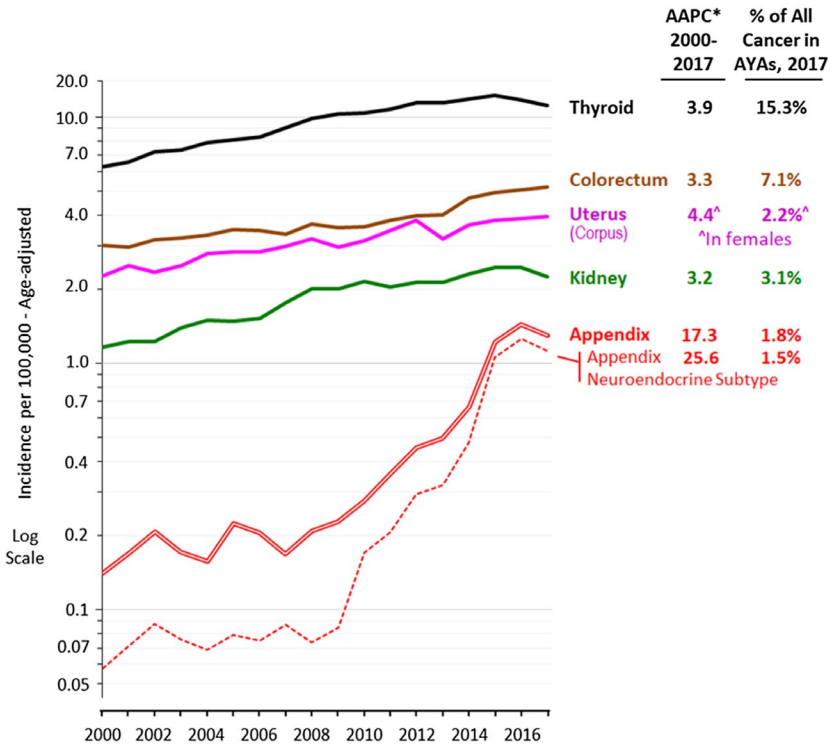

FIGURE 2. This chart illustrates the annual incidence of the 5 cancers with >1% prevalence among adolescents and young adults (AYAs) that had the greatest rates of increase during the period from 2000 to 2017 in US Surveillance, Epidemiology, and End Results (SEER) Program 18 areas. The incidence for appendix cancer includes the neuroendocrine subtype. *AAPC indicates average annual percent change (data source, SEER*Stat).

(benign) or /1 (borderline) in the ICD-O-3.0 (original) to /3; and the reverse. Most registries collect /3 (malignant) for all sites and /0 and /1 for CNS tumors. The reportability of in situ (/2) cases varies, depending on the reportability rules and dates of implementation of revisions to the ICD-O by individual registries. Cases may become nonreportable when the behavior code changes from /3 to /0 or /1 or /2.

An example in the ICD-O-3.2 is dermatofibrosarcoma protuberans (DFSP), most (but not all) variants of which have been downgraded from malignant (/3) to noninvasive (/1). Therefore, the category 4.4.3 *other fibromatous neoplasms* will lose the majority of its cases when the ICD-O-3.2 is implemented. One solution is to exclude all DFSP cases for all years. Yet this entity peaks in incidence in the AYA age group, specifically in those aged 34 to 39 years. Moreover, DFSP, although seldom metastatic, has a high rate of local recurrence after surgical resection if the tumor is not excised entirely with negative margins.[21] Chondrosarcoma grade 1 has been downgraded also in the ICD-O-3.2.

The latest Blue Book[22] on sarcomas and the ICD-O-3.2 have *gastrointestinal stromal tumor* as a /3, but it had been /1 unless it was stated as malignant in the ICD-O-3. Some registries may be hesitant to adopt changes affecting their reportability rules, especially if this means an end to reporting for a given neoplasm and changes the incidence trends over time, particularly if some of the tumors affected are fairly common. Different registries may adopt changes at different times. The challenge to cancer registries is to collect the same entities and data in the same way to allow meaningful aggregation and comparative analyses.

Another example is CNS neoplasms, which the WHO has redefined by both histologic and molecular features, including glioblastoma isocitrate dehydrogenase (IDH)-wild type and glioblastoma *IDH*-mutant; diffuse midline glioma *H3 K27M*-mutant; *RELA* fusion-positive ependymoma; and embryonal tumor with multilayered rosettes, *C19MC*-altered.[23] Some entities that are no longer diagnostically relevant, such as CNS-primitive neuroectodermal tumor, have been deleted.[23] For medulloblastoma, 4 distinct molecular subgroups—wingless (WNT), hedgehog (SHH), group 3, and group 4—have been integrated into the tumor's subclassification. The SHH subgroup is associated with *SMO* mutations. As

noted in a recent review,[24] SMO inhibitors have not yet been tested in randomized controlled trials.

Changes in the behavior code between versions of the ICD-O are inevitably difficult to deal with in analyses over time and between registries. Some entities became reportable, some became nonreportable, and some even became reportable in the ICD-O-3.1 and then not in the ICD-O-3.2, as exemplified by nondisseminated Langerhans cell histiocytosis.

Thyroid cancer is particularly problematic because its incidence has increased epidemically in the United States and other countries to become the most common cancer in AYAs—1 in 8 cases as of 2017 (Fig. 2). Part of the increase has been caused by overidentification (overdiagnosis)[25] in the detection of small tumors; the great majority of these are papillary thyroid cancer (PTC), of which classic PTC is the most common (70%-80%).[26] The follicular variant of PTC (FVPTC) is the second most common subtype. It has increased in incidence 4-fold in the last 3 decades, and now comprises approximately 20% of all thyroid carcinomas in Europe and North America.[27] A recommendation to consider the encapsulated variant of FVPTC as a nonmalignant neoplasm[25] and to rename this subclass noninvasive follicular thyroid neoplasm with papillary-like nuclear features (NIFTP; code 8349/1), was endorsed in the 2017 Blue Book.[28,29] This change intentionally removes *carcinoma* from the designation. NIFTP should be treated by lobectomy only, without completion thyroidectomy and radioiodine therapy.[30] It is estimated that 45,000 patients worldwide will be affected each year by this change in nomenclature,[31] approximately one-half of whom will be AYAs. Tumor registries are beginning to stop reporting NIFTP as invasive, which will probably lead to artefactual decreases in the incidence of FVPTC and PTC. There is a need to incorporate the new knowledge and to identify the many other forms of thyroid cancer in AYAs who are being overdiagnosed.

Nonspecific Cancers and Carcinomas of Unknown Primary Site

Cases not specified morphologically and just listed as neoplasms are in a separate category (category 11), except for those in the CNS (Table 3). Carcinomas for which the primary site is not specified (unknown primary) are in category 9.9.2. These categories may contain more cases diagnosed clinically and/or unconfirmed histologically. Analyses could be limited to histologically confirmed cases, thus reducing the frequencies of categories 11 and 9.9.2. The regular review of these cases by registries may prompt an examination of medical records or pathology reports to determine whether individual cases could be classified more appropriately.

Conclusions

The inclusion of comparative data for the group ages 15 to 29 years emphasizes the impact of extending the upper age limit for AYAs to 39 years. Germ cell tumors of the ovary are proportionately less common in individuals aged 30 to 39 years than in those aged 15 to 29 years, whereas the reverse is true for endometrioid tumors of the ovary. Although carcinomas of the breast, colorectum, and anus are proportionately more common in the group aged 30 to 39 years than in the group aged 15 to 29 years, the reverse is true for neuroendocrine tumors of the colon, rectum, pancreas, and lung. Whereas only 5% to 8% of all cancers occur in the group aged 15 to 39 years, approximately 20% of leukemias of mixed phenotype and of mast cell neoplasms occur in AYAs, as do 36% of invasive oligodendrogliomas, 61% of primary mediastinal large B-cell lymphomas, and nearly 69% of germ cell tumors of the testis.

The new classification supports the choice of the age range from 15 to 39 years, inclusive, for AYAs and straddles the International Classification of Childhood Cancer, third edition,[9] for children and classifications used for adults. Similar to the International Classification of Childhood Cancer, third edition, several diagnostic categories are emphasized for their morphology, such as leukemias, lymphomas, melanoma, and soft-tissue sarcomas. The new classification also separates neoplasms arising in blood and other vessels from sarcomas, because the former are not connective tissue tumors and are well represented in AYAs. For carcinomas, the emphasis is on primary site and, secondarily, on morphology. Benign and borderline tumors of the CNS are included because of their clinical significance, including high mortality rates, in pediatric and AYA patients, and in situ tumors are included because of their clinical significance in AYAs and older adult patients.

However, any new system is necessarily limited by the dynamic of increasing knowledge, both clinical and pathobiological, the latter including recent publications of work from the International Cancer Genome Consortium on a large number of whole cancer genomes.[32,33] Nonetheless, as with any new system, this classification of cancers in AYAs can be updated as terminology evolves. We anticipate that this proposal will serve its purpose for the near future, but modifications will continue to be necessary as important new biologic and clinical findings emerge and successive revisions to the WHO Blue Books and the ICD-O-3 are published.

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